What Doctors Must Learn
Volume 2

History is not His story, but much more

(ANALYSIS OF SYMPTOMS WITH CASE-BASED DISCUSSION)

DR. YESHWANT K. AMDEKAR
DCH, MD (PEDIATRICS),
FIAP (FELLOW OF INDIAN ACADEMY OF PEDIATRICS)

What doctors must learn – Vol 2

History is not His story, but much more

Analysis of symptoms with case-based discussion

Table of contents

jaundice
2.17 Oedema – fluid in wrong sites
2.18 Oedema – timely intervention a need!
2.19 Bulging abdomen – a sign of ill health
2.20 Abdominal distension – don't ignore
2.21 My child suddenly became stiff!
2.22 Seizure – must act fast
2.23 Headache an invisible problem
2.24 Headache – think beyond the brain
2.25 My legs ache
2.26 Leg pain may be a sinister symptom
2.27 Breathlessness – it is scaring!
2.28 Breathlessness – need early recognition and prompt action
2.29 Generalised weakness – a vague symptom
2.30 Generalised weakness – needs a cautious approach
2.31 My child is not gaining weight
2.32 Poor weight gain – needs a proper evaluation
2.33 My child is short
2.34 Everyone wants to reach "height"
2.35 Ensure child is developing well
2.36 Early pickup of developmental aberration for timely action

Section 3 Analysis of other symptoms seen at times in office practice

3.1 Skin rash a challenge
3.2 Fainting episode – a diagnostic challenge
3.3 Squint – a straight view

3.4 Mouth ulcers
3.5 Epistaxis
3.6 Sleep problems
3.7 Flatulence
3.8 Dental health – mostly neglected
3.9 Urine output – a marker of renal health
3.10 Voiding dysfunction
3.11 Abnormal movements – movement disorder
3.12 Excessive weight gain – obesity

Section 4 Symptoms we often forget to remember

4.1 Concept of untold symptoms

Preface

History taking is the most important part of a diagnostic process to which it contributes to more than 80%. However, it is possible only if a physician has acquired the art of history taking. History is not merely a reproduction of the patient's narrative but it is much more. This is where the skill lies to gather information in a manner that would lead to a probable diagnosis. It requires "thought in action" where the brain constantly interprets every bit of information as it is gathered, leading to the next relevant question. Thus, patient's reply to a question should lead to the next appropriate question. This is how entire history would be able to unfold the probable diagnosis.
Even a classic symptom pertaining to a particular system may also arise from other systems. For example, cough is a symptom of a primary respiratory system but it also may be secondary to cardiac, upper GI, neurological system or even

may be psychogenic in origin. Therefore, physicians should be conversant with basic concepts related to each symptom. Collected information if analysed properly can anticipate likely abnormal physical findings. When physical signs on clinical examination tally with what is expected on analysis of history, one is almost sure of the diagnosis. It can then be confirmed by relevant laboratory tests if necessary.

This book emphasises the need for every physician to develop adequate skills in history taking. It is important to follow simple rules and standard ways of history taking so as not to miss out any vital information. However, the relevant information is possible to gather only if a physician has clear concepts of the genesis of each symptom and understands what such a symptom conveys, then only, subsequent questions can be framed to obtain the best possible information. Correlation with other accompanying symptoms helps to define the anatomy and pathology of the disease, while etiology is guesswork based on rational thinking.
This book emphasises the importance of history taking, helps to understand the genesis of individual symptoms and demonstrates how such knowledge can be implemented to arrive at a bedside diagnosis in routine cases seen in office practice. Each chapter ends with MCQ for a short revision

Section 1 General view

1.1 Art of history taking

The diagnostic process in medicine is complex but should follow the standard sequence. It should be a patient-centric activity of gathering facts through patient hearing,

integration of collected information and interpretation (analysis of detailed history) that helps to guide afocused physical examination. It is the way to arrive at a bedside provisional diagnosis. This is a process of diagnostic refinement followed by diagnostic verification by appropriate tests, if necessary. It brings in rationality, confidence, and consistency, enables planning further management, eliminates mistakes, improves outcome, avoids misuse of laboratory tests and drugs, saves time as well as cost, offers satisfaction to patients and fulfillment to doctors. As detailed history analysis contributes to more than 80% of provisional diagnosis, enough time should be spent on gathering the right information**. History**is not **his story;**the patient focuses on what bothers him the most and does not necessarily report what the physician would want to know. History should follow the principle of “thought in action”. It means each question should be deliberate with a specific purpose, the answer to which should lead to the next relevant question. History should reveal not only anatomical diagnosis (site of the disease) but even microanatomical site (it is not enough to know the affection of the respiratory system but history must also suggest which part of the respiratory system may be involved, is it airways, lung parenchyma, interstitium or pleura). Once the history suggests the microanatomical site of the disease, further questions should try to unfold probable pathology (inflammation, infarction, infiltration or tumor, degeneration). Etiology is guesswork based on using adjectives (acute, subacute or chronic, static or progressive, continuous or recurrent, worsening or

improving). Thus, “thought in action” can arrive at a probable diagnosis.Every mother, however uneducated or illiterate, is observant about her child’s symptoms and physicians must develop skills to ask relevant questions to get the right information. Patient listening is important and time spent on history-taking is always worth it.

1.2 Simple rules of history taking

In the case of a single symptom, one may have to ask for other relevant symptoms if present. Such other symptoms may not be reported upfront as the patient may feel they are not important and highlight only a single symptom that bothers him most. For example, a patient suffering from fever may not mention mild running nose or cough that may denote an oncoming respiratory infection. Similarly, a patient complaining of general weakness has no other symptoms but on direct questioning, one may come to know other hidden symptoms such as polyuria, polydipsia and polyphagia that may suggest diabetes mellitus.In case of multiple symptoms, ask for a **chief complaint**. It is not rare for a patient to describe many symptoms such as fever, cold, cough, occasional vomit and one loose stool, headache and body ache, loss of appetite and disturbed sleep etc.Butone should enquire about a chief complaint (it is the most prominent complaint). If it is fever, one can understand all other symptoms are related to fever and so fevershould be a leading point to ask relevant history. Similarly, when the patient complains of abdominal pain, vomiting and fever, if the chief complaint is abdominal pain, it is likely to be acute

appendicitis, in which vomiting and fever are secondary symptoms., if the chief complaint is vomiting, viral A hepatitis is a probability. Thus, chief complaint offers a clue to a probable diagnosis and guide to a focused physical examination and relevant tests. But at times, the patient may consider all symptoms to be equally severe – means no single chief complaint. In such a case, enquire about the **sequence of symptoms** appearance and disappearance if any. For example. If a patient presents with fever followed the next day with a cold and then cough, it is likely to be a viral infection but if a patient presents with cold and cough first and two days later presents with mild fever, it is not an infection but allergy. Similarly,the sequence of the disappearance of symptoms is also important to note. For example, in a patient suffering from typhoid fever, fever is the last symptom to disappear but much before fever subsides, the patient shows improvement in appetite, general well-being and abdominal fullness. However, if the patient becomes afebrile before other symptoms disappear, one considers it to be a complication, such as septic shock. Thus, the sequence of appearance and disappearance of symptoms is also an important part of the inquiry.

1.3 Standard method of history-taking and its relevance

General information

Patient should be addressed by the first name which sets in a good rapport. Age is important to know as some diseases are more prevalent in particular age groups. The exact age in weeks or months is relevant to monitor growth in

infants.History should start with the first question
Was the patient completely well prior to the onset of the present illness? Often, the patient refers to the onset of the major symptom as the starting point of illness. However, when probed further, milder symptoms could be existing prior to the onset of the present illness. “Apparently” normalpatient may not really be normal prior to reported illness.This is important as milder symptoms are not voluntarily reported as the patient feels they are not important. For example, low-grade fever for a few days prior to the onset of high fever may not be mentioned. Missing such minor symptoms can lead the physician in the wrong direction. Similarly, acute onset of new symptoms in a previously healthy individual has a different interpretation than one presenting in a malnourished individual. A hidden disease, even without observed symptoms, does affect the general well-being of the patient and it is reflected by disturbed appetite, sleep, activity or energy level and behavior as well as change in bowel pattern or urination. Such minor changes are often not reported unless asked for. Thus, every effort must be made to make sure that the onset of the problem as reported is correct.

Chief complaint
It refers to the main complaint or reason for which a patient presents to a doctor. It may not always be the first symptom noticed by the patient. For example, the patient starts with a running nose followed the next day with a high fever. In such a situation, fever is the chief complaint, though not the first symptom perceived by the patient but the first symptom that

offers a clue to the respiratory system as the anatomical localization.

Onset, duration and progress

Hyperacute (in seconds) onset suggests mechanical cause such as trauma, inhaled foreign body, pulmonary embolism or pneumothorax, onset within minutes or few hours is often due to allergy or a vascular etiology such as thrombosis and onset over 2-3 days would suggest a probable infection. Thus, onset helps to consider probable etiology.

It is important to note the duration of symptoms. The short duration of fever may represent acute viral infection while fever presenting for two weeks or longer may be due to tuberculosis or non-infective inflammatory disease. Abdominal painlasting for months without any disturbance in health may be due to a functional disorder. It is important to note that the duration of a symptommay not coincide with the duration of the disease. Mild symptoms may persist well after the disease is cured but on the other hand, symptoms may disappear completely but the disease may still remain active as happens in tuberculosis or malignancy.Progress as noted in history helps to decide the type of disease. Symptoms may start improving as in common viral infection (recovering disease) or remain to the same extent for few days as in typhoid or tuberculosis (static disease, neither improving or deteriorating) or increase in severity or new symptoms may come up as in case of empyema following pneumonia or capillary leak syndrome following dengue fever (worsening disease). Viral infection may present with biphasic fever – recurrence of fever after the afebrile period

in which the second phase of fever is mild and short, it settles down by itself. However, if in an infection, the second phase of fever is severe and prolonged, it suggests immune complications as happens in leptospirosis. However, if the interval between two phases of fever is longer, say a few days, it is likely to be the same infection with a variable pattern of fever as happens in tuberculosis (fever off and on) and in such a situation, the patient remains sick during the afebrile period. On the other hand, the patient remains normal in case of recurrent fever due to recurrent infection.

Past history

Cough is a common recurring symptom in routine practice and the presence of past history of similar cough indicates a probable hyper-reactive airway disease as in the case of asthma. Thus, past history of similar disease (not merely similar symptoms) is important. For example, a child with nocturnal episodic cough in the past, presents this time, with progressively worsening cough with a whoop, obviously,the disease is different (pertussis) but has a similar symptom of cough. Similarly, recurrent episodes of fever may be due to different causes each time. Thus, past history of similar symptoms is different from that of a similar disease. It is also important to ask for past history of major illnesses. For example, if a patient presents with acute bacterial pneumonia but has a past history of meningitis and osteomyelitis at different times within the last one year, it suggests underlying immune deficiency disorder. However, if a patient presents with recurrent pneumonia in the same

lobe, it is likely to be due to congenital malformation in that lobe that predisposes to recurrent infection at the same site.

Family history

Few diseases may be genetically determined and hence recur in subsequent generations. Depending on the type of genetic inheritance (autosomal or sex-linked, dominant or recessive), one may get some clues to a diagnosis. For example. Hemophilia due to factor 8 deficiency is a sex-linked recessive disorder that means it occurs only in males but is transmitted by females. However, many disorders are autosomal recessive and hence occur in both males and females but both the parents are heterozygous – which means they harbor abnormal gene but are asymptomatic or minimally symptomatic which is often not noticed. Highly infectious diseases such as whooping cough often spread easily to other members in contact and hence, one may note the positive family history of a similar disease. Tuberculosis is another classic example. Thus, family history may also refer to a history of contact in the family.

Personal history

It refers to factors of general well-being as noted by the level of activity or energy, appetite, sleep, behavior, bowel movements, urination and weight record if available. Subtle or hidden illness may be present prior to the onset of the main complaint and it may be reflected in items of abnormal personal history as mentioned above. For example, polyuria may not be mentioned unless asked for and so one may miss diabetes mellitus or renal tubular disorder. As noted at the

beginning of this chapter, personal history indicates how well the patient was before reporting the present illness.

Drug history

Patient may be taking some drugs that may alter the presentation of his present disease as happens often in a febrile patient already on antibiotic therapy. Patients on any chronic drug therapy may present with symptoms of either excess of drug or due to missing a dose of a drug as may happen in case of diabetes. History of a drug allergy may be important to guide the further selection of drugs.

Diet history

Diet recall over a few days may bring about information of adequacy or inadequacy of nutritional intake. It would help in the management but also may offer a clue to the diagnosis. For example, a patient complaining of flatulence and gaseous abdominal distension may be consuming too much of sugar-containing foods including milk or change in bowel pattern may be related to consumption of wheat as in celiac disease.

Socio-economic history

Knowing the socio-economic status of the patient defines the risk of some of the diseases as well as affordability of therapy, based on which a physician must modify the diagnostic and therapeutic approaches to suit the needs of the patient. Of course, it has to be done without sacrificing the benefits of rational diagnostic and therapeutic measures. It is important to realise that the majority of problems in routine practice can be taken care of by clinical bedside medicine (analysis of detailed history and focused physical

examination) using minimum laboratory tests and/or drugs. (Essential drug list as suggested by the Government is sufficient for the treatment of 90% of patients in routine practice). There are scales available to assess the socio-economic status of patients. (Kuppuswami scale is commonly used)

Growth and development historyThis part of history is important in children during growing and developing years. Primary malnutrition due to poor intake of food or secondary malnutrition due to severe or chronic disease affects growth (weight is affected first and if the problem continues then the height is also affected and hence growth is measured by both weight and height). Even In absence of weight and height measurements, parents are able to inform growth patterns only if asked for. Many times, parents complain that younger sibling looks bigger than the elder or clothes don't require to change in size indicating poor growth. Similarly, inquiry about milestones offers clues to neurological or behavioral problems such as autism spectrum disorder or learning disorder. It should always be a practice to call an infant of about 10 months of age and see whether he responds to your calling his name. If he does not, one may suspect either hearing impairment, intellectual defect or autism spectrum disorder. It is thus possible to pick up such defects early enough for a better outcome.

Section 2

Analysis of common symptoms with case-based discussion

Analysis of individual symptoms

It is necessary to understand the basic concepts of the genesis of individual symptoms, nature's purpose behind such a response and how we should interpret the same and manage. A symptom may be common to any of the multiple systems in the body. Even an apparently specific symptom such as cough may primarily arise from the cardiac, neurological or upper GI system, though finally mediated through the respiratory system. Similarly, vomiting may be due to stimulation of chemoreceptor trigger zone in the brain as a result ofa variety of causes or even induced by severe cough and not necessarily only due to affection of GI system. A symptom may be so non-specific such as fever or generalized weakness that one needs to ask several leading questions for the right interpretation. Thus, knowledge of basic concepts should guide toasking relevant questions to seek desired information. It is equally important to realise that the symptom of the disease not only offers a clue to anatomical diagnosis (site of the disease) but also it is nature's intended way to protect the body. Nature is kind not to hurt but when faced with a disease, finds it necessary to present a symptom that not only protects but also alerts the patient and his doctor. It is well known that "fever is a friend and not a foe" and also pain is meant to protect the injured part from further damage.

In subsequent chapters of this section, you will find adetailed discussion of basic concepts related to individual symptoms, followed by clinical applicationbased on live case scenarios experienced in office practice and also a few MCQs for revision at the end of a chapter.

2.1. Fever a friend or a foe?

Back to basics – understanding fever

Why does body temperature rise?Fever is the body's immune response to the entry of organisms or tissue damage that may result from either an infection or non-infective inflammation due to autoimmune disorders, malignancy, allergy or trauma. Rarely, fever may result from failure of temperature regulating system as in case of hypothalamic disorder, autonomic disturbances or heat fever and also due to increased metabolism as in case of hyperthyroidism. At times, a drug may cause fever, though the diagnosis of drug fever depends on the exclusion of other possibilities and by the withdrawal of the offending drug.Rarely, fever may be a fictitious symptom. It is clear that fever does not equate to infection but there are many other causes of fever. However, infection is the most common cause of fever but not necessarily a bacterial infection. Viral infections are more common, especially in healthy young children. Thus,antibiotics should not be used in the treatment of every fever.

How does body temperature rise? Body's immune response is mediated through cytokines – chemicals produced by immune cells of the body such as monocytes/macrophages and neutrophils. These cytokines send signals to the hypothalamus that functions as a thermostat to raise body temperature to the desired set point. Hypothalamus acts through the autonomic nervous system and regulates body temperature. If the body needs to increase the temperature

to a moderate degree, it is achieved by preventing heat loss by peripheral vasoconstriction that manifests as chill in the patient and cold periphery as compared to the warm central part of the body. However, when the body needs to increase the temperature to a much higher degree, it does so by increasing heat production by excessive muscular contractions that manifest as rigors. Once the desired temperature is achieved, the thermostat shuts down and fever starts coming down either naturally or induced by the antipyretic drug. Cooling is facilitated by sweating, especially in case of high fever. One can understand that chills or rigors are not specific to any disease but simply correlate with the degree and speed of rising temperature. So, malaria may not present with rigors if the parasitic load is low but typhoid fever may present with rigors if the infection is more virulent.The degree of fever depends on the virulence of the organism and the immune status of the host. Neonate or severely malnourished child may not present with fever in spite of a serious infection. Similarly, the child may not present with a high fever to infection against which he is partially immunised. A child staying in a highly endemic area for malaria maynot present with high fever due to partial immunity derived from repeated exposure to malarial parasites. It is also true that the absence of fever may not rule out infection.

Is fever beneficial for the body? Fever is a friend and not a foe as it tries to prevent the multiplication of organisms. Raised body temperature leads to the increased blood supply to the damaged site that in turn brings in immune cells and

antibodies at the site of damage. It helps to contain the damage and promotes the healing process. Hence, fever is always helpful though when such a response crosses the intended limit, it could also harm the body as happens in hyperthermia, fortunately, it is very rare. Fever being an immune response is an indirect marker of good immunity or fighting power denoting nature's attempt at healing and hence fever is a desirable response, provided it is appropriate to the given situation. Much before science developed, it was known that serious patients with high fever often got better than those with low fever, who often succumbed. Hence in the olden days, doctors tried to induce fever hoping for better outcome. Of course, it is no longer tenable.

<u>If fever is helpful to the body, should it be controlled?</u>

Fever should be controlled only if it causes significant discomfort to the child in terms of extreme irritability or lethargy. Generally, discomfort caused by fever correlates with the peak of fever and it is the right time to use paracetamol. So, if a child's behavior is nearly normal in spite of fever, there is no need to bring down fever. Thus, we need to treat discomfort caused by fever and not fever itself.It is a myth and unfortunately a general belief, though wrong, that fever is harmful and so must be suppressed at any cost. It is important to dispel this myth with proper communication and counselling. Mild to moderate fever rarely produce significant discomfort and hence it need not be suppressed. Many children continue to be reasonably playful in spite of moderate fever. It is important to note that discomfort may be caused by factors other than fever that also need to be

addressed. Fever in a seriously ill patient in ICU may increase the energy demands of the heart and lungs in particular and so should be suppressed, even if fever is moderate.

What about risk of febrile convulsion?

Simple febrile convulsion occurs in 5% of normal children and it cannot be predicted. There is often a positive family history. Convulsion is short-lasting for less than a minute and self-limiting without risk to life or brain damage. It typically occurs within the first 24 hours of the onset of fever and rarely recurs in the same episode of fever. The first episode generally occurs in infancy or before the age of two years. However, recurrence in subsequent episodes of fever is likely up to the age of six years and can be prevented by a prophylactic drug – Clobazam that should be reserved for frequent recurrence. It is worth a note that control of fever does not necessarily prevent simple febrile convulsion though it is rational to administer paracetamol in case of high fever. In fact, many a time, parents notice fever only after the child develops a convulsion. Hence there is no need to worry about febrile convulsion though recurrence can be prevented. Convulsion in a brain-damaged child should not be considered as simple febrile convulsion and so also in a child with late-onset of the first episode and one occurring beyond the age of six years. Simple febrile convulsion does not increase the risk ofsubsequent epilepsy.

Which is an ideal antipyretic? Paracetamol is the ideal drug that controls discomfort withoutundue reduction of fever. It helps in diagnosis as fever pattern due to the natural

progression of the disease is not altered. Earlier generations of doctors categorised fever patterns as persistent, intermittent or remittent which was useful in diagnosing the cause of fever. Attempt to reduce fever by “strong” drugs, fever pattern is lost and is no longer useful for clinical diagnosis.If fever is suppressed unduly with a higher dose or more powerful antipyretic, it may lead to a false sense of disease controland one may miss worsening condition. Besides, it may also cause side effects. Paracetamol has an advantage over other antipyretic drugs in that it has a wide margin of safety and is devoid of gastric and renal side effects.Paracetamol is an ideal drug that relieves discomfort caused by high fever without disturbing the body’s helpful immune response. Thereby it helps to diagnose the cause of fever. Besides, it is the safest antipyretic**.**

What if paracetamol fails to control fever? If paracetamol is administered much before fever attains the desired peak, the fever will rise in spite of the drug. This is true for every other antipyretic because once the thermostat is set at a particular point, fever has to rise till that point is reached. So antipyretic is effective only when administered at the peak of fever which often coincides with significant discomfort.This fact is often ignored resulting in irrational therapy. Children are innocent and so they are often comfortable even at a moderate degree of fever. Discomfort is a subjective feeling and should be judged on an individual basis.However, if paracetamol fails to achieve desired comfort in spite of mild reduction of fever or otherwise, it in fact suggests probable serious infection that calls for judicious action. Thus, it helps

to diagnose serious disease in initial stages. Another condition in which every antipyretic would fail is “central” fever – fever caused by hypothalamic dysfunction. It is a rare condition but can be suspected with total failure of response to any antipyretic. Paracetamol rarely fails as ideal response is to relieve discomfort and not to control fever and it does achieve desired action. No antipyretic helps if administered too early in the course of rising fever. However, in case of genuine failure, one should rule out serious infection and consider Ibuprofenas the next alternative. Tepid water sponging is another temporary measure that is effective and safe. Cold compress over forehead is useless.

How does tepid water sponge reduce body temperature?

Temperature of tepid water should be around 25 degrees centigrade while body temperature in case of high fever may be around 40 degrees centigrade. When towel dipped in tepid water and squeezed is applied to skin surface and rubbed gently, heat from the skin surface is transferred smoothly to the towel due to physics principle of convection. It reduces temperature of that skin surface to which towel is applied. This process is repeated one by one on all four limbs, chest, abdomen, back and head. Temperature comes down by 2 degrees centigrade with two rounds of sponging the entire skin surface of the body. This method is useful when paracetamol fails to provide comfort or fever returns after paracetamol within a period of four hours. It is important to note that one should not use cold water. Application of cold water leads to vasoconstriction and in fact heat from the body is not allowed to dissipate. Further, child with high

fever cannot tolerate cold water application and becomes more uncomfortable. Tepid water sponge is effective in reducing body temperature only when it is done in an ideal way. Though normally it should be reserved when paracetamol does not produce desired action. While sponging, child should not be exposed to wind and skin should be dried well at the end of sponging.

When is urgent control of fever necessary?

It is only in case of hyperpyrexia – temperature > 105F that quick control of fever is necessary. Heat fever is one such condition in which entire body – central as well as peripheral parts of the body are equally hot. In every other condition of high fever, peripheral parts of the body are cold. In case of hyperpyrexia, physical methods are quicker than drugs and include ice-water enema or tepid water sponging. In addition, parenteral antipyretic may be used. Fortunately, hyperpyrexia is a rare event, endangering life and it needs diagnosis of cause of fever as well as proper treatment. Such a patient is ideally treated in intensive care facility.

Take home message

Fever is body's protective response and in fact it helps the patient to recover from illness. It is rarely harmful. Fever pattern and its progress helps physician to diagnose cause of fever, provided it is not suppressed by irrational therapy.

Next article deals with clinical application of basic facts highlighted in this article with discussion on live case scenarios, representing day-to-day problems faced by practitioners.

MCQs

1. Which of the following statement is right - Fever may be caused

A) By infection
B) Without infection
C) Due to inflammation
D) By all of the above

2) Which of the following statement is wrong? Causes of non-infective fever include

A) Malignancy
B) Collagen vascular disease
C) Toxoplasmosis
D) Drugs

3. Which of the following statements are correct – non-inflammatory causes of fever include

A) Heat fever
B) Hyperthyroidism
C) Central fever
D) All of the above

4) Which of the following statement is right? Fever does not respond to any antipyretic in

A) Acute bacterial infection
B) Malignancy
C) Central fever
D) Collagen vascular disease

5) Which of the following statement is wrong? Body temperature rises because of

A) Production of cytokines as a result of tissue damage
B) Temperature regulation system defect
C) Increased metabolism
D) None of the above

Answers to MCQs to be included in next article

Correct answers as follows
Q1 D, Q2 C, Q3 D, Q4 C, Q5 D

2.2. Can you believe? History leads to a probable cause of fever

Clinical application of basic concepts of fever

Cytokines mediate the rise in body temperature and so the degree of fever depends on the number of cytokines produced. A large number of cytokines are produced in localised severe acute bacterial infection, wide-spread acute viral infection and non-infective inflammation such as collagen vascular disorder or malignancy such as acute leukemia. Hence these diseases present with a high fever at the onset. The sudden release of malarial parasites in blood also results in high fever at the onset. Low-grade fever at onset suggests low-grade infection or chronic infection. A moderate degree of fever at the onset that increases to a higher degree over the next 3-4 days – step-ladder pattern –

suggests acute bacteremic bacterial infection in which initial bacteremia results in a moderate degree of fever that worsens when infection localises in a particular organ. Generally, acute bacterial infection responds poorly to paracetamol with persistent sick feeling even if fever is partially reduced while fever in acute viral infection responds better to paracetamol with the child feeling better even with minimal control of fever. Fever is often rhythmic in most infections except in the case of typical malaria. Fever in acute viral infections mostly settles by day 3-4 without any specific therapy, fever in untreated acute bacterial infection worsens by D3-4 while there is no change in fever pattern by D3-4 in case of malaria or non-infective inflammatory diseases. Accompanying symptoms if any help early localisation of the disease.

Following questions help in arriving at a provisional diagnosis in case of fever, even as early as by D 2-3.

1. Degree of fever at onset (within first 24 hours)
2, Response to paracetamol (in terms of discomfort)
3. Behavior during inter-febrile period (even with a small change in the reduction of fever)
4. Rhythm of fever (regular every 4-6 hours or irregular
5. Progress by Day 3-4 (without specific antibiotic therapy)
6. Any accompanying symptoms (often cold, cough or other)

Case-based study

Case 1

Two years old child presented with fever for a day. Fever was high at the onset, it responded fairly to paracetamol, child felt better during inter-febrile period, fever was rhythmic coming up every 4-6 hours. Physical examination did not reveal any abnormality.

High fever at onset – four possibilities – acute bacterial infection at the site of entry (tonsillitis, bacillary dysentery, UTI), acute viral infection, non-infective inflammatory disease (collagen vascular disease, malignancy) and malaria.Fair response to paracetamol and better during inter-febrile period rule out acute bacterial infection. Rhythmic fever is unlikely to be malaria. So, at this juncture, fever is likely to be due to either viral infection or non-infective causes. One has to wait to see further progress.

On day 2, child developed cold and cough. It suggests localisation to respiratory tract. Physical examination revealed fairly comfortable child, coryza, chest clear, no other abnormality. At this stage, you have already diagnosed it as acute viral respiratory infection.

One can anticipate quick recovery over next 2-3 days. Fever continued for another 2 days and then abated. However, cough continued for next few days.

Thus, it is **acute viral respiratory infection.** There is no need for antibiotic.It is anticipated that cough in acute viral infection may continue for few more days and it does not call for change in diagnosis or treatment.

Viral infection is most common cause of fever in healthy young children and it could also be frequent – few times a year. In spite of recurrence, as such episodes are self-limiting

and do not affect well-being and growth, parental counselling is necessary to allay their anxiety and refrain from unnecessary investigations and drugs.

Case 2

Two years old child presented with fever for a day. Fever was high at the onset, poor response to paracetamol with child remaining sick during inter-febrile period. Fever would rise every 4 hours. Physical examination revealed no abnormality.Poor response to paracetamol with continued disturbed behaviour during inter-febrile period would have warned of **oncoming acute bacterial infection** even before localisation. Commonly acute bacterial infection may localise to upper respiratory tract (tonsillitis, otitis media), lower respiratory tract (pneumonia), central nervous system (meningitis), intestinal system (dysentery) or less commonly to other sites such as cervical lymphnode or bone / joint. So, one has to observe development of any localising symptoms. On day 2, he developed abdominal pain and loose stools with blood and mucus. He was diagnosed as acute bacillary dysentery. He was treated with an antibiotic and got better. Bacillary dysentery is caused by gram negative bacteria and antibiotic of choice could be cotrimoxazole or cefixime. Amoxicillin would work but may be reserved for gram positive bacterial infection such as pneumonia.
It is possible that acute bacterial infection may be suspected even on first day of fever. However, one has to wait for localisation before starting an antibiotic.It is because infection at different sites vary in choice of antibiotic and also

its dosage.There is no single antibiotic that will treat effectively every bacterial infection. As a general rule, acute bacterial infections occurring in organs above the diaphragm are due to gram positive organisms while those below the diaphragm are caused by gram negative organisms. Deep-seated or serious infections such as meningitis, endocarditis or osteomyelitis must be treated with intravenous antibiotics and so also all bacterial infections in neonates, young infants and immunocompromised patients.

Case 3

Five years old child presented with fever for a day. Fever was low to moderate at onset, fair response to paracetamol with normal inter-febrile period and rhythmic pattern. As this child's fever was not high at onset, those four possibilities (acute bacterial infection at the site of entry, acute viral infection, non-infective inflammatory disease and malaria) are less likely though acute bacterial or viral infection of low virulence would still be possible. In such a case, we would have to wait and observe further course. Fever increased to high degree on day 4 with poor response now to paracetamol and child looking sicker during inter-febrile period. So, at this stage it looks like acute bacterial infection. Increasing trend of fever during first 3-4 days – moderate fever rising to higher degree - a step-ladder pattern - suggests bacteremic bacterial infection.This child did not develop any significant localising symptoms such as breathlessness (pneumonia) or headache, vomiting (meningitis), though did complain of vague abdominal pain. Physical examination showed mild

abdominal distension and sick child on D 4. **Typhoid fever** was suspected and blood culture sent for confirmation. In such a typical presentation, one may start antibiotic after sending out blood culture and not wait for results. This is because typhoid is a serious disease and early institution of antibiotic is ideal.Blood culture proved diagnosis of typhoid fever.Antibiotic of choice depends on local epidemiology but third generation cephalosporin is preferred and macrolide may also be used.

During bacteremic stage, child develops fever that increases once infection localises to some site. Three common localisation sites include lung (pneumonia), brain (meningitis) and intestine (typhoid). Though, lung can be infected also directly through droplet infection reaching through airways. One has to observe localising symptoms. Acute pyelonephritis is another bacterial infection without obvious localisation. However, it starts with high fever unlike a typical typhoid fever with step ladder pattern. Provisional diagnosis may not be possible in first 3-4 days in bacteremic bacterial infections. One has to wait for symptoms of localisation that may be observed on D2-3 such as vomiting in meningitis or mild tachypnoea in pneumonia. Typhoid fever may not present with any localising symptom except vague abdominal pain. One should not start an antibiotic unless there is reasonable clue to diagnosis, which is mostly observed by D3-4. If situation demands to start an antibiotic, one must order relevant tests before starting an antibiotic.

Case 4

Five years old child presented with high fever at onset with fair response to paracetamol and better during inter-febrile period. This would rule out acute bacterial infection. Fever was rhythmic and so malaria is unlikely. That leaves only two possibilities - acute viral infection or non-infective inflammatory disease. Physical examination did not reveal any abnormality. Fever continued for next 4 days without any change. Acute viral infection would have some accompanying symptoms and would have settled down by D4. Besides, fever in this child would rise every 12 hours and not every 4-6 hours as often happens in acute viral infection. Typically effect of paracetamol wanes off within 4-6 hours. At this stage, acute viral infection is also ruled out and hence it is most likely a non-infective inflammatory disease. Physical examination on D 4 did not reveal any abnormality. One may not be able to guess further course that may evolve over another few days to sometimes even weeks. But it is clear that it is not an infective disease and so investigations and therapy should not be addressed to infections**.** Here again, one must wait and observe for evolution of disease. In such a case, one must watch for joint involvement, skin rash, mouth ulcers (all suggestive of collagen vascular disease), lymphadenopathy, pallor, purpura and bony tenderness (all suggestive of probable haematological malignancy).
This child developed evanescent skin rash (rash appearing at the height of fever and disappearing when fever is controlled). It is a pointer to systemic inflammatory disease. Two weeks later, he developed joint swelling and pain suggesting the diagnosis of **systemic onset of Juvenile**

idiopathic arthritis. Non-infective inflammatory disease can be suspected as early as 4th or 5th day though diagnosis is not known till disease evolves in a recognisable pattern. However, there is no need to start empirical antibiotic therapy just because fever continues longer. Anti-inflammatory drug such as Naproxen is the drug of choice. Steroids are reserved only in selective cases, especially non-responders.Systemic onset inflammatory disorders simulate acute bacterial infection with high fever and neutrophilic leucocytosis. However, normal inter-febrile period and lack of localisation to any organ would suggest noninfective disorder. Unfortunately, diagnosis of systemic onset inflammatory disease is made only after antibiotics fail to improve the patient.

Case 5

Five years old child presented with high fever at onset (four possibilities) with erratic rhythm irrespective of paracetamol(mostly malaria) and normal during inter-febrile period. Physical examination on D 2 did not reveal any abnormality. Fever continued without any change or any other symptoms. In a typical situation, malaria may be considered at the end of first day of fever. Diagnosis of **malaria** due to plasmodium vivax infection was confirmed on peripheral blood smear. Chloroquine is the drug of choice in uncomplicated vivax malaria. Ideal dosage schedule should be adhered to. Early in the course of malaria, there are no positive findings on clinical examination as pallor and splenomegaly come up later and are more prominent in

recurrent malaria. However, history if well analysed can suspect malaria. It is equally important to keep in mind that malaria can present with atypical findings.Confirmation with peripheral blood smear is a must before starting anti-malarial therapy to avoid drug resistance. Rapid antigen tests have limitations though easy to implement and hence thin and thick peripheral blood smear continues to be the gold standard for diagnosis of malaria. It also defines parasitic index denoting severity.

Malaria being an endemic infection in India, it may present with wide variation of fever pattern. Hence, one may not expect each time fever with rigors but erratic fever pattern and normal inter-febrile period is a clue to diagnosis of malaria. Spleen may not be palpable in first attack of malaria. Epidemiology of infections must be taken into consideration for the etiological diagnosis of infections.

Case 6

Five years old child presented with low grade fever at onset. Low grade fever suggests infection with low virulence. Acute bacterial infection generally presents with high fever at onset but subacute or chronic bacterial infection may present with low grade fever as happens in tuberculosis**.** Acute viral infection may also present with low grade fever because host may be partially immune to such an infection either due to previous natural exposure to same virus or prior vaccination against same virus. Similarly, malaria may also present with low grade fever if host has had earlier exposure to malarial parasites. Systemic inflammatory diseases present with high

fever at onset. So, in this child acute bacterial infection and non-infective inflammatory diseases are ruled out. Physical examination at this stage did not reveal any abnormality. So, one must wait before considering any antibiotic or laboratory investigations. This is because one does not know which antibiotic and which test to order.Moreover, this child is not seriously ill. This child continued to run low grade fever for next one week without any other symptoms or signs. At this stage, viral infection as well as malaria are mostly ruledout. So, one starts thinking about subacute or chronic bacterial infections such as tuberculosis. Surely antibiotic is not necessary in this child as acute bacterial infection has been ruled out. Further tests may have to be addressed to rule out tuberculosis. This child showed pneumonia on chest x-ray and **tuberculosis** was confirmed by subsequent tests. Tuberculosis may also present with acute onset of high fever as seen in acute pleural effusion in a healthy person. This is because such a manifestation is immune mediated and hence acute presentation. Tuberculosis must always be treated with four drugs for first two months, followed by three drugs for next 4 months, irrespective of type and site of disease. However, therapy may have to be prolonged beyond 6 months, as in case of TB meningitis or disseminated TB. Empirical anti-TB therapy is irrational and it has resulted in drug-resistant TB.Thus,It is important to confirm diagnosis of tuberculosis by bacteriological testssuch as sputum culture (in young children, gastric aspiration can replace sputum for culture as sputum is often swallowed by children and AFB can be picked

up in gastric aspiration) or molecular tests such as GeneXpert that is available freely. It is also true that TB in a child is usually paucibacillary (a smaller number of bacteria) and so confirmation is not always possible. However, every attempt must be made to confirm the diagnosis. Contact screening of family members is an important tool in the diagnosis of TB in an infant. It is worth a note that there are enough facilities provided by the government for free diagnosis and treatment. Counselling a patient to ensure compliance of treatment is major responsibility of a doctor.
Tuberculosis presents with varied pattern of fever depending on the pathology of the disease. Acute onset pleural effusion due to Tuberculosis presents with short duration high fever, while typical primary complex presents with low grade fever without localisation to lungs even in an apparently healthy child. Typical presentation with low grade fever, loss of appetite and weight are classic but not always seen and are also not specific to tuberculosis.

Case 7

Two years old child presented with high fever at onset with poor response to paracetamol and continued to be sick during inter-febrile period. This would suggest acute bacterial infection. So, one may carefully look for localisation. (tonsillitis, otitis media, cervical lymphadenitis, UTI or bacillary dysentery). He did not develop any localisation over next 24 hours. Physical examination showed no abnormal signs. So, this rules out above-mentioned conditions except urinary tract infection that often has no specific localising

symptoms and for which one should order routine urinalysis and culture, so as not to miss UTI. Routine urinalysis showed large number of pus cells and urine culture confirmed diagnosis of **Urinary tract infection.** This child was treated with antibiotics covering gram negative infections such as cotrimoxazole or norfloxacin and thereafter subjected to further tests to rule out congenital defects in urinary system.

It is true that localisation of acute bacterial infection at the site of entry may get delayed beyond 2-3 days. However. non-localised fever in suspected acute bacterial infection in younger child demands ruling out UTI as early as possible. UTI is a serious disease in young children because it is likely to be due to congenital defects in urinary system and so, if not treated properly would damage kidneys due to recurrent episodes of infections. Chronic renal failure in children is often due to irrationally treated UTI and is a preventable disease with rational approach to fever. In older girls in particular, local unhygienic condition is the cause of recurrent UTI but it involves mainly lower urinary tract and hence less likely to damage the kidneys. It often presents without fever with local symptoms such as frequency of urination and burning or pain while passing urine.

Case 8

Five years old child presented with high fever at onset accompanied with headache. There was fair response to paracetamol and he appeared well during inter-febrile period. So, at this stage acute bacterial infection is unlikely. Does headache justify considering it as meningitis?

Meningitis is bacteremic bacterial infection as infection cannot reach meninges without going through blood stream. In such a case, localisation of infection occurs 2-3 days after the onset of fever. As headache appeared at onset, it is not likely to be meningitis. This child's headache was due to high fever itself as headache would go down as soon as fever was controlled.Child recovered within next 3 days and was diagnosed as viral infection.This child was treated without any specific therapy and was not subjected to any tests to rule out meningitis.

Headache is a non-specific symptom that accompanies any febrile disease. When such a symptom appears early in the course of disease, detailed history would reveal that headache disappears temporarily when fever is controlled by paracetamol. In meningitis, headache and vomiting continue irrespective of temporary control of fever. History of sequence of appearance and disappearance of symptoms gives a clue to such a problem and avoid undue tension.

Take home message

Analysis of detailed history of fever helps to arrive at a group diagnosis (bacterial or viral infection, malaria and non-infective inflammatory disorders (collagen vascular diseases and malignancy) within first 2-3 days of fever and at times even at the end of first day of fever. Until cause of fever is known, all that one needs to be sure is to rule out any serious problem. If child's behaviour is reasonably normal especially when fever is temporarily controlled with paracetamol, urine output is within normal limits and pulse / respiration are not

disproportionately fast, one can be confident to rule out seriousness. Once serious illness is ruled out, it is safe to wait for evolution of symptoms before an antibiotic is prescribed unless bacterial infection is suspected and preferably confirmed. Occasionally, localising symptoms in acute bacterial infection may evolve over a week (typhoid fever, leptospirosis, brucellosis and endocarditis) and also in acute viral infection (EB and CMV). Symptoms of non-infective inflammatory diseases may evolve over several weeks. In all such cases, counselling and documentation play important role in rational practice. Misuse of antibiotics is universal in the world and has posed a danger to life and increasing cost of health care due to development of antibiotic resistant organisms. This is an emergency and time is running out.

MCQs

1. Which of the following statement about acute viral infection is wrong?

A) It affects all parts of a system or multiple systems
B) It usually settles down by D 3-4
C) Child is sick during inter-febrile period
D) Often family history is positive

2. How early can one clinically suspect acute bacterial infection in most patients?

A) Day 3-4 after onset of fever
B) Only when localising symptoms appear
C) Within first two days of fever
D) When signs appear

3. Which bacterial infection may not have any localising symptoms?

A) Acute pyelonephritis
B) Typhoid fever
C) Tuberculosis
D) All of above

4. Which one of the following features strongly suggest malaria?

A) Fever with rigors
B) Erratic rhythm of fever
C) Sick during inter-febrile period
D) None of the above

5. Which are the diseases in which fever may persist for more than a week without any specific treatment?

A) Viral infection
B) Collagen vascular disease
C) Malignancy
D) All of the above

Answers to MCQs to be included in next article

Correct answers as follows

Q1 C, Q2 A, Q3 D, Q4 B, Q5 D

2.3. Cough – distress to patient, challenge to physician

Back to basics – understanding cough

“Cold and cough” – these terms are used by lay persons as per their perception and are often misinterpreted. Physician must confirm whether “cold” refers to nasal discharge or blocked nose and whether “cough” is a sound produced by an attempt at forceful expulsion during expiration. It is not surprising to find there may not be either cold or cough, even when complained. Another loose term used by lay people is congestion in chest. This term may simply refer to noisy breathing that may or may not be accompanied with cold or cough. Similarly, congestion in throat is another misrepresented term. After all, congestion is a physical sign and not a symptom and so, patient will not perceive congestion, it is just presumed. Hence, it is necessary for physician to confirm the intended meaning the patient wishes to convey. Without such clarification, physician may be misled in interpretation of such symptoms.

Do “cold and cough” go together?They are often together because most common cause of this combination symptoms is either a viral infection or allergy. Viral infection presents with fever followed by cold and cough while allergy starts with cold and cough but without significant fever (low grade fever is possible). In fact, sequence of events guides the doctor to diagnose the disease. Bacterial infection is mostly localised and does not present with cold and cough. However, allergy leading to cold and cough may be secondarily infected with bacteria and so may present as dual disease.

What is cough?Cough is a sound produced bysudden, forceful and often repetitive attempt at expulsion during expiration. It is a protective reflex that is expected to expel any irritant, secretions or foreign particles from larger airways – upper airways (pharynx, larynx and trachea) and lower airways (proximal or larger bronchi). Affection of other areas in respiratory tract such as nose, bronchioles, lung parenchyma, pleura and interstitium present with minimal or no cough. Cough arising from upper airways is dry while that from lower large airways is wet. Infants and young children may not be able to expectorate and wet cough at this age presents with noise produced by air moving in and out through increased secretions. However noisy chest may also present without cough.

How is cough generated? Cough starts with initial deep inspiration that is followed by brief powerful expiratory effort with closed glottis resulting in generation of pressure in the airways and then sudden opening of glottis with closure of nasopharynx and vigorous expiration through mouth. It produces sound and it all happens reflexly. It is clear that cough due to affection of bronchioles, lung parenchyma or interstitium cannot be effective because low luminal airflow and velocity at these sites fail to generate enough pressure and hence do not present with significant cough or often without cough. However, if child cannot initiate deep inspiration or powerful expiration as may happen in case of severely obstructed airways or respiratory muscle paralysis, he / she may not be able to cough even in presence of irritant or secretions in larger airways. Neonates and younger infants

are not able to cough effectively and it is obvious that child on mechanical ventilation would not be able to cough. Thus, absence of cough may not rule out large airway disease.

How is cough mediated?Cough is mediated exclusively via vagus nerve. It is worth noting that pharynx is not supplied by vagus and hence isolate affection of pharynx does not cause cough unless irritant or secretions trickle down to larynx. This has a clinical relevance in that isolated pharyngeal disease as in bacterial pharyngitis would not present with cough and so cough in pharyngeal disease suggests extension of disease beyond pharynx as happens in viral infection. Airways extending from larynx to larger or proximal bronchi contain rapidly adapting pulmonary stretch receptors that quickly adapt to persistent stimulus thereby interrupting bout of cough transiently, facilitating normal breathing in between bouts of cough. This is important as cough must be interrupted at least for few seconds to facilitate act of breathing. If single bout of cough continues for more than few seconds, it may lead to cessation of breathing – apnoea and may also be fatal as occasionally happens in young infant suffering from pertussis – whooping cough. Thus, pertussis is a serious disease in very young infant. However repeated bouts of cough cannot be prevented as nerve endings in mucosal epithelium sense the irritant or inflammatory secretions and produce cough. Finally vagal fibres enter brainstem from where cough reflex is generated via second order neurons. It also has clinical relevance as unconscious patient due to brainstem affection is not able to cough.

Factors in airways leading to coughThere are several factors in airways that can stimulate cough receptors to produce cough. These receptors are sensitive to mechanical factors such as inhaled foreign body or compression pressure on airways as in case of mediastinal tumour and these receptors also are sensitive to acid or isomolar solutions such as water. This explains cough due to gastro-esophageal reflux (GERD) or aspiration of water or food particles into the airways. Similarly, other events such as smooth muscle contraction as in asthma, vasodilatation and oedema as in cardiac conditions, mucus secretions as in bronchitis and reduced lung compliance as in pneumonia are responsible to produce cough. Inflammation commonly due to infection or non-infective causes are responsible for cough in routine practice. Drugs such as angiotensin converting enzyme inhibitors (ACE inhibitors) and non-steroidal anti-infammatory drugs (ibuprofen) are known to trigger cough, though exact mechanism is not known. Similarly psychogenic factors, especially in children, present with cough referred to as "habit cough".

Why drugs often fail to relieve cough?

There are several components such as neurogenic and mechanical factors that are involved in producing cough. Besides, most of the irritants or inflammatory products in airways are not easy to get rid of as consistency of mucus and efficiency of ciliary function determine ease of expulsion and so cough continues in spite of trial with different drugs. Even mere symptomatic relief is also difficult to achieve as there is no drug known to science that can control all these variable

factors. That is why most cough remedies are cocktails of cough-sedative, expectorant, mucolytics and antihistamines with the hope that one of the constituents may work. But it does not do so. Temporary relief may be possible with inhaled bronchodilator in case of bronchospasm. Transient relief may occur with hydration of airways as is done with steam inhalation or sips of warm water or chewable item in mouth secreting more saliva (dryness of airways increases cough). Reclining with head high position offers bit of relief and so also adequate ventilation and comfortable room temperature. Of course, removal of inhaled foreign body can "cure" cough.

<u>If symptomatic therapy does not work, what next?</u>

Significant cough though distressing to a patient, has a purpose of expelling the irritant. So, unless irritant is expelled, cough would never stop. Hence attempt should be made to offer comfort to a child rather than suppressing cough. During day time, most children remain reasonably comfortable in spite of cough, simply because they are too busy with play and other activities that they ignore cough. However, cough disturbs them during sleeping time and hence attempt must be made to ensure proper sleep within limits. One can use with discretion an antihistamine (first generation like chlorphenamine) containing cough suppressant in a single dose at night. Codeine is a powerful cough suppressant but is addictive besides it also causes constipation and hence best avoided. Pholcodiene may be an alternative. Mucolytics may be tried in wet cough but often are not beneficial. If cough suggests bronchospasm, inhaled

bronchodilator would offer relief better than oral medicine. There is no reason to use symptomatic drugs through the day and should only be administered when discomfort is intolerable. None of these modalities “cure” cough and this is the reason why treating cough is a big challenge to physician. Provisional diagnosis of the disease presenting with cough is vital, without which relief of cough may not be possible. Though luckily some of the causes of cough may be self-limiting but unfortunately, they are often recurrent.

Take home message

Cough is a reflex intended to expel the irritant in airways. Severe cough suggests affection of large airways (dry cough in upper and wet cough in lower airways) while mild cough arises from smaller airways and beyond (bronchioles, alveoli, interstitium and pleura). Pain is a symptom of pleural disease and may be present also in pneumonia (it is pleruo-pneumonia). Cough may also be a symptom of cardiac disease (left to right shunt or cardiac failure), upper GI system (GERD or aspiration syndromes), neurogenic (from ear drum or autonomic system) and psychogenic (habit cough). Drugs are not very useful to suppress cough except inhaled bronchodilator for spasmodic cough but cough suppressant with first generation antihistamine may be considered on SOS basis to relieve discomfort. Traditional home remedies are as good and must be tried.

Next article deals with clinical application of basic facts highlighted in this article with discussion on live case

scenarios, representing day-to-day problems faced by practitioners.

MCQs1**.** Which of the following statement is wrong? Mild cough is a feature of

A) Bronchiolitis
B) Pneumonia
C) Bronchitis
D) Pleural disease

2. Cough is generated with

A) Deep inspiration
B) Expiration with closed glottis
C) Forceful expiration with open glottis
D) All of the above

3. Which of the following statement is wrong? Cough may result from

A) Cardiac disease
B) Neurological disease
C) GI abnormality
D) None of the above

4. Which of the following statement is right? Cough may be absent if

A) Airways are affected
B) Airways are not affected
C) Heart is affected
D) All of the above

5. Which of the following statement is wrong? Cough is difficult to treat because

A) Correct diagnosis is often elusive
B) There are multiple mechanisms leading to cough
C) Irritant is not easy to expel
D) Cough receptors are not well developed

Answers to MCQs to be included in next article

Q 1 C, Q 2 D, Q 3 D, Q 4 D, Q 5 D

2.4. Cough should not be a challenge any more

Clinical application of basic concepts of cough

Cough is a localising symptom, unlike fever. It is often a primary respiratory disease though may be secondary to cardiac, GI or neurological disorder. Hence one must start with anatomical diagnosis – which system is involved and further which part of the system is affected – it is microanatomy of the disease. Significant cough localises disease to larynx, trachea or bronchi. In other areas of affection in respiratory tract, cough is mild and often accompanied with other symptoms such as chest pain in pleural disease, acute breathlessness in pneumonia or bronchiolitis. Palpitation and breathlessness suggest cardiac disease while chocking episode denotes probable gastro-esophageal reflux or neurological disorder. Of course, if one fails to localise anatomy of the disease, it may be psychogenic. Unless microanatomy is known, one can't proceed to pathology. Most common pathological processes

involved in production of cough are inflammation and allergy. Acute inflammation is characterised by fever while chronic inflammation presents with worsening systemic symptoms such as loss of weight and appetite. Allergy presents with sudden onset of cough that may also disappear suddenly but often would recur. Besides, there is often personal or family history of allergy. Respiratory allergy affects entire system so also in case of viral infection while bacterial infection is mostly localised to a part of the system and not generalised. Thus, probable etiology also can be guessed.

Following questions help in arriving at probable diagnosis in case of cough.

1. Is cough a major symptom?
2. If so, is there past history of similar illness?
3. Is cough worse at night as compared to day time?
4. Is there personal or family history of allergy?
5. Onset, duration and progress of cough
6. Are there other symptoms such as fever, cold or breathlessness?
7. Sequence of appearance of such symptoms

Case based study

Case 1

Two years old child presented with fever followed next day with cold and wet cough. Cough was severe with watery discharge from nose. There was no past history of similar disease. Physical examination revealed febrile child with

coryza but not looking very sick. Chest was clear.
Fever suggests infection and cough denotes airway disease along with involvement of nose as well. So this is generalised involvement of airways. This is typical of **viral infection** and so does not need any antibiotic or laboratory tests. Fever settled down within 3 days with paracetamol. Cold and cough also got better over next 2 days. In case of watery nasal secretions, one needs to wipe them and in case of nose block, instil normal saline drops in the nostrils. It helps to unblock the nose. Cough syrups are of no much use. Viral infection is often a generalised infection affecting entire system (cold and cough – upper and lower airways) and also at times multiple systems(cold, cough as well as diarrhoea) and also self-limiting within 3-4 days. Besides, viral infections spread fast and hence similar illness in other family members or prevailing in the community would offer a clue to probable viral infection.

Case 2

Two years old child presented with fever followed next day with cold and wet cough. Cough was severe with watery discharge from nose. There was past history of recurrent episodes of cough often without fever. However, there was no history of allergy in the family. Physical examination did not reveal any localising signs.
Onset of disease with fever suggests infection with generalised involvement of airways affecting nose down to bronchi. So, this is similar to previous case. However, in this child, fever subsided on its own within next 2 days but cough

worsened and continued for next two weeks. Physical examination during this period continued to show no other signs. As fever had disappeared, infection was definitely controlled. If so, why did cough continue for such a long time? This is referred to as hyper-reactive airway disease. It is known as **WALRI – wheeze associated lower respiratory infection.** It means this child is susceptible to recurrent episodes of cough that could be triggered by either viral infection or allergy. It is evident in history itself as this child had recurrent episodes of cough in the past. Cough did settle down by itself though it lasted too long. Here is the need for proper counselling rather than trying some medicines. Of course, there is no question of antibiotic in this child nor does this child deserve laboratory tests or chest x-ray. Inhaled bronchodilators help.

Generally, cold and cough in a respiratory viral infection settles soon after fever subsides. However, when cough in particular continues for few weeks well after fever subsides, one suspects inherent susceptibility of host to suffer from recurrent cough triggered by viral infection and also by multiple triggers other than infection.

Case 3

Six years old child presented with high fever, thick yellow nasal secretions and cough for last two days. Past history revealed repeated episodes of cold and cough since the age of 2 years, at times accompanied with high fever, treated with antibiotics. There was history of allergy in the family. Physical examination showed highly febrile child, looking sick,

adenoid facies, (open mouth suggesting chronic obstruction to breathing through nostrils due to enlarged adenoids), ears normal and no localising findings in chest. High fever in a sick child favours probable bacterial infection. However, in the background is hyper-reactive airways disease that is allergic as evident by family history of allergy in a child with repeated episodes of cold and cough. So, this child is suffering from **acute bacterial rhinosinusitis in the background of allergy.** It needs to be treated with antibiotic for the present but once infection is controlled, he should be treated for allergic rhinosinusitis. Amoxicillin with or without clavulanate is the drug of choice for respiratory infections.

Secondary bacterial infection is a result of retained secretions (as in rhinosinusitis, bronchiectasis, pyelonephritis or cholangitis) and significant immune suppression following measles or varicella infection, chemotherapy and immune deficiency disorders. While treating bacterial infection with appropriate antibiotic, primary background cause must be evaluated and managed well to prevent recurrences.

Case 4

Four years old child presented with high fever and mild cough. Fever was high at onset with mild response to paracetamol and high fever would recur every 4-5 hours as effect of paracetamol would wear off. Child always remained sick during interfebrile period. Cough was mild and not disturbing the child. Physical examination did not reveal any abnormality except sick looking child. At this juncture, one is expecting bacterial infection to localise mostly in lung

parenchyma as suggested by mild cough. Mild cough is also a feature of bronchiolitis, pleural or interstitial disease. Acute bronchiolitis presents with upper respiratory symptoms and often with mild fever followed by breathlessness while acute interstitial infection is often viral and so involves entire respiratory system with significant cough and so, both these diseases are unlikely in this child. Absence of chest pain rules out pleural disease. Though at times pleura may be involved in case of pneumonia and such a child may present with chest pain. Hence, even on day 2 of illness, **bacterial pneumonia** is most likely and one must watch carefully for further progress. A day later, he developed mild breathlessness and now diagnosis of pneumonia is confirmed by chest x-ray and CBC showing neutrophilic leucocytosis. Amoxicillin with clavulanate was started and child started responding with relief from breathlessness and fever was also in control. Though cough worsened by this time. Does it suggest complication or drug resistant infection? Neither of the two. In fact, as pneumonia starts improving, cough often worsens as inflammatory exudate liquefies and has to be expelled through airways. It results in temporary worsening of cough. It is nature's attempt to get rid of exudate to ensure complete healing. There was no need to change medicines and antibiotic was continued for 7 days with full recovery. There was no need of cough syrup as well. Recovering infection may worsen one of the original symptoms because of natural course of the disease (cough worsening in recovering pneumonia or diarrhoea continuing in recovering rota viral infection). In such a case, it is ideal to

communicate such an expected course of events ahead of its occurrence so that it allays anxiety on the part of patients

Case 5

Eight years old child presented with mild fever and cough for 2 days. Physical examination did not reveal any abnormality. At this stage, there is no clue to diagnosis, but acute infection seems unlikely due to onset with low grade fever and it is best to watch further progress without any tests or medicines. It is safe to wait as child is not sick looking. Child continued to run mild fever for next 10 days but cough was worsening. So, this is subacute infection of respiratory tract with gradual worsening. Physical examination at this stage showed localised crepitations on right side of chest and child had lost one kg of weight. Diagnosis of **tuberculosis** was possible and was proved with chest x-ray and confirmed with GeneXpert. This child was treated with four drug treatment (HRZE) for two months and HRE for next four months. Child showed improvement within 2-3 weeks, though treatment must be completed for 6 months.

Low grade fever may suggest acute viral infection of low virulence or one occurring in partially immune host. However, if low grade fever continues beyond 3-4 days, one should consider chronic low-grade infection such as Tuberculosis or fungal infection. Systemic fungal infection is seen in immune compromised host and not in normal person.

Case 6

Four years old child presented with mild fever and dry cough for two days but over next five days, cough started worsening though fever disappeared. Physical examination at this stage did not reveal any abnormality. It is clear that this is upper respiratory tract infection as evident by dry cough but further progress suggests worsening local condition in upper airways without deterioration of systemic symptoms such as fever. On direct questioning, two other family members were reported to be suffering from similar cough for last one month. So, this is highly infectious disease and diagnosis of **pertussis – whooping cough** was made. He was treated with Macrolide. It took another week before he got well. In absence of family history, pertussis can be suspected with worsening bouts of cough in severity and duration, especially in absence of past history of recurrent cough. It is treated with macrolide but antibiotic is effective only if started early in the course of the disease. Even if pertussis is diagnosed late in the course of the disease, it is rational to start an antibiotic as even it does not help the patient, it would prevent spread of infection to other members of the family. Progression of severe cough is a result of uncontrolled infection such as pertussis or gradual compression of airways as in case of mediastinal lymphoma. Both conditions may not present with any abnormal physical signs in the chest and must be suspected with consideration of other clues. Even chest X-ray is normal in pertussis but usually not in mediastinal lymphoma, though occasionally lymphnode may be hidden behind the heart and hence missed. Careful

assessment can visualise compression of large airways in such a case.

Case 7

Two years old child presented with severe cough for last 2 days. There were no other complaints. There was no past or family history of allergy. Physical examination did not reveal any obvious abnormality. Acute onset of severe cough without fever rules out infection and suggests either mechanical problem or allergy. As there is no history suggestive of allergy, it is likely to be mechanical problem. On direct questioning, parents recalled that cough started all of a sudden in a minute while he was eating. It gave away the diagnosis of **inhaled foreign body.** It was a piece of groundnut that was removed through bronchoscope. Inhaled foreign body may not be picked-up by clinical examination, especially if it is lodged in smaller bronchial segment and also missed by chest x-ray as vegetable foreign body is not visible on x-ray. However, one may pick-up localised signs of either atelectasis or emphysema due to complete or partial obstruction of a bronchial segment respectively on physical or radiological examination.

Basic rule to enquire about onset of the disease is so important in case of cough. Inhaled foreign body is missed for several weeks simply because a doctor had not asked for an onset. There is no other diagnosis in case of sudden onset of cough other than inhalation of foreign body. Inhalation of other material as in case of GERD presents with chocking episodes with recurrent cough.

Case 8

Eight years old child was seen for cough going on for last 6 months. There were no other symptoms. Physical examination did not show any abnormality. Several tests and trials with medicines including antibiotics, cough syrups and inhalers had failed to control cough. Finally, he was advised to undergo bronchoscopy that he refused. What was missing? On detailed questioning, it was clear that he was never disturbed by cough during sleep and play activities. Besides he was eating well and had gained weight during last 6 months. So, diagnosis was **habit cough**. Such a child often would oblige by controlling cough, if you insist that he should not cough while you examine him so that you would be able to diagnose his problem. It is due to emotional stress and one must go into depth to find out what bothers this child. Generally, both parents and the child refuse presence of any stress. Empathetic counselling with confidence of the diagnosis is the need and at times, one may have to take help of a psychologist.

Every chronic symptom is likely to affect well-being as evident by changes in weight, appetite, sleep, play activity or behaviour. Such symptoms appear early in the course of the disease and hence, should always be enquired. In absence of any such affection, diagnosis of functional disorder is obvious. It is only with such a rational approach to history taking that undue delay in the diagnosis and unnecessary investigation and drug therapy can be avoided.

Take home message

Treating coughshould not be a challenge as proper diagnosis is possible with analysis of detailed history and thorough physical examination. It leads to a probable anatomical and pathological diagnosis while etiological diagnosis is a rational guess. Tests are not commonly required unless specific diagnosis is being considered. CBC and eosinophilia in particular are not very useful for specific etiology. Chest X-ray is ideal for lung parenchymal disease but it is not so useful in airway or interstitial diseases. Chest USG is good for pleural effusion. CT scan should be avoided unless for specific reasons as it exposes the child to significant radiation with possible side effects in future. Symptomatic therapy has major limitations and parents must be properly counselled to accept time-bound natural relief, partially aided by drugs. It is doctor's duty to convince a patient and avoid prescribing unnecessary tests and drugs.

MCQs

1. In whichof the following condition, cough continues for about two weeks in spite of fever getting better within 3-4 days.

A) Acute viral infection
B) Hyper-reactive airways triggered by acute viral infection
C) Chronic viral respiratory infection
D) Secondary bacterial infection

2. Which of the following statements is wrong? Cough persisting for several weeks may be due to

A) Pertusis – whooping cough
B) Pneumonia
C) Tuberculosis
D) Inhaled foreign body

3. Which of the following statements is wrong? Primary complex due to tuberculosis may present with

A) Severe cough
B) Mild cough
C) No cough
D) Breathlessness

4. Which of the following statements is wrong? Recurrent respiratory infections occur in

A) Allergic rhinosinusoitis
B) Viral respiratory infections
C) Tuberculosis
D) Inhaled foreign body

5. Which of the following statements are right? Child may not look sick in spite of severe cough in

A) Asthma
B) Inhaled foreign body
C) Habit cough
D) All of the above

Answers to MCQs

Q1 B, Q2 B, Q3 D, Q4 C, Q5 D

2.5. Diarrhoea – when solid waste becomes liquid

Back to basics – understanding diarrhoea

What is diarrhoea?It is defined as change in bowel pattern that results in loose stools with increased frequency and child appears sick. Stool of normal consistency but passed more than 1-2 times a day may not justify definition of diarrhoea and so also an occasional loose stool with normal frequency does not mean diarrhoea. First part of the definition – change in bowel pattern - is also important. This is because young infant on exclusive breast feeds may pass as many as 10-15 loose, often watery stools per day and is not considered to be abnormal as there is no change in bowel pattern. Such infant has always been passing loose frequent stools and it is physiological, not pathological. It is substantiated by infant being happy in spite of many loose stools, feeds well and also gain weight well. Generally pathological stools – diarrhoea – makes a child sick and not happy, often with loss of appetite and dehydration depending on amount of water loss.

What is dysentery? Diarrhoea associated with stools containing mucus and / or blood is referred to as dysentery. It is usually a disease of large intestine – colon and is accompanied with abdominal pain. It represents severe inflammation and so child is quite sick. However small amount of mucus in stools without blood or abdominal pain may not be called as dysentery and such a patient may not be sick. Mucus in stools indicates any source of irritation and not necessarily an infection.

What is indigestion?

This term is used loosely by lay person and may represent different problems related to gastrointestinal dysfunction. It may not even relate to digestion problem and really does not connote any specific disease or defect. If digestion is affected, it should be ideally be called as maldigestion – meaning abnormal digestion. Digestion refers to breaking down of nutrients to smaller molecules that can be further absorbed. Proteins have to be hydrolysed to aminoacids, complex carbohydrates to simple sugars referred to as monosaccharide and fats to small molecules of fatty acids. Digestion starts with chewing of food and salivary amylase helping in first phase of carbohydrate digestion. Stomach helps in digestion of proteins and pancreatic enzymes and bile along with small intestinal enzymes are responsible for final process of digestion. If food is not digested, it cannot be properly absorbed and so results in diarrhoea. ,

<u>How does diarrhoea occur?</u>

Diarrhoea results when normal process of digestion or absorption is disturbed. Small intestine receives consumed food partly after digestion. If food is not digested properly, it is not absorbed and even if food is digested butIf small intestines are unable to absorb properly, diarrhoea results. Jejunum is the site of absorption of most nutrients except Vitamin B12 and bile acids that are absorbed in ileum. Besides amount of consumed food or fluid, much larger amounts of fluid is secreted by small intestine. Adult may consume about 1.5 litres of fluid in a day but small intestine of an adult secrets another 8.5 litres of fluid and hence small intestine has a load of about 10 litres of fluid to be absorbed

every day. Normally, 80-85% of fluids are absorbed by small intestine and only small amount is delivered to large intestine. If small intestine fails to absorb fluids, it results in large amount of watery diarrhoea. If small intestine secretes larger amount of fluids than normal due to any disease as happens in cholera, even normal small intestinal absorptive capacity cannot cope up with such a huge amount of secreted fluid, resulting in profuse diarrhoea with serious consequences such as hypovolemic shock. Large intestine receives only 10-15% of fluids that are further absorbed. Thus, small intestinal diarrhoea presents with large amount of watery stools while large intestinal diarrhoea presents with frequent small amount of stools often with mucus and / or blood.

How to know which nutrients are not absorbed?If weight loss is significant, it is calories that are not absorbed. Carbohydrates are main source of calories and its malabsorption results in diarrhoea with gaseous abdominal distension and anal excoriation due to acidic stools. This is due to fermentation of unabsorbed carbohydrates in colon by bacteria that produces gas and acidic stools. Protein malabsorption leads to oedema. Stools due to fat malabsorption are greyish white and foul smelling, often large in volume. Anaemia may be a feature of iron, vitamin B12 or folic acid malabsorption. Calcium deficiency causes tetany, vitamin D deficiency results in rickets and vitamin K deficiency leads to bleeding. Of course, all such deficiencies occur commonly due to deficient food intake of these substances rather than malabsorption.

Type of stools and resulting deficiency signs may suggest which nutrients are not absorbed from intestine. This helps especially when nutrient intake is normal and still deficiency signs appear suggesting abnormal intestinal function. At times, intestinal disease may result in deficiency of a specific nutrient while all other nutrients are well absorbed.

What are causes of diarrhoea?

Infective causes

Acute diarrhoea is commonly caused by enteroviral infections in healthy infants and toddlers. Such infections are rare beyond toddler age and are not preventable even with good hygienic care. They are referred to as democratic infections as they occur in all socioeconomic groups. Rota viral diarrhoea is a classic example. Bacterial infections such as E.coli, salmonella or campylobacter are seen in malnourished children as well those who are exposed to contaminated food. Toxins produced by vibrio cholera results in most severe form of diarrhoea. Diarrhoea is caused by toxins produced by bacterial infections such as shigella but also may result from infection at sites other than intestines such as urinary tract infection (UTI) and is referred to as parenteral diarrhoea – meaning other than enteral infection. It occurs typically in infants and young children. Other infections causing diarrhoea include parasites such as giardiasis and cryptosporidium and fungal infection mostly in immunocompromised patients. Tuberculosis does not present with diarrhoea as major symptom.

Non-infective inflammatory causes Autoimmune disorders

such as inflammatory bowel disease – commonly Crohn's disease in children and ulcerative colitis in adults are not uncommon in children. They mimic infections as they also present with fever, stool microscopy showing pus cells and neutrophilic leucocytosis. Kawasaki disease – generalised autoimmune multisystem disease – may also present with diarrhoea. **Non-inflammatory causes**

Acute malabsorption of carbohydrates as in case of transient lactose intolerance commonly occurs after viral infection. As lactase enzyme resides in most superficial part of mucosa, it is first to be disturbed resulting in lactose malabsorption as happens in viral infection. However, it is self-limiting and does not need lactose free diet. Congenital lactase deficiency is extremely rare. Children and adults who are not used to consume milk or milk products are likely to cause diarrhoea when exposed to excess of such products. It is due to disuse atrophy of enzyme – lactase. Drugs such as magnesium containing antacids, H2 receptor antagonist such as cimetidine and ranitidine, proton pump inhibitor such as lansoprazole and omeprazole, digoxin and methyldopa are known to cause diarrhoea. Irritable bowel syndrome refers to diarrhoea due to psychological factors such as stress, anxiety etc. Hunger diarrhoea occurs in infants who are poorly fed and greenish stools, besides intestinal hurry also produces greenish stools. Rarely hormonal disorders such as thyrotoxicosis and VIPoma also cause diarrhoea.

<u>Limitations of laboratory tests in acute diarrhoea</u>Routine stool microscopy is not helpful in diarrhoea as presence of

mucus, few pus cells or even RBCs are non-specific abnormalities that are present in infections, non-infective inflammation as well as any other cause of intestinal irritation. It may pick up parasitic infection though it is not related to acute diarrhoea. Stool should not be tested for lactose in acute diarrhoea as its presence does not equate to lactose intolerance. Stool culture has no place in routine clinical practice. Specific tests may be required for conditions such as celiac disease or inflammatory bowel disease. Stool examination may be necessary in chronic diarrhoea.

Should diarrhoea be controlled?Infections being commonest causes of diarrhoea, it is nature's attempt to expel irritants – germs and inflammatory exudates – for effective cure. Hence it is not logical to attempt control of diarrhoea and in fact it may be harmful. But useful constituents for the body that are lost in diarrhoea must be replaced and they include water and electrolytes. It is best done by oral rehydrating solution (ORS) or any equivalent home-made solution that contains ideal composition of sugar with sodium, potassium and bicarbonates as in "limbu-pani" – a glass of water with one teaspoonful sugar, pinch of salt and dash of lemon. Coconut water is another natural alternative. Soft drinks are harmful as they contain large amount of sugar that would worsen diarrhoea. Zinc is known to hasten recovery from diarrhoea and is the only drug to be prescribed in addition to ORS and can be given for 5 days in each episode of diarrhoea. It is most important in malnourished children. Probiotics may help to small extent but are not necessary in acute diarrhoea. There is no need for diet restriction. Soya milk formula is not

necessary in acute diarrhoea. Of course, specific therapy for bacterial or parasitic infections is obviously necessary.

Chronic diarrhoea

It is a multifactorial problem contributed by malnutrition, persistent infection, allergy to animal proteins, malabsorption, bile acid irritation and often worsened by drugs and hence must be properly evaluated. It is beyond the scope of this article. Recurrent episodes of infective diarrhoea (due to poor food hygiene) may be mistaken for chronic diarrhoea but a child with recurrent diarrhoea does recover completely in between episodes as against chronic diarrhoea that is persistent and so presents with progressive loss of weight.

Take home message

Diarrhoea may result from maldigestion (affection of stomach, liver or pancreas), malabsorption (affection of intestines) or intestinal hurry (affection of intestines through local, hormonal or neurogenic disorders) and hence it is rational to make an anatomical diagnosis before proceeding with pathological or etiological diagnosis. At times, chronic constipation can masquerade as loose stools as happens in habitual constipation or congenital megacolon. Detailed history and distended abdomen loaded with faeces can point to constipation as an underlying symptom. Evaluation of what is being lost in diarrhoeal stool is important such as water, electrolytes and nutritional elements. One should not attempt to stop diarrhoea with drugs that may also be dangerous.

MCQs

1. Which of the following statements is right? Stool pattern is considered abnormal if

A) 2-3 stools a day of normal consistency
B) Passing stools immediately after food
C) Change in bowel pattern
D) All of the above

2. Which of the following statements are right? Small intestinal disease may present with

A) Profuse watery stools
B) Stools with mucus
C) Stools with mucus and blood
D) All of the above

3. Diarrhoea is the result of

A) Increased intestinal secretions
B) Decreased intestinal absorption
C) Indigestion
D) All of the above

4. Which of the following statements is wrong? Diarrhoea may be secondary to

A) Urinary tract infection
B) Endocrine disorder
C) Tuberculosis
D) Bacterial otitis media

5. Which of the following statements is wrong? Lactose malabsorption presents as

A) Watery diarrhoea
B) Gaseous abdominal distension
C) Stools with mucus
D) Anal excoriation

Answers to MCQs

Q1 C, Q2 D, Q3 D, Q4 C, Q5 C

2.6. Diarrhoea beyond infections

Clinical application of basic concepts of diarrhoea

Exclusively breast-fed infant never suffers from diarrhoea with rare exception of immune deficiency disorders. However acute bacterial infection such as urinary tract infection may present with parenteral diarrhoea in young infants. Diarrhoea is more common in malnourished infants and children and in turn worsens nutritional status. Well - nourished infant and toddler often suffers from self-limiting viral diarrhoea as a result of small intestinal infection while older children often suffer from bacillary dysentery or parasitic infections such as amoebiasis or giardiasis as a result of large intestinal infection due to ingestion of contaminated food. Non-infective diarrhoea such as inflammatory bowel disease is more common in older children. Thus, age, nutritional status and type of stools suggest probable etiology and in routine practice, even stool examination is rarely necessary in acute diarrhoea. Physical examination is aimed at suspecting probable complications such as dehydration or local complications such as paralytic ileus as evident by abdominal distension.

Case-based study

Case 1

Two months old infant presented with loose stools for last two weeks. He was on exclusive breast feeds since birth. Prior to onset of diarrhoea, he used to pass stools 3-4 times a day which were golden yellow in colour. But since last two weeks, he passed 8-10 stools per day. However, he had been feeding well and also gained weight in spite of diarrhoea. Physical examination did not reveal any abnormality.
There has been change in stool pattern and so needs proper evaluation. However, infant is not sick and feeds well with normal weight gain. This may suggest it may not be pathological. What is then a probable cause of change in stool pattern? Breast milk consists of foremilk – first part of milk-feed which is watery and contains large amount of sugar while hindmilk – later part of milk-feed – contains fat. When infant takes a full feed – both foremilk and hindmilk – stools are formed and infrequent but when infant stops sucking after taking only foremilk and does so at every feed, stools are watery and frequent. So, this infant must be getting distracted while feeding and so takes only foremilk. As it is not sufficient for him, he feeds more frequently and takes again foremilk each time. This is the cause of this infant's loose stools – **foremilk without hindmilk.** There is no need of any tests or drug treatment but mother must be counselled to feed for a longer period at each feeding time without distraction so that infant consumes both foremilk and hindmilk.

Many young infants stop sucking even with half stomach full and then doze off to sleep. Such an infant should be gently tickled to wake up again from slumber and this is the way, infant can be trained to complete a full feed at one time. Detailed enquiry about feeding pattern is necessary to pick-up such problems.

Case 2

Eight months old infant presented with greenish loose stools since last one month. Prior to onset of this problem, he used to pass stools 1-2 times a day with normal colour. There were no other complaints. On direct questioning, mother informed that infant would refuse semisolid food even when she had been trying to introduce it since last two months and he was only on breast feeds. He had not gained weight over last two months. However, he was happy and playful but ever hungry. Physical examination did not reveal any abnormality.
Similar to previous case, there is change in stool pattern but infant is happy and so, it must not have been pathological. What then must be cause of diarrhoea in this child?
Green stools suggest unused bile getting excreted in stools. It could result from intestinal hurry due to infection that does not allow enough time for bile to mix with food but this infant is not sick and so it is unlikely to be an infection. Other cause is **inadequate food intake** because of which unused bile is excreted in stools. This infant is happy feeding on breast all the time, in fact, happy sucking even without adequate intake and is not used to intake any semisolid food. Few infants when exposed to semisolid food reject to begin

with but persistence makes the infant accept it. However, if mother gives in to frequent demand on breast feeding, infant does not learn to eat semisolid food He gets breast addicted and looks satisfied just by sucking even an empty breast resulting in inadequate intake. So this child does not need any tests or drug treatment for diarrhoea but just counselling parents to offer semisolid food while mother eats. It is a stimulus for the infant to imitate and learn to eat well. Children learn best by imitation.

Breast addiction is not uncommon and is the cause of underweight, constipation as well "hunger diarrhoea". Such a child is for ever hungry but refuses to accept other than breast milk. It is only on direct questioning that such a problem comes to attention.

Case 3

Ten years old child was seen for frequent stools for last two weeks. Stools were of normal consistency and colour but frequent. He used to pass stools once or twice a day but last two weeks, he has been passing stools 5-6 times a day. He continued to be happy and playful. Physical examination did not reveal any abnormality.

So, there is change in stool pattern in terms of frequency but colour is normal and so also child is happy and not sick. It indicates that food is well digested and his food intake is normal but there is intestinal hurry. As there is no evidence of infection, intestinal hurry is likely to be due to **stress.** Parents need counselling and no tests or drugs are required. Many may have experienced desire to pass stools when

stressed as happens in a child gong for examination. Similarly, strong gastro-colic reflex makes a child passes stools after eating but in both these situations, stools are normal though frequent.

Case 4

One year old child presented with recurrent episodes of loose stools for last 6 months. Each episode starts with fever and loose stools with mucus and foul smell. Every time he is prescribed different antibiotics and gets well but only to recur after few days. In between episodes, he feeds well, remains active and also gains weight. Though over last 6 months, he has gained only one kg of weight. Physical examination did not reveal any significant abnormality. It is likely to be repeated bacterial infections as suggested by fever and stools with mucus and foul smell. Parasitic infection such as amoebiasis is rare in young children besides it rarely presents as an acute diarrhoea. At any age, repeated bacterial infections strongly suggest either abnormality in the host such as immune deficiency or cystic fibrosis or due to unhygienic conditions. As this child remains well in between episodes and also gains weight, it rules out any significant host abnormality and so it must be due to **poor hygiene**. Thus, treatment of each episode by antibiotics is not going to solve the problem unless parents are counselled about taking adequate care of hygiene.

Bottle feeding is the main culprit for poor hygiene and so also ingestion of contaminated food. Hand hygiene is equally important to prevent food borne infections. Gastro-intestinal

infections are largely preventable and hence, besides treating infection, one must find the root cause behind such an infection. This is the only way recurrence can be prevented.

Case 5

Two years old child presented with abdominal distension, loose stools off and on and loss of weight and appetite over last one year. Frequency of stools varied from 2-4 per day with changing consistency. There was no fever or vomiting. He was treated with antibiotics, anti-parasitic drugs and also enzymes, digestives. Physical examination revealed chronically sick child with gaseous abdominal distension but without any other signs.

It is obviously a chronic progressive disease starting around one year of age. Absence of fever suggests non-infective etiology. However parasitic infections may present without fever but he has failed to get better in spite of anti-parasitic therapy. Parasitic infections are not difficult to treat though they may recur off and on due to poor food hygiene. So, they are recurrent but this child has a persistent disease, as evident by progressive loss of weight. This may be due to chronic malabsorption due to deficient intestinal enzymes. However, such a child would have normal or voracious appetite. As this child has loss of appetite, this malabsorption must have resulted from chronic inflammation. This inflammation is unlikely due to infection as it would have at least partially responded to antibiotics and so it is likely to be due to non-infective inflammation. As it has started around one year of age, on direct questioning to parents, it was

revealed that it coincided with introduction of wheat. This gives a clue that it may be **celiac disease** due to gluten sensitivity or allergy. Serum antibody test and intestinal biopsy can confirm the diagnosis and is managed by avoidance of wheat products as well as rye and barley.Celiac disease is not uncommon. It may present at any age even after months or years of consumption of gluten-containing foods, wheat, rye, barley besides canned fruits, flavoured milk or yoghurt and many processed food items.

Case 6

Eight years old child presented with loose stools with mucus, abdominal pain and poor appetite for last one month and fever off and on for last three weeks. Stools vary in number from 2-5 times a day. Abdominal pain was dull and generalised all over. Fever was mild to moderate. He had lost two kg of weight. Physical examination revealed sick looking child with abdominal distension and mild pallor. There was vague tenderness all over the abdomen. Other systems were normal.

This child has subacute progressive intestinal inflammatory disease as evident by mucus in stools but not so frequent suggesting probably both small and large intestine involved. It is unlikely to be infection as uncontrolled infection would have developed local or systemic complications. Thus, it is mostly non-infective inflammation such as **Crohn's disease.** Neutrophilic leucocytosis with thrombocytosis and hypo-albumenia are investigatory correlates of such a disease. Intestinal biopsy would prove the diagnosis and is treated

with steroids and anti-inflammatory drugs.
Inflammatory bowel disease resembles intestinal infection though subtle points and progress can differentiate one from the other. Etiology remains obscure and role of heredity, stress and diet is considered in causation of this disease. Ulcerative colitis is more common in adults.

Case 7

Eight years old child presented with loose stools and abdominal pain off and on for last 4 months. Stool frequency varies from 2-4 times a day and at times contains mucus. However, appetite activity, play and sleep are unaffected, He has gained one kg of weight over last 4 months in spite of loose stools. Physical examination did not reveal any abnormality. Several tests were carried out without clue to diagnosis and so also drugs were tried but failed.
There is change in bowel pattern since last 4 months but it has not affected general well-being of the child. This fact is important to note and it excludes all pathological conditions. Diagnosis of **irritable bowel syndrome** was made based on circumstantial evidence.
Irritable bowel syndrome is related to stress in a susceptible child and one must find out by discussion what stress the child at this age must be undergoing. Stress may arise from school, home or friends. Parents need to be counselled to observe common areas of stress with friends or at school and they should avoid undue stress at home.

Case 8

Four years old child presented with soiling underwear with small volume of loose stools few times a day for last one month. He was treated for loose stools without any benefit. Physical examination did not reveal any abnormality. On direct questioning, parents informed that besides frequent passing stools with soiling, he would pass good volume of hard stools once in 2-3 days. So, he was constipated as evident by-passing hard stools infrequently. But also. he was passing small volume of loose stools few times a day. As parents were more concerned about loose stools and soiling underwear, history of constipation did not come out as main problem. This is typical of **habitual constipation masquerading as diarrhoea.**Habitual constipation results from diet poor in roughage in a child who has irregular bowel habits. It is often compounded by hurry to go to school in the morning and unhygienic toilet facilities at school that makes a child hold back stools. Retained hard stool leads to stretching and weakness of rectal muscles that makes it more difficult to pass stools. It becomes a vicious cycle. Loose stool proximal to retained hard stool leaks from side of hard stool and keeps on soiling underwear. Diarrhoea is not the problem of this child and he needs management of constipation that is beyond the scope of present article on diarrhoea.

Take home message

First three cases are examples of diarrhoea due to non-pathological causes. What is common to all these cases is that child is not sick and happy. It is a clue to search for non-

pathological causes. Last case masquerades as diarrhoea but actually it is constipation due to poor habits. Other four cases can be diagnosed by analysing history though physical findings are mostly scarce. These cases emphasise importance of detailed history taking. Laboratory tests have limitation and should be reserved for specific diseases.

MCQs

1. Which of the following statement is wrong? Breast-fed infant may pass

A) watery stools
B) frequent stools
C) hard stools
D) Infrequent but soft stools

2. Green stools may suggest

A) infection
B) underfeeding
C) intestinal hurry
D) all of the above

3. Most common cause of recurrent GI infection in infant is

A) immune deficiency
B) cystic fibrosis
C) bottle feeding
D) All of the above

4. Which of the following statement is wrong? Blood and mucus in stools is seen in

A) bacillary dysentery
B) inflammatory bowel disease
C) intestinal viral infection
D) intussusception

5. Which of the following modality is most useful in evaluation of diarrhoea?

A) Detailed history
B) physical examination
C) Laboratory tests
D) All of the above

Answers to MCQs

Q1 C, Q2 D, Q3 C, Q4 C, Q5 A

2.7. Constipation- gut in slow motion, constant anticipation without culmination is constipation

Back to basics – understanding constipation

What is constipation?

Change in bowel pattern resulting in infrequent passage of stools, often hard and at times accompanied with painful defecation and may be stool smeared with blood. Incomplete evacuation of stool with frequent desire to pass stools must also be seen as constipation. Frequency of passing stools varies in normal individuals from 2-3 times a day to once in two days. It is the change in frequency and / or consistency that is to be considered. Individual passing stools three times a day, suddenly passes one stool a day is a change that needs

to be noted. Infant on exclusive breast feeding may pass a stool once in few days but it is soft, painless and infant is happy growing well, it is not considered as constipation because it is not hard and frequency has remained the same.

Physiology of bowel movements

After food is digested and absorbed by small intestines, it is passed on to large intestine. Colon and rectum function together to provide absorption of water, electrolytes and short chain fatty acids, to dehydrate the faecal matter, to store unwanted part and help in evacuation.in a socially appropriate manner when reflex urge sets in. If stool matter remains in colon and rectum for longer time, it becomes harder and more difficult to expel. Continence is maintained by coordinated function of pelvic floor, rectum and anal sphincters. Evacuation occurs through relaxation of pelvic floor. Rectum acts to store as well as expel stool matter, both actions require cortical sensory awareness acting in conjunction with intramural and spinal reflexes that ensure timely defecation. Anal sphincters act individually and in unison in response to rectal distension and sensation of rectal filling. There has to be adequate amount of stool matter for rectal distension enough to feel sensation of filling. Reflex relaxation of internal sphincter has an additional sensory function that allows rectal stool matter to move into upper anal canal. Voluntary control of external sphincter allows deferring evacuation till opportunity exists.

Prerequisites for normal bowel movements

Consumption of adequate amount of food is necessary to

form volume of stool enough to distend rectum so that sensation of rectal filling is perceived by cortex. There has to be enough insoluble fibre in diet to form bulk of soft stool matter so that it moves smoothly through colon and rectum and is expelled without difficulty. Bowel movements depend upon normal intestinal muscle function leading to peristalsis. Once rectal filling results in urge to pass stools, intact nervous system acts through reflex action to relax internal sphincter and pelvic floor muscles and move the stool matter into upper anal canal, possible with good anorectal tone. Finally intact voluntary control of external anal sphincter is necessary to defer evacuation to right moment. Thus, multiple factors contribute to normal bowel movements and disturbance of any of these factors may result in constipation.

Types of constipation *Primary constipation* that is due to intrinsic problem in colon and anorectum related to abnormal function of normally innervated and structurally intact muscle. This may result from dyssynergia between external sphincter and puborectalis muscle. In such a case, there is incomplete emptying of rectum in spite of soft stools. *Functional constipation* is a result of gut-brain interaction. It is contributed by altered motility, visceral hypersensitivity, altered mucosal and immune functions and change in intestinal microbiota, It could be worsened by stress. *Habitual constipation* is a type of primary constipation and is known as constipation of modern civilisation. It is due to improper eating habits (inadequate intake of fibre and water), poor physical exercise and defecation without

squatting (use of toilet seat) resulting in anorectal angle that is not conducive for smooth expulsion. Irregular bowel habits due to rush in the morning to go to school and holding back stools through school period adds to habitual constipation. As passage of stools become painful, child learns to hold back the stool in spite of an urge and it worsens the problem. *Secondary constipation* is due to structural defects (Hirschprung disease), chronic systemic illness (inflammatory bowel disease, intestinal TB, chronic diverticulitis), neurological (spinal or cortical disorders), hormonal (hypothyroidism, hyperparathyroidism), metabolic disorders (hypercalcemia, cystic fibrosis), lead poisoning and also side effect of few commonly used medications (calcium and iron supplements, anti-spasmodic like dicyclomine, pain relievers such as NSAIDs, propranolol etc)

Red flags in constipation

Following symptoms and signs are indicators of need for prompt action and further investigations. They include sudden development of constipation for no apparent reason, weight loss, severe abdominal pain, significant rectal bleeding and recent onset of severe constipation lasting for more than two weeks in spite of proper management. Delayed passage of meconium on first day of life may be initial symptom of secondary constipation such as in hypothyroidism or Hirschprung disease. Breast fed infant always passes soft stools irrespective of frequency and hence if constipated, always needs further investigations,

Harmful effects of persistent constipation

Constipation in children must be successfully treated without delay because persistent constipation results in loss of rectal tone and perpetuates worsening constipation. Urinary tract infection often results from constipation due to pressure over lower urinary tract by distended rectum. Besides, such harmful effects, constipation also causes significant discomfort due to bloating of abdomen and pain and appetite may be disturbed leading to undernutrition. Thus, constipation in children must be viewed as potential permanent harm and must be addressed promptly.

Management of constipation

Causes of secondary constipation should be ruled out clinically and in case of suspicion should be investigated as these problems can be treated effectively with appropriate intervention.

Immediate short-term aim in management of primary constipationis to ensure bowel movement every day to avoid permanent damage, for which drugs may be necessary. However, drugs must be used sparingly. Balanced diet with adequate fibre and intake of water, life style change with proper toilet habits and physical exercise and behavioural modification with coping with stress form the mainstay of long-term management.

Fibre is a natural bulk laxative that works by increasing bulk of stools. Prunes have lots of insoluble fibre and also has sorbitol that is laxative. Besides, it also has soluble fibre that is fermented in colon to produce short chain fatty acids that add bulk to stools. Prunes also stimulate beneficial gut

bacteria. Apple, pear, kiwi, figs, citrus fruits, spinach and greens, sweet potato, beans, peas, lentils, flaxseeds, whole grain and oats are some of the food items that are very useful. One must avoid fried food, chips, cookies, lots of dairy products and red meat.

There are different types of laxatives used in management of primary constipation. They include lubricating laxatives such as mineral oil, emollient laxatives or stool softeners such as lactulose, osmotic laxatives such as milk of magnesia, sorbitol and polyethylene glycol and stimulant laxatives such as senna. Increasing fibre in the diet as bulk laxative along with stool softeners are first line drugs. Osmotic laxatives may have to be added in case of stubborn constipation. Rectal suppository or enema should be reserved only for emergency situation for temporary use. They are not ideal for long-term use as they induce fear in children.

<u>Prevention of constipation</u>

Exclusive breast feeding for first 6 months followed by complimentary feeds consisting of suitably modified family food, continuing breast feeds for at least first year of life promotes ideal bowel movements. Good eating habits with consumption of vegetables and fruits, adequate water intake, physical exercise with outdoor activities and ideal toilet habits help in prevention of constipation. Once such habits are established in early childhood, they are sustained over subsequent years.

Take home message

Constipation is a common problem in children and is often ignored in initial stages till it becomes troublesome. Ideal habits promote good bowel movements. Secondary constipation must be suspected clinically and treated accordingly while primary constipation is managed by diet and life style modification with stool softeners or osmotic laxatives as per the need. Long term use of drugs must be avoided.

MCQs

1. What is constipation?

A) Change in usual bowel pattern
B) Soft stool with poor emptying in spite of straining
C) Hard stools with difficulty in evacuation
D) All of above

2. Which of the following mechanisms are not under voluntary control?

A) Rectal distension
B) Relaxation of internal sphincter
C) Relaxation of pelvic floor muscles
D) None of the above

3. Which of the following type of constipation presents with soiling of underwear?

A) Secondary constipation
B) Habitual constipation
C) Functional constipation
D) None of the above

4. Which of the following conditions need urgent action?

A) Acute onset of constipation
B) Severe abdominal pain
C) Weight loss
D) All of the above

5. Which of the following statement is wrong? To prevent constipation,

A) Diet must contain enough fibre
B) Plenty of dairy products are useful
C) Ideal physical exercise
D) Good toilet habits

Answers to MCQs

Q 1 D, Q 2 D, Q 3 B, Q 4 D, Q 5 B

2.8. Constipation – a challenge in diagnosis and management

Clinical application of basic concepts of constipation

Acute onset of constipation is brought to the notice of a doctor but patient complains of chronic constipation only when it becomes troublesome and painful. History in every case should include enquiry about stooling as a routine part of personal history. Incomplete emptying of rectum resulting in frequent urge to pass stools is also a part of constipation and at times, habitual constipation presents as frequent soiling of underwear with loose stools and mistaken for diarrhoea. Constipation is an emergency because of potential

dangers of chronic persistent constipation. Early diagnosis of constipation and timely successful management are important.

Case-based study

Case 1

Two years old child presented with history of progressive abdominal distension and constipation since early infancy. It was gradually getting worse. He had poor appetite and had lost weight over last one year. There was no vomiting though had occasional episodes of diarrhoea and had no other symptoms. His development (milestones) was normal. He was breast-fed during first 6 months followed by introduction of complimentary feeds.

It is chronic progressive constipation starting early in life. Infant on exclusive breast feeding never suffers from constipation and hence it is certainly a pathological disorder. Abdominal distension with constipation without vomiting suggests chronic colonic obstruction. Prolonged constipation may lead to occasional diarrhoea as stagnated excreta promotes infection.

Physical examination revealed marked abdominal distension loaded with faeces. Per-rectal examination led to ribbon like stool coming out indicating narrow colonic passage leading to obstruction. This is typical of congenital megacolon – **Hirschsprung disease**. It is due to absence of nerve cells in the muscles of colon because of which colon can't evacuate stool. Diagnosis can be confirmed by barium enema and rectal biopsy and is managed surgically.

Exclusively breast-fed infant never is constipated though stooling may be infrequent but soft. Chronic intestinal obstruction at that age is due to congenital megacolon. One may be able to suspect it even close to birth because of delayed passage of meconium. Normally meconium is passed within first 24-48 hours.Hirschsprung disease may be easily missed in initial years depending upon length of colon affected by absence of ganglion cells. Smaller the segment involved, later is the diagnosis made. This is because constipation is initially manageable but over time, abdominal distension and failure to gain adequate weight attract attention to facilitate correct diagnosis.

Case 2

Two years old child presented with gradually progressive abdominal distension and constipation since early infancy. He was gaining weight well though was short in length / height. He was lethargic, not active and had delayed milestones. At two years of age, he was not able to speak even few words. Physical examination showed dull looking child with puffiness of face, dry skin, hoarseness of voice, bradycardia, abdominal distension and umbilical hernia. Per rectal examination was normal. This child besides being chronically constipated, has delayed development and lethargy. It suggests **congenital hypothyroidism** or cretinism.

This case is similar to previous one but there are many subtle differences. Child with congenital megacolon fails to gain weight but mentally normal while hypothyroid child fails to gain height and mentally subnormal. Both conditions can be

suspected within first 2 days of birth by delayed passage of meconium. In case of congenital megacolon, meconium passage is delayed because of colonic obstruction while in hypothyroidism, it is due to sluggishness of intestinal muscle and so poor peristalsis. Both conditions should be suspected early in life to avoid permanent damage. It is most important in case of hypothyroidism as even small amount of delay in diagnosis and treatment may lead to permanent brain damage. In fact,in most centres in the country, cord blood is tested for TSH to screen for congenital hypothyroidism so as to prevent permanent brain damage. If TSH is high, further tests should be carried out to confirm the diagnosis. Incidence of congenital hypothyroidism is 1 in 2500 births and it is very high but preventable.

Case 3

Two years old child presented with gradually progressive constipation since the age of 9 months. He was consuming one litre of milk every day but was reluctant to eat solid food. His weight gain had slowed down in second year though he was very active and alert. Prior to development of constipation, he used to pass normal soft stools. Physical examination showed mild abdominal distension but no other abnormalities.

Constipation in this child has started later in first year of life and has been slowly progressive without much disturbance to child's health and activity. Thus, it is not likely to be due to any defect or disease but result of **excessive milk intake and**

poor eating habits. He has good appetite but his intake is restricted only to milk. This is because of feeding bottle addiction as he would consume 6-8 ounces of milk twice through the night while half asleep. Further attempt to force feed the child by mother had made this child stubborn and would just starve and cry for milk. Thus, sole cause of constipation in this child is faulty eating habits due to bottle addiction. This child would not improve unless his habits are changed. Temporary stool softeners may be necessary.
It is important to cultivate ideal eating habits right from 6 months of age. At that age, infant is eager and ready to put anything in mouth – it is a normal reflex. By about one year of age, most infants develop addiction either to breast or bottle feeding and this is the time to wean off gradually. If not done at that right time, child refuses to eat solid food and gets hooked on to milk feeds only. Further forced feeding should be always avoided. It is best to allow the child to starve but not to offer milk more than 2-3 times a day.

Case 4

Five years old child presented with gradually progressive constipation over last 2 years. Initially he would pass hard stools though each day, gradually frequency reduced to once in 2-3 days and stool was getting more difficult to pass with pain and occasional stool smeared with blood. He had no other significant symptoms. His appetite was average and so also his weight. He was active and playful. Physical examination did not reveal any significant abnormality. Constipation is this child has started around 3 years of age

and has been very slowly worsening but without significant disturbance in his health or activity. Thus, it is unlikely to be due to defect or disease. On direct questioning, it was found that he was consuming imbalanced diet as it contained very little fibre. He also drank little water. He was often consuming “junk” food and had poor intake of vegetables and fruits. This is known as **habitual constipation**. In addition, he would have to rush to school in the morning without passing stools and would withhold stool in school due to unclean toilet. This worsened his constipation. His diet needs drastic change and he should make a habit of passing stool every morning. Western toilet is not ideal to pass stools easily as against Indian type of squatting that maintains anorectal position straight facilitating easy expulsion of stool.Such constipation is a symbol of modern civilisation. This case is similar to previous one as eating habits came in the way of constipation in both these children. Parents are totally responsible for inculcating right eating habits in early childhood to ensure good health. Management of constipation in such children revolve around parental cooperation to change habits. However, it is ideal to ensure right eating habits right from one year of age, Otherwise, habits die hard.

Case 5

10 years old child presented with constipation and abdominal pain off and on for last one year. His stooling pattern would vary in frequency and consistency and also in timing of passing stools. Most of the time he would tend to be

constipated with occasional normal stool. He also had abdominal pain that also varied in frequency and severity with irregular pattern. His appetite was average and had not lost weight. His diet was essentially normal and so also his water intake. Physical examination did not reveal any significant abnormality.

Constipation in this child obviously is not due to any defect or disease but also not a result of poor diet or toilet habits. On direct questioning, it was realised that this child was always stressed and worried about his performance in every field. This is typically referred to as **functional constipation**. It is a result of gut-brain interaction. It is now well known that intestines govern brain emotions. If one sees something frightful, intestines cramp first before brain realises danger. When one goes to attend an examination or similar stressful event, one gets an urge to use toilet once more. And finally in colloquial language, we use the term – gut feeling that is superior to brain feeling. Intestinal complaints are most common due to such functional disorder and they may present with change in bowel pattern and abdominal pain. Such a child as well as parents need counselling and if necessary, psychological help.

Case 6

10 years old child presented with episodes of constipation, vomiting and abdominal distension three times over last 6 months. Each timehe received conservative treatment with IV fluids and rest to intestines that recovered him within 3-4 days. In between these episodes, he did not have any

symptoms referable to abdomen but had poor appetite, felt generally weak and had lost 2 kg weight over last 6 months.Physical examination during last episode showed abdominal distension with poor peristalsis and mild tenderness without any lump in abdomen. It suggested intestinal obstruction. Recurrent episodes of intestinal obstruction with intervening period of feeling unwell and loss of weight indicated persistent slowly progressive disease with intermittent symptoms of intestinal obstruction. It favours diagnosis of **intestinal Tuberculosis**. Barium meal would demonstrate narrowing of intestinal segments and biopsy may confirm diagnosis of tuberculosis.
Intestinal tuberculosis affects submucosa and not mucosa and hence does not present with diarrhoea. As disease progresses, attempt at natural healing leads to fibrosis of affected submucosal lesion resulting in partial intestinal obstruction. It may be relieved temporarily with conservative treatment only to recur again. Occasionally, constipation may alternate with loose stools due to transient mucosal inflammation though constipation is the major symptom in such cases.

Case 7

10 years old child presented with recurrent episodes of constipation, abdominal distension and vomiting three times over last one year. Each time he would recover with conservative treatment and in between episodes, he would be fully normal, had gained weight and remained active. Two years prior to these episodes, he had undergone abdominal

surgery for Meckel's diverticulum. Physical examination during last episode showed signs of intestinal obstruction. As this child has remained well during intervening period, it looks like a mechanical problem rather than inflammatory cause. Thus, it may be due to development of adhesions following previous surgery. Such a child may need re-exploration though with a possibility of **recurring adhesions**. This case is similar to previous one except that this child has been normal in between episodes as against previous child who was unwell during intervening period. It clearly indicates that this child has recurrent problem with recurrent symptoms whereas previous child had persistent problem with recurrent symptoms. Thus, it is important to differentiate persistent disease with recurrent symptoms from recurrent disease with recurrent symptoms. Sickness or wellness during Intervening period helps to differentiate the two conditions.

Case 8

10 years old child presented with constipation and abdominal pain over last two months. A year ago, he was diagnosed to be suffering from inflammatory bowel disease and was on treatment for the same. His main symptoms then were loose stools with mucus and abdominal pain. His disease was partially controlled with anti-inflammatory drugs and diet modification. Physical examination showed abdominal distension with colon loaded with faeces. It is clear that constipation in this child should be related to his pre-existent inflammatory bowel disease. It may be aggravated by drugs

prescribed for IBD or low fibre diet and / or poor intake of water. However, one must rule out any evidence of stricture formation due to intestinal inflammation that may need surgical intervention. It can be confirmed by intestinal imaging study.

This case illustrates possibility of constipation in diseases that generally present as loose stools such as inflammatory bowel disease or irritable bowel syndrome. On the other hand, diseases that present with constipation as major symptom may also present with occasional loose stools as happens in intestinal tuberculosis or hirschsprung's disease.

Take home message

Constipation may present with varied causes. Some of them may just need modification of diet or life style while others require specific intervention to cure it. Early diagnosis and treatment would avoid long-term damage. Thus, constipation should not be taken lightly and every attempt must be made to find right cause. Laxatives or stool softeners are only temporary measures till the problem is permanently solved.

MCQs

1. Which of the following statement is wrong? Constipation in early infancy may be caused by

A) Formula feeding
B) Poor intake of water
C) Congenital hypothyroidism
D) Congenital megacolon

2. Besides abdominal symptoms, evaluation of constipation must include

A) Diet and life style
B) General well-being
C) Mental milestones
D) All of the above

3. Which of the following statement is wrong? Modern civilisation has contributed to constipation because of

A) Junk food
B) Electronic media
C) Western toilet
D) None of the above

4. Which of the following statement is wrong? Constipation and loose stools may coexist in

A) Inflammatory bowel disease
B) Congenital megacolon
C) Congenital hypothyroidism
D) Intestinal tuberculosis

5. Which of the following statement is right? Drug of first choice in constipation is

A) Lubricating laxative such as mineral oil
B) Stimulant laxative such as senna
C) Stool softeners such as lactulose
D) Osmotic laxatives such as milk of magnesia

Answers to MCQs

Q 1 B, Q 2 D, Q 3 D, Q 4 C, Q 5 C

2.9. Vomiting - look beyond GI system

Back to basics – vomiting

What is vomiting

Vomiting is involuntary forceful expulsion of contents of stomach and intestine while regurgitation is return of ingested food from stomach into oesophagus and mouth without force. It is important to differentiate one from the other because regurgitation is a common natural event in young infants and should not be confused with vomiting. Nausea is merely a sensation of vomiting and may precede act of vomiting but may not end up with vomiting.

Pathogenesis of vomiting

Vomiting may be a symptom arising from many organs. Typically, vomiting due to gastro-intestinal system disorder is caused by obstruction or inflammation in upper GI tract. Besides GI disorders, receptors in the floor of fourth ventricle in the brain represents chemoreceptor trigger zone (CTZ) know as area of postrena which if stimulated causes vomiting. CTZ may be stimulated by either raised intracranial pressure or by abnormal metabolites arising from disturbed metabolic functions in the body as happens in liver disease (acute hepatitis), chronic renal disease and inborn errors of metabolism. Autonomic nervous system through its network may also lead to vomiting as may happen in case of acute stress of any kind such as acute cardiac disorder or extreme physical stress as in a long distance marathon and also due to adrenal disorders. Stimulation of vestibular part of 8th cranial

nerve is responsible for vomiting in motion sickness and stimulation of vagus – 10th cranial nerve due to pharyngeal or gastric mucosal irritation leads to vomiting. Some drugs also lead to vomiting, in particular, anti-cancer drugs. Severe bout of cough often ends up in vomiting. It is clear that cause of vomiting may lie in various different organs and not just in GI system, though vomit finally comes out of GI tract.

Natural response to vomiting

There is an increased salivation as nature's attempt to protect enamel of teeth from coming in contact with acid from stomach. Retroperistalsis starts from intestines sweeping all the contents upwards with relaxation of pyloric sphincter. Intrathoracic pressure lowers with deep inspiration and glottis is closed that prevents aspiration. Intra-abdominal pressure is increased with relaxation of lower oesophageal sphincter facilitating expulsion of stomach contents. It is often accompanied with sympathetic response leading to sweating and tachycardia.

Contents of vomitus

Commonly vomitus contains ingested food particles. Once stomach empties after repeated vomiting, all that vomitus contains is just small amount of mucus. Problem arising from obstruction beyond second part of duodenum results in bile-stained vomit. Repeated attempts to vomit with an empty stomach may cause retching that may also end up with greenish vomit due to forceful expulsion of duodenal contents. Occasionally, such a retching episode injures small blood capillaries in oesophagus resulting in streaks of blood.

It is referred to as Mallory-Weiss tear. It is caused by mechanical force of vomiting and is self-limiting. At times vomitus may contain large amount of blood as commonly happens in case of oesophageal varices due to portal hypertension. Coffee ground vomit indicates that blood has stayed in the stomach for some time before vomiting and is due to iron in blood getting oxidised. Blood in vomitus may also be a result of severe gastritis or in adults due to stomach cancer. Faecal vomit may suggest gastrocolic fistula.

Complications in vomiting

Frequent episodes of vomiting may lead to destruction of enamel of teeth due to acid contact. In young children, senior citizens and patients with neurological diseases, aspiration of vomitus is a risk. Severe vomiting may result in dehydration and electrolyte disturbances leading to hyponatremia, hypochloremia and metabolic alkalosis. Resultant metabolic alkalosis may cause hypocalcemia. Persistent vomiting for several days cause malnutrition and constipation.

Red flags in vomiting

Vomiting occurring few days after onset of other symptoms suggests a serious underlying cause that must be promptly diagnosed and treated appropriately. On the other hand, vomiting at the onset of the disease generally indicates local upper gastrointestinal problem and often self-limiting as in case of gastroenteritis. However persistent vomiting beyond two days in such a scenario indicates serious nature of disease. Reduced urine output suggests dehydration that needs correction and so also electrolyte disturbances.

Significant abdominal distension may be due to intestinal obstruction or paralytic ileus. Severe abdominal pain indicates either inflammatory or vascular disease (due to intestinal ischemia). Accompanying neurological symptoms such as headache, drowsiness or seizures signify serious neurological disease.

Management

it is most important to find out cause of vomiting that would lead to rational therapy of the disease. However, symptomatic control of vomiting may be tried for first one or two days. Generally symptomatic treatment does not work well though vomiting is controlled in benign conditions often on its own. Hence it is not rational to continue anti-emetic drugs for more than few days at most, exception being vomiting due to chemotherapeutic drugs. Ondansetron, metaclopramide or domperidone are the drugs used for symptomatic control and rarely injectable form may be used in case of drug not being retained orally. Common side effect of most antiemetic is extra-pyramidal syndrome leading to dystonia and abnormal movements.

Take home message

Vomiting occurring few days after onset of other symptoms usually has underlying serious disease while vomiting that starts at the onset of disease is usually of gastrointestinal origin and often benign though may continue as well. Persistent vomiting demands proper evaluation. Symptomatic treatment is not much effective and should not

be continued beyond first two days, instead demands proper diagnosis and management of primary disease.

MCQs

1. Vomiting is not a symptom of affection of this system in the body

A) Endocrine B) Haematology C) Respiratory D) Renal

2. Which of the following diseases trigger vomiting centre in the brain

A) Epilepsy B) Whooping coughC) Uraemia
D) Arterial stroke

3. This type of vomiting is not likely to be serious

A) Vomiting as first symptom with abdominal distension
B) Vomiting as first symptom without abdominal distension
C) Vomiting occurring after few days of onset of disease
D) Persistent vomiting beyond 2 days

4. Which of the following statement is right? Persistent / severe vomiting may lead to

A) Hypochloremia
B) Hypocalcemia
C) Hyponatremia
D) All of the above

5. Which of the following antiemetic drugs can stop vomiting completely?

A) Ondansetron
B) Metaclopramide

C) Domperidone
D) None of the above

Answers to MCQs

Q 1 B, Q 2 C, Q 3 B, Q 4 D, Q 5 D

2.10. Vomiting – need for urgentdiagnosis

Clinical application of basic concepts in vomiting

Vomiting may not be considered only as primary gastrointestinal disorder as it is caused by diseases of almost every system in the body, though finally it is mediated through GI system. Thus, attention should be given to every organ in the body that may be responsible for vomiting. Self-limiting vomiting due to transient gastritis generally stops within first two days.Vomiting persisting beyond 48 hours is unlikely to settle by itself and merely continuing anti-emetic drugs is irrational. Acute onset of vomiting due to metabolic disorders may not reveal any abnormal physical findings and need clinical suspicion based on exclusion of other causes.

Case-based study

Case 1

Two months old infant presented with history of vomiting for last two weeks. Infant was well till the onset of this symptom and was on exclusive breast feeds and growing well. Vomit is forceful and contains large amount of curdled milk. Prior to vomiting, infant is restless but feels relieved after a vomit and takes feeds well. However, vomiting recurs after few

hours again. Last two weeks, he passes stools infrequently and has not gained weight since then. Physical examination showed mild dehydration and distension of upper abdomen more in left hypochondrium and epigastrium. There were no other abnormalities.

In view of forceful vomiting containing curdled milk without bile and distension of upper abdomen, it suggests pyloric obstruction. As infant is otherwise asymptomatic, it is due to mechanical cause that must have been present since birth. Hence it is **congenital pyloric stenosis**. Question arises why it did not present at birth. It is not a complete obstruction as in atresia but partial obstruction and hence referred to as stenosis. Over first few weeks, due to partial obstruction, curdled milk is retained in stomach for longer time that causes chemical inflammation of pylorus due to acidic nature of curdled milk. Thus, over time, congenital partial obstruction becomes complete and hence there is delayed presentation. Rarely if pyloric obstruction is near complete at birth, neonate may present with pyloric stenosis within few days. Diagnosis can be easily confirmed by imaging study and needs surgical correction of the defect.

Absence of general symptoms such as fever in an otherwise well infant indicates mechanical cause. Forceful vomit suggests obstruction. Vomitus not containing bile localises obstruction proximal to second part of duodenum. Presenting so early in life favours a congenital lesion. Hence diagnosis is easy to arrive at and is totally curable with surgery. It is important to diagnose early enough to avoid dehydration and malnutrition. It is ideal to confirm the

diagnosis as other causes of obstruction proximal to second part of duodenum may simulate pyloric stenosis.

Case 2

Two months old infant presented with history of acute onset of vomiting followed by drowsiness. He was apparently well prior to onset of this illness. He was on exclusive breast feeding and was gaining weight well. Physical examination showed drowsy infant barely responding to painful stimuli. There were no other abnormal findings.
What starts acutely may be due to mechanical, neurological, vasogenic, metabolic or immune mediated disorder. There has been no history of trauma. Vascular etiology affects localised part of the brain related to the vascular supply and hence not likely in this child. Immune mediated disorders are rare at this age and are often accompanied with general symptoms such as fever or skin rash. Primary neurological condition that may result in acute onset of drowsiness is viral encephalitis but would have presented with fever, seizures and other prodrome of viral infection such as cold, cough etc. Thus, it is mostly a **metabolic disorder due to inborn metabolic defect.** It is not possible to guess which metabolic disorder it could be. This infant was hospitalised and put on IV fluids and some laboratory tests were ordered. Within 24 hours, infant improved dramatically and seemed to be normal. This is typical of metabolic disorder as oral feeds were stopped, offending agent coming through feed was withdrawn and resulting in fast improvement. Further laboratory tests confirmed it to galactosemia. Infant is born

with this type of metabolic error. It is treated with lactose and galactose free diet. Early diagnosis and treatment prevent permanent neurological handicap.
Acute onset of vomiting and drowsiness clearly denotes central cause of vomiting. Sudden onset and equally sudden recovery on withdrawal of oral feeds indicates metabolic disorder. Further laboratory tests help in pinpointing a cause.

Case 3

8 months old infant presented with vomiting since last one month and cough off and on. He was apparently well till onset of present problem. He would start feeding but would vomit while feeding and vomitus contained milk as such. Occasionally, he would suddenly chock while feeding with severe cough. It kept on happening over last one month. At times, milk would go down the stomach without vomiting. Physical examination did not show any abnormality.
As vomitus contains milk as such and not curdled milk, it is clear that milk does not reach stomach at all and so problem lies in oesophagus. When liquids cannot go down oesophagus, it is unlikely to be due to mechanical obstruction and hence it must be because of functional obstruction. It suggests malfunction of oesophageal muscle that does not allow liquids to travel down. Retained milk is mostly vomited out but occasionally would be inhaled into airways and result in severe chocking and cough. Further investigations demonstrated **achalasia cardia** – failure of oesophagus to relax and so remains mostly contracted. It is managed by repeated dilatation. When

solid food can't go down the stomach, it is likely to be mechanical obstruction but when solid food can go down due to gravity but liquids can't do down, it is often a symptom of muscle dysfunction, often of neurological abnormality. Achalasia is a condition in which oesophagus does not relax and so obstructs the passage of ingested material. It may lead to aspiration of ingested material into the airways. Chalasia is just the opposite of achalasia in which oesophagus remains dilated facilitating acid reflux from stomach and also aspiration into the airways. Older child and adult would also present with retrosternal burning sensation.

Case 4

8 months old infant presented with history of vomiting for last 2 months. He was well prior to onset of this illness. He was growing well. He vomits each time he feeds and in general reluctant to feed. He had lost 2 kg weight. Apparently, there was no other symptoms. Several tests and trials with medicines had failed. Physical examination revealed malnourished sick looking infant, weighing 5.2 kg but with no other abnormal findings. On direct questioning, it was found out that he was very irritable, constipated but passing lots of urine in spite of vomiting and severe anorexia. So, he had polyuria. Thus, it was due to **renal tubular disorder**. Further tests confirmed the diagnosis of hypercalcemia. It was a result of vitamin D toxicity. This case demonstrates importance of detailed personal history that pointed to polyuria. Generally, one tends to ask for oliguria as a marker of renal disease but renal tubular

disorder presents as polyuria besides other metabolic defects. There are no abnormal physical findings in such a metabolic disorder and it is only a detailed history that gives a clue to probable diagnosis. It is an abnormal metabolite that stimulates central trigger zone to cause vomiting.

Case 5

4 years old child presented with vomiting for last two months. He was well prior to onset of this illness. He would vomit off and on and vomitus would contain ingested food. There was no history of headache or any other symptoms. Physical examination did not reveal any abnormality.
It suggests central cause of vomiting. In absence of any other clue, one may have to investigate for metabolic disorder. Initial screening tests showed low serum sodium and high potassium. It suggests an adrenal disorder. Few days later this child developed neurological abnormality in the form of spasticity and deterioration of milestones. MRI of brain confirmed the diagnosis of **adrenoleucodystrophy.**It is a rare disease but this case emphasises a point that few diseases evolve over time to reveal a final diagnosis. However, it was easy to consider that vomiting in this child was due to chronic adrenal disorder. Acute adrenal disorder presents as shock. Rarely, adrenal tuberculosis may also present similarly but would have other symptoms such as low grade fever and loss of appetite and weight with evidence of primary lung disease. Diseases with multiple symptoms may start with only one symptom and then come out with other symptoms. In such a case, one may have to keep in mind an evolving

disease and even search for it with a single symptom at presentation.

Case 6

8 years old child presented with history of vomiting, severe abdominal pain and mild fever for last 24 hours. It started with vomiting followed by abdominal pain. Pain was periumbilical, dull and poorly localised. He had not passed stools for a day. Vomitus contained ingested material. He developed mild fever on D2. At this juncture, it was clear that as pain was the major symptoms, there was intra-abdominal inflammatory pathology evolving without obvious localisation. It is unlikely to be intestinal obstruction as there was no bile-stained vomit. By next day, pain had increased in severity and was localised to right iliac fossa. It clearly meant localisation now to appendix and hence diagnosis of **acute appendicitis** was made. It can be confirmed with USG. He was operated and recovered fully.

This case illustrates importance of enquiring about chief complaint. This child's chief complaint was abdominal pain but parents were worried about vomiting more than abdominal pain. Vomiting in such a case is related to autonomic response to pain. Visceral abdominal pain – related to viscera – is often periumbilical and dull with poor localisation till it localises to the site of inflammation with involvement of peritoneum and leads to increased severity of pain.

Case 7

10 years old healthy child presented with episodes of vomiting and headache on getting up in the morning that would ease over time only to recur next morning again. Severity and duration of these episodes went on increasing over next two weeks. There was no fever. These symptoms suggest increased intracranial pressure that is gradually worsening. And hence it is likely to be space-occupying intracranial lesion. Physical examination revealed papilledema on fundus examination and increased tone and brisk deep tendon reflexes in both lower limbs. It suggests slowly increasing hydrocephalus that was proved to be due to **cerebral aqueduct obstruction**. This child was operated for the same and had ventriculo-peritoneal shunt placed. Vomiting accompanied with headache is classic of raised intracranial pressure. Slow onset of raised ICP manifests first on waking up in the morning as during lying down position, there is venous stasis in intracranial compartment that adds to the borderline increased pressure and hence manifests on waking up. With routine movements, intracranial stasis gets reduced and so symptoms of vomiting and headache subside temporarily till pressure further increases and then symptoms get worst through the day.

Case 8

5 months old infant presented with vomiting since last 2 months. Prior to onset of vomiting, he was happy and growing well on exclusive breast feeds. For first three months, mother was at her mother's place with her infant. Since she came back to her home, vomiting started and it

continued. Physical examination did not reveal any abnormality though infant had not gained any weight over last two months and was irritable. Several tests done were all negative and so also empirical therapeutic trials had failed. History of change in place coincided with onset of symptoms and hence it was decided to go into further details.
On further discussion with the family, psychologist found out the cause of **disturbed mother-infant relation** as the cause of vomiting. Elder aunt of the infant had taken charge of the baby and mother was allowed to handle the baby only during breast feeding. This infant was craving for mother's bonding. Father of the infant was taken into confidence and he managed to send his elder sister out of town on some pretext and from the very day, infant stopped vomiting. So, it was a functional disorder and not any organic disease.
This case illustrates possibility of functional disorders even in a young infant. Mother-infant relationship is vital for infant's secured feeling and it promotes not only sound health but also ideal psychological development. Health is not just physical well-being but also mental, psychological and emotional well-being. Factors disturbing any of these components would affect health and manifest with physical symptoms simulating organic disorder.

Take home message

Vomiting is a symptom that may arise from almost every system in the body. Thus, anatomical diagnosis is a must. Disease may evolve over first few days to suggest a diagnosis As a rule, persistent vomiting beyond first two days definitely

needs further evaluation and mere continuation of anti-emetic drug is not justified. Vomiting due to metabolic disorders present often without any abnormal physical findings and diagnosis depends on strong clinical suspicion based on analysis of history.

MCQs

1. Which of the following statement about congenital pyloric stenosis is wrong?

A) May present at birth
B) May present at 4 weeks of age
C) May present at 8 weeks of age
D) May present after one year of age

2. Which of the following statement is wrong? Vomiting due to upper GI obstruction may be caused by
A) Congenital malformation
B) Muscle dysfunction
C) Infection
D) Neurological disorder

3. In this condition, child recovers completely within a day if not fed orally but recurs shortly

A) Pyloric stenosis
B(Inborn error of metabolism
C) Acute appendicitis
D) None of the above

4. In this condition, vomiting is the most predominant symptom

A) Raised intracranial pressure
B) Acute appendicitis
C) Metabolic disorders
D) All of the above

5. Which of the following symptoms must be enquired when child presents with vomiting?

A) Oliguria
B) Polyuria
C) Both of the above
D) None of the above

Answers to MCQs

Q 1 D, Q 2 C, Q 3 B, Q 4 C, Q 5 C

2.11. Abdominal pain – a varied pathogenesis

Back to basics – abdominal pain

What is pain?

Painis an unpleasant sensory and emotional experience arising out of an underlying cause. It helps an individual to guard against it. Pain stimulates pain receptors that are present in all organs (except brain, lung parenchyma). It is transmitted via nerves to spinal cord and then to brain where it is processed. Brain thereafter sends message down the spinal cord via specific nerves to initiate desired action. It is also important to note that pain can arise from diseases of nerves as well and such a pain is burning or like an electric shock.

Anatomy and physiology related to abdominal painAbdominal contents are divided into foregut, midgut and hindgut. Foregut represents pharynx, oesophagus, stomach, liver, gall bladder, pancreas while midgut represents duodenum, caecum, and appendix while hindgut represents colon, rectum and part of anal canal. Each section has visceral afferent nerve that transmits pain to spinal cord via autonomic sympathetic nerve. Visceral afferent nerve thus represents non-specific area. There is an overlap between visceral nerve and somatic nerve that is specific to a particular area. That is how pain from visceral organ is first felt at the distribution of somatic afferent and hence pain from inflamed appendix is first felt at periumbilical region – T10 level and then shifts to right iliac fossa at T12 level where appendix is situated.
Specific quadrant pain is related to the organ situated in that quadrant. However, some quadrants contain more than one organ. At times, pain in right hypochondrium may represent lower lobe pneumonia and may be mistaken for pain arising from liver. Similarly, as kidney lies in posterior compartment, pain manifests at renal angle on the back.

Type of abdominal pain**Visceral pain** arises from abdominal organs as a result of stretch, inflammation or ischemia. It is dull, poorly localised superficial, often referred to distant area from the site of involved organ. It is accompanied with autonomic symptoms such as nausea, vomiting, sweating and feeling sick. As against **parietal pain** represents involvement of peritoneum, it is sharp, severe and well localised to the site of involved organ. This is how as mentioned above,

appendicular pain starts in midline and gets localised to right iliac fossa with increasing severity once the peritoneum is affected. If hollow organ is involved, pain is colicky with intermittent pain-free period and if solid organ is involved, pain is dull and continuous.

Pathogenesis of pain

Most common cause of pain is **inflammation**. Inflammation is mostly a result of infection but also due to non-infectious causes such as systemic inflammatory diseases and malignancy. **Vascular pain** results from ischemia and is localised to the part supplied by affected artery. **Stretch** leads to pain as happens if liver or lymph node capsule is stretched. **Psychogenicabdominal pain** is common in older children and is caused by stress. **Referred pain** may present at the distant site from affected organ as happens in back pain arising from renal disease or shoulder pain in adults due to myocardial ischemia. **Neurogenic pain** is localised to area supplied by affected nerve and usually seen in affection of peripheral nerves, thus not applied to abdominal pain (rarely herpes zoster may present with severe superficial burning pain in a localised part innervated by a single nerve and diagnosis becomes apparent only after vesicular skin rash appears a day or two later)

Onset, duration and progress a good guide

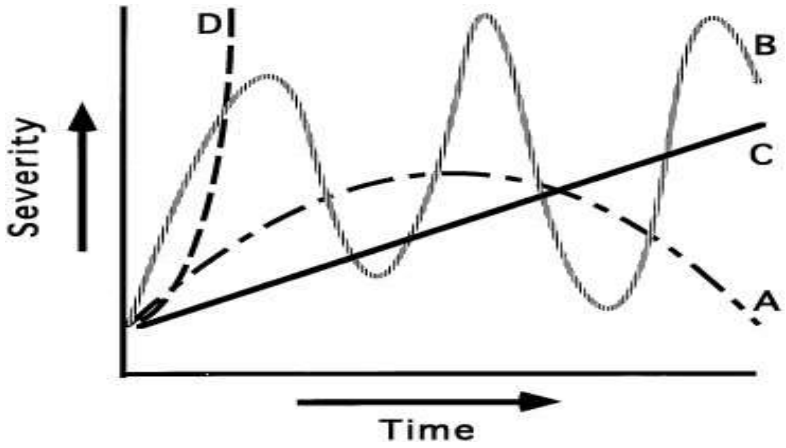
Sudden onset of severe pain is often mechanical (renal or biliary calculus, intestinal obstruction, ovarian torsion – mostly surgical) or vascular (include five types of lesions - vasculitis, vasculospasm, thrombosis, embolism and

haemorrhage – mostly medical). Onset of pain over few days preceded by fever is typically an inflammatory pain and may also have additional symptoms depending on which organ is involved. (Loose stools with blood and mucus in bacillary dysentery, nausea, vomiting followed by jaundice in viral hepatitis, severe periumbilical pain with vomiting in appendicitis).

Severity of pain suggests severe inflammation as in case of liver abscess or appendicitis while viral hepatitis often has mild pain.

Recurrent pain with intervening normal period suggests colicky pain coming from hollow tubular structures such as intestines, ureter or bile ducts.

Persistent pain worsening over few hours may indicate surgical cause while persistent pain over days or weeks may be due to gastritis or peptic ulcer (rare in children) and may be psychological, especially if it does not hinder routine activities. It is generally “pain of convenience” – it comes whenever need arises and disappears completely when child is engrossed in what he likes to do. Worsening pain or one that disturbs health needs cautious evaluation.



A – Self-limiting mild-moderate pain B – colicky pain
C – slowly worsening pain D – acute severe worsening pain

Physical examination

Inflammatory pain is accompanied with tenderness besides other localising signs such as guarding or rigidity and such a child is very sick looking. Mechanical causes may or may not present with abnormal physical findings. There are no physical findings in renal or biliary colic so also lead colic. Vascular pathology causing pain is devoid of any local tenderness and they include vasculitis, intestinal ischemia as happens in dengue capillary leak syndrome and shock, sickle cell haemolytic anaemia or Henoch-Schoenlein purpura (IgA vasculitis). In adults, diabetes may present with abdominal pain without any local physical abnormality though in children it is a rare manifestation of diabetes. Abdominal migraine and abdominal epilepsy also present without any physical findings. Generally. these two conditions are considered when other causes are excluded.

Besides local abdominal examination, findings away from abdomen should not be ignored. Degree of sickness, dehydration or electrolyte – acid-base imbalance must be

noted in every child. Affection of other systems offers clue to a disease with multiorgan involvement as seen in inflammatory bowel disease with arthritis or sickle cell anaemia with pallor and splenomegaly.

When physical examination is normal!

In presence of abnormal physical findings such as localised tenderness, guarding and rigidity or a palpable lump make diagnosis rather easy. But real challenge is when there are no abnormal physical findings. In such situations, one must look at personal history in details. Appetite, sleep, playfulness, energy and activity, behaviour besides bowel movements and urination. If all these points in the history are negative, one is nearly certain it to be a functional pain. However. any of the above mentioned symptoms are subtly upset, one needs to be cautious. Severity of pain and especially if pain awakens the child, one must find organic cause. Rare conditions such as Meckel's diverticulum, lead poisoning, porphyria, abdominal migraine and abdominal epilepsy are some of the conditions where there may be paucity of abdominal findings.

Red flags

Sudden acute severe abdominal pain with abdominal distension and vomiting is usually an emergency and there is a need to find the cause to initiate appropriate action. For example, there may be an urgent need for surgical intervention as in case of intestinal obstruction. Bile-stained vomit signifies intestinal obstruction till proved otherwise. Blood in vomit, blood passed per rectum or purpuric skin

rash needs proper assessment as it often signifies underlying serious illness. Unanticipated severe abdominal pain presenting few days after the onset of fever indicates complication as happens typically in dengue fever. When fever in such a case is on the wane, sudden appearance of severe abdominal pain suggests capillary leak syndrome with shock during which intestines are not perfused with adequate blood and so suffer ischemic pain. If neglected for a day, it may endanger life. Loss of weight in persistent abdominal pain needs evaluation. Finally, if child wakes up from deep sleep with severe pain, it definitely deserves further investigations even in absence of abnormal physical findings. Though one may come across a child who wakes up happily only to fake severe abdominal pain. Guarding, rigidity, toxic or sick look and signs of shock or sepsis are obvious serious signs.

Investigations

Tests must be ordered only after considering a provisional diagnosis. Though unfortunately tests are carried out because one has no clue to any diagnosis. But in such a case, which tests would you order?

"So called routine" tests are irrational. CBC and routine stool examination mostly do not add any value to a diagnosis. In an inflammatory pain. CBC is bound to be abnormal but one has made out inflammation even without CBC. And CBC in such a case fails to define cause of inflammation. Serum amylase or lipase are markedly increased in acute pancreatitis but moderate increase may also be seen in intestinal ischemia or strangulated hernia. Similarly, stool examination has little

value though presence of occult blood may need cautious interpretation. Stool microscopy showing worm infestation may not be related to the cause of abdominal pain. Abdominal USG is not necessary if liver or spleen in enlarged on physical examination as it adds not much greater value to diagnosis unless one suspects liver abscess or any other space occupying lesion. Same is true with CT or MRI scan. USG showing “probe tenderness on right iliac fossa” or distended loops of intestine or thickened bladder wall need correlation with clinical profile and recently it is an established trend that all tests come with a rider “correlate clinically”. Imaging report often have “?” as prefix to a probable interpretation! But clinicians are supposed to offer definite opinion and not a probable diagnosis. Thus. prerequisite for ordering tests is provisional bedside diagnosis and test results must be correlated with clinical profile.

Take home message

Abdominal pain is often a challenging symptom. Acute onset of severe abdominal pain needs cautious attempt at early diagnosis for better outcome. Similarly, one must rule out potential serious diseases. Tests must be ordered with specific aim of getting more relevant information. There are no “routine” tests. Management has to be more specific for rational outcome. Empirical treatment must have a justification and should not be polytherapy hoping one of the drugs may work.

MCQs

Q 1 Which of the following statement is wrong?
Pathogenesis of pain includes

A) Vasogenic
B) Neurogenic
C) Degenerative
D) Inflammatory

Q 2 Disease involving this system may cause abdominal pain

A) Respiratory system
B) Hematological system
C) None of the above
D) Both of the above systems

Q 3 Which of the following statement is wrong? There are no abdominal findings in this condition

A) Lead colic
B) Abdominal migraine
C) Vasculitis
D) All of the above

Q 4 This may not suggest “Red flag” in case of abdominal pain

A) Blood per rectum
B) Bile-stained vomit
C) Severe pain
D) Pain disturbing sleep

Q 5 Which of the following test would you order first when there is no clue to the cause of abdominal pain?

A) Stool microscopy
B) Urinalysis
C) None of the above
D) Both the tests

Ans to MCQs

Q1 C, Q 2 D, Q 3 D, Q 4 C, Q 5 C

2.12. Beware of abdominal pain – it is in a pandora's box

Clinical application of basic concepts of abdominal pain

Detailed history offers a probable clue to the diagnosis. Comfortable or sick looking child, distended or flat abdomen, tender or not can differentiate few conditions from others. Other signs of specific organ involvement would help to narrow down anatomy of the disease. It is not difficult to decide probable pathology as mechanical and vascular causes have sudden onset while inflammatory disorders would have onset over few days. Progress and duration add to analyse probable cause.

Case based study

Case 1

Eight months old infant presented with acute onset of vomiting that was followed within hours with recurrent

severe crying episodes. It was followed with passing blood and mucus per rectum. There was no fever. Physical examination showed mild abdominal distension and no other abnormal findings. Considering acute bacillary dysentery, he was treated with antibiotics. Acute onset of severe abdominal pain with vomiting without fever or diarrhoea suggests surgical cause and blood and mucus passed through rectum indicates vascular compromise as in **intussusception.** Over next few hours, he looked sicker with increasing abdominal distension. USG confirmed intussusception and he was operated. Fortunately, intestines did not show gangrene and so could be salvaged.

Acute onset of abdominal pain with vomiting and abdominal distension is clear indication of surgical problem Absence of fever and sudden onset rules out medical cause of inflammation such as acute bacillary dysentery. Blood and mucus passed through rectum was not accompanied with passage of stools and so it was not intestinal infection. Delay in diagnosis may result in gangrene that may endanger life and need resection of some part of the bowel.

Case 2

Eight years old child presented with fever and loose stools for 2 days, better with symptomatic treatment for few hours but fever recurred with stools with blood and mucus and abdominal pain. Physical examination showed mild abdominal distension and vague tenderness all over abdomen. CBC showed neutrophilic leucocytosis and RBCs and pus cells in stool microscopy. Considering it to be acute

bacillary dysentery, he was treated with antibiotics without benefit. Afebrile period after initial fever rules out acute infection and so it is non-infective inflammation. Diagnosis of **inflammatory bowel disease** was confirmed.
Fever in acute infection does not subside unless infection gets cured. As this child's fever abated for few days before coming on again, it is not an infection. However, fever, blood and mucus in stools and abdominal pain are all symptoms of inflammation and hence diagnosis of non-infective inflammation would be considered. Rational analysis of history is necessary for proper assessment.

Case 3

Three months old infant was diagnosed as inguinal hernia and was scheduled for surgery in next few days. Meanwhile he suddenly started crying and inguinal swelling increased. He also had mild fever. Surgeon tried to reduce hernia and he succeeded but crying would not stop. Thinking about rare situation where hernia is reduced but without relieving obstruction, urgent surgery was considered. At that time paediatrician's opinion was asked for to rule out any other condition responsible for continued crying. As this child had fever at onset of crying, acute intestinal infection was possible. Per rectal examination led to passing of stools with blood and mucus confirming diagnosis of **acute bacillary dysentery.**Any disease with fever at onset indicates most likely an infection. Obstructed hernia should not have presented with fever. Abdominal pain in this child led to crying that in turn increased scrotal swelling that was easily

reduced and so it was not an obstructed hernia. Acute bacillary dysentery may start with fever and at times followed by generalised convulsion due to toxic encephalopathy even before onset of abnormal stools with blood and mucus.

Case 4

Eight years old child presented with episodes of recurrent severe abdominal pain over last one year. Each episode started with poorly localised abdominal pain, not controlled by any drugs but getting relieved by itself. Mild abdominal discomfort remained for few more hours. Physical examination was totally normal even during the attack. Routine tests were also normal. Poorly localised abdominal pain suggested visceral affection likely to be intestinal, without peritoneal involvement and it has been recurrent but non-progressive. It has not been a colicky pain but dull ache. It suggests persistent intestinal lesion with recurrent pain. It was finally diagnosed as **Meckel's diverticulum**. It represents a localised acidic patch in some part of intestine, presenting same way as gastric ulcer does. It is suspected by microscopic blood in stools and further confirmed by radionucleide scan.It is not a common disorder but sound analysis of history paves the way to a probable diagnosis even in absence of any physical signs. Recurrent abdominal dull pain is rare and is likely to be non-infective inflammatory disorder. It may be due to immune-mediated vasculitis or chemically mediated disorder. Vasculitis is a general disease and so absence of involvement of other sites makes it less likely.

Microscopic blood in stools was a clue in this child suggesting an inflammatory disorder.

Case 5

Three years old child presented with acute onset of severe abdominal pain for last two weeks. There was moderate fever at the onset that settled within two days but pain continued though varying in severity but present most of the times and gradually getting more severe. There were no other symptoms. Physical examination showed healthy child but miserable with pain and mild abdominal distension without tenderness.

Sudden onset of severe generalised abdominal pain worsening over time suggests progressive disorder of non-infective etiology. Infection does not present with sudden onset abdominal pain and hence fever in this child would favour immune mediated disease. Such a disease could be vasculitis. It was proved to **Hennoch-Schonlein purpura**. Within next few days, this child developed purpuric skin rash and also developed arthritis and haematuria.

Sudden onset of a symptom could be either mechanical (traumatic), vascular, neurogenic, metabolic or immunological but not infective. Abdominal pain in this child is obviously not traumatic, neurological or metabolic. Fever leads to a probable immunological cause and hence diagnosis of general vasculitis, Primary vascular lesion is restricted to small area supplied by affected blood vessel as happens in myocardial infarct but this child had generalised vasculitis

affecting multiple organs – intestine, kidney and skin. It could also involve any other organs as well.

Case 6

Ten years old child developed severe abdominal pain localised to lumber region of abdomen on one side that was described by him as burning. There were no other symptoms. Physical examination did not reveal any abnormality. Abdominal pain if visceral is dull ache and poorly localised while if it is parietal, it is sharp and well localised. This did not fit in either of these two types. This was well localised but burning. It suggests neurogenic pain. Next day this child came up with vesicular skin rash over the area of abdominal wall where child was describing burning pain. Diagnosis of **herpes zoster** was evident.
Character of pain gives a clue to probable diagnosis. Colicky pain arises from tubular structures such as intestine. Ureter or bile duct. Dull ache usually refers to solid organs and burning pain characteristic of neurogenic pain.

Case 7

Eight years old child presented with history of abdominal pain, loose stools with mucus and fever off and on for last three months. He was treated with antibiotics without improvement. Physical examination showed chronically sick looking child with abdominal distension and vague generalised abdominal tenderness. Finally, endoscopy was performed and biopsy showed changes suggestive of intestinal tuberculosis. He was put on anti-TB treatment with steroids and he improved. As steroids were tapered over

next few weeks, his symptoms recurred in spite of continuing anti-TB treatment. He was reviewed again with another endoscopy and biopsy that this time revealed diagnosis of **inflammatory bowel disease.**Anatomical lesion in intestinal tuberculosis is submucosal and not mucosal and hence loose stools is not the primary manifestation of such a disease. In fact, classically, intestinal tuberculosis presents as subacute intestinal obstruction resulting in vomiting and at times overflow loose stools. But primary symptom is constipation and vomiting and not loose stools – described as alternating constipation and diarrhoea. This case also highlights limitation of laboratory results if considered without clinical correlation. Histopathology of intestinal tuberculosis is often confused with that of inflammatory bowel disease but clinical profile of two diseases is different.

Case 8

Six years old child presented with acute onset of high fever and severe abdominal pain that was followed within next eight hours with generalised convulsion. Physical examination showed highly febrile, drowsy child with abdominal distension. Next day he passed stools with mucus and blood. Diagnosis of **acute bacillary dysentery** with toxic encephalopathy was made and child recovered after treatment with antibiotics.

Generalised convulsion within few hours of onset of fever rules out meningitis that would have presented as headache, irritability and vomiting without convulsion. So, convulsion is likely to be due to either metabolic or immunological

disorder. Metabolic disorders do not start with high fever with abdominal pain and so unlikely. Combination of high fever, abdominal pain and convulsion all within few hours is rare in immunological disorder. Hence one must consider toxin mediated disorder as was in this child. Shigella infection produces toxin that may result from several complications and at times complication presents before local intestinal symptoms. Such a presentation of shigella dysentery is not uncommon.

Take home message

It is important to follow simple basic rules of history taking. Site of abdominal pain, type and severity, besides origin, duration and progress and accompanying symptoms offer a clue to diagnosis. Detailed history alone can be helpful even in the absence of abnormal physical findings. Investigations need clinical corelation, especially in case of abdominal USG that often reports non-specific findings such as mesenteric lymph nodes, dilated local intestinal loop or thickened bladder wall.

MCQs

1. Which of the following statement related to abdominal pain is wrong?

A) It may be due to primary renal disease B) It may be due to primary lung disease C) It may be due to primary haematological disease D) None of the above

2. Which of the following symptoms may not bedue to primary intestinal problem?

A) Abdominal distension B) Constipation
C) Loose stools
D) All of the above

3. Child passing blood in stools often suffers from pain except in this condition

A) Bacillary dysentery
B) Intussusception
C) Intestinal polyps
D) Inflammatory bowel disease

4. Physical examination of abdomen is normal except in this condition

A) Inflammatory bowel disease
B) Meckel's diverticulum
C) Intestinal vasculitis
D) Renal calculus

5. These abnormal test results may not correlate with cause of abdominal pain

A) Stool microscopy showing ova of roundworms
B) USG showing mesenteric lymph nodes of 1.5 cms in size
C) Plain X-ray of abdomen showing calcified spot in the liver
D) None of the above

Ans to MCQs

Q 1D, Q 2D, Q 3 C, Q 4 A, Q 5 D

2.13. My child looks pale!

Pallor – back to basics

What is pallor?

Pallor refers to paleness or whitish (mild yellowish) hue instead of pinkish hue to skin and is at times a complaint of the patient. It is not equivalent of anemia. It results most commonly due to reduced hemoglobin – anemia but it may also be caused by peripheral vasoconstriction as happens in rising fever or due to sudden stress or shock. At times, stretched skin due to severe edema also looks pale to simulate anemia and is referred to as waxy pallor. Carotenemia (due to excess of consumption of carrots) also gives a pale yellow hue to skin due to its pigment. Occasionally, severe pallor is mistaken for mild jaundice because of pale yellow hue to skin.

What is anemia?

Anemia refers to decreased red blood cell mass lower than age-appropriate standard norms that is accompanied with reduced hemoglobin. It results in decreased oxygen carrying capacity. It is important to note that hemoglobin in normal newborn is as high as 16-17 Gm%, at 3 months of age it comes down to as low as 10 Gm% and then slowly rises to adult level over next few years. So, 10 Gm% hemoglobin in newborn is anemia and at 3 months it is normal because it is physiological dip as a result of decrease erythropoiesis due to increased tissue oxygenation and reduced production of erythropoietin.

How to make sure that pallor is due to anemia

It is easy to note that paleness due to rising fever or shock is restricted only to periphery and also skin looks cold with child looking sick. Severe edema is obvious not to miss.
Besides skin looking pale, mucus membranes are also pale due to anemia. However, conjunctiva may not appear pale in spite of anemia in case of conjunctivitis and so also mucus membrane in mouth not looking pale due to glossitis. Hence it is important to look for anemia in multiple sites for correct interpretation. All other conditions simulating pallor due to anemia reveal normal mucosal color.

Is it possible to assess severity of anemia on clinical examination?

It is difficult to assess degree of anemia in dark skinned individual. However, creases on the palm may offer some clue. Normally creases are well seen in normal individuals while they are not much visible in anemic person as they also become pale. It is best to compare your own palm with that of the patient to get fair idea of anemia and also to some extent degree of anemia. Though one must note that such a clinical sign is not very sensitive and not a substitute to hemoglobin estimation.

Is it possible to judge duration of existing anemia on clinical examination?

Acute severe anemia presents with cardiac decompensation in terms of signs of cardiac failure. Chronic anemia rarely shows such signs because nature has time to compensate for poor hemoglobin and patient restricts his activities.

Physiology of anemia

It is important to know basic physiology that would help in the diagnosis of type of anemia.
Stem cell in bone marrow go through several maturation phases (progenitor cell to erythroblast to normoblast to reticulocyte to mature RBC). Hemoglobin is incorporated in late stages of normoblast and continues to be present in reticulocytes and RBCs. Life of reticulocyte in peripheral circulation is just one day while life of RBC is about 120 days. Naturally there are many RBCs in peripheral blood smear but very few reticulocytes. Routinely there are no normoblasts in peripheral circulation.
When RBCs are lysed at the end of their life, hemoglobin is released and broken down to heme and globin. Globin a protein is stored in body and heme is further broken down to iron and biliverdin, iron is also stored in the body and biliverdin forms bilirubin that needs to be conjugated by liver enzyme to make it water soluble and excreted via urine and stool.

Clinical relevance of physiology of anemia

It is obvious from physiology that increased hemolysis of RBCs will produce lots of unconjugated bilirubin that cannot be handled by liver and so result in jaundice. It is hemolytic jaundice in spite of liver being normal but can't handle excessive load of lysed RBCs. This being unconjugated bilirubin is not water soluble and so can't be excreted in urine. Thus, urine is not high colored in hemolytic anemia and physical findings include enlarged spleen and liver – a

result of increased work on the part of these organs. Increased number of reticulocytes or presence of normoblasts in peripheral blood smear suggests increased bone marrow production to compensate for increased RBCs destruction as happens in hemolytic anemia. Thus, jaundice is not always a liver disease.
On the other hand, if there are small number of RBCs along with decreased number of WBCs and platelets, it would suggest bone marrow suppression as happens in aplastic anemia. Presence of blast cells in peripheral blood smear would suggest leukemia.

Types of hemoglobin

There are three types of hemoglobin – HbA (98-99% of total hemoglobin in adults) seen in normal individuals beyond the age of 2-4 months, consisting of alpha and beta chains.,HbA2 consisting of alpha and delta chains that is also present in normal individuals beyond 2-4 months but is in small amount just 1-2% of total hemoglobin. HbF (fetal hemoglobin) found up to the age of 2-4 months of age, consisting of alpha and gamma chains and it disappears thereafter in normal individuals. Persistence of fetal hemoglobin beyond 4 months of age or increased amount of HbA2 (beyond 2% of total hemoglobin) suggest abnormality.

Nutrients necessary to form hemoglobin

Iron, vitamin B12 and folate are major nutrients required to form hemoglobin, deficiency of which leads to significant anemia. Less than 10% Iron in food is absorbed through intestines in iron-sufficient individuals as process of

absorption is hindered by presence of calcium and phytates in diet. However, nature maintains balance by increased iron absorption to an extent, in iron-deficient individuals. Heme iron (from non-vegetarian food) is better absorbed than non-heme iron. Vegetarian food lacks in Vitamin B12 unless person consumes adequate amount of dairy products. It is absorbed in ileum and not jejunum (most other nutrients are absorbed through jejunum). Other factors contribute to smaller extent and include vitamin C, few micronutrients and thyroxin, deficiency of which cause mild degree of anemia.

Types of anemia and their clinical correlates

1.Deficiency anemia is most common type of anemia. It is mainly caused by diet deficient in iron, B12 or folate. At times it may also be a result of deficient digestion and / or absorption in spite of adequate intake as happens in intestinal diseases. It may also be due to loss of blood as happens in hook work infestation or intestinal bleeding as in local intestinal pathology. Thus, mere supplementation may not suffice in case of intestinal pathological conditions. Such patients present with refractory anemia. Iron deficiency anemia presents with platenychia or koilonychia while B12 deficiency has knuckle pigmentation. There is no hepatosplenomegaly and patient does not look so sick.

2. Bone marrow disorders result either from marrow suppression as in aplastic anemia or due to infiltration of marrow as in leukemia or other similar disorders. Such patients present with purpuric spots or other bleeding manifestations and are sick looking. Infiltrative marrow

disorders present with hepatosplenomegaly while aplasia without organomegaly.

3. Hemolytic anemia is either due to congenital abnormality as in thalassemia, other hemoglobinopathies or spherocytosis while it may be acquired due to antibody mediated destruction as in autoimmune hemolytic anemia. Such patients present with mild jaundice besides severe anemia and have hepatosplenomegaly (thalassemia major is an exception as it does not present with jaundice in spite of excessive hemolysis. This is because red blood cells are destroyed in bone marrow before hemoglobin is added into them) Those with congenital disorder are not sick looking, short with abnormal facial characteristics due to extramedullary erythropoiesis necessary to compensate excessive hemolysis.

Following table summarises above findings

	Liver	Spleen	Jaundice	Sick	others
Iron deficiency	-	-	-	no	nail signs
B12 deficiency	-	-	-	no	knuckle sign
Co ngnital hemolytic	++	++	+	no	facial signs
Aquired hemolytic	++	++	+	mild	-
Aplastic	-	-	-	sick	-
Leukemia	++	++	-	sick	purpura

Blood basic investigations

	WBC	Platelets	MCV	RDW	Others

Iron deficiency	N	N	low	high	micro-hypo
B12 deficiency	low	low	high	high	macro
Hemolytic	N	N	variable	N	micro-hypo
Bone marrow	low	low	variable	variable	blasts

Treatment

Deficiency anemia needs relevant supplementation. Oral iron therapy works well as iron absorption is enhanced in deficient state. Parenteral therapy is not indicated and packed cell blood transfusion is not necessary unless child presents in cardiac failure that is rare. B12 deficiency often needs parenteral therapy as absorption depends on intrinsic gastric factor. However oral treatment may be tried first. Folate deficiency is treated with oral supplements. Diet modification is important to prevent recurrence.
Other types of anemia need specialized care and should be left to specialists.

Take home message

Pallor is not equivalent of anemia. Subjective assessment in mild or moderate anemia is not dependable, especially in dark skinned individuals. Creases on palms offer clue to severity of anemia to some extent. Simple algorithm depicted above helps in clinical diagnosis of type of anemia. CBC and peripheral blood smear almost confirms type of anemia and may need further specalised tests to fine-tune diagnosis. Oral supplementation in deficiency anemia is adequate and in case of refractory deficiency anemia, one must rule out

intestinal disorders. Specialised care is necessary for other types of anemia.

MCQs

1. Anemia with hepatosplenomegaly is feature of

A) Iron deficiency anemia
B) Thalassemia
C) Aplastic anemia
D) Vitamin B12 deficiency anemia

2. Anemia with jaundice is feature of

A) Leukemia
B) Thalassemia
C) Autoimmune hemolytic anemia
D) Iron deficiency anemia

3. Which of the following cells ARENOT seen in blood peripheral smear in normal individual?

A) Reticulocyte
B) Platelet
C) Normoblast
D) Monocyte

4. Which of the following condition shows microcytosis (small size RBCs) besides iron deficiency anemia?

A) Vitamin B12 deficiency
B) Aplastic anemia
C) Thalassemia
D) Leukemia

5. RDW (red cell width) is increased in this conditions

A) Aplastic anemia
B) Leukemia
C) Vitamin B12 deficiency
D) Hemolytic anemia

Answers to MCQs

Q1 B, Q2 C, Q3 C, Q4 C, Q5 C

2.14. Bedside diagnosis of anemia possible!

Clinical application of basic concepts of pallor

Pallor is not equivalent to anemia, though in routine practice, pallor is mostly due to anemia. Acute severe anemia presents as shock as happens in case of large amount of blood loss while chronic severe anemia may present as congestive cardiac failure. Slowly progressive anemia may be well compensated without cardiac dysfunction.
Three major groups of diseases present with anemia – deficiency, hemolysis and bone marrow dysfunction. It is possible to differentiate these groups by history and physical examination. CBC and peripheral blood smear are excellent screening tests that nearly confirm group diagnosis. Further specialized tests may be necessary to define subgroup final diagnosis. This approach helps to minimize laboratory tests instead of ordering battery of tests.

Case based study

Case 1

One year old child presented with pallor noticed by mother over last few weeks. There were no other significant symptoms. He was exclusively breast-fed for first 6 months and thereafter mother introduced diluted cow milk while continuing breast feeds and occasional cereal.

As there are no symptoms of cardiac decompensation, it appears to be chronic severe anemia. Absence of jaundice mostly rules out hemolytic anemia (though thalassemia major has no jaundice). In absence of purpura (indicating low platelets) and any significant sickness, bone marrow disease is not possible. This leaves us with deficiency anemia. This child consumes milk only that is poor in iron and so this is mostly iron deficiency anemia.

Physical examination showed not sick looking, pallor ++, koilonychias +,wt 8.5 kg, length 74 cms (both normal meaning that this is getting enough calories and proteins), temp normal, HR 120 RR 28 (mild proportionate increase in heart and respiratory rate in absence of fever suggesting mild strain on heart), liver 3F +, firm, not tender, liver span 8 cms, spleen not palpable, signs of rickets +

This child has signs of iron deficiency anemia as suggested by koilonychias and also had rickets. Milk is poor source of both iron and vitamin D and hence it is compatible with our thinking. However deficiency anemia does not present with hepatomegaly. Considering mild increase in HR and RR, it denotes congested liver though without congestive cardiac failure as yet. This child would present with cardiac failure if not treated.

Thus, clinical diagnosis is **iron deficiency anemia on the brink**

of cardiac failure. Blood smear showed microcytic hypochromic anemia with low MCV and high RDW, characteristic of iron deficiency anemia. There is no need to further confirm with serum iron studies. Oral iron supplement with ferrous sulphate is ideal (choice depends on tolerance in individual child) and will show rise of one gram of Hb in one week.

Iron deficiency anemia is most common in the community, especially in children and women. Iron and vitamin D deficiency go together in a child on predominant milk diet. It is due to poor intake of iron but also due to poor absorption of iron from food due to calcium and phytates in the diet. At times, persistent intestinal microscopic bleeding would cause iron deficiency, which is resistant to treatment because of continued loss of blood.

Case 2

Two years old child presented with severe pallor and no other symptoms. Six months ago, he was seen for severe pallor for which blood transfusion was given without arriving at any definite diagnosis. He was exclusively breast-fed for first 5 months and then mother started complementary feeds gradually increasing to family vegetarian diet over next 6 months. After the age of one year, he was on family food with small amount of cow milk. He has grown well over last two years. No family history of similar disease. No history of consanguinity.

This child has recurrent anemia in spite of one blood transfusion 6 months ago for similar complaints.

Analysing present episode, it suggests deficiency anemia

because of absence of any other symptoms suggestive of either hemolytic anemia or bone marrow involvement. He is consuming very little milk after first 6 months of life though he is on full family diet. Lack of dairy products in a vegetarian diet runs a risk of vitamin B12 deficiency. So, this may be B12 deficiency anemia. However odd point is recurrence of anemia within 6 months. This is explained on the basis of absence of B12 supplementation during last episode wherein blood transfusion improved his hemoglobin temporarily only to go down again.

Physical examination showed comfortable child not sick looking, wt 11 kg, length 87 cms, pallor ++, pigmented knuckles, mild icterus, no abnormal facies, liver and spleen not enlarged and other systems normal.

Severe anemia without hepatosplenomegaly in a comfortable child with pigmented knuckles suggests B12 deficiency anemia. However odd point is presence of icterus that is normally seen in hemolytic anemia. Absence of enlarged liver and spleen and abnormal facies (due to extra-medullary hemopoiesis) ruled out hemolytic anemia. 5% of B12 deficiency anemia patients may have mild jaundice. So, diagnosis stays to be **vitamin B12 deficiency anemia.** This is suggested by macrocytic anemia with increased MCV and RDW and further confirmed by serum Vitamin B12 level.

Ideal treatment consists of parenteral Vitamin B12 as oral absorption may be erratic due to insufficient intrinsic factor in stomach. As this child was not treated with Vitamin B12 last time, anemia recurred.

Vitamin B12 deficiency is typically seen after the age of one

year as milk intake goes down. It is often missed as iron deficiency is far more common. At times, intestinal malabsorption or diseases of ileum in particular lead to vitamin B12 deficiency. (Most nutrients are absorbed through jejunum but vitamin B12 is absorbed in ileum.

Case 3

Two months old infant presented with pallor noticed since last one month and focal seizure involving left upper limb one hour prior to hospitalization. There was no jaundice or purpura or bleeding. He was born after full term with forceps delivery. Baby cried immediately and was on breast feeds throughout.

Anemia in two months old infant is not a deficiency anemia as fetus derives all the nutrients from mother that last for few months after birth and continued exclusive breast feeds will nourish the infant ideally. This was a full term born infant and not preterm who may present with anemia so early in life due to short period of nutrient transfer from mother to fetus. It is also unlikely a hemolytic anemia as it would present with jaundice. It may be therefore bone marrow problem, however there is no clue in absence of purpura or bleeding (due to thrombocytopenia). With this discussion, one is not sure about type of anemia on history alone.

Focal seizure suggests space occupying lesion in motor cortex. Brain tumors are rare at this age and so correlating with anemia, this may be a hematoma – could be attributed to trauma caused by forceps delivery. Such a hematoma may account for loss of blood and hence anemia. Typical subdural hematoma may accumulate blood over time to lead to

anemia as well as focal seizure.
Physical examination showed conscious infant, weight 3.5 kg (birth weight 2.5 kg), length 55 cms, head O 40 cms, pallor ++, no icterus, liver 2F +, soft, spleen not palpable, mild weakness of left upper limb, anterior fontanel mildly boggy suggesting mile increased intracranial tension.
Anemia without hepatosplenomegaly or purpura and comfortable infant suggested deficiency anemia a result of slow progressive subdural bleed. Hence diagnosis of **subdural hematoma** with significant anemia is certain.
It can be proved by USG of skull (CT scan may not be necessary) and deficiency anemia by CBC and peripheral smear. It is worth noting that deficiency anemia may result from slow bleeding anywhere in the body, at times bleeding is hidden – commonly in intestines as in hook work infestation or any other bleeder such as polyposis.
Treatment may need evacuation of hematoma and antiepileptic drug such as phenobarbitone for about 6 months and iron supplements.

Case 4

Two months old infant presented with pallor noticed since last few days and irritable since then, reluctant to feed. Born after full term normal delivery and has been on exclusive breast feeds.This infant has been sick as evident by loss of appetite and irritability and has presented with recent onset of anemia. It is obviously not a deficient anemia at this age and also unlikely to be congenital hemolytic anemia as infant is sick. So most likely this is bone marrow disease – either aplastic anemia or any other infiltrative disorder such as

leukemia. Physical examination showed sick looking infant, weight 4.3 kg, length 53 cms, head O 39 cms, pallor++, liver 2 F+, soft, spleen just palpable, deformity of left forearm This anemia is without hepatosplenomegaly (liver 2 f+ and spleen just palpable at this age is within normal limits) in a sick irritable infant with congenital malformed limb suggests congenital aplastic anemia. Presence of any congenital malformation seen on physical examination demands search for any other malformations that may as well be hidden. So clinical diagnosis is **congenital aplastic anemia.**
CBC will show pancytopenia and diagnosis can be confirmed by bone marrow aspiration. There is no specific drug treatment and marrow transplant is indicated.

Case 5

Six years old child presented with lump in left upper quadrant of abdomen noticed and feeling fatigue since last few months. Past history of one episode of severe abdominal pain lasting for a day and settled down by itself without definite diagnosis. There is history of consanguinity.
Lump in left upper quadrant of abdomen suggests enlarged spleen and feeling fatigue over few months indicates chronic anemia. Chronic anemia with splenomegaly is either due to hemolytic anemia or bone marrow disease such as leukemia or storage disorder. One episode of severe abdominal pain that lasted for a day does not suggest a colic. Abdominal pain may be inflammatory, vasogenic, neurogenic, referred or psychogenic. Inflammatory pain is not possible as there are no symptoms of inflammation and neurogenic pain is ruled

out as it is localized to direction of a nerve. So, this may be vasogenic pain. Bone marrow disease would not cause vasogenic pain while hemolytic anemia that may cause vasogenic pain is sickle cell disease. Thus, diagnosis of sickle cell anemia is most likely. History of consanguinity favors such a diagnosis.

Physical examination showed weight 18 kg, height 102 cms, pallor +, liver 3F+, not tender, spleen 4F+, firm, no icterus, no ascites, rest of the systems are normal. It supports diagnosis of hemolytic anemia in view of hepatosplenomegaly with anemia but without jaundice. (Jaundice in hemolytic anemia is classically seen in congenital spherocytosis and acquired autoimmune hemolytic anemia. Jaundice is absent in thalassemia. However sickle cell disease may affect liver and cause jaundice that is not hemolytic but due to affection of liver). Thus diagnosis in this child is **sickle cell anemia.**

Diagnosis can be confirmed by hemoglobin electrophoresis that would show hemoglobin S. It is called sickle cell anemia because RBCs are sickle shaped instead of spherical that impedes smooth flow of blood in capillaries.

Treatment is symptomatic.

Sickle cell anemia rarely presents with severe anemia but manifests with vascular obstruction (occlusion of blood vessels) in the form of dactylitis, hemiplegia due to middle cerebral artery occlusion or abdominal or chest pain. Such events get aggravated by dehydration.

Case 6

Four years old child presented with fever and irritability for one week and pallor noticed since last two days. There are no

other complaints. Child was well prior to onset of symptoms. Fever suggests either infection or inflammation. Viral infection presents with cold, cough and most often self-limiting.**,** so it is unlikely. Bacterial infection usually has localizing symptoms which this child does not have. So, it may be non-infective illness. Pallor indicates hematological illness and irritability may suggest pain that has not been localized and hence likely to be generalized. Generalised pain may be either muscle or bony pain. Pallor with bony pain may indicate possibility of leukemia.

Physical examination showed sick looking child, irritable, pallor++, liver 4F +, soft, not tender, spleen 2F +, large cervical lymphnodes on both sides, firm, not tender.

In view of hepatosplenomegaly and lymphadenopathy with pallor, **acute lymphoblastic leukemia** is most likely. Diagnosis is confirmed by peripheral blood smear showing blast cells along with low hemoglobin and platelet count with lymphocytosis and if necessary, by bone marrow examination. Further studies are necessary to define more details for which specialized tests are necessary. These specialized studies can tailor-make chemotherapy and also help in prognostication.

Early diagnosis of leukemia is important. However, steroid therapy and / or blood transfusion given for severe anemia without proper diagnosis can not only result in delayed diagnosis but also be the cause of poor outcome in case of leukemia. In fact, steroid therapy without proper diagnosis and justification is always fraught with danger.

Case 7

Two years old child presented with gradually increasing pallor over last 4 months and deviation of angle of mouth noticed over last two days. He had been otherwise well.Pallor in this child may be due to deficiency anemia, hemolytic anemia (without jaundice) or chronic bone marrow disease due to storage. Deviation of angle of mouth suggests facial nerve palsy. In absence of any other neurological symptoms, facial nerve must be involved at its exit from the skull or beyond. If it is at the exit, it must be due to compression of enlarged bone while if it is due to lesion beyond exit, it may be unrelated to anemia. General rule is to ascribe all symptoms to a single disease and hence it may be safe to assume that facial nerve is caught at its exist from skull due to enlarged bone. Bone may be enlarged due to abnormal storage in the bone as happens in osteopetrosis. Physical examination showed comfortable child not sick looking but stunted, pallor ++, liver 4F+, not tender, spleen 2F +, lower motor neuron facial palsy. Anemia with hepatosplenomegaly in absence of icterus and abnormal facies rules out hemolytic anemia and favors bone marrow storage disease, such as**osteopetrosis** in which bone is thickened that leads to compression of cranial nerves leaving skull bone.

Diagnosis can be confirmed by x-rays showing dense bony structure without differentiation between cortex and marrow cavity. There is no specific treatment and transplant is the only possibility.

This case illustrates a type of problem that involves not only anemia but also involved neurological system that would make a search for unusual cause of anemia. Similar problem

of anemia with neurological affection has been discussed in one of the earlier cases - subdural hematoma presenting as anemia and a focal seizure.

Case 8

Eight years old child presented with gradually progressive abdominal distension, loss of appetite and weight over last 6 months and pallor noticed over last two months.
Progressive abdominal distension over long period suggests enlarged liver with or without enlarged spleen. Any other space occupying lesion is also possible such as tumor or cyst. Loss of appetite and weight denotes catabolic state often representing generalized disease. Absence of fever rules out infective or inflammatory disease. Pallor indicates anemia that has developed over last two months and so suggests complication of generalized disease now affecting bone marrow. Thus, this is most likely to be storage disorder due to abnormal metabolism. Exact nature of storage is not possible to define on history or physical examination.
Physical examination showed weight 18 kg, height 105 cms, pallor ++, no icterus, liver 5F +, firm, not tender, spleen 5F +. Firm. This child is undernourished and stunted suggesting chronic catabolic disease involving liver and spleen and lately affecting bone marrow as well. It favors **storage disorder.**
Further specialised tests can prove exact metabolic defect. One of the common storage disorder at this age is Gaucher's disease result of specific enzyme deficiency. It can be treated with enzyme replacement.
Storage disorder due to accumulation of abnormal metabolite results in organomegaly without functional

disturbance in the affecting organ till late in the course of disease. It is only when bone marrow is involved that hematological manifestations point out to the diagnosis.

Take home message

It is important to make sure that pallor is due to anemia though it is almost always true. Few pointers in the history such sickness, short stature, malnutrition, abnormal facies, abdominal distension, jaundice, purpura and affection of other systems in the body can easily suggest group diagnosis of anemia – deficiency, hemolytic or bone marrow disease. Physical examination can nearly support diagnosis inferred by the analysis of history and further confirmed with specific tests instead of battery of tests.

MCQs

1. Jaundice is not seen in this hemolytic anemia

A) Autoimmune hemolytic anemia
B) Thalassemia
C) Congenital spherocytosis
D) G6PD deficiency

2. Jaundice is rarely seen in

A) Iron deficiency anemia
B) Vitamin B12 deficiency anemia
C) Aplastic anemia
D) Leukemia

3. Hepatosplenomegaly is not a feature of

A) Congenital spherocytosis
B) Thalassemia
C) Vitamin B12 deficiency anemia
D) Leukemia

4. Pancytopenia is seen in

A) Aplastic anemia
B) Leukemia
C) Vitamin B12 deficiency anemia
D) All of them

5. X-ray of bones nearly confirms diagnosis of this condition

A) Thalassemia
B) Leukemia
C) Osteopetrosis
D) Iron deficiency anemia

Answers to MCQs

Q1 B, Q2 B, Q3 C, Q4 D, Q 5 C

2 15. Every yellowis not jaundice

Back to basics – jaundice

What is jaundice?

It is yellow discoloration of sclera, skin and mucus membranes. It is visible in sclera as icterus, only when serum bilirubin exceeds 2 mg%. (Icterus is a bird with yellow beak) Normal serum bilirubin level varies between 0.2 to 1 mg and

hence jaundice is not visible early in the course of rising bilirubin till it crosses 2 mg%.

Clinical correlate of jaundice

Patients report yellow colored urine before icterus is visible in eyes. However, urine is high colored in many other conditions such as concentrated urine due to dehydration, hematuria and drug induced urine color. Though shade of high colored urine is different in all these conditions, urine is dark yellow in jaundice. Yellow color of urine is due to excretion of water-soluble conjugated bilirubin while unconjugated bilirubin being not water soluble, cannot be excreted in urine and so, urine is not yellow in hemolytic jaundice even when sclera is yellow. Thus, urine color differentiates hemolytic jaundice from hepatobiliary jaundice. Mild icterus is not easy to make out as sclera may appear muddy due to environmental exposure in non-jaundiced individuals or also in case of vitamin A deficiency and is often confused with icterus. Skin and mucus membranes appear yellow only when jaundice becomes severe. In fact, common cause of yellow skin is carotenemia (due to consumption of carrots in normal individuals) wherein eyes are not yellow and so it cannot be confused with jaundice. Mild yellowish hue is also seen in anemia.

Physiology of jaundice

Natural breakdown of RBCs at the end of their life span of about 120 days, hemoglobin is released. Hemoglobin consists of heme and globin, heme is iron and biliverdin. Globin a protein and iron are stored in the body while biliverdin is

converted to bilirubin that is bound to albumen and transported to liver where it is conjugated to water soluble form. It is excreted in duodenum via bile and then excreted in stools and urine.
On and average, 1% of RBCs break down each day and amount of bilirubin formed can be easily handled by metabolic processes in the liver. In fact, liver has lots of functional reserve but to a limit. So, when this limit is exceeded, jaundice develops due to accumulated bilirubin.

Types of jaundice

1. Hemolytic jaundice

Excessive breakdown of RBCs results in large amount of bilirubin that cannot be handled by normal liver. Hence unconjugated bilirubin accumulates in the body. Unconjugated bilirubin is not water soluble and so cannot be excreted. Thus, urine is not high colored in hemolytic jaundice. Unconjugated bilirubin may accumulate in brain leading to dysfunction (bilirubin encephalopathy due to immature blood-brain barrier) especially in a newborn baby or if large amount is produced as in case of rare disorder – Crigler-Najjar syndrome. It typically happens in severe hemolysis due to Rh blood group incompatibility in a neonate. Besides blood group incompatibility in a neonate, increased hemolysis occurs due to congenital defects as in case of Thalassemia, sickle cell disease, spherocytosis and G6PD deficiency (jaundice occurs with exposure of some of the drugs) and also in acquired autoimmune hemolysis.

2. Hepatocellular jaundice due to liver cell damageIt is a result of hepatic cell damage. Liver has lots of reserve and only 15% of liver cells are enough to handle normal amount of bilirubin. Thus, when more than 85% of liver cells are involved, even small amount of unconjugated bilirubin produced by normal RBCs breakdown cannot be handled resulting in jaundice. Depending upon degree of conjugation defect, part of bilirubin that gets conjugated is excreted in urine and so urine is high colored. As liver cells are damaged, other liver functions (production and metabolism) are also affected.

3. Hepatocellular jaundice due to isolated functional defect

Deficient enzyme responsible for conjugation fails to convert normal amount of unconjugated bilirubin that accumulates in the body (simulating hemolytic jaundice) and as it is not water soluble, urine is not high colored. (Crigler-Najjar or Gilbert syndrome)

Deficient enzyme responsible for bilirubin excretion results in accumulation of conjugated bilirubin that colors urine yellow. (Dubin-Johnson and Rotor syndrome)

4. Biliary jaundice

Normal amount of bilirubin produced is conjugated by liver but due to obstruction to excretion into the intestines, conjugated bilirubin accumulates in the body. However as conjugated bilirubin is water-soluble, it is excreted in urine and so urine is high colored. As bilirubin cannot enter intestines, stool is clay colored. (Normal stool is brownish or yellowish in color due to presence of stercobilin – excretory

form of conjugated bilirubin in intestines). Biliary obstruction may be due to congenital defect such as biliary atresia or acquired conditions such as biliary calculus, cholangitis or due to external compression of biliary tract as in malignancy.

5. Hepatobiliary or biliaryhepatic jaundice

Liver cell – hepatocyte disease if not controlled in time spreads to biliary tract (compression of biliary radicals due to oedema of inflamed hepatocytes) and often patients present as hepatobiliary disease – meaning both the components of liver – hepatocyte and biliary tract are involved. Similar situation arises when primary biliary tract disease spreads to hepatocytes (as happens in intrahepatic biliary obstruction) thus involving both the structure. It is important to assess primary disease in such hepatobiliary disorders. Thus, it is clear that timely intervention of hepatocyte or biliary tract disease may prevent spread of disease to other parts of the liver and complicate the problem.

Clinical approach to jaundice

Confirm jaundice

Mild degree of jaundice may not be easily discernable unless one examines eyes against natural sunlight. At times, muddy sclera may be mistaken for jaundice. Mild jaundice does not give yellow hue to skin or mucus membranes. Yellowish skin without yellow eyes suggest carotenemia and not jaundice. High colored urine may also be due to other causes. Urine is not high colored in hemolytic jaundice. Similarly, clay-colored stools may be due to fat malabsorption or giardiasis besides obstructive jaundice. Once clinical jaundice is confirmed, high

colored urine suggests either hepatocyte or biliary tract disease, Clay colored stools and itching favor biliary obstruction though may also be seen in severe hepatocyte disease. Normal urine color indicates hemolytic jaundice that is also characterized by pallor. However, pallor and yellowish tinge may look similar and may be mistaken for one another. In hemolytic jaundice, pallor is more prominent than jaundice which is mild. Hepatocyte or biliary tract disease may also look pale due to co-morbid conditions but jaundice is presenting finding. In a short duration of jaundice, (hepatitis A or E infection) sickness disproportionate to degree of jaundice favors hepatocyte disease while high degree of jaundice with reasonably maintained health status indicates biliary disease. In a long duration jaundice as in case of hepatitis B or C infection, metabolic liver diseases (Wilson's disease) and cirrhosis, nutrition and growth are affected. Hepatomegaly with or without splenomegaly are seen in almost all of these cases of jaundice. In chronic hepatocyte disease such as cirrhosis, there may not be jaundice until late stages, at which time ascites is also often present. However, absence of hepatosplenomegaly in presence of jaundice suggests specific enzyme deficiency and such patients are otherwise healthy without any liver dysfunction or pallor.

Complications

Portal hypertension is seen in chronic liver diseases and manifests as hepatosplenomegaly with ascites. Liver cell failure is characterized by encephalopathy and bleeding.

Investigations

Serum bilirubin – direct (conjugated) and indirect (unconjugated) componentsDirect bilirubin more than 2 mg% is considered as conjugated bilirubinemia irrespective of total and indirect bilirubin levels. When total bilirubin is less than 5 mg%, direct component more than 20% of total is also considered as conjugated bilirubinemia. For example, when total bilirubin is 2 mg% and direct component is 0.5 mg%, it denotes direct bilirubinemia even when indirect bilirubin is higher than direct bilirubin.

SGPT (ALT) and SGOT (AST)These are intracellular enzymes and so liver cell destruction leads to increase in enzymes level in blood. SGPT is more specific to liver pathology as SGOT also increases in other diseases such as heart or skeletal muscle affection. SGPT is higher than SGOT in primary liver disease while SGOT is more than SGPT in systemic diseases with liver involvement. Hence it is ideal to order both enzymes in assessment of liver disease. SGPT is very high in acute destruction of liver cells as seen in acute viral A hepatitis, in which SGPT may be in thousands. Per se, it does not suggest any serious illness but indicates acute disease.

Serum proteins – albumen and globulin

Serum albumen level goes down in chronic liver cell disease but not in acute liver cell disease as half-life of albumen is three weeks and takes time to go down. Serum globulins are often increased in immune mediated disorders.

Alkaline phosphatase Serum levels of alkaline phosphatase increase in biliary obstruction. However it is not specific to biliary disease as it is also increased in muscle diseases.

GammaGT – gamma glutamyl transferase
High level of this enzyme is more specific in biliary obstruction as it is not increased in muscle disease.
Prothrombin time
As hepatocyte produces coagulation factors, prothrombin time is increased in severe liver disease and hence it is a measure of severity of disease. It indicates liver cell failure.
Viral markers
In suspected viral hepatitis (A,B.C,E), viral markers may help in confirming diagnosis. Hepatitis B virus has surface, core and e antigen and host responds with antibodies to surface and e antigen. Core antigen is present only in liver cells and so no antibodies are seen in blood. Other viruses may also affect liver such as CMV or HIV.
Urinalysis
Presence of bile salts and pigments confirms diagnosis of conjugated bilirubinemia.
CBC is not much useful except in cases where one suspects acute bacterial infection as in acute cholangitis.
Imaging
USG helps in delineating patency of bile ducts and presence of bile in gall bladder. It also may suggest echo-structure of liver that may be corroborative.
Histopathology of liver
Liver biopsy is reserved for diagnosis of cirrhosis and chronic hepatitis.

Management

Most of liver diseases are treated with symptomatic therapy and have no specific therapy except hepatitis B and C can be treated with drugs but should be best managed by specialists. Autoimmune hepatitis is treated with steroids. Biliary tract disorders may be surgically treatable. Liver transplant is now possible for many diseases.

Take home message

Jaunice may be due to conjugated bilirubinemia (high colored urine) or unconjugated bilirubinemia (normal urine color). Jaundice is mild in proportion to sickness in hepatocyte disease while jaundice is severe in an apparently normal child in an extra-hepatic biliary obstruction. Absence of hepato-splenomegaly with jaundice suggests enzyme disorders. Laboratory tests in primary liver diseases offer specific interpretation – serum bilirubin denotes extent of the disease, ALT (SGPT) the acuity, serum albumen the chronicity and prothrombin time the seriousness, GGT the biliary tract obstruction.

MCQs

1. Icterus is visible only when serum bilirubin level increases beyond

A) 0.5 mg%
B) 1 mg%
C) 1.5 mg%
D) 2 mg%

2. Which of the following levels suggest direct bilirubinemia?

A) Total bilirubin 3 mg% - direct 0.5 mg%
B) Total bilirubin 1.6 mg% - direct 0.8 mg%
C) Total bilirubin 1 mg% - direct 0.2 mg%D) Total bilirubin 10 mg% - direct 1.5 mg%

3. Liver is not enlarged in this jaundice

A) Hepatitis
B) Cirrhosis
C) Enzyme disease – Gilbert disease
D) Hemolytic jaundice

4. Which of the following statement is wrong?

A) High colored urine in hepatitis
B) Urine may not be high colored in cirrhosis
C) High colored urine in autoimmune hemolytic anemia
D) High colored urine in biliary obstruction

5. Very high levels of SGPT are common in

A) Severe liver disease
B) Chronic liver disease
C) Acute liver disease
D) Liver cell failure

Answers to MCQs

Q1 D, Q2 B, Q3 C, Q4 C, Q5 C

2.16. Jaundice without liver disease, liver disease without jaundice

Clinical application of basic concepts

Icterus in sclera is evident in most cases except in early phase of the disease. High colored urine is easily noticeable though seen only in conjugated bilirubinemia. Once jaundice is suspected, color of urine and stool offer clue to type of jaundice. High colored urine is characteristic of hepatocyte disease while clay-colored stools of biliary obstruction. However, severe hepatocyte disease also may have clay-colored stools. Sick look in spite of mild jaundice favors hepatocyte disease while deep jaundice in apparently healthy individual suggests biliary obstruction. Normal urine color indicates indirect bilirubinemia and if due to hemolysis, pallor is the main symptom with milder jaundice. Thus, history alone can diagnose probable group of disorder. Physical examination nearly can confirm diagnosis. General appearance – sick or not sick, enlarged liver and / or spleen, other signs of liver disease such as edema or ascites, significant pallor offer clues to diagnosis. Thus, history and physical examination can help order specific tests to confirm diagnosis and avoid battery of test that may not be necessary.

Case based discussion

Case 1

8 years old child presented with moderate degree of fever for two days followed by high fever for next 3 days without any other symptoms. On D5, mother noticed high colored urine and mild icterus.

Onset with moderate fever followed by high degree of fever suggests bacterimic bacterial infection that has not localized

so far until D5. Non-localising bacterimic infection typically is typhoid fever. Now that child develops jaundice, it is mostly a complication of typhoid fever. Viral A hepatitis classically starts with prodrome of nausea, vomiting and anorexia followed in a day or two with jaundice and so it is not viral hepatitis. Had it been irregular pattern of fever, one may have considered malarial hepatitis.
Physical examination showed sick looking child, febrile, mild icterus, tumid abdomen, liver 3F+, span of 9 cms, spleen just palpable.
CBC showed Hb 11 Gm%, WBC 3000 P 30 L 60 M10 E0, pl 80000, total bilirubin 4.3 mg% Direct 3.4 mg% SGPT 170 SGOT 250 Alk phos normal
CBC is typical of typhoid fever – leucopenia, lymphocytosis, monocytosis, thrombocytopenia. SGOT > SGPT indicates systemic extra-hepatic infection with secondary liver involvement and not primary liver disease.
Blood culture grew S.Typhi
Typhoid fever with hepatitis was confirmed.
Jaundice appearing few days after onset of fever indicates extra-hepatic infection with subsequent involvement of the liver. It could be a result of direct extension of primary infection (typhoid or malaria) or immune complication of primary infection (leptospirosis or different viral infection including Covid– part of multisystem inflammatory disease). Jaundice presenting early in the course of a disease is usually due to primary liver disease.

Case 2

6 years old child's mother accidentally noticed yellow tinge in the eyes but child was quite normal without any complaints. On direct questioning, urine color was normal.
Normal urine color rules out hepatocyte and biliary disease. It may be hemolytic jaundice but pallor was not noticed.
Physical examination showed mild icterus but no pallor or hepatosplenomegaly. Growth and nutrition were normal.
It rules out hemolytic jaundice. This is a normal child with mild unconjugated (indirect) bilirubinemia. It suggests enzyme deficiency – **Gilbert disease.**
Diagnosis may be confirmed by liver biopsy but it is not indicated as clinical diagnosis strongly suggests a benign disease and so invasive investigation may be avoided.
This is a specific enzyme deficiency but all other liver functions are normal. It is a benign condition and child will lead normal life without any problem but with jaundice.
This case illustrates an example of jaundice without any evidence of liver or hemolytic disease in a healthy asymptomatic child. Such a presentation is typical of an enzyme defect that is benign though persistent and has no treatment. However Crigler-Najjar syndrome is another enzyme defect causing indirect bilirubinemia but is severe and has a risk of brain damage (typically basal ganglia) .

Case 3

8 years old child was accidentally noticed to have yellow eyes while he was getting ready to go to school. As he was fine, he insisted to go to school. On his return, his doctor diagnosed it as viral A hepatitis and suggested no specific treatment.

Viral A hepatitis presents with a prodrome of nausea, vomiting and severe anorexia before developing jaundice and child does feel weak. So, this is most unlikely to be viral A hepatitis. At this point we need to follow the child closely for further progression. Next morning when he got up to brush teeth, he suddenly fainted and mother noticed severe jaundice. He was rushed to the hospital.

It is evident that this child has developed fulminant liver disease that has worsened just over a day. Sudden fainting may suggest severe anemia or syncope. So, this child seems to be suffering from acutely worsening liver disease with or without anemia.

Physical examination showed a sick child, deep icterus, severely pale, liver 5F+ firm, spleen not palpable, no ascites. This child has combination of acutely worsening jaundice with severe anemia. Acute onset of severe anemia may be due to hemolysis or occult hemorrhage. Sudden onset of any symptoms may be either immune mediated or metabolic. Autoimmune hemolytic anemia and autoimmune hepatitis don't go together and child in either of these two conditions is not very sick. So, this may be metabolic disorder. Most common metabolic liver disorder > 5 years of age **is Wilson's disease.** It was confirmed by low serum ceruloplasmin. Wilson's disease is a genetic disorder and presents with a wide spectrum of symptoms of varied duration. It is treated with chelating agents such as D-penicillamine and zinc. This child had developed acute fulminant liver disease and so had no time for drugs to act. He underwent acute liver transplant and survived.

Case 4

8 years old child presented with high fever, severe body ache and headache for last 4 days. Fever would respond poorly to paracetamol and he would look sick throughout the illness. Body ache was so severe that he could not walk because of pain. On D6, he developed high colored urine and jaundice. Analysing fever before onset of jaundice, these symptoms are common to viral infections, malaria and also typhoid or any other severe bacterial infection. Severe body ache – myalgia – to an extent of inability to walk is unusual feature in this child. Typical viral infection would often have cold, cough etc and usually settle within 3-4 days. Malaria presents generally with erratic fever pattern and typhoid typically starts with moderate degree of fever that rises over next few days. Hence it may be evolving bacterial infection that has not yet produced any localizing symptom. Such infections include bacterial endocarditis, leptospirosis, rickettsia or brucellosis. Development of jaundice suggests hepatic complication of extra-hepatic disease and leptospirosis is one of such diseases presenting with liver and kidney affection. Physical examination showed sick looking child, highly febrile, congested eyes, oral mucosa and throat, icterus +, liver 3F+, soft, not tender, spleen not palpable, other systems normal. Jaundice developing few days after high fever with severe myalgia and headache favors diagnosis of leptospirosis. One may have to look for involvement of other organs, if not clinically visible, by relevant laboratory tests.
Investigations showed Hb 9 Gm%, WBC 18000 P 72 L 25 M 3 E 0 Pl 2.5 Serum bilirubin total 4 mg% Direct 3.2 mg%, SGPT

250 SGOT 375 Alk Phos 85 Serun proteins 5.7 Gm% Alb 3.4 Gm%, serum creatinine 1.8 mg%
Laboratory tests demonstrate neutrophilic leukocytosis favoring acute infection, conjugated bilirubinemia with increased enzymes suggestive of hepatocyte disease and high serum creatinine indicates nephritis.
Leptospirosis with liver and kidney involvement was considered. It was confirmed with IgM antibody to leptospira Amoxycillin or Doxycycline are drugs of choice.
Leptospirosis is a bacterial infection contracted through injured skin exposed to contaminated urine of rodents or other animals while walking in a water-logged area. It presents with high fever with severe myalgia and congested mucosa and few patients develop immune mediated complications affecting commonly liver and kidney but also any other organ may be involved.

Case 5

One month old infant presented with jaundice that started on D3 of life and was considered to be physiological jaundice that would settle down by itself. However, it persisted over next four weeks and hence child was brought to a doctor. Infant had normal delivery and had been on exclusive breast feeds. He had gained one kg of weight in last one month and was happy in spite of jaundice. Urine color was normal. Normal urine color rules out conjugated bilirubinemia and hence hepatocyte and biliary tract diseases are unlikely. It may be hemolytic jaundice but pallor would have been a major complaint that is not so in this infant. Thus, it is

unconjugated bilirubinemia but without hemolysis, It could be an enzyme deficiency such Crigler-Najjar syndrome that presents with severe progressive jaundice or Gilber syndrome that presents with mild jaundice later in childhood. Physical examination was normal.
Serum bilirubin was 3.2 mg% direct 0.4 mg%, other tests N
So by exclusion, we need to look at other causes.
Other causes of such jaundice are breast milk jaundice or breast feeding jaundice. Breast feeding jaundice is due to inadequate breast milk intake resulting in decrease in enterohepatic circulation. Such an infant would not gain adequate weight. Breast milk jaundice is due to a chemical substance present in breast milk that leads to jaundice that is self-limiting. As this infant had grown well, **breast milk jaundice** is the diagnosis.
There is no need to stop breast feeding as in spite of continuing breast milk, jaundice is known to disappear over next few weeks. One may prove diagnosis of breast milk jaundice by withdrawing breast milk for few days and demonstrating clearing of jaundice. However, it is not necessary as transient withdrawal may disrupt breast milk secretions besides mother gets wrong impression that her breast milk is not suiting the infant.

Case 6

One month old infant presented with jaundice since D 3 of life and was considered to be physiological jaundice. However, jaundice persisted over next four weeks. Urine color was normal. Baby was born after full term normal

delivery with birth weight of 2.5 kg, infant was on exclusive breast feeds. Mother complained about lethargy as baby would not cry for a feed and had to be coaxed to feed. Jaundice with normal urine color suggests unconjugated bilirubinemia. It is unlikely to be hemolytic jaundice as pallor would have been a major symptom. So, it is non-hemolytic indirect bilirubinemia. Infant with enzyme deficiency would not have been lethargic unless jaundice went on increasing fast. In which case, it would have resulted in convulsions or refusal of feeds. So, we need to think beyond common causes. This infant is lethargic that may suggest brain involvement that is likely to be slowly evolving disease as there are no acute symptoms of brain disease such as convulsions or refusal of feeds.

Physical examination showed lethargic infant, weight 3.4 kg, length 49 cms, head O 36 cm, heart rate 70 / minute, no pallor, no hepatosplenomegaly, no other abnormality

This infant has gained weight well but has bradycardia and his length is short for this age. This suggests delayed bone growth characteristic of **congenital hypothyroidism**. Lethargy and bradycardia are other typical features of hypothyroidism. Diagnosis can be confirmed by serum T3, T4 and TSH levels. Treatment with thyroid hormone supplement would be necessary for life.

It is important to diagnose congenital hypothyroidism right at birth to avoid permanent brain damage. It is ideal to order TSH level on cord blood on every infant at birth to suspect hypothyroidism even before symptoms develop. Prevalence of congenital hypothyroidism is one in 3500 live births and so

quite high to justify routine screening for hypothyroidism at birth. Most obstetric centers in India now screen for congenital hypothyroidism. Bone age depicts bone maturation and is estimated on x-ray by special charts (Grulich-Pyle chart). Absence of lower femoral and upper tibial epiphysis at birth suggests delayed bone maturation that is the hallmark of congenital hypothyroidism.

Case 7

8 years old child presented with fever for two days followed by jaundice. Urine color was normal. Child had been healthy without any prior illness.

Jaundice with normal urine color is due to unconjugated bilirubin. So, this is likely to be either hemolytic jaundice or due to enzyme deficiency. Enzyme deficiency is not triggered by fever but hemolysis may and so it may be acute hemolytic jaundice. Presence of pallor may favor hemolysis and absence of pallor enzyme deficiency.

Physical examination showed healthy child, well grown, pallor++, mild icterus, liver 1F +, soft, spleen 2F +, no other abnormality.

Pallor with splenomegaly suggests hemolysis and so this is hemolytic jaundice. As onset of this illness was with fever, it is likely to be acquired autoimmune hemolytic anemia and jaundice.

Investigations showed Hb 8 Gm%, normal total and differential WBC and platelets, serum bilirubin 3 mg% with direct 0.4 mg%.

This confirms anemia with unconjugated bilirubinemia.

Coomb's test is positive indicating antibody related hemolysis.
This condition may be self-limiting and if not, need to be treated with steroids and if necessary, with packed cell transfusion. Blood transfusion may itself aggravate further antibody destruction and so reserved only if necessary. In such a case, there has to be proper match between donor's and recipient's rare blood groups.

Case 8

8 years old child presented with jaundice and abdominal distension for last one week. On direct questioning, he was not well over last 6 months. He had poor appetite, loose stools at times, feeling weak and had lost 2 kg weight.
This suggests chronic progressive illness. Prior to developing jaundice and abdominal distension, his past history would have made us search for chronic evolving disease. As only localizing symptom of loose stools relate to gastrointestinal tract, one would think of GI disease. However loose stools are not frequent and there are no other symptoms of GI disturbances such as vomiting, flatulence or abdominal pain. Such a disturbance may denote indigestion which may be due to liver, biliary system or pancreatic disorders. Biliary or pancreatic disorders would present with pain. Absence of pain in this child may therefore suggest evolving hepatocyte disease. Abdominal distension noticed over just a week suggests ascites and in ascites in chronic liver disease indicates decompensation with portal hypertension.
Physical examination showed weight 16 kg, height 110 cms,

icterus +, liver 4F +,firm, liver span 10 cms, spleen 3 F+, firm, ascites +, other systems normal
Investigations showed Hb 10 Gm%, serum bilirubin 5.2 mg Direct 4.1 mg%, SGPT 100 SGOT 60 Serum proteins 4.2 Gm% Albumen 2.3 Gm%, INR 1.8
Laboratory tests prove chronic liver disease (low serum albumen) that has recently decompensated (bilirubinemia with mild raised enzymes but raised INR). Diagnosis of **cirrhosis** can be confirmed by liver biopsy that may be dangerous at this stage with liver beginning to fail. It may not offer much more information and so may be deferred. Etiology may not be apparent with liver biopsy.
Treatment is symptomatic.
Cirrhosis is different from fibrosis. In cirrhosis, besides fibrosis, there is an attempt at regeneration of liver tissue. This is the end result of chronic hepatocyte damage caused by various disorders such as hepatitis B or C infection, autoimmune hepatitis, metabolic disorders such as Wilcon disease and also by toxins.

Take home message

Detailed history can initially differentiate between conjugated bilirubinemia (high colored urine) and unconjugated bilirubinemia (normal urine color) and further between hepatocyte (sickness in spite of mild jaundice) and biliary tract disease (normal health in spite of deep jaundice). Ultimately both hepatocyte and biliary tract disease involve other areas and present as hepatobiliary disease. Pallor is the

major feature of hemolytic jaundice. Jaundice without hepatosplenomegaly denotes enzyme defect.

MCQs

1. Which of the following statement is wrong?

A) High colored urine, clay colored stools is a biliary disease
B) High colored urine, and clay colored stools may be a hepatocyte disease
C) High colored urine and normal color stools may be hepatocyte disease
D) Normal urine and clay colored stools is a biliary disease

2. Which of the following statement is wrong?

A) Jaundice in malaria is unconjugated
B) Jaundice in malaria is conjugated
C) Jaundice may not occur in Malaria
D) None of the above

3. Nausea, vomiting and anorexia are features of

A) Chronic hepatitis
B) Cirrhosis
C) Typhoid hepatitis
D) Viral A hepatitis

4. Jaundice without enlarged liver suggests

A) Wilson’s disease
B) Biliary tract obstruction
C) Chronic hepatitis
D) Malarial hepatitis

5. Jaundice without splenomegaly is seen in this condition

A) Malaria hepatitis
B) Cirrhosis
C) Enzyme deficiency
D) Typhoid hepatitis

Answers to MCQs

Q1 D, Q2 A, Q3 D, Q4 B, Q5 C

2.17. Edema – fluidin wrong sites!

Back to basics

Edema refers to increased collection of fluids in interstitial spaces including third space (pleural and peritoneal). It results from imbalance between pressures in intravascular and extravascular compartments. In health, two thirds of total body fluid is intracellular (part of extravascular compartment) and remaining is divided into intravascular (plasma and lymph), interstitial and small amount in transcellular space (ocular, cerebrospinal fluid, fluid in pleura, peritoneum and joint space). Transcellular fluid is static and not in balance with other compartments.

Pressure between compartments

Intravascular pressure pushes fluid out of vessels. Mean arterial pressure is 70-90 mm Hg, at arterial capillary end 30 mm Hg and at venous end. Intravascular osmotic pressure pulls fluid into vessels. 19 mm Hg pressure is exerted by albumen and 9 mm Hg by cations in plasma. Interstitial osmotic pressure of 8 mm Hg is contributed by interstitial

albumen. Interstitial fluid also exerts negative pressure of 3 mm Hg also helps in pulling the fluid into vessels.

Net effect

Pressure at arterial capillary end (pushing out) is more than at venous end (pulling in). This difference results in leaking of fluid at arterial capillary end. However, 90% of leaked fluid is reabsorbed at venous end because of large venous surface and big pores and remaining 10% is absorbed through lymphatics back into circulation. This is the way balance is maintained in health and no edema results.

Genesis of edema

It is clear from above mentioned physiology that edema results when there is imbalance in pressures. Increase in hydrostatic pressure and decrease in osmotic pressure pushes fluid out of intravascular compartment. Edema also results in case of capillary leak (as happens in dengue fever) and also due to lymphatic block (lymphedema).
In renal conditions, fluid retention and excess of sodium leads to leakage into interstitial spaces and hence edema results. In case of protein malnutrition, decrease in albumen causes edema and so also in liver disease in which excess fluid accumulates in peritoneal cavity due to portal hypertension. In congestive cardiac failure, edema results due to increase in hydrostatic pressure. Capillary leak besides edema, also results in fluid collection in pleural or peritoneal cavity. Lymphatic obstruction as well as venous obstruction leads to localized edema related to site of obstruction. In angioedema, allergic inflammation leaks out fluid locally out

of capillaries resulting in local edema (sudden swelling of eyes or lips).

Common causes of edema

Renal - acute nephritis and nephrotic syndrome presents with sudden onset of periorbital edema. Patient typically complains of eyelid edema noticed on waking up in the morning. This is because of acute onset edema is first visible in loose connective tissue around eyes and venous congestion around face during sleep makes it appear typically on waking up in the morning.

Angioedema – this is another condition where onset of edema is sudden and so again noticed around eyes in addition to swelling of lips with redness and itching.

Chronic liver disease – presents with edema feet and collection of fluid in peritoneal cavity due to portal hypertension. As onset of edema in liver disease is slow, it manifests on dependent parts on feet. In acute liver disease such as acute hepatitis, there is no edema as half-life of albumen is 2-3 weeks and hence it takes that much long time before edema manifests.

Congestive cardiac failure presents with edema feet as edema develops slowly and so seen on dependent parts over feet in an ambulant patient. In non-ambulant patient, it may be seen on sacral area – dependent part of the body in lying down position. Though sacral edema is not noticed by patient

Protein malnutrition (PEM) presents as edema feet often of acute onset. This is because subclinical hypoproteinemia exists over time but a trigger such as acute diarrhea

manifests oedema simulating acute onset. Unlike acute onset oedema in an apparently normal individual in a renal disease, acute onset oedema in PEM manifests in a chronically sick individual.

Capillary leak syndrome presents with edema in dependent parts but spread fast to serous cavities.

Lymphatic or venous obstruction at a particular site presents with localized edema (say of one leg only) as against generalized edema (both legs)

Myxedema is seen in hypothyroidism and lipiedema in morbid obese patient. Edema is non-pitting in these conditions and so also in chronic lymphatic obstruction due to thickening of accumulated lymph (as in chronic filariasis)

Clinical approach

Detailed history

Asses whether edema is generalised or localised. Obviously causes would vary.

Onset of edema – acute onset edema noticed around eyes is mostly due to acute renal disease and also may be seen in angioedema (it is localised and with itching, redness). Capillary leak syndrome may also develop oedema in a short time including in serous cavities. Slow onset of edema is classical in other conditions such as liver, cardiac diseases and PEM (latter condition seem to manifest acute oedema but in reality, it is subclinical oedema triggered by an event such as diarrhoea)

Degree of edema – severe sudden onset edema is typical of nephrotic syndrome referred to as anasarca. Rarely it may be

seen in late stages of other disorders in young children.
Urine output – oliguria is seen in acute nephritis along with high colored urine due to hematuria. Oliguria may also appear in dehydrated state due to poor intake or loss of fluids as in diarrhea and also seen in cardiac failure due to poor renal perfusion and in severe intravascular constriction due to massive edema in liver disease or capillary leak.
High colored urine may be due to hematuria, jaundice and also concentrated urine due to dehydration.
Accompanying symptoms would pinpoint to a specific organ involvement such as jaundice in liver disease, breathlessness and palpitation in cardiac disease and irritability or lethargy in PEM.
Physical examination
It is necessary to confirm pitting edema by transient pressure over bony surface such as shin of tibia. Edema is pitting in most conditions though it may be non-pitting in hypothyroidism and chronic lymphatic obstruction.
State of health offers clue. Comfortable child with massive edema is typical of nephrotic syndrome. Child is acutely sick looking in capillary leak syndrome while chronic sick look is seen in liver, cardiac diseases or in PEM.
Ascites is typical of chronic liver disease and may be seen in nephrotic syndrome and capillary leak along with pleural effusion.
Other signs such as systemic hypertension would suggest a renal disease, jaundice and hepatomegaly a chronic liver disease, enlarged liver also in cardiac disorder due to congestive cardiac failure, cardiomegaly and heart murmur

indicative of cardiac disease and signs of PEM such as skin, hair and mental changes denoting severe protein malnutrition.

Investigations

Provisional diagnosis would help in ordering specific investigations. Urinalysis and biochemistry in renal disease, urine for bile salts and pigments and liver function tests in liver disease, chest x-ray, ECG and 2D echo cardiogram in cardiac disease are main investigations. Other tests would depend on primary conditions.

Management

Edema per se rarely needs drug therapy and so management is focused on primary organ disease. Diuretics should not be routine prescription for every edema and choice of diuretic may also depend on type of disease. In severe edema due to nephrotic syndrome, diuretic is rarely required but it may lead to intravascular constriction with further damage to kidneys and hence cautious approach is necessary. Diuretics may be useful in cardiac failure and certainly not in PEM.

Take home message

Oedema is a presenting symptom of a renal disease, angioneurotic oedema or localized venous or lymphatic obstruction. Oedema is not a presenting symptom in liver or heart disease and protein malnutrition as these diseases present with other symptoms in which oedema is often noticed by the doctor. Sudden weight gain may be due to

oedema that is missed on physical examination. It is not difficult to find the cause of oedema clinically.

MCQa

1. This fluid is not in balance with fluid in other sites

A) Interstitial
B) Intracellular
C) Transcellular
D) Intravascular

2. Which of the following statement is WRONG? Edema results from

A) Increased capillary pressure
B) Increased capillary permeability
C) lymphatic obstruction
D) None of the above

3. Which of the following statement is WRONG? Ascites in case of generalized edema is seen in

A) Cirrhosis of liver
B) Protein malnutrition
C) Nephrotic syndrome
D) Capillary leak

4. Which of the following statement is WRONG? Oliguria may be present in

A) Nephrotic syndrome
B) Cardiac failure
C) Capillary leak
D) Myxedema

5. This patient is well and happy in spite of edema

A) Capillary leak
B) Nephrotic syndrome
C) Protein malnutrition
D) Angioedema

Answers to MCQs

Q 1 C, Q 2 D, Q 3 B, Q 4 D, Q 5 B

2 18. Edema – timely intervention a need!

Clinical application of basic concepts

Edema results from various factors such as increased hydrostatic pressure, reduced osmotic pressure, increased permeability of capillaries and lymphatic or venous obstruction. Fluid collects in interstitial spaces and serous cavities. Understanding pathogenesis helps in defining probable cause of edema and relevant timely intervention. Acute onset edema in a normal individual is first perceived in loose connective tissue around eyes before it is noticed elsewhere as happens typically in acute renal disease and angioedema. Chronic cardiac disorders present with edema on dependent parts – legs in an ambulatory persons while chronic liver disease has localizing fluid collection in peritoneal cavity – ascites besides edema of legs. Protein-calorie malnutrition may manifest edema suddenly, triggered by minor illness such as diarrhea. Edema in an acutely sick child may represent capillary leak as seen in dengue fever. Localised edema is due to venous or lymphatic obstruction.

Case-based discussion

Case 1

1.Eight years old child presented with acute onset of edema of eyelids noticed one morning on waking up. On next day, he developed breathlessness. On direct questioning, he had oliguria and cola colored urine.
Acute onset of periorbital edema with oliguria and cola colored urine suggestive of hematuria is in favor of acute glomerulonephritis - AGN. Breathlessness in this child may be due to hypertension a result of AGN.
Physical examination showed mild edema feet and around eyes, HR 140/min, RR 35/min, BP 150/100 mm Hg, systemic examination normal. It is diagnostic of **Acute glomerulonephritis**. Investigations – urinalysis RBCs ++, granular casts +, proteins +, serum creatinine 1.8 mg%, ASO high titer.
In children > 5 years of age, it is most likely due to post-streptococcal infection and if so, prognosis is very good. Improvement is quick with symptomatic treatment though microscopic hematuria may continue for few weeks. To confirm improving situation, serum C3 level should be ordered. It is low at the peak of illness, starts rising during recovery. If C3 level continues to be low, it may indicate chronicity of the disease and needs referral to a specialist. If AGN presents in younger child, it may be due to other infections and prognosis may be guarded.

Case 2

Four years old child presented with acute onset of edema around eyes. There were no other symptoms. On direct questioning, urine output was normal and urine was colorless. However, child had itching and redness around the swelling that also spread to lips.
This suggests **angioedema** – allergic process and not renal disease.
Physical examination showed edema around both lower eyelids looking pink. There was no edema anywhere else, Blood pressure was normal and so also heart and respiratory rate, Systemic examination was normal.
There is no need for any investigations or any drug therapy. It settles down by itself. Antihistamines and steroids are not necessary unless child presents with laryngeal edema causing inspiratory obstruction, it is rare.

Case 3

One year old infant presented with loose stools for two days followed by acute onset edema of face and feet. He was born after full term and normal delivery with birth weight of 2.6 kg He was on exclusive breast feeding for first 6 months and thereafter mother started dilute cow milk and occasional watery mixture of rice and dal.
Diet history clearly suggest inadequate intake of calories and proteins for last 6 months and hence edema is likely to be due to nutritional deficiency – PEM. His subclinical deficiency must have been precipitated by episode of loose stools and thus edema developed acutely though he must have had low serum proteins even before episode of loose stools.

Physical examination showed wt 9 kg, length 70 cms, head O 45 cms, lethargic but irritable on disturbance, edema feet ++, abdomen distended, liver 3F +, firm, liver span 7 cms, spleen not palpable, no ascites, other systems normal,
Diagnosis of **protein energy malnutrition** is evident.
There is no need of investigations to prove the diagnosis, however one may look for other deficiencies such as anemia and also evidence of any occult infection such as tuberculosis or urinary tract infection.
Treatment revolves around resuscitation in case of severe PEM, restoration of deficiencies, rehabilitation and prevention by proper counselling.
It is important to note that acute onset oedema in this child occurred in a poorly growing child and hence, it suggests occult hypoproteinemia that became manifest with acute diarrhea. Thus, this is a chronic disease manifesting acute oedema as against a renal disease with acute onset oedema in a healthy child.

Case 4

Ten years old child presented with oliguria and cola colored urine for last few days. Physical examination showed minimal edema feet and investigations suggested diagnosis of glomerulonephritis.
While diagnosis of glomerulonephritis is acceptable, it seems to be different than classical post-streptococcal acute glomerulonephritis. Hence it was decided follow this child clinically and also with repeat serum C3 level.
Physical examination showed persistent mild edema feet,

oliguria and hematuria. Also, his blood pressure revealed increasing though on 95th centile for age. It suggests development of hypertension.
C3 level continued to be low even at the end of 6 weeks of persistent glomerulonephritis. It suggested diagnosis of **chronic glomerulonephritis**.
This child should be referred to a specialist for further management that may need renal biopsy to define probable cause and prognosticate the disease.
This case illustrates the need for clinical suspicion of unusual type of glomerulonephritis and value of serum C3 level in follow-up of acute glomerulonephritis. Besides clinical abnormality, persistent low C3 level is a marker for need for referral. This case represents the way a generalist should pick-up an atypical course of a disease with timely referral to a specialist. If missed, patient often presents with chronic persistent disease which is obvious but too late.

Case 5

Two years old child presented with acute onset edema noticed first around eyelids that was followed 12 hours later with generalized edema all over the body. There was no history of oliguria, high colored urine or any other symptoms. Sudden development of generalised edema without any prior symptoms and edema starting around eyes suggests renal glomerular pathology. In view of absence of oliguria or high colored urine and massive edema favor glomerular epithelial pathology such as nephrotic syndrome.
Physical examination showed comfortable child, happy and

active, blood pressure 90/50 mm Hg with massive generalized edema, abdominal distension with ascites but without hepatosplenomegaly. There were no other findings. These findings suggest severe hypoproteinemia that has led to massive generalized edema and also ascites but without endothelial glomerular involvement as evident by absence of oliguria, high colored urine or hypertension.

Investigations showed urine protein +++, urine – creatinine protein ration of 3, serum proteins 4.2 Gm%, albumen 1 Gm%,, globulin 3.2 Gm%, serum cholesterol 320 mg and creatinine 0.5 mg.

Massive proteinuria and albuminuria, hypercholesteremia and normal serum creatinine favor diagnosis of **minimal lesionnephrotic syndrome.**

Management consists of oral prednisolone 2 mg/kg/day for 4-6 weeks followed by two third total dose on alternate day for 4 weeks. Such a prolonged therapy during first attack offers better chance of complete remission without further relapse. However in case of relapse, similar therapy needs to be given but for shorter period.

Minimal lesion indicates pathology restricted to epithelium of glomerulus. It typically presents for the first time between age of 2 and 5 years of age and carries usually good prognosis in spite of relapses. If this child develops diarrhea, there may also be oliguria due to dehydration and urine color also may be high colored. Diarrhea in such a child may be due to edema of intestinal mucosa that leads to malabsorption but may also be due to infection.

However, if such a child presents in later childhood (unusual

age) or has oliguria and hypertension but with massive edema, it may suggest primary glomerular epithelial disease (because of massive edema) that has also involved glomerular endothelium (because of oliguria and hypertension). This is referred to as complicated nephrotic syndrome and needs referral to a specialist.

Case 6

Two years old child presented with history of fever and macular skin rash over last three days. Considering it to be viral infection, symptomatic treatment was given and fever abated. However very next day, child started vomiting and had severe abdominal pain. This was followed by edema feet and abdominal distension. On direct questioning, child had not passed urine for last 12 hours.
Initial symptoms do suggest viral infection but it did lead to unexpected symptoms of vomiting and abdominal pain. It indicates immune mediated complication and not extended viral infection as original symptoms had totally disappeared. Sudden abdominal pain along with vomiting suggests probable vascular complication involving intestines that is immune mediated. As child had not passed urine, it may denote shock and so abdominal pain is due to severe intestinal ischemia. This suggests capillary leak syndrome. Physical examination showed child in shock with cold extremities, pale skin, marked tachycardia, low blood pressure, mild edema feet, ascites and also mild pleural effusion. This is classical of **capillary leak syndrome** following probable dengue viral infection.

Investigations confirmed dengue fever. Child was treated aggressively for shock and recovered in next two days.
This case illustrates how primary infection may get better but lead to immune mediated complications within a day or two of apparent cure of primary infection. Such a situation cannot be anticipated but every child should be observed for 2-3 days after apparent cure for such a possible complication. If diagnosed in time, child can be saved. Rarely, such immune mediated complications may manifest even after few weeks of primary viral infection (Covid 19 pandemic is an example).

Case 7

8 years old child presented with history of progressive abdominal distension over last 6 months and edema of feet noticed over last two months. He had lost 3 kg weight and had poor appetite.
This looks to be chronic progressive disorder as evident by loss of weight over few months. Abdominal distension could be due to organomegaly (commonly enlarged liver with or without spleen) or ascites. Further development of edema feet in this case indicates chronic liver disease. In absence of jaundice, it may be well compensated liver disease (means liver has not failed though poorly functioning).
Physical examination showed chronically sick looking child, edema feet +, abdominal distension, liver 3 cms +, firm, liver span 9 cms +, spleen 2 cms +, no ascites, no jaundice, other systems normal
Firm and enlarged liver with splenomegaly favors chronic liver disease with portal hypertension. Absence of ascites,

bleeding or encephalopathy suggests well compensated chronic liver disease such as **cirrhosis.**
Investigations – Hb 9 Gm%, serum proteins 5.2 Gm%, albumen 2.4 Gm%, globulin 2.8 Gm%, ALT (SGPT) 210 AST (SGOT) 160 Serum bilirubin 0.9 mg
There is marked hypoalbuminemia with mild rise in liver enzymes with normal bilirubin. (Bilirubin is increased only when more than 85% of liver cells are damaged and so in chronic liver disease, increased bilirubin or jaundice suggests liver gradually failing. However in acute hepatitis, increased bilirubin does not mean failing liver).
Diagnosis of cirrhosis can be confirmed by liver biopsy.
Management is palliative.
Hypoalbuminema in a patient with enlarged liver is a marker of chronic liver disease and should be picked up even before oedema manifests. It is rare to find hypoalbuminemia in acute liver disease unless such an acute liver disease occurs in an individual with occult asymptomatic liver pathology such as obesity, undernutrition and hepatitis B infection carrier state.

Case 8

10 years old child presented with history of breathlessness and edema feet over last few days. On direct questioning, he used to feel breathless on exertion but had ignored the same. There was no past history of any major disease.
This child seems to be having slowly progressive cardiac disease and now presenting with failure as evident by breathlessness and edema feet.

Physical examination showed sick child, HR 120 / min, RR 27/min, BP 120 / 60 mm Hg, mild edema feet, engorged neck veins with positive hepatojugular reflex, apex beat on 6th intercostal space outside midclavicular line, systolic murmur at mitral area conducted to axilla.
These findings suggest **mitral regurgitation with CCF**.
It is mostly due to Rheumatic disease though there is no past history of throat infection or arthritis. (Throat infection may often be mild or even asymptomatic and so not reported)
Diagnosis can be confirmed by chest x-ray, ECG and 2D echocardiogram (showing active valvulitis) and etiology indicated by high anti-streptolysin O titre.
If this is active c rditis, it is treated with steroids followed by aspirin and further long acting penicillin to prevent relapses.
Besides, symptomatic therapy for CCF is necessary. Oedema in cardiac failure denotes right sided failure. It is not a common finding in congenital heart disease but may be seen in chronic acquired heart disease due to rheumatic fever. Most other cardiac conditions present with either left ventricular or biventricular failure. Isolated right sided failure is nature's way to avoid left sided failure and is possible only in chronic disorders where nature has time to compensate.

Take home message

Analysis of oedema illustrates importance of chief complaint. If oedema is a chief complaint, it is mostly renal in origin whereas most other conditions present with other symptoms in which oedema is often noticed by the doctor. Angioedema is an exception and presents acutely in a healthy child. Final

diagnosis is easy in terms of system affected and further can be confirmed by physical examination and necessary investigations.

MCQs

1. Which of the following statements is wrong related to classical post-streptoccocal acute glomerulonephritis?

A) Typical age group is beyond 5 years
B) Microscopic hematuria may persist for weeks
C) Edema and oliguria improve quickly
D) Prognosis is guarded

2. Urinalysis is completely normal in spite of edema

A) Acute nephritis
B) Cardiac failure
C) Angioedema
D) Liver disease

3. Child with edema may be breathless in

A) Acute nephritis
B) Nephrotic syndrome
C) Angioedema
D) All of the above

4. Which of the following statement is wrong? Relapse is possible in

A) Nephrotic syndrome
B) Cardiac failure
C) Cirrhosis
D) Protein malnutrition

5. Blood pressure may be abnormal in this child with edema

A) Nephrotic syndrome
B) Capillary leak
C) Cardiac failure
D) All of the above

Answers to MCQs

Q 1 D, Q 2 C, Q 3 D, Q 4 C, Q 5 D

2.19. Bulging abdomen –sign of ill-health!

Back to basics – abdominal distension

What is abdominal distension?

Distension refers to enlargement, dilation or ballooning effect and when applied to abdominal distension, it commonly relates to intra-abdominal space occupation or swelling but also may be related to abdominal wall. It is a subjective observation by the patient or a doctor but abdominal girth can be measured, specially to monitor progress in generalized abdominal distension. Patients often complain of bloating – a sensation of fullness and tightness attributed to “gas”. Localised abdominal distension usually presents as swelling in a part of abdomen. Abdomen of normal young infant is protuberant due to liver and spleen being accommodated in the abdomen unlike in an adult in whom these organs are mostly in the chest.

Causes of abdominal distension

There are several causes of abdominal distension that can be grouped – most of the words describing these groups start from alphabet F – faeces flatus, fat, fetus, fluid, flab (flabby abdominal muscles), food and functional. Other causes include organomegaly (enlarged liver and / or spleen) and any other space occupying lesions such as tumor or cyst. Each of these disorders may result from varied causes..

Defining causes in each group

Faeces (often along with accumulation of flatus) leading to abdominal distension may be acute as in intestinal obstruction or paralytic ileus due to sepsis or hypokalemia and also may be chronic as in case of subacute intestinal obstruction due to congenital megacolon or intestinal tuberculosis. One of the most common cause is habitual constipation due to low fiber in the diet and poor bowel habits. It may also be caused by inadequate food intake or intestinal disorders such as irritable bowel syndrome or inflammatory bowel disease, dyspepsia (indigestion) or systemic disorders such as diabetes and hypothyroidism.
Flatus (retained gas) may also be caused by all above mentioned disorders besides lactose intolerance and aerophagy.
Fluid in abdomen may lie free in peritoneal cavity (ascites) or may be localized (mesenteric or ovarian cyst). Ascites fluid may be transudate (non-inflammatory) as in case of portal hypertension and nephrotic syndrome or exudate (inflammatory) as in case of peritonitis or malignancy.
Organomegaly (enlarged liver with or without splenomegaly)

may be due to primary liver or hematological diseases, systemic infections or storage disorders.

Flab (flabby abdominal muscles) may result from malnutrition due to loss of muscle mass or generalised poor muscle tone. Stretched abdominal muscles for longer period as in pregnancy may also result in flabby abdominal muscles.

Food may lead to abdominal distension due to indigestion, constipation, food intolerance or allergy.

Functional disorders may result in abdominal distension due to gut-brain interaction. This is a result of mental stress or anxiety in a susceptible individual who cannot cope up with it. It is now clear that emotions are controlled by gut while brain is responsible for action. During anticipated stress situation (student appearing for examination), one may get urge to pass stool or urine once more. If one sees something frightful, intestines cramp, it is a result of gut-brain interaction. And in colloquial English language, we say “it is my gut feeling”. This is how gut-brain interaction results in various intestinal disorders. Bloating – a sensation of fullness in abdomen may not be due to accumulation of gas (flatus) but even without gas. This is due to contraction of diaphragm (result of gut-brain interaction) that pushes liver and spleen into abdomen resulting in abdominal distension.

Fat and fetus are easy to make out.

Localised swelling if very large, especially retroperitoneal, may present as abdominal distension but if lies anteriorly in the abdomen, it is complained of as lump in abdomen. Typically enlarged spleen presents as lump in left hypochondrium.

Clinical approach

Detailed history analysis and focused physical examination can offer provisional diagnosis.

Onset may be acute in surgical or metabolic diseases while gradual in many other conditions.

Progression helps to define probable cause, Waxing and waning abdominal distension suggests either constipation or gaseous distension. Progressive generalized distension for few days but remaining stable thereafter indicates probable ascites while organomegaly or other tumors / cysts may present as slowly progressive localized abdominal distension over several weeks.

Accompanying symptoms such as vomiting, constipation, diarrhea, abdominal pain, pallor, jaundice help to localize the disease to a specific organ and also define probable pathology.

Personal history of loss of appetite or weight, sleep and behavior disturbances offer clue to diagnosis and so also past, family and drug therapy history.

Site of abdominal distension – generalised or localized.

Shape of distended abdomen in case of generalized abdominal distension, flank fullness suggests fluid (ascites) while distension mainly in upper part of abdomen as depicted by downward displacement of umbilicus indicates enlarged liver (distance between xiphisternum and umbilicus is much mcore than distance between umbilicus and symphasis pubis – normally umbilicus is placed midway between xiphisternum and symphasis pubis).

Percussion can differentiate between gaseous distension

(tympanic note) and fluid (dull note) and further ascites is confirmed by demonstration of shifting dullness whereas absence of shifting dullness suggests encysted fluid as in case of mesenteric or ovarian cyst..

Localised distension in a particular quadrant of abdomen is related to organs in that quadrant. Thus, enlarged spleen presents as lump in abdomen in left hypochondrium and swelling increasing across to right iliac fossa. Enlarged kidney presents as lump in lumber region and appendicular lump in right iliac fossa.(Neuroblastoma and Wilms tumor are other causes of localizes swelling. Wilms tumor does not cross midline while neuroblastoma often spreads across the abdomen. (Tumor due to neuroblastoma may not be palpable and at times even not picked up on routine imaging). Large lymph node mass present in the middle of abdomen. Enlarged liver may not present as localized abdominal distension unless large enough, especially in older children and adults.

Consistency relates to duration and pathology of disease. Firm hepatomegaly suggests chronic process while hard liver or any mass indicates probable malignancy. Soft swelling may suggest cyst or softening lymph nodes.

Pain and Tenderness indicate inflammatory process or stretching of capsule as in case of enlarged liver or lymph nodes. Congested liver as in congestive cardiac failure is also tender, though chronic congestion may not cause pain.

Other systems affection may offer clue to probable cause. Hematological diseases may present with pallor, purpura, bony tenderness or generalised lymphadenopathy. Tender

enlarged liver with engorgement of neck veins and hepatojugular reflex suggests congestive cardiac failure while congested liver due to constrictive pericarditis is often missed as it presents as enlarged liver without hepatojugular reflex and obvious cardiac symptoms. Neurological manifestation may suggest neurometabolic disorders that present with hepatosplenomegaly and abdominal distension as in case of Gaucher or Nimman-Pick disease.

Investigations

Provisional diagnosis is a prerequisite to planning investigations that would help in minimizing tests. Acute abdominal distension demands abdominal x-ray in erect position to rule out intestinal obstruction. USG of abdomen is useful in variety of conditions causing abdominal distension. Biochemical tests are reserved for specific organ involvement. Ascites tap can help in differentiating exudate from transudate by serum ascites albumen gradient. Histopathological diagnosis is important in chronic liver disease as well as tumors.

Treatment

It is possible only when final diagnosis has been reached. Few conditions may be amenable to surgery or drugs while palliative therapy is possible for some other conditions.

Take home message

Acute onset of abdominal distension is usually a surgical disease in a sick individual while one that presents over few days (may be in a sick person) or weeks (often in non-sick

individual) is usually due to medical disorders. Localised swelling relates to the organs occupying in a particular quadrant while generalized distension is due to accumulation of gas (tympanic note on percussion) or fluid (dull note on percussion and shifting dullness). Occasionally, very large mass may present as generalized abdominal distension.

MCQs

1. Which of the following statement is WRONG? Abdominal distension exists without intra-abdominal pathology in

A) Normal infant
B) Malnourished child
C) Obese child
D) None of the above

2. Which of the following statement is WRONG? Abdominal distension waxes and wanes over short time in

A) Aerophagia
B) Malnutrition
C) Subacute intestinal obstruction
D) Habitual constipation

3. This condition presents with chronic abdominal distension due to constipation

A) Congenital megacolon
B) Acute intestinal obstruction
C) Paralytic ileus
D) Intestinal tuberculosis

4. Ascites is a feature of

A) Abdominal tuberculosis
B) Capillary leak syndrome
C) Portal hypertension
D) All of the above

5. Most useful parameter in examination of ascites fluid is

A) Cell count
B) Protein content
C) Serum ascites albumen gradient
D) Sugar content

Answers to MCQs

Q1 D, Q2 C, Q3 A, Q4 D, Q5 C

2.20. Abdominal distension –don't ignore

Clinical application of basic concepts

Abdominal distension may be generalized due to accumulation of gas, collection of fluid or chronic constipation with accumulation of faeces. Distension caused by flatus or faeces wax and wane while there is no variation in other conditions. Typically, enlarged liver presents as upper abdominal distension that displaces umbilicus downwards from its usual mid-position. Enlarged spleen or any other mass in abdomen presents as lump in a specific quadrant of abdomen. Surgical conditions leading to abdominal distension are usually acute while medical conditions are often chronic. Ofcourse, there are exceptions on both side such as, congenital megacolon a chronic surgical

problem and paralytic ileus following diarrhea as acute medical problem. Hypotonia of abdominal muscles and excess of abdominal fat may also lead to abdominal distension though it is not a primary presentation.

Case based discussion

Case 1

8 years old child presented with periumbilical pain and generalized abdominal distension for 2 days. It was accompanied with mild fever, occasional vomit and loose stool. Vomitus contained food particles and not bile stained. Sudden onset of abdominal pain and abdominal distension suggest acute inflammatory pathology and occasional vomit and loose stool rules out primary intestinal disease but related to structure near to intestine. Had it been a primary intestinal disease, loose stools and / or vomit would have been prominent symptoms. Acute infection in a healthy child generally presents with high fever, though low grade infection may cause low grade fever. It may also be non-infective inflammatory disease.
Physical examination on D2 showed a sick looking child and generalized abdominal distension, tenderness all over and guarding. It denotes oncoming acute inflammatory condition. Next day pain shifted to right iliac fossa and became severe. Diagnosis of **acute appendicitis** was considered and proved on USG.
Parents refuse surgery and next day, pain disappeared. Does it suggest sudden natural recovery?
On physical examination, child looked much more sick, HR

140, BP 80/50 mmHg, CRT > 4 seconds, child was in shock. This suggests appendix had developed **gangrene** and so pain disappeared but child went into shock endangering life.
This case illustrates a fact that symptom relief may not be all well and we should go by overall condition.
Visceral abdominal pain is diffuse, not severe and often accompanied with occasional vomiting or loose stools while parietal pain is severe and localized. This is why pain in appendicitis starts near umbilicus (appendix originates from midgut and hence initial location of pain) and then when peritoneum is involved, pain shifts to right iliac fossa and becomes severe. It is classic of acute appendicitis.

Case 2

One year old infant presented with gradually progressive abdominal distension. He was breast-fed for first 7 months and then also on semisolid food. On direct questioning, he was constipated from first month, had not gained weight over last few months. His development was normal. Breast-fed infant is never constipated unless there is something abnormal. Progressive abdominal distension and failure to gain weight are both a result of worsening constipation.This is likely to be chronic progressive lower intestinal obstruction, mostly congenital megacolon**.**Physical examination showed weight 7.2 kg, length 70 cm, moderate pallor, abdomen loaded with faeces, per rectal examination revealed empty rectum and ribbon like stool coming out, suggesting colonic obstruction proximal to rectum. Diagnosis was conformed as **congenital megacolon**by barium enema

and rectal biopsy showing absent ganglion cells. He was surgically treated.

Intestinal obstruction generally presents as an acute event but it may also be intermittent as in case of subacute intestinal obstruction due to intestinal tuberculosis. It is important to note that breast-fed infant is never constipated and this case is an example of chronic progressive intestinal obstruction in a breast-fed infant. Constipation in congenital megacolon may be initially considered to be due to habitual constipation though failure to thrive and persistent abdominal distension are the clues to a pathological problem.

Case 3

One year old infant presented with loose stools and abdominal distension for 2 days. Loose stools stopped suddenly but abdominal distension worsened. There was no vomiting or any other symptom.

When any symptom suddenly and unexpectedly gets better, it is often indicative of a complication. In this child, loose stools suddenly stopped, it may suggest either intussusception or paralytic ileus. Intussusception presents as abdominal pain and vomiting and so not likely in this child. Paralytic ileus may be a manifestation of sepsis or hypokalemia. As this child has had no fever, sepsis is not likely and so it may be hypokalemia.

Physical examination on D3 showed a malnourished child, HR 120 / min RR 25 / min generalized abdominal distension, poor peristalsis, muscle hypotonia, deep tendon reflexes

sluggish. Disproportionate tachycardia, muscle hypotonia and sluggish DTR suggest hypokalemia. Potassium is intracellular ion and so its deficiency affects skeletal and heart muscle besides intestinal muscle. That explains tachycardia and sluggish DTR. Serum potassium was low and diagnosis of **hypokalemia** was confirmed. Child was treated with IV potassium – it is only in extreme situation that one needs to use IV potassium and that too with caution. Oral potassium supplements are ideal in the form of coconut water to prevent such hypokalemia especially in malnourished children and elderly persons.

If a healthy child suffers from diarrhea, transient hypokalemia settles down by itself but as this child was malnourished having low potassium, hypokalemia after diarrhea may be life threatening due to cardiac rhythm dysfunction or severe muscle paralysis affecting respiratory muscles. Such a complication may need potassium supplements urgently. Potassium is excreted by kidney only if it is in excess but it is also secreted continuously by renal tubules irrespective of potassium pool in the body. That is why person may feel tired even after short duration of illness such as viral fever or diarrhea. In a healthy individual, it settles down by itself in next 2-3 days.

Case 4

Two years old child presented with fever and skin rash for 2 days. Fever abated but child developed abdominal distension and became sicker.

Fever and skin rash suggests viral infection that seemed to

settle by itself as expected. Whenever new symptoms appear unexpectedly while original symptoms get better, this is likely to be either metabolic or immune mediated complication. As this child also got sicker, it is likely to be immune reaction to viral infection. Sudden development of abdominal distension denotes fluid collection – ascites due to capillary leak. Sickness in such a case is due to development of shock. Physical examination showed signs of shock, mild edema of feet and ascites. It suggests diagnosis of **capillary leaksyndrome** following dengue viral infection that can be confirmed with NS1 antigen and IgM antibodies.
Such a complication must be diagnosed in its early stage of compensation as denoted by poor urine output and change in behavior of the child in spite of fever abating. At this stage, there are no obvious signs of shock and child can recover with IV fluids maintaining intravascular volume till capillary leak stops naturally. However once uncompensated shock develops with low blood pressure, it may irreversible.
Any infection may trigger excessive immune response that may appear few days after fever disappears or even may occur with continuation of fever. Any unexpected course of events in a given infection should alert a physician about such a complication.

Case 5

8 years old child presented with abdominal distension for a week. There were no other symptoms.
Acute onset of abdominal distension would have other symptoms such as vomiting / constipation in case of

intestinal obstruction or fever, skin rash in case of capillary leak syndrome or diarrhea preceding abdominal distension. As this child did not report any such symptoms, on direct questioning, it was revealed that he was not well for past 5 months with loss of appetite and weight as well as mild abdominal distension. So, it is clear now that this child has chronic disease that seems to have developed worsening abdominal distension over last one week. It suggests ascites in a child with chronic liver disease.

Physical examination showed weight 23 kg, height 120 cm, mild pallor, edema feet, no jaundice, liver 4F +, firm, not tender, span 9 cm, spleen 2F +, ascites +

These findings are in favor of chronic liver disease – cirrhosis with ascites. Cause may be either exposure to poisons such as alcohol or previous infections such as HBV or HCV.

Laboratory investigations showed serum bilirubin 1.2 mg D 0.6 mg ALT 230 AST 180 Alk phos normal Serum proteins 5.3 Gm% Albumen 2.1 Gm%

Diagnosis of **cirrhosis** is made that can be proved by liver biopsy and further tests to assess probable cause.

Treatment is merely symptomatic.

This case illustrates importance of detailed history that should always begin with assessing whether the patient was genuinely normal prior to onset of recent problem. This is because minor symptoms existing before onset of major symptom are often ignored by patients or relatives and not reported unless asked for it specifically. Best way to ensure that a patient was completely well prior to onset of present problem is to enquire about activity, energy, appetite, sleep,

behavior, bowel and urination – in short detailed personal history. Minor symptom does present with change in one or more of these factors.

Case 6

10 years old child presented with gradually increasing generalized abdominal distension over last 6 months. There were no other symptoms. He remained healthy during this time.

This child seems to have slowly progressive space occupying lesion in abdomen that has not caused any organ dysfunction. So, it must be a benign condition. As abdominal distension was generalized, it may be either ascites or very large mass. Ascites would have presented with other symptoms as generalized edema in nephrotic syndrome or other symptoms as in chronic liver disease. Both are unlikely in this child. Very large benign mass is possible though it could have produced pressure on some organs with some symptoms. So at this stage, history clearly suggests benign intra-abdominal mass.

Physical examination showed healthy normal child with generalized abdominal distension with dullness all over but without shifting dullness. Though rarely, shifting dullness may be absent in case of very large ascites. So, this child has fluid containing mass that means cyst. Abdominal USG confirmed it to be a **mesenteric cyst.** It was removed surgically.

This case illustrates a fact that patient or in this case, mother of a child was giving a clue by stating that there was slowly

progressive abdominal distension over last six months. Ascites does not present with this way over such a long period. It is always right to believe what patient says as it is rarely proved wrong. It is especially true when mother reports about her child.

Case 7

8 years old child presented with gradually progressive abdominal distension over last one year, loss of appetite and weight over last 6 months and feeling tired over last 3 months.

This is slowly progressive disease with probable hepatosplenomegaly and lately has developed severe anemia as suggested by feeling of tiredness. So disease must have started in liver and / or spleen and now spread to bone marrow. Such a disease is likely to be due to storage of abnormal metabolites – metabolic disorder.

Physical examination showed wasted and stunted child with severe pallor, liver 3F +, firm, span 10 cm, spleen 5F +, no ascites or jaundice.

Such a massive enlargement of liver and spleen without any liver dysfunction is typical of storage disorder that has spread to bone marrow and hence severe anemia.

Laboratory tests showed Hb 5 Gm%, WBC 2300 Pl 0.4 lakhs, LFT normal, USG showed large liver and spleen, no evidence of portal hypertension.

Further specific enzyme studies confirmed diagnosis of **Gaucher disease.**

If diagnosed early before significant damage, enzyme

replacement is possible though such a therapy is very costly. Once disease spread to brain, there would be no use of enzyme replacement therapy.
Progressive abdominal distension over months are broadly of three types – present without disturbed health as in case of mesenteric cyst / benign tumor or with hepatosplenomegaly and growth failure but without liver dysfunction as in storage disorder or with abnormal liver functions as in cirrhosis with portal hypertension and ascites.

Case 8

8 years old child presented with abdominal distension off and on for last 6 months. Abdominal distension would be waxing and waning and he also had occasional loose stools. He had not lost weight or appetite.
Waxing and waning abdominal distension is either due to flatus or constipation. As this child is not constipated, it must be flatus. It is necessary to find out cause of excessive flatus. As he has remained healthy, it is unlikely to be any significant pathology. It could be due to irregular food habits, life style or due to stress or anxiety – **functional disorder.**
Physical examination did not reveal any abnormality except moderate abdominal distension with tympanic note on percussion suggestive of gas.
This child does not need any tests but needs counseling.
It is well known that intestines sense the stress first and sends message to brain to execute appropriate action. Hence, functional disorders are often related to gastrointestinal system and may present as dyspepsia,

abdominal pain or distension, change in bowel habits such as diarrhea or constipation, anorexia, nausea and vomiting. Diagnosis of such disorders depend on high index of suspicion in a typical situation aided by exclusion of other pathological conditions. Irritable bowel syndrome is one such example, diagnosis of which is based on standard criteria.

Take home message

Acute onset of abdominal distension represents mostly a serious problem unless it is waxing and waning. Chronic persistent abdominal distension is caused by either fluid, organomegaly or tumor. Accompanying symptoms guide to a probable cause and physical examination can finetune the diagnosis with relevant specific tests. Non-specific abdominal USG findings such as lymph nodes, free fluid, gaseous distension of intestines, calcification in the liver or thickened urinary bladder need cautious clinical correlation.

MCQs

1. This surgical condition is not chronic

A) Appendicitis
B) Intestinal obstruction due to TB
C) Congenital megacolon
D) Obstructed hernia

2. This condition with abdominal distension does not present with pain

A) Inflammatory bowel disease
B) Intestinal TB

C) Storage disorder
D) Functional problem

3. Which of the following statement is WRONG?

A) Ascites may be due to liver disease
B) Ascites may not be due to liver disease
C) Ascites may occur without liver disease
D) None of the above

4. Paralytic ileus may result from

A) Acute diarrhea
B) Sepsis
C) Electrolyte disturbance
D) All of the above

5. Which test may be abnormal in abdominal distension due to functional disorder?

A) Stool microscopy
B) Abdominal USG
C) Colonoscopy
D) None of the above

Answers to MCQs

Q 1 D, Q 2 C, Q 3 D, Q 4 D, Q 5 B

2.21. My child suddenly became stiff!

Back to basics – seizure

What is seizure?

Brain is like an electrical network in which neurons (nerve cells) communicate with each other through electrical signals to generate multiple functions. Disturbance in this electrical circuit leads to a sudden abnormal event that is referred to as seizure. It is different than a convulsion. Convulsion is a seizure affecting motor cortex with tonic (stiffness) or clonic (jerky movements).

What goes wrong in electrical circuit?

Biochemically, it is the ionic imbalance that leads to either too much of excitatory transmission response or too little of inhibitory response and results in a seizure. Most often ionic balance is restored by nature itself and so seizure stops by itself within 1-2 minutes. Nature rarely fails, but if does happen then seizure continues for longer time and it would need a drug to stop it.

What is the difference between seizure and convulsion?

Convulsion is a type of seizure which manifests as jerking movements or stiffness of some parts of the body. However, seizure may present without such movements or stiffness because manifestations depend upon which part of the brain is responsible for sudden electrical discharge. Convulsion results when part of the brain that controls voluntary body movements (motor system) is involved. Involvement of other parts of the cerebral cortex do not manifest with convulsion but presents as sudden attack of emotional disturbances if temporal lobe is the site of abnormal electrical discharge or when parietal lobe is involved, result is sensory manifestations. Thus, seizure may not present with

convulsion though convulsion is most common type of a seizure and many times, both terms are used interchangeably, though not scientifically correct.

Types of seizures

They are classified into two major groups – generalized and localized (focal) depending on extent of involvement. Each group is further divided into motor (presents as movements - convulsion) and non-motor (without movements). Generlised seizure always present with loss of consciousness even for a brief period while localized seizure may or may not lose consciousness (focal with impaired awareness or focal aware). At times, localized seizure may spread to other parts and become generalized. Hence it is the onset of seizure that decides focal or generalized. At times it is difficult to classify seizure type as it may be mixed and also may change.

Presentation of seizure types

Gemeralised motor seizure may present as stiffness (tonic), jerky movements (clonic) or both (tonic-clonic), atonic (sudden loss of muscle strength) and myoclonic (muscle spasm). In tonic seizure, spasm of respiratory muscles leads to apnea and cyanosis. In-built neural mechanism breaks this phase, breathing starts and clonic phase begins. Generalised non-motor seizure (absence seizure) presents with very transient lapse of awareness when person stops momentarily and in few seconds. resume what he was doing. Localised (focal) motor seizure presents as abnormal movements of a part of the body with or without loss of consciousness. Focal

non-motor presents as emotional change without loss of consciousness.

What is epilepsy?

Unprovoked recurrent seizures are referred to as epilepsy. Unprovoked means seizure occurs without any apparent trigger factor. It may be idiopathic (cause not known), often genetic or at times secondary to previous brain damage.

Etiology of seizure

It is classified into acute symptomatic (due to present existing disease), remote symptomatic (due to brain damage resulting from previous disease) or idiopathic (cause not known).

Acute symptomatic seizure

Most common cause is simple febrile seizure. It occurs due to sudden rise in body temperature in a genetically susceptible child. Around 5% of normal children suffer from simple febrile seizure. First episode usually presents between age of 6 months and 2 years though such a seizure may recur even up to the age of 6 years. Seizure typically occurs during first 24 hours of onset of fever, lasts for a minute or less, is self-limiting and child becomes normal without any sequelae. There is no need for treatment though recurrence can be prevented by use of clonazepam. (2.5 mg twice a day for 3 doses orally) only during first 24-36 hours of onset of fever along with an antipyretic. (Though control of fever may not prevent a simple febrile seizure and at times, seizure may present before fever is noticed).

Intracranial infection may occur at any age and is accompanied with persistent fever, vomiting and change in

sensorium. Such a child needs hospitalization.

Hypocalcemia is another cause that may present in an infant or young toddler having active vitamin D deficiency rickets. This is a seizure that does not end with post-ictal (at the end of seizure) drowsiness. Neonate born with low birth weight may also suffer from hypocalcemia.

Remote symptomatic seizure It is a result of brain damage caused by previous illness that may have resolved completely though leaving behind epileptogenic focus. Obviously one clue to such remote symptomatic seizure is a child with developmental delay. Birth injury, significant serious head injury, severe brain infections and congenital brain malformations are common causes of remote symptomatic seizure. Treatment consists of using anti-epileptic drugs.

Idiopathic seizure It occurs in normal brain and may be triggered by subtle event such as minor illness. Often such a seizure may not recur. If recurs, it is termed as idiopathic epilepsy. Modern science may find specific cause for such idiopathic seizure or epilepsy in the form of genetic or biochemical defect. Hence, what has been referred to as idiopathic epilepsy is now being proved to be a genetic disorder.

Seizure mimics

There are several conditions that mimic a seizure and it is important to differentiate them. Such conditions include movement disorders (dystonia) or benign sleep myoclonus, sudden fall (syncope, vertigo), breath-holding spasm. Each of

these conditions have typical presentation that is different from a seizure. Most of them retain consciousness.

Clinical approach

Detailed history of the event is most important. It should be obtained from one who has actually observed the event. Ideally it could be captured on video that helps a lot. This is because onset of seizure is vital to understand nature of seizure. If onset is missed, vital information is lacking. Besides onset, duration, progress and accompanying symptoms offer a clue to probable diagnosis. Seizure preceded by fever may be simple febrile seizure or fever triggered hypocalcemic seizure (both of them recover quickly) or intracranial infection (that is accompanied with vomiting and change in sensorium). Seizure without fever may be remote symptomatic (abnormal brain function) or idiopathic (cause unkown).

Physical examination can detect abnormal neurological findings if any and whether these findings denote active disease or old disease. Normal neurological status suggests either simple febrile convulsion or hypocalcemia that leaves no sequelae. Child with active meningitis is sick besides neck rigidity and drowsiness whereas those with remote symptomatic seizures also have abnormal neurological findings but child is not sick and has delayed development.

Investigations

Probable diagnosis decides whether tests are required or not. Simple febrile seizure does not need any test, it is a clinical diagnosis based on characteristic presentation and exclusion

of other causes. Meningitis is proved by CSF examination, hyppcalcemia due to vitamin D deficiency by serum calcium, phosphorus and alkaline phosphatase and bone X-ray. Brain damage due to previous disease may need neuro-imaging. EEG is required only in case of epilepsy.

Management

Most often seizure stops on its own before any intervention. Only when patient presents during active seizure, one may use diazepam or midazolam to control the event. However, as one can't anticipate how long the seizure may last, it is best to attempt to control the seizure. Nasal puff of midazolam is easy to administer even by parents that aborts the attack quickly. Long term anti-convulsant drugs are not necessary except in case of epilepsy or when active symptomatic seizure has a risk of recurrence. Simple febrile seizure needs no treatment but recurrence can be prevented by clobazam given at the onset of fever along with antipyretic drug. Three doses at 12 hours interval suffice as chance of seizure is only in first 24 hours of onset of fever.

Take home message

Occurrence of a seizure is frightening to parents and a challenge to a doctor. However, only action doctor needs to undertake is to stop a seizure and a decision about the need of immediate hospitalization (meningitis, severe head injury or uncontrolled seizure) and subsequent need for investigations (in case of recurrent seizures). Most of the times, a seizure stops by itself but if patient presents with active seizure, diazepam or midazolam may be required.

MCQs

Q 1 Which of the following statement is wrong?

A) Seizure may not present with movements
B) Seizure may occur without loss of consciousness
C) Seizure is always preceded by some symptoms
D) Seizure may not recover by itself

Q 2 This type of generalized seizure is not associated with movements

A) Clonic
B) Tonic / Clonic
C) Myoclonic
D) Absence

Q 3 Which of the following statement is wrong

A) Focal motor seizure presents without loss of consciousness
B) Focal motor seizure presents with loss of consciousness
C(Focal non-motor seizure presents with loss of consciousness
D) None of the above

Q 4 These seizures end with sleepiness except

A) Simple febrile seizure
B) Seizure due to hypocalcemia
C) Seizure due to meningitis
D) Idiopathic epilepsy

Q 5 Transient loss of consciousness may be seen in this condition even when it is not a seizure

A) Vertigo
B) Breath holding spasm
C) Syncope
D) Benign sleep myoclonus

Answers to MCQs

Q 1 C, Q 2 D, Q 3 C, Q 4 B, Q 5 B

2.22. Seizure – must act fast!

Clinical application of basic concepts

Must find out - Is it a seizure or seizure-mimic? Is seizure, is it generalized or focal? If generalized, is it motor or non-motor? is it tonic, clonic, tonic-clonic, atonic or myoclonic? If focal, is it focal aware or focal with impaired awareness? Is it acute symptomatic seizure, remote symptomatic seizure or idiopathic seizure? Is it epilepsy, if so is it epileptic syndrome?

Seizure is a paroxysmal event that may mimic non-seizure disorder. Hence one must confirm an event to be a seizure before embarking on its cause. Most seizures are recognized by jerky movements of the limbs and transient loss of consciousness with staring spell. However, seizure also can present without loss of consciousness and it needs to be recognized. Movement disorders present with typical pattern without change in sensorium. Syncope occurs mostly in standing position and recovers in seconds. Breath-holding spasm it typically triggered by emotional stress as child holds

breath in expiration and may turn blue or pale. One must enquire about sequence of events from a witness and if possible, captured video record. It helps to define nature of the event. Feeling giddy or rotational movements may suggest vertigo. Once we know it is a seizure, next is to find out its cause. Preceding events, type of seizure and post-seizure events offer clue to diagnosis. Thus, detailed history is important. Age group helps to define common causes, simple febrile seizure is common in early childhood and so also hypocalcemia due to rickets. Idiopathic epilepsy is diagnosed in children > 5 years of age. Physical examination may be totally normal as in case of simple febrile convulsion, idiopathic epilepsy or hypocalcemia. Localising neurological signs may represent either recent or old brain lesion responsible for seizure. Thus, it is important to consider whether it is acute or remote symptomatic seizure.

Case-based discussion

Case 1

One year old infant presented with high fever followed in next 18 hours with a generalised seizure that lasted for a minute. Child remained bit dazed thereafter for few minutes and then recovered completely without any neurological sequalae. He had no prior illness. He had grown well and had achieved normal milestones. There was no family history of similar or any other neurological disorders.
This child has had normal brain development and has remained neurologically normal after the seizure that was self-limiting and triggered by high fever.
Physical examination showed no neurological abnormality

and cause of fever was considered to viral infection that settled within next two days without any specific therapy. It supported diagnosis of **simple febrile seizure.**
This child does not need any drug therapy and this being the first seizure, there is no need for prophylactic therapy. Parents were counselled about possible recurrence during first 24 hours of onset of fever. However, in case of recurrence, one may suggest midazolam nasal spray as an immediate rescue drug to stop a seizure. It can be used by parents at home and seizure stops immediately.
Simple febrile seizure never occurs beyond age of 5-6 years and has no relation to future epilepsy.
It is important not to prescribe an antibiotic for suspected simple febrile seizure as acute bacterial meningitis may be masked. If antibiotic is considered for whatever reason, CSF examination is a must before starting antibiotic.

Case 2

One year old infant presented with high fever followed in next 18 hours with generalised seizure that lasted for few minutes and stopped on its own. It was not followed by any post-ictal event in the form of drowsiness and he remained totally normal. However, within next two hours, he had another similar episode that lasted just for few seconds. He was fine thereafter. Fever disappeared over next two days. He had normal growth and development. There was no family history of similar disorder. On direct questioning, he was bottle fed and consumed about a liter of milk each day with very little solid food – he was bottle addicted.
This seizure did not result in post-ictal events and it is a clue

to probable diagnosis of hypocalcemic seizure. As he was mainly on milk diet, he had a risk of developing vitamin D deficiency as milk is poor source of vitamin D and this infant would not have been exposed to sunlight. So rickets may have contributed to hypocalcemia triggered by viral infection.Physical examination showed signs of rickets without any neurological abnormality. It supported diagnosis of **hypocalcemia** as a cause of seizure.

Vitamin D deficiency rickets can be confirmed by bone X-ray and high serum alkaline phosphatase with either low or normal serum calcium and low phosphorous level. Serum calcium may be normal in this child because nature moves available calcium from bone to blood and thereafter to tissues. However, it takes few hours to replenish calcium at tissue level and that is the reason that this child had another seizure though it was for shorter duration. But once calcium is replenished in neurological tissues, seizure stops.

If one saw this child during a seizure, IV calcium could have been administered for quick replenishment. However, if one saw this child after second seizure, one may consider oral replacement of calcium along with vitamin D. it is ideal to delay starting vitamin D supplement for few days before administration of oral calcium because vitamin D is likely to deposit calcium into bone from whatever is available in tissues and it may precipitate a seizure. Once rickets is fully cured, there isno risk of hypocalcemia.

Case 3

One year old infant presented with high fever followed by irritability and vomiting for two days. On D3, he developed a

generalised seizure that lasted for few minutes and was followed by drowsiness for few minutes. Fever and irritability continued with occasional vomiting.
This child has developed a seizure on D 3 of onset of fever and so this is certainly not a simple febrile seizure or fever triggered hypocalcemia. Irritability and vomiting suggest probable increased intracranial pressure. It favors diagnosis of bacterial meningitis. Initially the disease is localized only to meninges but subsequently cortex is affected if not treated in time and presents as seizure.
Physical examination showed highly febrile sick looking child. He was drowsy but when disturbed was irritable. He had meningeal sings in the form of neck stiffness. There were no localizing signs. Anterior fontanel was open and bulging. It favors the diagnosis of **bacterial meningitis**.
It was confirmed by CSF showing 400 cells 90% polymorphs, proteins 250 mg% and sugar 20 mg%. Culture showed pneumococcus sensitive to all antibiotics.This child was treated with IV Ceftriaxone and luckily recovered completely.
Older child with acute bacterial meningitis rarely presents with seizures as disease remains localized to meninges for a long period. On the other extreme of age, neonate presents with generalied seizure in acute bacterial meningitis as disease spreads quickly from meninges to brain cortex.Neuroimaging is justified in acute bacterial meningitis only in presence of focal signs that may suggest brain abscess or subdural empyema.

Case 4

Two years old child presented with high fever and drowsiness followed a day later with generalised seizure. On direct questioning, he was not keeping well over last two months with recurrent episodes of fever that were treated each time with antibiotic but without diagnosis and would get temporarily better. He had lost 2 kg of weight and felt sick. There were no other significant illnesses prior to onset of this problem two months ago. There was no significant family history and he denied contact with any contagious disease.
This child has subacute onset of disease with recurrent fever. It suggests continuous disease with recurrent illness as evident by his progressive loss of weight and sickness. So, it is same disease that suddenly developed high fever, drowsiness and seizure. Such a neurological manifestation came up suddenly that suggests immune reaction to existing infection and in the background of subacute disease. It favors probable diagnosis of TB meningitis. Unlike bacterial meningitis, this child has had brain cell affection early in the course of the disease – encephalopathy without much increased intracranial pressure.
Physical examination showed malnourished child, drowsy, mild neck stiffness, positive Macewan's sign – cracked pot sound suggestive of raised intracranial tension, left sided hemiparesis with left sided facial palsy with spasticity, brisk deep tendon reflexes and extensor plantar reflex.
Pathologically these signs represent meningeal inflammation with hydrocephalus, vasculitis affecting middle cerebral artery as evident by hemiparesis with facial palsy and brain cell edema as shown by drowsiness. These pathological findings are typical of TB meningitis.

CSF showed 120 cells, mostly lymphocytes, 120 mg% proteins and 35 mg% sugar. CT scan showed meningeal enhancement with hydrocephalus and brain edema. CSF culture for TB was negative and so also GeneXpert. Chest x-ray was normal. Even in absence of proof, **TB meningitis** was diagnosed on circumstantial evidence. This child was treated with anti-TB drugs and steroids and made a recovery but was left behind with brain damage.

Family screening for tuberculosis with a chest X-ray is ideal to suspect tuberculosis in young children as tuberculosis is a paucibacillary disease in children (bacilli are small in number) that makes confirmation difficult. This child presented in late stage of the disease with drowsiness and convulsion and hence was left with brain damage. In fact, at such a stage, disease is often fatal If picked up before developing any change in sensorium or convulsion, there could be complete recovery without damage. Thus, it is vital to diagnose tubercular meningitis in early stage for better outcome. Theoretically, fungal infection can also present in the same way but it is usually seen in immune-compromised patients.

Case 5

Two years old child presented with high fever and drowsiness that worsened over next 8 hours followed by generalised seizure that lasted for 10 minutes that came under control with drugs. He was apparently well prior to onset of present illness. He maintained normal growth and development. There was no significant past or family history or contact with any contagious disease.

This history denotes fast progression of neurological disease

with brain cell affection in the form of progressive brain cell edema with deepening consciousness and seizure. It could be viral encephalitis or autoimmune encephalopathy. Both conditions look similar with very small difference that may not be discernable. Autoimmune encephalitis generally precedes with febrile illness by at least 2-3 days with an interval of apparently normal period before onset of neurological disease. As this child did not have any recent illness, it may favor viral encephalitis.

Physical examination showed febrile child, unconscious responding to painful stimuli with no localizing signs. It suggests generalised brain cell edema of probably due to viral infection. However, any other cause such as toxic encephalitis or fever triggered metabolic encephalopathy cannot be ruled out.

Investigations – Hb 11 Gm%, WBC 4000 P 55 L 43 M 2 E 0 Pl 1.8 lakhs, malarial parasites not detected, CSF cells 80 mostly lymphocytes, 60mg% proteins, normal sugar, culture –ve, MRI scan of brain showed diffuse brain edema.

Diagnosis of **viral encephalitis** was made and treated with IV acyclovir –antiviral antibiotic mainly acting against herpes viral infection, though administered empirically. Child also received anti-edema measures. Including anti-convulsant drugs. He improved luckily though etiology could not be proved.

Diagnosis of viral encephalitis is often made without a final proof though modern technology can help to diagnose the type of viral infection. Not all viral infections can be treated with anti-viral drugs but herpes virus can be successfully treated only if therapy is started in first two days. Hence the

need for early definitive diagnosis. In absence of such a facility, empirical treatment with acyclovir is justified.

Case 6

Ten years old child presented with jerky movements of left upper limb that lasted for 5 minutes and stopped by itself. There was no loss of consciousness. He was well prior to onset of this event and remained normal thereafter as well. There was no significant past or family history.
This history suggests focal seizure (focal aware). It is not a movement disorder as it occurred suddenly and also stopped by itself. Cause of this seizure cannot be assessed on history, except a fact that there must be local lesion in motor cortex. Physical examination showed no abnormality. It meant an apparently silent lesion that must have manifested suddenly without any obvious trigger factor. Such a lesion may be any local structural malformation that must have existed since birth or it may have resulted from focal swelling that may have resulted from local inflammation or infection. But its exact cause cannot be guessed at this stage and needs investigations.
CT scan of brain showed ring enhancing lesion in right motor cortex that had surrounding edema suggestive of inflammatory origin and not a congenital malformation. Such a local inflammatory lesion is commonly due to chronic infection such as tuberculosis or neurocysticercosis – a parasitic infection that gets transmitted from consuming uncooked meat. CT scan often fails to differentiate between these two lesions. Based on presence of multiple lesions on CT scan, diagnosis of tuberculosis – **tuberculoma** was

made.Child was treated with standard anti-TB therapy with steroids. On follow-up, this child showed disappearance of lesion on repeat CT scan.

Diagnosis of tuberculoma is often circumstantial. If CT scan finds a scolex of the parasite, diagnosis of neurocysticercosis can be confirmed. In absence of such clues, spectroscopy may be useful to differentiate these two conditions. Without definitive diagnosis in such a situation, management is debatable. This child certainly needs anti-convulsant drug – carbamazepine is the drug of choice and one may decide to observe further course. However, there is always a risk of tuberculosis spreading to affect brain cell with dire consequences. On the other hand, treatment of neurocysticercosis is short for few weeks as against minimum 6 months for tuberculosis. So, decision should be taken after discussing with parents and explaining pros and cons of all options. If this child is purely vegetarian, chances of cysticercosis is very low unless he consumed vegetarian food contaminated by uncooked meat without knowing. Thus, many such patients end up with anti-TB treatment with steroids.

Case 7

Ten yeasr old child presented with sudden onset of generalised seizure that lasted for few minutes and stopped on its own. He was healthy child with normal brain development and had no symptoms prior to onset of this event. History suggests unprovoked generalised seizure. Physical examination was normal and so diagnosis of **probable idiopathic epilepsy** was considered though not

labelled at this stage as very definition of epilepsy is recurrent seizures. One may have to wait for further course. As it is the first episode, one may consider clinical follow-up without any tests. EEG at this stage may not be interpretive as normal EEG does not rule out epilepsy and abnormal EEG may be found in small number of normal children. There is no risk even if there was another similar seizure. Though one can be prepared with nasal midazolam spray to control seizure. Most of such seizures are short lasting and self-limiting without causing any brain damage.
EEG may help to rule out focal epilepsy with generalization as many times, no one else has observed onset of a seizure that may have been focal which spread to all the areas resulting in generalized seizure as the presentation. EEG was normal in this child, confirming it to be generalised seizure. CT scan of brain is not justified in this child. Parents were counselled and no treatment was advised.

Case 8

Thirteen years old child suddenly fell down while standing for morning prayers in school. He was well till this event happened. He had arrived at school as usual after normal breakfast, No one had noticed onset of this event as it was only when he fell down that everyone around him noticed. By the time his friends offered help, he had got up, was bit dazed for a few minutes but became normal quickly. One cannot be sure whether this was a seizure that made him fall down or whether it was pseudo-seizure – syncope. However, fact that he got up within few seconds would go in favor of syncope. Though difference between the two is evident only

if one knows the onset of this event.
Physical examination was normal as expected and so diagnosis of **syncope** was considered.
As this event occurred without any known provocation, it may be a result of neurogenic syncope – vasovagal attack due to unknown stress, such as fear, starving or lack of sleep, the event that typically occurs in standing position. It is a clinical diagnosis and does not justify any tests such as tilt-table test where sudden tilt of position may induce the attack. However, it such an event occurred while exercising or during sport activities, one may have to take a serious note of it as it may represent cardiogenic syncope. It may be fatal if not diagnosed properly unlike vasovagal attack. If cardiogenic syncope is suspected, one must order ECG and echo-cardiogram to pick up any functional or structural abnormality – one must rule out long QT – it is often familial and so history of sudden death in the family should arouse suspicion of cardiogenic syncope.

Take home message

Seizure must be differentiated from seizure-mimic (pseudo-seizure). Detailed history from an observer helps to confirm a seizure. Seizure may manifest without a convulsion. Non-convulsive seizure is not easy to pick-up. What presents as a generalized seizure may have a focal origin that is easily missed unless there is an observer around when a child develops a seizure. As drug of choice is different, in case of doubt, EEG helps to rule out a focal seizure. Acute symptomatic seizure is triggered by an active disease such as an intracranial infection, metabolic disorder or severe head

injury.Remote symptomatic seizure results from pre-existing brain damage and idiopathic seizure is unprovoked, cause is not evident but may be genetic in origin. Most often, a seizure stops by itself but brain damage is likely if it continues for more than five minutes. Hence it is ideal to control a seizure by midazolam, if possible. Parents or care-takers can be trained to administer the drug via a nasal puff that is easily available.

MCQs

1. Which of the following statement related to simple febrile seizure is WRONG?

A) It may occur with any degree of fever
B) It may occur anytime up to 6 years of age
C) It may recur many times
D) None of the above

2. This seizure is not accompanied with any post-ictal drowsiness

A) Simple febrile seizure
B) Idiopathic epilepsy
C) Hypocalcemia
D) TB meningitis

3. This seizure is preceded by fever and irritability

A) Simple febrile seizure
B) Bacterial meningitis
C) TB meningitis
D) Fever triggered hypocalcemia

4. Which of the following seizure needs EEG?

A) Simple febrile seizure
B) TB meningitis
C) Focal seizure
D) Hypocalcemia

5. Which of the following statement related to neurogenic syncope is TRUE?

A) It may occur in any position
B) It may occur at any age
C) It recovers within seconds
D) It may end with drowsiness

Answers to MCQs

Q 1 D, Q 2 C, Q 3 B, Q 4 C, Q 5 C

2.23. Headache - an invisible problem

Back to basics

Pathogenesis of pain in general

Most commonly, pain is a result of inflammation that is accompanied with swelling, warmth and redness besides pain or tenderness – not all the features are always present. However non-inflammatory types of pain may be neurogenic, vascular, psychogenic or referred. They are not accompanied with symptoms and signs of inflammation and so present without swelling, warmth or redness. Neurogenic and vasogenic type of pain is localized to their respective areas of supply. Psychogenic pain is not due to any physical disease

but patient does feel it genuinely. It is a pain of convenience that does not disturb health, sleep or activities of choice. Pain at one site may originate from the other site because these two sites share same nerve connections – it is a referred pain. So, one may have to be careful to assess origin of pain that may have either no signs or hidden signs.

What is headache?

Pain or discomfort arising from any part inside or around the head (skull) is referred to as headache. What makes it worrisome is the fact that it is mostly invisible to an outsider including a doctor as it is not accompanied with swelling. And if there is a swelling, it needs to be carefully detected as it is at obscure places. It poses a challenge though luckily most causes of headache are not serious. It is important to note that infants and toddlers up to the age of around 4 years are not able to localize pain and they present with excessive crying or irritability, depending on severity of pain. In general, headache is a complaint of older children and adults.

Brain does not feel pain

Brain cells are devoid of sensory nerve supply and so also lung parenchyma. These are the only two structures in the body without sensory nerve endings. Though brain does help to perceive and localize the pain. Pain is carried by sensory nerve endings from the site of affection up to thalamus – the receiving center that in turn relays the information to somatosensory cortex. It is the cortex that would take appropriate action to guard against pain.

How does intracranial pathology cause pain?

It is the surrounding structures in the intracranial cavity that have sensory nerve endings and pain arises from them and not the brain cell. Thus, meningeal irritation or inflammation leads to headache and so also due to increased intracranial pressure as happens in progressive hydrocephalus or brain tumor,

Other areas in or around skull that cause headache

Temporomandibular joint, ear, paranasal sinuses and teeth are parts included in skull that may cause a person to report headache. Eye resides in the socket in the skull and so eye strain can be mistaken for headache. Though it is not strictly pain but just discomfort due to strain of extra-ocular muscles. it is common to ascribe headache to refractive error but that is not true. Cervical neck pathology with muscle spasm and also tightness of facial muscles (tension headache) lead to headache. It may also be caused by temporomandibular joint or dental pathology. Pain may also arise from the mind itself – psychogenic headache.

Causes of headache

Acute onset of high fever is associated with headache that can be severe in dengue, influenza or other viral infections as well as in Typhoid and malaria. In such cases, headache is temporarily relieved with reduction of fever though may increase again with rising fever. This is important as it rules out intracranial infection such as meningitis that also starts with high fever followed by headache that continues even if

fever is temporarily controlled. So, always enquire about relation between fever and headache. Obviously, there are no neurological signs in non-neurological febrile patients with headache, though subtle signs during initial stage of meningitis may be easily missed.

Intracranial infection such as bacterial meningitis must be ruled out in case of fever with headache. Typically, such a child presents also with irritability and vomiting, headache persists in spite of temporary reduction of fever. This fact must be assessed carefully so that diagnosis of meningitis can be suspected even before clinical signs appear such as neck stiffness and other signs of meningeal irritation.

Acute sinusitis is not common in children and chronic sinusitis is difficult to diagnose but certainly is a cause of headache. It may be suspected in case of persistent nasal discharge in spite of proper treatment. Maxillary sinuses appear by the age of 3 years and frontal sinuses by 7 years. Ethmoidal and sphenoidal sinuses are present at birth but rarely involved. Child under the age of 4-5 years, cannot localize pain and hence sinusitis is considered only in older children. Migraine is another common cause of headache in older children and is suspected by recurrent attacks of headache with strong family history.

Intracranial space occupying lesions is always at the back of mind especially when headache is progressive and often presents on getting up in the morning with natural relief thereafter through the day in early stage of the disease. Thereafter ofcourse, headache is continuous and accompanied with vomiting.

Hypertension is another cause of headache as it contributes to increased intracranial pressure.
Trauma as a result of head injury may be superficial in the scalp or deep intracranially. Blood-shot eyes or bleeding through nose or ear are warning signs of brain injury, besides change in behavior and vomiting. However, vomiting occurring immediately after head injury is due to fright and crying and not indicative of brain injury. Superficial injury presents with swelling on the scalp and may not be visible but palpable.
Tension headache is due to tightness of facial muscles as a result of inability to relax as may happen due to stress or anxiety.
Psychogenic headache is pain of convenience as it disappears completely during happy hours and does not anyway affect health and well-being.
Rarely, TM joint, ear or dental pathology may also present with headache and easily overlooked.

Clinical approach to headache

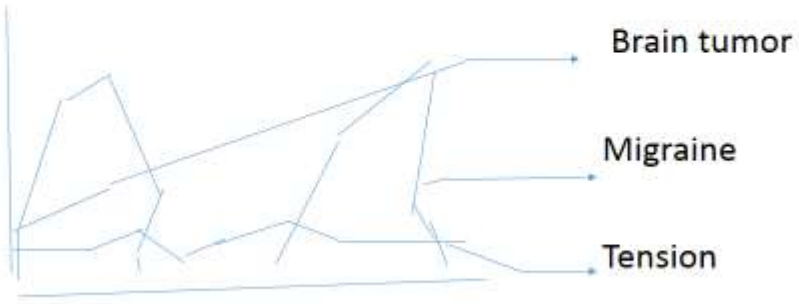
Site and type of headache may offer some clues. Patient often complains of bitemporal headache while localized headache may be due to disease at underlying site. Dull continuous pain is more common though throbbing pain may be of vascular origin.
Onset – fever associated with headache is easy to differentiate between presences or absence of intracranial infection as mentioned above by simple question – does fever and headache are relieved temporarily with antipyretic

or whether headache continues in spite of fever control. Of course, headache in intracranial infection is preceded by irritability and vomiting. Headache without associated fever may be due to migraine that is often recurrent with positive family history and also due to hypertension, often secondary to renal disease.

Duration and progress – short duration headache with fever disappears once disease is controlled. Migraine typically lasts for some hours and settles by itself to recur again after few days. At the height of headache, vomiting along with intolerance to sound and light are other manifestations before headache resolves by itself or after an analgesic. Slowly progressive headache over few days to weeks often accompanied with vomiting suggests probable brain tumor that may begin with morning headache on getting up and relieved after an hour or so. This is because intracranial pressure is temporarily increased to a small extent in normal persons due to venous congestion during sleep. In normal individual, such an increase in intracranial pressure is small enough not to manifest but in presence of developing brain tumor, small increase of intracranial pressure in the morning is good enough to manifest headache only to be relieved on its own as venous congestion disappears with increasing activity. However once brain tumor enlarges, it would manifest with headache for increasing periods and not restricted only to mornings.

Tension headache is also chronic but at steady level with mild exacerbations and remissions. Psychogenic headache is of convenience and does not disturb sleep or play and growth

and well-being is well maintained.
Physical examination reveals abnormal neurological findings in case of meningitis and brain tumor. In other conditions causing headache, there are no clinical abnormalities except in case of hypertension and subtle signs of chronic sinusitis



For simplification, chronic headache progression is represented by the graph above that easily can differentiate between brain tumor, migraine and tension headache. Of course, brain tumor would also present with neurological signs. It is ideal to examine other areas around skull – TM joint, teeth / gums and ear as well as neck.

Investigations

Acute bacterial meningitis is confirmed by CSF showing high number of WBCs – mostly neutrophils and high proteins with low sugar and culture can detect organism responsible for meningitis, provided test is ordered prior to starting an antibiotics. Antigen can still be detected in patients who are

already on antibiotics.
Neuroimaging – CT scan is diagnostic of brain tumor though etiology needs to be assessed. Brain tumors are also caused by infections such as tuberculosis or cysticercosis besides benign and malignant diseases. Etiology can be assessed only by biopsy or usually diagnosis is considered with circumstantial evidence in case of tuberculosis or neurocysticercosis, supported by spectroscopy.
X-ray of paranasal sinuses has low sensitivity to demonstrate changes in sinusitis and ideally CT scan is necessary. However, in view of radiation exposure, diagnosis of chronic sinusitis can be made on clinical basis and treated.
Diagnosis of migraine and tension headache is entirely clinical and it is important to rule out other conditions in case of doubt.

Management

Analgesics are mainstay of symptomatic treatment. Besides, several drugs are tried in Migraine but there is no single drug therapy that can be strongly recommended. Flunarizine, cyproheptadine and beta-blockers are tried with variable success and are reserved for those patients who suffer frequent severe attacks difficult to alleviate with analgesics. One should search for offending trigger factors that also vary in individual patients such as coffee, nuts or sleep deprivation. Infections are treated by specific antibiotics.

Take home message

Headache presenting with fever does not pose a diagnostic challenge and so also chronic progressive headache due to

intracranial space-occupying lesion. However chronic persistent or recurrent headache needs proper evaluation. Chronic sinusitis presents without fever and is easily overlooked, though persistent nasal purulent discharge and generalized vague ill-health offer clue to the diagnosis of chronic sinusitis. Tension headache and psychogenic headache are not easy to differentiate from one another. Migraine may present with atypical pattern with transient paresis. It is ideal to seek help of a specialist in such cases.

MCQs

1. Localised headache may be a feature of

A) Meningitis
B) Brain tumor
C) Migraine
D) Tension headache

2. Headache should not be ascribed to this organ

A) Teeth
B) TM joint
C) Neck
D) Eyes

3. Which of the following statements related to sinusitis is RIGHT?

A) Diagnosis is based on history of persistent nasal discharge > 2 weeks
B) Sinusitis occurs only in older children
C) Localised tenderness is not easy to demonstrate
D) All of the above

4. Headache due to severe head injury is often caused by

A) Subarachnoid hemorrhage
B) Subdural hemorrhage
C) Extradural hemorrhage
D) All of the above

5. Hypertension in children may present as headache and may result from

A) Renal disease
B) Adrenal disease
C) Vascular disease
D) All of the above

Answers to MCQs

Q 1 C, Q 2 D, Q 3 D, Q 4 B, Q 5 D

2.24. Headache – think beyond the brain!

Clinical application of basic concepts

Onset, duration and progress of headache in addition to site and type offer enough clues to come to a reasonable diagnosis. Accompanying symptoms add to make correct decisions. Acute onset headache is usually due to infection or migraine and rarely due to acute onset of hypertension as in case of acute glomerulonephritis. Headache due to meningitis starts with fever and irritability before headache and vomiting appear and headache continues in spite of temporary control of fever with paracetamol. Headache accompanied by general extra-cranial infections such as

influenza, malaria, typhoid etc are related to high fever and so headache disappears along with fever only to recur again as fever comes up. Chronic headache is usually due to tension headache or psychological – pain of convenience but also may be due to chronic sinusitis that is difficult to diagnose. Seen only in older children after frontal sinuses appear by 7-8 years of age and persistent nasal discharge in spite of usual treatment is the clue to diagnose chronic sinusitis. Recurrent headache is classically an attack of migraine with intervening period absolutely normal. It is the progressive headache and headache associated with vomiting that needs proper evaluation. Headache due to brain tumor starts often as morning headache on waking up that gets better by itself within next couple hours and rest of the day goes fine. This is the beginning of the problem that may get worst over time. It is important to recognize it in time to take a proper action. It is easy to note that investigations are often not required to find out the cause of headache and so CT scan should be reserved only for suspected raised intracranial pressure and should not be considered just to allay anxiety keeping in mind radiation exposure. There is no clarity about safe limit of radiation exposure in children and it is best avoided if possible.

Case 1

6 years old child presented with high fever and headache since last two days. There were no other symptoms. He was normal prior to onset of this illness and had no significant past illnesses. Another sibling had similar illness.

Physical examination showed no localizing signs and child did look better when fever was temporarily controlled with paracetamol. It favors diagnosis of **viral infection.**
Child got well within next two days without specific therapy. As fever and headache appeared together on very first day of illness, meningitis is unlikely. One may decide to observe further trend of fever as it may resolve by itself if it is viral infection or it may get worse. Specific investigations may be necessary to diagnose cause of fever but not for headache.

Case 2

Six years old child presented with high fever for two days followed by irritability, headache and vomiting over next 24 hours. He was reported to be sick with poor intake of fluids and was lethargic. Irritability followed by headache and vomiting two days after onset of fever clearly suggests intracranial infection. Physical examination showed sick looking child, lethargic with mild neck stiffness but no other localizing neurological signs. This suggests bacterial meningitis. CSF showed 130 cells most of them neutrophils with 100 mg of protein and 35 mg of sugar – consistent with diagnosis of **bacterial meningitis**. CSF culture was negative. Child was treated with IV ceftriaxone and recovered completely without any sequalae.
First two days of fever denote bacteremic phase and thereafter as bacteria settle in meninges, localizing symptoms appear such as irritability, headache and vomiting. Other disease such as viral meningitis (often termed as aseptic meningitis) may present in similar way though child is

not sick and shows improving trend over two days. CSF examination is a must to prove diagnosis of meningitis and CSF culture often would identify bacteria causing meningitis. In absence of positive CSF culture, one should choose antibiotic based on age-related epidemiology.

Case 3

Eight years old child presented with puffiness of eyelids and reduced amount of urine that was high colored over last 24 hours followed by severe headache. There was no fever. Two weeks ago, he had suffered from throat infection that was treated with antibiotics and got well.

Puffiness of eyelids and oliguria with probable hematuria indicates acute onset of glomerulonephritis. Headache following this episode is likely to be due to acute onset hypertension – accompaniment of glomerulonephritis.

Physical examination revealed puffy eyelids, mild edema of feet and blood pressure of 160 / 100 mm Hg. There were no neurological or cardiac findings – so there were no other complications of hypertension. Fundus examination was also normal.Urinalysis showed RBCs, granular casts and proteins, serum creatinine and urea were raised. This confirmed diagnosis of **acute glomerulonephritis**. Child was treated with anti-hypertensive drug and other symptomatic measures and he improved over next two days

Mostly, such a disease runs a favorable course especially if it has followed streptococcal throat infection. However other infections also cause similar illness that may not share good prognosis. Hence it is ideal to order serum C3 level that is low

at the peak of illness and gets back to normal as disease improves. Persistence of low C3 level after 2-3 months of apparent cure suggests incomplete improvement and needs referral to a specialist. Thus, serum C3 level is important to monitor every patient of acute glomerulonephritis

Case 4

Ten years old child presented with acute onset of severe throbbing headache that got worse over next few hours to an extent that he could not tolerate light or sound. He vomited twice at the peak of headache. He did not find much relief with analgesic but headache gradually subsided and he was back to normal. He never had headache in the past though his mother often had episodes of severe headache for which she always took analgesics as all investigations ordered by her doctor were negative.

This history is classical of **migraine**with positive family history. It is a bed-side diagnosis but based on classical history of an episode and supported by family history. Physical examination was completely normal.

Investigations were not ordered as there was no suspicion of any other disease and analgesic was prescribed on sos basis. Patient suffering from migraine should be advised to watch for any obvious trigger factors that could be avoided to prevent an attack. Though it is not easy to pinpoint to trigger factors. It is important to counsel about possible recurrences and symptomatic management with analgesics. Frequent attacks may justify trial with some drugs like flunarizine or cyproheptadine or betablocker given for few months to

assess if they would help. It is important to note that migraine can present in different ways such as transient paresis or severe abdominal pain. Diagnosis of migraine in such presentations is difficult and all other conditions need to be ruled out before labelling it. Migraine is considered to be of vascular origin and hence such a bizarre presentation. But in all such presentations, a common factor is self-limiting episode within few hours with complete recovery but often recurrent.

Case 5

Ten years old child presented with low grade fever and running nose with mild to moderate headache off and on for two weeks. He reported poor appetite and had lost 1 kg weight over last two weeks. There was no vomiting or lethargy. He always suffered from repeated cold and cough at times with fever and he was diagnoses as allergic rhinitis with at times secondary infection needing antibiotic therapy. He was not on any long-term treatment for allergy. Superficially to look at, low grade fever with loss of appetite and weight may suggest probable tuberculosis but cold has been a predominant symptom that is against tuberculosis. Headache in this child is another clue that is obviously not a neurological symptom as it is not accompanied with lethargy or vomiting. Thus, this looks to be chronic sinusitis. Physical examination showed congested nasal mucosa with mucopurulent discharge in a chronically sick looking child. Tenderness over frontal or maxillary sinuses could not be demonstrated as such finding is often difficult to assess,

especially it being chronic disease. In acute sinusitis, it could be possible to detect sinus tenderness. Diagnosis of **chronic sinusitis** is again based on interpretation of history. This child was treated with a course of antibiotics for 10 days along with long term therapy for allergic rhinitis in the form of intranasal steroids.

Routine investigations are not helpful to prove the diagnosis of chronic sinusitis. CBC, ESR and x-ray of paranasal sinuses are mostly inconclusive. CT scan of paranasal sinuses may prove the diagnosis though radiation exposure may be a consideration and so one can depend on bed-side clinical diagnosis unless patient does not improve as expected. In such a case, further management is best left to a specialist.

Case 6

Eight years old child presented with headache every morning on waking up that lasted for an hour but over next few days, headache was increasing in duration as well as intensity though getting better with analgesic. He also had occasional vomiting along with headache on some days. He was advised to take an analgesic as and when necessary, considering probability of migraine as his mother suffered from migraine. And it was ascribed to his habit of going to be pretty late at night. However, when headache started worsening, he was advised for further tests.

This symptom is classical of gradually increasing intracranial pressure, due to space-occupying lesion, either tumor or hydrocephalus. Physical examination showed mild intentional tremors. This findings in the background of history of

gradually worsening headache suggest **cerebellar tumor.** CT scan confirmed presence of tumor and luckily it turned out to be benign mass that could be removed.
Supratentorial brain tumor presents with localizing symptom such as a focal seizure in initial stage, much before it can lead to headache due to increased intracranial pressure while infratentorial tumor presents with raised intracranial pressure and headache along with many other symptoms because infratentorial area is a small space crowded by multiple structures.

Case 7

Twelve years old child complained of low-grade headache off and on for last two months. It was nearly constant though severity fluctuated to some extent but it was never severe. There were no other accompanying symptoms like vomiting or fever. There were hardly any days without headache. It disturbed the child and parents as no cause was found even after investigations including CT scan of brain.
This symptom of chronic headache has never been severe or worsening over weeks and without any other symptoms. Physical examination was normal. This is typical of **tension headache.**
It is due to tense muscles of face and neck, often a result of stress and inability to relax. Tension headache is a diagnosis of exclusion and based on history analysis and normal physical examination as well as absence of any impact on general health. There is no need for any investigations though most patients have had even CT scan – just to make

sure that one is not missing anything.
There is no drug treatment though analgesic may relieve headache for a wihile but frequent use should be avoided as all analgesics produce side effects especially on kidneys. Prolonged use of analgesics is the most common cause of chronic renal failure in adults. Counselling is an important aspect of management and parents must participate in destressing the child in every way possible.

Case 8

Twelve years old child presented with headache off and on for last four months. It was sporadic, at times severe but without any other symptoms. His health status was well maintained and so also his food intake, play activities and sleep. On detailed analysis of history, there was no clue to causation of headache .
Physical examination was completely normal.
There was no need for any tests though they already had done CT scan of brain that ruled out any brain pathology.
Parents were told to document timing, duration and severity of headache episodes as well as aggravating and relieving factors if any and assured that there was no major cause to worry. It was clear that headache never came in the way of those activities but headache surfaced when he was made to undertake any act that he hated to do so. So, this was headache of convenience – **psychogenic headache.**
Parentswere counselled. Placebo was prescribed and child improved over time.
It is worth noting that headache is genuinely felt by such a

patient though there is no organic cause and this is not malingering. Diagnosis is made with exclusion of other causes and only in case of suspicion, neuro-imaging may be necessary though routinely should be avoided for the fear of radiation exposure. Counselling is an important part of management and help of an expert may be necessary.

Take home message

Headache is a common symptom in general population including older children. Even when benign conditions are amongst the most common causes, patients always worry about headache if it is severe, recurrent or long-lasting. Refractive error is commonly blamed for headache though it is not true but most patients undergo eye check and many opt for CT scan to allay their anxiety. Detailed history and physical examination almost always can define probable cause and investigations should be reserved only for few conditions. However treating benign persistent or recurrent headache such as tension headache, psychogenic headache or migraine is often a challenge and need a referral to a specialist.

MCQs

1. This symptom can differentiate headache caused by meningitis from extra-cranial infections such as viral infection

A) Degree of fever
B) Occasional vomiting
C) Irritability
D) Interferbile period

2. Which of the following statements related to neck stiffness are RIGHT?

A) It suggests meningitis
B) It may not suggest meningitis
C) It may be seen in typhoid fever
D) All of the above

3. Which of the following statement related to cause of hypertension is WRONG?

A) Renal glomerular disease
B) Renal tubular disease
C(Coarctation of aorta
D) Adrenal disease

4. Chronic sinusitis in children is best suspected by

A) Tenderness over sinuses
B) High fever
C) Persistence of mucopurulent nasal discharge > 2 weeks
D) None of the above

5. Migraine can present as

A) Headache
B) Abdominal pain
C) Transient brain stroke
D) All of the above

Answers to MCQs

Q 1 D, Q 2 D, Q 3 B, Q 4 C, Q 5 D

2.25. My legs ache!

Back to basics

Introduction

Leg ache is the most common complaint of localized limb pain. It is so because we use legs more than upper limbs and hence leg ache may be a part of generalised body pain though maximally felt in legs. However, pain may arise from any extremity. There are multiple layers of tissues in a limb and pain may arise from any of these structures.

Pathogenesis of pain – a revision

Pain is commonly a result of inflammation but also may be vasogenic, neurogenic, psychogenic or even referred to a distant site other than the site of a disease. Inflammation is easy to make out as it is accompanied with swelling, redness, tenderness and warmth besides pain. Vasogenic pain in the limb may arise from affection of draining veins. Neurogenic limb pain may be shooting or burning. Both vasogenic and neurogenic pain are localized to the area of supply. Tired muscles accumulate metabolites that cause pain while sustained contraction of muscles cause severe cramps.

Anatomical parts of a limb

It consists of skin, soft tissue, muscles, bones, tendons, ligaments, joints, blood vessels and nerves. Obviously, pain may arise from any of these parts or may involve more than one part. It is important to define anatomy of the disease as pathology and etiology are different in affection of each part.

Common causes of pain in limbs

Trauma – impacted foreign body or splinter, injection site, sports injuries to muscle, tendon, bone or joint
Infection – bacterial cellulitis, abscess, osteomyelitis and arthritis, as well as myositis, viral myositis, syphilis
Non-infective inflammation – rheumatic fever, Juvenile idiopathic arthritis and other rheumatological disorders, dermatomyositis, bone malignancy
Vitamin deficiency – rickets, osteomalacia, scurvy
Vascular – vasculitis, sickle cell disease, deep vein thrombosis, aseptic necrosis of bone, osteochondrosis
Hematological – leukemia, hemophilia
Neurogenic – herpes, peripheral neuritis
Idiopathic – growing pain, restless leg syndrome
Psychogenic or functional – pain amplification syndrome
Referred pain – hip joint pain from psoas abscess

Clinical approach to pain in limbs

History

Age group – children are at risk of injuries. Young infant present with accidental birth injury or one caused by vigorous massage or pulled elbow while lifting the child. Children under the age of 4-5 years cannot localize pain and may be difficult to assess.
Localised or generalised – trauma, bacterial infection, herpes, arthritis are causes of local pain while viral infection with myositis, vitamin C and D deficiency, leukemia are conditions that result in more wide spread pain in all the limbs.
Origin – Acute onset is typical of trauma or at times vascular

or neurogenic pathology. Infection and inflammation are never very acute, they manifest over 2-3 days or even longer.
Duration and progress – traumatic and infective conditions are usually short lasing as they get diagnosed and treated early. Other conditions may have prolonged course. Most conditions result in continuous pain but growing pain manifests in later part of evening. Hemophilia may cause recurrent arthritis.
Relation to rest and activity – morning stiffness or pain is typical of inflammatory arthritis while pain in the evening after daily activity suggests degenerative arthritis (not seen in children). Arthritis may present with arthralgia in initial stages and so pain may be the only factor. Similarly, growing pain manifests after day's activity and is relieved by massage and rest. (Inflammatory pain is aggravated by touching or pressing but if it is relieved by massage, it denotes pain arising from tired muscles as in case of growing pain. Pain caused by muscle cramps is relieved by trying to relax the affected muscle by suitably changing position.
Degree and type of pain – severe pain suggests severe inflammation as in fracture or acute cellulitis or osteomyelitis. (In fact, in children, cellulitis should be viewed as probable osteomyelitis). Burning pain is characteristic of herpes zoster while dull ache may suggest mild inflammation or non-inflammatory disorders. Shooting pain down the leg may suggest a pinched nerve due to a vertebral disease.
Accompanying symptoms – swelling suggests inflammatory disease – either trauma, infection or non-infective disorders. Fever indicates wide spread disease as viral myositis or

severe localized inflammation as in osteomyelitis or abscess. Significant pallor or purpura denote hematological disorder.
Past history – similar illness is seen in few rheumatological disorders and hemophilia
Family history – may guide to genetic or familial disorders such as hemophilia, sickle cell disease or some of the rheumatological disorders.

Physical examination

Sick or not sick – Child is sick looking in cases of Infection and non-infective inflammation while trauma, hemophilia, growing pain etc do not present with significant sick look.
Local swelling denotes local inflammation. Generalised pain is not accompanied with swelling except in polyarticular disease. Localised bony swelling is seen in osterochondrosis and also in bone tumors.
Local warmth and tenderness signifies severe inflammation as in case of septic arthritis or osteomyelitis.
Restriction of movement – Severe painful conditions do not permit any movement of affected limb. Pain due to periarticular disease is evident on active movement of the affected joint but not on passive movement. It differentiates articular from periarticular disease. Pain in arthritis is aggravated by active as well as passive movement.
Localised muscle wasting signifies chronic pathology as seen in wasting of quadriceps in chronic arthritis of knee. Similarly, deformities and contractures are seen in chronic arthritis.
Hematological signs such as pallor or purpura denotes probable leukemia. It often presents as vague limb pain but

sickness and other signs including hepatosplenomegly and lymphadenopathy would suggest correct diagnosis. It is often missed as rheumatological disorder and wrongly prescribed steroids – it is a fatal mistake. In fact, steroids are rarely necessary in management of limb pain. Bleeding gums suggest scurvy though it may also be a manifestation of other hematological disorders such as leukemia. However, scurvy presents as pseudoparalysis (severe pain resembling paralysis).

Cardiac signs may be seen in rheumatic fever and at times in other rheumatolgical disorders.

Absence of any abnormal signs may suggest diagnosis of growing pain, pain amplification syndrome or functional pain.

Investigations

CBC helps in suspected acute osteomyelitis or arthritis as well as hematological disorders such as leukemia or sickle cell disease. ESR / CRP detects degree of inflammation but not the cause. Suspected rheumatological diseases need specific tests and it is not rational to ask for ANA (it is specific for SLE with positive dsDNA) or RA factor (positive in older female child with polyarticular disease) in every case of arthritis. Such tests are better reserved for specialists. X-ray detects signs of rickets, scurvy and also bone injury.

Management

Specific treatment depends on final diagnosis. However symptomatic treatment with analgesic may be tried till then. Neurogenic pain such as in case of herpes zoster is not easy to control with analgesics. Steroids should never be used in

undiagnosed limb pain lest leukemia is missed with dire consequences of poor outcome.

Take home message

Lower limb pain is a common complaint in most benign conditions such as growing pain or one caused by hypermobility of joints or restless leg syndrome but they pose a challenge in treatment. Diseases presenting with pain can be simply treatable as in case of vitamin D or C deficiency. Limb pain can be a part of generalized body pain as in case of viral myositis that is self-limiting. However, limb pain may be an initial symptom of a serious disease such as leukemia or chronic persisting rhematological disorder. In these cases, steroids should never be used without proper diagnosis and referral to a specialist is ideal.

MCQs

1. This condition presents with pain without swelling

A) Neurogenic pain
B) Growing pain
C) Functional pain
D) All of the above

2. Infection causing limb pain is often associated with swelling but not in this condition

A) Cellulitis
B) Herpes zoster
C) Syphilis
D) osteomyelitis

3. This rheumatological disorder presents with severe pain with minimal or no swelling

A) Juvenile idiopathic arthritis
B) Vasculitis
C) Rheumatic fever with arthritis
D) Juvenile dermatomyositis

4. Pain associated with bleeding disorder is seen in

A) Leukemia
B) Scurvy
C) Vasculitis
D) All of the above

5. Which of the following statement is RIGHT?

A) Pain and swelling are severe in septic arthritis
B) Swelling is much more than pain in juvenile idiopathic arthritis
C) Pain is more than swelling in juvenile dermatomyositis
D) All of the above

Answers to MCQs

Q 1 D, Q 2 B, Q 3 C, Q 4 D, Q 5 D

2.26. Leg pain – may be a sinister symptom!

Clinical application of basic concepts

Pain in both lower limbs represents a generalized disease while pain in one limb may be due to local pathology. Pain without swelling denotes non-inflammatory disease

.Generalised pain in the limbs with fever may be due to myalgia caused by viral infection, malaria, leptospirosis or typhoid fever or due to bony pain as in leukemia. Pain as the only symptom without swelling or fever may be due to physical as well as functional disorders that pose a diagnostic challenge. Etiology is not known in few of these conditions such as growing pains, fibromyalgia or restless leg syndrome. Non-organic functional disorder is diagnosed by analysis of detailed history but only after ruling out other conditions.

Case 1

Six years old child presented with generalised limb pain and headache for a day followed by high fever. There were no other symptoms. He was temporarily better for few hours after paracetamol but pain and fever would recur. Three days later, he became well without any specific treatment. Physical examination showed no localizing signs and as child got better without any specific treatment, it was labelled as **viral infection with myalgia.**

There is no need for any investigations in such a case

On first day when pain in limbs is the only symptom, one may not be sure of further evolution of the disease though it is often a forerunner of fever. One should wait for fever to appear in such situations. Once fever comes up, one is still not sure about the cause of fever. However, if the child looks better during inter-febrile period, it is mostly a viral infection and one would expect quick improvement.

Case 2

Six years old child presented with fever and severe leg ache along with headache for last two days. Pain was so severe that he could not walk and had to be lifted to reach the doctor. There were no other symptoms. On detailed questioning, this boy had waded through knee-deep water during heavy rains few days prior to onset of this illness. Physical examination showed severe congestion in conjunctiva – almost red eye – characteristic of leptospirosis. Diagnosis of **leptospirosis** was confirmed by presence of IgM leptospira antibodies. It is important to order liver and renal function tests as these two organs are often involved in immune phase of the disease and one may find biochemical abnormality before symptoms may ensue.

He was treated with doxycycline and recovered completely. Leptospirosis presents with symptoms similar to many other infections. Severe myalgia with conjunctival congestion is also common to many diseases. This case illustrates the fact that history taking is an art of asking questions based on rational thinking and if history of wading through knee-deep water is missed, one may not consider the diagnosis of leptospirosis. Such a history is unlikely to come forth from parents. Etiology of febrile illness during rainy season should consider specific diseases such as malaria, typhoid besides common viral infections and leptospirosis.

One must watch carefully for immune phase involving liver and kidney besides other organs such as brain that may overlap with fever or follow after a short afebrile period.

Case 3

Three years old child presented with fever and excessive irritability for last three days. There were no other specific symptoms. He was apparently well prior to this illness.
Fever suggests probable infection or inflammation and excessive irritability denotes pain; site of pain is not clear as child at this age can't localize the pain. In absence of any other localizing symptoms, it is likely to be a generalized pain. It may be either myalgia or bony pain. It can be decided easily on physical examination.
Physical examination showed significant pallor without any other signs such as lymphadenopathy or hepatosplenomegaly. It may suggest the possibility of leukemia as marrow aplasia does not cause pain. Diagnosis of **acute lymphoblastic leukemia** was confirmed by blood tests and further referred to specialist for management.
Acute leukemia in early stage may present with fever as the only symptom and physical examination may elicit pallor, purpura or bony tenderness. It is not easy to document bony tenderness in a crying child. High index of suspicion can help diagnosis of acute leukemia in early stage. Further immunological tests are necessary for complete diagnosis.

Case 4

Three years old female child presented with pain on standing and walking for last four days. Initially it was thought to be due to unnoticed fall or injury though child denied any such accident. There were no other symptoms. As pain persisted, parents noticed that child resisted movements of right leg and there was fullness of right knee joint.

This seems to be pain arising from either knee joint or periarticular tissues around the joint or bone. As there was no significant fever, acute infection is unlikely. So, it may be non-infective inflammation and leukemia should also be kept in mind.

Physical examination showed knee joint swelling with restricted active and passive movements. This confirms articular involvement. Periarticular disease presents with free passive movements though active movements are restricted. So, diagnosis of arthritis is made though etiology is not evident. One may have to wait to see further progress over days or weeks. Till then clinical follow-up is the most rational way forward. On detailed physical examination, few small joints of hand were also affected though minimally. So, this was not monoarthritis as thought of initially but it was pauciarthritis with three other joints involved. This pointed strongly to **Juvenile idiopathic arthritis.** Investigations – CBC was within normal limits though ESR was 75 mm that indicated inflammation. It pointed to a probable juvenile idiopathic arthritis. Child was treated with non-steroidal anti-inflammatory drugs and weekly methotrexate.It is important to note that patient may complain of limb pain while walking, in the initial stage of developing arthritis and subsequently, affection of small joints may be overlooked with a focus on a single large joint.Besides, examining all the joints, it is also important to look beyond the joints as other organs are also often involved in JIA such as iridocyclitis**.**

Case 5

One year old severely malnourished infant presented with excessive irritability and paucity of limb movements for last two days. There was no fever or any other significant symptoms. This child was fed with dilute milk without any solid food till date. He had past history of two episodes of loose stools.

Paucity of movements may suggest neurological disease but excessive irritability may denote possibility of severe painful conditions affecting all limbs that has resulted in pseudo-paralysis. So, it may represent generalised limb pain. In absence of fever, it is less likely to be infection or non-infective inflammation, though severely malnourished infant may not respond with fever in spite of serious infection or inflammation. Leukemia is a possibility and so also scurvy as this child is severely malnourished.

Physical examination showed severe malnutrition – weight 4.5 kg, length 65 cms, head O 43 cms, significant pallor, painful movements of all limbs, no purpura or bleeding gums, no hepatosplenomegaly or lymphadenopathy. Other systems were normal including nervous system.

Absence of enlargement of liver, spleen or lymph nodes favor diagnosis of scurvy in this child. Bleeding gums are seen only when many teeth erupt and as this child had only two teeth, this sign may not appear so early. Bony tenderness is seen in both scurvy and leukemia but is difficult to note in a crying infant and so not dependable.

Investigations ruled out leukemia and so diagnosis of scurvy was made. It can be proved by low serum vitamin C levels

though such a test is not routinely available. So best diagnostic test is radiological demonstration of subperiosteal hemorrhage along with other epiphyseal changes.
This child was confirmed to be suffering from **scurvy** besides multiple nutritional deficiencies.
He was treated with 500 mg of Vitamin C per day and within three days, pain had disappeared and he could move his limbs well and was no more irritable.
Scurvy manifests clinically only in absence of vitamin C in diet for more than six months. Thus, it is seen beyond the age of six months and only in severely malnourished child.
Symptoms improve dramatically within few days on oral administration of vitamin C and it is also a therapeutic test for scurvy. Vitamin C is not stored in the body and so body depends on daily intake of vitamin C.

Case 6

Ten years old male child presented with pain and swelling of left knee joint for two days. He was apparently well prior to present problem. He started getting pain in left knee region within half an hour after a trivial fall while walking. It was considered to be a sprain but got worst over next two days. There was no fever.
Apparently, it would look like a traumatic injury but it is worth noting that fall was trivial though outcome of injury was much more severe. It suggests underlying pathology. Such preexisting pathology may be congenital malformation in the joint itself or functional disorder like hemophilia or acquired condition like leukemia.

On detailed history, it was revealed that he would bleed easily after an injury that would take time to control. This in a male child suggested hemophilia and further confirmed by history of similar disease in maternal uncle.
Physical examination showed significant swelling of left knee joint with tenderness and restriction of movements – both active and passive – but without warmth or redness. This suggests diagnosis of **hemophilia**.
It was confirmed by blood tests showing very low levels of factor 8. Management consists of factor 8 replacement to stop further bleeding. Physiotherapy is necessary to prevent contractures but only when risk of bleeding is minimized and even then, one need to exercise caution while doing physiotherapy. Such a child is prone to trivial injury and so should be advised not to participate in physical sports.

Case 7

Eight years old child presented with history of pain and paucity of movements of both legs for last four days. There were no other significant symptoms. He had suffered from simple viral infection few days prior to this illness but had recovered completely without any specific drugs.
It is important to decide whether this is true paresis or pseudoparesis due to severe pain. On direct questioning, it was revealed that he could not move his limbs at all and so it was true paresis. Neurological condition that presents with pain may be due to affection of nerve roots or also due to spasm of affected muscles as a result of anterior horn cell irritation as in case of polio or polio-like viral infection..

Physical examination showed lower motor neuron involvement with symmetrical affection of both lower limbs without involvement of upper limbs or other areas. Further on raising leg straight, pain was exaggerated suggesting root involvement. This favored diagnosis of ascending polyneuritis – Guillen Barry syndrome. He was treated with IV gamma globulin and recovered completely over next few days. It is an autoimmune disease that may worsen over next few days and involve respiratory muscles requiring mechanical ventilation or also involve cranial nerves and autonomic nervous system. Most of the patients recover though few may remain with residual paresis.

Case 8

Ten years old child presented with history of pain over both legs mainly in the evenings for last two months. Through the day, he would remain active without pain but by late evening especially just before going to bed, he would complain of aches in both legs. Pain would get relieved with massage and he would fall asleep to get up and remain normal through next entire day. There was no relief with pain killers though massage made him feel relieved. This history of typical of growing pain. Physical examination in this child was totally normal. There is no need for any investigations in such a child. Parents were counselled about diagnosis of **growing pain.** It suggests tired muscles with accumulation of metabolic products that are absorbed following increase in blood supply with massage or application of heat. Exact cause is not known though it is a benign condition and gets

well by itself over time. One must avoid repeated use of analgesics that may lead to renal damage. Physical measures are good enough to offer relief.

Take home message

Pain in limbs may arise from many of the structures and it is important to determine the site of a lesion. Even a localized joint pain may begin with non-localising pain in the limb and what looks like a unilateral disease may also be a generalized disease. Pain cannot be localized by children under the age of 4-5 years and irritability may be the only presenting symptom. Generalised limb pain may arise from muscles or bones and differentiation is not easy especially in young children. Cause of limb pain may vary from very benign condition of unknown etiology to a very serious hematological vascular or neurological disease.

MCQs

1. Leptospirosis is suspected in presence of

A) Severe myalgia
B) Conjunctival congestion
C) increased serum creatinine
D) All of the above

2. This is the drug of choice in the treatment of leptospirosis

A) Gentamycin
B) Erythromycin
C) Doxycycline
D) None of the above

3. This factor differentiates generalised muscle pain from generalised bony pain

A) Severity of pain
B) High fever
C) Tenderness
D) None of the above

4. Pain may be so severe in this condition that it presents with pseudoparalysis

A) Leptospirosis
B) Scurvy
C) Both of them
D) None of them

5. This finding is classical of periarticular disease

A) Active and passive movements are painful
B) Active movements are painful but not passive
C) Passive movements are painful but not active
D) Both movements are not painful

Answers to MCQs

Q 1 D, Q 2 C, Q 3 C, Q 4 C, Q 5 B

2.27. **Breathlessness - It is scaring!**

Introduction

Breathlessness refers to increase in efforts of breathing. Normally air contains 20% oxygen and volume of air inspired and expired in every breath without extra efforts in a healthy

individual at rest amounts to about 6-7 ml/kg body weight. It provides adequate oxygen and gets rid of carbon dioxide. Oxygen concentration is low at high altitudes where even a normal person feels breathless. When oxygen requirements are increased as in case of physical exercise, healthy individual can easily compensate to an extent by merely increasing respiratory rate. However, beyond certain degree, person has to make extra efforts by using accessory muscles of respiration that normally do not come into play. However, time may come when muscles of respiration get tired and cannot sustain efforts of breathing that may endanger life.

Body's compensatory mechanisms

Unfortunately, oxygen cannot be stored in the body and hence must be made available all the time. Nature has provided mechanisms that can compensate to an extent for increasing needs or disturbed respiratory function. First step would be to increase respiratory rate. There is a limit to increasing respiratory rate as there has to be enough time to exhale carbon dioxide as well. Normally, expiratory time is 2-3 times of inspiratory time – IE ratio is 1:2 or 1:3. So when respiratory rate cannot be further increased, nature tries next step of using accessory muscles of respiration to push in more air at each breath. Intercostal muscles retract in the hope of sucking in air with more power and even smaller muscles contribute to such an effort. It reflects as increased effort at breathing and person feels uncomfortable and distressed. Concurrently, heart rate also increases and with increasing respiratory and cardiac activity, oxygen demands

also increase resulting in further imbalance. Increase in heart rate also has limitations as oxygen is supplied to heart during diastole and there has to be enough time for diastole as well. In normal adults, systolic time is about 60% of diastolic time though in young infants with faster heart rate, systolic and diastolic time may be equal or at times, even systolic time may be more than diastolic time. Initially by increasing efforts of breathing, oxygen requirements are barely met with but if disease worsens by then or respiratory muscles get tired, tissues suffer from lack of oxygen and retained carbon dioxide. This stage demands urgent intervention as body's available resources have failed.

Effects of lack of oxygen and retained carbon dioxide

Effects of oxygen deficiency and / or retained carbon dioxide are felt by brain the most as brain needs continuous supply of oxygen. Retention of carbon dioxide results in vasodilation of cerebral vessels that cause increase in intracranial pressure and poor cerebral perfusion resulting in deficient oxygenation to brain cells, as also happens if oxygen is not available in adequate amount from the lungs. It is how brain gets affected. Next to suffer is heart, followed by all other organs. Disturbed gas exchange results in change in pH of blood resulting in acidosis which in turn affects heart function resulting in rhythm disturbances. Heart dysfunction adds to the trouble – typically heart and lungs work together – it is cardiorespiratory system. Disease of one of them affects the other as well. Other systems have a functional

reserve and so can tolerate abnormal respiratory functions to an extent but not the brain or heart.

Common causes of breathlessness

Though diseases of respiratory and cardiac systems predominate in leading to breathlessness, affection of other systems also may cause breathlessness though mediated through cardiorespiratory system. Neurological diseases that control breathing can also cause breathlessness. Metabolic acidosis results in tachypnoea – deep and rapid breathing and may be mistaken for breathlessness though there is no increase in efforts of breathing. Cardiac disease may originate in renal system as a result of hypertension and adrenal disease may also be the cause of hypertension. Similarly severe anemia may cause cardiac dysfunction and thus, directly or indirectly, many systems may be involved and clinician must be aware of it. Common respiratory causes include pneumonia, pleural effusion, asthma, chronic bronchitis with emphysema in adults, bronchiolitis in children, interstitial lung disease and upper airway obstruction as in case of laryngitis. Common cardiac diseases in children are congenital heart defects and rheumatic heart disease (less common recently), myocardial diseases more in adults and so also hypertension. Neurological disorders presenting with breathlessness include lower motor neuron diseases such as ascending polyneuritis and polio-like illness. Central nervous disease may affect breathing rate, rhythm and depth but these are not presenting features.

Diabetic ketoacidosis presents with tachypnea mimicking breathlessness.

Clinical approach

Analysis of detailed history

First step in clinical approach is always to define anatomy of the disease – which system is involved. It is often possible to define microanatomy. Thereafter one should find out pathology and etiology is often a guess based on anatomy and pathology.

Age – Young infant in first three months has high chance of cardiac disease but in later infancy and thereafter, pulmonary diseases are more prevalent.

Origin, duration, progress – Respiratory diseases are often acute with exception of chronic interstitial lung disease while cardiac disorders are chronic with exception of acute myocarditis. Asthma is typically episodic and nocturnal. There is a difference between episodic and recurrent. Episode refers to similar presentation at similar time while recurrence may occur at any time.

Accompanying symptoms – Fever is common in respiratory diseases with exception of asthma while cardiac conditions are rarely accompanied with fever. Cough followed by breathlessness is classical in respiratory disease though cough may be absent in bronchiolitis or interstitial lung disease while recurrent cough is feature of heart defect with pulmonary congestion such as left to right shunt in which presence of mild tachypnea is often overlooked. Palpitation may be complained by older child suffering from heart

disease and occasionally an observant mother may give a history of rapid precordial pulsations. Hypophonia or aphonia in a breathless child suggests paralysis of respiratory muscles. Often on direct questioning, one can get idea of abnormal audible respiratory sounds. Stridor, wheeze orgrunt denote upper airway inspiratory obstruction, lower airway expiratory obstruction and lung parenchymal disease respectively. Hissing sound suggests nasal obstruction.

Behavior of the child is an important early indicator of lack of oxygen or retained carbon dioxide. Confusion or irrelevant behavior denotes lack of oxygen while irritability is a feature of hypercarbia.

Physical examination – nutritional and growth parameters help in differentiating acute from chronic conditions. Degree of breathlessness is evident by presence or absence of chest retractions and accessory muscles in action. Respiratory and heart rate and blood pressure changes offer clue. Generally breathless child has tachypnea and tachycardia but if tachypnea is proportionately more as compared to tachycardia, it would suggest primary respiratory disease. While, if tachycardia is proportionately more as compared to tachypnea, it denotes cardiac disease. Hypertension may be due to renal or endocrinal disease and also due to coarctation of aorta. Behavior change, blood pressure and pallor if any must be looked for. Chest signs such as restricted movements, change in percussion note, diminished or abnormal breath sounds and foreign sounds denote pulmonary disease while enlarged heart and murmur indicate cardiac disease. Chest signs may be absent in

interstitial lung disease as well as upper respiratory obstruction in laryngitis. Similarly, absence of murmur does not rule out cardiac disease. Cyanosis is a late sign in respiratory diseases while it may be seen in an otherwise comfortable child with cyanotic heart defect. Paralysis of respiratory muscles are easy to note. Measurement of oxygen concentration by pulse-oximete is now considered as part of physical examination as it can detect seriousness early enough to take proper corrective action.
In most of the situations, clinical diagnosis is possible with analysis of detailed history and focused physical examination.

Investigations

Provisional diagnosis is a must before planning investigations. Tests are only to confirm or rule one or two similar diseases that are considered. Of course, test results must be interpreted in the light of clinical profile.
CBC and chest x-ray are useful primary investigations in pulmonary diseases. While chest x-ray and ECG can give some clue to cardiac disease, echocardiogram is much more useful to get accurate information. One may need blood gas analysis in serious situations.

Management

Breathlessness is an emergency where patient needs hospitalization except an asthmatic who may be relieved of breathlessness in a short time with nebulized drugs. Specific treatment depends on final diagnosis but supportive therapy includes oxygenation, mechanical ventilation and hydration. Cardiac defects need surgical intervention for correction.

Neurological disease such as ascending polyneuritis is treated with IV immunoglobulins and may need physiotherapy.

Take home message

Breathlessness (increase in work of breathing) and tachypnoea are not synonymous though often used interchangeably. Tachypnoea or increase in respiratory rate is the first compensatory mechanism by the body before breathlessness ensues and so it is important to pick up. While breathlessness is an emergency requiring immediate resuscitation by oxygenation and further action decided by probable diagnosis. Behavior change is the early manifestation of hypoxemia in acute respiratory diseases and irritability in case of retained carbon dioxide.

MCQs

1. Oxygen deficit may result from

A) Low oxygen concentration in the environment
B) In spite of normal oxygen concentration in environment
C) Pollution in the environment
D) All of the above

2. Which of the following statement is WRONG?
Breathlessness may be the presenting symptom of

A) Renal disease
B) Endocrinal disease
C) Hematological disease
D) Vascular disease

3. Which of the following statement is WRONG? Hypoxic patient may have

A) Rapid and shallow respiration
B) Rapid and deep respiration
C) Slow and irregular respiration
D) All of the above

4. Which of the following statement is WRONG?

A) Pulse-ox may be abnormal in case of high fever
B) Pulse-ox may be abnormal in cardiac disease without breathlessness
C) Pulse-ox may be abnormal in a comfortable child
D) None of the above

5. Chest x-ray is not likely to be useful in this respiratory disease

A) Acute bronchiolitis
B) Chronic interstitial disease
C) Pleural effusion
D) Pneumonia

Answers to MCQs

Q 1 D, Q 2 C, Q 3 B, Q 4 D, Q 5 B

2.28. **Breathlessness – Need early recognition and prompt action**

Clinical application of basic concepts

Breathlessness is a symptom but much before it ensues, there are subtle clues for physicians to observe. Mild increase in respiratory rate is obvious especially in older children, only if carefully looked for. Mothers of young children often notice fast abdominal movements. Thus, tachypnea is earliest sign and it is at this stage, one can find out probable cause. Increase in heart rate is not evident to a patient unless older child complains of palpitation. If this early phase is missed, respiratory distress manifests with chest retractions – evidence of increased work of breathing. By now effects of hypoxia and / or hypercarbia are seen and earliest symptom of such abnormality is change in behavior – state of confusion, drowsiness or irritability. If this is missed, cyanosis develops a late sign indicating respiratory failure.

Case-based discussion

Case 1

Two years old healthy child presented with fever for a day followed by dry cough, noisy breathing and breathlessness that worsened over next 12 hours and fever continued.
Dry cough suggests involvement of upper airways and noisy breathing denotes obstruction to passage of air that must have resulted in breathlessness. Fever indicates infection and as other symptoms have followed quickly after the onset of fever; it is most likely a viral infection.
Physical examination revealed not sick looking but uncomfortable child with stridor, RR 40/min, HR 115/min, mild fever, marked suprasternal retraction, normal breath-sounds, no foreign sounds and other systems were normal.

These findings are in favor of acute laryngeal obstruction due to viral infection – **viral laryngitis - croup**. This child was treated with oxygen and hydration and recovered within next two days.

While this case is typical of croup, one must make sure that it is not a bacterial infection leading to upper airways obstruction as may happen in retropharyngeal abscess or rarely bacterial tracheitis. Patient suffering from such diseases are sick and highly febrile. At times, upper airway obstruction may be caused by angioneurotic oedema or laryngeal spasm due to tetany, in which case, there are other clues in history and physical examination.

.

Case 2

Two years old healthy child presented with mild fever and cold followed a day later with breathlessness for which child required hospitalization.

This history is almost similar to croup but this child has not reported any cough or noisy breathing. Absence of cough and noisy breathing rules out affection of larger airways but smaller airways such as bronchioles do present without cough. It also may be a cardiac disease such as acute viral myocarditis. Further physical examination would differentiate between these two possibilities.

Physical examination revealed no sickness but discomfort, rapid shallow breathing, RR 60/min, HR 120/min, chest shows bilateral emphysema with diminished breath sounds without foreign sounds, liver 3F +, liver span 7 cms, not

tender, hepato-juguar reflex absent, spleen just palpable, no cardiomegaly or murmur.
These findings favor diagnosis of **acute bronchiolitis**. It is a clinical diagnosis and generally no tests are necessary. Treatment is symptomatic with oxygenation and hydration. Prognosis is good and recovery is expected within few days. This disease is caused by respiratory syncytial virus and typically seen in healthy infants. Such a disease in a child with congenital cardiac defect or immune deficiency may progress into respiratory failure.

Case 3

Four years old child presented with history of fever and cough for two days followed by breathlessness that worsened over next 12 hours. Child was apparently well prior to this illness. Fever was high with sick interfebrile period. Cough was mild. High fever with sick interfebrile period suggests bacterial infection and mild cough points to probable lung affection. Breathlessness starting two days after onset of fever denotes progression of infection likely to be a developing pneumonia. Physical examination showed sick looking child, RR 40/min HR 105/min, intercostal chest retraction, chest movements reduced on right lower part anteriorly, TVF / VR increased, impaired note on percussion, bronchial breath sounds with few crepitations, other systems were normal.
These signs are typical of pneumonia of right middle lobe.
Investigations – Hb 11 Gm%, WBC 18000 P 78 L 22 E 0 Pl N
Chest X-ray revealed haziness on right side suggesting

diagnosis of **acute bacterial pneumonia**.
Child was treated with Amoxycillin-clavulanate and recovered well.
Sequence and progression of symptoms are important to note as much as severity of each symptom. Breathlessness appearing two days after onset of fever and mild cough is typical of pneumonia. With proper treatment, breathlessness is first to improve followed by fever but cough increases in severity. This is because damaged part gets liquified and nature tries to expel it through airways. It is not a worsening situation and parents must be counselled appropriately.

Case 4

Eight years old healthy child presented with high fever for a day followed by breathlessness that increased over next few hours requiring urgent hospitalization.
Breathlessness in this child has come up within a day of onset of fever and increased in severity quickly. It suggests immune response and not just the extension of infection that would have taken longer time to manifest more so in older child. This suggests development of severe pleural effusion. As there has been no history of any preceding infection, it is likely to be occult infection like tuberculosis to which this child has reacted immunologically.
Physical examination showed highly febrile child, uncomfortable, RR 45/min HR 120/min, chest movements absent on left side, trachea and apex beat shifted to right, dull note on percussion up to second intercostal space, absent breath sounds, no foreign sounds.

These findings are classical of large pleural effusion. Etiology is a guess – most likely tuberculosis.
Investigations – Hb 12 Gm%WBC 18000 P 80 L 15 E 2 M 3 Pl N Chest X-ray marked haziness all over left side with mediastinal shift to right, pleural fluid WBC 5000 P 75 Proteins 2.6 Gm suggestive of acute inflammation. Pleural fluid was negative for GeneXpert and also bacterial culture Thus, diagnosis in this child is based on local epidemiology and ruling out other infections. So final diagnosis was **pleural effusion due to tuberculosis.**
It is worth to note that neutrophilic leukocytosis in blood and pleural fluid denotes acute inflammation – either infective or non-infective immune inflammatory in origin. Progress of symptoms in a short time suggests non-infective immune inflammation. It is not unusual to confirm the diagnosis of tuberculosis in such a patient because focus of tuberculosis in such a child is expected to be very small situated in subpleural region that sets up a large immune reaction in the form of pleural effusion. Though every effort must be made to prove the diagnosis including contact tracing.

Case 5

Six years old child presented with acute onset of cough and breathlessness over last 12 hours. Cough was severe and dry. There was no fever. Child had similar episode few months ago and was treated with some medicines – details not known. There was family history of asthma in father.
This history clearly suggests bronchial asthma. Even in absence of past and family history, one would have

considered same diagnosis because acute onset of severe cough suggests airway disease and absence of fever denotes non-infective cause.
Physical examination showed uncomfortable child but not sick, RR 35/min HR 105/min, subcostal chest retraction, audible wheeze, chest movements bilaterally symmetrical, rhonchi + on auscultation. Other systems normal.
Diagnosis is clear – **bronchial asthma.**
There is no need for any tests in this child. Eosinophilia is not diagnostic of asthma. Chest X-ray may reveal prominent bronchovascular markings and at times patchy haziness due to small areas of atelectasis due to bronchial obstruction. Serum IgE is also non-specific and does not add value to diagnosis. Pulmonary function tests are not reliable at this age as child may not be able to perform the test optimally. Allergy tests are reserved only in resistant cases as majority patients are allergic to house dust mites, pollens and not commonly to food items. Other irritants such as smoke, perfumes and at times stress worsen the disease.
Acute attack is best treated with nebulised salbutamol and steroids. Adrenaline is used only in case of inadequate response. Maintenance therapy may be required in a child with persistent asthma.

Case 6

Eight years old child presented with history of breathlessness over last 2 weeks, gradually increasing and cough off and on. She was apparently well two weeks ago when she felt out of breath while coming back from school. However, on rest, she

felt better. Gradually she found it difficult to move about for which she sought medical attention. There was no significant past or family history.

Gradually progressive breathlessness with cough may be due to respiratory or cardiac disease. Significant cough indicates airway affection and considering breathlessness along with it, could it be asthma? But asthma is typically episodic and not gradually worsening. Intestitial lung disease may present with breathlessness gradually increasing, though such increase is over long time and cough is not a feature. Lung parenchymal disease often is infective and so presents with fever. So, this child may be suffering from cardiac disease. In absence of significant past history, congenital heart defect is less likely. So, it is likely to be an acquired heart disease. Most common cause of acquired heart disease in children is due to complications of rheumatic fever. Though there is no past history suggestive of rheumatic fever in terms of arthritis, such an episode may be incomplete and not even diagnosed as rheumatic fever. Gradually worsening breathlessness suggests increased pulmonary venous congestion as in case of mitral valve affection – either stenosis or regurgitation or both. Physical examination revealed uncomfortable child, RR 34/min HR 120/min BP 100/50 mmHg, peripheral pulses well felt, precordial pulsations, apex beat in 6^{th} intercostal space just outside mid-clavicular line, hyperdynamic, cardiomegaly+, pansystolic murmur best heard at apex and conducted to axilla, P2 loud, liver 2F+, firm, mild tenderness, hepatojugular reflex +ve, spleen not palpable, chest occasional crepitations, other systems normal. It was

diagnosed as **Rheumatic mitral regurgitation**. There was no evidence of active carditis. This was confirmed by chest x-ray, ECG and echocardiogram. Child was treated with oxygen and anti-failure line of therapy followed by long term penicillin prophylaxis.

It is important to differentiate cardiac diseases from respiratory disorders. Breathlessness in a respiratory disease is either acute or very slowly progressive while in cardiac disease it evolves over few days to weeks. Such a difference makes a child with respiratory disease more hypoxic as compared to a cardiac disease. However, there are always few exceptions.

Case 7

Two years old child presented with history of cough and breathlessness off and on and occasional episodes of fever since last one year. History of not gaining weight.

Absence of fever each time rules out primary infective disorder. As cough and breathlessness in this child suggests progressive disease as evident by faltering in weight and hence asthma is ruled out. Chronic interstitial lung disease would not present with significant cough and hence not likely. So respiratory causes are ruled out and so it must be a cardiac problem. Recurrent cough and breathlessness suggest pulmonary congestion and so probably left to right shunt defect such as VSD – it being the common defect.

Physical examination revealed wt 8 kg, length 82 cm RR 40/min HR 120/min BP 90/50 mmHg, no cyanosis, peripheral pulses well felt, precordial pulsations, apex in 5^{th} intercostal

space outside midclavicular line, hyperdynamic, systolic murmur best heard in parasternal area on right side 3rd intercostal space, P2 loud, liver 3F+, firm, mild tenderness +, spleen not palpable, chest few creps+, other systems normal. These findings are in favor of ventricular septal defect with congestive cardiac failure – **VSD with CCF.**

Investigations include echocardiogram that confirmed moderate size VSD with pulmonary hypertension. Chest X-ray and ECG add value to final diagnosis as lung markings and other chest defects and rhythm disturbances are not detected on echocardiogram.

Child was treated with anti-failure line of therapy and would need surgical correction at optimal time.

Congenital heart defect may present at variable time depending on severity of defect. Left to right shunt defects present only after few weeks after birth when pulmonary pressures are reduced and so blood can flow across the defect from high pressure on left side to lower pressure on right. Smaller the defect, later is the presentation. Atrial septal defect may remain undiagnosed even in adult life. Most of the cardiac defects are successfully repaired.

Case 8

Four years old child presented with breathlessness for last 12 hours that worsened over time and child became drowsy. He was suffering from asthma and was on regular compliant treatment and well controlled.

On the face of it, it may appear to be an attack of asthma but there was no cough at all, besides, the fact that he was well

controlled. This should raise suspicion about the diagnosis. Physical examination showed RR 45/min, deep and rapid breathing, chest clear no foreign sounds, HR 110/min, drowsy child responding to being called. Other systems N. Deep and rapid breathing with clear chest denotes metabolic acidosis and one common cause that is often hidden to begin with is diabetic ketoacidosis.

On direct questioning, this child did have polyuria, polydipsia and polyphagia for last one week prior to onset of this illness. Investigations confirmed the diagnosis of **diabetes mellitus.** Child was treated accordingly.

Metabolic acidosis presents with deep and rapid respiration and may simulate breathlessness, though there is no increased effort of breathing, hypoxia or any chest signs. Metabolic acidosis results commonly in case of severe diarrhea and also renal tubular disease. Both these conditions present with other symptoms and signs but diabetes mellitus presents with hidden symptoms that are often not reported or observed.

Take home message

Onset of breathlessness offers a clue to the diagnosis. Sudden development of breathlessness in minutes is often due to mechanical causes such as inhaled foreign body or pneumothorax or rarely pulmonary embolism in adults. Breathlessness occurring in few hours is usually immunological as in asthma or immune mediated pleural effusion, that presenting in few days may be due to infections like pneumonia, bronchiolitis or myocarditis.

Cardiac breathlessness due to heart defects may present in short period but in the background of symptoms of heart disease.

MCQs

1. Which statement related to breathless child is WRONG?

A) Cough is always present in respiratory disease
B) Cough is present in left to right shunt
C) Cough may not be present in respiratory disease
D) Cough may not be present in cardiac disease

2. Cough is a predominant symptom in this disease

A) Bacterial pneumonia
B) Acute bronchiolitis
C) Left to right shunt defect
D) Chronic interstitial lung disease

3. This is the only physical sign in acute bronchiolitis

A) Bronchial breath sounds
B) Hyper-resonant note on percussion
C) Mediastinal shift
D) Fine crepitations

4. Which of the following statement related to chest retraction in breathless child is WRONG?

A) Not seen in acute bronchiolitis
B) Not seen in chronic interstitial lung disease
C) Not seen in pleural effusion
D) None of the above

5. Which of the following statement is RIGHT?

A) Wheeze is heard in asthma
B) Wheeze is heard in cardiac failure
C) Wheeze may be present in case of inhaled foreign body
D) All of the above

Answers to MCQs

Q 1 A, Q 2 C, Q 3 B, Q 4 D, Q 5 D

2.29. General weakness – a vague symptom

Introduction

General weakness is not only a subjective feeling of the patient but vague enough that demands seeking clarification by the physician before embarking on its analysis. It may connote different meaning to different people. When a mother complains about general weakness in her child, it depends on her subjective understanding of what child has. It may be far from what the real problem may be. Often a mother equates thin stature of a child to weakness.

Basic concepts

Generalised weakness is a complaint that mostly does not refer to neurological disorder. However, one must make sure that weakness does not relate to paresis. Rarely though, severe hypokalemia in a malnourished child or in a sick elderly individual may present with generalised paresis and so also in case of progressive ascending polyneuritis. So, one must confirm that there is no paucity of movements in a child presenting with generalised weakness.

Generalised weakness refers to tired muscles. Muscles are short of desired energy required for movement and so such a patient wants to move and also can move but finds it very tiring and so reluctant to move.

What do muscles need for adequate functioning?

Muscles need oxygen that is provided by lungs, pumped by heart and transported through hemoglobin to the required sites. Thus, a child with slowly developing lung or heart disease or anemia may fail to deliver adequate oxygen to muscles and so present with generalised weakness. If lung or heart disease or anemia develops rapidly, child presents with acute breathlessness rather than generalised weakness. Thus, generalised weakness develops over time and rarely in a short time. Besides oxygen, muscles need glucose for energy. It is not hypoglycemia but hyperglycemia that fails to deliver enough glucose to muscles. If a child is hypoglycemic, presentation is acute with seizure or change in behavior. But in case of hyperglycemia as in diabetes, muscle cells cannot utilize glucose in spite of its availability and hence remain deprived of necessary glucose. Thus, diabetes may present with generalised weakness. However, if a child with diabetes often presents acutely with diabetic ketoacidosis and preceding phase of weakness may be short and missed. Energy comes from calories – though carbohydrates are major source of calories and thus child with severe protein-energy malnutrition (PEM) also has generalised weakness. He is reluctant to play and prefers to be inactive. Muscles in such a child are wasted that causes generalised weakness, besides

the fact that nutrients are also not available.
Muscles also need intracellular ions such as potassium and magnesium for adequate function. However, hypokalemia or hypomagnesemia are mostly acute and when severe, they manifest paresis – neurological weakness. Though rarely renal disease may present with chronic hypokalemia presenting with generalized weakness.

Causes of generalised weakness

As mentioned in the discussion above, severe PEM is a common cause generalised weakness though major symptoms pertain to acute infection that brings the child to a health facility. In addition, loss of weight and appetite predominate. In older children, PEM is secondary to chronic progressive disease as in case of end-stage renal disease. However generalised weakness may the only presenting symptom in chronic lung diseases such as interstitial lung diseases (chronic bronchitis with emphysema is rare in children), chronic heart conditions (congenital heart defects, rheumatic heart disease and myocardial disorders) and chronic anemia of various etiologies (iron or Vitamin B12 deficiency, congenital hemolytic anemia and marrow aplasia). Diabetes as a cause of generalised weakness should be kept in mind and will be evident only if one asks for polyuria, polydipsia and polyphagia. As generalised weakness is a prominent symptom, other symptoms are often not complained of, unless asked for.
Chronic hypokalemia is a rare to present as generalised weakness.

Clinical approach

History – it is important to confirm that the child can move limbs in all directions, thus ruling out paresis.
Onset and duration – acute onset within few days is typical of diabetes that is evident by asking about thirst, increased urine output and increased appetite. Most other conditions present subtly over days or weeks.
Progress – fast deterioration is a risk if one misses diabetes while other conditions worsen slowly.
Localising symptoms – chronic lung disorders are easily missed as patients get adapted to hypoxia and do not complain about breathlessness though they do have tachypnea that is not noticed and complained by the patient. Palpitation may be a symptom in chronic cardiac conditions in older children while mother may notice precordial hyperactivity in younger child. Cough may be a symptom in both cardiac disease but often absent in chronic interstitial lung disease. Significant pallor is rarely missed by parents and often is a main compliant along with generalised weakness. So, it is easy to make out in history itself.

Physical examination

General appearance – tired and chronically sick look is common to all except child with diabetes who may look acutely sick especially if initial stage is missed. Child with PEM may be irritable or lethargic depending on severity of protein or calorie deficiency.
Growth parameters – growth faltering is the hallmark of PEM but also abnormal in other chronic conditions to a variable

extent. Child with diabetes may have normal growth.
Pallor is striking in case of severe anemia while many other conditions may also be marginally pale.
Heart rate and respiratory rate – are important to pick up subtle cardiac and respiratory disorders. Mild tachypnea is easily missed unless one counts respiratory rate over a full minute and same is true with mild tachycardia in a cardiac disease. Such basic signs can pick-up chronic but otherwise silent cardiac or respiratory diseases that are otherwise missed. Pulse-oximetry is now considered as bedside clinical maneuver as much as use of blood pressure in routine clinical practice. This is because it offers great help in early pick-up of respiratory and cyanotic heart defects. In fact, it is now being used routinely in every newborn for early diagnosis of congenital heart defect.
Systemic examination – murmur, cardiomegaly and signs of cardiac failure would suggest a cardiac disease. Mild increase in respiratory rate may be the only finding in chronic interstitial lung disease that is easily overlooked. Pulse-oximetry will show low oxygen saturation in such a case. One must make a habit of noting pulse and respiratory rate judiciously. In case of severe pallor, presence of hepatosplenomegaly suggests hemolytic anemia while absence indicates either deficiency anemia or marrow aplasia. Child with marrow aplasia looks sick while one with deficiency or hemolytic anemia are not sick looking.

Investigations

Specific investigations are planned according to the system involved. At times, there is no clue on history or physical examination. In such a case, chest x-ray and echocardiogram / ECG may offer clue. However if both these tests are normal and one is sure about the problem being pathological, further tests may be necessary to rule out cardiac disease (stress test) and interstitial lung disease (CT scan). Detailed hematological tests can define specific cause of anemia (serum iron or B12, hemoglobin electrophoresis).

Management

It would depend on specific diagnosis. What is important to note is a fact that generalised weakness is a vague symptom and cannot be passed on by prescription of a “tonic”. There is a risk of missing life-threatening diseases. A child with PEM needs not only nutritional supplements but counselling for rehabilitation and ruling out comorbid conditions. Specific treatment depends on final diagnosis.

Take home message

Generalised weakness as a presenting complaint needs cautious approach because some of the conditions present with subtle clues on history as well as physical examination that are easily overlooked. Diabetes mellitus is easy to miss unless thought of and so also chronic renal tubular disease. Polyuria is common to both and personal history mostly focuses on oliguria and not polyuria. Thorough physical examination should start with actual count of respiratory and heart rate so as not to miss mild abnormality.

MCQs

1. Weakness to lay people may mean

A) Thin stature
B) Loss of weight
C) Paresis
D) Any of the above

2. Choose the RIGHT statement. General weakness to a doctor should mean

A) Wants to move but cannot
B) Reluctant to move due to severe pain
C) Reluctant to move in spite of no pain
D) Feels tired easily

3. Pseudoparalysis is characterized by

A) Severe pain
B) Normal power
C) Normal deep tendon reflexes
D) All of the above

4. Choose WRONG statement. General weakness in PEM is due to

A) Poor muscle mass
B) Hypoglycemia
C) Hypokalemia
D) None of the above

5. Choose the WRONG statement. Hypoglycemia may present as

A) Generalised weakness
B) Seizure
C) Unconsciousness
D) All of the above

Answers to MCQs

Q 1 D, Q 2 D, Q 3 D, Q 4 D, Q 5 A

2.30. General weakness – needs a cautious approach

Clinical application of basic concepts

Thin stature is often considered equivalent to weakness. Weakness is a vague symptom and refers to lack of energy. Energy is provided through nutrition and oxygen. Naturally, many organs are involved in providing energy and undue expenditure of energy also would lead to weakness. However, when generalied weakness is the only complaint, one needs to go into the details of history to find out presence of subtle symptoms that may have been overlooked. Weakness is also a term used for paresis and one must make sure to rule out neurological disease.

Case 1

Ten years old child presented with generalised weakness for last two weeks. There were no other symptoms. On direct questioning, this child had stopped going out to play for last few months though for no apparent reason. He would prefer to be sedentary. There is no history of palpitation or breathlessness or polyuria, polydipsia and polyphagia.

Parents had not noticed any significant pallor.
It is obvious that this problem has existed for several months though may have been aggravated over last two weeks and hence presented now. This history suggests that this child has been getting progressively tired over months. Absence or palpitation, breathlessness and symptoms of diabetes rules out chronic lung, heart conditions and chronic anemia. So, we have no clue to diagnosis on history.
Physical examination showed no abnormality on casual cursory examination but on detailed observation, his respiratory rate was 30 per minute at rest and it was certainly abnormal. Oxygen saturation at rest was 92% that meant he was hypoxic. Though he was apparently comfortable. This finding is in favor of chronic interstitial lung disease. Investigations showed Hb 15 gm%, CBC otherwise normal, chest x-ray normal, HRCT showed evidence of interstitial lung disease. Thus, diagnosis of **chronic interstitial lung disease** was confirmed. Etiology is often not evident but may be related to environmental factors triggering immune response. Steroids are considered on such a probability.
History should start with confirmation of period of onset of the symptoms as subtle symptoms are often not reported. This case emphasises need for thorough physical examination including counting respiratory and heart rate for a minute. Chronic hypoxia may often be tolerated by patients as they get adapted to it as polycythemia often compensates mild chronic hypoxia and hence delay in the diagnosis.

Case 2

15 years old child presented with generalised weakness and on direct questioning he reported tiredness that was gradually increasing. Apparently, he had no other symptoms except mild cough. At the age of 6 years, he had suffered from rheumatic fever with arthritis, from which he had recovered completely.

Gradually increasing tiredness suggests slowly progressive disorder. In the background of rheumatic fever in the past, one may consider heart condition such as valvular damage. Tiredness in this child indicates deficient oxygen supply to muscles as a result of cardiac disease. It denotes pulmonary venous congestion due to back pressure from left side of heart. It may favor diagnosis of mitral stenosis. Mitral regurgitation may have been more symptomatic with breathlessness.

Physical examination revealed heart rate 92 / min at rest, respiratory rate 27 / min (both mildly increased), no obvious cardiomegaly but loud first heart sound and mid-diastolic murmur at mitral area. This suggests diagnosis of **rheumatic mitral stenosis.** Echocardiogram confirmed the diagnosis of mitral stenosis. This child needs surgical intervention.

Slowly worsening left sided heart function may present with exertional dyspnea that patients perceive as tiredness and hence complain of general weakness. This case illustrates how this vague symptom needs to be analysed.

Case 3

Four years old child presented with generalised weakness for last four months that made him not to participate in play

activities. Apparently, there were no other symptoms except mild cough. On detailed questioning, he would feel tired after playing for few minutes and would rest in between such activities. It suggested slowly worsening tiredness over several months. It could be either lung or heart disease. If it is a heart disease, it is unlikely to be congenital heart defect because he was normal during first three years of life. Rheumatic heart disease also can be ruled out as he has been too young to have suffered from damage to heart as a result of rheumatic fever. However other types of heart diseases such as primary myocardial or pericardial conditions cannot be ruled out. Chronic interstitial lung disease is a possibility as interstitial lung diseases have different etiologies including damage due to previous viral infections and also due to environmental exposure to toxic or pollutants.

Physical examination showed heart rate 104 / min and respiratory rate 28 / min (both on higher side), precordial pulsations, mild cardiomegaly, soft systolic murmur at mitral area not conducted with normal heart sounds. Chest examination revealed few crepitations at the base of lungs. This suggests probable primary myocardial disease. Echocardiogram confirmed diagnosis of **cardiomyopathy**. Etiology of such a disease is not easy to correlate as it may result from events that have occurred in the past such a viral infection. Further investigations are necessary to rule out metabolic or storage disorders.

Case 4

Eight years old child presented with generalised weakness and tiredness over last two months that was gradually increasing. On direct questioning, he reported mild fever off and on during this period. But it did not disturb him and so was ignored. There were no other symptoms.

Mild fever off and on suggests probable infection that has affected either lung or heart. Absence of cough almost rules out infective lung disease. Valvular or myocardial diseases do not present with low grade long duration fever but pericardial affection is likely. It may be in the form of mild pericardial effusion or constrictive pericarditis. Mild pericardial effusion may not be symptomatic and if fluid accumulation increases, it may lead to breathlessness. However constrictive pericarditis may deteriorate slowly as fibrosis develops slowly in chronic infection such as tuberculosis. So, one must look for evidence of constrictive pericarditis in this child.

Physical examination showed mild tachycardia and tachypnea but no other cardiac findings. However, liver was enlarged and firm but not tender and neck veins were engorged without hepatojugular reflux. This suggests superior and inferior vena caval obstruction and so diagnosis of **constrictive pericarditis due to tuberculosis** was considered. Echocardiogram confirmed the diagnosis.

Constrictive pericarditis hampers free heart movements during cardiac cycle. As the process evolves slowly, heart function does not deteriorate over short time as may happen in progressive pericardial effusion with cardiac tamponade.

Hence patient feels increasingly tired over time and presents with weakness.

Case 5

Four years old child presented with generalised weakness and reluctant to participate in play activities over last six months. It was gradually increasing to an extent that he would refuse to go out to play. Earlier he used to be very active child. There were no other symptoms.
Gradually increasing tiredness indicates slowly worsening oxygenation either due to chronic lung or heart disease. Absence of cough rules out lung disease as well heart disease with pulmonary congestion. So, it may be due to heart condition with pulmonary oligemia – less of blood flowing through lungs resulting in lack of oxygen. However, such a child should develop cyanosis but if pulmonary obstruction is mild, cyanosis may not be noticed by parents. Though on direct questioning, parents may reveal probable cyanosis seen on crying. However, it can be easily overlooked.
Physical examination showed mild clubbing of nails that is suggestive of chronic hypoxia, systolic murmur at pulmonary area without cardiomegaly or cardiac failure. This is in favor of **pink Fallot's tetralogy.** Echocardiogram confirmed the diagnosis. He would need surgical correction.
This case illustrates the fact that mild cyanosis is difficult to be noticed by the patient and also by the doctor. Pulse oximeter is now a part of physical examination that would pick up mild hypoxia that can be further evaluated.

Case 6

Six years old child presented with slowly progressive tired feeling over last 6 months. He was normal prior to onset of present symptom and was very active. Initially he would run about but would need to rest – it was unusual for him though was ignored. When this problem increased over time to an extent that he would refuse to go out to play, parents sought medical advice. On direct questioning, he had developed cough over last two months that was also slowly worsening. It suggests slowly worsening tiredness and cough. It suggests probable space occupying lesion in the chest now compressing over airways leading to increasing cough. Physical examination showed impaired note on percussion on right anterior side of the chest but it did not correlate with either lobes of lungs or pleura. It denotes probability of mediastinal mass in the chest. Chest x-ray confirmed **large mass in anterior mediastinum** and was proved to be benign. It was surgically removed.

Physical findings in the lung lesion are restricted to surface area of lobes of lungs – it means right upper lobe lesion has findings only in upper half of the chest anteriorly and right middle lobe only in lower half of chest anteriorly. Lower lobe is represented only posteriorly. Physical findings in pleural disease are seen below a particular intercostal space but all over the chest anteriorly, laterally and posteriorly. This is referred to as lobar or pleural distribution of signs respectively. When physical signs do not corelate with either of these two patterns, one considers space occupying lesion.

Case 7

Ten years old child presented with generalised weakness and tiredness for last one week. He was very active prior to onset of this symptom and in fact used to attend coaching for tennis. One week ago, he felt tired while playing though coach insisted on his continuing to play and next day he refused to go to play. Initially parents thought he was just being stubborn. But when his problem persisted, they visited a doctor. It was only on direct questioning that polyuria, polydipsia and polyphagia was apparent and so diagnosis of **diabetes mellitus** was confirmed. He would have come in diabetic ketoacidosis with unconsciousness within next few days had he been further ignored.

This case illustrates importance of detailed personal history. As oliguria is more common than polyuria, such a symptom is often missed while eliciting history. Polyuria may be caused by diabetes mellitus, diabetes insipidus, renal tubular disorders or it may also be psychogenic.

Case 8

Two years old child brought by mother complaining of generalised weakness since the age of one year. According to mother, this child was doing well till the age of one year. He had grown well and weighed 10.5 kg at one year, his food intake was good. But thereafter he started making fuss about eating and in spite of trying various methods and tonics, there was no improvement. He had gained just 1.5 kg through entire second year. So, she was worried about his generalised weakness.

On direct questioning, mother reported that he maintained

his activity level and there were no physical complaints. Active child almost rules out any significant pathology. Physical examination showed happy and active child, weight 11.5 kg, length 87 cms and there was no abnormality detected. So, this child was completely **normal**.
This mother was interpreting “poor” intake of food and “poor” weight gain during second year as weakness.
She required to be counselled rather than child needing any investigations or treatment. So, mother had to be managed. Health status is better judged by activity level and growth by length / height and not by weight. Weight gain is variable in a normal healthy children and all that we need to ensure is no weight loss and some weight gain over months. Weight gain in a normal child during second year is one third of that gaind in first year while length gain is half of what is gained in first year. Thus, every normal child looks thin during second year as compared to first year due to differential gain in length and weight. This is often reported by mothers as weakness.

Take home message

This case emphasises the need to understand terms used by the patient that may not corelate with real problem. There are several terms used by patients while describing their symptoms, meaning of which may be misinterpreted by the doctor. Weakness is one such term that may connote paresis or tiredness but also just a thin stature. Cold and cough are other such terms used to connote noisy breathing. Patients also use medical terms to describe their symptoms such as acidity, chest congestion or throat problem. Unless doctor

tries to decipher correctly what patient wants to convey, further evaluation may proceed in wrong direction.

MCQs

1. Generalised weakness is the presenting complaint in this infectious disease

A) Acute UTI
B) Hepatitis A viral infection
C) Acute URTI
D) Acute meningitis

2. Which statement is RIGHT? Reason for generalised weakness in hepatitis A infection is

A) Sudden destruction of large number of hepatocytes
B) Development of jaundice
C) Liver failure
D) All of the above

3. Generalised weakness may be the presenting complaint in this chronic disease

A) Chronic sinusitis
B) Chronic UTI
C) Tuberculosis
D) All of the above

4. Which statement is RIGHT? Reason for generalised weakness in above diseases is

A) Low grade fever
B) Prolonged fever

C) Increased catabolism
D) All of the above

5. Generalised weakness in interstitial lung disease is easily missed because

A) Absence of cough or breathlessness
B) No chest signs
C) Respiratory rate not counted
D) All of the above

Answers to MCQs

Q 1 B, Q 2 A, Q 3 D, Q 4 C, Q 5 D

2.31. My child is not gaining weight!

Introduction

Children during their growing period must show age-appropriate weight gain, though undue weight gain at other age groups may be pathological. Rate of weight gain varies in different phases of life. Weight gain is maximum during adolescence when a child gains about 15–20 kilogram weight over four years till puberty is achieved and continues to gain about 8-10 kilograms over next four years. Average weight gain in first year is about 5-6 kilogram and thereafter it slows down to about 2 to 2.5 kilogram per year till adolescent spurt starts. Around 7-9 years of age, there may be a small increase in weight over the average, amounting to about 3-4 kilogram per year during those two years. While adults are expected to maintain weight but small gain in weight may continue

even in adult life. However, rate of weight gain not only depends on different periods of growth but also varies in individuals depending on genetic endowment, environmental influences and life style. Thus, if one child grows 3 kilogram a year after first year and the other 2 kilogram a year, at the end of another 10 years, they end up with difference of 10 kilogram in their comparative weight but both may be equally healthy. It is the trend noted by periodic weight record that is more relevant. This is important to note that weight is not the only criterion of health.

Make sure what parents mean

Most parents don't know what the average weight should be at a particular age. It is often their perception that the child is not putting on adequate weight. This is a common complaint especially in toddler age group. It is because infant gains about 6 kilogram weight and 25 centimeter length in first year that drops to 2 to 2.5 kilogram weight and 12 centimeter length in second year. Hence there is a disparity between weight and length gain in second year as compared to first year. Weight gain is 40% and length 50% of the respective gain in first year. Thus, child in second year is perceived to be thinner in relation to length as compared to stature during first year. Such a change is physiological. In addition, forced feeding that inevitably results in second year compounds the problem. During subsequent years, child's weight gain follows individual pattern that may be different from the average. Parents often compare child's weight with that of other children around same age but they forget that

two children of same parents also differ in their stature and at times even twins differ. So, doctor must make sure whether there is a real problem of poor weight gain or just an imaginary one.

Confirm “not gaining weight?”

Healthy child is always active, playful and happy without any symptoms. This is much more so in younger children. As age advances, behavioral issues may come in the way of happiness in particular and also may affect activity level. However, most complaints about weight gain exist in case of toddlers and younger children and this is in spite of children remaining active and happy. With too much focus on weight instead of health, poor weight gain is often a universal complaint especially in middle and upper socioeconomic groups. Objective way to monitor weight is to maintain growth chart. As every child has different birth weight and weight velocity. Growth chart in a normal child is expected to maintain around same centiles through growing period. In simple words, growth of the child follows the line on growth chart which he / she has always been before. It indicates normal growth pattern irrespective of actual weight. Growth chart is also useful to monitor length / height till puberty is reached and head circumference in first two years of life as brain growth is nearly over by end of second year. Each child should have a growth chart and parents should maintain periodic charting, initially with the help of a doctor in infancy but later by themselves. Normal growth pattern as shown in linear records in a chart can assure parents about child’s

growth being normal. It also provides confidence to a doctor not to indulge in unnecessary laboratory tests.

Clinical approach

First step is to confirm whether child is losing weight or weight is stationary for more than expected period or whether child gains weight but less than expected at that age. Loss of weight certainly is abnormal and needs explanation. In first six months of life, if an infant's weight remains the same even for a fortnight, it is considered abnormal because this is an age of fast growth. In subsequent months of infancy, if weight has remained the same over a period of one month, one needs to look into probable cause. Beyond infancy, absence of weight gain over three months period should be considered abnormal.
Once it is confirmed that weight gain has faltered, next step is to find probable cause. In an otherwise asymptomatic infant, most common cause is inadequate nutrition, mainly due to faulty feeding such as over-diluted milk or delayed introduction of complementary feeds. In an otherwise asymptomatic toddler, feeding problems such as bottle or breast addiction and forced feeding by parents are common reasons for absence of weight gain. In such an infant or toddler, appropriate correction of faulty feeding settles the problem and there is no need for any medical help.
However, in an older child in spite of being asymptomatic, weight loss or absence of weight gain over three months may need proper assessment to exclude pathological causes. Of course, if child is symptomatic at any age with weight loss, it

clearly calls for finding the cause. Such children are sick to a varying extent, have poor appetite and are cranky. So, they are easy to recognise.

Pathological causes

In every acute short-lasting illness, there may be weight loss recorded but such a child would regain weight in a short time again. All that one needs to do is to confirm regaining of weight. It calls for no more intervention. However. if a child after a weight loss fails to regain original weight, one must observe for some more time and take appropriate action if weight does not pick-up. It may indicate incompletely cured disease or underlying silent disease. In a chronic illness, significant weight loss is expected and it would take longer time to regain lost weight but on recovery, child should stop further loss of weight and then start, at least, gaining little weight. Regaining weight to original level may not occur after severe loss of weight during prolonged illness though on all other counts, child should have improved especially in terms of energy level and appetite.

Weight loss in absence of obvious illness

It is a real challenge. One must get into details of history to find any clue to a hidden problem. Such problems could be low grade infection such as tuberculosis or slowly evolving organ dysfunction. Loss of appetite, behavior change such as irritability or lethargy, disinterest in play or activities that he / she loved earlier are some of the signals of hidden disorders. It may be ideal to monitor body temperature by thermometer to pick-up any low-grade fever that would

necessitate investigations for infections. In absence of documented fever, hidden organ dysfunction must be scrutinized. Subtle symptoms often are not reported by patients or parents of children and doctor must enquire about them. Mild puffiness of eyelids in the morning may indicate renal disease. Vague abdominal pain or distension and change in bowel pattern may signify hidden intestinal disease such as lymphoma or inflammatory bowel disease. Unexplained pallor is due to hematological disorder and may present only with mild enlargement of spleen. Undue tiredness after accustomed exertion may suggest either a lung or heart defect or diabetes mellitus. Acquired or inborn metabolic disorders may present initially without any obvious signs on physical examination. Doctor must enquire about subtle symptoms or look for subtle signs on physical examination to pick up a hidden cause of poor weight gain or loss of weight.

Take home message

Parents often complain of children not gaining weight but it may be just a wrong perception. It could easily be evident when you see such a child being active, playful and happy. Further, growth chart helps in objective assessment. However, once the problem is confirmed to be genuine, there would be a need to pinpoint the cause. At times, cause is hidden and detailed enquiry and focused physical examination can offer a clue. It is irrational to prescribe a “tonic” without proper evaluation for a child who is not gaining weight. Anyway “tonic” mostly does not help.

MCQs

1. This symptom is seen in both healthy and diseased child

A) Active
B) Playful
C) Loss of appetite
D) Happy

2. This is most dependable in deciding whether problem is genuine or not

A) History
B) Physical signs
C) Investigations
D) Growth chart

3. Indicator of a genuine problem on growth chart is

A) Weight at first visit
B) Velocity of weight
C) Both
D) None

4. Which of the following statement related to infant's weight gain is RIGHT?

A) Must gain every month
B) Must gain every two months
C) Must gain every week
D) All of the above

5. Which of the following statement is WRONG? This may be the hidden cause of not gaining weight

A) Acute infection
B) Chronic organ dysfunction
C) Chronic infection
D) All of the above

Answers to MCQs

Q 1 C, Q 2 D, Q 3 B, Q 4 A, Q 5 A

2.32Poor weight gain – needs a proper evaluation

Basic concepts for clinical application

Healthy children of the same age vary a lot in their weight and hence a record of weight at one single point may fail to judge whether child is failing to gain weight. Growth chart shows a range of normal weight expected at a particular age, upper level being at 97th percentile and lower at 3rd percentile, 50th percentile being the mean. A child who starts at 3rd centile and maintains same level over years is considered normal. It is only downward trend that signifies weight loss. It is different than not gaining weight. Weight of children beyond infancy may remain the same even over three months' period and it is normal. This is because normal weight gain at this age may be as small as 500 grams over three months that is not even easy to assess. Hence when child presents with not gaining weight, doctor must make sure whether it is true or just perceived. Most of the times, it is a perception of parents without monitoring child's weight.

Case-based discussion

Case 1

Mother of 18 months old child complained of poor weight gain for last 8 months and poor appetite for last 6 months. There were no other symptoms. Child was gaining weight well in first six months and thereafter gained less weight over next 4 months. But last 8 months, there has been hardly any gain in weight. On direct questioning, child remained active, had normal bowel movements and urination and slept well. On this history, it is clear that this child is unlikely to have any significant disease as he has been active and playful. So, we need to look at his growth chart to decide whether he has been faltering in his weight centiles. It would help to decide probable cause of faltering if any.

Physical examination did not reveal any abnormality. Growth chart showed that he had maintained his weight centiles and so also his length and head O. Hence this is **a normal child**. Many mothers perceive poor weight gain around second year of their child. This is physiological in the sense that weight gain slows after first year. But add to the problem of misconception, most mothers start forcing to feed the child and it becomes a vicious cycle. More the mother forces the child, more he refuses and starts hating food. This pattern ends up in poor eating in third year and child may even start losing weight. Counselling is an important part of management of such a problem and not the “tonics”.

Case 2

Four years old child was brought by parents because of poor weight gain after first year of life and poor appetite. There

were no other symptoms. Previous growth records showed that his weight centiles had come down from 75th to 25th level though his height was maintained above 50th centiles through last four years. He remained active and playful and had normal development.

History suggests that this child has genuinely faltered in his weight centiles without any symptoms or change in his activity and energy level. It mostly rules out any active disease. As his height has maintained centile level, it may suggest recent poor intake of nutrition. However, parents complain about poor intake of food since the age of one year. Had it been poor intake of nutrition over last three years, his height also would have started showing decline in centiles. This kind of disparity suggests that his intake of food is sufficient for his energy and activity level though parents perceive it to be poor. After all, metabolism decides how much you use out of what you eat. It is similar to a fact that amount of study is not proportional to success achieved but lot depends on how much you use.

Physical examination did not reveal any abnormality.

Thus, we can conclude that this child is deriving enough energy from his food irrespective of what parents consider it to be poor intake. So, it is likely to be constitutional thin stature. This child is “programmed” to be thin but healthy. It may be a familial tendency. On direct questioning, it was revealed that his father used to be thin during childhood though now he is obese. This is a **normal child** and in fact should take adequate care not to become obese later in life. This child does not need any drugs or investigations but

parents should be properly explained and counselled.
This case illustrates a fact that weight below the expected average may not be pathological but just a constitutional or genetic factor. Growth chart helps to allay anxiety of parents.

Case 3

One year old infant presented with poor weight gain since birth. He was born at the end of full term with birth weight of 1.4 kg. His cry was poor at birth and required bag and mask ventilation with oxygen for half an hour. He was fed with nasogastric tube for 2 days followed by spoon feeding and then on breast feeding, He did not need any other intervention and was discharged after 10 days. His weight has always been below 3rd centile, his length on 5th centile and head O on 10th centile. His motor milestones are delayed as he has started to sit but with support but his other milestones are near normal. His mother had suffered from prolonged febrile illness during second trimester and had lost 6 kg weight in this illness.
This history suggests lack of fetal nutrition and explains his poor birth weight as well as remaining below the expected centiles at one year of age. In view of absence of any other symptoms, it is unlikely that this child has any other disease.
Physical examination at one year – weight 6.8 kg, length 71 cm, head O 44 cm, mild pallor +, no other abnormal findings.
He has gained 5.4 kg in one year which is within normal limits. Birth length was not recorded.
So, this child is **normal** and expect him to catch up over time to near normal adult parameters.

In malnutrition, weight goes down first and if nutrition continues to be poor, length starts faltering after few months. However, head O remains within normal limits. This is the way nature protects brain even in malnutrition. Only exception is when malnutrition starts early in first trimester of pregnancy.

Case 4

Two years old child presented with poor weight gain since last one year. He was born after full term delivery with birth weight of 3 kg, was breast-fed exclusively for first 6 months and thereafter was on complementary feeds along with breast feeding. Mother continued to breast-feed this baby even now though milk yield had been very scanty. The child gradually started fussing to eat but would demand breast milk. Initially forced feeding worked for a short time but he learnt to vomit food if forced. Thus, his intake of food had gone down drastically over last few months and was surviving only on small quantity of breast milk. He had started going to playgroup and since then used to get frequent episodes of cold and cough with occasional fever. He was constipated and suppressed urge of passing stool to avoid painful defecation. His sleep was disturbed and he was getting cranky.

This history clearly suggests significant deficiency of nutritional intake. Recurrent episodes of cold, cough and occasional fever relates to contact with probable viral infections and aggravated by malnutrition. It is unlikely to be malnutrition secondary to chronic disease and so is likely to

be due to primary malnutrition because of breast addiction and refusal to eat solid food.
Physical examination showed significant pallor, signs of rickets in terms of fronto-parietal bossing and beading of costochondral junctions. There was generalised hypotonia of muscles. No other abnormal findings.
Laboratory tests confirmed severe iron deficiency anemia and x-ray of wrist showed signs of rickets.
This child's poor weight gain is due to **breast addiction** that led to fussy eating resulting in mother forcing the child to eat and it started a vicious cycle ending with poor nutrition and poor weight gain. This child needs to de-addict from breast feeding. Mother should be counselled about it though it is a challenge at this stage. After all, any addiction is difficult to get rid of without strong will power.
This case illustrates importance of ideal feeding practices. Breast addiction and bottle addiction are not uncommon but forced feeding during early life is a rule with many parents. Such habits come in the way of proper weight gain. Best time to prevent breast addiction is when it begins. While breast feeding may be continued in second year, one must consider risk of addiction leading to malnutrition and such an addiction is difficult to change.

Case 5

Two years old infant presented with poor weight gain since the age of 3 months. He was born after full term normal delivery with birth weight of 2.5 kg. He was exclusively breast-fed during first four months but was supplemented

with formula feed because of suboptimal weight gain during fourth moth. In spite of supplementation, child did not gain weight as per expected standard. He was also seen to lag behind milestones in all domains. He suffered from recurrent respiratory infections since the age of 9 months.

This history suggests multiple problems in this child besides poor weight gain. So, it must be a disorder that has disturbed multiple systems but has not been worsening. Such a static disorder may be either congenital malformation or chromosomal disorder or one time damage to brain early in life. There is no history suggestive of perinatal or postnatal events and so is unlikely to be one time damage to brain. It may be a syndromic disorder.

Physical examination revealed weight 8.5 kg (birth wt 2.5 kg), length 84 cm, head O 45 cm, generalised hypotonia and dysmorphic features with developmental delay suggestive of chromosomal abnormality.

It suggests the diagnosis of Down's syndrome that is confirmed by chromosomal study. Thus, poor weight gain in this child is due to **Down's syndrome** and needs no intervention as far as weight is considered. However, this child requires physiotherapy and occupational therapy.

Syndrome refers to conglomeration of several abnormalities. It is often a genetic disorder and may involve defects in multiple organs. Growth failure is common in many of them besides abnormal facies and congenital defects.

Case 6

15 years old child presented with fever followed by severe anorexia, vomiting and jaundice for last 4 days, Fever subsided over another few days but jaundice kept on worsening over next one month. At the end of a month, entire skin became yellow and there was severe itching. Child had lost appetite completely during this month and had lost 4 kg weight. Jaundice subsided thereafter slowly and so also itching disappeared and appetite was slowly returning to normal. However, child had not started gaining weight for which mother brought the child for opinion. On direct questioning, it was found out that mother had restricted many items from routine diet as advised by many friends. Physical examination showed no abnormality.

Laboratory investigations showed no active liver disease though serum proteins were lower than normal and IgG HAV was positive confirming diagnosis of hepatitis A recovered infection. So this child's poor weight gain was due to **undue restriction i**mposed by the family.

Mother needs to be counselled about importance of good nutrition during the stage of recovery from hepatitis A disease and harm caused by undue restriction of diet. It is a myth to restrict items of routine diet in a child suffering from acute hepatitis due to hepatitis A virus.

This case illustrates that prolonged illness would result in significant loss of weight as expected but on recovery, a normal child should start gaining weight. At such a time, patient should be encouraged to eat a balanced food to an extent possible without undue restriction. Myths prevail

about restriction of food during and after illnesses affecting intestines and liver in particular. It is often harmful.

Case 7

8 years old child presented with fever off and on and loss of appetite for last one month. There were no other symptoms. He was treated with two antibiotics over last one month but without any response. Several laboratory tests and chest x-ray were normal except mild anemia and ESR 75 mm.
History suggests subacute infection or non-infective disorder that has not shown any localisation even at the end of one month. It is probable that this is infective disease with hidden localisation such as TB or hidden lymphoproliferative disorder such as lymphoma. Such a child needs review of focused physical examination and repeat relevant tests.
Physical examination at the end of one month revealed enlarged liver, firm, not tender without signs of liver dysfunction in the form of pedal edema or splenomegaly. Neck veins were engorged but hepatojugular reflux was absent. This suggested block in superior vena cava and enlarged liver a manifestation of block in inferior vena cava. This gave away a diagnosis of constrictive pericarditis most likely to be due to Tuberculosis.
There was no easy way to confirm diagnosis of tuberculosis in this child as BAL was also negative.
Based on circumstantial evidence, anti-TB therapy was started and child was afebrile within two weeks.
Final diagnosis in this child was **tubercular constrictive pericarditis.**Unlike in younger children, poor feeding / eating

habits lead to poor weight gain in apparently normal and active children, weight loss or initially no weight gain of unknown origin is always a challenge in older children and adults that is likely to be due to a pathological cause. Hidden cause may be a chronic infection, organ dysfunction or malignancy. Often there are clues enough to suspect one of these conditions only if one is careful in history taking and physical examination.

Case 8

One year old child presented with poor weight gain and loss of appetite over last four months. This child was born after full term and normal delivery with birth weight of 3 kg. Was exclusively breastfed till the age of 6 months and then started on complementary feeds consisting of home-made gruel of cereals and pulses. Child was gaining weight well till the age of 8 months but had not gained any weight thereafter in last four months. Mother complaind of abdominal distension and occasional loose stools for which no medical help was sought. There were no other symptoms. Only localizing symptoms in this child were abdominal distension and occasional loose stools. On direct questioning, mother informed that abdominal distension waxes and wanes at times and so abdominal distension is due to accumulation of gas and hence suggestive of intestinal disease. As there is no history of fever, infection is unlikely. However, as this child developed loss of appetite, it must be non-infective inflammatory disorder that has resulted in malabsorption. As onset of this problem nearly coincides

with introduction of complementary feeds, it may be related to one of the food items, wheat being the common culprit. So, it may be celiac disease.
Physical examination revealed gaseous abdominal distension and moderate pallor. There were no other abnormal findings. This supports intestinal disorder but cause should be found out by further investigations.
It was confirmed to be **celiac disease** with presence of IgA tTG antibodies. Further confirmation can be achieved by duodenal biopsy though avoiding wheat may show improvement and is a rational way of proving the diagnosis in absence of duodenal biopsy.
Malabsorption is a hidden cause of poor weight gain or weight loss. If malabsorption is immune mediated, loss of appetite is a predominant symptom with or without fever. However appetite may be normal or even voracious in spite of loss of weight if malabsorption is due an enzyme defect,

Take home message

Constant weight in infancy should be taken as loss of weight. However, in subsequent years, constant weight for a few months is not abnormal though throughout childhood, weight must increase over time. Poor weight gain in infants and toddlers is often a misconception and in such a situation, child is always active and happy. Even hidden pathology leads to disturbed well-being and is noted in terms of poor appetite, lack of energy and change in behavior such as lethargy or irritability. Growth chart is the best objective measure that helps not only to differentiate normal from

abnormal but also suggests a probable diagnosis by analysing onset, duration and progress of weight loss.

MCQs

Q 1 In assessing poor weight gain, this is the most important point in the history

A) Mother's perception
B) Appetite
C) Activity and energy level
D) Change in behavior

Q 2 In assessing poor weight gain, this is the most important point in clinical examination

A) Present poor weight
B) Tracing weight centiles
C) Abnormal physical findings
D) All of the above

Q 3 Weight gain may be poor at one year in this situation

A) Birth wt 1.8 kg in a neonate born after 32 weeks gestation
B) Birth wt 1.2 kg in a neonate born after 32 weeks gestation
C) Birth wt 2.4 kg in a neonate born after 38 weeks gestation
D) All of the above

Q 4 Which of the following statement is WRONG? Weight gain may be poor in a normal child because

A) Breast addiction B) Myths and beliefs about feeding
C) Constitutional D) None of the above

Q 5 Which of the following statement is WRONG? This cause of poor weight gain may be hidden

A) Tuberculosis
B) Insufficient breast milk
C) Celiac disease
D) Syndromic child

Answers to MCQs

Q 1 C, Q 2 B, Q 3 B, Q 4 D, Q 5 B

2.33. My child is short!

Introduction

Height (of) achievement is always a dream and hence short stature in a child is a concern for parents. Length / height depends upon nutrition in infancy, thyroid and growth hormone beyond infancy in childhood and sex hormone during adolescence besides genetic factors. Once puberty is achieved, growth nearly stops but timing of puberty varies in individuals. If puberty is delayed, it ends in tall stature while if puberty is achieved early, it results in comparatively a short stature. Girls active in sports tend to achieve puberty bit later than those who tend to be sedentary. In an adequately nourished infant with normal status of thyroid function, growth hormone and sex hormone, height will be decided by genetic factors alone and it cannot be changed. Thus, a child of short parents is most likely to be short. It is a myth that height increases with pull-ups or any other exercises or supplements.

Is there any difference between length and height? How are they measured?

For practical purposes, both are nearly equal though length is slightly more than height by 0.5-0.7 cm. This is because on standing, there is a small reduction in intervertebral spaces. Measuring length or height accurately is not as simple as one thinks. For accurate measurement of height, person must stand erect with occiput, buttocks and heels touching against stadiometer (ruler on a stand) and horizontal headpiece resting on head. Length is measured with infant lying supine on an infantometer (horizontal ruler with a movable foot-end piece) with head placed at one end and foot-end moved to touch heels with outstretched legs. Ideally measurement is considered accurate only if three consecutive readings are same. However, in routine practice, single reading suffices as interpretation does not vary significantly. Though for research purposes, utmost accuracy is vital.

Average length / height gain in childhood

Infant gains 25 cm in length in first year of which 15 cm are gained in first 6 months and 10 cm in later half of first year. In second year, length is increased by 12 cm and thereafter 5-6 cm per year till adolescence. During adolescence, height increases by 10-12 cm per year. Though there are many variables and so, even normal children grow at different velocity.

Can adult height be predicted in childhood?

In normal healthy child, Tanner formula can roughly predict adult height taking into account mid-parental height with a range of 8.5 cm on either side.
Target height = (father's height + mother's height) divided by

2 + 6.5 cm for boys and minus 6.5 cm for girls with a range of 8.5 cm upwards or downwards. Indian Academy of Pediatrics has published simplified growth chart for calculation of mid-parental height.

Confirm short stature

Height below 3rd centile on growth chart or 2nd standard deviation below the mean for that age is considered short stature meaning child is short in height. The best way to judge is to monitor height velocity over time period or to follow the trend of height gain. It is only when short stature is confirmed that one must try to find out probable cause.

Common causes of short stature

Early onset of chronic malnutrition is the most common cause seen especially in underprivileged community. In such a situation, child will never attain normal adult height. Other pathological causes include endocrine disorders such as Cushing's, hypothyroidism and growth hormone deficiency, syndromic abnormalities (Down, Noonan, Russell-silver, Prader-Willi and Turner syndrome n girls),chronic systemic diseases, chondrodystrophy such as achondroplasia and rickets. In absence of any such pathology, short stature may be genetic or constitutional. A child with constitutional short stature does attain normal adult height with spurt in height gain during adolescence while a child with genetic short stature does remain short in adult life.

Clinical approach to short stature

First step is to confirm short stature. Parents often compare growth of their children with those of the peers or other children in extended families. Even two children of the same parents vary in their growth pattern.
Once short stature is confirmed, it is important to find out time period of origin of the problem. If it has started very early in life, it is mostly primary nutritional or at times secondary nutritional due to severe systemic diseases. Child with systemic disease resulting in short stature is sick looking with many other symptoms and signs besides short height. Most of other conditions presenting with short stature do not manifest sickness. Growth hormone deficiency typically presents after the age of 3-4 years of age. Delayed cognition is typical of hypothyroidism along with abnormal facies and are also in mucopolysaccharidosis with hepatosplenomegaly and bony defects. Bony defects are typical features of chondrodystrophy and rickets. Abnormal facies, delayed development and structural malformations in different organs are features of syndromic problems. Proportionate or disproportionate trunk and limbs may help in assessing bony abnormalities. In absence of any of the above-mentioned abnormalities, short stature may be genetic or constitutional. Assessment of mid-parental height and periodic growth monitoring including height velocity would help differentiating one from the other.

Investigations in a child with short stature

Bone age is estimated radiologically by Tanner-Whitehouse method that can report bone age in weeks. Routinely, X-ray

of one hand is used to estimate bone age because it includes 20 bones in one single plate. Routine X-ray of bones fails to assess bone age correctly as it can at best offer a range in years that is not useful. Bone age denotes bone development or maturity. Height age is the age at which normal child achieves the said height. It denotes bone growth. For example, if six year old child has height of 100 cm then his height age is 4 years because normal child attains 100 cm at 4 years of age. Chronological age is an actual age in years and months. Comparing chronological age with bone and height age and assessing height velocity gives a clue to probable diagnosis. Bone age is delayed in hypothyroidism and growth hormone deficiency and may be mildly delayed in malnutrition. If chronological age is more than height age which is more than bone age, it suggests hormonal deficiency that can be further proved by specific tests. In fact, hypothyroidism should be suspected at birth by cord blood tested for TSH and further confirmed by free T4. Early detection of hypothyroidism is important to avoid delayed brain development as thyroid supplements can be started before brain function is affected. If chronological age is equal to height age but more than bone age, it is likely to be genetic or due to small birth weight for gestational age (both the conditions have normal height velocity) or chromosomal disorder (abnormal height velocity). If bone age and height age are equal but less than chronological age, it is likely to be constitutional (normal height velocity) or chronic diseases (abnormal velocity). Once a probable diagnosis is made, specific tests can be planned to confirm the diagnosis.

Management of short stature

There are no supplements or physical exercise that increases height. So, management depends on the cause. Growth hormone replacement therapy is now freely available and would help a child with GH deficiency. It is also approved for idiopathic short stature and in children with some of the syndromic problems such as Turner syndrome in girls. It may also be useful in severe systemic diseases with growth failure. Thyroid replacement should start at the earliest as mentioned above. Vitamin D deficiency rickets can be treated with Vitamin D supplements. Systemic diseases may have specific treatment if available.

Take home message

Short stature, once confirmd, is best assessed by periodic monitoring of growth and plotted on standard chart. It helps to pick up early deviation in growth parameters and would lead to rational management if possible. Treatable conditions should not be missed and in particular, hypothyroidism is best diagnosed at birth with cord blood screening. Growth hormone replacement is now possible and economically affordable. After all, height (of) achievement is everyone's aim in life.

MCQs

Q 1 Which of the following statement is WRONG? Short stature is considered if

A) Single height measurement falls below 3rd centile on growth chart

B) Single height measurement falls below 2nd standard deviation from the mean at that age
C) Only if height velocity is abnormal
D) All of the above

Q 2 Which of the following statement is RIGHT? Growth is proportionate in

A) Hypothyroidism
B) Growth hormone deficiency
C) Rickets
D) Mucopolysaccharaidosis

Q 3 Delayed development is a feature of

A) Hypothyroidism
B) Syndromic problems
C) Mucopolysaccharaidosis
D) All of the above

Q 4 Which of the following statement is WRONG? Bony defects are seen in

A) Rickets
B) Achondroplasia
C) Mucopolysacchariadosis
D) None of the above

Q 5 There are no abnormal physical findings in

A) Genetic short stature
B) Constitutional short stature
C) Idiopathic short stature
D) All of the above

Answers to MCQs

Q 1 C, Q 2 B, Q 3 D, Q 4 D, Q 5 D

2.34. Everyone wants to reach “height”

Basic concepts for clinical application

Short stature is defined as height 2 standard deviation below the mean for the age and must be confirmed before embarking on finding the cause. Cause of short stature can be guessed clinically. Detailed history should include sick or not sick, proportionate or disproportionate, bony defect of not, mental development normal or abnormal, abdominal distension or not and family history of short stature and physical examination for malnutrition, abnormal facies and other congenital malformations Linear growth measurements including velocity of growth and mid-parental height offer clues to probable cause of short stature. Further, comparison between chronological age, height age and bone age (as measured radiologically) along with height velocity can almost confirm the cause of short stature. Hormonal estimation or other specific tests for storage disorders may be necessary to stamp the diagnosis.

Case based discussion

Case 1

Two years old child was referred for not gaining weight and height as compared to previous child. He was irritable and not very active since last one year. There were no other

specific complaints. He was growing well till about 8 months of age but thereafter his weight and height gain was very poor. On direct questioning, this child was exclusively breast-fed till one year of age and thereafter mother tried to introduce family food but child would refuse but insist on breast feeding. His intake of semisolid food was very little. His elder brother and parents were of average height.
History of gradual faltering of weight and height after the age of 8 months and irritability with poor nutritional intake suggests **primary malnutrition** as the cause of growth failure.
Physical examination confirmed poor weight and height and there were no other significant abnormal findings.
There is no need for any investigations in this child and all that is required is to de-addict him from breast feeding and ensuring adequate intake of family food together with supplements of vitamins and minerals.
Parents often complain of poor weight and subnormal length is noted by the doctor. On direct questioning, parents may have observed poor length. It is clear that when both weight and length are faltreing but weight more affected than length, problem is related to poor nutrition – either primary or secondary. Primary malnutrition is considered in children below age of two years. Generally, no investigations are necessary in primary malnutrition but It is important to note that bone age is minimally delayed in malnutrition but it is easy to rule out endocrine disorders.

Case 2

Two years old child was referred for recurrent fever, cough and loose stools since the age of 3 months and also not gaining weight and length. He was born after full term normal delivery with birth weight of 3 kg and was exclusively breast-fed in early months. However, while on exclusive breast feeds, he developed first episode of fever and cough and thereafter continued to suffer infections at periodic intervals. Over time, parents noticed poor weight and height gain. This infant on exclusive breast-feeding started suffering from recurrent febrile illnesses and so it suggests abnormal host status. In view of two systems involved – respiratory and intestines – it may be either immune deficiency disorder or cystic fibrosis. It is likely to be progressive disorder as evident by poor growth. So **secondary malnutrition** due to host abnormality is the cause of poor growth.

Physical examination revealed sick looking child, irritable with evidence of respiratory tract infection besides poor weight and length, weight affected more than length. It is typical of severe malnutrition. This child needs investigations to diagnose exact cause of recurrent illnesses. This child was proved to be suffering from cystic fibrosis resulting in secondary malnutrition.

Malnutrition starting in early infancy in an infant born with normal birth weight and on exclusive breast feeds is always due to abnormality in the host, either immunological, structural (congenital malformation specially in urinary tract or lungs) or functional (as in cystic fibrosis). There may be few clues on history and physical examination but investigations are necessary to make a final diagnosis.

Case 3

Eight years old female child presented with not gaining height noticed over last few years. Parents realized it when younger sibling outgrew this child's height. There were no other complaints. This child was healthy, active and doing well in his studies. Parents were of average height and there was no family history of short stature.

History rules out genetic cause, chronic malnutrition and hypothyroidism. Growth hormone deficiency, constitutional delay, turner syndrome and skeletal disorders are possibilities that can be differesntiated on physical examination.

Physical examination showed weight 22 kg, height 110 cms, proportionate limb-trunk growth. Child looked happy and active and there were no abnormal findings.

This rules out turner syndrome (short neck, increased cubital angle, wide spaced nipples, high arch palate) and also skeletal disorders. So now we are left with growth hormone deficiency and constitutional delay. This can be differentiated on height velocity on growth chart. He was diagnosed as **constitutional delay** based on maintaining same height velocity each year.

Growth hormone deficiency results in decreasing height velocity (height velocity. Normal height gain each year beyond 2 years of age till adolescence is around 5-6 cm per year and in GH deficiency, it goes on decreasing every year, generally starting from 2 years of age.

It is not uncommon to find that one of the parents were short in height in childhood but picked up to normal height

during adolescence growth spurt. This is typical of constitutional short stature.

Case 4

Eight years old child was referred for short stature. Parents had noticed it since the age of 3-4 years but thought he might catch-up with improving nourishment and exercise. He has no other symptoms; he was active and good in studies. His both parents were short.

History suggests genetic short stature though we must make sure that there is no other cause because genetic or familial short stature is a diagnosis of exclusion.

Physical examination showed weight 20 kg, height 110 cm and no other abnormality. We need to look at growth chart. It showed height velocity of 4.5 cm each year and it was maintained the same over years. Mid-parental height revealed that this child's target height (height expected at adulthood) fell within mid-parental height. This confirms diagnosis of **genetic or familialshort stature**. There is no need for any investigations, however in case of doubt, one may estimate bone age and compare it with chronological and height age.

A child born to parents with short stature may not necessarily be short in adulthood. Hence it is always necessary to confirm absence of any other cause for short stature in such a child.

Case 5

Six years old child presented with developmental delay noticed since the age of 9 months and short stature noticed

since the age of 3 years. Parents were more worried about developmental delay and so did not seek advice about height. Unfortunately, doctor had not measured height either and so it went unattended. On direct questioning, he was constipated and passed hard stools once in 2-3 days. Combination of developmental delay and constipation along with short stature suggests multisystem affection starting early in infancy. It favors the possibility of hypothyroidism or storage disorder such as mucopolysaccharides.

Physical examination showed weight 21 kg, height 96 cm, dysmorphic features, coarse dry skin, hoarse voice, puffiness of eyelids, umbilical hernia, abdominal distension loaded with feces and developmental delay mainly in cognition. These findings are typical of **congenital hypothyroidism**. His chronological age is six years, height age is near 4 years.

It was confirmed with low T3, low T4 and high TSH. Bone age in this child was 3 years – lower than height age which in turn was lower than chronological age.

He needs thyroxin supplements for life. However, his mental condition would not improve, height may pick up a little but he would always remain short though his constipation would improve on treatment.

Congenital hypothyroidism ideally is diagnosed at birth by cord blood screening as it offers a chance for normal mental development as well as target adult height. Such screening is now done routinely in most of the general hospitals in India. It is cost effective. In absence of such screening, one must look for clues for early diagnosis such as delay in passing meconium at birth, subnormal length at birth with wide

posterior fontanel suggestive of delayed bone growth. If not picked up at that age, permanent damage is most likely.

Case 6

Six years old child presented with short stature noticed two years back when younger sibling had surpassed elder brother's height. There were no other complaints. Initially, parents were not much worried as they both were also short but now the child himself was concerned about it. So, parents sought medical advice. On direct questioning, parents were sure that this child looked normal in height in first few years. History suggests height faltering after first few years and progressively going down but child maintained good health. This history may suggest familial or genetic stature but if height velocity is progressively going down, it may be due to growth hormone deficiency. Growth chart would offer clues. Physical examination showed weight 21 kg, height 102 cm without any other abnormality. His height age is four and half years. However height velocity was going down every year from the age of 3 years – initially it was 6 cm in fourth year, 5 cm in fifth year and now 4 cm in last year. It strongly suggests **GH deficiency**. However, one should confirm it with appropriate tests as familial short stature is a possibility. Investigations ordered for him included serum IGF 1 – insulin like growth factor that correlates well with level of growth hormone. Growth hormone can be estimated with a stimulation test in which basal level is assessed followed by a level after a challenge with drug like clonidine. Absence of increase in GH after a

challenge is diagnostic of GH deficiency.
Growth hormone is now freely available, very safe though cost is a factor though it is getting more affordable. Ideally therapy should be continued till target height is achieved.

Case 7

Two years old child presented with poor height and weight since early infancy. This child was born after full term with birth weight of 1.6 kg. There was no major problem at birth though direct breast feeding was not possible for first few days and had to be fed with spoon. Over next few months, mother noticed poor growth and after the age of six months, he suffered recurrent respiratory or intestinal infections. History suggests growth failure that started in utero and further contributed by recurrent infections. It was not an isolated short stature but chronic severe malnutrition. Physical examination showed weight 5.5 kg, length 70 cm, head O 44 cm, moderate pallor, poor subcutaneous fat, poor muscle mass. There were no other localizing signs.
He had gained 4 kg weight (normal neonate gains 4 kg in first 6 months) so his weight age is just 6 months. We did not now his length at birth, however considering it to be around 45 cm he had gained 25 cm (normal neonate gains 25 cm length in 12 months), so his height age is around 12 months. His head O at birth is not known but considering it to be near normal or slightly lower than normal, it may have been 32 cm, so he has gained 12 cm (it corresponds to one year of age). Thus. he is lagging behind in all three growth parameters. He is also developmentally subnormal. It is

typical of intrauterine growth restriction. However, his weight is much lower as compared to his height and head O, it is because of his recurrent illnesses with poor nutritional intake. So his growth failure is due to **IUGR** further worsened by **poor nutrition.**

Ideally, he does not need any specific investigations though one must make sure that there is no other cause for recurrent infections such as immune deficiency.

Fetal malnutrition starting in first trimester of pregnancy affects brain growth as evident by subnormal head O. This would result in learning disability in such a child besides remaining short in adulthood.

Case 8

Six years old female child presented with short stature noticed over last four years and also mild developmental delay. There was no history of constipation or any frequent illnesses. Short stature with developmental delay suggests abnormal growth and brain development. One may consider hypothyroidism but absence of constipation is against it. However not every child with hypothyroidism would have all typical symptoms. Other possibility is any syndromic abnormality that shares both growth and brain dysfunction. Physical examination showed weight 18 kg, height 98 cm, wide webbed neck, low set ears, wide cubital angle, presence of heart murmur and mild developmental delay. Dysmorphic features with heart murmur in female child is typical of **turner syndrome**.

This was confirmed by chromosomal study showing 45 X

pattern. Such children are often detected late in childhood or later because of delayed sexual maturity. There are many other syndromes with short stature along with other abnormalities. As a rule, short stature in a female child demands to rule out Turner syndrome.

Take home message

Growth monitoring with periodic measurements of weight, length / height (till puberty) and head O (during first two years of life) is important in all children. Of course, weight should be monitored periodically throughout life. Growth chart with periodical plotting of measurements help not only in the diagnosis of short stature but also in evaluating cause of many other diseases as origin, duration and progress of diseases is obhectively evident.

MCQs

Q 1 Which of the following statements is WRONG? This child with short stature also has developmental delay

A) .Hypothryoridism
B) Turner Syndrome
C) Constitutional delay
D) Mucoplolysaccharide defect

Q 2 This child of short stature has proportionated limb-trunk growth

A) GH deficiency
B) Hypothyroidism
C) Chondrodystrophy
D) Mucopolysaccharide defect

Q 3 This child of short stature is likely to catch up to normal target height

A) Constitutional delay
B) Familial short stature
C) Hypothyroidism
D) Turner syndrome

Q 4 Weight is more affected than height in this child of short stature

A) Chronic malnutrition
B) GH deficiency
C) Hypothyroidism
D) Constitutional delay

Q 5 This child of short stature does not need any investigations and / or therapy

A) Turner syndrome
B) Constitutional delay
C) GH deficiency
D) Rickets

Answers to MCQs

Q 1 C, Q 2 A, Q 3 A, Q 4 A, Q 5 B

2.35.Ensure child is developing well

Introduction

Childhood is the period of not only just the growth but also of development. Growth refers to increase in physical size while

development denotes maturity of function. As much as growth is mainly measured by increase in weight, height and head circumference reflecting increase in brain size, development is assessed in four major domains – gross motor, find motor, cognition (social-adaptive) and language. At times, only one of the domains is delayed, it is referred to as differential developmental delay. If two or more domains are affected, it is referred to as global developmental delay. Global developmental delay is a result of brain dysfunction but may not be equal in all domains. However isolated developmental delay in one domain may result in spite of normal brain function as in case of gross motor delay due to musculo-skeletal disorder and language delay due to hearing impairment.

Maturity of brain functions mostly occurs in first two years of life and to a small extent may continue till five years of age, thus it is most vital to assess development in first two years of age though mild dysfunction may manifest later in childhood.

Developmental milestones

There occurs sequential achievement of milestones in each domain.

<u>Gross motor domain</u>

Head control, rolling over from prone to supine and thereafter in both the directions, sitting with support and thereafter without support, crawling (some children may skip crawling), stand with support and thereafter without support, walking, going up the staircase and thereafter down

the staircase, running, hopping and jumping

Fine motor domain

puts hands to mouth, holds object given in the hand, reaches for objects when shown, transfer object from one hand to other, releases object when asked for, pincer grasp (picking up small size object with two fingers), drinks from a cup, turns pages thereafter even a single page at a time, can thread a bead, use scissors to cut a paper

Cognitive domain

keeps quiet when picked up, focuses on eyes, follows moving object, recognizes mother, social smile (smile on response), recognizes other family members, responds when called, pick-a-boo, understands 'no', says bye bye, follows simple orders, points to body parts, scribbles and sequentially achieve ability for strokes, circle, cross, square, triangle etc, puts on one block over the other thereafter tower of several blocks, imitation play, turns pages of a book thereafter able to turn each page separately, recognizes shapes and colors, identifies objects in pictures

Language domain

Receptive language develops before expressive language. Receptive language refers to understanding the language while receptive language refers to response either in the form of gestures (non-verbal) or spoken words (verbal) that is speech. Expressive non-verbal language (gesture or sign language) develops before verbal language (speech). Even a newborn responds to human words especially high-pitched sounds of the mother and stops crying when talked to. This represents early receptive language. Cooing, babbling follow

thereafter. When called by name, infant of 4-5 months of age looks towards the direction of the caller. It represents expressive language. This is followed by monosyllables, bisyllables and then on to words, meaningful words, sentences. Many of these milestones involve more than one domain such as cognition, language and fine motor together.

Developmental assessment in office practice – a screening test

Detailed developmental assessment is necessary only when developmental delay is suspected on screening test. Screening test can evaluate developmental status at 3 months of age (head control, recognizing mother, social smile, cooling / babbling, hand to mouth), at 12 months of age (sitting definitely and also may be standing, pincer grasp, response when called by his name, pick-a-boo) and finally at 18 months of age (walking, few words a must).
Assessment at 3 months of age picks up developmental delay if any arising from antenatal or perinatal events (usually head size is smaller than expected), that at 12 months helps to pick up minor defects and autism spectrum disorder (disorder of social communication presents as language delay, absence of eye to eye contact, purposeless hyperactivity and repetitive behavior) and that at 12 months of age to confirm autistic spectrum disorder and other minor delays.

Developmental delay – static or progressive?

Static developmental delay results from one time damage to the brain that may have occurred before, during or after birth, especially during first two years of life. Early

recognition and prompt management can improve developmental status at least to some extent. Progressive developmental delay is due to active disease and would worsen over time if not recognized early enough and managed in time. Ideally such defects need to be picked up on routine screening at birth or early in life. However not all such progressive disorders are amenable to diagnosis, control or correction and hence outcome are usually poor.

Common causes

Prenatal – congenital malformations of brain, chromosomal disorders, congenital infections (maternal infection transferred to the fetus through placenta) and adverse effects of drugs or radiation exposed to mother during pregnancy.
Perinatal – hypoxic ischemic brain injury, bilirubin enephalopalhy (result of severe unconjugated bilirubinemia) problems in preterm neonate such as periventricular hemorrhage, periventricular leucomalacia, hypoglycemia, polycythemia), hypoglycemia in neonate born to diabetic mother
Postnatal – intracranial infections, severe head injury, hemorrhage due to coagulation defects, inborn errors of metabolism, hypothyroidism, Autism spectrum disorder

Investigations

MRI of brain helps to detect site of damage that may offer a clue to probable etiology. Genetic tests including chromosomal study and metabolic tests are necessary in suspected conditions. Classical presentation of cerebral palsy

and autism spectrum disorder may not need any specific tests. Assessment of vision and hearing may demand tests such as VER – visual evoked response and BERA – brainstem evoked response audiometry.

Prevention

Low birth weight and preterm delivery may be preventable with proper antenatal care at least in some situations. Blood group Rh incompatibility is preventable by administration of antibody to Rh –ve mother immediately after the birth of Rh +ve neonate. Ideal intranasal care may prevent damage such as due to hypoxic ischemic encephalopathy. Prevention of brain damage due to inborn errors of metabolism can be prevented by neonatal screening for specific disorder and now available for about 6 common conditions including hypothyroidism in many centers in India. However western countries routinely test for 20 conditions.

Management

Physiotherapy to prevent contractures, occupational therapy to make a child independent as much as possible, speech therapy and sensory integration (coordinated use of sensory inputs) are the mainstay of management that should be started as early as possible for better outcome. Behavioral modification may be necessary. Other areas that deserve attention are constipation, gastro-esophageal reflux, nutritional deficiencies – especially vitamin D deficiency, general hygiene, immunization, visual and hearing aids if necessary and special education.
Thyroid hormone replacement therapy should be started at

birth or as early as possible. Drugs are necessary only in case of epilepsy or rarely to control excessive hyperactivity in a child with autism spectrum disorder.

Take home message

Developmental screening is important for early detection of delay if any. Earlier in life a developmental delay is pricked-up, more is the chance of better outcome. Cause of developmental delay can be guessed on clinical parameters and may need confirmation by specific tests. Management mainly consists of non-pharmacological therapies and drugs are rarely needed. Outcome of progressive disorders is generally poor. Ideal antenatal and perinatal care may prevent some of the causes of static developmental delay. Prevention of damage due to metabolic defects is possible only when there is an index case in the family or by universal neonatal screening.

MCQs

Q 1 This gross motor milestone may be skipped in a normal child

A) Rolling over
B) Sitting without support
C) Climbing staircase
D) Crawling

Q 2 This is the earliest age at which child may accidentally inhale foreign body

A) One year
B) 8 months

C) 5 months
D) 3 months

Q 3 Global developmental delay is considered when

A) Any of the two domains are affected
B) Any of the three domains are affected
C) All domains are affected
D) All of the above

Q 4 Which of the following statement is WRONG? This condition may manifest with language delay in spite of normal hearing

A) Tongue tie
B) Global developmental delay
C) Autism Spectrum disorder
D) All of the above

Q 5 This disorder can be diagnosed and prevented at birth

A) Down's syndrome
B) Congenital malformation of brain
C) Congenital hypothyroidism
D) Birth injury

Answers to MCQs

Q 1 D, Q 2 B, Q 3 D, Q 4 A, Q 5 C

2.36Early pickup of developmental aberration for timely action

Clinical application of basic concepts

Development refers to functional maturation and is assessed in terms of four domains – gross motor, fine motor, cognitive or social adaptive and language. Brain is the major organ responsible for development in all domains. However, in spite of normal brain function, developmental delay could be due to affection of other systems. Gross motor delay could be due to musculoskeletal problems and language delay from hearing impairment. Visual defects can lead to pseudo-developmental delay. First two years of life are crucial for brain development and also to small extent up to five years. Thus, major defects in development manifest early in life though minor defects such as learning disability may be detected later. It is important to monitor development in early years of life. Beyond five years of age, school performance can detect aberrations if any. Grossly, developmental assessment at the age of three months can detect major abnormality arising out of antenatal or perinatal events, at one year of age, autism spectrum disorder and hearing impairment can be made out and further confirmed at eighteen months of age. Developmental delay in two or more domains is referred to as global developmental delay. It is important to decide whether the delay is static (milestones are achieved at same slow speed) suggests onetime damage or progressive (development speed is slowing down and so worsening) indicates ongoing damage.

Case-based discussion

Case 1

Mother of one year old infant complained about her child not able to sit without support, did not speak any words and was not gaining weight well. On further enquiry, mother was 35 years old when this child was born after full term with normal delivery but his birth weight was only 2 kg. He did not cry at birth but had spontaneous breathing on stimulation and was normal thereafter. Mother on direct questioning said that he appeared to be bit slow in his activity but it did not bother her much. He had been constipated. He held his head at 6 months and rolled over at 10 months. He recognized her at 3 months of age, could reach for the object by 6 months of age, he is very social and goes to anyone who picks him up and is a loving happy child.

History suggests that he has a developmental delay in all four domains that is static and so it must be due to onetime damage. There has been no significant problem at birth and so cause of this delay must be due to antenatal factors. This mother delivered at the age of 35 years that carries high risk of chromosomal disorder.

Physical examination revealed weight 7 kg, length 70 cm, head O 40 cm, dysmorphic features suggestive of Down syndrome, microcephaly, generalised hypotonia, developmental quotient around 50%, no other neurological abnormality, grade 3 systolic murmur, no other abnormality in hear or any other systems.

This child has static global developmental delay with congenital heart defect and dysmorphic features suggest diagnosis of **Down syndrome**. It can be confirmed with chromosomal analysis and parents must be counselled about

avoiding future pregnancies. In fact, they should have been counselled before or during early pregnancy about prenatal tests to diagnose such defects.
There is no specific treatment but child can do better with physiotherapy and occupational therapy. At present, heart defect does not need any intervention except periodic monitoring for timely action. He may need diet advice and relief for constipation. Such children have borderline developmental quotient and can be educated to an extent. But ultimately, they should be trained in areas that need more physical than intellectual inputs.

Case 2

Mother of one year old infant complained about developmental delay in her child. He had started rolling over at 11 months of age, barely reached for the object, could recongise mother and other family members but had no vocalization. He was generally irritable and constipated. This child was born after full term with normal delivery in a primipara at the age of 22 years and had no problem at birth but his weight was 1.5 kg. Mother suffered from severe vomiting during first three months and later developed eclampsia for which she had to be hospitalised.
History suggests severe intrauterine growth restriction as a result of maternal illness during pregnancy. As there was no significant birth problem, cause of developmental delay must be due to antenatal factors and one of such factors could be poor nutrition right from early fetal life. Any other cause needs to be considered based on physical examination.

Physical examination showed weight 6 kg, length 68 cm, head O 39 cm, signs of calorie malnutrition and anemia, irritable child, generalised hypotonia with poor muscle mass, no localizing signs in any other systems.
This child is severely malnourished – wasted (very poor weight) and stunted (poor length) with microcephaly suggesting severe malnutrition that must have started very early in fetal life as also evident by history. So, this child's developmental delay is due severe fetal malnutrition **IUGR.** There is no need for any investigations. Treatment is to improve nutrition, though this child will remain short and small in stature and also will have some brain dysfunction. However, with improved nutrition and adequate stimulation, this child's disability can be minimized to an extent possible. It is important to realize that it is only when malnutrition sets in so early in fetal life that results in developmental delay. Otherwise, any severe malnutrition resulting during later part of pregnancy or after birth is not likely to affect brain function severely though mild defect such as learning disability may manifest especially if malnutrition started early during infancy.

Case 3

One year old adopted child presented with developmental delay. Parents had taken custody of this child two months ago and were concerned about this child not standing without support and not speaking any words. Though he seemed to understand spoken words. He did not show stranger anxiety. Otherwise, he appeared to be happy,

feeding well and had gained one kg weight in last two months. Apparently, they were not informed about earlier milestones but were told that the child was normal. History suggests that this child is lagging behind in milestones and his present development is like a 9-month-old child. However, there are no clues to probable cause.

Physical examination showed weight 8 kg, length 72 cms, head O 42 cm, no other significant abnormality.

This child's weight and length is appropriate for 9 months and his head O is on the lower limit of normal.. His milestones are not much behind the expected level and considering that this child was reared for first 10 months in a shelter home, there may have been lack of stimulation and it could explain mild delay, especially in speaking. Language development occurs first in receptive domain and then follows with expressive ability. This child did respond to spoken words which means that he had developed receptive language. Thus, our impression would favor a normal child with **lack of stimulation** in early infancy. It is expected that this child would pick up lagging milestones within short time with interaction with parents.

This case illustrates importance of early stimulation in terms of communicating with the neonate and infant. It is the best way to achieve maximum developmental potential. This is equally important in normal and abnormal neonates and infants. In fact, communication with the fetus during pregnancy has shown benefits, though not easy to evaluate.

Case 4

One and half years old child presented with complaints of not able to walk without support. He was born after full term with normal delivery, birth weight of 3 kg and had been normal without any significant problems. He was exclusively breast fed for first 6 months and then was started on complementary feeds while continuing breast feeds that he takes even now. He gained weight well in first year but had not gained only 800 gm in last 6s months. His had achieved all milestones in time during first 6 months but had started sitting without support only at one year and then standing at 14 months. Doctor had said that it was within normal limits. But now that he was not able to walk without support, parents were worried. His other milestones were all normal as he could speak words with meaning and respond appropriately to his age.

History suggests well-nourished child without any disease and is lagging behind only in gross motor milestones sometime after first 6 months and his other domains are all normal. So, he has an isolated delay in gross motor milestones starting sometime after the age of 6 months in spite of normal brain function. Obviousl,y fault lies in musculoskeletal system and not in peripheral neurological system as he can stand and so does not have any paresis. Acquired motor delay in a well-nourished child is often due to vitamin D deficiency rickets contributed by predominantly milk diet. Physical examination showed weight 11.5 kg, length 78 cm, head O 48 cm, signs of rickets, no other abnormality. Diagnosis of **vitamin D deficiency rickets** was confirmed by biochemical (low or normal serum Ca, low

serum P and high serum alkaline phosphatase) and radiological tests (cupping and fraying at the epiphyseal end of bones with demineralization)
He needs therapeutic dose of vitamin D– 6 lakhs unit to heal rickets and repeated twice more to replenish store. Ideally. vitamin D supplements should be given every day for next three months but often lacks compliance and so massive dose is often prescribed. His parents should be counselled about diet. He may have been breast-addicted and so consumes mainly milk without much solid food.

Case 5

Two years old child presented with inability to speak and not responding age-appropriately. Though his motor milestones were normal. He was born after full term with normal delivery and had achieved all the milestones in time except speech development. When consulted for this delay, doctor had said that such a delay was familial in this child as his uncle also was a late talker. They were advised to check hearing ability and was found to be normal. But now a year had passed without any improvement and so parents were worried. On direct questioning, it was realized that he was hyperactive, often kept on moving without purpose, engrossed in activities with repeated maneuvers, had no eye-to-eye contact, would not respond when called by his name and would not follow any orders. However, he would sit for long time in front of television. Unfortunately, this aspect remained ignored. So, this child has communication disorder with attention deficit.

Physical examination confirmed his abnormal behavior but had no abnormality in any other system.
Diagnosis of **Autism spectrum disorder** was confirmed by standard tests done by psychologists – DSM 5 – developmental manual test.
He would need long term therapy under the guidance of developmental pediatrician. Surely, he would improve at least to some extent but only with strong commitment from parents. Ideally, he should have picked up at about one year. Simple screening test for autism spectrum disorder is to see whether child responds to his name being called at least by one year. If there is no response, one should check hearing ability and brain function in other domains and if found to be normal, such a child should be referred to a developmental pediatrician. If diagnosed early and treated properly, there would be much better outcome. There are no drugs for such a problem though occasionally hyperactivity may need some medications. Such disorders are on the rise, may be due to awareness but also may be due to nuclear families with limited social interaction with inappropriate use of TV, mobile phones, computers.

Case 6

Mother of one year old child complained of inability to hold head, not recognizing her, no smile or cooing. This child was born after full term cesarean section delivery done for fetal distress. He did not cry at birth, required oxygenation and ventilation for next 4 days, had few seizures on D2, was on IV fluids and subsequently fed through nasogastric tube. He also

developed sepsis that was treated with antibiotics. He was discharged on D 20. He was prescribed anti-convulsant drugs. He had three more episodes of seizures and had poor weight gain. He was on physiotherapy without much benefit.
History clearly suggests severe **hypoxic-ischemic birth injury** resulting in severe developmental delay and seizure disorder.
Physical examination showed malnourished infant with global developmental delay, microcephaly and severe spasticity with signs of bilateral upper motor neuron lesion.
It confirms diagnosis of spastic cerebral palsy with remote symptomatic epilepsy (due to one-time damage) and severe malnutrition.
In such a typical presentation, there is no need for neuro-imaging as it does not add to any more relevant information. Neuro-imaging is reserved in a child suspected to have cerebral palsy only in case of absence of microcephaly, presence of abnormal birth history or family history of similar problem. Treatment is palliative in the form of physiotherapy to prevent contractures, occupational therapy and anti-epileptic medications, nutritional rehabilitation and care of non-neurological problems such as constipation, gastro esophageal reflux, skin and bladder care and vitamin D supplements. Prognosis in such a child is guarded.
Hypoxic-ischemic birth injury is preventable to an extent with adequate prenatal and intranatal care.

Case 7

Two years old child presented with delayed standing and inability to walk without support. He was born after 30 weeks

of gestation with normal delivery. He did not cry at birth and required resuscitation with bag and mask ventilation and oxygenation. He also required tube feeding for one week but thereafter was discharged. He was under regular follow-up and had gained initial milestones bit delayed than normal. He held head at 5 months, rolled over at 7 months, sat at 10 months. Cognitive and language milestones were also developed a bit late but he was talking now at two years of age and also had normal understanding. He had no other complaints. This history suggests mild global delay that has improved to near normal except inability to walk without support at 2 years of age that is disproportionately delayed. We may have to find an answer on physical examination.
Physical examination showed normal nutritional status with cognition and language development within normal limits, bilateral upper motor neuron signs in lower limbs but not in upper limbs. There were no other abnormalities.
Bilateral pure motor UMN lesion without sensory loss rules out spinal disease. As pyramidal fibers innervating both lower limbs are affected, the lesion in the brain is periventricular. Lesion is static and so diagnosis is **spastic diplegic cerebral palsy.** This must have resulted from probable milder grade of periventricular hemorrhage. This type of cerebral palsy has near normal brain function.
There is no need for neuro-imaging as it is unlikely to give any more relevant information.
Physiotherapy would improve this child and prognosis is good.
Cerebral palsy is a static brain disorder of tone and posture in

a developing brain with or without affection of domains other than gross motor. Thus, in cerebral palsy, motor delay is always a predominant finding.

Case 8

Mother of one year old child complained of developmental delay. This child was born after full term and normal delivery. Child had cried immediately and there was no problem at birth. He was discharged on D 3 with exclusive breast feeding. However,,, on D 5, he developed acute episode of vomiting followed by seizures and unconsciousness for which he was hospitalized. There was no fever. He was put on IV fluids and anti-convulsant drugs and some investigations were sent. Over next 2 days, he improved and looked normal. Pending reports, he was discharged without any medicines. However, he returned with similar events within next two days.

This history rules out any birth injury as his problems started only after four days with normal birth events. Sudden onset of severe neurological symptoms in an apparently normal neonate and then quick recovery within two days on IV fluids, suggest **inborn error of metabolism.** In such a disorder, abnormal metabolite is formed from one of the constituents of ingested feed due to lack of a specific enzyme and this abnormal metabolite is toxic to the brain. Specific abnormality can be diagnosed only on detailed metabolic investigations. If not recognized in time, it results in ongoing brain damage that is fatal. Even if abnormality is diagnosed and offending agent from ingested feed is withdrawn,

irreversible damage may have occurred and one can hope to prevent further damage. Ideally such diseases are best diagnosed by neonatal screening to prevent damage but can be considered mostly if index case exists in the family. However neonatal screening is carried out routinely in western countries for more than 20 such diseases, selected on the basis of its incidence in the community. In India, many centers have started neonatal screening program for 5-6 common diseases and is cost-effective considering common prevalence of these diseases in the community.

Take home message

Developmental screening must be done by every doctor at 3, 12 and 18 months and if found to be delayed, detailed testing is necessary. This is the only way one can pick-up early deviation in development and take appropriate measures to achieve maximum developmental potential. This is an important part of monitoring young child in initial years. Subsequently school performance can be a guide to any such delay .

MCQs

Q 1 Malnutrition at this period is likely to result in severe developmental delay

A) Early in fetal life
B) Late in fetal life
C) Early in infancy
D) Late in infancy

Q 2 Global developmental delay refers to delay in

A) Two domains
B) Three domains
C) All domains
D) Any of the above

Q 3 Which of the following statement regarding cerebral palsy is RIGHT?

A) Head O is normal
B) Head O is small
C) Head O is big
D) Any of the above

Q 4 Delayed speech development may be seen in

A) Hearing impairment
B) Autism spectrum disorder
C) Normal child
D) Any of the above

Q 5 Autism spectrum disorder is suspected by

A) Poor social communication skills
B) Purposeless hyperactivity
C) Delayed speech
D) Any of the above

Answers to MCQs

Q 1 A, Q 2 D, Q 3 B, Q 4 D, Q 5 D

Section 3 Analysis of other symptoms seen at times, in office practice

There are few other symptoms that may not be as common as those discussed in previous section but may present as an only complaint or at times, be a part of multiple symptoms. Thus, they also need to be addressed. Some of these problems are ignored by patients and doctors alike and at times not even be recognized as an important contributor to the diagnosis or general ill-health.

3.1.Skin rash – a challenge

Basics

Skin rash may appear red, swollen, itchy or irritated and also may present change in texture (rough – sandpaper rash in scarlet fever). Rash is not pathognomonic of any disease as similar rash is seen in many diseases. We need to look at morphology (macular, popular, vesicular, nodular, pustular, purpuric, confluent), progression (spread, peeling – dying skin, pigmentation), areas involved (hands and feet, trunk and back, face - rashes that spare hands and feet are scarlet fever and impetigo), blanching (becoming white or pale after pressure as in inflammatory rash), time of appearance of rash (usually appear in first few days (often an infection), but if after a week, mostly inflammatory diseases), accompanying symptoms and signs, history of allergy and drugs, family history of similar disease.

Common causes related to morphology Centrally distributed maculopappular rash (viral infections, rickettsia, inflammatory diseases, drugs), confluent blanching erythema (kawasaki, scarlet fever, toxin induced rash), vesicular rash (varicella, herpes simplex, enteroviral infection, skin-scalded

syndrome, Steven-Johnson syndrome), urticarial rash (coxasackie, HBV,HCV, vasculitis, malignancy, drug, allergy), purpuric rash (HUS, DIC, ITP, leukemia, meningococcemia, vasculitis, hemorrhagic viral fever),nodular (TB, fungal, rickettsial, erythema nodosum), circular scaly rash with raised border, skin in the middle of the lesion appears normal (ringworm), red, itchy with prominent borders, scaly, blisters that may ooze and form crusts (contact dermatitis, allergic eczema), red blisters, itchy (hand, foot and mouth disease), red, wet and irritating (diaper rash), silvery scaly with sharp defined borders, may be itchy, often on scalp, elbows, knees and back (psoriasis), butterfly rash on face sparing nasolabial fold (Systemic lupus erythematosus), painful swollen red rash with fever (cellulitis), pimples or blisters, extremely itchy, scaly, raised white lines (scabies), white or yellow scaly patches that flake off, areas are red, greasy or oily (seborrhea), red skin all over sparing hands and feet, sandpaper rash (scarlet fever).

Clinical approach to skin rash<u>Does it represent underlying serious disease?</u> In every disease, physician must rule out serious disease even before embarking on finding he cause. In such cases, patient appears sick with change in behavior and often febrile accompanied with other symptoms besides skin rash.

Purpuric rash suggests severe and often life-threatening diseases such as meningococcemia, disseminated intravascular coagulation abnormality as in sepsis or hemolytic uremic syndrome with renal involvement, vasculitis of different etiologies including malignancy,

leukemia and bone marrow aplasia. Gangrenous rash is typical of rickettsial infection. Vesiculo-bullous rash in a sick looking child denotes probable drug reaction as in Steven-Johnson syndrome. These conditions merit hospitalisation and prompt management.

Does it represent a potential evolving serious disease?

Skin rash with inflammation of mucus membranes, fever and / or multisystem involvement indicate diseases that may worsen over time as in case of kawasaki disease (may develop coronary artery dilation leading to myocardial infarction) and toxic reactions to staphylococcal or streptococcal infections (prolonged fever with risk of fatality). Such suspected diseases ideally need referral for expert opinion for investigations and management.

Once seriousness is ruled out, is it primay skin disease or systemic disorder?

Such diseases are usually benign though they need correct diagnosis to prevent chronicity (scabies, eczema, contact dermatitis- need proper treatment or referral to a dermatologist) and close monitoring to rule out complications if any (infectious disease such as measles, varicella, mumps).

Rash itches or itch rashes?

It is important to assess what started first – was it a rash or an itch? Unless asked for, such a history does not come forth and even then, most patients or parents of children might not have noticed the sequence of appearing itch and rash as both may follow in quick succession. Itch at the onset without a rash may suggest autonomic nervous system

abnormality (worsened by anxiety or stress), diabetes or increase in bile salts as in obstructive jaundice. Such primary conditions causing itching are often missed. Rarely Hodgkin lymphoma or other immune disorders may present with itching before other symptoms and signs manifest.

Management

Life threatening and potential serious conditions are managed by experts. Infections may be self-limiting or may need antibiotic as in case of scarlet fever or skin infection such as scabies, cellulitis or impetigo. Many other conditions are managed with palliative measures more than curative treatment. It is ideal that dermatologists manage primary chronic skin diseases. Oral antihistamines are necessary for severe allergic or itching disorders. Local application of steroids, antibiotics, antiseptics, moisturisers and soothing agents may be required.

Ointment contains 80% oil and 20% water whereas cream contains 50% of each oil and water. So, ointments are preferred for dry skin as they provide moisture and also possess antiseptic properties. Lotion is dilute form of cream.

Home remedies – cold compress help temporarily to offer relief from severe itching. Oatmeal bath is recommended as it works as anti-inflammatory and antioxidant. Aloe vera has medicinal properties and is present in many skin creams. Coconut oil is a moisturizer and also has antibacterial activity.

Take home message

Physician must rule out life threatening and potentially serious conditions that may present with skin rash and then

try to be rational in approach to probable etiology. It is important to differentiate primary skin disease from a systemic disease (fever and other symptoms) presenting with skin rash. Timely referral for hospitalization in serious disease is important to save life. Potentially serious conditions must be suspected in time with necessary investigations. Referral to a dermatologist is important especially in chronic primary skin conditions.

MCQs

Q 1 This helps the most in the diagnosis of skin rash

A) Morphology
B) Progression
C) Distribution
D) Accompanying symptoms and signs

Q 2 Skin rash in relation to fever may appear on

A) First day
B) Within 2 -3 days
C) After a week
D) All of the above

Q 3 Which of the following statement is WRONG? Peeling of skin is seen in

A) Varicella
B) Kawasaki disease
C) Severe inflammation
D) Severe infection

Q 4 Which of the following statement is WRONG? This type of skin rash suggests potential seriousness

A) Purpuric
B) Gangrenous
C) Vasculitic
D) None of the above

Q 5 This type of application is ideal for dry skin

A) Cream
B) Lotion
C) Ointment
D) All of the above

Answers to MCQs

Q 1 D, Q 2 D, Q 3 A, Q 4 D, Q 5 C

3.2. Fainting episode – a diagnostic challenge

Basics

Sudden fainting referred to as syncope presents with sudden brief loss of postural tone and consciousness due to transient cessation of cerebral blood flow as a result of either hypotension or bradycardia. Typically attack occurs while standing due to blood pooling in lower limbs due to gravity resulting in fall in blood pressure resulting in transient hypo perfusion of brain. It is frequently preceded by feeling of warmth or nausea. If attack is prolonged, seizure may occur. Skin is pale and sweating, eyes are closed and there is no bladder or bowel incontinence. Symptoms other than

fainting may also be present such as drowsiness, headache, unsteadiness or feeling weak. Several mechanisms are involved in causation of such
an episode and include autonomic nervous system dysfunction, auto-dysregulation of cerebral blood flow, endogenous vasodilator and low serum ferritin. Autonomic nervous system exercises control over many functions such as heart rate, blood pressure, bowel and bladder and such control is involuntary.

Causes

It may occur in healthy individual as a vasovagal attack triggered by anxiety, fear, stress, pain or hunger and is typically experienced by an older school-going child while standing at morning assembly. But sudden fainting attack may also be caused by variety of disorders such as heart defects (rhythm disturbances, arrhythmias including tachycardia, WPW syndrome or long QT syndrome, myocardial dysfunction, hypertrophic cardiomyopathy, outflow tract obstruction), nervous system disorder (seizure, stroke, transient ischemic attack, migraine, normal pressure hydrocephalus. autonomic nervous system dysfunction), severe anemia, hypoglycemia, dyselectrolytemia, sepsis, toxins and drugs Other situational syncope include dehydration, hyperventilation, cough and micturition syncope. Postural hypotension is seen in elder people due to sudden change in body stance.

Differential diagnosis

Syncope has to be differentiated from primary seizure

disorder or vertigo. Besides, underlying cause of syncope has to be established. History is most important to be obtained from a witness with second-to-second details of the event. Such detailed history is often unavailable but attempts must be made to obtain as much information as one can get. This assumes far more important because often physical examination does not reveal any abnormality. Subsequent investigations and management depend on provisional diagnosis.

Clinical approach

First step is to confirm a fainting episode as syncope. A single episode of a standard pattern in a healthy child occurring in a typical situation does not pose a problem. Prolonged attack or recurrent episodes need careful assessment.

Seizure may occur in any position, eyes are open and attack is followed by post-ictal drowsiness, at times with bladder incontinence of tongue biting as against eyes are closed, there is no bladder incontinence, tongue biting or drowsiness following syncope. In fact, a child with a syncope stands up immediately after a momentary period of unconsciousness.

Vertigo presents with feeling of spinning, unsteadiness, dizziness without loss of consciousness or feeling surrounding objects rotating. It is worsened with change in position and is often accompanied with nausea or vomiting.

Next step is to find the cause, once syncope is confirmed.

A single typical episode with a quick and complete recovery in an otherwise healthy child without any physical examination abnormalities or obvious disorder does not need any investigations and patient or parents must be assured of

its benign nature. However, in case of first attack occurring during physical exercise such as playing indicates probable heart defect. Besides, prolonged attack or recurrent episodes justify further assessment.

Physical examination may reveal evidence of heart or neurological disease, severe anemia or other acute conditions and further relevant tests can be ordered or patient referred for specialty opinion. Repeated physical examination and also in different positions may detect autonomic nervous system abnormality.

Real challenge is to plan investigations in absence of any clue on physical examination. It is important to rule out cardiac rhythm disturbance especially long QT syndrome or any other silent heart defect, for which ECG or Holter monitoring and echocardiogram would be necessary. EEG may be abnormal in a seizure disorder, however normal EEG does not rule it out. Video-EEG may be considered if seizure is highly suspected that could record tracings for longer hours. Neuro-imaging is rarely necessary. Tilt-table test can detect changes in heart rate and blood pressure on sudden changing position from lying down to sitting and standing. Fasting blood sugar is considered to rule out hypoglycemia.

Management

Appropriate therapy depends on specific cause of syncope and is best left to a specialist. In an apparently benign and situational syncope, preventive measures may be considered. They include diet modification such as small frequent feeds, adequate amount of salt and water, sleep with elevated head, slow change of body position and avoiding caffeine or

alcohol. Biofeedback may be tried. It consists of mind-body technique that involves using auditory or visual feedback to gain control over involuntary body functions such as blood flow, heart rate and blood pressure. Essentially it is a form of relaxation technique and meditation and it is useful in cases of chronic stress, anxiety or pain.

Take home message

Diagnosis of syncope is based on history obtained from a witness and cause is commonly benign. However, if syncope occurs during physical exercise or if it is prolonged or recurrent, it needs further investigations and specific management. In benign syncope, relaxation technique and meditation is useful.

MCQs

Q 1 Which of the following statement is WRONG? These are other symptoms of poor cerebral blood flow

A) Drowsiness
B) Headache
C) Nausea
D) Feeling weak

Q 2 Which of the following statement related to syncope is WRONG?

A) Eye are open
B) It occurs in standing position
C) It lasts for very short time
D) It takes time to revive consciousness

Q 3 Physical examination in a child with syncope may be normal in

A) Presence of heart defect
B) Presence of neurological defect
C) Both of the above
D) None of the above

Q 4 Which of the following statement is WRONG?

A) Every child with syncope must be investigated
B) Syncope occurring during physical exercise must be investigated
C) Recurrent syncope must be investigated
D) All of the above

Q 5 Syncope due to autonomic nervous system abnormality can be suspected by

A) Changes in heart rate
B) Changes in blood pressure
C) Changes in body temperature
D) All of the above

Answers to MCQs

Q1 C, Q 2 D, Q 3 C, Q 4 A, Q 5 D

3.3. Squint - a straight view

Introduction

It is a condition in which both eyes do not look in the same direction. This happens when eye muscles of both eyes do

not work together. When one eye is focusing at an object, other eye is off direction. Affected eye does not look at the direction of gaze. Affected eye is turned inwards, outwards, upwards or downwards though most commonly eye turns inwards or outwards and rarely up and down.
It is also termed as strabismus. It may be mild and not easily noticeable. Such a child may turn head or neck to one side to suppress the vision from the affected eye to avoid double vision. It may be intermittent or present all the time. Intermittent squint is normal in first two months of life and It disappears by the age of 4 months. It may develop transiently in a normal person when tired.

Types of squint

It is referred to as esotropia when affected eye is turned inwards, exotropia when eye is turned outwards, hypertropia when tuned upwards and hypotropia when tuned downwards. It may be constant or intermittent squint. It may manifest when eye is open or when eye is covered referred to as latent squint. In concomitant squint, degree of squint is same all the directions which means all eye muscles are working but not synchronously as their movements are not aligned. Non-concomitant squint manifests only in one direction but not in the other direction.

Causes of squint

Some babies are born with a squint, if it persists beyond the age of four months,it is permanent. Exact cause of such a congenital squint is not known. Often it is familial. However, squint may develop anytime later in life as a result of

refractive errors (myopia, hypermetropia), diseases of eye muscles (including paralysis of muscles), retinoblastoma, brain defects (midbrain lesions involving 3rd, 4th and 6th cranial nerve nuclei, cerebral palsy, abnormalities such as Down's or Noonan syndrome) and also affection of nerves.

Is squint harmful?

A child with a squint may stop focusing with the affected eye called as amblyopia or lazy eye resulting in disuse of neural functioning and is at the risk of permanent loss of vision if not corrected in time. Binocular vision is due to vision focused from both the eyes together and it results in three-dimensional view of the object as perceived by brain. Thus, a child with a squint may not develop binocular vision. However, in case of children with squint, both eyes focus on different spots but brain learns to ignore out of focus image and so there is usually no double vision unlike in adults. Squint in a child also is a cosmetic problem that affects the child psychologically causing diffidence.

Clinical approach

Generally, it is the parents who report a squint in their child but a physician also should look for it, especially if it is mild and ignored by parents to seek advice or when it is latent. Squint persistent beyond the age of four months or worsening squint at any age needs proper evaluation. First step is rule out neurological disorder. Signs of raised intracranial pressure, paresis including that of eye muscles and cerebellar signs must be looked for and so also developmental delay in a child with abnormal physiognomy.

Once neurological disease or defect is ruled out, it is ideal to refer the child to an ophthalmologist. It is not easy to check refraction or fundus examination in a young child.
Routine testing of vision at birth and again at 6-8 weeks of age, thereafter at preschool and school age is ideal. Squint and amblyopia should be diagnosed as early as possible for better outcome.

Management

Glasses may be required for refractive errors. Amblyopia is treated with a patch on a normal eye so as to force the brain to use the affected eye. Vision therapy is to improve visual skills and abilities, it is a training with the help of lenses, prisms, filters, patches, electronic gadgets and balance boards. It is used for improving lazy eye and improve binocular vision problems. At times, corrective surgery may be necessary though under-correction or over-correction is a possibility.

Take home message

Persistent squint beyond four months of age in an infant should not be ignored. Sudden occurrence of a squint may be due to neurological disorders and need appropriate investigations and management. Non-neurological squint should be referred to an ophthalmologist as early as possible for proper evaluation and treatment to avoid long term vision problems.

MCQs

Q 1 Which of the following statement is WRONG? Squint

A) Is normal in first two months
B) It may be evident when tired
C) May disappear by itself
D) Is always fully correctable

Q 2 Which of the following statement is WRONG?

A) Squint may be present since birth
B) Squint may develop anytime in life
C) Squint may not be evident
D) None of the above

Q 3 Which of the following statement regarding squint is WRONG?

A) It may result from myopia
B) It may result from hypermetropia
C) It may result from Amblyopia
D) No cause may be found

Q 4 Clinical examination in a child with squint should include

A) Refractive error
B) Movements of eyes in all directions
C) Fundus examination
D) All of the above

Q 5 Early diagnosis of squint is important to prevent

A) Loss of vision
B) Loss of binocular vision
C) psychological disturbance
D) All of the above

Answers to MCQs

Q 1 D, Q 2 D, Q 3 C, Q 4 D, Q 5 D

3.4. Mouth ulcers

Introduction

Mouth ulcers are common in the community seen at all age groups, while most of them are benign and self-limiting, one must be cautious to rule out any systemic disease.in the background. Mucus membrane of the mouth is very delicate and is easily damaged even by trivial factors. These lesions are seen in all the parts of mouth including base of gums. They are painful and cause difficulty in eating, drinking and speaking.

Causes

Two most common causes include local trauma and aphthous ulcers. Local mechanical injury may be induced by accidental biting of cheeks or lips while chewing hard food items, sharp tooth, poorly fitting dentures or braces, chemical injury by spicy food or thermal injury due to hot drink. Ulcers caused by local injury heal within 1-2 weeks. Aphthous ulcers are also localized to mouth but they recur every few months and may take longer time to heal. While exact cause is not known, it is likely to be triggered by genetic predisposition, stress, Vitamin or iron deficiency and hypersensitivity to tooth pastes or smoking. Both these causes are restricted to mouth only without much systemic symptoms.
Next common causes include infections and vitamin / irondeficiency. Viral infections such as coxasachie A (hand,

foot, mouth disease), herpes, varicella, EB virus, HIV, bacterial infections such as scarlet fever, fungal infections, syphilis etc. Deficiency disorders include vitamin B12, folate and other B complex vitamins and iron deficiency. Rare but serious causes include autoimmune disorders such as SLE, juvenile idiopathic arthritis, dermatomyositis, vasculitic syndromes, inflammatory bowel disease, drug reactions such as Steven-Johnson syndrome and occasionally leukemia. Some of these disorders may start with mouth ulcers before other manifestations appear and, in such situations, correct diagnosis is difficult in initial stages. However, such patients are often disproportionately sick. Mouth represents transition between skin and GI tract and hence mouth ulcers are often associated with skin lesions or GI disturbances

Clinical approach

First step is to differentiate between localized benign lesions and mouth ulcers representing a probable systemic disorder. Systemic disorders generally present with sickness besides other symptoms such as fever, skin rash, joint involvement etc. Sickness is different than discomfort caused by painful lesions and though it is a subjective impression, experienced physician can make out a difference between discomfort and sickness. However, at times it could be tricky and one has to be cautious in pronouncing a benign lesion.
Once a benign disorder is considered, detailed history may suggest local injury and if not, one may look at family history to consider aphthous ulcers and also assess probable vitamin

B12 or folate deficiency (knuckle pigmentation) and iron deficiency (koilonychia). Past history of similar disease favors diagnosis of aphthous ulcers as they are recurrent. History of drug therapy may be a clue to cause of mouth ulcers.
However one must be cautious to keep in mind that isolated mouth ulcers may be the only initial presentation of systemic disorders and so must look for evidence of systemic disease on physical examination such as evidence of viral infection, skin rash, nail abnormalities, joint swelling, restriction of joint movements or pain, pallor, lymphadenopathy, hepatosplenomegaly and signs of other organ involvement.

Investigations

Isolated mouth ulcers without evidence of any other physical findings in an otherwise normal child does not call for any investigations. In case of accompanying pallor, peripheral blood smear along with CBC can suggest deficiency anemia. Aphthous ulcers is a clinical diagnosis based on circumstantial evidence and do not justify any tests.
If mouth ulcers don't heal within two weeks or in case of doubt about probability of systemic disorder, CBC, ESR / CRP are initial tests that may support a systemic disease – either infective, autoimmune or malignant diseases. Further investigations depend on initial test results. In case of suspected fungal infection, work up for immune deficiency including HIV is necessary as fungal mouth ulcers are seen in immune-compromised individuals with an exception of a neonate or an individual on long term antibiotic therapy.

Treatment

Benign lesions are self-limiting and don’t need any specific drug therapy. It is best to avoid spicy and hot food, drink plenty of fluids, keep mouth clean and rinse mouth with warm water with pinch of salt by keeping water in mouth for few minutes. Local application may be tried such as ghee or butter or local anesthetic agents. Local steroids are not helpful but analgesics may be tried. Withdrawal of probable offending drug is necessary if suspected to be drug induced mouth ulcers. Replacement therapy is necessary for vitamin and iron deficiency disorder.
Viral infections are also self-limiting and don’t need any specific treatment. However, disseminated herpes infection needs to be treated with acyclovir or ganciclovir. Streptococcal infection is treated with first generation cephalosporin or amoxicillin. Fungal infection is treated with local anti-fungal drug unless it is disseminated that needs systemic therapy. Treatment of other serious disorders must be left to a specialist.

Take home message

Mouth ulcers are usually benign and self-limiting but we must look for any systemic disorder in the background that may not be initially evident, unless specially looked for. However, if lesions persist beyond two weeks, we must investigate to rule out a systemic disorder. Local lesions heal on their own and while local application may be tried, healing takes a natural course.

MCQs

Q 1 Which if the following statement is WRONG? Mouth ulcers may be

A) Progressive
B) Migratory
C) Both likely
D) One of them likely

Q 2 Which of the following statement is WRONG?

A) Single ulcer is due to local cause
B) Single ulcer is due to systemic cause
C) Multiple ulcers are due to systemic cause
D) Multiple ulcers are due to local cause

Q 3 Which of the following statement is WRONG? Self-limiting mouth ulcers may be

A) Due to local cause
B) Due to systemic cause
C) Never associated with fever
D) Always associated with pain

Q 4 Which of the following statement is WRONG? Fungal infection in mouth may be seen in

A) Immunocompetent individual
B) Immunocompromised individual
C) Normal neonate
D) None of the above

Q 5 This helps the most in healing of mouth ulcers

A) Local anesthetic agent
B) Local steroid

C) Nature
D) Analgesic

Answers to MCQs

Q 1 C, Q 2 B, Q 3 B, Q 4 D, Q 5 C

3.**5.Epistaxis**

Introduction

It is a common condition, often due to benign causes but at times may represent more serious diseases. Epistaxis refers to bleeding from nostrils, nasal cavity or nasopharynx, mostly from anterior part of nasal cavity. Blood dripping from posterior nasopharynx may be due to epistaxis, but occasionally, massive epistaxis may be confused with hemoptysis or hematemesis.

Causes

Fragile capillaries in the anterior part of nose are vulnerable to rupture due to extreme environmental temperatures and is one of the most common benign cause of epistaxis. Nose pricking may also lead to epistaxis and so also accidental injuries to nose or head injuries with basal skull fracture. Foreign body in the nostril may also present with epistaxis but it is unilateral. Epistaxis may be the manifestation of underlying hematological or vascular disorder. Common hematological diseases include platelet disorders (destruction or lack of production of platelets or platelet dysfunction in spite of normal number of platelets) and

coagulation disorders (congenital or acquired deficiency of coagulation factors), vascular disorders such as vasculitis and different types of vascular malformations. Hypertension is not the cause of epistaxis in children as hypertension is mostly acute unlike in adults in whom chronic progressive hypertension may present with epistaxis.

Clinical approach

First step is to make sure that it is blood. Many times, blood-stained kerchief is shown to the doctor as evidence of blood. Rarely, a child can intentionally cheat the parents and doctors both. Next step is to locate exact site in the nasal cavity. History should find out whether epistaxis was unilateral or bilateral, time taken to stop bleeding, recurrence of bleed within short time, relation to environment or injury, any accompanying symptoms such as fever, bleeding from other sites, past history of similar episodes and family history of bleeding disorder. Physical examination should first ensure that child is hemodynamically stable and then start with inspection of nasal cavity, ideally with a nasal speculum. During active episode, local examination may not be possible unless bleeding stops but throat examination may reveal dripping of blood from nasopharynx. In such case, firm pressure should be applied over the nostril for two minutes to stop bleeding. If it fails or takes time to control bleeding, further evaluation or referral is mandatory. Bleeding is easily controlled by pressure in common benign causes and physical examination should rule out presence of foreign body.

Physical examination should focus on abnormal findings such as sick look, purpura, and ecchymosis, bleeding from other sites, hepatosplenomegaly or lymphadenopathy.

Investigations

Tests are necessary only when one suspects hematological or pathological vascular causes of bleeding. Sick child, bleeding from other sites, difficulty to control bleeding and presence of abnormal physical findings are pointers to justify further evaluation with relevant tests. Hemoglobin level can estimate degree of blood loss. Besides CBC, platelet count and PT / PTT are basic screening tests to give a clue to type of bleeding disorder. Further tests can define subtypes such as bone marrow in case of low platelet count and coagulation factor assessment and liver function tests in case of coagulation defects. In case of suspicion of vasculitis, special tests may be required.

Management

Usually firm pressure over nostrils controls bleeding. However, if it fails, topical epinephrine 1:10000 and 4% lidocaine may help to control bleeding. Nasal packing with gauze has been a traditional method that is now replaced with balloons, tampons or compressed sponge.
Specific treatment is necessary in case of hematological or vascular causes.

Take home message

Occasional self-limiting epistaxis in a healthy child is often a benign problem and does not call for any specific

intervention. However, it could be the first and only a single manifestation of hematological or vascular disorder that would need proper evaluation. Usually, detailed history and thorough physical examination does point to a probable systemic disorder.

MCQs

Q 1 Epistaxis could be

A) Unilateral
B) Bilateral
C) Variable
D) All of the above

Q 2 Which of the following statement is RIGHT? Degree of blood loss can be assessed with

A) Detailed history
B) Area of kerchief / towel soaked with blood
C) Pulse rate
D) All of the above

Q 3 Which of the following statement is WRONG? Epistaxis in a healthy child may be due to

A) Local cause
B) Platelet disorder
C) Coagulation disorder
D) Vasculitis

Q 4 Which of the following statement is WRONG?
A) Recurrent epistaxis should be investigated
B) Recurrent epistaxis may not need investigations

C) Isolated first episode should not be investigated
D) None of the above

Q 5 Epistaxis in such a child must always be investigated

A) Recurrent nose bleeds
B) Family history of nose bleeds
C) Difficult to control bleed
D) All of the above

Answers to MCQs

Q 1 D, Q 2 C, Q 3 D, Q 4 D, Q 5 D

3.6. Sleep problems

Introduction

Sleep is not a luxury but a necessity. Adequate sleep is necessary to boost health and work performance. Neonate sleeps for 18-20 hours a day including day time naps, infant about 12-13 hours, toddler around 10-12 hours and adult should sleep for 7-8 hours a day. Day nap should not be more than 20-30 minutes if at all taken by adults. However, majority adults and many children are sleep-deprived because of changes in life styles, especially in cities. Sleep pattern is often ignored as an important part of health and so rarely discussed unless it poses a significant problem.

Inculcating ideal sleep habits

Most of the neonates sleep very well during the day and remain comparatively awake during night hours. This pattern settles down to natural rhythm by next few weeks.

Thereafter, babies sleep for 3-4 hours at a time and get up for a feed to sleep again. However, as infant grows and develops ability to interact with the surroundings, sleep is easily disturbed and infant remains awake for more time. Many times, infant fights sleep in order to be with parents and other family members. It is at this time around 6 months of age that parents should try to set healthy sleep pattern. Parents should set a fixed sleep schedule that should be followed each day to an extent possible. It is important to put the baby on the bed when sleepy but awake so that infant learns to fall asleep. Mother should maintain physical contact with the baby while putting the baby to sleep and even sing a song to sooth the baby but not form habit of breast feeding or rocking to get to sleep. Warm water bath often helps to induce the baby to sleep. Sleep hygiene consists of providing comfortable quiet environment avoiding light or sound. Once this routine is repeated every night, infant gets habituated to sleep at that particular time. Initially baby shares parent's bed and subsequently a toddler could share parent's room but separate bed. Such healthy habits need to be sustained in subsequent years. Older children and adults must plan a dinner time at least two hours prior to sleep time and also avoid ingestion of stimulant like coffee and exposure to electronic gadgets to facilitate good sleep. It is known that bright light including TV screen disturb initiation of sleep because melatonin - hormone from hypothalamus – is secreted in poor light inducing sleep process. Meditation and other relaxation techniques such as concentrating on breath

or body scan, light music or reading a book help to calm the mind and relieve stress.

Common sleep problems in children

Normal infant in initial months does get up for a feed but goes back to sleep immediately at the end of the feed. Most infants towards latter half learn to sleep through the night without need for a feed. Occasional sleep disturbance is common in children and is due to minor illness or environmental disturbance. However, when it happens frequently, one must attend to it as a problem such as difficulty in initiation of sleep, interrupted sleep and day time sleepiness.

Initiation of sleep

Parents may find it difficult to put the baby to sleep. This is usually due to failure of timely instilling sleep habit. Even older child may also face same problem. Most households are busy till late in the night with many activities and this is deterrent for young child to go to sleep. Older children may be stressed with studies or fights that make them not get to sleep quickly. They waste time in bed trying to sleep and then find it difficult to get up in time.

Interrupted sleep – arousal disorders

Infant may wake up several times due to hunger but often due to habit formed by mother to feed on breast or by bottle to put him back to sleep. Once this habit is formed, infant wakes up for sucking pleasure rather than for hunger. Older children may get up due to obstructed airway presenting as snoring or sleep apnea – transient cessation of breathing in

sleep that may go unnoticed but results in day time sleepiness. Nightmares are other reasons for waking up in sleep, they occur after a dream and children are able to remember the event when fully awake. At times, older child wakes up half-asleep, is confused and agitated and performs simple activities such as sitting up or mumbling, complex activities such as screaming and becoming aggressive – night terrors or even more complex activities such as sleep walking. Children have no recall of such activities as against in nightmares where the child remembers entire event. Restless leg syndrome is another problem where a child gets severe urge to move legs and hands due to perception of aching, itching, tingling or creeping sensations. This is a result of stress, anxiety or depression.

Day time sleepiness

Inadequate sleep due to difficulty in initiation of sleep and need to get up in time for school or work and interrupted sleep due to variety of reasons lead to day time sleepiness. School going children burn late night oil to study specially during examination time and it is a common cause of day time sleepiness. Some of them are habituated to go to bed at late hours at all times.

Common sleep problems in adults

Most of the problems faced by older children are also prevalent in adults and mainly arise from stress and tension at work place and at home. Chronic respiratory disorders such as chronic bronchitis and emphysema, asthma, sinusitis, chronic cardiac diseases, diabetes, chronic GI problems,

other degenerative disorders of joints and physiological old age issues such as aches and pains as well as enlarged prostate in males are other reasons for sleep problems.

Consequences of sleep problems

Sleep disorders can take a toll on mental and physical health besides on mood, energy, memory issues, weight gain, ability to handle stress and poor performance at study and work. It is a silent problem but has serious negative impact of life.

Management of sleep problems

Prevention is better than cure and it is important for parents to inculcate healthy sleep habits that are as vital as other habits related to diet and exercise. It is also necessary to develop emotional and psychological health that helps to cope up stresses in life effectively.

Sleep problems must be recognized early to manage well. Parents should try to change wrong habits in children with cooperation and firmness. Pathological conditions are tackled appropriately such as adenoidectomy and tonsillectomy for obstructed sleep apnea. Stress, tensions, depression need proper counselling and at times expert referral.

Rarely investigations may be necessary to diagnose correctly the cause of sleep problems. Sleep laboratories are now available in major centers for sleep study that involves continuous monitoring of several physiological parameters during sleep. Such a study can reveal apnea episodes and other breathing disorders and may need special intervention.

Take home message

Sleep problems are common in children and adults but remain unrecognized or ignored. Inculcating healthy sleep habits and providing sleep hygiene for infants and children are important steps for parents. While transient disturbance in sleep is common, frequent disturbances in sleep need early recognition, proper evaluation and management.

MCQs

Q 1 Ideal amount of sleep every day for adults is

A) 5-6 hours
B) 7-8 hours
C) 9-10 hours
D) Amount that keeps an individual happy

Q 2 Day time sleep

A) Is not recommended
B) Is a must
C) Is ideal for 20 minutes
D) Is fine as per the need

Q 3 This organ in the body is responsible for sleep initiation

A) Cerebral cortex
B) Hypothalamus
C) Thalamus
D) All of the above

Q 4 This system dysfunction is common cause of disturbed sleep

A) Respiratory
B) Cardiac

C) Neurological
D) GI

Q 5 A child with nightmare wakes up with

A) Inconsolable crying
B) Aggressive behavior
C)Confusion and disorientation
D) Normal behavior

Answers to MCQs

Q 1 B, Q 2 C, Q 3 B, Q 4 A, Q 5 D

3.7. Flatulence – "gas" problem

Introduction

Flatulence refers to accumulation of gas in gastrointestinal tract. Normally, gas is formed in the large intestine as a result of bacterial (helpful bacteria exist in intestine) action on digested food (bacterial fermentation) that produces gases like methane, nitrogen and carbon dioxide which exit out. Gas also may be swallowed while eating or drinking rapidly, eating gas-producing food items, soft aerated drinks and chewing gums. Flatulence may also result from indigestion caused by food intolerance, food allergy or gastrointestinal diseases. Whenever excessive gas is produced or normally produced gas does not exit, it results in retention of gas in alimentary canal. Flatulence leads to bloating of abdomen as well as frequent farting and burping. It produces discomfort and embarrassment.

Gas-producing food items

It is mainly the carbohydrates in food that produce gas as a result of their digestion while proteins and fats contribute to a little gas. Amongst sugars, lactose (dairy products) and fructose (some fruits like mangoes, grapes, oranges, sweet lime, figs) cause excessive gas though other sugars also may do so especially if ingested in excess. Complex sugars such as starch are difficult to break down and so produce excessive gas as happens in consumption of potatoes, corn, noodles and wheat. Rice produces very little gas. Beans, cabbage, broccoli, sprouts, green leafy vegetables produce excessive gas. However, it is important that many of such gas-producing food items are also beneficial in many other ways and hence food intake should be balanced with proteins, fats, carbohydrates, vegetables, fruits and dairy products based on what suits individual's digesting system the most.

Common causes of flatulence

Flatulence is often a result of wrong eating habits including imbalanced food consumption and constipation that upset digestive processes causing flatulence. This does not represent any disease process. However gastrointestinal diseases also lead to flatulence and at times may be the only symptom of developing disease.

Food intolerance is an individual person's inability to handle food effectively and while it is not a disease but an isolated defect that demands avoiding food items to which a person is intolerant.

Food allergy results from abnormal response to ingested

food that worsens over repeated exposure to an offending agent unlike in case of food intolerance in which there is no worsening symptoms. Gluten induced allergy – Celiac disease is due to allergic reaction to wheat, barley, oats is a classic example of food allergy that is often seen.GERD – gastroesophageal reflux disease – may present with burping in addition to retrosternal discomfort (heart burn), vomiting and aspiration into airways leading to cough.
Lactose intolerance is due to lack of lactase – an enzyme that is necessary to break down lactose – that presents with flatulence, abdominal bloating and pain often with watery stools. This is seen often secondary to acute intestinal infection in children, is transient and self-limiting. It is also more prevalent in old age, especially in those individuals who are not used to consume dairy products.
Inflammatory bowel disease may present with flatulence in addition to abdominal pain, abnormal stools often with blood and deteriorating health. In such cases, other systems may also be involved such as joints, eyes, kidneys etc.
Irritable bowel syndrome presents in similar way but with normal health status and is mainly due to stress rather than any intestinal pathology.
Chronic intestinal infections including parasitic disease such as amoebiasis and giardiasis also present with flatulence. In fact, chronic intestinal disorders of any cause including GI malignancy may result in flatulence.

Clinical approach

First step is to note whether symptoms pertain to frequent passage of gas (farting or burping) or just abdominal bloating with discomfort without passing gas. Acute onset bloating of short duration with severe abdominal pain is the hallmark of surgical abdomen such as acute intestinal obstruction and presents with vomiting.

Those who present with excessive farting also have bloating and abdominal discomfort. If such a person is otherwise healthy and not sick, it may be due to imbalanced diet or irritable bowel syndrome. However, if such a person is sick, one needs to evaluate more sinister causes that include inflammatory bowel disease (IBD), chronic intestinal infections including TB, parasitic diseases and even malignancy. Physical examination often fails to diagnose such conditions and would need further relevant investigations.

If burping is a main presenting symptom, one may consider upper GI disorders such as GERD, esophagitis or malignancy in old age with other accompanying symptoms or simply an imbalanced diet in an otherwise healthy person.

Investigations

Healthy individual with chronic problem does not justify any investigations as problem lies often in eating habits. Similarly, no tests are necessary in case of strong clinical suspicion of irritable bowel syndrome.

Stool microscopy may reveal evidence of chronic parasitic infection. Occult blood in microscopic stool examination or fecal calprotectin may indicate presence of intestinal inflammation. Reducing substance in stool (lactose) is seen in

every loose stool and does not suggest per se lactose intolerance and hence not routinely asked for.
CBC may show neutrophilic leukocytosis with thrombocytosis in IBD along with low serum albumen. ESR may be useful in monitoring course of the disease rather than diagnosis of a specific condition. Serum IgA anti-tTGantibody is a screening test for celiac disease.
Imaging may be necessary in selective conditions such as GERD, IBD, intestinal TB, malignancy but with limited use.
Colonoscopy and intestinal biopsy help in diagnosing IBD, celiac disease. Upper GI endoscopy with biopsy is necessary to diagnose GERD along with 24 hour esophageal pH monitoring.
However, tests for celiac disease or GERD must be further supplemented with therapeutic trial as all such tests have limitations and at best are highly suggestive but nor confirmative by themselves.

Management

It depends on the cause. Life style and diet modification is the key to many benign conditions. Home remedies include eating low carbohydrate containing fruits such as berries (blue, black, straw), apricots, grapefruit, peach, watermelon and low carb vegetables such as carrots, tomatoes, green beans. Rice and fermented items like Idli produce less gas. Knee-chest position, hot water, ginger help to relieve gas.

In pathological disorders, diet modification to avoid offending agent is the standard management in allergic disorders as in celiac disease in which wheat, barley and oats

should be avoided. Diet change (elemental diet) is also necessary in other intestinal disorders such as IBD besides immune-suppressive drugs. Prokinetic drugs, H2 blockers and PPI –proton pump inhibitors may help in case of GERD. Chronic intestinal infections are treated with specific antibiotics.
Surgical treatment may rarely be necessary in GERD as fundal plication or rarely in IBD – especially ulcerative colitis with fistula formation as seen more commonly in adults.

Take home message

Flatulence is a common problem in the community presenting as abdominal bloating or discomfort, excessive farting or burping. Diet and life style related issues predominate in apparently healthy persons. However, flatulence of sudden onset or as a persistent symptom in a sick individual demands further evaluation to rule out significant gastrointestinal disorder.

MCQs

Q 1 Which of the following statement related to symptoms caused by flatulence is WRONG?

A) Abdominal bloating
B) Abdominal pain
C) Excessive burping
D) None of the above

Q 2 Which of the following statement is WROMG?

A) Farting may be normal
B) Burping may be abnormal

C) Farting is always abnormal
D) None of the above

Q 3 This type of carbohydrate is responsible for excessive gas production

A) Lactose
B) Fructose
C) Starch
D) All of the above

Q 4 This produces least gas

A) Rice
B) Wheat
C) Sprouts
D) Dairy products

Q 5 This condition needs drug treatment

A) Inflammatory bowel disease
B) Irritable bowel syndrome
C) Chronic intestinal infections
D) Gastroesophageal reflux disease

Answers to MCQs

Q 1 D, Q 2 C, Q 3 D, Q 4 A, Q 5 B

3.8. Dental health – mostly neglected!

Introduction

In general, teeth are the most neglected part of the body in Indian population with few exceptions. Parents

of infants are concerned about timely eruption of teeth and thereafter blame every disease either to erupting teeth or even during pre-erupting stage. Younger generation is concerned with cosmetic aspects of teeth with emphasis on well aligned sparkling white teeth. But almost no one is concerned about dental health.

Basic information

First tooth may appear anytime from 6 months to as late as 18 months of age. Rarely neonate is born with a tooth that may have to be extracted because of fear of slipping into throat and downwards but almost there is no human without teeth. (Though such a condition is reported). Primary teeth have shorter and thinner roots and thin enamel that looks white. They are 20 in number. They primarily act as “space-maker” for permanent teeth to be placed. Around 6 years of age, deciduous teeth start falling to create a space for permanent teeth and this process continues till all 20 teeth are replaced by 32 teeth. Wisdom teeth may appear as late as 18-25 years and often at least one of them is impacted in many individuals. Though appearance of primary as well as permanent teeth may vary a great deal in normal individuals but number remains universally the same. Though, extra teeth or appearance of permanent teeth before falling of primary teeth are also known but rare.

Dental care

Care in early childhood

It should start right from the time first primary tooth erupts. It is ideal to use a finger or finger-brush with rice-grain size smear of fluorinated toothpaste and by the time all primary teeth erupt, one may need to use a pea size dollop of toothpaste. In place of toothpaste, tooth powders of various types can be used as per the prevalent cultural practices. Bottle feeding, prolonged breast feeding or night feeding should be avoided to prevent tooth decay.

Care at other age groups

Soft bristled brush is ideal and must be used with gentle pressure. Vigorous pressure damages the enamel. One must follow correct technique of brushing. Brush must be held at 45-degree angle to the gum line and use rotational movement. Molars – the teeth at the back must be properly brushed as most food particles are stuck there. Foods and drinks with acidic pH tend to damage enamel if you brush immediately after consumption of such items, so brushing must be delayed by 30-60 minutes. Brushing should be only two times a day and not after each meal. Cleaning gums with finger and mouth wash are important to maintain hygiene. It is important to clean the teeth, gums and mouth with water each time immediately after eating anything. Sugary drinks should not be consumed in a lingering way. Eat raw fibrous fruits such as apple, pear,

carrot, cucumber that help to keep teeth white. They help to remove plaque over the teeth that are responsible for yellow staining of teeth. Such fibrous foods also need extra chewing and so generated saliva neutralises acid that may erode teeth.
Water fluoridation helps to prevent tooth decay by 20-40%. It is ideal to use fluorinated tooth paste. Permanent teeth start getting yellowish over time and one may use a whitening tooth paste that has an abrasive ingredient such as silica that scrubs the surface of teeth and make it white. Periodic floss or interdental cleaning helps maintain oral hygiene.

Dental diseases

90% of adults in India have some tooth decay after the age of 20 years. It is seen also in younger children with temporary teeth but usually starts during adolescence due to lack of proper maintenance.

Tooth pain

It is a common problem that may result from infection of tooth or its structures but also may indicate other conditions such as cracked tooth (fractured tooth), chipped tooth, impacted tooth and exposure of nerve endings in a cavity.

Dental caries or cavities

Commonly known as tooth decay, it is caused by breakdown of the tooth enamel as a result of bacteria

on teeth that acts on foods and produce acid that destroys enamel and results in tooth decay. With worsening destruction, nerve endings are exposed resulting in sensitivity to cold items that leads to sudden and severe pain. Cracked tooth is a fracture that may cause pain on biting or on release of biting pressure and at times may result in cold sensitivity with inflammation of pulp. Pain may also be due to infection in a decaying tooth.

Gingivitis

It is the infection of gums that is caused by a plaque formed of sticky bacteria. It may localise to form an abscess. Such infections may spread to nearby areas and may even be the cause of brain abscess in a child with congenital cyanotic heart defect. They also may cause halitosis – bad smell to breath and mouth. If left untreated, it causes bone loss and tooth may become loose and shift.

Impacted wisdom tooth

50% of adults may have at least one impacted wisdom tooth. It may damage the adjacent tooth, may result in gum disease, a cavity or misalignment.

Stained teeth

Teeth are often stained due to pigments in food or drugs. Iron deficiency anaemia being so common in India, oral iron is the cause of staining of teeth. Tobacco chewing is also common in India and is a cause of

staining.

Crooked teeth

Teeth may be crooked and poorly aligned or there may be a gap between teeth that may come in the way of good hygiene.

Bruxism

It refers to grinding of teeth. It may occur in sleep or even at day time. It may result from stress, sleep problems, malaligned teeth or respiratory allergy and may result in headache, cracked tooth and jaw pain.

Monitoring dental health

Periodic dental check is ideal right from the time first tooth erupts. Infants and toddlers commonly visit paediatricians or health facility for immunisation and hence there are opportunities to not only monitor growth but also dental health. Subsequently parents need to be sensitised to continue maintaining good oral hygiene. Visit to dentist happens only when routine measures fail. Early detection of dental problems is the only way to preserve good teeth. Unfortunately, this aspect is universally ignored in India.

Management of dental problems

Dental care has advanced a great deal applicable even for young children and it is out of scope of this article. Aim is to prevent dental diseases and correct naturally

present defects at the right time. Treatment of neglected diseases always would leave behind permanent damage though at least would prevent further worsening.

Take home message

Dental health is a neglected area and community including doctors need to be sensitised to attend to it. Prevention of dental diseases is easy and cost-effective. Early detection is ideal but needs a periodic visit to a dentist even in absence of any symptoms.

MCQs

Q 1 Which of the following statement is WRONG?

A) Permanent teeth are always 32 in number
B) Timing of appearance of teeth is variable
C) Primary teeth are whiter than permanent teeth
D) Impacted third molar is rare

Q 2 Which of the following statement is RIGHT? Eruption of primary teeth may cause

A) Loose stools
B) Fever
C) Mild discomfort
D) Convulsion

Q 3 Brushing of teeth should start when

A) First permanent tooth erupts
B) First primary tooth erupts

C) Set of primary incisors erupt
D) Child can spit out

Q 4 Which of the following statement is WRONG? Tooth pain may be caused by

A) Infection
B) Injury
C) Impacted tooth
D) Crooked teeth

Q 5 Which of the following statement about bruxism is WRONG?

A) It may be due to stress
B) It may cause headache
C) It may cause tooth pain
D) It occurs only in sleep

Answers to MCQs

Q 1 D, Q 2 C, Q 3 B, Q 4 D, Q 5 D

3.9.Urine output – a marker of renal health

Introduction

Urine output is an important parameter that is easy to judge on direct enquiry and offers useful bedside information. (It is difficult to judge in young infants but measurement of 24 hour urine output is rarely necessary) Patients can also keep a note of it and therefore it forms a part of personal history

that every doctor must enquire. Reduced amount of urine – oliguria – is the common occurrence in disease states but excessive amount of urine – polyuria may also be a manifestation of a disease process. Urine output less than 400-500 ml per 24 hours in adults is considered as oliguria. In infants, it is less than 1 ml/kg body weight/hour and in children less than 0.5 ml/kg/hour. (Urine output more than 2.5 litres per day in adults is considered polyuria and it is more than 5 ml/kg/hour in infants and 4 ml/kg/hour in children).

Basics revisited

Glomerular filtration is the first step to make urine through which excess of water and waste products from the blood are excreted out of the body. Kidneys are supplied with 20% of cardiac output that amounts to more than one litre per minute in an adult. Afferent arterioles deliver blood to a glomerulus for filtration while efferent arterioles carry the filtrate into excretory system and venules carry the blood back into the circulation. Renal autoregulation can dilate or constrict afferent arterioles which counteracts changes in blood pressure within limits. Rate at which kidneys filter blood is called glomerular filtration rate (GFR). Serum creatinine increases only after GFR is reduced considerably to less than 30 ml/min from normal 100-120 ml/min and hence is a late indicator of impaired renal function.

Common causes of oliguria

Reduction in blood supply to kidneys, impairment of glomerular function and obstruction to urine outflow are

main groups of mechanisms that produce oliguria.
Poor perfusion of kidneys may result from dehydration due to various causes (poor intake of water specially in hot weather, severe diarrhoea or vomiting, severe burns, shock), capillary leak (dengue shock syndrome), poor cardiac output, systemic hypertension or renal artery disease (stenosis or arteritis).
Glomerular disfunction may be caused by glomerulonephritis (endothelial and interstitial diseases) and renal failure of different etiology.
Obstruction to urine outflow presents with oliguria in spite of normal renal function as in case of bilateral pelvic-ureteric junction defect or posterior urethral valve. Neurological disorders may also lead to retained urine.

Clinical approach to oliguria

First step is to confirm reduced urine output. It may be obvious or rarely may need to measure urine quantity especially in ICU setting. Poor intake of fluids in a sick individual may also have oliguria but it is not severe and also not a presenting feature.
Enlarged bladder denotes either mechanical obstruction or neurological disorder, in spite of normal urine formation.
Once obstruction is ruled out, one must look for signs of dehydration (tachycardia, dry mucosa, loss of skin turgor) or shock (cold extremities, hypotension, increased capillary refill time and encephalopathy) or cardiac failure (pedal oedema, engorged neck veins, cardiomegaly).
Haematuria (high coloured urine), facial puffiness, systemic

hypertension suggests endothelial glomerular disease such as acute glomerulonephritis due to different causes or signs of renal failure (encephalopathy with deep rapid breaths suggestive of metabolic acidosis) of various etiology.

Investigations

Routine urinalysis is a simple test that may reveal primary renal glomerular disorder. Urinary specific gravity and electrolytes are important in specific conditions. GFR can be estimated roughly – referred to as eGFR by following simple equation. (there are other complicated methods of calculation)

eGFR = 0.5 X (height in cms divided by serum creatinine mg%)
Serum creatinine and blood urea are other renal function tests. Relation of blood urea and serum creatinine may vary between 10:1 to 20:1. Increased ratio suggests pre-renal conditions due to poor perfusion as in case of dehydration or cardiac failure.
Other relevant tests would depend on probable diagnosis and include estimation of electrolyte and acid-base imbalance, imaging (abdominal USG, upper GI study, echocardiogram) and diagnostic tests for infections.

Management

It would depend on final diagnosis. Not every patient with oliguria would need intravenous fluids. It would be necessary only in case of dehydration or intravascular constriction. It may be harmful in case of cardiac failure and useless in case of obstructed urinary system. Similarly diuretic drugs are not

required for every oliguric patient. Other details are out of scope of this article.

Clinical approach to polyuria

It is important to ask for polyuria because physicians generally focus on oliguria, it being much more common. Polyuria is at times confused with frequency without increased volume of urine. Thus, it is not merely the number of times urine passed but quantity of urine passed at each time. Young infants tend to pass urine more often and it is dilute, hence polyuria is not easy to assess. Common causes include diabetes (polyphagia and polydipsia along with polyuria), renal tubular disorders (child is sick with acidosis and other metabolic abnormalities) and rarely psychological disorders (compulsive water-drinking).

Take home message

Oliguria – reduced urine output may be due to insufficient formation of urine or due to obstruction to outlet of urine in spite of adequate production. If it is due to reduced formation, one needs to differentiate between prerenal condition from renal pathology. It can be assessed easily on the basis of history and physical examination before embarking on investigations. Polyuria should be differentiated from frequency due to local irritation and is easy to miss in young children unless specially asked for.

MCQs

Q 1 For routine office practice, oliguria must be

A) Estimated by periodic physical collection over 24 hours
B) Measured by 24 hours collection by catheterisation
C) Assessed by frequency and amount passed each time
D) Any of the above

Q 2 Which of the following statement about oliguria is WRONG?

A) It may result due to renal glomerular disease
B) It may be due to renal tubular disease
C) It may occur in spite of normal kidneys
D) None of the above

Q 3 Pre-renal oliguria may be due to

A) Cardiac disease
B) Intestinal disease
C) Capillary leak syndrome
D) All of the above

Q 4 Oedema in case of oliguria is seen in

A) Dehydration
B) Capillary leak syndrome
C) Obstruction to urine outflow
D) None of the above

Q 5 Hypertension in case of oliguria is seen in

A) Glomerulonephritis
B) Cardiac failure
C) Renal failure
D) All of the above

Answers to MCQs

Q 1 C, Q 2 B, Q 3 D, Q 4 B, Q 5 D

3.10Voiding dysfunction

Introduction

Voluntary control on passing urine develops over first few years with child remaining dry initially during the day and thereafter also by night. Depending on toilet training, most children achieve control during day by 3-4 years and night by 5-6 years. Though few children continue to wet the bed at night for longer duration. Ideal time for toilet training in children should start only after child demonstrates urge to pass urine by gestures or manoeuvres and should proceed slowly as per the child's response. Urination appears to be a natural and simple process but a complex mechanism is involved for bladder to do what brain orders. However, the problems may arise at any age. Voiding dysfunction is a broad term used to describe conditions in which there is inconsistent coordination between urinary bladder and urethra. Normally urinary bladder allows filling of urine coming from the kidneys, stores it and releases it at appropriate time under voluntary control within limits. Voiding dysfunction occurs when there is a problem with either filling, storage or emptying.

Back to basics

Once bladder starts filling to its normal capacity, it sends a message to the brain requesting an order to release. If facility does not exist, brain orders to wait for some more time and orders release as soon as possible. But there is a limit to

which bladder can distend and hold on. If order to release does not come in time, bladder decides to release urine by itself without the brain order, even without proper facility and incontinence results. In normal health, such a neurological network works well and maintains dry state. This network includes sympathetic and parasympathetic nerve, their connections in spinal cord, centre of micturition in pons that relays information to brain cortex to get final order and pass on to spinal centres. Several muscles are involved such as bladder detrusor, external sphincter muscle and pelvic floor muscles. When detrusor contracts, sphincter must open and pelvic floor muscles must relax to allow urine exit. Any abnormality in this circuit results in voiding disturbances.

Symptoms

Voiding problems manifest in various ways such as frequency (passing urine number of times more than usual), urgency (strong urge to pass urine), hesitancy (difficulty in initiating act of urination), straining, slow, weak or interrupted stream of urine, retention (not emptying completely), overflow incontinence, dribbling and bed wetting (enuresis beyond the expected age of control). Such symptoms may be transient as in case of UTI or may also be intermittent or persistent. If left untreated, may cause renal damage.

Causes

Broadly causes can be divided into neurogenic and non-neurogenic. Neurogenic problems arise from cerebral cortex, pons, spinal cord or nerves due to various causes that result

in detrusor-sphincter dyssynergia (incoordination between detrusor and sphincter – when detrusor contracts, sphincter also contracts resulting in functional obstruction to passage of urine). Non-neurogenic problems present as overactive, underactive or dysfunctional bladder and may arise from weak pelvic muscles, use of alcohol, caffeine or drugs like antihistamines or atropine, urinary tract infection, overweight and life style issues. Anatomical obstructive malformation in lower urinary tract and constipation are other important causes of voiding problems and recurrent UTI. Chronic constipation results in distention of rectosigmoid that presses on urethra and also may be due to common sharing of neural pathways. Enlarge prostate is a common cause in old age. Young children tend to hold urine for long by ignoring urge to pass urine while busy with playing or at times avoiding unclean wash rooms in schools. It may lead to daytime wetting. Giggle incontinence results while laughing in children who are susceptible to detrusor instability and generally it disappears as child grows. Urethral irritation due to local fungal infection may cause frequent urination and at times urgency or incontinence.

Clinical approach

Detailed history of voiding helps to define the type of a problem. Urgency is the hallmark of overactive bladder, chid often has daytime urinary incontinence and tries to hold urine by standing on tiptoes, crossing of legs or squatting with heels pressed into perineum.
Infrequent urination is the hallmark of underactive bladder

and such children strain while voiding.
History of constipation is often overlooked as a contributory factor to voiding problems.
Neurological examination may pick-up lower motor neurone lesion in lower limbs with distended bladder with overflow incontinence or continuous dribbling without bladder distention. Examination of lower spine may reveal subtle signs such as tuft of hair indicating lower spinal cord defect. Upper motor neurone diseases may also result in voiding problems but major presentation is other than just voiding issues such as change in sensorium or seizures.
It is important to observe flow of urine and patient's manoeuvres while passing urine that gives clues to type of voiding problems.

Investigations

Routine urinalysis may reveal evidence of UTI that should further be confirmed with urine culture. Imaging studies include USG to assess post-void residual urine volume and rarely CT scan or micturating cystourethrogram may be necessary. EMG of pelvic muscles may help to define status of these muscles. Urodynamic studies are required in selected cases of suspected detrusor-sphincter dyssynergia. In case of neurological disorders, relevant investigations are necessary.

Management

Non-neurological voiding problems are treated with behaviour modification, bladder retraining, biofeedback and Kegel exercises to strengthen pelvic floor muscles. Drugs are

rarely necessary but may be required in selected cases such as prophylactic antibiotics for patients with high infection risk or anticholinergics for temporary use.
Neurological causes need relevant surgical correction or medical palliative management such as repeated bladder catheterisation to avoid residual urine retention.

Take home message

Voiding problems are often not reported in time by patients, especially in case of non-neurological issues. These problems need timely intervention to prevent permanent renal damage besides disrupting normal life. Toilet training is important in early childhood and delayed or too early enthusiastic attempts may both lead to problems.

MCQs

Q 1 Toilet training in children

A) Should start as early as possible
B) Urination is a natural process, training is not necessary
C) Should start only when child shows urge to pass urine
D) Should start when child can follow oral instructions

Q 2 Which of the following statement about voiding problems is RIGHT?

A) It may be physiological
B) It may be pathological
C) It may be functional
D) All of the above

Q 3 Which of the following statement about symptoms of voiding problem is WRONG?

A) Incontinence
B) Oliguria
C) Urgency
D) Hesitancy

Q 4 Nocturnal enuresis may be a symptom

A) In a normal child
B) In a child with neurological disorder
C) In a child with anatomical urinary tract malformation
D) All of the above

Q 5 Voiding problem may need

A) Surgical procedure
B) Antibiotics
C) Behaviour modification
D) Any of the above

Answers to MCQs

Q 1 C, Q 2 D, Q 3 B. Q 4 D, Q 5 D

3.11. Abnormal movements – movement disorder

Introduction

Movements are under voluntary control that help us perform different tasks efficiently including maintaining body balance and working with hands. Such control is exercised mainly by basal ganglia and other related structures. Movement

disorders result from loss of voluntary control – referred to as involuntary movements. However, at times, abnormal movements may also occur in spite of normal neurological system and then are considered to be physiological. Movement disorders often present as isolated or major symptom but may also be accompanied with affection of many other brain areas.

Types of movement disorders

They may often be involuntary but occasionally voluntary and may be either hyperkinetic (increased movements) or hypokinetic (decreased movements as in Parkinsonism). Hyperkinetic abnormal movements may include ataxia, dystonia, tremors, chorea, athetosis, myoclonus, hemiballismus, tics or Tourette syndrome.

Characteristics of movement disorders

Hyperkinetic

Ataxia – loss of truncal balance resulting in instability and swaying and / or intentional tremors (tremors on intentional movement of limbs – when made to move the limbs) and incoordination (as demonstrated by finger to nose test and many other clinical maneuverers) with hypotonia.
Dystonia – increased muscle tone with twisting seen at rest or brought out on stimulation, sustained for variable time.
Tremors - distal, fast, rhythmic, small amplitude movements, may be seen at rest or on stimulation or stretching hands out. May be physiological due to anxiety, stress or fear and are also seen in hyperthyroidism and liver cell failure.

Chorea – proximal, chaotic, fast, non-rhythmic, large amplitude with emotional instability and hypotonia
Athetosis –writhing movements, distal, slow, rhythmic, large amplitude, often together with chorea – choreoathetosis.
Myoclonus – shock-like jerky movements of group of muscles, fast, arhythmic. At times, such movement may be benign as in sleep myoclonus with normal neurological system.
Hemiballismus – wide flinging movements of half side of the body (fly removal movement – movement while removing a fly coming on to your face), large amplitude.
Tics – suppressible, paroxysmal stereotype muscle contractions
Tourette syndrome – frequent, repetitive, jerky localised movements often involving face and neck with vocal tics in the form of grunting, throat clearing, shouting or barking, usually getting better by the age of 20 years

Hypokinetic

Slow movements as shuffling gait in Parkinsonism with other features such as tremors, stiffness of limbs and trunk, incoordination and impaired balance.

Causes

Can be etiologically classified as follows
Immune mediated (rheumatic chorea)
Infection (TB meningitis with choreoathetosis and hemiballismus)
Genetic (degenerative disorders)
Metabolic disorders (Wilson disease)

Vascular (strokes)
Tumour (cerebellar tumour)
Toxins (chronic hepatic disease or inborn error of metabolism)
Drugs (anti-emetic, anti-convulsant)

Clinical approach

First step is to characterise abnormal movement based on following criteria (as depicted above).
proximal or distal, fast or slow, rhythmic or arhythmic, small or large amplitude. This may be confirmed by observation or supporting clinical maneuverers. Typical characteristic may help guess probable etiology based on common presentation.
Second step is to confirm that abnormal movement is definitely pathological as tremors or myoclonus may be physiological.
Next step is to ascertain whether abnormal movement is the only abnormality or whether there is evidence of affection of other areas of brain. Presence of other symptoms and signs such as change in sensorium, pyramidal tract signs, cerebellar signs, meningeal signs, cranial nerve affection and evidence of involvement of other systems such as liver or thyroid disease. Thus, anatomy of disease can be found out.
Pathology and etiology are guessed on history of presentation (acute, subacute or chronic, static or progressive) as well as past history (rheumatic fever), family history (consanguinity as in Wilson disease) and drug history.

Investigations

Provisional diagnosis is a prerequisite to ordering tests. Obviously specific test may vary in each case. CT or MRI scan may show basal ganglia lesion and affection of other areas if any. But may not be necessary in each case. For example, diagnosis of rheumatic chorea is often clinical with typical presentation and neuro-imaging will not help in defining the cause. Stroke may be confirmed with neuro-scan and if performed in early stage, damage can be reversed. CNS infection can be diagnosed with CSF examination, Wilson disease is confirmed with biochemical tests. Tics do not justify any tests.

Treatment

Symptomatic therapy

Abnormal movements may be controlled by drugs such as Haloperidol for rheumatic chorea, levodopa or beta-blocker for Parkinson tremors and other drugs like antidepressants and anticholinergics. Duration of symptomatic treatment depends on natural progress of the condition. Abnormal movements in Rheumatic chorea are usually controlled in few weeks but at times they may recur. Prolonged treatment is necessary in degenerative conditions.

Specific therapy

It depends on the cause. For example, Penicillamine for Wilson disease, anticoagulant and aspirin for strokes, anti-infective for infections and penicillin prophylaxis for rheumatic chorea as per the standard guidelines.

Take home message

Abnormal movements may be physiological or pathological. Characteristics of abnormal movement help in classifying movement disorder and guessing probable etiology. Brain imaging is necessary in selective conditions depending on availability of specific therapy. Symptomatic therapy is necessary for variable period.

MVQs

Q 1 Which of the following statemen is WRONG? Abnormal movements may be

A) Voluntary
B) Involuntary
C) Physiological
D) None of the above

Q 2 This abnormal movement can be suppressed

A) Dystonia
B) Tremors
C) Tics
D) None of the above

Q 3 This abnormal movement can occur in sleep

A) Tremors
B) Myoclonus
C) Tics
D) All of the above

Q 4 Which of the following statement is RIGHT?

A) Tremors are distal and slow movements
B) Chorea is proximal and repetitive movement

C) Dystonia is increased tone with twisting
D(None of the above

Q 5 This abnormal movement disappears over years by itself

A) Tourette syndrome
B) Tics
C) Tremors
D) Ataxia

Answers to MCQs

Q 1 D, Q 2 C, Q 3 B, Q 4 C, Q 5 A

3.12. Excessive weight gain – obesity

Introduction

Malnutrition exists in the form of undernutrition as well as overnutrition. Unfortunately, India faces a dual burden of malnutrition. Prevalence of obesity has increased over last decade, more so in urban areas, but also in rural parts of the county and in all socioeconomic groups. Obesity in children is likely to persist in adulthood with its dire consequences that manifest in early adult life with significant morbidity and shorten productive life. Childhood obesity is mostly preventable with timely intervention to inculcate ideal life style.

Difference between overweight and obesity

Overweight is defined as weight between 85^{th} and 95^{th} percentile for same age and sex while obesity refers to weight above 95^{th} percentile for same age and sex. Both the

conditions usually have abnormal or excessive fat accumulation in the body that carries risk to health. Overweight if not controlled progresses into obesity. Severe degree of obesity is referred to as morbid obesity.

Methods of measurement

Periodic charting of weight and length / height right from birth up to puberty (completion of growing period) and thereafter weight is most essential to pick up early deviation in these parameters. This is the simplest way to track growth pattern though weight within normal range does not exclude excess accumulation of fat.

Body Mass Index (BMI) is calculated by dividing weight in kilograms by height square in meters. Body composition of growing children vary with age and sex but also is different in Indian children as compared to children in western countries. Hence, we must use Indian growth charts for assessing growth parameters including BMI. Indian Academy of Pediatrics has developed these charts, latest version is published and can be easily downloaded from IAP website as well as one can search them on internet. However, BMI is also a rough guide but most practical way of measurement and so is widely used. IAP charts are user-friendly to an extent that one need not calculate BMI by the equation mentioned above but charting weight and height of a child on BMI chart can give an answer. There are three lines on BMI friendly chart – upper line – OB - represents obesity, middle line – OW - overweight and lower line – UW - underweight. Weight and height are charted and the point

where they meet decides BMI. If this point falls at or below UW, person is underweight, between UW and OW, person is normal. If this point lies between OW and OB, he is overweight and if point lies at or above OB, he is obese. It suffices for appropriate action. Waist-hip ratio is another practical and simple way to judge not only fat accumulation but also fat distribution. Waist circumference is divided by hip circumference is below 0.9 in males and below 0.8 in females DEXA scan is the best way to assess fat content of the body. Many Indians have excess of fat, even in an apparently thin body stature and are referred to as “thin fat Indians”. Such individuals are also at high risk of complications.

Causes of overweight / obesity

Balance between consumption of energy (calories from food) and expenditure of energy (metabolic state and physical exercise) decides the outcome. Excess intake of calories and / or lack of physical exercise are the most common causes of overweight / obesity in the population. This is referred to as exogenous obesity. Onset of such an imbalance may start even in infancy, especially in infants fed on formula feeds or may start at later age due to wrong life style.

Endocrine disorders also present with obesity and they include primary hypothyroidism and Cushing’s syndrome or secondary to pituitary disorders.

Children with developmental delay tend to be obese due to either lack of physical exercise or as in syndromic disorders such as Prader-Willi or hypothalamic disorders. Leptin

deficiency may present in early infancy with obesity. (Leptin hormone controls intake of food)

Clinical approach to obesity

First step is to confirm overweight / obesity by charting weight, height, BMI and waist / hip ratio. Once it is confirmed, next step is to find out probable cause.
As a general rule, tall and obese is due to exogenous factors (excess intake of energy and / or lack of physical exercise) while short and obese are due to endocrine causes.
Developmental delay or mental retardation points to hypothyroidism (lethargy, constipation are other features) or syndromic abnormalities (dysmorphic features and other associated malformations).
Rapid weight gain over short time indicates primary adrenal disorder such as tumour or hyperplasia (hypertension is a feature) and it may also be secondary to pituitary disorders (evidence of other hormonal disorders and increased intracranial tension in case of pituitary tumour)

Investigations

Exogenous obesity does not justify any tests to confirm the diagnosis. However due to inherent risk of consequences of obesity, periodic check on blood pressure, blood sugar, lipid profile and relevant tests for assessment of function of other organs is ideal.
Endocrine disorders should be confirmed by relevant tests. Imaging studies may be necessary in case of suspected pituitary or adrenal tumour as well as for determination of

bone age as in case of hypothyroidism.
Genetic tests may help to confirm syndromic abnormalities.

Complications

Obesity is a multisystem disease and severity of which depends on duration and degree of obesity. Many organs are affected though the functional disturbances are not evident clinically, they need to be monitored by relevant tests. Diabetes, hypertension, myocardial dysfunction, hypoventilation, fatty liver, osteoporosis, constipation, indigestion, immune dysfunction and mental health disturbances are common and need periodic monitoring.

Prevention

Exogenous obesity can be prevented to a large extent by healthy life style that should be inculcated from infancy. Periodic growth charting (both weight and height) right from birth onwards up to puberty and thereafter monitoring weight every month is a simple way to keep a track of your weight and therefore health status. With it, one can take necessary action if weight increases to higher centiles. Weight for height is a better index to monitor and ideally, both weight and height should maintain similar centiles. Change in weight to higher centile needs explanation and appropriate action such as diet control and increase in physical exercise. Ideal life style is far more important in case of genetic propensity to exogenous obesity. It is sad to see the child being attended by the doctor since early infancy without growth monitoring and then come to attention, only

when obesity has already been established. At that stage, outcome is usually poor in spite of good efforts.

Management

Exogenous obesity is managed by controlling excessive or imbalanced intake of energy and increasing energy expenditure by appropriate physical activities. Ideally, entire family should follow the same regime. Sustaining efforts is difficult for most patients and hence outcome is poor with its future consequences.

Endocrinal obesity would be treated according to the cause while syndromic obesity poses challenge in management due to multiple issues other than obesity.

Associated problems of obesity require proper management as in case of diabetes, hypertension and other organ dysfunctions.

Bariatric surgery is a debatable method of management and should be reserved for morbid obesity.

Take home message

Prevalence of obesity in all socioeconomic groups is fast increasing. Exogenous obesity must be prevented by ideal life style and by monitoring growth parameters regularly. Management of established obesity has poor outcome as sustained compliance is near impossible. Endogenous obesity develops more rapidly and is accompanied with many other symptoms and physical signs. Early diagnosis and prompt management is the way to expect better outcome.

MCQs

Q 1 Which of the following statement is WRONG?

A) Obesity and overweight always go together
B) Overweight person may not be obese
C) Thin person may have excess of fat
D) All of the above

Q 2 Which of the following parameters is the best indicator of obesity?

A) Weight
B) Height
C) Weight for height
D) All of the above

Q 3 Which is the most reliable parameter of excessive body fat?

A) Weight for height
B) Body mass index (BMI)
C) Waist-hip ration
D) Dexa scan

Q 4 Rapid onset of excessive weight gain is likely to be

A) Exogenous obesity
B) Hypothyroidism
C) Cushing's syndrome
D) Syndromic obesity

Q 5 Which of the following statement is WRONG? Obesity may affect

A) Kidney
B) Brain

C) Liver
D) None of the above

Answers to MCQs

Q 1 A, Q 2 C, Q 3 D, Q 4 C, Q 5 D

Section 4 Remember not to forget

Concept of unreported symptoms

History taking is a process of "thought in action" in which, leading questions can unfold hidden physical symptoms to offer a clue to a diagnosis. However, there exist symptoms that are never expressed by patients and so, never taken into consideration by doctors in the management of diseases. Patients do not voice them as they feel these symptoms do not relate to the disease process in the body. But they don't know that these very symptoms have direct relevance to the outcome of a disease. Such symptoms arise from the mind and they include anxiety, fear, uncertainty, depression, frustration, self-pity and even self-blame. The disease affects both the body and mind. It is natural both need to be addressed in the management of diseases. While science (drugs) helps to treat the body, doctors must take care of the mind by ethical practice with empathy, proper communication and counselling, it is a measure of doctor's concerns, commitment, accountability, responsibility, transparency and honesty. It instills faith in the mind of a patient, calms his mind and improves compliance with treatment with a better outcome.
Every doctor must take note of such untold symptoms that

are universally present in every patient, though to a varying extent and provide holistic care with the help of brain (science), heart (compassion), mind (commitment) and soul (inner conscience).
