Clinical Signs and Syndromes in Surgery

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Foreword
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Foreword

It had been an extremely pleasant experience going through the pages of *Clinical Signs and Syndromes in Surgery*.

In an era, where technology is rapidly trying to replace clinical skills, like recording a detailed history, eliciting clinical signs, etc., this work nudges you as a gentle reminder of the unquestionable relevance of clinical examination of a patient. I feel it has been a long-felt need of both undergraduate and postgraduate students, to have a ready-reckoner like this, and its utility is not restricted to students of surgery alone. It is of immense value to students of all disciplines of modern medicine.

In an examination scenario, to be able to group your findings and/or to know the names of various 'named signs' and syndromes while presenting a case, is definitely a great advantage and would impress an examiner no end. Also, it will have immense utility in viva voce as well, and will make a topper standout from a mediocre. I am convinced that Dr Shivananda Prabhu has worked hard and researched well to collect this wealth of information and I am sure the medical students

community will appreciate the value of this collection for years to come.

I congratulate and compliment Dr Shivananda Prabhu for this effort and wish him all the best in all his future academic endeavors.

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Preface

Ever since I was an undergraduate student, eliciting of clinical signs has always fascinated me. I remember watching in awe as seniors demonstrated clinical signs.

This wonderment at such skills reached its peak during discussion of central nervous system (CNS) disorders in the medical wards. Neurological disorders are nothing but a collection of signs, one used to think. Such thoughts brought anxiety with them as one was not sure how to cope.

Those times are long gone, but the fascination with signs remains. Having chosen general surgery as my field, it is only natural that I would now be interested mainly in signs pertaining to surgical conditions. Life of a surgeon is in many ways easier than that of a physician inasmuch as many of surgical conditions produce distinctive symptoms and signs unlike most medical illness. Also, surgical conditions most often than not lead to some anatomical and physiological distortions discernible by clinical examination as clinical signs. Only there have not been many books dedicated to this aspect of clinical examination. There are many excellent books dealing with clinical examination as a whole, but they do not segregate clinical signs from rest of the process of clinical evaluation. Hence, for a student preparing for clinical examination, it becomes a tough task to brush up his knowledge. Hence, the need for a book dealing

exclusively with clinical signs. Also, while we do know about a particular sign as an indication of a particular disease often we do not really know the best way to elicit the sign. Easy access to diagnostic imaging has only made the ignorance deeper. This book attempts to address this problem. This should hopefully help not only students preparing for examinations but also practising surgeons.

I have included a brief account of syndromes, as I found these to be the scourge of exam-going students. Examiners revel in catching students off-guard by throwing questions at them about some obscure syndrome or the other. I hope to reduce such a threat by touching upon them. While this book may not have anything new in its content, I am sure the idea of presenting "signs and syndromes" in a concise book is a novel one.

Shivananda Prabhu

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Introduction

Clinical examination is the most exciting as well as challenging part of a surgical residents' daily routine. Even in today's world where the advancement in the fields of laboratory sciences and diagnostics make the diagnosis of disease conditions less difficult than before, clinical acumen retains its importance. One needs sound clinical judgement to be able to make proper use of diagnostic technology. Hence, students of surgery should endeavor to acquire a level of clinical skills which allows them to narrow down the diagnostic possibilities and order for investigations accordingly.

Often, while examining a patient one relies on certain clinical finding elicited during examination to arrive at a plausible conclusion. Of course a detailed history taken from patient by a sympathetic and astute clinician will go a long way in pointing towards the pathology that the patient has. It is beyond the scope of the book to go into details of history taking. Good history along with well-detected clinical findings, when analyzed together will make the clinical picture clearer. If all the symptoms and clinical observations could be explained by a single pathological entity, then the diagnosis is near certain. Hence, only one diagnosis need be put forth and investigation asked for just to confirm or rule it out. On the other hand if all facets of the case cannot be explained by a single pathological lesion, then

differential diagnosis should be thought of and investigations ordered accordingly.

What are these clinical findings which help us reach a definitive conclusion? These are observations made by the clinician during inspection, palpation, auscultation, or percussion. They are objective findings which can be corroborated by any clinician. There is no subjective element in them.

In other words, they are called "clinical signs".

A clinical sign when properly elicited gives a clue to underlying pathology. Its presence makes the diagnosis more of a probability and less of a guess. When many such observations or signs are put together it is possible to arrive at a conclusive opinion regarding the disease process. It all looks simple and straightforward at first look. But one is well advised to keep the following facts in mind before embarking on the pursuit of this art of eliciting clinical signs.

- Just knowing the theory underlying a clinical sign is not enough. One should be familiar with the exact technique of eliciting the sign. One may not be able to demonstrate a sign, even when it is present if one employs incorrect technique. Even books will help only to a limited extent. There is no substitute for observing an expert clinician eliciting the sign.
- If an attempt at eliciting a sign is likely to cause discomfort to the patient then it is necessary that clinician explains to him the nature of the test and

enlists his cooperation, e.g. rebound tenderness. An uncooperative and distressed patient is sure recipe for failure. If one fails to elicit such a sign within one or two attempts it is better to let it go as inconclusive or absent. One should persist in trying to elicit a sign only if it is vital for the diagnosis. There are very few signs of such singular clinical importance.

- If a simple laboratory test can avoid prolonged clinical examination and laborious analysis then choose it, especially in an emergency setting, e.g. chest X-ray with domes of diaphragm to check for free gas under diaphragm will clear the diagnosis immediately and should not be unnecessarily delayed pending detailed examination.
- One should be able to elicit the sign even when the diagnosis is as yet unclear. Anybody can elicit the sign once the diagnosis is established and known, e.g. even a beginner will be able to observe visible gastric peristalsis once endoscopy has revealed the presence of gastric outlet obstruction. But that observation will only serve academic purpose. On the other hand, if visible gastric peristalsis is observed by an astute clinician in the OPD itself, patient will be saved a lot of time and of course money. Such skill at observing the signs is especially useful while one is working in mofussil areas and not a city.

- Remember a particular sign need not be present in all cases of particular pathology. Atypical presentation of a disease condition is quite common and one needs to maintain a high degree of clinical suspicion to be able to diagnose a condition even in the absence of typical signs.

With these few facts in mind let us now acquaint ourselves with clinical signs, system-by-system.

Chapter ONE

Abdominal Signs

It is only natural that we are discussing abdominal signs first. It has been rightly said that abdomen is a 'Pandora's Box'. Even the most experienced clinicians often stumble when it comes to abdomen. There are times when the final truth about abdominal pathology is only revealed at laparotomy. True, ultrasound, contrast CT, MRI, etc. have made diagnosing abdominal pathology less difficult but not yet easy.

There are many reasons why an abdominal pathology is more difficult to diagnose than lesions anywhere else. The foremost of the reasons is the fact that abdomen is the seat of so many organs of varied anatomy and physiology. That being so, ordinarily when any of these organs is involved it should produce distinct signs and symptoms, but unfortunately most of these organs are interlinked both anatomically as well as function-wise. Hence, a disease in any one organ will disrupt not only its function but also that of the others that are linked to it, thereby confusing the picture. And nature adds to the confusion by introducing its own signs and symptoms via body's protective mechanism. For example, vomiting caused by protective pylorospasm in case of acute appendicitis. Another reason why abdomen is still an enigma is that a proper evaluation of abdomen requires not only a skilled clinician but also a relaxed and cooperative patient. Many a time to expect a patient having severe pain abdomen to be relaxed is to expect too much from him.

Clumsy attempts at palpation will only serve to raise his anxiety. Hence, it is really necessary to make a patient feel at ease before starting any examination. A few minutes spent in sympathetic explanation will go a long way in making the examination worthwhile.

Before starting to look for abdominal signs one has to make the patient lie comfortably on the examination couch breathing deeply but steadily. It goes without saying that the whole of the abdomen should be exposed right from nipple level to mid thigh level. Of course, one has to provide adequate privacy. Ensure that a lady assistant is present if the patient is a female, for obvious reasons.

SIGNS ON INSPECTION

Looking for abdominal signs should start with careful inspection of the abdominal wall as well as its movements. For ease of presentation we will discuss the inspectory signs first followed by palpatory ones and so on.

The signs have not been listed in any particular order of their perceived importance. They are discussed in alphabetic order to eliminate any personal bias regarding their clinical significance.

Auenbrugger's Sign

This sign is said to be present when there is an epigastric prominence produced by marked pericardial effusion.

The differentiation should not be difficult as underlying pericardial effusion will definitely produce other signs and symptoms referable to the poor cardiac function like features of congestive cardiac failure, muffled heart sounds, etc. When in doubt, simple investigation like X-ray chest AP view or echocardiography should clear the doubt.

Beevor's Sign

When the infraumbilical portion of both rectus abdominis muscles is paralyzed umbilicus moves closer to the xiphisternum. This is due to the lack of tone of the lower part of muscles.

Cullen's Sign (Umbilical Black Eye)

This is usually seen in hemorrhagic pancreatitis. There is periumbilical discoloration due to seepage of blood either transperitonially or along the falciform ligament.

It should be borne in mind that this is relatively late sign in the process of pancreatitis and hence should not he sought as an aid to diagnose acute pancreatitis. While its presence indicates grave prognosis for the patient its absence in no way rules out pancreatic inflammation. Diagnosis of acute pancreatitis is essentially clinical based on detailed history and careful examination of the abdomen followed by laboratory tests like serum amylase.

One should remember that the sign is not exclusive for pancreatitis. Any massive intraperitoneal bleed also can lead to the development of this sign. For example, ruptured ectopic.

Grey Turner's Sign

This is bluish discoloration of the flank seen most commonly in acute hemorrhagic pancreatitis. Once again this is due to hemorrhage into retroperitoneal space due to acute pancreatic inflammation. This blood dissects through tissues and appears in flanks. It goes without saying that this is another sign of grave prognosis indicating the need for urgent resuscitation.

One has to remember that this sign may also be associated with other equally serious condition like leaking abdominal aortic aneurysm (AAA), retroperitoneal bleed due to trauma, etc.

Fox Sign

Occasionally there is discoloration of inguinal region in cases of hemorrhagic pancreatitis due to trickling of hemorrhagic fluid.

Hippocratic Facies

Evident during advanced stages of any acute peritoneal inflammation. Patient has in drawn but bright eyes, anxious look with pinched face and cold skin.

Ransohoff's Sign

Yellow pigmentation of umbilicus and periumbilical region in rupture of common bile duct. The extravasated bile traverses along the falciform ligament to reach the umbilical region.

Sign de dance (Dance's Sign)

This is seen in some cases of intussusception, especially the iliocolic type. There is emptiness in the right iliac fossa because of progressive telescoping of the ileum and cecum in to distal colon leaving the right iliac fossa empty. There may be some in drawing of the parietal wall noticeable during inspection. This can be confirmed by palpation. Also on palpation one may be able to feel a sausage shaped mass, with its concavity towards the umbilicus. The consistency of the mass might change from time-to-time depending upon peristalsis. One may be able to appreciate visible peristalsis in these individuals. Barium enema is confirmatory and shows the claw sign, which is discussed later. History from the patient might reveal intestinal colic, obstruction and *red current jelly* stools.

Tanyol's Sign

Normally umbilicus lies midway between symphysis pubis and xiphisternum. But a mass arising from the pelvis may lead to displacement of umbilicus upwards

nearer to xiphisternum. The opposite happens when there is gross ascites. This is known as Tanyol's sign.

Visible Peristalsis

While inspecting the abdomen for any abdominal condition one is supposed to look for movements, be it respiratory, peristaltic or pulsatile. Of these, peristaltic movements, if made out during clinical examination, are more likely to be of clinical significance.

Peristalsis is normal forward propulsive movements of entire gastrointestinal tract. Whenever there is any mechanical obstruction to the forward propulsion of its contents these peristaltic waves become stronger and more frequent in an effort to overcome the obstruction. Patient is likely to have colicky abdominal pain associated with vomiting. These strong peristaltic waves are often visible in not so obese patients. Depending upon the site of obstruction, the clinical nature of visible peristalsis changes. For example, if the obstruction is at pylorus of the stomach, as occurs in chronic duodenal ulcer patient has epigastric pain and peristalsis is visible in upper abdomen. To induce peristalsis, whenever gastric outlet obstruction is suspected on history given by the patient, he is made to drink substantial amount of water and asked to lie down. The clinician should preferably sit beside patient's bed and watch for peristaltic wave starting in the left hypochondrium and moving slightly downwards and to the right. The wave

appears as an area of fullness preceded by constriction. Patient will complain of colicky pain during the test. Other clinical tests like succussion splash and auscultopercussion are done to confirm the dilated state of stomach following outlet obstruction. Succussion splash is the splashing sound of retained gastric contents heard in the epigastrium with the help of stethoscope when patient is gently shaken. Auscultopercussion involves marking the borders of a dilated stomach with the help of change in sound heard when the bell of stethoscope is kept on the epigastrium and abdominal wall is scratched in a radial fashion moving away from the stethoscope. As long as the scratching finger lies over the dilated stomach there will be tympanic note which will abruptly change in character when the finger moves beyond the boundaries of the stomach. One can mark multiple such points, which when joined will indicate roughly the position of the dilated stomach in the abdomen.

Visible peristalsis can also be induced by flicking the abdominal wall with the fingers or putting a few drops of ether on the abdominal wall.

Peristaltic wave will travel in a step ladder pattern progressively moving downwards on case of obstruction of small bowel. In case of obstruction of distal colon the peristaltic wave may be seen to pass from right to left at or just above the level of umbilicus. The direction of movement and other signs of colonic

obstruction help to differentiate this from visible gastric peristalsis.

SIGNS ON PALPATION

Whenever a case of abdominal pathology presents itself in the OPD or casualty the tendency amongst surgical residents is to go and start palpating the abdomen even before a decent history is taken. This practice is improper, insensitive and often counter productive.

Before palpating the abdomen one should try to determine the nature of the pathology that the patient is mostly likely to have. This necessitates taking a good history eliciting details of all the symptoms. Any doubts that the clinician might have should be clarified asking necessary questions. At the end of the history taking clinician will have developed a rapport with the patient. Then the examination proper should start, beginning with general physical examination. Inspection of the abdomen should be done after exposing the abdominal wall fully and allowing the patient to breathe regularly. Any inspectory sign should be recorded for corroboration during palpating. If it is deemed that palpation is likely to be painful then percussion and auscultation should be done before going in for palpation. It is a good practice to explain to the patient the nature of palpatory maneuver that will be needed.

Aaron's Sign

This sign is said to be present when the patient expresses a feeling of discomfort in the epigastric region or the precordial region on applying sustained pressure over the McBurney' point. But it should be noted that there are other signs and symptoms, which cause much less discomfort at the same time pointing towards appendicular inflammation. Hence, this sign is rarely sought when one is suspecting acute appendicitis.

Alder's Sign

This is a useful sign in diagnosing acute appendicitis during the later half of pregnancy.

In a pregnant patient with pain in the right iliac fossa the source of pain can be differentiated by asking her to lie on the left side. After waiting for a few minutes if tenderness is again sought for, if it is of uterine origin then the point of maximum tenderness will be move to the left with the uterus while if it is of appendicular origin then it will stay where it was.

Baid Sign

It is one of the abdominal signs described by an Indian surgeon.

In a thin patient suffering from pseudocyst of the pancreas, the stomach is often pushed anteriorly towards

the abdominal wall. In such a patient if a Ryle's tube is in place it is often palpable beneath the thin abdominal wall. This fact can easily be confirmed by a lateral X-ray of the abdomen.

Bapat's Bed Shaking Test

An ingenious method to elicit inflammation of the parietal peritoneum is to gently shake the bed. The resultant body movement is enough to induce pain in a patient with peritonitis. This test is considered more humane than repeated palpation of the abdomen in a patient with peritonitis.

Boa's Sign

It is a sign of acute cholecystitis. An area of hyperesthesia can be detected over the posterior abdominal wall between T_{11} and L_1 and starting from 1 inch lateral to the midline to posterior axillary line. This is an example of referred pain.

Carnett's Sign

Whenever there is tenderness over any part of the abdomen, if the tenderness decreases on contracting the abdominal muscles then the source of pain is intraperitoneal. If it remains the same then the pathology is in the parietal wall.

Cope's Psoas Test

This is elicited in evaluating a case of possible retrocecal appendicitis. When the appendix lies retrocecally, as it does very often, it lies in close proximity to psoas major muscle. Hence, when such an appendix is inflamed psoas muscle may get irritated enough to go into spasm. In such cases hyperextension of the hip joint will cause pain to the patient. In well-established cases patient might have fixed flexion deformity of the right hip joint.

Cope's Obturator Test

Cope's obturator test has the same underlying principle as Cope's psoas test only, it is positive in pelvic appendicitis when the appendix is in close proximity to obturator internus muscle. Due to the proximity to the inflamed organ the muscle fibers irritated. Hence, when such patient is asked to internally rotate the right hip joint he will experience pain.

Fothergill's Sign

Presence of a tender mass overlying one of the recti which does not cross the midline and is palpable even when the muscle is made taut is indicative of rectus sheath hematoma.

A careful history will tell the clinician that the swelling was of acute onset and had etiological factors

like a bout of severe cough, or any sudden strain on the rectus abdominis.

Klein's Sign (Cf. Alder's Sign)

Useful in differentiating acute appendicitis from mesenteric lymphadenitis. In many instances the clinical features of acute appendicitis are indistinguishable from those of mesenteric adenitis. In such cases if the patient is asked to lie on the left side the point of maximum tenderness will more to the left side in mesenteric adenitis but not in acute appendicitis. But one has to remember that in Meckel's diverticulum also the tenderness might move to the left in a similar manner.

Mallet-Guy Sign

It sometimes looked for in chronic pancreatic inflammation. Patient is asked to lie on his right side. Deep palpation in the left subcostal and epigastric region will elicit pain if pancreas is inflamed.

Murphy's Sign

This is a sign of acute cholecystitis. This helps to clarify the situation when doubt exists as to the underlying pathology causing pain in the right hypochondrium.

Patients asked to take a deep breath while the clinician does deep palpation in the right

hypochondrium. Patient will hold his breath midway through inspiration as the inflamed gallbladder touches the abdominal wall. Admittedly this causes discomfort to the patient and hence the test should not be repeated without sufficient reason.

Rebound Tenderness (Blumberg's Sign)

All the abdominal signs this one is perhaps the most significant inasmuch as its presence usually indicates problem of a serious nature. Often the patient would require a laparotomy as rebound tenderness is an indicator of inflammation of the parietal peritoneum. And most cases of peritoneal inflammation or peritonitis do need surgical intervention. Hence, it is imperative that we become adept at this sign.

One should remember that rebound tenderness is looked for only when there are other clinical features of peritoneal pathology like pain abdomen of acute onset which is exaggerated by cough or movement, vomiting, constipation, etc. Hence, one should be gentle in trying to elicit rebound tenderness. If the diagnosis is already beyond doubt (e.g. X-ray showing gas under the diaphragm, etc.) one should not inflict more pain on the patient just to satisfy one's curiosity.

Once it has been decided that rebound tenderness should be looked for to clarify the situation, the nature of the test should be explained to the patient. Then he/she should be requested to keep the abdomen as relaxed

as possible. A round of superficial palpation will usually point to the site of maximum tenderness. Here the clinician should palpate deeply watching patients face all the while. Then he should maintain the pressure for a brief while then release the pressure abruptly. If one were to observe patient's face during the whole maneuver one can notice that patient winces during initial palpation, shows sign of less discomfort during the phase of sustained pressure and winces in pain again when the palpating hand is abruptly lifted.

The pain during the test is due to the movement of the sensitive parietal peritoneum. During initial deep palpation movement of the parietal peritoneum causes pain, but during sustained pressure, as there is no further movement there is a dip in the painful stimulus. But when the hand is released abruptly parietal peritoneum springs back into it original state causing even more pain to the patient.

Gently performed this test can clarify the diagnosis in condition like acute appendicitis, perforative peritonitis in its early stages, pelvic inflammation, etc. But one should remember that it needs a highly cooperative patient for the successful eliciting of this sign. An apprehensive patient would simply resist any palpation by keeping the abdomen guarded. Of course beyond a certain threshold of pain, nature takes over by making the abdomen wall rigid precluding any further palpation.

While checking for rebound tenderness the active part of the test is the abrupt release the palpating hand. One should resist the temptation to make exaggerated movements during the release. Often it is seen that after release of the pressure the forearm is taken back in a dramatic arc by the clinician. But one would do well to remember that once off the abdominal wall forearm movement has no effect other than dramatics. Such antics only succeed in making the patient anxious.

Rosenbach's Sign

Loss of abdominal reflex when the viscera are inflamed is called Rosenbach's sign. Normally, when skin of the abdominal wall is stimulated there is contraction of the abdominal wall muscles. This is called superficial abdominal reflex. Contraction of the same muscles when the neighboring bony points are tapped is named deep abdominal reflex.

Abdominal reflex is also sometimes called epigastric reflex or supraumbilical reflex.

When the underlying intraperitoneal viscera are inflamed abdominal muscles tend to go into spasm as a protective response. Hence, there is loss of abdominal reflex.

Rovsing's Sign

This is a sign which is sometimes present in acute appendicitis. If the clinician presses the abdomen in the left iliac fossa patient may complain of pain in the

right iliac region. Displacement of air and fluid within the colon proximally may be the reason for this phenomenon. The displaced air stimulates the inflamed appendix and cecal mucosa thus causing pain. Direct displacement of the abdominal contents, thus disturbing the inflamed appendix may be another factor contributing to the increased pain.

Rovsing's sign can be elicited with less discomfort to the patient when compared to rebound tenderness but it is not very reliable. Absence of Rovsing's sign does not rule out appendicitis.

Ten Horn's Sign

This sign is said to be present when the patient feels pain on gentle traction of the right spermatic cord. If present, it indicates the presence of acute appendicitis. Traction applied on the right spermatic card produces downward movement of the cord contents some of which like gonadal vessels lie in close proximity to the viscera in the right iliac fossa. The disturbance then produced of these viscera especially cecum and inflamed appendix may induce pain. Of course one has to rule out funiculitis before seeking out this sign.

SIGNS ON PERCUSSION AND AUSCULTATION OF ABDOMEN

These are considered together as the number of signs under these headings are comparatively small. But one

should remember that both auscultation and percussion cause much less discomfort to a patient with abdominal pain than palpation. Hence, auscultation of the abdomen has to be completed immediately after inspection followed by percussion keeping potentially pain inducing palpation to the last.

Let us examine a few signs under this category.

Balance's Sign

This refers to persistent dullness in the left hypochondrium and left lumbar regions and shifting dullness in the right flank typically present in rupture of the spleen. When there is bleeding due to splenic trauma there is hemoperitoneum which is the reason for shifting dullness in the right flank, but left flank will have persistent dullness due to the presence of perisplenic hematoma and clots.

Relevant history of trauma, signs of internal hemorrhage, external evidence of injury like lacerated wounds, contusion, rib fractures, etc. should make the diagnosis clear without having to wait for Balance's sign to appear. This is a surgical emergency and earlier the bleeding is controlled surgically the better. Hence, if splenic injury is suspected, once the airway, breathing and circulation are stabilized the diagnosis should be confirmed by imaging studies like USG and /or CT and arrangements are made for immediate blood transfusion and operative management.

Federici's Sign

In massive pneumoperitoneum, cardiac sounds are clearly heard with the stethoscope kept over the abdomen wall. This is due to unhindered conduction of sounds through air contained within the peritoneal cavity.

Kehr's Sign

It is the pain felt in the left shoulder by the patient who has ruptured spleen. The underlying mechanism is the irritation of left dome of diaphragm by the blood. The resultant pain stimulus traveling via the phrenic nerve is referred to the left shoulder also as supraclavicular nerves share the same root value as the phrenic nerve.

This sign is almost always present in a case of splenic rupture. But it is for the clinician to ask about it and analyze it. Patient is unlikely to volunteer this information as he is more likely to be bothered about the severe abdominal pain he would certainly have because of ruptured spleen. Also in a case of polytrauma there are likely to be other direct injuries on the thorax and upper limb. Hence, presence of Kehr's sign gets masked.

This sign is discussed here for convenience of reference even though it is not elicited on percussion.

Kenawy's Sign

It is the venous hum heard when one auscultates just below the xiphoid process. It is due to splenic vein engorgement associated with portal hypertension with splenomegaly. The hum gets accentuated during inspiration as the flow to the splenic vein increases due to compression of the spleen by the descending diaphragm.

Puddle Sign

This is used to detect minimal ascites. As low as 150 ml of intraperitoneal fluid can be detected by this test, if carefully done. Patient is asked to lie prone for 5 minutes then assume knee elbow position. The free intraperitoneal fluid collects in the dependent periumbilical area. This usually manifests as dullness in the periumbilical region or in the most dependent part of the abdomen.

The level of the fluid can be deduced by keeping the bell of the stethoscope on the most dependant part and auscultating while one of the flanks is flicked regularly with fingers. Stethoscope is gradually moved towards the opposite flank with the patient maintaining the knee elbow position. A sharp increase in the intensity of the sound indicates the level of the fluid.

Toma's Sign

This is used to distinguish between inflammatory ascites from ascites due to noninflammatory causes like portal hypertension, cardiac failure, hypoproteinemia, etc.

In inflammatory ascites there is gradual contraction of the mesentery which ultimately draws most of the intestines to the right. This is logical to expect, as the line of attachment of the mesentery is from left to the right, starting just to the left of L2 vertebra to the upper end of right sacroiliac joint. Hence, when the patient with inflammatory ascites lies supine one can elicit tympanic note on the right side whereas the note is more uniformly dull on the left side.

RADIOLOGICAL SIGNS IN ABDOMEN

In evaluating a case of abdominal pathology, either acute or chronic, many a time clinical examination proves inconclusive. One has to resort to investigations in such cases to arrive at a diagnosis. One of the simplest investigation that is can be done easily in any hospital at any time of the day or night is an X-ray of the abdomen, taken both in erect and supine position. A chest X-ray with domes of diaphragm is also useful in classifying doubtful cases of upper abdominal pathology. Let us now examine a few important and relatively easy radiological signs so as to finetune one's

diagnostic acumen without having to resort to radiologist's help all the time.

Benz's Sign (Crow Foot or Seagull Sign)

It is indicative of gallbladder stones. About 90% of the gallbladder stones are radiolucent and hence do not show up on plain X-ray. But when they do contain enough calcium to be radiopaque one might be able to appreciate lucent areas within the radiopaque stones indicating gradual build-up of the stones around an inflammatory focus. If the lucent area is tri-radiate then it is called Mercedes Benz sign or crowfoot sign after the emblem of the famous car company. If it is biradiate it is called "seagull in flight" sign, as it resembles seagull with its wings spread apart.

Claw Sign

It is a sign indicative ileocolic intussusception on barium enema study. The barium enema casts a claw like opaque shadow around the apex of the intussusceptum. It is safe to do a barium enema study whenever large bowel obstruction is suspected, one might emphasize here.

Coffee-bean Sign (Omega Sign)

This is seen on plain X-ray in a case of sigmoid volvulus. There is a huge loop of bowel occupying

almost the whole of the abdomen with a central bifurcation resembling one-half a coffee-bean. Occasionally, the gas shadow may resemble the Greek letter Omega. Then it is called Omega sign.

In both the above cases it is apparent that the loop in question is arising from the pelvis.

Colon Cut-off Sign

It is a sign detected on plain X-ray of the abdomen taken in a patient suspected of acute pancreatitis preferably in erect posture. The radiolucent shadow cast by the gas in the transverse colon seems to come to an abrupt end near the mid transverse colon due to the proximity of this part of the bowel to the inflamed pancreas, producing the classical appearance of colon "cut-off".

Cupola Sign

It is a sign of free gas within peritoneum. If a chest X-ray including both the domes of the diagram is taken in erect posture then the free gas if present will collect beneath the domes of diaphragm as a dark radiolucent crescent.

Free air in the peritoneal cavity may be due to perforation of a hollow viscus, postlaparotomy or laparoscopy or may follow certain gynecological procedures live vaginal douche, hysteroscopy, etc.

It is said a good quality radiograph can detect as little as 1 ml of free gas in the peritoneum.

Double Bubble Sign

This is a sign of duodenal obstruction. A plain X-ray in such a case will show two bubbles of gas in the upper abdomen. This is due to distension of the stomach as well as the proximal duodenum, separated by the pylorus.

Duodenal obstruction is not so uncommon in the newborn due to duodenal atresia. X-ray helps in early diagnosis of this conduction.

In the adults, stricture, annular pancreas, neoplasm, etc. can be the cause of duodenal obstruction. Here barium study, upper GI endoscopy, etc. can be done to clarify the diagnosis further.

Football Sign

In young children all the free air might collect under the anterior abdominal wall, which in a supine abdominal X-ray will cast a football like shadow. This is called football sign.

Medusa Lock Sign

This is characteristic of intestinal obstruction due to roundworms. These worms will have narrow columns of gas in their alimentary canals. These may show up on a plain film as coiled locks of hair when there are large numbers of worms obstructing the lumen. Hence,

the name "Medusa Lock sign" after the Greek mythological character.

Reverse 3 Sign

It is barium study sign seen in cases of carcinoma of ampulla of vater. Barium in the widened C loop of duodenum with the lesion in the ampulla contribute to this appearance.

Stierlin's Sign

It is a radiological sign discernible on barium meal follow through study.

There is constant emptying of the terminal ileum and the cecum with barium remaining either proximal to it or distal. This is due to the intestinal hurry caused by inflammatory process most commonly tuberculosis.

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Chapter TWO

Thyroid Signs

Clinical examination of the thyroid gland is another fascinating experience for students of surgery. A goiter is a quintessentially surgical problem which excites any surgeon worth his salt. Hence, a patient who happens to have a goiter is likely to be popular amongst students, through he/she is not likely to relish such popularity.

SIGNS IN THYROTOXICOSIS

All the clinical conditions affecting thyroid gland thyrotoxicosis perhaps has the most florid features. A properly taken history and careful examination make the diagnosis clear enough in most cases. But somehow more than the effects of excess thyroid hormone on the target organs, exophthalmos or ophthalmopathy that occurs simultaneously has been the source of most of the signs associated with thyrotoxicosis. While there is a long list of such "eye signs" which impresses beginners no end, one has to realize that the presence of these signs not really necessary to diagnose and treat a case of thyrotoxicosis. The names that these myriad eye signs go by are of academic interest only. Students should try to understand the underlying pathology which results in the particular sign. Whether or not one can recollect the name should not matter. But unfortunately in our exams sometimes more stress is laid on the students' recollection of such names than his grasp of the underlying mechanism. Hence, an

attempt has been made to list as many eye signs as one could collect arranged in alphabetical order. If readers can send me more signs with adequate references it will be gratefully acknowledged.

Abadie's Sign

This refers to spasm of levator palpebrae superiors due to excessive sympathomimetic activity.

Ballet's Sign

External ophthalmoplegia without internal ophthalmoplegia. External ophthalmoplegia can be partial or complete.

Becker's Sign

This refers to abnormal pulsation of the retinal vessels. Needless to say, this necessitates endoscopic examination.

Boston's Sign

This refers to the jerky lowering of upper eyelid when the eyeball is rotated downwards. One has to differentiate this from Von Graefe's sign where the reference is to upper lid lag and not to its jerky movement.

Dalrymple's Sign

There is widening of palpebral fissure such that the upper sclera is clearly visible, in Grave's disease. This is primarily due to spasm of the levator palpebral superioris causing retraction of the upper eyelid.

Enroth's Sign

This refers to abnormal fullness of eyelid seen in Grave's disease. This is due to edema and is more manifest in the upper eyelid than lower eyelid.

Gifford's Sign

In thyrotoxicosis due to Grave's disease upper eyelid cannot be everted easily. This is again due to spasm of levator palpebrae superioris.

In cases of proptosis of the eye due to intraorbital tumors though the outward clinical picture may appear similar, Gifford's sign is absent and upper eyelid can be everted easily.

Graefe's Sign

Syn: Von Graefe's Sign

In Grave's disease when the patient is asked to follow a finger moved slowly up and down in front of his upper eyelids persistently lag behind. One can confirm the

presence of this lag by noticing the corneoscleral limbus clearly moving down ahead of the upper eyelid.

This sign has to be differentiated from 'pseudo-Graefe's sign which is the lagging behind of the upper eyelid due to paralysis of oculomotor nerve.

Griffith's Sign

This is the opposite of Graefe's sign. When the patient is asked to follow a finger moved in upward direction in front of his eyes the lower lid lags behind the corneoscleral limbus persistently.

Grocco's Sign

In Grave's disease whenever the patient puts in a muscular effort there is acute dilalation of the heart.

Jellinek's Sign

Brownish pigmentation of the upper eyelid seen in cases of Grave's ophthalmopathy is referred to as Jellenek's sign.

Jendrassik Sign

This refers to the paralysis of extraocular muscles. The difficulty in moving the eyeball upwards and outwards is the most commonly noticed defect.

Joffroy's Sign

When a patient with exophthalmos is asked to look upwards with the head is bent downwards there will be no noticeable wrinkling of the forehead as would occur in normal individuals.

Knies' Sign

Inequality in pupillary dilatation seen Grave's ophthalmopathy.

Kocher's Sign

Mean's Sign, Globe Lag Sign

Here when examiner's hand held at the level of patients' eyes is suddenly raised higher the upper eyelid moves up more rapidly than the eyeball.

Loewi's Sign

Pupils dilate readily with epinephrine in primary thyrotoxicosis.

Mobius's Sign

Inability to achieve convergence of the eyeballs while looking at a near object. This is due to paresis of the medial rectus muscle.

Mann's Sign

Syn: Dixon Mann's sign

In Grave's ophthalmopathy the two eyeballs do not appear to be at the same level.

Plummer's Sign

If refers to patients inability to walk up a stair case. This is mainly due to proximal myopathy weakening the muscles necessary for such activity. This is one sign in primary thyrotoxicosis which is not related to ophthalmopathy.

Rosenbach's Sign

It is the fine tremor of the upper eyelid, noticeable especially when the eyes are gently closed. This sign shares its name with the abdominal sign where the abdominal reflex is lost due to inflamed viscera.

Sainton's Sign

In Grave's ophthalmopathy, frontalis muscle contracts after the cessation of levator action during upward gaze. Normally, these two muscles act simultaneously when the patient gaze upwards.

Stellwag's Sign

This refers to infrequent blinking of the eyelids in Grave's disease. Also, the blinking is incomplete. The reasons are obvious.

Snellen's Sign

Syn: Reisman's Sign

Bruit heard over closed eyes in Grave's ophthalmopathy.

Topolanski's Sign

There is pericorneal congestion in Grave's ophthalmopathy.

SIGNS ASSOCIATED WITH THYROID PATHOLOGY OTHER THAN THYROTOXICOSIS

Berry's Sign

Disappearance of the carotid pulse in a patient with goiter is an ominous sign. If usually means the infiltration of carotid sheath by thyroid malignancy.

Pemberton's Sign

This sign is present when the patient has a retrosternal goiter. Patient is asked to raise both his/her hands above

the head and maintain that position for a few minutes. In case of retrosternal goiter patient will develop facial congestion and engorgement of major neck veins. This is due to obstruction to venous drainage occurring at the thoracic inlet due to pressure by the goiter. One has to keep in mind that patient might actually faint during this maneuver. Hence, the test should be done only till the first signs of congestion are apparent.

Kocher's Test

This has to be differentiated from Kocher's sign seen in thyrotoxicosis.

This test helps to detect tracheomalacia in patients with long standing large goiters. Mild pressure with thumb and fingers of examiners right hand on both the lobes will produce strider due to loss of elasticity of the tracheal rings. Such patients might have strider and breathlessness postoperatively following removal of the gland.

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Chapter

THREE

Signs Pertaining to Other Organ Systems

These signs are discussed together as they are comparatively fewer in number. Once again students of surgery are urged to understand the mechanism behind these signs rather than just trying to memorize them. And these signs are more useful when analyzed together with relevant history and proper clinical examination. One should not rely on any one clinical sign only to include or rule out pathology of a particular organ.

Let us now examine a few of these signs:

SIGNS IN TORSION TESTS

Angell's Sign

This sign is present in cases of torsion of testis where the predisposing factor is the presence of a long mesorchium.

The presence of mesorchium cannot be made out in the already torted testis, but can easily be made out on the opposite side. The opposite testis lies horizontally. One can also feel the gap between the testis and epididymis palpation.

Deming's Sign

This is another sign of torsion tests which helps to differentiate if from acute epididymorchitis. In case of torsion testis the affected testis lies at a higher level

than its fellow. This is because of the shortening of the cord due to the twist in it. Cremasteric muscle spasm also contributes to the higher placement of the testis.

Prehn's Sign

Elevation and support to the scrotum on the side of the pathology relieves the pain in epididymorchitis but not so in torsion testis.

The reason for this is obvious, i.e. in epididymorchitis at least part of the pain is due to the dragging effect of having an edematous testis on the spermatic cord. It gets relieved with support. But in torsion testis the pain is due to ischemia which remains unaltered even in elevated position.

Differentiating torsion tests from acute epididymorchitis is of great importance because of the danger of gangrene of the testis in case of torsion if prompt surgical correction is not done. Hence, the significance of these signs cannot be over emphasized. However, if one is not sure about the diagnosis in an emergency setting it is better to follow that time tested adage "when in doubt, open and see". Of course, if it can be arranged promptly Doppler ultrasound study of the testicular blood flow will be helpful, but it should not be at the cost of too much valuable time. A negative exploration is never as important as delayed surgery resulting in loss of testis.

SIGNS IN LATENT TETANY

Tetany is a state of abnormal muscular spasm induced by deficiency of ionic calcium in blood. It can occur in hypoparathyroidism, alkalosis, massive blood transfusion, etc. Since it is a potentially life threatening condition one has to recognize tetany when it is still in its latent stage. This is where knowledge of signs of latent tetany will help. A few of the more important signs have been discussed below:

Chvostek's Sign (Syn: Weiss Sign)

This is present whenever there is hypocalcemic tetany. Normally, this sign is elicited by tapping lightly in front of the tragus to stimulate the branches of the facial nerve. In case of tetany there is muscular twitching of the facial muscles most notably at the angle of the mouth.

The presence of these signs should alert the clinician to the lurking danger of cardiac problems. Hence, immediate parenteral calcium supplementation is necessary whenever this sign is evident. Hence, its clinical significance cannot be over emphasized.

Escherich's Sign

This is another, albeit less popular sign of latent tetany. Tapping the skin near the angle of the mouth leads to protrusion of the lips.

Peroneal Sign

Tapping ever the peroneal nerve at the fibular neck will trigger dorsiflexion and abduction in latent tetany.

Trousseau's Sign

This refers to the induction of carpopedal spasm by temporarily blocking arterial supply to the hand. This is done by the application of sphygmomanometer cuff to the arm, inflating above the systolic pressure and maintaining it for about 3-5 minutes. During this time due to lack of blood supply latent tetany becomes overt and the intrinsic muscles of the hand go into and spasm producing the so-called "obstetrician's hand (accoucheur's hand)". Of course do not forget to release the pressure after you have confirmed the presence of the sign.

One has to remember that the name Trousseau's sign also refers to another phenomenon, i.e. occurrence of migrating thrombophlebitis in patients with visceral malignancies especially with carcinoma of the pancreas.

SIGNS IN DEEP VEIN THROMBOSIS

Deep vein thrombosis is a condition which can develop silently in a hospitalized patient and has the potential to kill the patient via pulmonary embolism. It is widely accepted that presence of deep vein thrombosis is much wider than is commonly believed. Hence, clinicians

have always made efforts to detect it in time. Hence, the different signs. Students are warned that some of these signs are highly subjective and mostly elicited clumsily. Occasionally vigorous attempts at eliciting a sign may cause more harm than good by dislodging a hitherto quiescent thrombus. In today's world where most well equipped hospitals have access to color Doppler study of the venous system, these signs are slowly losing their significance. However, they are mentioned here for the sake of completion.

Homan's Sign

Dorsiflexion of the foot with the knee bent produces pain in the calf whenever there is established thrombosis in the veins of the leg. One has to dorsiflex the foot slowly and gently for fear of dislodging the clot. Also sudden dorsiflexion might induce calf pain even in otherwise normal individuals.

Rupture of a popliteal cyst also produces a similar picture with positive Homan's sign. Inadvertent initiation of anticoagulants in this clinical scenario is, needless to say, disastrous.

Moses' Sign

Squeezing of the calf muscle produces pain when there is established deep vein thrombus.

This sign has been deleted from most of the textbooks now as repeated attempts to elicit it are disastrous to the patient.

Whenever there is any doubt about the presence it is better to perform duplex scan to confirm or exclude deep vein thrombosis.

The most prudent approach for a surgeon would be to take prophylactic measures like low dose anticoagulants, early mobilization, calf muscle exercises, elastocrepe bandage application, etc. in the immediate postoperative period. Better be safe than sorry!

Payr's Sign

This is a sign of local thrombophlebitis and not of deep vein thrombosis. It has been included here for convenience of study.

In cases of thrombophlebitis of the foot vein the fact may not be clinically obvious on inspection due to the thick skin of the sole. Pain on palpation over the sole indicates the presence of thrombophlebitis.

SIGNS OF VISCERAL MALIGNANCY

Leser-Trélat Sign

This refers to the sudden appearance and rapid increase in the number of seborrheic keratosis in patients

harboring internal malignancy. The condition is usually associated with pruritus.

Trousseau's Sign

Patients harboring internal malignancies like carcinoma of the pancreas, stomach, etc. often have migrating thrombophlebitis even before the primary lesion becomes symptomatic. Hence, a clinician with high index of suspicion should look for these life threatening conditions when a patient presents with innocuous looking thrombophlebitis.

It may be noted that this sign shares its name with the obstetricians hand sign of latent tetany.

Troisier's Sign

It is the enlargement of Virchow's node in the left supraclavicular region in malignancies of intraperitoneal organs or genitalia. Though it may rarely occur as the presenting symptom in these cases, it invariably signifies poor prognosis.

SIGNS IN PERIPHERAL VASCULAR DISEASE

Sign of Disappearing Pulse (De Weese Sign)

This sign may be present in cases were peripheral vascular deficit is present but clinically the pulses are palpable. If in such a patient ischemia is strongly suspected based on history then the patient is asked to

exercise or walk to the point of claudication and then the pulse is again looked for. The pulse which was earlier present will now be absent. This is due to vasodilatation that results from exercise. The deficient blood flow which was enough for pulse to be felt at a resting stage will now become insufficient to impart a palpable pulsation.

Harvey's Sign

This is an indirect method of inferring reduced arterial flow to the limbs. In case of arterial deficit obviously the venous flow is sluggish as well. This can be deduced by assessing venous refilling. A segment of vein is emptied of the blood by placing index fingers of both hands firmly on the vein, initially side by side and later moving them apart. Now if the finger farther from the heart is lifted the refilling of vein is slower in an ischemic limb than normal.

A similar deduction can be drawn by lifting the limb above the level of the heart to empty the veins and then placing it horizontal. In a normal limb veins fill up within 5 seconds in a severely ischemic limb this may take up to 30 seconds.

Fuchsig Sign

This helps to detect the presence of popliteal pulse. This pulse is difficult to palpate even is normal individual. Hence, this indirect method is sometimes employed to detect its presence. Patient is asked to sit on a chair

cross legged with the leg in question over its fellow, knee crossing knee. In a relaxed patient if the popliteal pulse is present then the upper leg will oscillate synchronous with the pulse. A negative test may mean either a very weak pulse or absent one.

SIGNS IN ARTERIAL ANEURYSM

In today's world of increasing subspecialization a general surgeon is rarely called upon to manage a known case of aneurysm. Hence, the signs which are going to be described are rarely, if ever, going to be seen in a surgical ward. But in a patient who is in the surgical ward for some other complaint, if any of these signs are noticed it is worth while ascertaining the cause. Early diagnosis of a major artery aneurysm might mean the difference between life and death for the patient concerned.

Again the signs are described in alphabetic order.

Bozzolo's Sign

Visible pulsations of the arterioles in the nasal mucous membrane is believed to indicate the presence of aneurysm of thoracic aorta.

Cardarelli's Sign

In aneurysms the aortic arch there will be transverse pulsations of an endotracheal tube, if the patient has

been intubated. This is due to transmission of arterial pulsation to the nearby trachea.

Dorendorf's Sign

In aneurysm of the aortic arch there will be fullness of one of the supraclavicular fossae.

Delbet's Sign

In case of aneurysm of the main artery of a limb, absent distal pulses, if nutrition of the distal limb is maintained then the collateral circulation is deemed to be sufficient for maintaining the vascularity of the distal limb. The nutrition of the distal limb can be deduced by looking for signs of chronic ischemia like thin and shining skin, loss of subcutaneous fat brittle nail, etc. Loss of distal pulse alone does not indicate poor vascularity.

Drummonds Sign

Puffing sound synchronous with cardiac systole heard from the nostrils especially when the mouth is closed in some cases of aortic aneurysm.

Glasgow Sign

Systolic murmur heard over the brachial artery in aneurysm of the aorta.

Porter's Sign (Oliver's Sign)

Tracheal tug noticeable in aneurysms of arch of aorta.

Similar tug may be there when carcinoma of the main bronchus becomes adherent to arch of aorta.

Queenu Muret Sign

This is another sign indicating the maintenance of distal blood supply in a case of aneurysm of main artery of a limb proximally. If in such a case a needle prick is made distal to the aneurysm, free flow of blood indicates satisfactory collateral circulation.

Both this as well as Delbet's sign have to be necessarily looked for if the proposed treatment for the aneurysm is ligation of the artery. Of course, with the advent of aneurysmorrhaphy, grafts and stents, ligation of an aneurysm is only of academic interest now.

Perez's Sign

Crepts heard over the upper chest when the arms are alternatively raised and lowered in case of aneurysm of aortic arch.

This is also positive in case of fibrous mediastinitis.

Branham's Sign (Nicoladoni's Sign)

Strictly speaking, this is not a sign of any aneurysm but of arterio-venous malformation.

Whenever arteriovenous fistula's is suspected, if the feeding artery to the lesion is compressed then not only

the size of the AV malformation reduces but the associated bruit disappears. Also there is a fall in pulse rate and normalization of pulse pressure. Of course in these days of duplex scanning this sign is hardly ever sought for. Blocking the feeding vessel will interrupt the hyperdynamic circulation caused by shunting of blood. This results in all the changes in hemodynamics listed above.

SIGNS OF HERNIA

It is indeed surprising that diagnosing a hernia should need the assistance of signs. Of course diagnosing a hernia is a straightforward act most of the time but not always, especially when dealing with rare types or early stages of hernia.

Silk Glove Sign

This helps the clinician to distinguish between a normal cord and a cord with empty inguinal hernial sac. Sometimes especially in indirect inguinal herniae the contents may not prolapse into the sac at the patient's will nor after standing. In such cases, if one palpates the cord, the cord will feel as if it is wrapped in silk. The cord contents cannot be felt distinctly. This is due to the empty sac made of parietal peritoneum.

Howship-Romberg Sign

This may point to an obturator hernia but needs a very high index of clinical suspicion to detect its presence. In cases of obturator hernia pressure on the obturator nerve by the hernia contents might produce pain on the inner aspect of patient's knee.

Border Sign

This radiological sign indicates the occurrence of ventral hernia in the early postoperative period. If a barium study is done in the early postoperative period the presence of a ventral hernia or incisional hernia is detected by the sharp outlining of the inferior and lateral borders by the barium within the level loops while medial and upper borders are not well delineated.

This occurs due to the dependant state of either the lateral for inferior border of hernia in various positions of the body.

MISCELLANEOUS SIGNS

Here we discuss a group of signs which are not interconnected but are interesting all the same. They are listed under the heading 'miscellaneous' only for convenience. This arbitrary clustering of signs in no way diminishes their significance in the appropriate clinical setting.

Battle's Sign

This refers to ecchymosis over the mastoid process in cases of fracture of the middle cranial fossa. In pre CT era this sign was a very significant clinical finding indicating the gravity of the trauma.

Boyee's Sign

This is a sign of esophageal diverticulum. Often in an esophageal diverticulum food residue and saliva can be found.

If one presses the side of the neck, the diverticulum might empty the contents with a gurgling sound.

Eyelash Sign

This is used to differentiate true unconsciousness from malingering, hysteria and such other functional states. In the latter case if the eyelashes are stroked gently there will be movement of eyelids, but not in a true organic or traumatic brain lesion.

Hoover's Sign

Another sign which might help detect genuine organic lesion from functional one. If a patient having genuine paralysis one lower limb, is asked to lift that leg he/she involuntarily makes counter pressure with the heel of the other leg against the bed in an effort to lift the paralyzed leg. This counter pressure can be noticed

whether the affected limb could be moved or not. No such counter pressure is discernible in hysteria or malingering.

Macewan's Sign (Cracked Pot Sign)

Percussion over the skull of a child with hydrocephalus will give the sound skin to tapping a cracked pot.

Nikolsky's Sign

In pemphigus vulgaris rubbing the skin with slight pressure might lead to pealing off of the epidermis.

Milian's Sign

This differentiates between erysipelas and streptococcal cellulitis. Erysipelas is a streptococcal infection of the skin while cellulitis is inflammation of the subcutaneous tissue. In the head and neck region the two can be distinguished by the fact that erysipelas spreads to the external ear but cellulitis does not as there is no subcutaneous tissue in the pinna.

Kanavel's Sign

In inflammation of the ulnar bursa the point of maximum tenderness lies between the transverse palmar creases.

Ripault's Sign

It is a sign of death. It refers to permanent change in the shape of pupil by unilateral pressure on the eyeball.

Tinel's Sign (DTP Sign)

If the skin over the point of injury to a peripheral nerve is gently tapped and patient complains of tingling or "pins and needles" along the distribution of the nerve, it means that the regeneration of the nerve is under way. Similar "Distal tingling on percussion" (DTP) also occurs when the injury to the nerve is only partial.

Setting Sun Sign

As the name suggests this is a sign of grave prognosis in conditions like hydrocephalus, intracranial bleed, brain tumor, etc.

The iris appears to set behind the lower eyelid exposing large area of sclera above the corneoscleral junction.

Slip Sign

This is a popular "sign" amongst students. All it refers to is that in a lipoma the edge of the swelling slips away from the palpating finger. This palpating feel has attained the status of a 'sign' with time.

Wrist Sign

Present in Marfan's syndrome the abnormally long fingers of the patient will make thumb and little finger overlap each other when the opposite wrist is gripped.

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Chapter

FOUR

Clinical Syndromes

A clinical syndrome can best be defined as a collection of signs and symptoms which are commonly associated with a particular morbid process. For example, if any pathological condition is associated with a particular clinical picture more often than not, then all the signs and symptoms which contribute to such clinical picture may be said to constitute a clinical syndrome, e.g. Murphy's syndrome in acute appendicitis.

But a particular set of signs and symptoms in termed a syndrome only if it is consistently associated with the same morbid process and knowledge of such as association between the clinical picture and disease process helps in arriving at a diagnosis, e.g. Meig's syndrome in fibromyoma of the ovary.

Often it has been noticed that our students are worried about not knowing a particular syndrome. One who knows the most syndromes is likely to be considered the most well read amongst his peers. This state of affairs is partly due to the undue importance attached by some examiners to the recalling of names of vague clinical syndromes. But it is necessary to remember that it is all very well if one knows about these obscure syndromes after knowing common conditions. If not, it is far better to know only common conditions now, only then notice syndromes. After all, one's clinical career involves treating common conditions 99% of the time. One is not likely to be accused of missing a rare syndrome. But even a master

of syndromes is likely to be held responsible if a common disease process is overlooked in the search for exotica. Some one has rightly said "if you make a rare diagnosis you are rarely correct".

Why then should we have a section on syndromes? The reason is simple. I have observed students spending hours in the library looking up reference books just to get the name of a particular syndrome. Many books, especially the western ones mention only more significant syndromes and skip the rest. In order that an interested student gets basic information on most surgical syndromes at one place this chapter has been included. Once again I remind my students that knowing these syndromes is secondary to a proper study of common clinical conditions.

Here too the syndromes have been discussed in the alphabetical order of their names. I have tried to discuss in brief only fen amongst them which I think are most relevant. Wherever possible synonyms have been mentioned within brackets.

AURICULOTEMPORAL NERVE SYNDROME

Syn: Frey's syndrome: Gustatory Sweating

This refers to flushing, sweating and hyperaesthesia in the region of the ear and cheek in response to mastication of food. This occurs due to erroneous transmission of parasympathetic stimulus meant for

stimulating salivation through the sympathetic channels innervating skin and its integuments. Often such an error follows trauma to the region or surgery on the parotid gland. Following such insult there is cross connection between parasympathetic fibers from otic ganglion and sympathetic fibers from the superior cervical ganglion. Hence, whenever the patient eats the impulses meant to stimulate the parotid gland actually go to the skin stimulating sweat glands, blood vessels etc. Hence the clinical picture of sweating and flushing.

If the syndrome follows surgery or trauma, then only reassurance and local anti perspirant spray is sufficient. Usually symptoms improve in 6 months. Occasionally, if the symptoms become annoying enough, then surgical section of Jacobson's nerve or tympanic branch of glossopharyngeal nerve or vidian nerve.

AFFERENT LOOP SYNDROME

This occurs following gastrectomy and Billroth II anastomosis. If there is obstruction to the flow of content of afferent loop into the gastric stump, then there is stagnation and increased pressure within the proximal loop. Immediately after surgery this might result in duodenal blow out. If blow out does not happen then the patient will complain of pain and bloating sensation in the epigastrium. The bilious contents might suddenly get discharged into the gastric stump when the pressure

builds up beyond a threshold level, resulting in vomiting. Increased pressure within the duodenum might lead to acute pancreatitis due to contents entering pancreatic duct.

If left untreated continued dilatation of the proximal jejunum and duodenum may lead to gangrene of the bowel.

Afferent loop obstruction needs surgical treatment most of the time. Common operation done are enteroenterostomy between proximal dilated and efferent loops. Alternatively the distal portion of the afferent loop can be resected and the anastomosis converted to a Roux-en-Y type.

The common causes for afferent loop syndrome are too long an afferent loop, kink in the loop, volvulus, jejunogastric intussusception internal herniation etc. Hence it is advisable to keep the afferent loop as short as possible and close all potential spaces for internal herniation.

BECKWITH-WIEDEMANN SYNDROME

Syn: EMG Syndrome

In this neonatal syndrome, there is association between exomphalos, macroglossia and gigantism. Hypoglycemia is another dangerous feature of this syndrome which needs to be expected and promptly dealt with to avoid fatal outcome.

This syndrome has an autosomal recessive inheritance. It is also called the "EMG Syndrome", an acronym for its three important clinical features, i.e., exomphalos, macroglossia and gigantism.

BLAND-WHITE-GARLAND SYNDROME

This syndrome refers to the anomalous origin of left main coronary artery from the pulmonary artery; there is flow of deoxygenated blood from pulmonary artery to the left main coronary. Patient will have exertional dyspnoea due to resultant ischemia and cardiac failure. The infant may survive into adulthood only if there is good collateral circulation between right and left coronary arteries.

Cardiac catheterization and angiography are diagnostic as the clinical picture is usually not specific. Treatment is surgical re-implantation of left main coronary artery directly to the aorta. Mortality in untreated cases is very high.

BLIND LOOP SYNDROME

This is the clinical picture usually seen in patients who have undergone bowel surgery, most classically end to side anastomosis. If the proximal residual segment of either limit of an end to side anastomosis be it jejunum, ileum or colon is allowed to be more than 1.5 cm beyond the anastomosis then that segment will not drain

properly leading to stagnation and bacterial overgrowth. This change in intestinal microflora will lead to malnutrition. Patient will have malabsorption especially for fat, vitamins and other micronutrients. Patient will have symptoms of dyspepsia, flatulence, abdominal pain, anemia, etc. and may even suffer from frank intestinal obstruction.

Treatment involves giving rest to the bowel keeping the patient nil by mouth and inserting a Ryle's tube especially if there is subacute intestinal obstruction. If the patient can take oral feeds, then broad spectrum antibiotics like doxycycline with or without metronidazole should be given to control the luminal bacterial overgrowth. This coupled with probiotics like lacto bacillus spores given orally should be able to restore the intestinal micro flora. The only problem with this line of management is that the patient is likely to have only a temporary relief before the condition recurs. The final court of appeal is surgery wherein the blind loop is resected if technically feasible. Hence one has to prevent the occurrence of blind loop syndrome during the first surgery itself, by keeping the blind loop as short as possible under the circumstances.

Similar clinical picture might be seen following side to side anastomosis by-passing a non-resectable growth or in a particularly difficult case of adhesive obstruction where the exact point of obstruction can not be reached due to dense adhesions. Since these are unavoidable

situations one has to warn the patient about possibility of the blind loop syndrome and he should be advised about appropriate nutritional support.

Another condition where occasionally a blind loop like picture develops spontaneously is intestinal tuberculosis or any other condition causing multiple strictures of the bowel. The segment between two consecutive strictures is partially obstructed and hence leads to stagnation and all its associated problems. Here the treatment involves resection of the entire segment or strictureplasty to relieve obstruction.

BOERHAAVE'S SYNDROME

This refers to an acute surgical emergency where in there is perforation of the lower oesophagus due to barotrauma. It usually follows a violent bout of vomiting and retching. Patient complains of severe pain in the epigastrium and retrosternal region. There might be symptoms of the pleural effusion. On examination patient appears toxic and is most cases has surgical emphysema. This condition needs a high index of suspicion on the part of clinician for diagnosis. And unless emergent management is instituted patient likely to succumb to massive thoracic inflammation that ensues.

If suspected then a contrast film with water soluble contrast confirms the diagnosis in most cases. Only rarely thin barium may have to be used. One has to

bear in mind that too much time should not be wasted in arriving at a diagnosis. A relevant history along with the clinical picture including surgical emphysema is almost confirmatory.

Management usually involves emergency thoracotomy to deal with the contamination and to institute drainage. The usual and prudent approach is to do an oesophageal diversion and toilet. In very early cases, direct two layers repair of the oesophagus can be attempted. Attempting oesophageal resection in an already compromised patient is a very risky affair and hence not usually practised.

In a few cases where the diagnosis has been made but the clinical picture is mild a conservative approach can be followed by keeping the patient nil per orally starting TPN and higher antibiotics. Of course if the condition of the patient does not improve rapidly then surgical intervention becomes unavoidable.

BUDD-CHIARI SYNDROME

Syn: Robi Tansky's Disease

This syndrome refers to the clinical picture which results from obstruction to the hepatic veins. The cause of obstruction is thrombosis in most cases and only occasionally is the venous obstruction due to web or neoplastic encasement of the hepatic veins. The underlying pathology for spontaneous thrombosis within the hepatic veins may occasionally be apparent

like thrombocytosis, polycythemia or oral contraceptive pills but in most cases idiopathic.

The onset can be acute or more insidious. In its acute form there will be gross hepatomegaly associated with severe right hypochondrial pain, portal hypertension with gross intractable ascites and may rapidly progress to hepatic coma and death.

When the onset is more protracted patient may develop cirrhosis, portal hypertension, oesophageal varices etc resulting in slow deterioration in health.

Diagnosis depends on high index of suspicion and getting relevant investigations like color Doppler, contrast CT, liver function tests, etc.

Definitive treatment may be possible in rare cases when the obstruction is due to a web obstructing vascular flow. Endovascular meatotomy is the procedure of choice and is done transatrially under guidance. In other cases porta systemic shunts can be done if the patient's general condition permits it but the results are not always satisfactory.

CARCINOID SYNDROME

As the name suggests this syndrome is associated with carcinoid tumors. More than 95% carcinoid tumors occur in the gastrointestinal tract and produce the syndrome complex only when they metastasize to the liver. These tumors arising from Kulchitsky cells produce substances like serotonin, kinins, histamine,

prostaglandins, etc. which are responsible for the signs and symptoms of carcinoid syndrome. The classical picture occurs in about 10% of the patients and includes flushing, angiomas, bronchospasm, diarrhoea and occasionally mental variations. There might be stenosis of tricuspid and pulmonary valves leading to cardiac dysfunction. The symptoms are sometimes induced by intake of alcohol, chocolate cheese, etc.

Diagnosis can be clinical by detecting high levels of 5-Hydroxy indole acetic acid (5-HIAA) in urine. Nuclear scintigraphy may help in localizing the tumor.

Treatment is usually symptomatic as the established syndrome usually means metastatic disease. However, it worth while resecting the primary if it is feasible.

CHILAIDITI'S SYNDROME

This syndrome causes diagnostic confusion during evaluation of acute abdomen cases. Occasionally, especially in children and in the elderly there will be interposition of transverse colon between the right lobe of the liver and the right dome of the diaphragm. In erect abdominal and chest x rays it appears as gas under the diaphragm causing diagnostic dilemma.

Chilaiditi's syndrome can be distinguished from free gas by the following features.

The gas in chilaiditis syndrome is within the bowel and hence does not form a sharp crescent. It is borders

may show slight undulation and haustral markings if seen carefully.

There will be no gas under the left dome of diaphragm in chilaiditi's syndrome.

Classical clinical picture of perforative peritonitis is absent.

COMPARTMENT SYNDROME

It is a situation wherein there is increased pressure within a confined anatomic space leading to obstruction of vascular flow. If not treated early there may be threat to the function and viability of the tissues supplied. Classically, such compartment syndrome occurs in the lower limb following trauma and hemorrhage. Infection and gangrene is another life threatening condition that can contribute to such patients death. Increased collection of extracellular fluid in infection and of blood in trauma leads to increased pressure within the musculofascial compartments of the lower limb leading to reduced blood supply to the distal limb. There will be pallor, pulselessness and paresthesia of the foot along with severe pain.

The only way to prevent gross limb oedema and gangrene of distal limb is to do an urgent decompressive fasciotomy of the deep fascia, thus relieving the pressure builds up.

Similar pathology may occur in the upper limb, peritoneal cavity or any other closed body space.

Treatment almost always involves immediate surgery if one wishes to avoid distal ischemia.

Also one has to remember the possibility of crush syndrome if the aetiology involves severe crushing trauma.

CRUSH SYNDROME

This is a shock like state that follows major crush injury of the muscles. In its classical form, it occurs after the release of the compressive force crushing the muscles, as is often the case in case of road-traffic accidents, mining accidents, earthquake, etc. The sudden release of myoglobin into the circulation may damage the renal tubules resulting in oliguria and renal shutdown. Occasionally similar unfortunate result may follow the release of tourniquet.

CRST SYNDROME

It is a syndrome usually observed in scleroderma. It is characterized by:

- Calcinosis cutis — C
- Raynaud's phenomenon — R
- Sclerodactyly — S
- Telangiectasia. — T

Scleroderma is a disease in which there is progressive thickening of the skin due to thickening of fibrous tissue with eventual atrophy of the epidermis.

CRONKHITE-CANADA SYNDROME

It is a rare condition in which there are gastrointestinal polyps, alopecia and nail dystrophy.

CRIGLER-NAJJAR SYNDROME

It is a type of familial non hemolytic jaundice wherein there is unconjugated hyperbilirubinemia. There is a congenital defect in conjugation of bilirubin to form bilirubin diglucuronide due to absence of glucuronyl transferase.

During early infancy it can cause permanent brain damage resembling kernicterus, especially when glucuronyl transferase is completely absent (type I Crigler-Najjar syndrome). In type II disease, there is only partial deficiency of the enzyme and hence the disease runs a milder course. Type I disease is fortunately autosomal recessive whereas Type II is dominant.

Phenobarbitone has been used in the treatment to induce glucuronyl transferase in the liver.

CUSHING'S SYNDROME

This refers to the clinical picture observed when there is inappropriately elevated plasma cortisol levels. The increase in plasma cortisol levels may be:

a. ACTH dependent like in the case of pituitary adenoma (also called Cushing's disease) external

administration of excess of ACTH, ectopic ACTH secretary tumors, etc.

b. Independent of ACTH, e.g. hyperfunctioning neoplasm of adrenal cortex or excessive exogenous administration of corticosteroids.

Clinical features include a diabetes like status, hypertension myopathy, osteoporosis cutaneous striae, central obesity including moon face and buffalo hump etc. Women may develop hirsutism, acne and amenorrhoea. In some patients mental depression is apparent.

Treatment depends on the underlying cause. Further details are beyond the scope of this book.

CRUVEILHIER-BAUMGARTEN'S SYNDROME

In cirrhosis of the liver umbilical and paraumbilical veins open up and there will be varicosities of the periumbilical veins. The clinical picture of dilated and tortuous veins around the umbilical is called caput medusae (medusa head appearance) after the Greek mythological character. The flow of blood will be away from the umbilical. This forms one of the sites of portosystemic anastomosis. This whole clinical entity is named Cruveilhier-Baumgarten's disease or syndrome.

CAROTID STEAL SYNDROME

Signs and symptoms of vertebrobasilar insufficiency resulting from siphoning of the blood from the vertebral artery to the external carotid artery are referred to as the carotid steal syndrome. Patient has transient ischemic attacks and spells of dizziness and loss of balance.

DUBIN-JOHNSON SYNDROME

It is a type of congenital non-hemolytic hyperbilirubinemia. There is defective transport of conjugated bilirubin into the biliary canaliculi and hence conjugated hyperbilirubinemia occurs.

Diagnosis is established by the presence of bilirubinemia poor bromsulphthalein (BSP) clearance and typical picture on liver biopsy. Prognosis is generally good.

DUMPING SYNDROME

Syn: Postcibal syndrome

It is the most distressing post gastrectomy clinical syndrome. It is of two types. In the early dumping syndrome patient suffers from vasomotor symptoms like hypotension, tachycardia, sweating and giddiness within a few minutes of food ingestion. There may be

colicky abdominal pain followed by diarrhoea. The underlying mechanism is simple. It is basically a disorder of a carbohydrate metabolism. In a patient who has had gastrectomy especially Billroth II type, there is sudden presentation of large quantities of carbohydrate to the small bowel as the "antral pump" no longer exists. This large carbohydrate load leads to sudden transient hyperglycemia which prevents further carbohydrate absorption. The carbohydrate thus left behind in the bowel, lumen draws in water from the mucosa along osmotic gradient. This results in increased bowel activity, diarrhoea as well as features of hypotension. Ingestion of more carbohydrates worsens this condition. Patient is advised to take small, dry meals poor in carbohydrates at frequent intervals. Octreotide may be of use in preventing symptoms but it is too expensive for routine use. A few cases might need surgery in the form of conversion of Billroth II to Billroth I if possible or interposition of antiperistaltic bowel segment between the stomach and the jejunum.

On the other hand, late dumping is less common as well as less distressing. Here patient develops features of hypoglycemia about 2 hrs after food intake. Here the initial hyperglycemia stimulates exaggerated insulin secretion leading to hypoglycemia. Ingestion of more food usually relieves the symptoms. This syndrome is usually treated conservatively.

GARDNER'S SYNDROME

It is one of the polyposis coli syndromes. It is inherited as an autosomal dominant trait and is associated with multiple tumors like osteomas, fibromas, desmoids, and epidermoid cysts early in life followed later by the development of multiple colonic polyps. There is increased risk of colorectal malignancy in this condition. The only definitive surgical procedure which eliminates the risk of colorectal malignancy is total procto-colectomy followed by permanent ileostomy or ileoanal anastomosis. Ileorectal anastomosis after resection of the whole colon sans rectum is less morbid but also less protective.

GILBERT'S SYNDROME

It is a type of familial non hemolytic jaundice. It generally inherited as autosomal dominant and presents itself only during adolescence. Patient develops mild jaundice (usually below 6 mg/dl) and may have anorexia, malaise and upper abdominal pain. Such episodes may be triggered by any stress like infection, starvation, etc.

Investigations reveal increase in indirect bilirubin. There is no evidence of hemolysis nor any bilirubinuria.

The underlying pathology is a deficiency of glucuronyl transferase. Occasionally the uptake of

unconjugated bilirubin from the plasma itself is impaired.

Mild cases do not require any treatment. In some cases treatment with phenobarbitone has been used to stimulate liver enzymes including glucuronyl transferase thus relieving symptoms.

HORNER'S SYNDROME

It occurs when there is involvement of cervical sympathetic pathway anywhere from hypothalamus to post ganglion nerves. It consists of ptosis (drooping) of the upper eyelid, miosis (constriction) of the pupil, enophthalmos (small, indrawn eyeball) and anhidrosis (absence of sweating in the ipsilateral hemiface. When the Horner's syndrome is due to central cause like a brain stem tumor or infarct usually other localizing signs appear making the diagnosis clear. Most of the common surgical conditions like cervical rib, Pancoast's tumor, brachial plexus trauma carcinoma thyroid etc involve the preganglionic nerves.

In post-ganglion Horner's syndrome, nerves beyond the superior cervical ganglion are affected by base skull fracture, tumor, cavernous sinus thrombosis, etc. It is noteworthy that sweating is normal when postganglion fibers only are involved.

Horner's syndrome is important clinically in that it draws one's attention to the underlying pathology which is almost always more sinister.

HEPATORENAL SYNDROME

This term is now used to indicate any renal failure occurring in patients with parenchymal liver disease when no other cause can be found. This is especially so in the immediate post operative period after surgery on the biliary tract in a jaundiced patient. Liver cirrhosis is another cause making patients vulnerable for hepatorenal syndrome. It may be precipitated by gastrointestinal bleeding, surgical intervention, abdominal paracentesis and sometimes even forced diuresis. Initially, it resembles pre-renal azotemia but does not respond well to correction of volume depletion. It may rapidly progress to acute tubular necrosis.

It is important to rule out other causes of simultaneous liver and renal damage like sepsis, shock, drugs like halothane and aminoglycoside, toxemia etc which can mimic the clinical picture of hepatorenal syndrome.

It is interesting to note that the kidneys in such patients are histologically normal and function normally when transplanted onto a patient without liver disease. Only late in the progression of hepatorenal syndrome changes of acute lobular necrosis (ATN) set in.

It is not clear what causes hepatorenal syndrome. Some believe it is the direct damage caused by bilirubin and bile salts to renal tubules. Others believe there is an element of ischemic damage in the development of this process. It is possible that there are toxins in the

serum of jaundiced patients which are noxious to the renal tubules.

Prevention as well as treatment consists of maintaining high effective plasma volume both before and after surgical intervention. As long as there are no changes of ATN then the kidneys recover fully. Once ATN is established then the treatment has to be altered accordingly. It goes without saying that one has to rule out or eliminate other causes of acute renal damage.

JOB SYNDROME

It is a variant of hyperimmunoglobulin E syndrome seen in fair skin individuals. Patient starts developing symptoms from the first year of life itself. They include recurrent seborrheic eruptions of the scalp, dystrophic nails, follicular eruptions resembling herpes and an unexplained tendency to develop multiple staphylococcal 'cold' abscesses. There are no inflammatory signs perhaps due to defective erythema response. Patient may also have recurrent otitis media, respiratory tract infection and liver and lung abscesses.

This syndrome is named after the famous biblical character Job who is supposed to have suffered from chronic furunculosis and multiple boils.

KEARNS-SAYRE SYNDROME

It is disorder of mitochondrial function. It consists of hypoparathyroidism, progressive external

ophthalmoplegia, pigmentary retinopathy and cardiomyopathy. Patient presents with ophthalmic symptoms at an early age, but the possibility of hypoparathyroidism should be kept in mind. This syndrome has been noted to occur with diabetes mellitus.

KLIPPEL-TRENAUNAY-WEBER SYNDROME

Elephantiasis Congenita Angiomatosa

Usually seen in the extremities wherein there is combination of angiomatosis and localized gigantism due to hypertrophy of bone and muscle. Hence, it is sometimes called angiosteohypertrophy. The tissue hypertrophy is presumably due to abundant vascularity of the part resulting from angiomatosis.

KOENIG'S SYNDROME

This refers to the alternating attacks of constipation and diarrhea as well as intestinal colic and gurgling in the right iliac fossa associated with ileocecal tuberculosis. Ileocecal TB, especially the ulcerative type often leads to multiple strictures in the distal ileum. This initially tends to cause sub acute obstruction and stasis (i.e. constipation). The resultant bacterial over growth ultimately causes enteritis and diarrhea.

LAMB SYNDROME

The name is an acronym for the following clinical features, i.e.

- **L** — **L**entiginosis
- **A** — **A**trial myxoma
- **M** — **M**ucocutaneous myxoma
- **B** — **B**lue naevi

Its recognition is important as there is increased risk of melanoma in these patients.

LAMBERT-EATON SYNDROME

Syn: Eaton-Lambert Syndrome

It is the proximal myopathy associated with internal malignancy. The muscle weakness is progressive and not associated with any cutaneous lesions of dermatomyositis.

Its clinical significance lies in the fact that it draws one's attention to the underlying internal malignancy.

LERICHE'S SYNDROME

First described by Leriche in 1923. Typical symptom complex as described by Leriche affects men between the age of 35 to 60 in whom there is thrombotic occlusion of aortic bifurcation. It includes extreme fatiguability of both lower limbs, symmetrical atrophy

of both lower limbs, pallor of lower limbs and most typically inability to achieve stable penile erection. The last symptom occurs due to reduced flow through internal pudendal artery and hence poor filling of corpora cavernosa.

The most common cause for thrombotic occlusion of aortic bifurcation is atherosclerosis. And as the process of narrowing develops slowly over a period of months, acute symptoms are rare.

Finally, there may be ulceration and gangrene of the lower limbs.

Diagnosis is established by color Doppler studies and arteriography. Treatment involves by passing the occlusion in most cases though in selected cases thromboendarterectomy might give satisfactory results.

LYNCH SYNDROME

Syn: 1. Familial Atypical Multiple Mole Melanoma (FAMMM) Syndrome
2. BK Mole Syndrome (after the families studied)

It is a familial disorder with multiple dysplastic naevi, transmitted as dominant trait. Many of these patients develop melanoma in these naevi. These patients are also at risk of developing primary melanoma. When two or more members of the same family are affected the chance of melanoma is deemed to be more than 50%.

MUNCHAUSEN'S SYNDROME

Syn: Hospital Hopper Syndrome

It is a term coined by Lord Asher in 1951 to describe factitious disorders wherein the patient either exaggerates or simply invents varied symptoms in order to gain attention. Typically he will have a long past history, sometimes interspersed with multiple operations, and history of having consulted many doctors.

The description of symptoms by the patient is often very convincing as the patient will have learnt over time how to present his case. Munchausen's syndrome needs to be differentiated from plain malingering wherein the patient's motivation is purely external, e.g. sickness absenteeism.

Baron Munchausen was an German army officer in the 18th century who, it was believed, told fantastic tales about his own exploits just to gain attention.

MURPHY'S SYNDROME

Syn: Murphy's triad

This phrase is sometimes used to refer to the symptom complex which accompany is early acute appendicitis i.e. the sequence of pain, vomiting and fever (hence the name Murphy's triad).

Initially, the pain is felt around the umbilicus as the source of pain is visceral. As the parietal peritoneum in the right iliac fossa becomes inflamed later in the course of acute appendicitis pain gets localized to right iliac fossa. Pain is usually followed by vomiting and not vice versa. The vomiting is the result of protective pylorospasm. Increased body temperature may be present as a result of inflammation but rarely beyond 38-39°C.

It has to be kept in mind that diagnosis of acute appendicitis is not always easy and the aforementioned classical syndrome may not always be present. One may have to depend on one or more of the factors like tenderness over McBurney's point including rebound tenderness, increased WBC count, sonological evidence of appendicitis etc to arrive at a plausible diagnosis. Some cases might even warrant CT abdomen and/or diagnostic laparoscopy.

MEIGS' SYNDROME

This is an infrequent accompaniment of benign solid ovarian tumors, most commonly ovarian fibroma. The syndrome comprises of hydrothorax (more common on the right side) and ascites.

Believed the ascites is due to seepage of fluid from the tumor into the peritoneal cavity. Hydrothorax results when such peritoneal fluid reaches the thorax via diaphragmatic lymphatics.

One has to remember that malignant ovarian tumors can also cause ascites and pulmonary involvement. There is no need to emphasize that the outlook for such patients is bleak unlike those having Meig's syndrome.

MALLORY-WEISS SYNDROME

In this there is longitudinal tear in the mucosa at the cardioesophaged junction following a bout of severe vomiting and retching. It can cause significant hematemesis and melaena. The vomiting episode usually follows a binge of alcohol but may be due to any other cause.

Typical history of vomiting first of gastric contents followed later by nonproductive retching and hematemesis should alert the clinician to the possibility of Mallory-Weiss syndrome. Fiber optic endoscopy shows the mucosal tear confirming the diagnosis. The tear is rarely deeper than the submucosa. But the possibility of through and through perforation (Boerhaave's syndrome) should be kept in mind and the same ruled out.

Treatment is conservative initially as for any other cause of upper GI bleed. Sengstaken tube tamponade is contraindicated as it may further extend the tear. Vasopressin analogues or direct adrenaline injection around the tear might be needed in some cases.

Surgery may be needed in rare cases and involves a long gastrotomy and under running the tear with

non-absorbable suture. One has to remember that occasionally there may be more than one tear.

MARFAN'S SYNDROME

It is an autosomal dominant disorder with high penetrance and variable expressivity. The genetic defect is said to be in chromosome 15. There is an inherited defect in collagen formation and function in these individuals.

The leading defects are skeletal, ocular and cardiovascular. The skeletal deformities are abnormally wide arm span, greater pubis to sole length than vertex to pubis length, arachnodactyly, hyper extensible joints, kyphoscoliosis pectus excavatum and flat feet. Great toes may be longer than other toes and skull deformities like dolichocephaly may be seen. Large deformed ears, ectopia lentis, aortic aneurysm, cutaneous striae, etc. are some of the other defects seen in Marfan's syndrome.

Mental retardation is not a feature of this syndrome. In fact Abraham Lincoln one of the greatest presidents of USA is said to have suffered from this syndrome.

Cardiovascular involvement is the most common cause of death. Propranolol is said to be of some benefit in limiting progression of the vascular defects. Surgery might be needed for aneurysm, kyphoscoliosis or ocular defects.

MALABSORPTION SYNDROME

This is a clinical condition caused by poor absorption of various nutrients from the gastrointestinal tract. Symptoms include weight loss, lassitude, anorexia, anemia, oedema due to protein deficiency etc apart from abdominal symptoms like diarrhoea, flatulence and discomfort. Specific symptoms of deficiency of specific nutrients may be present, e.g. night blindness due to vitamin A deficiency, peripheral neuropathy caused by vit. B_{12} deficiency.

Etiology includes failure of digestion as in chronic pancreatitis, cholestasis, gastric bypass surgeries, blind loop syndrome etc; or failure of mucosal absorption as in gluten enteropathy (celiac sprue) or hypogammaglobulinemia. Short bowel syndrome causes malabsorption due to rapid transit of bowel contents. Radiation enteritis and mesentenie ischemia are some of the other causes.

Investigation include endoscopic jejunal mucosal biopsy, breath tests for lactose and lactulose, barium studies and immunologic tests for hypogammaglobulinemia.

Treatment depends on the cause and has not been discussed here for obvious reasons.

MIRIZZI'S SYNDROME

It is one of the unusual causes for obstructive jaundice. Here a stone in the Hartmann's pouch presses on the

bile duct producing obstruction. In many cases there may be a cholecysto choledochal fistula at the site of obstruction. Cholangiography will show the site of obstruction but differentiation from cholangiocarcinoma may be difficult.

Treatment usually involves surgery, but one has to bear in mind that the separation of gall bladder from the common hepatic duct may damage the latter.

Another variant of Mirizzi's syndrome is when a large stone in the cystic duct just above the junction with the bile duct, obstructs common hepatic duct.

This is commonly misdiagnosed as stone in the bile duct. ERCP will clarify the picture.

MAFUCCI'S SYNDROME

It is a clinical condition in which multiple enchondromas are associated with hemangiomas, usually cavernous type. It is not a genetic disorder and hence not hereditary. There is thought to be severe mesodermal dysplasia. Hemangiomas of the skin or soft tissues is the presenting feature. Phleboliths may be seen on X-ray. Enchondromas affect metacarpals, phalanges or even long bones, not necessarily on the same limb as hemangioma.

Though there is an increased risk of chondrosarcoma in these patients, they usually live fairly long as the tumors are low grade and are more likely to die of non skeletal causes than the tumor itself.

It is worth while noting here that multiple enchondromatosis without hemangiomas is referred to as Ollier's disease.

MENDELSON'S SYNDROME

This refers to the clinical picture due to aspiration the gastric contents into the respiratory tract especially in obstetric patients following vomiting or regurgitation.

During pregnancy there are several factors which might induce vomiting or regurgitation. One is the obviously increased intra-abdominal pressure. Progesterone also is known to induce vomiting and regurgitation by its relaxing action on the gastric esophageal sphincter. Hormonal influences (probably chorionic gonadotrophin) predominate during the early pregnancy in the causation of emesis. Also psychogenic and neurogenic factors are believed to play a role in aggravating emesis.

Aspiration of gastric contents leads to a chain reaction with obvious deleterious effect. There will be a deteriorating clinical picture representing the ill effects of lower respiratory infection, pulmonary oedema and finally respiratory distress syndrome. Added to this will be the ketoacidosis which might result from severe vomiting itself.

The major decision to be made in this situation is when to terminate the pregnancy. The guiding principle

is to make sure that the crisis should not be allowed to worsen till both mother and foetus are at risk.

NAFFZIGER'S SYNDROME

Scalenus Anticus syndrome, Adson-Caffey syndrome

In this syndrome vascular and neurological symptoms are produced by pressure on the subclavian artery and brachial plexus passing through the scalene triangle. Scalene triangle is bounded anteriority by the scalenus anticus muscles, posteriorly by the scalenus medius and inferiorly by first rib.

In some people because of the excessive resting tone of the scalenus anticus muscle the first rib is slightly elevated producing compression on the subclavian artery and nerve roots especially C_8, T_1.

This is one of the causes for a symptom complex called thoracic outlet syndrome. Other causes are cervical rib, long transverse process of C_7, malunion of fracture of the first rib or clavicle, tumors or exostosis in the region, etc. Symptoms include:

1. Neurological symptoms like pain, paraesthesia and numbness usually in the region of ulnar distribution in the hand and fingers. The symptoms might later spread to the rest of the upper limb and may finally produce severe sensory motor loss and muscular atrophy.

2. Symptoms of arterial compression like claudication pain fatigue, coldness and weakness of upper limb. These symptoms are aggravated by exercise or exposure to cold.
3. There may be symptoms of embolization into end arteries of the fingers. Emboli are thrown by the thrombus that forms in the dilated part of the subclavian artery beyond the compression (post stenotic dilation). Patient might suddenly develop pain, pallor, paresthesia of the distal finger suggesting acute ischemia. Frank gangrene may develop later.
4. Venous compression occurs less often but can produce edema and cyanosis.

Diagnosis is by clinical tests like scalene maneuver (Adson's test) costoclavicular compression maneuver (exaggerated military posture) and three minutes elevated arm stress test, etc. Clinical examination will reveal signs of ischemia as well as neurological deficit. Conformation can be sought by X-ray, Doppler study, angiography, nerve conduction study, electro-myography, etc.

Management depends on the specific cause if one is detected. Otherwise thoracic outlet decompression through supra clavicular approach usually benefits more than 90% of the patients. It involves removal of the scalene muscles, resection of cervical rib if present and resecting first rib if thought to be the culprit.

NELSON'S SYNDROME

Syn: Postadrenalectomy Syndrome

This syndrome follows total adrenalectomy. Total adrenal ablation was being practised for Cushing's syndrome of non-adrenal origin. Not only was the surgery associated with significant mortality and morbidity, it was noticed that postoperatively there as pituitary hypertrophy, abnormally high ACTH levels and hyperpigmentation. The morbidity levels used to reach so high as to need pictuitary ablation. This clinical picture was referred to as Nelson's syndrome. It is rare nowadays.

OGILVIE'S SYNDROME

Idiopathic Megacolon

It is otherwise called acute intestinal pseudo obstruction.

It is etiology is not fully understood. It usually occurs in elderly individuals following trauma or pelvic surgery. Excessive pelvic sympathetic discharge is believed to be the cause.

Patient has acute distension of the abdomen associated with vomiting and pain. Usually patient has constipation but occasionally there may be diarrhea. X-ray shows massive dilatation of cecum and ascending colon without evidence of organic obstruction.

If the cecal diameter is < 10 cm then conservative gentle enema or flatus tube insertion might relieve the obstruction. Of course, electrolyte imbalance or other causes of paralytic ileus should be ruled out.

Patient should be monitored by serial X-rays and clinical evaluation. If the cecal diameter is measured to be > 12 cm then either endoscopic or operative intervention is needed as otherwise chances of cecal perforation are high. Endoscopic decompression needs experience and is potentially dangerous. Also there is a chance of recurrence of obstruction. Thus surgery may be required in case endoscopy is not available or is unsuccessful, if there are more than two recurrences and when there is endoscopic evidence of ischemia and necrosis. Patient might need colostomy or colonic resection depending on the operative findings.

ORMOND'S SYNDROME

Syn: Ormond's Disease

If refers to idiopathic retroperitoneal fibrosis. Some cases may have positive family history. In such cases other fibrosing conditions like sclerosing cholangitis, Reidel's thyroiditis etc may coexist.

Patient usually presents with oliguria. The cause of obstruction is dense periureteric fibrosis especially at or below the pelvic brim. Without treatment the obstruction progresses and may lead to renal failure.

Intravenous urogram shows both ureters obstructed and displaced towards the midline. CT scan is diagnostic.

Ureteric stenting or percutaneous nephrostomy relieves the obstruction temporarily. Though high dose steroids have been advocated by some, definitive treatment involves ureterolysis (i.e. releasing of fibrous adhesions) and omental wrapping around the ureters. The only other alternative is hemodialysis.

ORTNER'S SYNDROME

This is one of the unusual reasons for recurrent laryngeal nerve palsies. Here the recurrent laryngeal nerve gets paralyzed by the enlarged heart. The commonest cause for such cardiomyopathy is the left atrial enlargement seen in mitral stenosis.

It may be added here that pre-existing recurrent laryngeal nerve palsy should be sought for via indirect or direct laryngoscopy before posting any patient for thyroid surgery. It discovered post operatively, it is difficult for any surgeon not to get blamed for the palsy, however impeccable the surgery may have been.

POLAND'S SYNDROME

It refers to a rare condition where there is absence of one breast ipsilateral pectoralis major and sometimes

pectoralis minor associated with absent costal cartilages. In some cases there may be syndactyly.

In some patients the defect may only be restricted to muscle and breast. In them the anomaly becomes obvious only as the child grows up. In patients who have full blown syndrome with hypoplastic chest wall early reconstruction might become necessary.

PENDRED'S SYNDROME

Described by Vaughan Pendred in 1896. It refers to a dyshormonogenetic goiter associated with sensorineural deafness. The hormone deficient is peroxidase which is involved in the iodine binding of tyrosyl units within the thyroglobulin. Usually the deficit is moderate and the patient is euthyroid.

The condition is autosomal recessive and hence a family history should be sought.

If hypothyroid state exists, then hormone replacement may be needed. On the other hand, the sensorineural deafness may need further evaluation and expert management to ensure normal development of the infant.

PICKWICKIAN SYNDROME

This syndrome is named after the famed fictional character created by Charles Dickens.

It refers to hypoventilation and its related events occurring in morbidly obese individuals especially in lying down position.

Obesity leads to poor compliance of chest wall and increased intra-abdominal pressure. This in turn reduces the functional residual volume of the lungs. This state of affairs may result in ventilation perfusion mismatch especially in the lung bases. Patient may develop hypercapnia and hypoxemia. Many patients have obstructive sleep apnea. All these may ultimately result in polycythemia pulmonary hypertension and right heart failure. The term "Pickwickian" is especially applied when the patient is somnolent even during day time.

Needless to say weight loss forms the mainstay of treatment. Correction of sleep apnoea may be of benefit. Progesterone has been tried in the treatment in an effort to improve respiratory drive.

This clinical entity has been discussed here because morbid obesity is increasingly being recognized as a surgical problem.

PLUMMER-VINSON SYNDROME

Syn: Paterson Kelly Syndrome

It is one of the less common causes of dysphagia. Most of the patients are women and hence this condition has to be differentiated from globus hystericus wherein there is no mechanical obstruction in the oesophageal lumen.

Plummer Vinson syndrome is invariably found in patients suffering from iron deficiency anemia. There will be a mucosal web in the oesophagus in the post cricoid region, obstructing the passage of food bolus. Elsewhere also the oesophageal mucosa is unhealthy and friable. Patients present with dysphagia and choking sensation. Other features of sideropenic anemia like koilonychia, bald tongue, angular cheilosis etc may be present.

If diagnosed accurately, treatment is dilatation of the pin hole opening in the esophageal web. Great care is needed as the mucosa is friable and may bleed. Simultaneous treatment of iron deficiency anemia completely reverses the oesophageal changes. Untreated this condition may progress to malignancy. Hence Plummer Vinson syndrome is considered a precancerous state of the oesophagus.

PEUTZ-JEGHERS SYNDROME

This syndrome is often mistakenly considered as part of the familial adenomatous polyposis syndrome. Actually, it is an autosomal dominant condition causing the development of hamartomas and not adenomas through out the gastrointestinal tract. Small intestine is most commonly affected though stomach and colonic polyps are also well documented.

Patients become symptomatic early in life and may suffer from colicky pain, hemorrhage and anemia. There

may be circumoral and labial pigmentation. Unlike in familial adenomatous polyposis syndrome, the risk of malignant change in the polyps is significantly less but is present nevertheless.

If the symptoms are troublesome, then conservative resection might be needed. Per operative enteroscopy to detect and snare other lesions, if present will reduce the need for early reoperation for recurrent symptoms.

POSTCHOLECYSTECTOMY SYNDROME

Following cholecystectomy, though a large percentage of patients are relieved of their symptoms a significant number continue to have symptoms of dyspepsia and reflux disease. In about 5 percent of patients these symptoms are severe enough to seek medical attention. This symptom complex has been arbitrarily learned as post cholecystectomy syndrome.

In some of these patients common bile duct stones may be the cause for symptoms. These patients need endoscopic retrieval of stones. In a few others biliary dyskinesia and sphincter of Oddi dysfunction may be the underlying pathology. In these patients endoscopic sphincterotomy may be of benefit.

It is worth while to note that occasionally biliary tract disease coexists with irritable bowel syndrome or diverticulosis. In such cases colonic pathology might very well be the cause of residual symptoms. Hiatus hernia is another such coexisting condition.

Even after ruling out all possible organic causes for dyspepsia a definite subset of patients remains wherein there is no apparent cause. The only hope for these patients is counseling to divert their focus off the abdominal symptoms.

PRUNE BELLY SYNDROME

Syn: 1. Eagle Barret syndrome
2. Triad syndrome

In this condition there is a typical triad of abnormalities. They are deficiency of anterior abdominal wall (hence "prune belly") cryptorchidism and malformations of urinary tract. Surgical correction of the abdominal wall laxity is difficult. Many children die during infancy mostly due to uncontrolled urinary tract infection.

The exact etiology is unknown.

PARKER-WEBER SYNDROME

In this condition there are multiple arteriovenous malformations in the lower limb and sometimes in the adjoining pelvis. These congenital AV malformation may be localized to a small area or may involve the whole limb. There may be local gigantism. The AV fistula will cause hyper dynamic circulation and may progress eventually to cardiac failure. Branham's sign can be demonstrated in many cases (i.e. heart rate falls

when the feeding artery to the AV malformation is blocked.)

Skin pigmentation and ulceration are the other symptoms of this rare condition.

POSTCONCUSSION SYNDROME

This follows head injury resulting in cerebral concussion. If there is no focal lesion demonstrable on CT then these patients are supposed to have suffered diffuse axonal injury. Most patients recover fully, but some patients continue to have problems with memory, dizziness, nausea, hypersensitivity to stimuli and diminished concentration.

POSTSPLENECTOMY SYNDROME

Following splenectomy patients become vulnerable to bacterial infection, especially by capsulated organisms like *Pneumococcus, Meningococcus* and *H. influence.* Aggressive treatment is mandatory to prevent overwhelming postsplenectomy septicemia.

In elective splenectomy it is prudent to vaccinate the patients at least 2 weeks before surgery against capsulated organisms. In emergency splenectomies especially in children perioperative antibiotic cover is necessary till vaccine becomes effective. During surgery for traumatic rupture of spleen all attempts should be

made to salvage at least a part of the spleen (splenorrhaphy). Splenunculi, if found should be meticulously saved. Of course, splenunculi cannot be saved when splenectomy is done for hematological causes.

POSTPHLEBITIC SYNDROME

Syn: Post-thrombotic Syndrome

This term refers to the long-term complications of deep vein thrombosis (DVT) DVT can occur due to a variety of causes which impair mobility like serious illness, trauma, major surgery, pregnancy, etc. There may not be any symptoms in many patients.

Some may develop limb edema, pain and pyrexia. With time if life threatening complications like pulmonary embolism do not develop the clot is lysed and veins recanalize but the valves are damaged permanently resulting in deep venous incompetence. The resultant venous hypertension leads to edema, lipodermatosclerosis, eczematous lesions and finally venous ulcer. Treatment of this condition is mostly conservative. Valvuloplasty is the surgical option for these patients.

One has to remember that deep vein incompetence is not a must for venous ulceration. Pure superficial vein varicosities also cause the same picture and can be treated by simple ligation procedures.

PARANEOPLASTIC SYNDROME

This refers to a symptom complex which associated with a neoplasia but not caused directly by the tumor. The neoplasia cause the clinical syndrome mostly by the production of either a hormone or any other systemically active factor.

In some causes these paraneoplastic syndrome might not be of life threatening nature like polycythemia in renal cell carcinoma and acanthosis in gastric and uterine cancers. But some other neoplasia produce syndromes which add significantly to the morbidity and mortality. For example, syndrome of inappropriate secretion of ADH (i.e. SIADH) seen in small cell carcinoma of the lung. Small cell tumor of the lung can also produce a Cushing's syndrome like condition by secreting an ACTH like substance. Dermatomyositis in lung and breast cancers, carcinoid syndrome in bronchial adenoma, migrating thrombophlebitis in carcinoma pancreas are some of the other examples of paraneoplastic syndrome.

PIERRE-ROBIN SYNDROME

Consists of micrognathia, small tongue, cleft palate and eye defects. Eye defects may be in the form of congenital glaucoma, myopia or retinal detachment. The smallness of the mandible and the resultant posterior

displacement of the chin will result in tongue falling back and obstructing the airway. In its severe form the airway obstruction may be life threatening for the newborn child. The infant might also have choking episodes during feeding most of these unfortunate children need early surgical correction

The exact etiology is not known.

PSEUDO-ZOLLINGER-ELLISON SYNDROME

This is seen sometimes in multiple endocrine neoplasia (MEN) type I due to exaggerated gastrin response to hypercalcemia due to hyperparathyroidism. Though the pattern of ulceration and symptomatology resembles that of true Zollinger-Ellison syndrome, this condition is termed 'pseudo' because it disappears as soon as parathyroid hyperfunction is dealt with.

Rendu-Osler-Weber Syndrome

In this hereditary syndrome there are multiple telangiectasias of gastrointestinal tract, nose, skin, liver, CNS, etc. Epistaxis is the most common symptom but gastrointestinal bleed can be life threatening. Since the telangiectasias are small they are not detectable by imaging. Hence, angiography or endoscopy is the mainstay of diagnosis. A positive family history is often suggestive. Photocoagulation or electrocoagulation of

these lesions has been tried with mixed results. Treatment is difficult especially when there are multiple lesions. Massive GI bleed may necessitate major resection.

ROTOR'S SYNDROME

It is a type of congenital nonhemolytic jaundice in which there is conjugated bilirubinemia and bilirubinuria.

Liver is normal histologically and usually the jaundice is mild. The underlying pathology is said to be poor storage of bilirubin in the liver cells.

Prognosis is excellent compared to other types of conjugated hyperbilirubinemia syndromes.

RAPUNZEL SYNDROME

This term is sometimes used to describe the presence of extensive trichobezoar occupying the stomach, duodenum and down into the jejunum.

Patient is usually mentally challenged and will have alopecia, halitosis along with an epigastric mass. History of eating ones own hair may be available if elicited carefully. Patient might have features of intestinal obstruction. Gastrointestinal bleeding and perforation are other serious complications.

If diagnosed in time surgery relieves the symptoms completely.

RAYNAUD'S SYNDROME

This refers to the sequence of local pallor, cyanosis and erythema that occurs when a limb with peripheral vascular disease is exposed to cold.

It was earlier classified as primary or Raynaud's disease which was idiopathic, and secondary or Raynaud's phenomenon which occurred in association with other diseases like scleroderma, cervical rib, SLE, carpal tunnel syndrome, etc. This distinction is not very clear. Hence has been given up and the term Raynaud's syndrome is used for all cases.

This condition is due to the abnormal sensitivity of the arterioles to cold. When exposed to cold these arterioles constrict and the part becomes blanched or pale (local syncope). The hypoxia thus induced initially results in capillary dilatation. These dilated capillaries get filled with deoxygenated blood leading to cyanosis (local asphyxia). Finally, the arterioles themselves dilate and fresh oxygenated blood again flows leading to erythema and engorgement.

The condition may slowly to progress to ischemic ulceration and local gangrene.

SEZARY SYNDROME

It is a distinct variety of mycosis fungoides or cutaneous T-cell lymphoma.

It presents with erythroderma, pruritus and lymphadenopathy. Peripheral blood shows specific cells call Sezary cells.

Clinical course is protracted over years. Lymphadenopathy is an indicator of poor prognosis.

If the disease is limited to the skin, then total skin electron beam irradiation and topical chemotherapy may be tried. PUVA has also been tried.

SIADH (Syndrome of Inappropriate Secretion of ADH)

This is seen in some cancers (like small cell cancer of the lung) burns and head injury. There is excess secretion of ADH hormone leading to concentrated urine, expanded extracellular fluid volume and dilutional hyponatremia. Patient may suffer from lethargy and dizziness. If correctly diagnosed, fluid restriction is all that is necessary. Use of loop diuretics like furosemide and isotonic saline may be needed occasionally.

SIPPLE SYNDROME

Syn: Multiple Endocrine Neoplasia type II

In this familial condition, there is association of medullary carcinoma of thyroid, phaeochromocytoma and parathyroid adenomas.

Sipple syndrome or MEN type II is again subdivided into type II a and II b. In MEN type II b in addition to the neoplasms mentioned above patients will have multiple mucosal neuromas especially of the lips and the tongue (bumpy lips) diffuse ganglioneuromas of the gastrointestinal tract, skeletal abnormalities and marfanoid features. Sometimes MEN type II b is also referred to as "MEN type III".

The clinical significance of Sipple syndrome lies in the fact that medullary carcinoma thyroid that occurs is more aggressive than its counterpart in MEN type II a. Unless the diagnosis is made early and total thyroidectomy done, the disease rapidly becomes fatal. But one has to be conscious of the possibility of coexisting phaeochromocytoma in order to avoid catastrophe on operating table.

Phaeochromocytoma in MEN II is always benign and restricted to adrenal medulla. It may or may not be symptomatic.

Parathyroid adenomas are much rarer in MEN type II b than II a.

Sipple syndrome has a autosomal dominant inheritance.

STEIN-LEVENTHAL SYNDROME

Polycystic Ovary Syndrome

Typical triad of Stein-Leventhal syndrome is large polycystic ovaries, secondary amenorrhea and

infertility. Other features commonly observed are obesity, hirsutism and dysfunctional uterine bleeding.

The exact etiology is not known. It is thought there is excessive androgen production from the enlarged ovaries.

The surgical importance lies in the presence of a pelvic mass. Awareness of this syndrome makes recognition of its typical features easy. Of course the management of the condition has to be left for gynecologists.

STURGE-WEBER SYNDROME

Syn: Sturge-Klischer-Weber Syndrome

It consists of a triad of angiomas namely congenital cutaneous angioma in the region of trigeminal nerve, ipsilateral meningeal angioma and angioma of the choroids.

While the cutaneous angioma (flame nevus) is obvious at sight, the presence of other two angiomas can be suspected by the focal neurological signs and secondary glaucoma that they produce. Surgeon's role is restricted to recognizing the syndrome thereby paving the way for specific management by experts in the field. Hence, details have not been discussed.

SHEEHAN'S SYNDROME

It is hypopituitarism resulting from pituitary infarction following severe postpartum hemorrhage. Patient may

later present with features of hypothyroidism, loss of libido, features of adrenal insufficiency, hypoglycemia due to increased sensitivity to insulin, etc.

If not properly treated patients rapidly progress to coma. Treatment involves replacement of deficient hormones.

STEWART-TREVES SYNDROME

In its classic form it refers to the lymphangiosarcoma occurring in a lymphoedematous upper limb following radical mastectomy. Some people include other causes of lymphedema also in the above definition, e.g. postradiation lymphedema.

The possibility of this long-term complication should be borne in mind and the patient asked to report any ulceration or local pathology especially when the lymphedema is significant. Lymphangiosarcoma is a very aggressive tumor and may need a major amputation.

SJÖGREN'S SYNDROME

It is an autoimmune disorder characterized by dry eyes (kerato-conjunctivitis sicca) and dry month (xerostomia). It may be primary, otherwise called "sicca syndrome" wherein the clinical features are severe but there are no other associated connective tissue disorder. On the other hand, secondary Sjögren's syndrome refers

to that which is associated with other connective tissue disorders like rheumatoid arthritis, systemic lupus erythematosus, myasthenia gravis, primary biliary cirrhosis.

Investigation usually involves assessing the level of damage to parotid and lacrimal glands. Schirmer's test for lacrimation, other tests like sialography, FNAC of the gland, technetium scan, etc. are also occasionally necessary to clarify the diagnosis.

Autoantibody estimation might point to any associated connective tissue disorder like rheumatoid arthritis which needs specific therapy.

The treatment of this syndrome mainly symptomatic and basically involves moistening the mucous membranes with artificial tears, saliva, etc. Occasionally transplantation of parotid duct to lacrimal sulcus may be necessary to save vision when severe keratitis endangers it.

SCHEUERMANN'S SYNDROME

Syn: Sarcoidosis

It is a multisystem granulomatous disorder producing lesions similar to that of tuberculosis.

It can run subacute and self limiting course or may be chronic with multiple extrapulmonary manifestations including parotid involvement, erythema nodosum, polyarthralgia, hepatosplenomegaly nephrocalcinosis,

etc. The last named manifestation occurs due to disturbed calcium metabolism leading to hypercalcemia.

For definitive diagnosis either a lymph node or a skin lesion has to be biopsied. Transbronchial lung biopsy is always confirmatory. The granulomas are noncaseating unlike in tuberculosis. Kveim intradermal test using antigen from human sarcoid tissue is suggestive.

Though many subacute cases resolve spontaneously, chronic and more severe forms need prolonged corticosteroid treatment to suppress the manifestations.

Involvement of parotid gland by sarcoidosis with associated facial palsy, fever and anterior uveitis is some times referred to as "Heerfordt's syndrome".

STEVENS-JOHNSON SYNDROME

Syn: Erythema Multiforme Bullosum

A severe form of bullous erythema multiforme involving large parts of the body surface and the mucous membranes. It is a potentially life threatening rare complication of sulphonamides. Needless to say the drug has to be stopped immediately.

Occasionally, Stevens-Johnson syndrome is associated with ocular lesions like conjunctivitis, iritis, etc. It is then referred to as "ocular-mucous membrane syndrome".

SANDIFER'S SYNDROME

It is a peculiar phenomenon where a patients of gastroesophageal reflux disease has spasmodic contortions of the head. Fortunately, it disappears with the elimination of reflux either medically or by surgical intervention.

SUMP SYNDROME

This is a syndrome that develops many years following a choledochoduodenostomy. Patient develops clinical features of recurrent cholangitis like fever, pain in the right hypochondrium, etc.

The reason for this syndrome is the collection of biliary sludge and vegetable matter in the retroduodenal part of the bile duct. Partial stenosis of the anastomotic site might be a contributory factor in the causation.

Treatment by way of endoscopic sphincterotomy and/or balloon dilatation of the anastomosis has been tried, but the results are not always satisfactory.

SHORT-BOWEL SYNDROME

As the name suggests this clinical condition results from resection of too much small bowel.

Such massive bowel resection might be necessary in superior mesenteric artery infarction or superior mesenteric venous thrombosis. Other causes include necrotizing enterocolitis, intestinal agenesis in neonates,

radiation enteritis, inflammatory bowel disease, massive trauma, etc.

Characteristic features of this syndrome reflect the altered physiology of the GI tract. Patient will have diarrhea, steatorrhea weight loss, nutritional deficiency, etc. due to short and inadequate bowel length for adequate digestion or assimilation.

Treatment involves use of antidiarrheal drugs and H_2 receptor antagonists. H_2 receptor blockers reverse the hypergastrinemia that is seen following short bowel syndrome. Antibiotics may occasionally be needed if gut sepsis is diagnosed but may worsen diarrhea.

Surgical treatment involves interposition of a reversed (anti-persistatic) intestinal segment or intestinal lengthening. The drawbacks of interposition surgery are the technical difficulty in a patients with short bowel. On the other hand intestinal lengthening operation (Bianchi operation) involves sectioning the available bowel longitudinally using stapler and then anastomosing them end to end. This relies on the fact that the two sides of the bowel are independently supplied by mesenteric vessels.

Intestinal allo-transplantation offers hope but is as yet in experimental stage only.

SEAT-BELT SYNDROME

This occurs in road traffic accident due to an improperly worn seat-belt. The massive deceleration force will

result in the seat-belt impinging on the abdominal wall. This can lead to avulsion of rectus abdominis from the pubic bone. Needless to say such an injury will result in trauma to the soft viscera within the abdominal cavity.

Another feature of this syndrome is the fracture of the posterior process and/or body of the lumbar vertebrae. This results from hyper flexion of the spine.

The contusion caused by improperly applied seat-belt may be seen across the abdomen (seat-belt sign).

Treatment is as for any other blunt abdominal trauma with willingness to do laparotomy at the slightest doubt of visceral trauma.

TURCOT'S SYNDROME

It is a part of the familial adenomatous polyposis syndrome. In Turcot's syndrome gastrointestinal polyps are associated with brain tumors like gliomas or medulloblastomas. It is inherited as autosomal dominant and is definitely a premalignant state as far as the GI tract is concerned. By the second decade of life there are multiple polyps mostly in the colon but also in the stomach and small bowel. The higher the number the greater is the risk of malignancy.

Patient may have loose stools, bleeding per rectum or features of overtmalignancy like weight loss, intestinal obstruction, etc.

Family history might suggest the diagnosis. Colonoscopy will confirm the diagnosis.

Total proctocolectomy with ileoanal anastoneosis is the only way to prevent an almost certain malignant transformation of one of the polyps. If conservative resection sparing the rectum has been done then regular follow-up for endoscopic screening for rectal polyps is advisable.

Sulindac has been used in the treatment of this condition. This NSAID is apparently successful in suppressing these polyps.

TUMOR LYSIS SYNDROME

This sometimes occurs following chemotherapy. Lysis of tumor cells releases large quantities of intracellular substances like uric acid, potassium phosphate, etc. into circulation. Hyperkalemia may induce life threatening cardiac arrhythmias. Hyperkalemia is worsened by renal failure caused by hyperphosphatemia. Increase in phosphate also causes hypocalcemia leading to cramps, cardiac arrhythmia and tetany.

This syndrome is more likely to occur is large tumors highly sensitive to chemotherapy, e.g. lymphomas.

Anticipating this syndrome in the appropriate setting and taking precautionary measures is very important. Acid-base balance, hydration, electrolytes, uric acid should all be montored and if necessary corrected before initiation of chemotherapy. Once manifest treatment is as for individual abnormality. Some patients might need hemodialysis.

TORRE SYNDROME

Syn: Muir-Torres Syndrome

This is another syndrome associated with visceral tumor especially small bowel neoplasm.

Patient might have sebaceous cysts, sebaceous adenomas, fibromas, lipomas, etc. together with visceral malignancy. Occasionally, the patient might have double malignancy, e.g. fibrosarcoma and small bowel cancer.

The clinical significance of this syndrome lies in the fact that awareness of its existence might lead to an earlier diagnosis of the visceral tumor in an occasional patient even when the symptoms are not suggestive. Of course it needs a very high index of suspicion on the part of the clinician.

TIETZE'S SYNDROME

It is a condition of unknown etiology wherein there is painful non-supperative inflammation of costochondral cartilages. It is sometimes referred to as peristernal perichondritis. Local tenderness may be the only symptom. There is no gender predilection.

The disease usually self-limiting and hence needs only symptomatic treatment. Occasionally steroids, either local or systemic may be needed. If the symptoms continue then excision of the involved cartilage may be needed.

TAKAYASU'S SYNDROME

Syn: Takayasu's Disease; Pulseless Arteritis

This is an arteritis of probably immunological origin that affects the branches of aortic arch, most commonly the subclavian artery. Most of the patients are young women.

Patient presents with claudication pain in the upper limb with absent pulses. Constitutional symptoms like fever, vomiting myalgia, arthralgia may be present. If the cerebral circulation has been involved then symptoms of cerebrovascular insufficiency appear. Ischemic gangrene of the upper extremity and myocardial infarction are lurking dangers in this condition.

Treatment involves immunosuppressive drugs like prednisolone or cyclophosphamide. Direct arterial surgery has proven disappointing as reocclusion is very common.

VERNER-MORRISON SYNDROME

Syn: WDHA Syndrome

- Watery diarrhea hypokalemia, achlorhydria
- Pancreatic cholera syndrome
- Vipoma syndrome

Vasoactive intestinal peptide secreted by vipoma, an islet cell tumor produces severe diarrhea which is resistant to

conventional treatment. This in turn leads to hypokalemia, hypochlorhydria and hypovolemia. Patients have abdominal cramps and weakness. Occasionally, there may be hypercalcemia. Diagnosis can be confirmed if fasting plasma VIP level is more than 500 mg /ml.

Definitive treatment is surgical removal of the tumor. But electrolyte and acid-base balance has to be restored before surgery. Octreotide has revolutionized the presurgical management. Type of surgery will depend upon the exact nature of the tumor and its location within the pancreas.

WILKIE'S SYNDROME

Syn: Superior Mesenteric Artery (SMA) Syndrome

This refers to the compression of distal part of the duodenum between superior mesenteric artery anteriorly and the aorta and vertebral body posteriorly. Sudden weight loss and immobilization in bed have been cited as triggering factors. Abnormal attachment of ligament of Trietz is another contributing factor. It is rare in obese patients.

It is onset is insidious and patient presents with vomiting. Contrast study will reveal dilated stomach and duodenum proximal to the crossing of superior mesenteries artery.

If conservative measures fail then duodeno-jejunostomy is the surgical procedure of choice for relieving obstruction. Simple gastrojejunostomy may fail to relieve duodenal obstruction adequately.

WERMER'S SYNDROME

Syn: MEN – Type I

It is the less commonly known name for multiple endocrine neoplasia type I. The commonest feature of this autosomal dominant condition is hyperpara-thyroidism. Other features are pancreatic islet cell tumors and pituitary adenomas. Rarely adrenal tumor are also seen.

The pathology in parathyroid glands is usually a hyperplasia and not adenoma whereas elsewhere it is adenoma. Pancreatic adenoma may occasionally produce excess gastrin leading to Zollinger-Ellison syndrome.

Treatment is as for the individual tumor depending on site histopathology and clinical features.

WEAK VEIN SYNDROME

This is one of the causes for gross primary varicosities of the lower limb venous system, especially of the superficial system. The wall of the veins is congenitally

defective and lacking in tone. Even the valves within the vein are weak and likely to be incompetent. Patient may not have overt complications of these varicosities as long as the deep system is efficiently functioning.

WALTMAN WALTER SYNDROME

This is a shock like state that can rarely occur in the early postoperative period following CBD exploration or even simple cholecystectomy. After about 2-3 days after the procedure patient suddenly complains of severe epigastric and right hypochondrial pain and may have circulatory collapse. Sometimes, the features may mimic myocardial infarction.

The etiopathogenesis of this syndrome involves bile leak either due to ligature slipping from cystic duct stump or due to malfunctioning T-tube. The leaked bile first gets collected in the Morison's pouch and then in right subphrenic space and paracolic gulter. The liver gets pushed downward and to the left thus impinging on the inferior vena cava. The reduced venous return to the heart may cause circulatory collapse and angina like picture.

Diagnosis should be suspected on clinical grounds alone as it is an acute emergency. Only if patient is relatively stable, ultrasound can be done to confirm the diagnosis. One should not unnecessarily delay re-exploring the patient. T-tube should be repositioned and

cystic duct stump checked after drainage and wash. Abdomen should be closed after keeping a drain in the Morison's pouch.

VON HIPPEL-LINDAU SYNDROME

It is an autosomal dominant condition. The genetic defect found in this condition is known to suppress expression of endothelial growth factor. Hence, the main characteristic of this syndrome is vascular tumors of the central nervous system especially cerebellum and retina. Adenocarcenoma of the kidney is another commonly associated condition whose incidence increases with age. Phaeochromocytoma is the other neoplasm sometimes seen in this condition.

ZOLLINGER-ELLISON SYNDROME

Syn: Gastrinoma Syndrome

This syndrome was described in 1955 in association with pancreatic islet cell tumor secreting gastrin. Many of these patients (up to 20%) will have multiple endocrine neoplasia type I.

The characteristic triad of symptoms of this syndrome is jejunal ulceration, hypersecretion of gastrin and pancreatic islet cell tumor. This condition is more common is males and occurs in middle aged people. ZE syndrome associated with MEN type I occurs in

the young. Patients have multiple peptic ulcers even at unusual sites like jejunum. Gastroesophageal reflux disease and secretory diarrhea are seen in some. Peptic ulcer perforation is a life threatening complication of this syndrome.

Diagnosis is by measuring serum gastrin level (> 100 pg/ml). But one should stop all antiacid secretory drug at least 1 week before estimation of gastrin levels. Acid secretion studies also may give a clue to the diagnosis.

Treatment involves excision of the offending neoplasm. But one should remember that at least half of these tumors may be malignant and many a time they are on the duodenal wall. If there are no demonstrable metastasis then exploration is done and tumor localized with the help of intraoperative ultrasound. However, the results of surgery are not always good. But at least it offers the hope of complete cure if a single benign tumor is the cause of the syndrome. Alternatively, proton pump inhibitor may be able to control symptoms of peptic ulceration adequately. Occasional patient might need hear total gastrectomy for symptom relief.

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