

An Illustrated Atlas of All India PG Entrance Examination 2010

2010

# All India Express



**Dr Saraswata Das**

MD Radiodiagnosis (Std)

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# PREFACE

It is with great joy that I bring out the EBook of All India PG Entrance Examination 2010. I think it will prove useful in giving a cross-sectional view of all the questions asked in this exam, and that too with a minimum acquisition time. As it has been named "Express", its purpose is to enable the readers to get the maximum information in minimum time, which I think will be extremely valuable during the last few days before the exam. Explanations of repeat/easy questions have not been included for obvious reasons. State-of-the-art illustrations have been added wherever necessary for better understanding and retention of the concepts as well as to make the reading pleasurable.

As always, do send your comments, criticisms, suggestions & feedback.

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**Jiyo Utho Badho Jeeto**

**All The Best!!!**

Question Recall Acknowledgement : PPM Unique Question Recaller

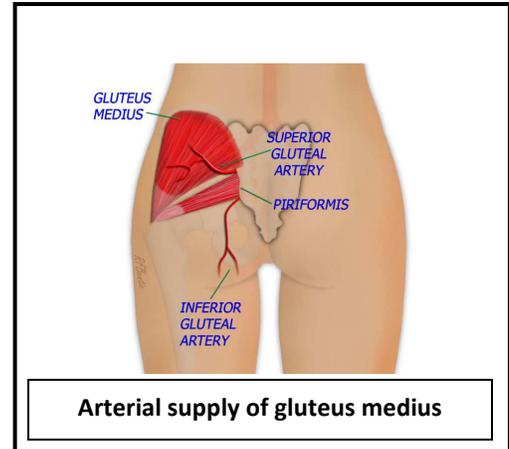
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# ANATOMY

1. Gluteus medius is supplied by :

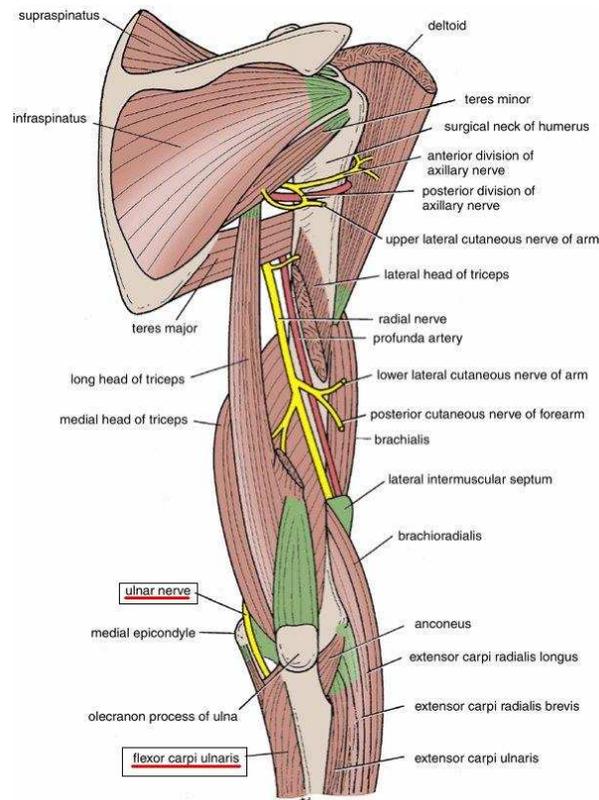
- Superior gluteal artery
- Inferior gluteal artery
- Obturator artery
- Ilioinguinal artery

The main supply to gluteus medius is from the deep branch of the superior gluteal artery.



2. All are composite muscle except :

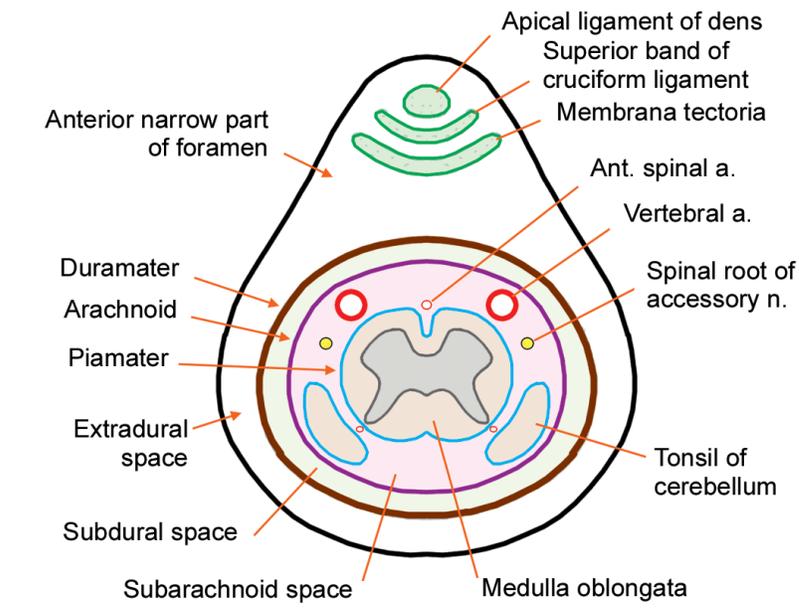
- Flexor carpi ulnaris
- Flexor digitorum profundus
- Pectineus
- Biceps femoris



Flexor carpi ulnaris is supplied by **ulnar nerve only**. All the other muscles mentioned have dual nerve supply and hence are composite muscles.

3. Which of the following passes through foramen magnum :
- Vertebral artery**
  - 12<sup>th</sup> nerve
  - Sympathetic chain
  - Internal carotid artery

**Scheme to show the structures passing through foramen magnum**



**Vertebral arteries**

At the **C1 level** the **vertebral arteries** travel across the posterior arch of the atlas before entering the foramen magnum.

Inside the skull, the two vertebral arteries join up to form the **basilar artery** at the base of the medulla oblongata. The basilar artery is the main blood supply to the brainstem and connects to the Circle of Willis to potentially supply the rest of the brain if there is compromise to one of the carotids. At each cervical level, the vertebral artery sends branches to the surrounding musculature via anterior spinal arteries.

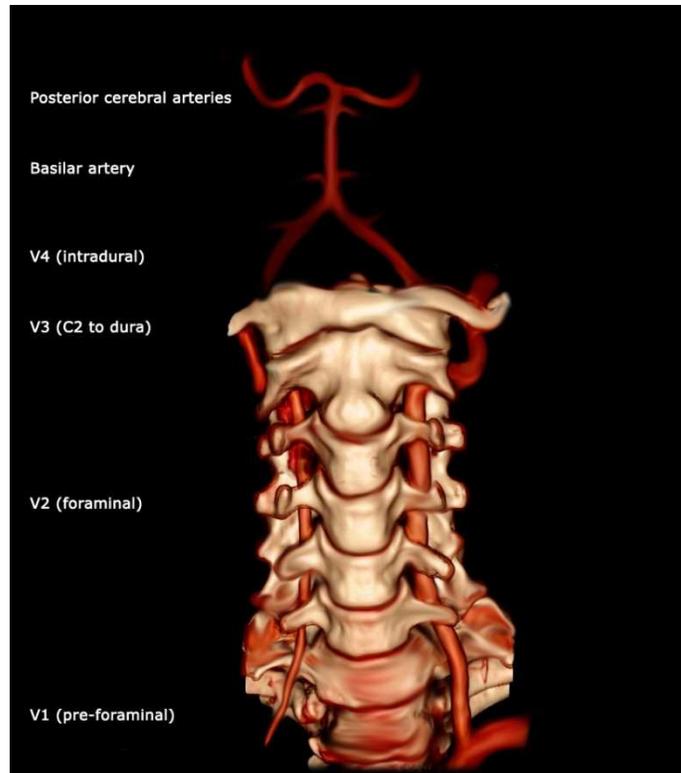
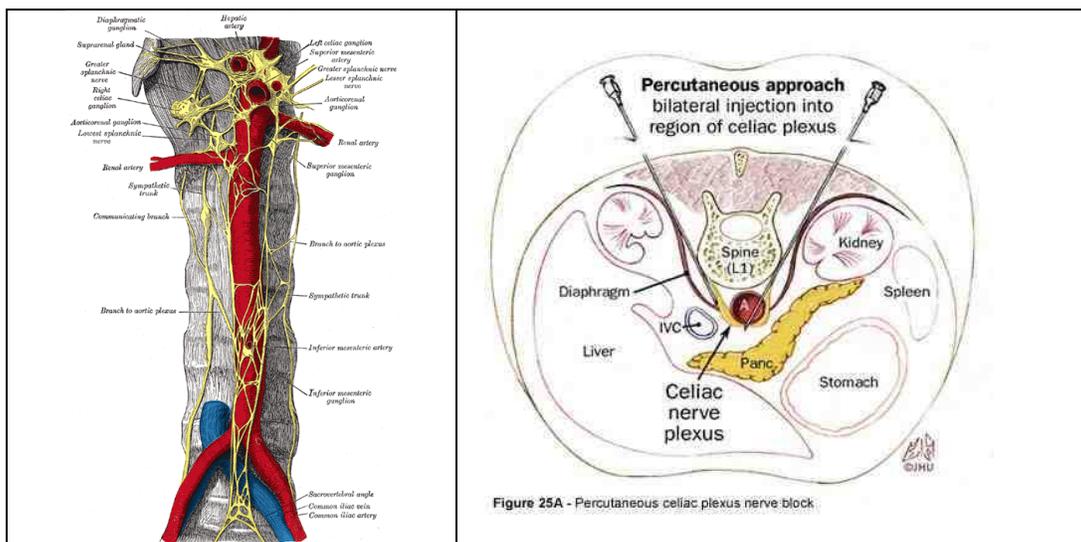


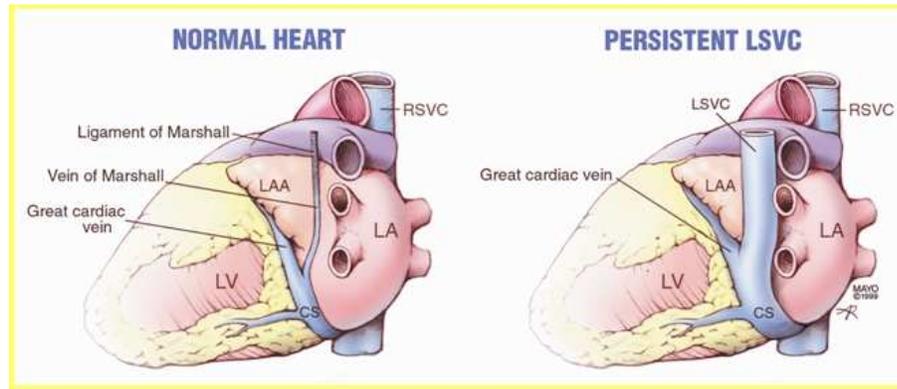
Figure showing parts of vertebral artery & its course

4. Celiac plexus is located at :
- Anterolateral to aorta
  - Posterolateral to aorta
  - Medial to sympathetic chain
  - Lateral to sympathetic chain



The celiac plexus is located **anterior to the aorta** at the level of **L1 vertebra**.

5. Left sided SVC drains into :
- Right atrium
  - Left atrium
  - Coronary sinus**
  - Pericardial space



#### Classification of Persistent Left Superior Vena Cava

A. 90% of the cases:

- Persistent left superior vena cava connecting to the right atrium via coronary sinus.

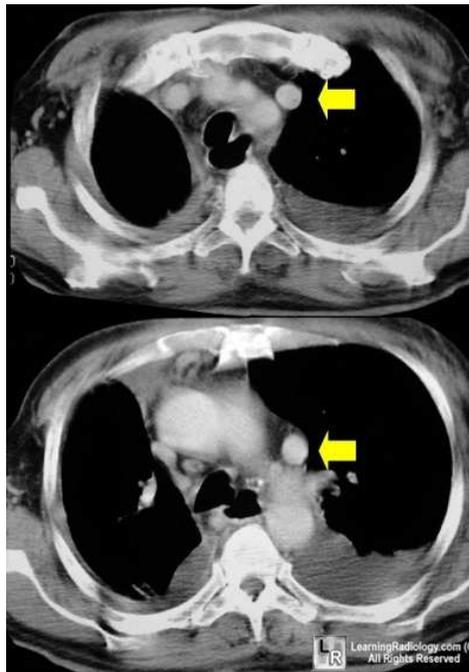
B. 10% of the cases:

- Persistent superior vena cava that connects to left atrium :
  - Often associated with:
    - Atrial septal defect.
    - Heterotaxy syndromes.

## Persistent Left Superior Vena Cava: Quick Review

- Incidence-uncommon
  - 0.3% of general population;
  - 4.3-11% of patients with CHD
- Two types
  - Persistent left SVC connecting to right atrium **via coronary sinus** is only common anomaly of SVC (90% of this anomaly)
  - In other 10%, persistent SVC connects to left atrium
    - Most with connection to left atrium have associated ASD or heterotaxy syndromes

- This produces a right-to-left shunt of a rather small magnitude
- Etiology
  - Failure of regression of left anterior and common cardinal veins and left sinus horn
- Course of persistent left SVC
  - Draining into right atrium
    - Starts at junction of left subclavian vein and left internal jugular
    - Passes lateral to aortic arch
    - Receives left superior intercostal vein
    - Anterior to left hilum
    - Joined by hemiazygous system
    - Crosses posterior wall of left atrium
    - Receives great cardiac vein to become **coronary sinus** (common)



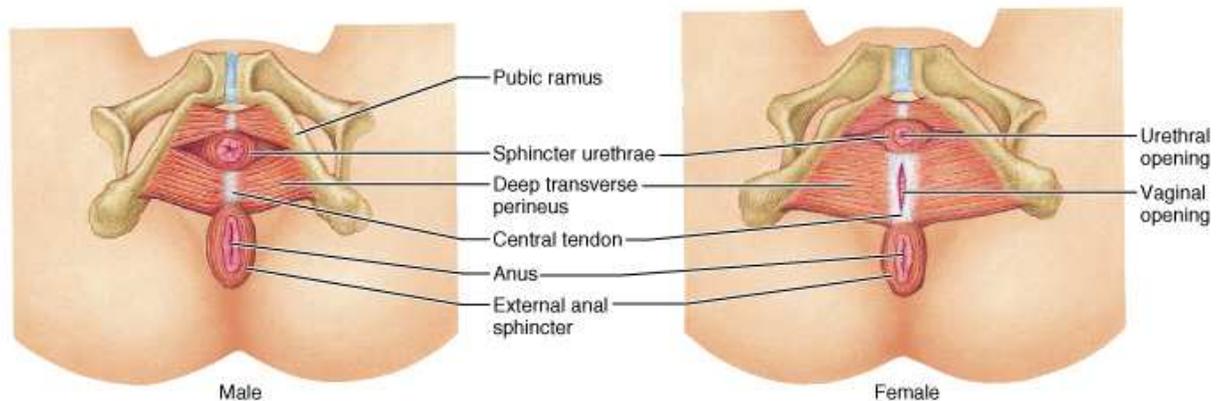
*Yellow arrows point to left-sided persistent SVC passing lateral to aortic arch and anterior to left hilum*

- Draining into left atrium
  - Starts at junction of left subclavian vein and left internal jugular
  - Passes lateral to aortic arch
  - Receives left superior intercostal vein
  - Anterior to left hilum
  - Joined by hemiazygous system
  - Passes between the left atrial appendage (anteriorly) and the left superior pulmonary vein posteriorly
- Absent / small left brachiocephalic vein (65%)
- Really this abnormality produces bilateral SVCs
- In small percentage, right SVC is absent (10-18%)

6. Hypogastric sheath is a condensation of :
- Scarpa's fascia
  - Colles' fascia
  - Pelvic fascia**
  - Inferior layer of urogenital diaphragm

“The subperitoneal endopelvic fascia consists of a connective tissue matrix around the pelvic organs. This fascia is continuous with both visceral & parietal layers of **pelvic fascia**. The condensation of this fascia forms the **hypogastric sheath** containing the vessels to the pelvic viscera from the lateral wall, the ureters & in male the vas deferens.” – *Grant's Atlas of Anatomy, 10e/205*

7. Urogenital diaphragm is formed by all except :
- Colles' fascia**
  - Deep transverse perinei
  - Perineal membrane
  - Sphincter urethrae



(b)

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**Urogenital diaphragm is formed by:**

- Deep perineal muscles (**Sphincter urethrae & Deep Transverse Perinei**)
- Superior fascia of urogenital diaphragm
- Inferior fascia of urogenital diaphragm (**Perineal membrane**)

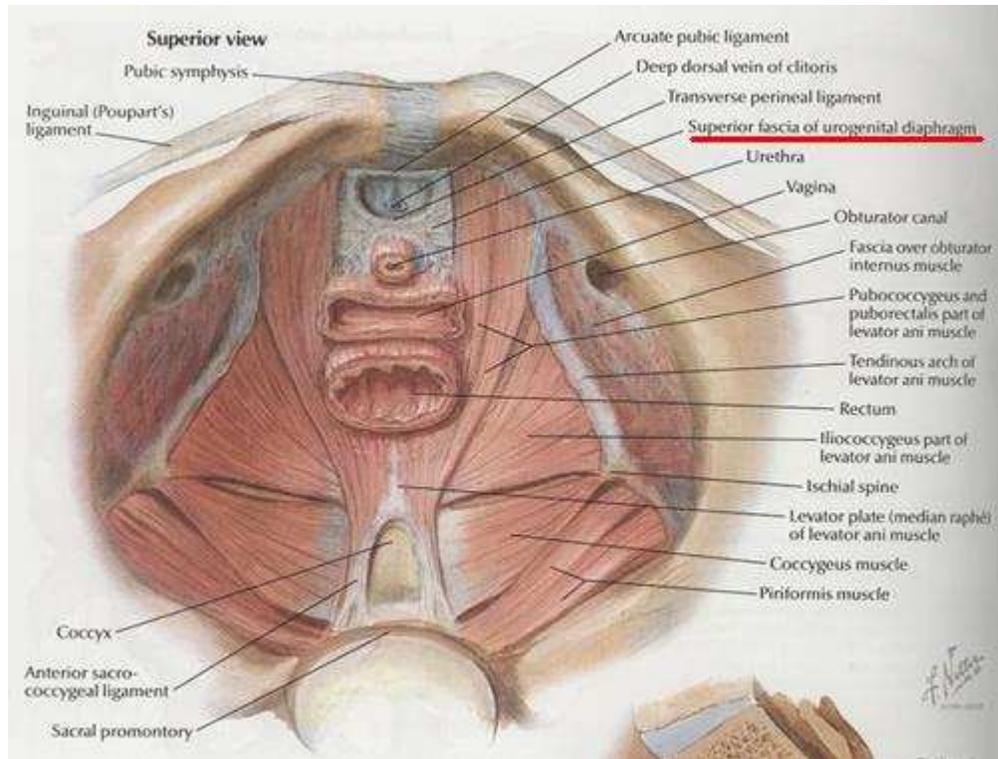


Figure showing the superior fascia of urogenital diaphragm

8. Most common renal anomaly seen is :
- Supernumerary arteries
  - Supernumerary veins
  - Double renal arteries
  - Double renal veins

The most common renal vascular anomaly is **multiple renal arteries** (17% - 34%). For details, see the journal extract given below.

## Images in Endourology

### Anomalous Renal Vasculature: A Laparoscopic Perspective

Arni Rice, M.D., Amanjot S. Sethi, M.D., and Chandru P. Sundaram, M.D.

A MINIMALLY INVASIVE APPROACH TO EXTERPATIVE and reconstructive renal surgery warrants a thorough understanding of renal vascular anatomy and its aberrations. Laparoscopy affords little room for error when employed for the management of renal masses, kidney donation, and ureteropelvic junction obstruction (UPJO). Hence, laparoscopic renal surgeons cannot forgo the meticulous dissection and identification of all renal vessels including their potential anomalies.

The reported prevalence of renal vascular anomalies ranges from 25% to 40%.<sup>1,2</sup> The most common renal vessel aberration is multiple renal arteries (17%–34%).<sup>1,2</sup> Historically, supernumerary renal arteries are found more often on

the left.<sup>1</sup> Anomalous arteries can travel with the hilar vessels or alternatively feed directly into the renal parenchyma. Right-sided lower pole arteries commonly cross anterior to the inferior vena cava (Fig. 1). Accessory polar arteries can cross anterior to the UPJ on either side and contribute to obstruction. Ectopic kidneys tend to exhibit a higher prevalence of vascular irregularity.<sup>1</sup> The renal arteries supplying ectopic moieties can originate from various branches of the aorta including the celiac trunk, superior mesenteric artery, iliac arteries as well as from different points along the aorta itself.

The overall reported prevalence of supernumerary renal veins (11%–20%) is lower than that of multiple renal arter-

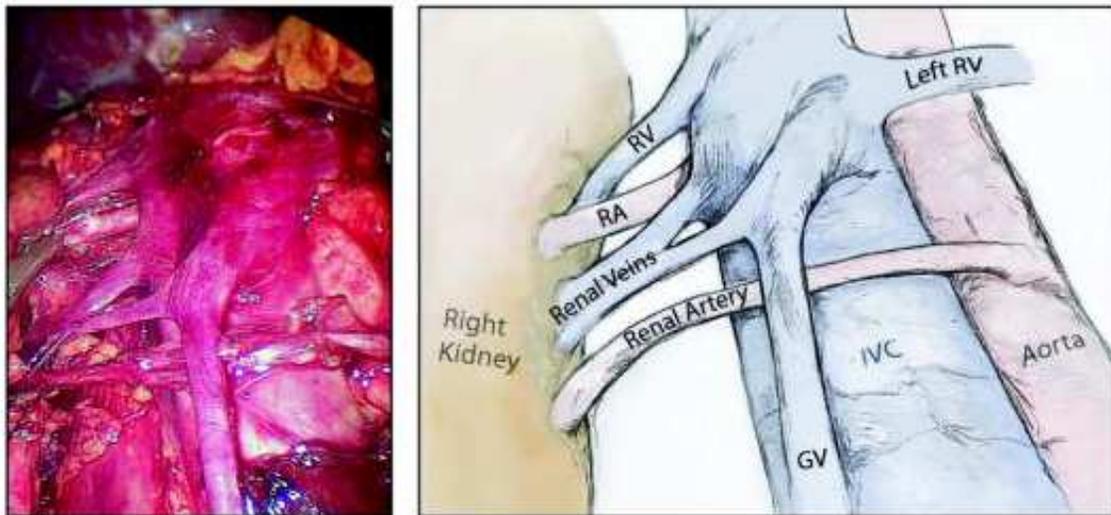


FIG. 1. Multiple right renal vessels with a lower pole renal artery.

Department of Urology, Indiana University School of Medicine, Indianapolis, Indiana.

9. Lymphatics from the spongy urethra drain into the following lymph nodes:
- Superficial inguinal lymph nodes
  - Deep inguinal lymph nodes
  - External iliac lymph nodes
  - Para-aortic lymph nodes

“Lymphatics from spongy part of urethra pass mostly to the **deep inguinal lymph nodes.**” – *BDC,3e/309 (Vol.2)*

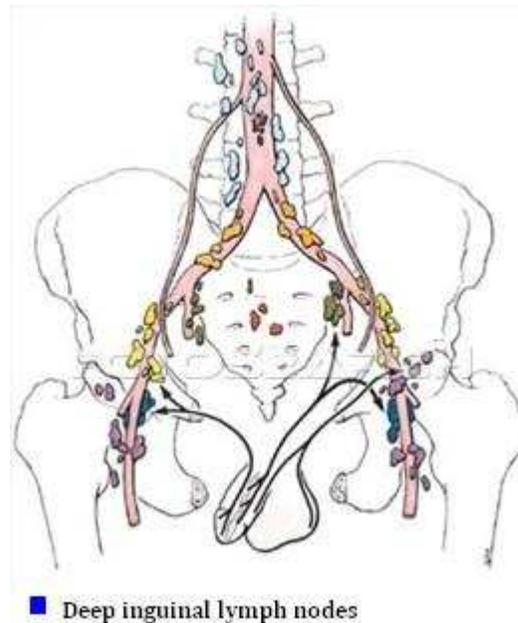
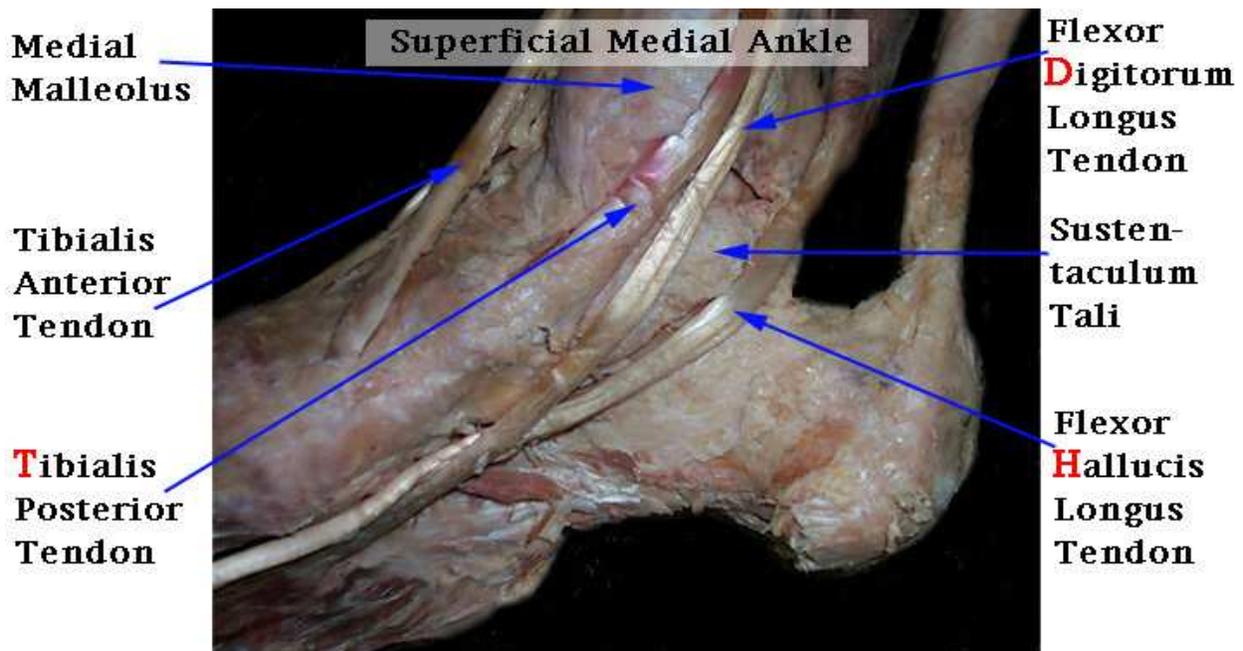
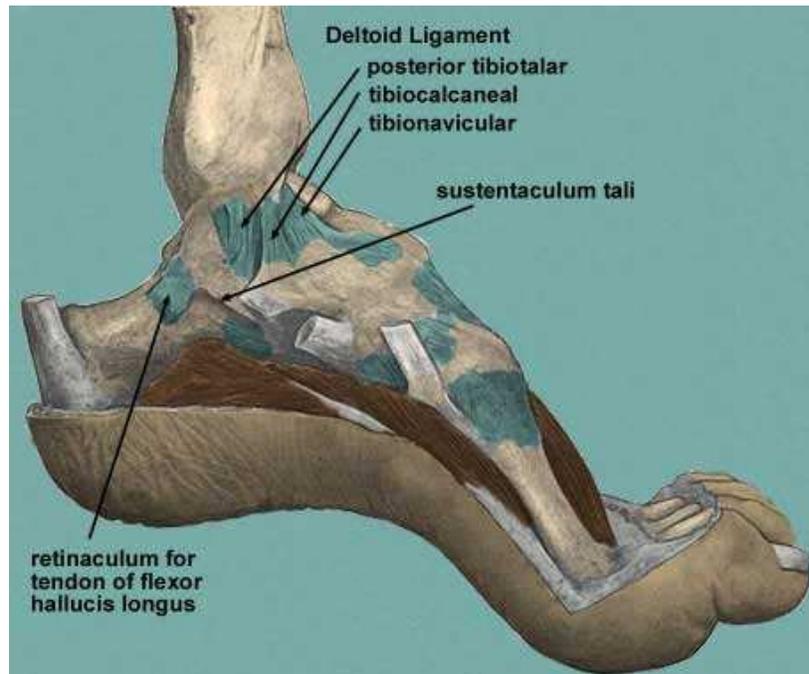


Figure showing lymphatic drainage of male pelvis

10. Attachment of sustentaculum Tali, is closely related to :
- Tibialis anterior
  - Tibialis posterior
  - Flexor hallucis longus
  - Flexor digitorum longus

At the upper and forepart of the medial surface of the calcaneus is a horizontal eminence, the **sustentaculum tali** (alternatively, the talar shelf), which gives attachment to the plantar calcaneo-navicular (spring) ligament, tibiocalcaneal ligament, and medial talocalcaneal ligament.

This eminence is concave above, and articulates with the middle calcaneal articular surface of the talus; **below, it is grooved for the tendon of the Flexor hallucis longus**; its anterior margin gives attachment to the plantar calcaneonavicular ligament, and its medial, to a part of the deltoid ligament of the ankle-joint.

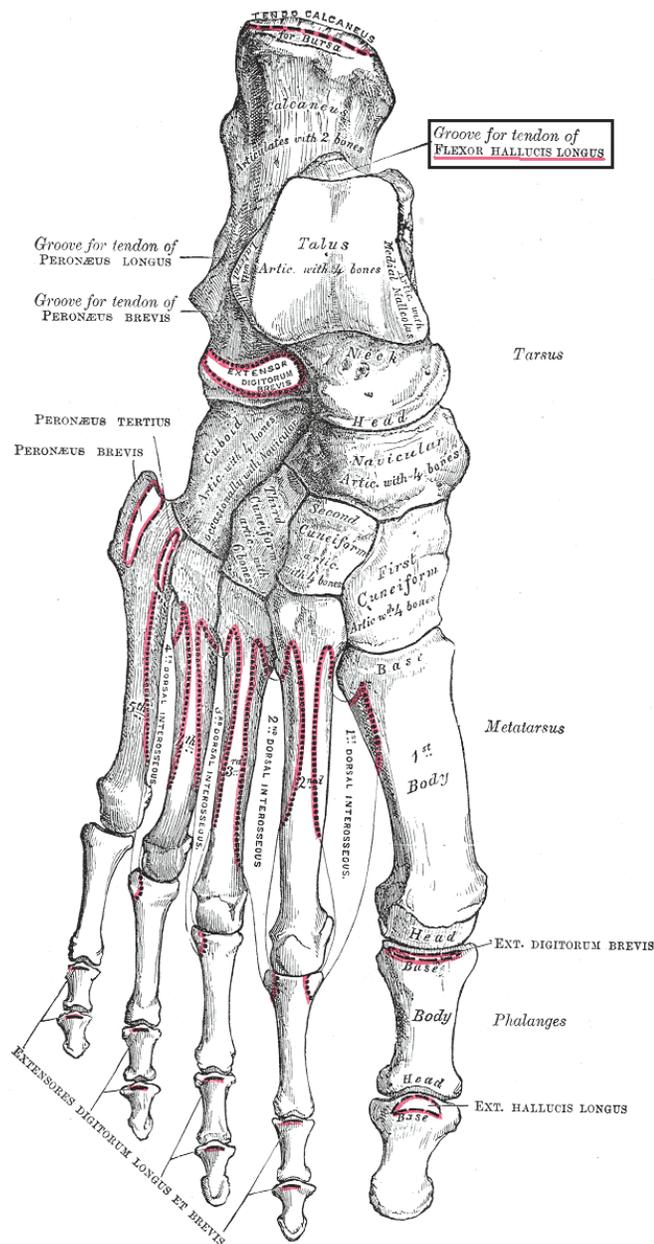


#### **Also know**

The **medial surface** (of calcaneus) is deeply concave; it is directed obliquely downward and forward, and serves for the transmission of the plantar vessels and nerves into the sole of the foot; it affords origin to part of the Quadratus plantæ. At its upper and forepart is a horizontal eminence, the **sustentaculum tali**, which gives attachment to a slip of the tendon of the **Tibialis posterior**. This eminence is concave above, and articulates with the middle calcaneal articular

surface of the talus; below, it is grooved for the tendon of the **Flexor hallucis longus**; its anterior margin gives attachment to the plantar calcaneonavicular ligament, and its medial, to a part of the deltoid ligament of the ankle-joint.

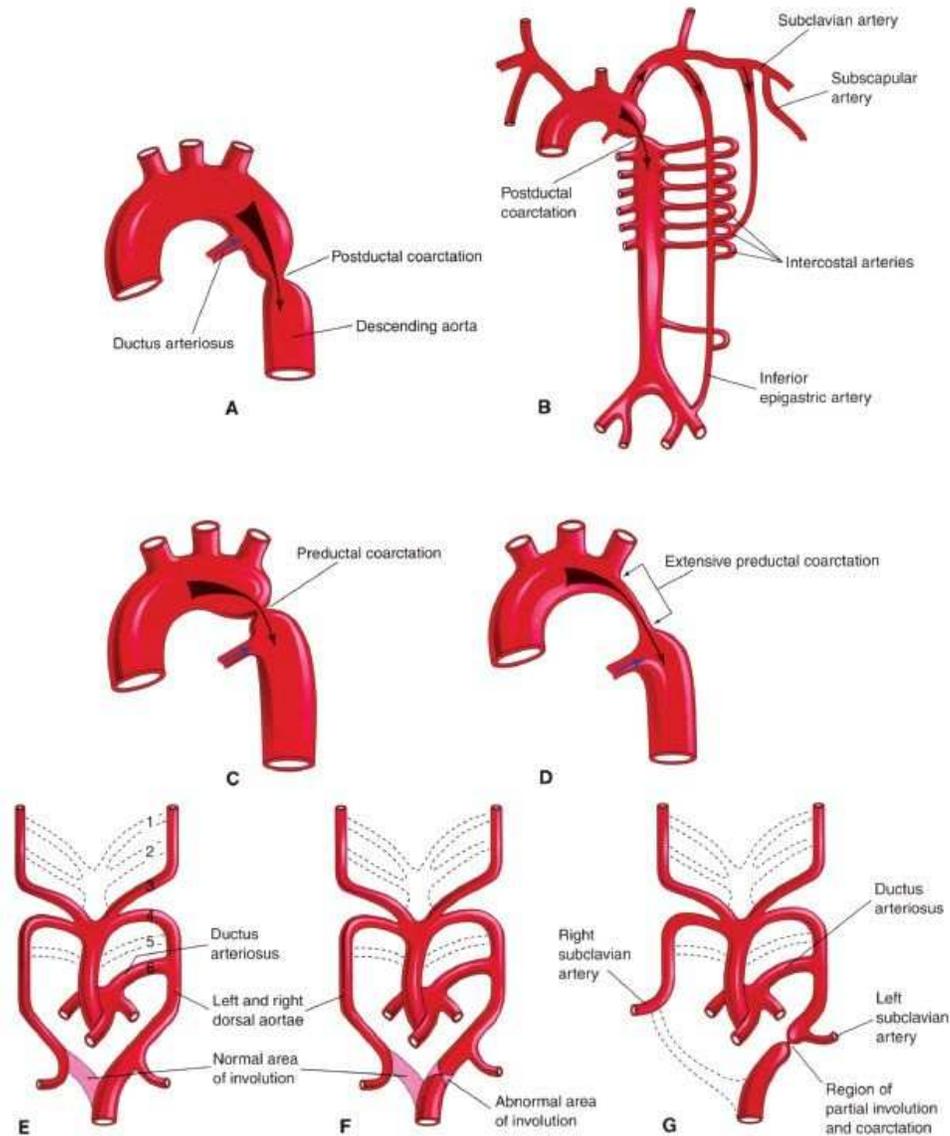
Know both the facts and answer according to what is being asked in the exam.



11. In post ductal Coarctation of aorta collaterals are formed by all except :
- Vertebral artery
  - Suprascapular artery
  - Subscapular artery
  - Posterior intercostal artery

The major pathways of collateral flow include the following:

- Subclavian artery to the internal mammary artery to the intercostal arteries
- Subclavian artery to the costovertebral trunk to the intercostal arteries
- Transverse cervical and **suprascapular arteries** to the intercostal arteries



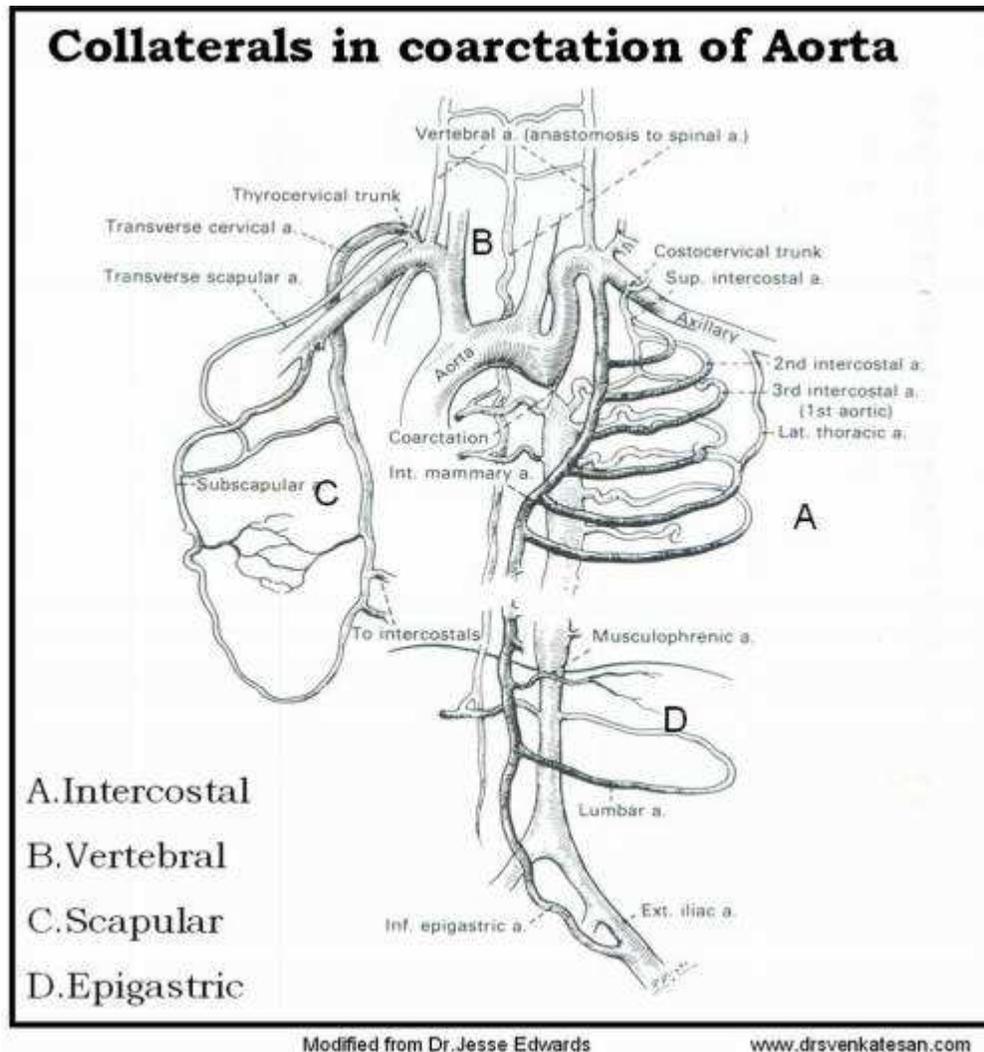
From: Moore, K. L., & Persaud, T. (2008). *The developing human—clinically oriented embryology*, (8th ed., p. 323).

**Coarctation of the aorta.** A, Postductal coarctation of the aorta. B, Diagrammatic representation of the common routes of collateral circulation that develop in association with postductal coarctation of the aorta. C and D, Preductal coarctation. E, Sketch of the aortic arch pattern in a 7-week embryo, showing the areas that normally involute. Note that the distal segment of the right dorsal aorta normally involutes as the right subclavian artery develops. F, Abnormal involution of a small distal segment of the left dorsal aorta. G, Later stage, showing the abnormally involuted segment appearing as a coarctation of the aorta. This moves to the region of the ductus arteriosus with the left subclavian artery. These drawings (E to G) illustrate one hypothesis about the embryological basis of coarctation of the aorta.

**Also know**

Vertebral artery *may* also be involved in collateral formation. See the following figure yourself.

Ref: Dr.Jesse Edwards (*Fowler's Text book of Cardiology* )



12. Movements taking place during abduction of shoulder joint are all except :

- axial rotation of humerus at acromioclavicular joint
- elevation of humerus
- Medial rotation of scapula**
- Movements at the clavicular end of sternoclavicular joint

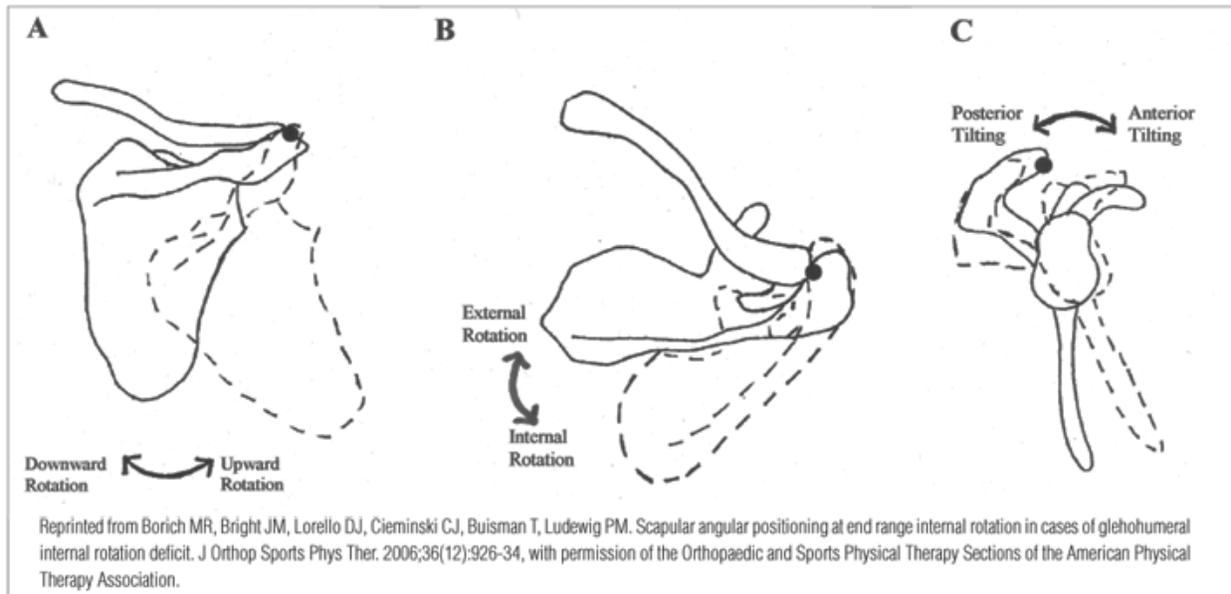
Arm abduction occurs when the arms are held at the sides, parallel to the length of the torso, and are then raised in the plane of the torso. This movement may be broken down into two parts: **True**

**abduction** of the arm, which takes the humerus from parallel to the spine to perpendicular; and **upward rotation of the scapula**, which raises the humerus above the shoulders until it points straight upwards.

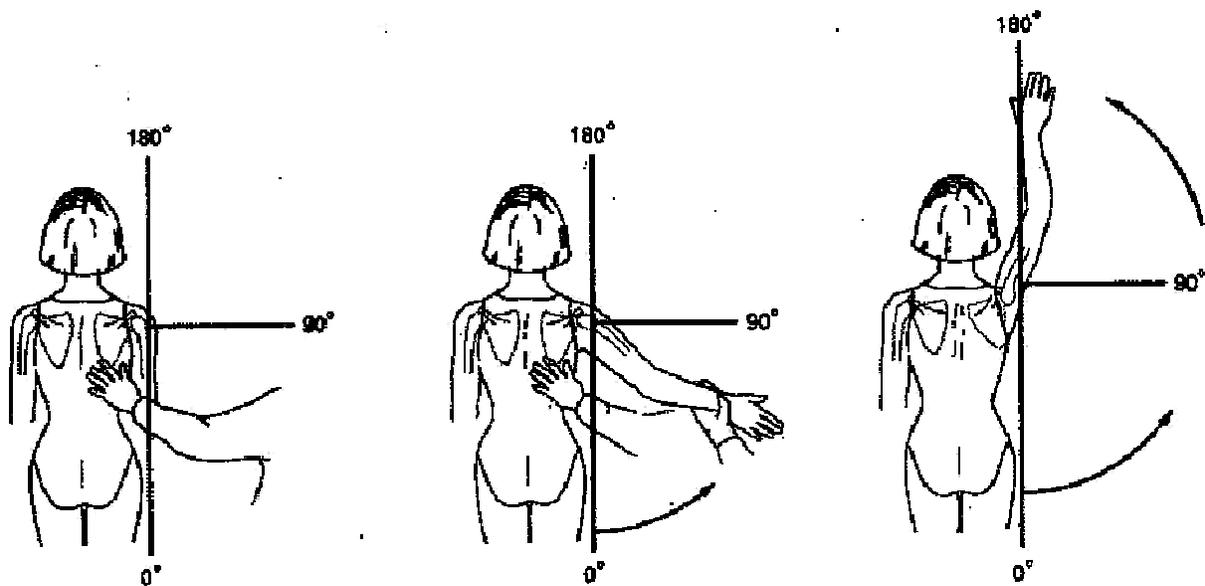
**Muscles:**

True abduction: supraspinatus (first 15 degrees), deltoid

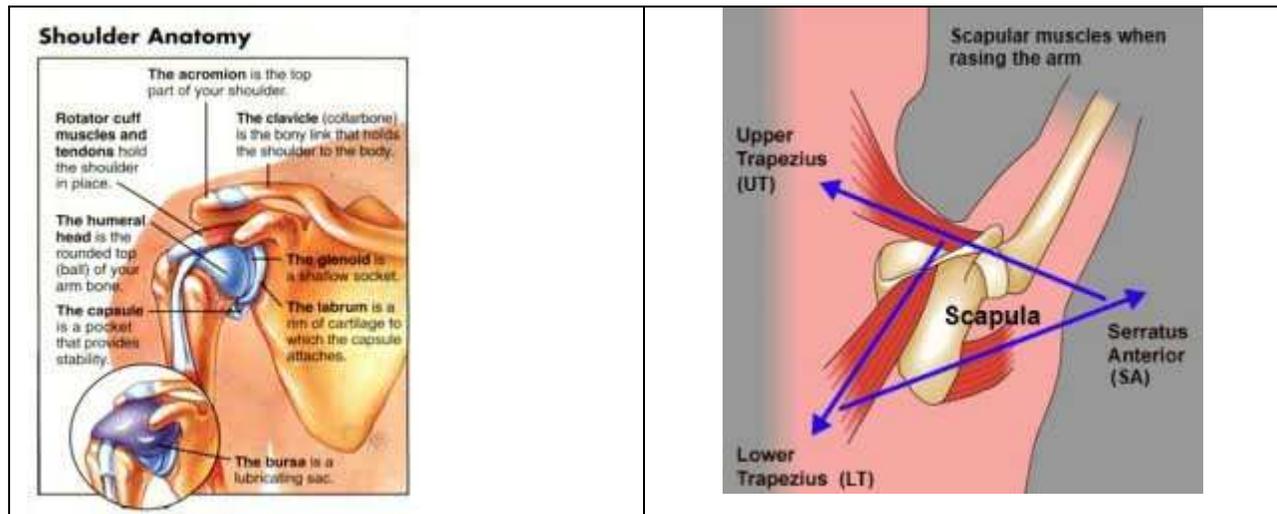
Upward rotation: trapezius, serratus anterior



**Figure 1.** Scapular Motions. A) Upward/downward rotation about an axis perpendicular to the plane of the scapula; B) Internal/external rotation about a superiorly directed axis; and C) Anterior/posterior tilting about a laterally directed axis.



**Figure 2.** Shows lateral rotation of scapula during abduction of arm



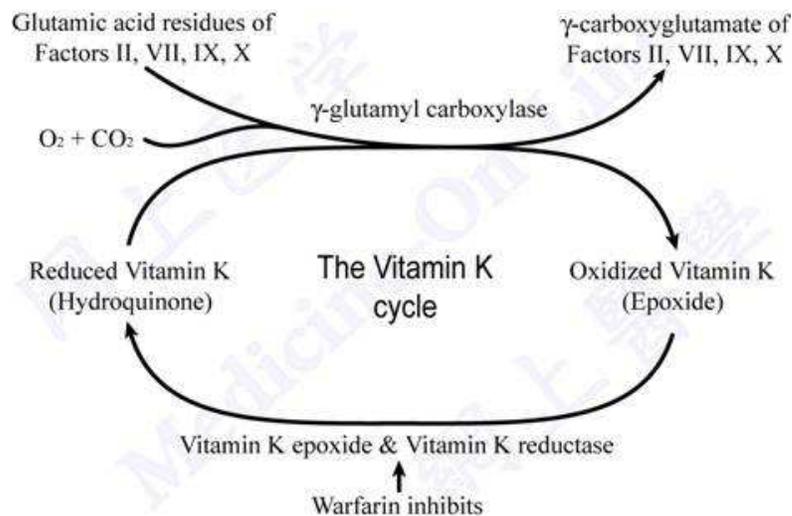
**Figure 3.** Shows the anatomy of shoulder joint and scapular movement while raising the arm

# PHYSIOLOGY

13. Vitamin K dependent clotting factors are :

- IX, X
- VIII, X
- VII, XI
- XI, XII

Vitamin K dependent clotting factors are factors **II, VII, IX and X**.



## Extra Edge

### Familial deficiency of vitamin K-dependent clotting factors

Combined deficiency of **vitamin K-dependent clotting factors II, VII, IX and X** (and proteins C, S, and Z) is usually an acquired clinical problem, often resulting from liver disease, malabsorption, or warfarin overdose. A rare inherited form of defective gamma-carboxylation resulting in early onset of bleeding was first described by McMillan and Roberts in 1966 and subsequently has been termed '**vitamin K-dependent clotting factor deficiency**' (VKCFD). Biochemical and molecular studies identify two variants of this **autosomal recessive** disorder: **VKCFD1**, which is associated with point mutations in the gamma-glutamylcarboxylase gene (**GGCX**), and **VKCFD2**, which results from point mutations in the vitamin K epoxide reductase gene (**VKOR**). Bleeding ranges in severity from mild to severe. Therapy includes high oral doses of vitamin K for prophylaxis, usually resulting in partial correction of factor deficiency, and episodic use of plasma infusions or prothrombin complex concentrate.

14. Which of the following is non – respiratory function of lung :

- Excretion of anions
- Sodium exchange**

- c) Potassium exchange
- d) Calcium exchange

### Non-Respiratory Lung Functions

Whilst the main function of the lung is for respiratory gas exchange, it has several other important physiological roles.

These include:

- reservoir of blood available for circulatory compensation
- filter for circulation:
  - thrombi, microaggregates etc
- metabolic activity:
  - activation:
    - angiotensin I to II (this helps in Na reabsorption)
  - inactivation:
    - noradrenaline
    - bradykinin
    - 5 H-T
    - some prostaglandins
- immunological:
  - IgA secretion into bronchial mucus

15. Maximum water absorption occurs in

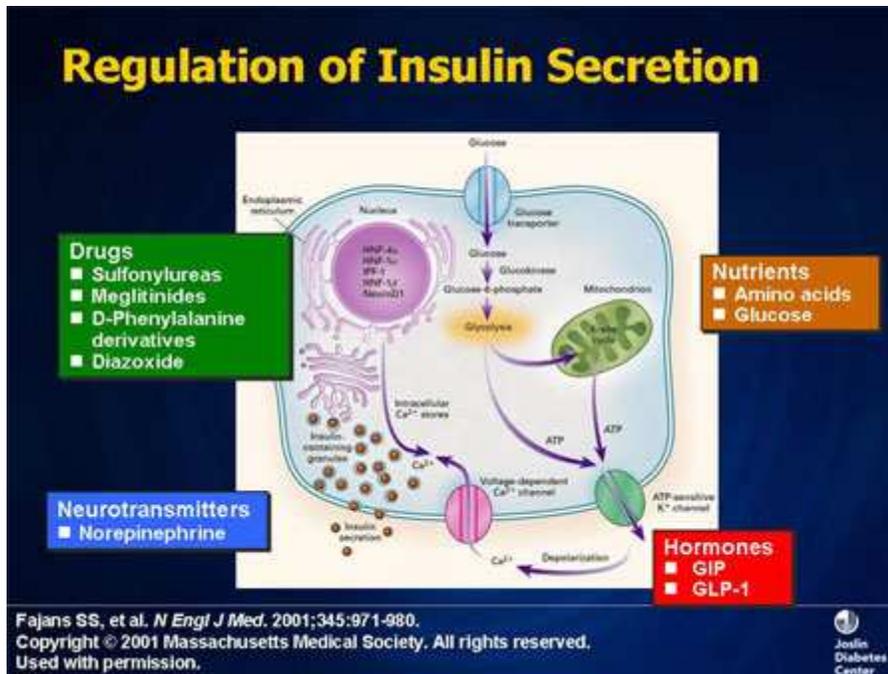
- a) Stomach
- b) Jejunum
- c) Ileum
- d) Colon

Jejunum absorbs 5.5 L/day of water in the GIT.

16. All of the following increase insulin secretion except :

- a) GH
- b) Epinephrine
- c) Secretin
- d) Gastrin

Epinephrine ↓ insulin secretion.



17. Action of NO on smooth muscle cells in intestine :

- Vasodilatation
- Inhibition of smooth muscle motility
- Act through cGMP
- Vasoconstriction

Non-cholinergic non-adrenergic neural mechanisms involving nerves containing NO have been shown to modulate smooth muscle in the gastrointestinal tract, and it has been suggested that release from tonic NO inhibition may be important in the regulation of cyclical fasting **small intestinal motility**.

Leukocyte-derived inducible **NO inhibits gastrointestinal motility** after manipulation and plays an essential role in the initiation of intestinal inflammation.

18. Cardiac motion is least at :

- Early systole
- Mid systole
- Mid diastole
- Early diastole

Motion artifacts in electrocardiographically gated cardiac CT are usually minimized by reconstructing images at **phases of near quiescence in mid-diastole**.

“The period in which the heart has the least motion is usually (but not always) in mid diastole, near a phase between 55% and 75%.” - *MDCT: a practical approach* -Sanjay Saini, Geoffrey D. Rubin, Mannudeep K. Kalr

19. In standing position , venous return is affected by all except :

- a) Calf muscle
- b) Arterial pressure
- c) Fascial planes
- d) Competent valves of perforators

#### Major factors influencing venous return

1) **Respiratory cycle** - Central venous pressure (CVP) decreases with inspiration thereby increasing venous return. This is explained by the negative intrathoracic pressure originated at inspiration, which is transmitted to the great veins of the thorax; moreover, the downward diaphragm movement during this phase helps the pulling of blood toward the heart by increasing the intrabdominal pressure. At expiration, the mechanisms reverse. 2) **Venous tone** - is governed by autonomous system. 3) **Right heart function** - The blood reaching the right ventricle is pumped to the pulmonary circulation and therefore will not be damped backward in the venous system. 4) **Gravity** - Venous pressure increases by approximately 0.77 mmHg for each centimeter (cm) below right atrium In a standing person, the venous pressure around the ankle is about 90 mmHg. Gravity actually causes **blood pooling** in the legs. 5) **Muscle pump - Muscle contractions** helps venous return by **compressing** the surrounding veins - the so-called **muscle pump**.

20. In heavy exercise, cardiac output is increased to five times but there is little change in pulmonary artery pressure , why ?

- a) Exercise induced hyperventilation causes constriction of pulmonary vasculature
- b) Upper lobe perfusion increases more than ventilation
- c) Sympathetic stimulation decreases tone of vasculature
- d) Opening of parallel vessels

As exercise progresses, pulmonary blood flow increases, indicating an abrupt fall in pulmonary vascular resistance (PVR). This ↓resistance indicates an ↑in cross-sectional area of the pulmonary vasculature as **more capillaries are recruited**, especially in the lung apices, and with distension of the already recruited capillaries.” - *ACSM's advanced exercise physiology* By Charles M. Tipton, American College of Sports Medicine

21. BMR depends on :

- a) Lean body mass
- b) BMI
- c) Obesity

d) Body surface area

22. Decreased BMR is seen in :

- a) **Obesity**
- b) Hyperthyroidism
- c) Feeding
- d) Exercise

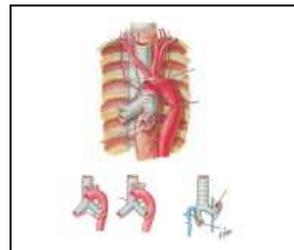
23. Low CSF protein is seen in all except :

- a) Infants
- b) Recurrent lumbar puncture
- c) **Hypothyroidism**
- d) Pseudotumor cerebri

Low CSF protein levels can occur in conditions such as **repeated lumbar puncture** or a chronic leak, in which CSF is lost at a higher than normal rate. Low CSF protein levels also are seen in **some children between the ages of six months and two years**, in acute water intoxication, and in a minority of patients with **idiopathic intracranial hypertension (pseudotumor cerebri)**. CSF protein levels do not fall in hypoproteinemia.

24. True about bronchial artery is all except :

- a) 2% of systemic circulation
- b) **Helps in gaseous exchange**
- c) Causes venous mixing of blood
- d) Nutritive function to lung



Gaseous exchange is done by **pulmonary arteries**, not by bronchial arteries.

25. Hot water bottles used to relieve intestinal spasm acts by :

- a) Stimulation of adrenergic receptors
- b) Stimulation of cholinergic receptors
- c) Inhibition of cold receptors
- d)  $\uparrow$ temperature of peritoneal fluid

*I am giving the fact below . Read them & decide for yourself. Option A appears to be the most probable answer.*

Therapeutic heat has physiologic effects that are mediated via neurologic, vascular, and biophysical mechanisms. The neurologic mechanisms of pain relief are thought to occur through the **–gate control” theory of pain inhibition**. This theory is based in part on the inhibition (–gating”) of noxious signals in the substantia gelatinosa of the dorsal horn of the spinal cord. **Topical heat increases the temperature of the skin and underlying tissues, leading to stimulation of thermoreceptors**. The resultant **increase in small**

**nonmyelinated c-fiber activity** in first-order afferent neurons inhibits the concurrent nociceptive signals in other first-order neurons. This interaction is thought to occur at the synapse between first- and second-order neurons, and is termed **presynaptic inhibition**. Functional brain imaging has shown that gentle warming of the skin activates the thalamus and posterior insula. Touch stimulation of the skin activates the thalamus and S2 region of the cerebral cortex as well. These data suggest that some of the potential benefits provided by topical heat may be mediated by higher centers in the brain in addition to the effects shown in the substantia gelatinosa. Feelings of comfort and relaxation associated with topical heat therapy also mitigate the integration and coherence of the pain experience in the brain. **Locally, vascular mechanisms contribute by increasing blood flow to the tissues, resulting in dilution of prostaglandins, bradykinins, and histamine, which are modulators or instigators of nociception.** This local increase in blood flow brings improved oxygenation and nutrients that enhance tissue repair.

Biophysical mechanisms come into play when heat is used to treat musculoskeletal pain. Topical heat increases connective tissue extensibility, supporting a biophysical mechanism of thermotherapy. This phenomenon is called **plastic deformation** or elongation. Skeletal muscle spasm is relieved through a reflex action via thermoreceptors and a decline in muscle spindle  $\gamma$  fiber activity, decreasing the muscle spindles' sensitivity to stretching. Heat may also relieve muscle spasm by activating descending pain-inhibitory systems via an unknown mechanism. Decreased motor neuron activity from the dorsal horn of the spinal cord, resulting in a lowering of muscle tone, has also been postulated.

# BIOCHEMISTRY

26. Sphingomyelinase deficiency occurs in :

- a) Gaucher's disease
- b) Tay-Sachs disease
- c) Fabry's disease
- d) **Niemann-Pick disease**

Disease	Enzyme deficiency
Gaucher's disease	$\beta$ -glucosidase
Tay-Sachs disease	Hexosaminidase A
Fabry's disease	$\alpha$ -galactosidase
Niemann-Pick disease	Sphingomyelinase

27. Rothera test is done for :

- a) Glucose
- b) **Ketone bodies**
- c) Bile pigments
- d) Protein

28. In electrophoresis which lipoprotein moves the least :

- a) LDL
- b) HDL
- c) VLDL
- d) **Chylomicron**

**Order of mobility in electrophoresis:**

**HDL > LDL > VLDL > Chylomicron**

Memory Aid : **High** density lipoprotein is **highly** mobile on electrophoresis.

29. Branch chain amino acid oxidation is defective in :

- a) **Maple syrup urine disease**
- b) Hartnup disease
- c) Phenylketonuria
- d) Alkaptonuria

**Maple syrup urine disease** (MSUD), also called **branched-chain ketoaciduria**, is an autosomal recessive metabolic disorder affecting branched-chain amino acids. It is one type of organic acidemia. The condition gets its name from the distinctive sweet odor of affected infants' urine.

MSUD is caused by a deficiency of the branched-chain alpha-keto acid dehydrogenase complex (BCKDH), leading to a buildup of the branched-chain amino acids (leucine, isoleucine, and valine) and their toxic by-products in the blood and urine.

The disease is characterized in an infant by the presence of sweet-smelling urine, with an odor similar to that of maple syrup. Infants with this disease seem healthy at birth but if left untreated suffer severe brain damage and eventually die.

From early infancy, symptoms of the condition include poor feeding, vomiting, dehydration, lethargy, hypotonia, seizures, hypoglycaemia, ketoacidosis, opisthotonus, pancreatitis, coma and neurological decline.

30. Mitochondrial liver enzyme is :

- a) Alkaline phosphatase
- b) SGOT
- c) SGPT
- d) GGT

ALT (SGPT) – Cytoplasmic enzyme

AST (SGOT) – Found in both cytoplasm & mitochondria

31. Method used to differentiate the genetic expression of tumor cell vs. normal cells :

- a) Northern blot
- b) Southern blot
- c) Western blot
- d) Microarray

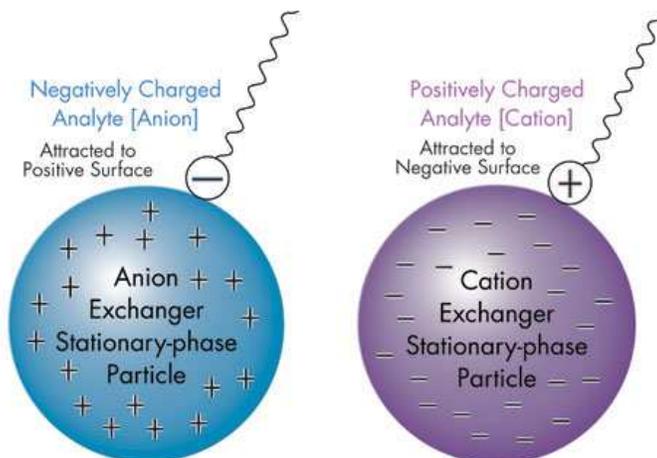
A **DNA microarray** is a multiplex technology used in molecular biology. It consists of an arrayed series of thousands of microscopic spots of DNA oligonucleotides, called features, each containing picomoles ( $10^{-12}$  moles) of a specific DNA sequence, known as *probes* (or *reporters*). These can be a short section of a gene or other DNA element that are used to hybridize a cDNA or cRNA sample (called *target*) under high-stringency conditions. Probe-target hybridization is usually detected and quantified by detection of fluorophore-, silver-, or chemiluminescence-labeled targets to determine relative abundance of nucleic acid sequences in the target. Since an array can contain tens of thousands of probes, a microarray experiment can accomplish many genetic tests in parallel. Therefore arrays have dramatically accelerated many types of investigation. DNA microarrays are often used to determine the differing patterns of gene expression in two different types of cell, e.g. normal & cancer cells.

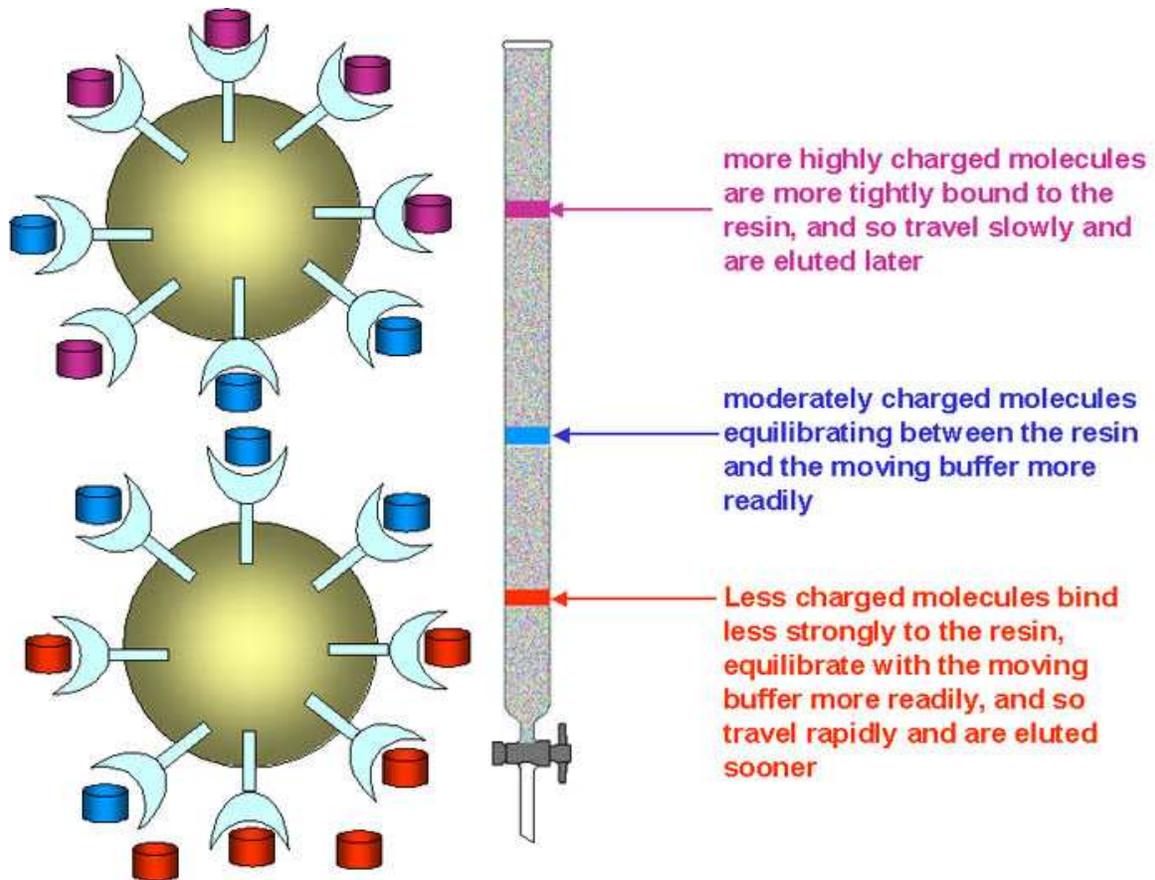
32. The procedure of protein separation in which a charged moving ion collides with a static ion is known as :

- Ion exchange chromatography
- Absorption chromatography
- Partition chromatography
- Electrophoresis

**Ion-exchange chromatography** (or *ion chromatography*) is a process that allows the separation of ions and polar molecules based on their charge. It can be used for almost any kind of charged molecule including large proteins, small nucleotides and amino acids. The solution to be injected is usually called a *sample*, and the individually separated components are called *analytes*. It is often used in protein purification, water analysis, and quality control.

Ion exchange chromatography retains analyte molecules on the column based on coulombic (ionic) interactions. **The stationary phase surface displays ionic functional groups (R-X) that interact with analyte ions of opposite charge.** This type of chromatography is further subdivided into cation exchange chromatography and anion exchange chromatography. The ionic compound consisting of the cationic species  $M^+$  and the anionic species  $B^-$  can be retained by the stationary phase.





# PATHOLOGY

33. Pathogenesis of all of the following is granulomatous except :

- a) Wegener's granulomatosis
- b) **Buerger's disease**
- c) Takayasu arteritis
- d) Churg-Strauss syndrome

**Thromboangiitis obliterans** (also known as **Buerger's disease**) is a recurring **progressive** inflammation **and** thrombosis (clotting) of small and medium arteries and veins of the hands and feet. It is strongly associated with use of tobacco products, primarily from smoking, but also from smokeless tobacco.

34. Marker of GIST is :

- a) CD 34
- b) **CD 117**
- c) CD 15
- d) CD 45

The diagnosis of GIST is often suspected histologically, but such tumors can have a broad morphologic spectrum. A panel of immunohistochemical markers is used to aid in correct diagnosis. A total of 91 to 98% of GISTs stain positively for **KIT (CD117)**, which is a major factor in the initial identification of GIST and therefore is often the inclusion criterion into many reported series. Staining of GISTs for other standard laboratory immunomarkers is more variable, including **CD34** (~70%), **smooth-muscle actin** (~35%), **S-100** (~10%) and **desmin** (~5%). **PKC theta** is an immunohistochemical marker found in two studies to be expressed in GISTs, KIT-negative GISTs and schwannomas, but not in other mesenchymal tumors of the GI tract.

35. Paraganglioma are associated with:

- a) Glycogen deposition
- b) Enlarged mitochondria
- c) Shrunken mitochondria
- d) **Dense core neurosecretory granules**

Most paragangliomas are either asymptomatic or present as a painless mass. While all contain **neurosecretory granules**, only in 1–3% of cases is secretion of hormones such as catecholamines abundant enough to be clinically significant; in that case manifestations often resemble those of pheochromocytomas.

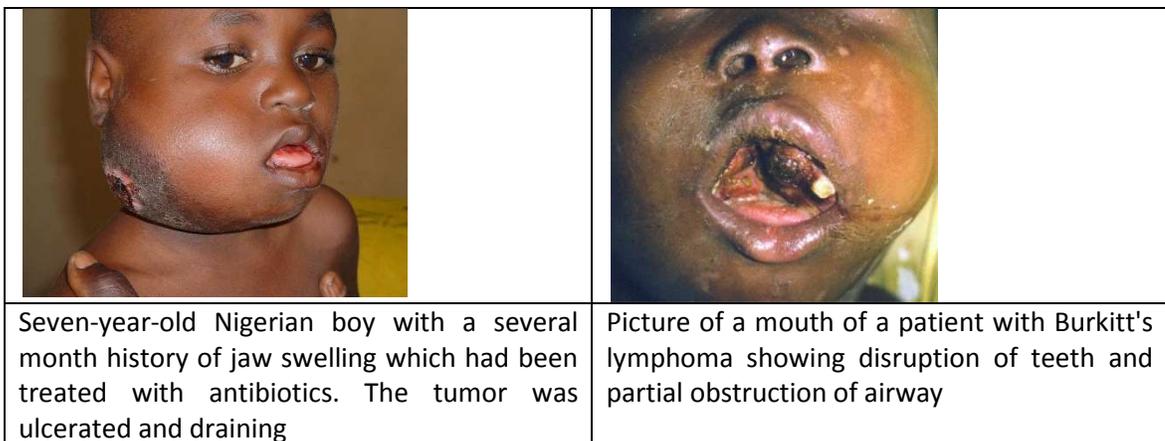
36. About Lung carcinoma all is true except :
- 70% are SCC
  - Oat cell ca is central in distribution
  - Oat cell ca cause cavitations
  - Oat cell ca shows perihilar lymph nodes
37. Burkitt's Lymphoma is associated with :
- t(8:14)
  - t(11:14)
  - t(15:17)
  - t(14:18)

Almost by definition, Burkitt's lymphoma is associated with a chromosomal translocation of the c-myc gene. This gene is found at 8q24.

- The most common variant is t(8;14)(q23;q32). This involves c-myc and IGH@. A variant of this, a three-way translocation, t(8;14;18), has also been identified.
- A rare variant is at t(2;8)(p12;q24). This involves IGK@ and c-myc.
- Another rare variant is t(8;22)(q24;q11). This involves IGL@ and c-myc

38. t(2;8)(p12;q24) is seen in :
- T cell ALL
  - Burkitt mature B
  - Pre B cell ALL
  - AML

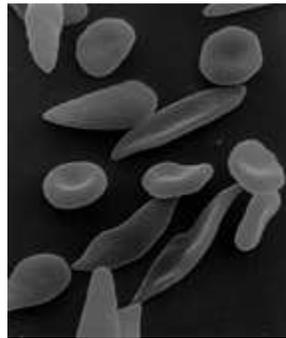
A rare variant of Burkitt's lymphoma is at t(2;8)(p12;q24). This involves IGK@ and c-myc



39. Heterozygous sickle cell anemia gives protection against :

- G6PD
- Malaria
- Thalassemia
- Dengue fever

Those with only one of the two [alleles](#) of the sickle-cell disease are more resistant to malaria, since the infestation of the malaria plasmodium is halted by the sickling of the cells which it infests.



Normal and sickle-shaped red blood cells

40. False about Pseudo –hypoparathyroidism is :

- Low PTH levels
- Low calcium & high phosphate levels
- End organ resistance
- Associated with Albright osteodystrophy

**Pseudohypoparathyroidism** is a condition associated primarily with resistance to the parathyroid hormone. Patients have a low serum calcium and high phosphate, but the **parathyroid hormone level (PTH) is actually appropriately high** (due to the hypocalcemia). Its pathogenesis has been linked to dysfunctional G Proteins (in particular, Gs alpha subunit).

41. Low calcium & high Phosphate is seen in :

- Hyperparathyroidism
- Hypoparathyroidism
- Hyperthyroidism
- Hypothyroidism

- The diagnosis of hypoparathyroidism is supported by **hypocalcemia, hyperphosphatemia, and low parathyroid hormone levels** in the absence of renal failure or intestinal malabsorption.
- Both total and ionized calcium are decreased. Normal total serum calcium levels range from 9-10.5 mg/dL (2.2-2.6 mmol/L). Normal ionized calcium levels are 4.5-5.6 mg/dL (1.1-1.4 mmol/L).

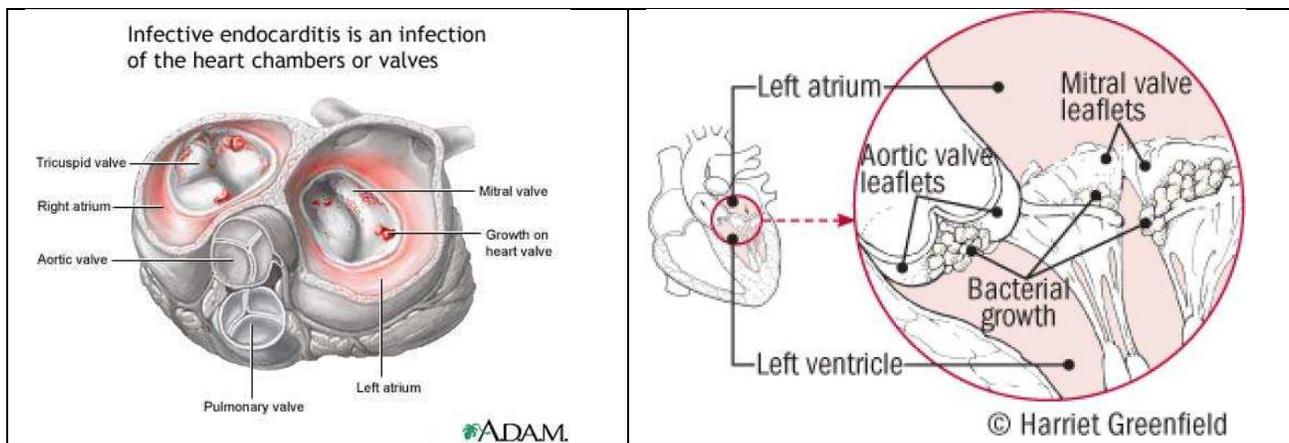
- Serum magnesium level can be low, high, or normal.

42. Which of the following have most friable vegetations :

- Infective endocarditis
- Libman sacks endocarditis
- RHD
- SLE

The hallmark of IE is presence of friable, bulky, potentially destructive vegetations containing fibrin, inflammatory cells and bacteria or other organisms.

Aortic and mitral valve most common sites, valves of right heart may be involved particularly in intravenous drug abusers.



43. Marker of Langerhans cell histiocytosis

- CD 1a
- CD 10
- CD 30
- CD 56

The Langerhans cell displays immunoreactivity (cytoplasmic and nuclear) for S-100 protein and the monoclonal antibody CD1a, HLADR (Ia) and peanut agglutinin.

Ultrastructurally, the Birbeck (or X) granule is straight or curved pentalaminar rod of variable length, often vesicular at one end frequently resembling a racquet.

44. Bradykinin in acute inflammation causes :

- a) Vasodilatation
- b) Vasoconstriction
- c) Pain
- d) ↑vascular permeability

There is compelling evidence linking bradykinin (BK) with the pathophysiological processes that accompany tissue damage and inflammation, especially the production of pain and hyperalgesia. Several mechanisms have been proposed to account for hyperalgesia including the direct activation of nociceptors as well as sensitization of nociceptors through the production of prostanoids or the release of other mediators. In keeping with this, antagonists of the BK B2 receptor are efficacious analgesic and anti-inflammatory agents in acute inflammatory pain. More recently it has been suggested that when inflammation is prolonged, BK B1 receptors, which are not expressed in healthy tissues to a significant degree, also play an important role in the maintenance of hyperalgesia. This may be one of a number of adaptive mechanisms that occur peripherally and centrally following the prolonged activation of nociceptors during inflammation or injury

45. Cavitory lesions in the lung are seen in :

- a) Primary pulmonary TB
- b) Staph. aureus infection
- c) Pneumococcal infn
- d) Pneumocystis carinii

46. Lymphoplasmacytoid lymphoma is associated with :

- a) IgA
- b) IgG
- c) IgD
- d) IgM

**Waldenström's macroglobulinemia** (also known as lymphoplasmacytic lymphoma) is cancer involving a subtype of white blood cells called lymphocytes. The main attributing antibody is IgM. It is a type of lymphoproliferative disease, and shares clinical characteristics with the indolent non-Hodgkin lymphomas.

47. Somatic mutation which leads to PNH is :

- a) BAF
- b) DAF
- c) MIRL
- d) GPI

Paroxysmal nocturnal hemoglobinemia (PNH), a hematopoietic stem cell disorder, arises from a somatic mutation of the phosphatidylinositol glycan-class A (*PIG-A*) gene. The gene product is required in the biosynthesis of a glycosylphosphatidylinositol (GPI) structure and serves as an anchor for a group of membrane proteins. The PNH cells are characterized by a total or partial lack of the GPI-anchored membrane proteins. Without this structure, intravascular hemolysis occurs due to the inability to regulate the lytic and cell-stimulatory activities of complement on the membrane surface of hematopoietic cells. Two proteins, CD55-decay accelerating factor and CD59-membrane inhibitor of reactive lysis, are known to be tethered to the cell membrane by the GPI-anchor. Additionally, platelets lacking CD59 may cause venous thrombosis with possible Budd-Chiari syndrome. The development of PNH requires a hypoplastic bone marrow, somatic mutation restricted to the *PIG-A* gene in the stem cell, and clonal expansion of the hematopoietic stem cell pool.

48. Which special stain is used to study fungal morphology in tissue sections :

- a) PAS
- b) Von Kossa
- c) Massan's trichrome
- d) Alizarin red

49. In a person with mutation in Rb gene if retinoblastoma is not seen then the probability of finding which carcinoma is highest :

- a) RCC
- b) Osteosarcoma
- c) Pinealoblastoma
- d) Chondrosarcoma

50. Caspases are involved in :

- a) Cell division
- b) Apoptosis
- c) Cell necrosis
- d) Inflammation

51. Which of the following cannot be used in aneuploidy detection :

- a) FISH
- b) RT-PCR
- c) Nested PCR
- d) Microarray

Microarray can detect point mutation, small deletions & insertions but not numerical or structural abnormalities in chromosomes.

52. During light microscopy , the characteristic feature seen in apoptosis is :

- a) Cell membrane normal
- b) Cell appears swollen
- c) Nuclear moulding
- d) Eosinophilic cytoplasm

**Chromatin condensation** is the most characteristic feature of apoptosis.

53. Down's syndrome is most commonly associated with :

- a) Maternal non-disjunction
- b) Paternal non-disjunction
- c) Balanced translocation
- d) Mosaicism

54. Genetics of Chromophobe cell variant of RCC ?

- a) Mutant VHL gene
- b) 3p deletion
- c) 5p gain
- d) Trisomy of 7, 17

*None of the options seem to be correct. Read the information given below & decide for yourself.*

Chromophobe RCCs generally have a tendency to grow very slowly in vitro in comparison to all other type of renal tumors. This may be a reason why cytogenetic reports are scarce and usually few metaphases of poor quality were available for investigation. A low chromosome number ranging between 32-39, without discernable structural changes was the most frequent cytogenetic finding. **Chromosomes 1, 2, 6, 10, 13, 17 and 21 were most frequently lost.** Additional structural aberrations have been described. Endoreduplication of the cells with hypodiploid karyotype has been observed. It is of interest, the presence of an hypodiploid clone can be disclosed by a DNA index of 0.86. The low chromosome number has been confirmed by other techniques such as flow cytometry, comparative genomic hybridization (CGH), restriction fragment length polymorphism (RFLP) analysis, and polymorphic microsatellite markers.

55. AFP most likely to be raised in :

- a) Seminoma
- b) Hepatoblastoma
- c) Choriocarcinoma
- d) Down syndrome

**"Hepatoblastoma** – **Most patients (98%) present with raised AFP,** often at extremely high concentrations (e.g.  $10^6$   $\mu$ g), which can assist in diagnosis. Thereafter, AFP can be used to

monitor the therapy & follow-up of these patients.” – *Clinical Biochemistry: Metabolic & Clinical Aspects* by William J.Marshall, Stephen K.Bangert, p.907

56. AFP is not raised in :

- a) Teratoma
- b) Yolk sac tumor
- c) Pure choriocarcinoma
- d) Embryonal cell ca

Alpha-fetoprotein (AFP) is secreted by yolk sac elements; elevated levels of AFP are consistent with NSGCT. Choriocarcinoma could be a component of such a tumor, but AFP is within the reference range in pure choriocarcinoma. AFP has a serum half-life of between 5 and 7 days.

57. Cystic fibrosis is an autosomal recessive disorder . A normal couple is having two daughters out which one is affected with the disease. What is the chance of her sister being carrier of the disease ?

- a) 0
- b)  $\frac{1}{2}$
- c)  $\frac{1}{4}$
- d)  $\frac{2}{3}$

C-Normal allele, c-abnormal allele

Cc x Cc gives CC(normal), Cc(carrier), Cc(carrier) and cc(affected).

So chance of her sister being a carrier =  $\frac{2}{3}$

58. Most common cause of abdominal aortic aneurysm is :

- a) Marfan syndrome
- b) Syphilis
- c) Atherosclerosis
- d) Cystic medial necrosis

59. Males are more commonly involved than female in :

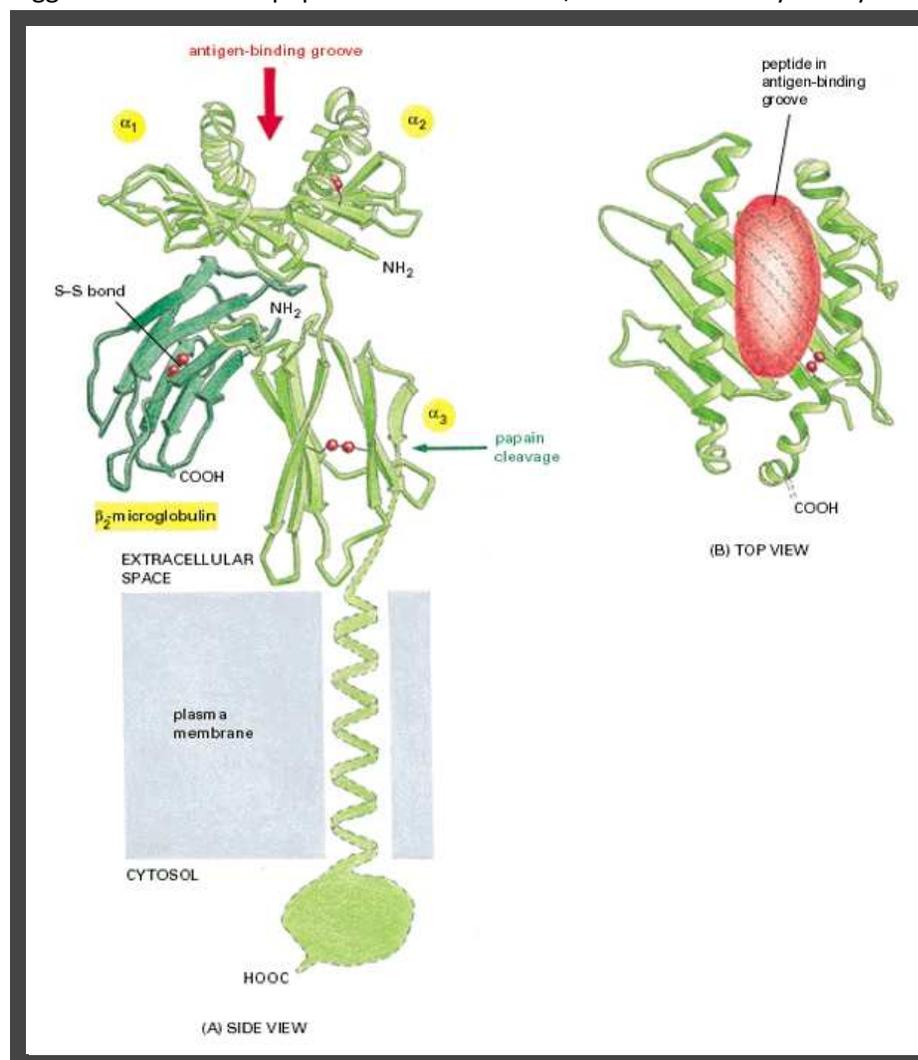
- a) AD
- b) AR
- c) XD
- d) XR

60. Type I MHC ( HLA)presents peptide antigen to Tcells (CD8+) so that the peptide binding site is formed by :

- a) Distal domain of  $\alpha$ -subunit

- b) Proximal domain of  $\alpha$ -subunit
- c) Between  $\alpha_1$  &  $\beta_1$
- d) Between  $\alpha_2$  &  $\beta_2$

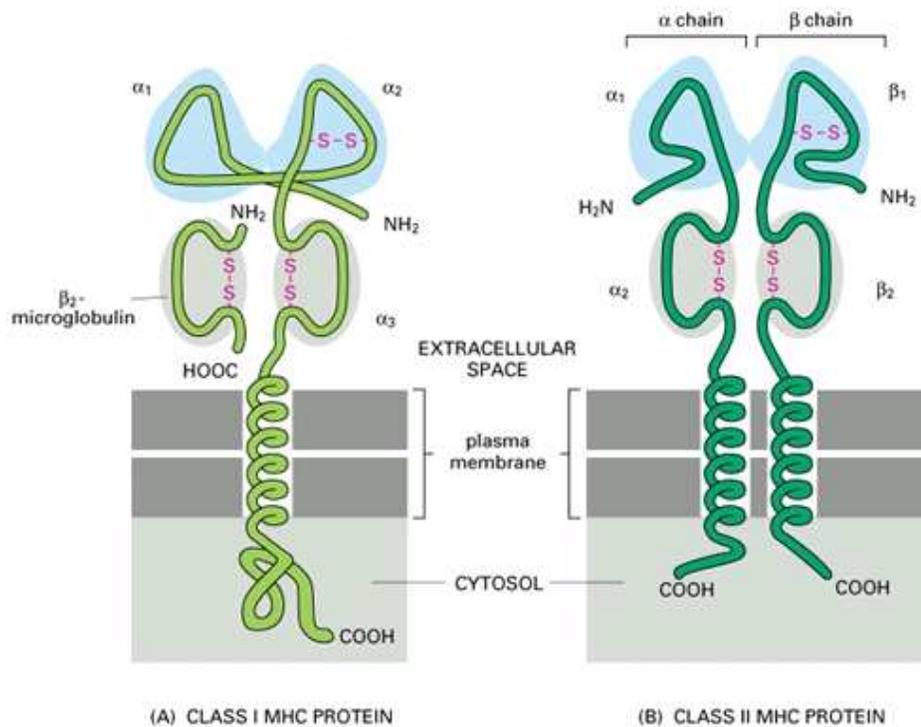
Class I MHC protein has a single peptide-binding site located at one end of the molecule. This site consists of a deep groove between two long  $\alpha$  helices derived from the nearly identical  $\alpha_1$  and  $\alpha_2$  domains; the base of the groove is formed by eight  $\beta$  strands derived from the same two domains. The groove is large enough to accommodate an extended peptide of about 10 amino acid residues. In fact, when a class I MHC protein was first analyzed by x-ray crystallography, this groove was found to contain a small density, suspected to be bound peptide that had co-crystallized with the MHC protein. This finding implicated the groove as the antigen-binding site and suggested that once a peptide binds to this site, it dissociates very slowly.



### Summary

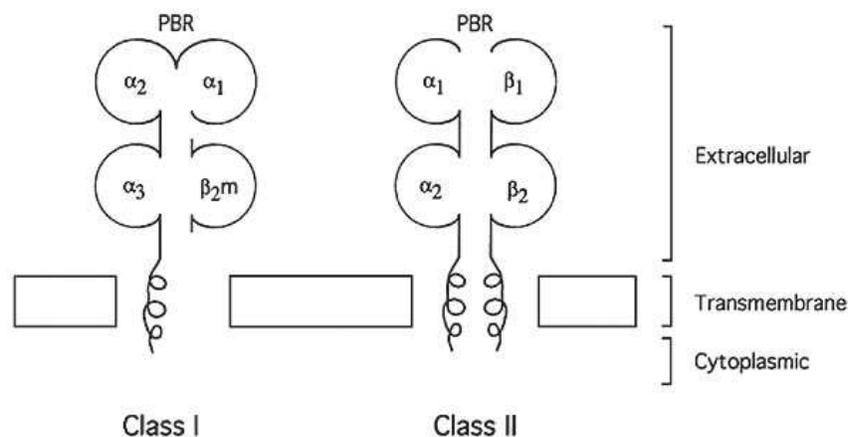
Peptide binding region of MHC I – Between  $\alpha_1$  and  $\alpha_2$

Peptide binding region of MHC II – Between  $\alpha_1$  and  $\beta_1$



### Class I and class II MHC proteins

(A) The  $\alpha$  chain of the class I molecule has three extracellular domains,  $\alpha_1$ ,  $\alpha_2$ , and  $\alpha_3$ , encoded by separate exons. It is noncovalently associated with a smaller polypeptide chain,  $\beta_2$ -micro-globulin, which is not encoded within the MHC. The  $\alpha_3$  domain and  $\beta_2$ -microglobulin are Ig-like. While  $\beta_2$ -microglobulin is invariant, the  $\alpha$  chain is extremely polymorphic, mainly in the  $\alpha_1$  and  $\alpha_2$  domains. (B) In class II MHC molecules both chains are polymorphic ( $\beta$  more than  $\alpha$ ), mainly in the  $\alpha_1$  and  $\beta_1$  domains; the  $\alpha_2$  and  $\beta_2$  domains are Ig-like. Thus there are striking similarities between class I and class II MHC proteins. In both, **the two outermost domains** (shaded in blue) interact to form a groove that binds foreign antigen and presents it to T cells. All of the chains are glycosylated except for  $\beta_2$ -microglobulin (not shown).



**Figure 2.** Schematic presentation of the structure of MHC class I and class II molecules. PBR = peptide-binding region. (Reprinted, with permission, from the Annual Review of Genetics, Vol. 32 ©1998 by Annual Reviews, www.annualreviews.org).

61. Lupus anticoagulant is not associated with :

- a) Bleeding
- b) Thrombosis
- c) Recurrent abortion
- d) Anti-phospholipid syndrome

**Lupus anticoagulant** (also known as **lupus antibody, LA, or lupus inhibitors**) is an immunoglobulin that binds to phospholipids and proteins associated with the cell membrane. Since interactions between the cell membrane and clotting factors are necessary for proper functioning of the coagulation cascade, the lupus anticoagulant can interfere with blood clotting as well as in-vitro tests of clotting function. **Paradoxically, lupus anticoagulants are also risk factors for thrombosis.**

Conceptually, lupus anticoagulants overlap with the antiphospholipid antibody syndrome. Lupus anticoagulants can be understood as the tendency of antiphospholipid antibodies to prolong the clotting times, especially in phospholipid rich clotting testing such as the dilute Russell's viper venom time.

Often, the lupus anticoagulant is diagnosed on asymptomatic patients by a routine blood testing prior to surgery. Patients with a lupus anticoagulant are **prone to** thrombosis **and** habitual abortion (repeated miscarriages).

Both words in the term "lupus anticoagulant" can be misleading:

- Most patients with a lupus anticoagulant do not actually have lupus erythematosus, and only a small proportion will proceed to develop this disease (which causes joint pains, skin problems and renal failure, amongst other complications). Patients with lupus erythematosus are more likely to develop a lupus anticoagulant than the general population..
- **The term "anticoagulant" accurately describes its function in vitro, but in vivo, it is now known that it functions as a coagulant.**

62. What is the cause of hypercoagulation in nephrotic syndrome :

- a) Loss of anti-thrombin III
- b) ↓fibrinogen
- c) ↓metabolism of Vit K
- d) ↑protein C

**“Hypercoagulable states: Nephrotic syndrome** – The reason here is **loss of natural anticoagulant proteins, especially antithrombin,** in the urine.” – *Diagnostic Hematology by Norman Beck, p.434*

63. Which of the following regarding carcinoid heart is true ?

- a) Calcification of tricuspid valve

- b) Intimal fibrosis of RV, tricuspid valve & pulmonary valve
- c) Thickening of RV
- d) Tricuspid stenosis

### **Carcinoid Heart Disease:**

Carcinoid heart disease is a rare, metastatic disease.

Cardiac involvement in carcinoid syndrome is mainly seen in the endocardium and valves of the right heart.

Gross appearance : There are pearly-gray, uniform plaque-like thickenings on the endocardium of tricuspid and pulmonary valves and on the right ventricle.

Microscopic appearance : The plaques are composed of mainly smooth muscle cells and some collagen fibers, embedded in an acid mucopolysaccharide matrix.

The lesion is thought to be due to serotonin (5-HT) , which is inactivated during the passage of blood through the lungs by the monoamino oxidase present in the pulmonary vascular endothelium.

Cardiac involvement (due to systemic effects of circulating vasoactive amines produced by a metastatic carcinoid tumour) is detected in 50 -70% of patients.

Prognosis is poor due to development of heart failure and death.

Most patients present with right-sided heart valve dysfunction since pulmonary and tricuspid valves lesions are the most common (>95%) cardiac pathology.

Left-sided valvular involvement, and angina associated with coronary vasospasm occur in 10% of patients. Lesions on the left side are seen in patients with defective atrial or ventricular septum.

### **Note:**

Carcinoid tumours are rare neuroendocrine malignancies mostly arising within the gastrointestinal system, particularly the ileum and appendix.

The carcinoid syndrome, characterised by cutaneous flushing, secretory diarrhoea, and bronchospasm, occurs secondary to the paraneoplastic effects of vasoactive tumour products such as serotonin and only occurs in the presence of metastatic spread.

Raised mean 24 hour urinary excretion of 5-hydroxy-indole acetic acid (5-HIAA), an end product of serotonin metabolism, is a key diagnostic finding.

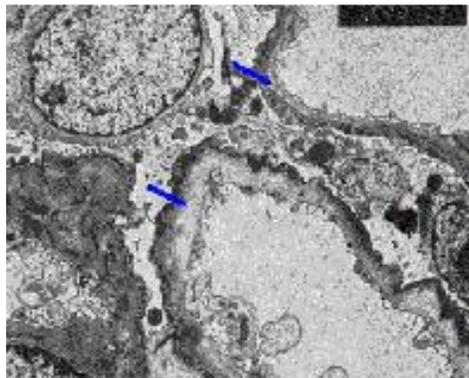
Carcinoid heart disease occurs in approximately 50% of patients with the carcinoid syndrome and usually heralds a worsening prognosis.

Palliation of symptoms and prolonged survival can be achieved with appropriate medical treatment and valvar surgery in selected patients with carcinoid heart disease.

64. Electron microscopy is diagnostic in :

- a) Goodpasture syndrome
- b) Alport syndrome
- c) Wegener's granulomatosis
- d) Churg Strauss syndrome

Electron microscopy reveals diffuse thickening and splitting of the basement membrane in 60-90% of patients. Diffuse thinning is observed in some patients with Alport syndrome. A normal ultrastructure of the GBM makes a diagnosis of Alport syndrome highly unlikely.



**Electron micrograph of kidney biopsy from a patient with Alport syndrome. Note the splitting and lamellation of the glomerular basement membrane (see arrows).**

65. A female presents with progressive breathlessness . X-ray shows patchy fibrosis .Histology shows increase fibroblastic foci . What is the most probable diagnosis :

- a) Cryptogenic organizing pneumonia
- b) Hemorrhagic edema
- c) Non-specific interstitial pneumonia
- d) Usual Interstitial Pneumonia (UIP)

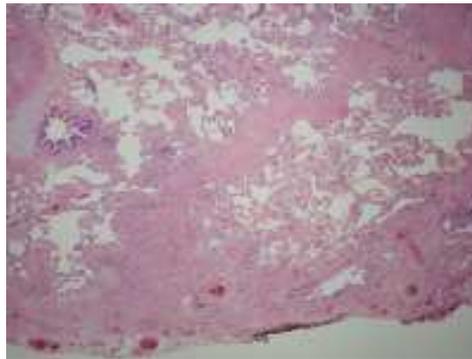
Most patients with idiopathic pulmonary fibrosis present with a gradual onset, often greater than 6 months, of dyspnea and/or a nonproductive cough. A chest radiograph typically reveals diffuse reticular

**opacities**; however, it lacks diagnostic specificity. High-resolution computed tomography (HRCT) findings are significantly more sensitive and specific for the diagnosis of idiopathic pulmonary fibrosis. On HRCT images, usual interstitial pneumonia is characterized by the presence of reticular opacities often associated with traction bronchiectasis. As idiopathic pulmonary fibrosis progresses, **honeycombing** becomes more prominent. Pulmonary function tests often reveal restrictive impairment and reduced diffusing capacity for carbon monoxide.

As mentioned above, **idiopathic pulmonary fibrosis is an idiopathic interstitial pneumonia characterized by usual interstitial pneumonia on histopathology**. The hallmark pathologic feature of usual interstitial pneumonia is a heterogeneous, variegated appearance with alternating areas of healthy lung, interstitial inflammation, **fibrosis**, and **honeycomb change**.

**Fibrosis predominates over inflammation** in usual interstitial pneumonia. **Fibroblastic foci** represent microscopic zones of acute lung injury and are randomly distributed within areas of interstitial collagen deposition and consist of fibroblasts and myofibroblasts arranged in a linear fashion within a pale-staining matrix. Although fibroblastic foci are not specific for usual interstitial pneumonia, they represent an important diagnostic criterion.

Another important diagnostic criterion for usual interstitial pneumonia is **honeycomb change**. Microscopically, honeycomb change is defined by **cystically dilated bronchioles lined by columnar respiratory epithelium in scarred, fibrotic lung tissue**. Dense eosinophilic collagen without associated honeycomb change signifies fibrotic scars and is also characteristic of usual interstitial pneumonia. Interstitial inflammation, consisting of patchy alveolar septal infiltrates of mononuclear cells, is not predominant in usual interstitial pneumonia.



**Patchwork distribution of abnormalities in a classic example of usual interstitial pneumonia (low-magnification photomicrograph; hematoxylin and eosin stain; original magnification, X4).**

# PHARMACOLOGY

66. Pancreatitis occurs with :

- a) Abacavir
- b) Zidovudine
- c) Lamivudine
- d) **Didanosine**

*Table II: Characteristic time duration and dosage range over which drug-induced pancreatitis occurs with some commonly implicated drugs. Although most cases have been reported within the listed time duration and dose range, cases outside of these ranges have been described.*

Drug	Duration (refs.)	Dose (refs.)
Acetaminophen	2–9 days (32, 33, 37, 38)	Toxic dose range (36)
Amino-salicylates	2 days to 1 month (39)	800–1,600 mg (40)
Atypical antipsychotics	Within 6 months (41)	Variable doses (41)
Azathioprine	13–21 days (17, 21)	50–150 mg/day (17)
Codeine	A few hours (42, 43)	40 mg (42, 43)
<b>Didanosine</b>	10–20 weeks (17)	>10 mg/kg (17)
Erythromycin	Within hours (19, 35, 36)	Toxic doses (19, 35, 36)
Estrogens	Within 3 months (17)	Variable doses
Furosemide	2–5 weeks (12, 36)	40–100 mg/day (17, 44, 45)
Interferon 2b	4–21 days (14)	3 million units three times weekly (14)
L-asparaginase	2 days after initiation to 10 weeks after discontinuation (46)	6–21 doses (47)
Mercaptopurine	7–32 days (48)	50–125 mg/day (48)
Pentamidine	6–11 days (49)	200–240 mg/day (49)
Sulfonamide	1–4 weeks (36, 50)	2–4 g/day (51)
Tetracycline	Within 3 weeks (17)	Toxic doses (17)
Thiazides	2–65 weeks (36, 44)	25–1,000 mg/day (44)
Valproate	3–12 months (52, 53)	>30 mg/kg (25, 53)

When didanosine (ddI) was approved by the FDA in 1991, a warning against ddI-associated pancreatitis was included in the labeling. That warning was based on experience in phase III trials with late-stage AIDS patients who took high doses of the drug.

67. Which is true regarding prebiotic fructans :

- a) Cannot be digested due to absence of enzymes in upper GIT
- b) **Because of  $\beta$ -conjugation of the anomeric C2 in their fructose monomers, they resist hydrolysis by intestinal degradative enzymes**
- c) Resistant to acidic medium of stomach
- d) They are dietary supplement containing potentially beneficial bacteria with lactic acid bacteria

## Prebiotics

Prebiotics promote the growth of probiotics gut micro flora, exert positive effect on digestive health and regularity, improve mineral absorption, enhance immune functions and promote overall health. Low in calories and suitable for diabetics. Prebiotics such as oligosaccharides are found naturally in certain fruits and vegetables including asparagus, bananas, chicory, garlic, onion, wheat and tomatoes

### *Effect on mineral absorption*

The nondigestible carbohydrates (dietary fiber) have been reported to impair the small-intestinal absorption of minerals because of their binding or sequestering action.

### *Fate in the gastrointestinal tract*

Because of the  $\beta$ -configuration of the anomeric C-2 in their fructose monomers, inulin-type fructans resist digestion in the upper part of the gastrointestinal tract. Moreover, there is evidence that they are not absorbed to any significant extent. Thus, it has been proposed that they be called a colonic food (ie, a food entering the colon and serving as a substrate for the endogenous bacteria, thereby directly providing the host with energy and metabolic substrates).

### *Effect on the metabolism of lipids*

The effects of inulin-type fructans on triglyceridemia have been studied in both animals and humans. In rats, a decrease in serum triglyceridemia (in both the fed and the fasted state) was consistently reported in several studies.

Feeding rats a diet supplemented with oligofructose (10% in the diet) significantly lowers serum triacylglycerol and phospholipid concentrations but does not modify free fatty acid concentrations in serum

### *Reducing disease risk*

For inulin-type fructans, claims that they reduce the risk of disease include the following:

- Constipation relief resulting from fecal bulking and possible effects on intestinal motility.
- Suppression of diarrhea, especially when associated with intestinal infections.
- Reduction of risk of osteoporosis if inulin-type fructans improve the bioavailability of calcium and if this functional effect is followed by a more physiologic change in peak bone density and mineral bone mass.
- Reduction of the risk of atherosclerotic cardiovascular disease associated with dyslipidemia, especially hypertriglyceridemia, and insulin resistance,

which in particular is known to be associated with hyperenergetic, high-carbohydrate feeding regimens. The reduction of risk via a hypocholesterolemic effect needs further investigation as does the proposal of a sound mechanistic hypothesis to be tested in humans.

- Reduction of the risk of **obesity** and possibly of **type 2 diabetes**, both of which are known to be associated with insulin resistance.

Cancer is a last area for further research on the ability of inulin and oligofructose to reduce risk of disease. In 2 studies, feeding rats with inulin significantly reduced the incidence of the so-called aberrant crypt foci induced by such colon carcinogens as azoxymethane and dimethylhydrazine.

<b><u>Nomenclature</u></b>	<b><u>Description</u></b>
Prebiotic	Prebiotics are <b>nondigestible food ingredients</b> that benefit the host by selectively stimulating the growth or activity of one or a limited number of bacteria in the colon. Modification by prebiotics of the composition of the colonic microflora leads to the predominance of a few of the potentially health-promoting bacteria, especially, but not exclusively, <b>lactobacilli</b> and <b>bifidobacteria</b> .
Probiotic	Probiotics are <b>microorganisms found naturally in foods</b> that stimulate the growth of beneficial organisms within the gastrointestinal(GI) tract. Probiotics are widely used to prepare fermented dairy products such as yogurt or freeze-dried cultures. In the future, they may also be found in fermented vegetables and meats. Several health-related effects associated with the intake of probiotics, including alleviation of lactose intolerance and immune enhancement, have been reported in human studies. Some evidence suggests a role for probiotics in reducing the risk of rotavirus-induced diarrhea and colon cancer. The probiotics bacteria most commonly used are Lactobacillus, <b>Bifidobacterium</b> , <b>Streptococcus</b> and <b>Enterococcus</b> species.
Symbiotic	The <b>combination of probiotics and prebiotics</b> is called as a synbiotics. This combination might improve the survival of the bacteria crossing the upper part of the gastrointestinal tract, thereby

	enhancing their effects in the large bowel. In addition, their effects might be additive or even synergistic.
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*Prebiotics & Probiotics are an important topic for exam. You can read more about them at <http://www.pharmainfo.net/reviews/probiotics-and-prebiotics-functional-food-promotion-health>*

68. True of Trientine is all except :

- a) **More potent than penicillamine & orally absorbed**
- b) Alternative to penicillamine in non-tolerant
- c) Not given with iron within 2 hrs of ingestion
- d) Causes Fe deficiency anemia which is reversible with oral iron supplement

Trientine is **less potent** than Penicillamine.

69. Ethosuximide is the drug of choice for :

- a) GTCS
- b) CPS
- c) **Absence seizures**
- d) Myoclonic seizures

70. A 14 year old adolescent girl was treated for acne with Tetracycline, Minocycline , Doxycycline and Azithromycin . Following the use of antibiotics , she noticed development of hyper – pigmentation over her nails. Which of the following drug was responsible for hyper – pigmentation ?

- a) Tetracycline
- b) **Minocycline**
- c) Doxycycline
- d) Azithromycin

Certain drugs, including the antibiotic **minocycline**, can cause **streaking of the nails** as a side-effect.

71. Proton pump inhibitor is :

- a) Ranitidine
- b) Misoprostol
- c) **Omeprazole**
- d) Loxatidine

72. Which of the following newer drug has activity on both HER1 and Her2neu receptors :

- a) Gefitinib

- b) Imatinib
- c) Lapatinib

## Molecularly Targeted Therapies for Breast Cancer

### Lapatinib (GW572016)

Lapatinib is a small molecule inhibitor of the TK domain of both HER1 and HER2. Preclinical studies have demonstrated inhibition of growth and induction of apoptosis in breast cancer cells lines driven by EGFR or HER2 expression through decreased phosphorylation of the TK domains of both receptors. Lapatinib has a significantly longer half-life than gefitinib or erlotinib. Burris et al reported a phase 1b trial of lapatinib in EGFR and/or HER2 expressing cancers. Sixty-six patients were treated, including 30 patients with metastatic breast cancer. Four of the patients with metastatic breast cancer (13%) had an objective response. All four had progressed through prior trastuzumab. Ten patients (33%) were believed to have stable disease at a median duration of therapy of 5 months. All 10 had EGFR expression by IHC, and 8 of these 10 over-expressed HER2.

Preliminary results of a phase II trial of lapatinib in trastuzumab-refractory HER2+ metastatic breast cancer have been reported. Of 41 patients evaluable at the time of this report, the objective response rate was assessed as 10%, though only a 5% response rate was confirmed by an independent review. The same independent review confirmed disease stabilization at 16 weeks in 25% of patients. Grade 3 toxicity was modest and consisted of diarrhea, rash, and fatigue in 10% or less of patients.

Based on the modest single-agent activity of this drug in patients with multiple prior trastuzumab-containing therapies, several phase III trials are planned or ongoing to study this drug in addition to capecitabine, the taxanes, and letrozole. These are mainly in the metastatic setting, but a new adjuvant (and neoadjuvant) trial is under development by the NCCTG and the North American Breast Intergroup incorporating this agent for HER2+ patients.

### Pertuzumab (2C4)

Pertuzumab is a well tolerated monoclonal antibody designed to target HER2 at an epitope distinct from trastuzumab. It blocks formation of HER2 heterodimers with other members of the HER family and thus reduces signaling through the multiple pathways associated with HER activation. A phase II study of this agent in metastatic breast cancer has completed accrual but has yet to be reported.

### Canertinib (CI-1033)

Canertinib is an orally available pan-HER TK inhibitor. In addition, its binding to the TK sites is irreversible, which may be an advantage compared to other TK inhibitors that bind reversibly. Canertinib has been well tolerated in a variety of phase I schedules with some disease stabilization in patients with refractory metastatic breast cancer. A phase II trial of this agent in metastatic breast cancer has completed accrual.

## HER1 (EGFR) Inhibition

Two main classes of agents have been developed that specifically target the EGFR. **Gefitinib** and **erlotinib** are small molecule inhibitors of the EGFR TK, and cetuximab is among a group of monoclonal antibodies that target EGFR. A phase II trial of erlotinib similarly demonstrated a low clinical benefit rate (less than 5%) in refractory patients.[22] These studies demonstrate some single-agent activity of the small molecule TK inhibitors in refractory metastatic breast cancer, but they are somewhat disappointing. A significant problem with anti-EGFR therapy is that a good predictive marker for benefit is not readily available to ensure that patients treated are those in whom the target is an important part of the tumor biology. No studies have been reported as yet with anti-EGFR monoclonal antibodies, and it should be noted that there has been little activity of the small molecule TK inhibitors in metastatic colon cancer, yet reproducible activity has been observed with monoclonal antibodies such as cetuximab, which may have multiple mechanisms of action beyond inhibition of the EGFR TK.

There are good preclinical and some clinical data regarding the synergy of anti-EGFR therapy and chemotherapy, and this strategy is being pursued in breast cancer. The NCCTG has completed a trial of **gemcitabine and erlotinib** and will soon open a trial of **irinotecan and cetuximab** in the setting of refractory metastatic breast cancer. Other research strategies ongoing with these agents include combinations with other targeted therapies, including endocrine therapy.

Table 2. — Agents Targeting HER Signaling

Drug	Target				Tyrosine Kinase Inhibitor	Monoclonal Antibody
	HER1	HER2	HER3	HER4		
Trastuzumab	-	+	-	-	-	+
Gefitinib, erlotinib	+	-	-	-	+	-
Cetuximab	+	-	-	-	-	+
Lapatinib	+	+	-	-	+	-
Pertuzumab*	+	+	+	+	-	+
CI-1033	+	+	+	+	+	-

\* Binds to HER2 but inhibits HER2 dimerization with HER1, HER3, HER4.

73. Fetal hydantoin syndrome is caused by :

- Phenytoin
- Alcohol
- Tetracycline
- Valproate

**Fetal Hydantoin Syndrome** is a rare disorder that is caused by exposure of a fetus to the anticonvulsant drug **phenytoin** (Dilantin). The symptoms of this disorder may include abnormalities of the skull and facial features, growth deficiencies, underdeveloped nails of the

fingers and toes, and/or mild developmental delays. Other findings occasionally associated with this syndrome include cleft lip and palate, having an unusually small head (microcephaly) and brain malformations with more significant developmental delays.

### Signs

1. Microcephaly
2. Growth delay
3. Mental retardation and developmental delay
4. Craniofacial dysmorphic features
  1. Flat nasal bridge
  2. Epicanthic folds
  3. Prominent upper lip over wide mouth
5. Distinguishing characteristics
  1. Nail and distal phalanx hypoplasia
  2. Hypertelorism



74. Which is incorrectly matched :
- a) Phenytoin – cleft lip & palate
  - b) Zidovudine – skull defect
  - c) Valproate – NTD
  - d) Warfarin – nasal bone dysplasia

Zidovudine is not teratogenic, and so is used to prevent mother to child transmission of HIV infection.

75. Tyrosine kinase inhibitors are used in :

- a) **GIST**
- b) Receptor mediated endocrine tumor
- c) Neurofibromatosis
- d) AML

76. A 10 year old boy presented with unilateral throbbing headache which was relieved by sleep . Which of the following drug cannot be used for prophylaxis ?

- a) **Sumatriptan**
- b) Verapamil
- c) Amitriptyline
- d) Verapamil

77. Allopurinol is used in the treatment of :

- a) Osteoarthritis
- b) **Gout**
- c) Rheumatic arthritis
- d) Ankylosing spondylitis

78. The drug used to treat Chlamydia infection in pregnancy :

- a) Tetracycline
- b) Doxycycline
- c) **Erythromycin**
- d) Penicillin

79. True about Penicillin binding protein are all except :

- a) It is mechanism of resistance for only gram +ve organism
- b) PBP is essential for cell wall synthesis
- c) **PBP is a constituent of cell wall**
- d) PBP acts as carboxypeptidases & transpeptidases

PBP is a constituent of **cell membrane** (not cell wall).

80. Amphotericin B toxicity can be lowered by :

- a) Giving it with glucose
- b) ↓ the dose
- c) **Using liposomal delivery system**
- d) Combining with Flucytosine

81. About thalidomide not true is :

- a) Reintroduced for its activity in ENL
- b) Developed as an antiemetic in pregnancy but withdrawn because of phocomelia

- c) Used for new & relapsed cases of multiple myeloma
- d) MC S/E is diarrhea & euporia

MC S/E of Thalidomide – Sedation & constipation

Most serious S/E of Thalidomide – Sensory neuropathy

Most important S/E of Thalidomide – Phocomelia

82. All are true about Phenytoin except :

- a) Does not ↓insulin release
- b) Follow saturation kinetics
- c) No CNS S/E at therapeutic dose
- d) Teratogenic

Phenytoin ↓insulin secretion → Hyperglycemia & glycosuria

83. Mechanism of action of Theophylline in bronchial asthma is :

- a) Phosphodiesterase inhibition
- b) ↑calcium from sarcoplasmic reticulum
- c) Adenosine receptor inhibition
- d) β2 agonist

84. MAO inhibitor is not used with :

- a) Pethidine
- b) Pentazocine
- c) Buprenorphine
- d) Morphine

MAO inhibitor if given to a patient taking Meperidine congeners (Pethidine, Demerol) → It will cause Serotonin syndrome.

85. Which of the following drug is given i.v. for the treatment of HIT

- a) Abciximab
- b) Lepirudin
- c) Plasminogen
- d) Alteplase

Lepirudin is a Direct Thrombin Inhibitor (DTI) given IV in pts with HIT.

86. Regarding Mycophenolate mofetil incorrect is :

- a) MC S/E is nephropathy
- b) Used in transplant rejection
- c) It is a prodrug & converted to Mycophenolic acid

d) Can't be used with Azathioprine

MC S/E are – GI (Diarrhoea, vomiting) & Hematologic (Leucopenia).

87. Urokinase and streptokinase are contraindicated in :

- a) Malignancy
- b) A-V fistula
- c) PE
- d) Thrombophlebitis

**Intracranial neoplasm** (malignancy) is an absolute C/I for these agents.

88. Drug not given in myoclonic epilepsy is :

- a) Topiramate
- b) Zonisamide
- c) Carbamazepine
- d) Valproate

Juvenile myoclonic epilepsy can be aggravated by Phenytoin or Carbamazepine.

89. Along with Rivastigmine for Alzheimer disease which drug cannot be given ?

- a) SSRI
- b) RIMA
- c) TCA
- d) Atypical antidepressant

Coadministration of cholinesterase inhibitors (e.g. Rivastigmine) & drugs having anti-cholinergic activity (e.g. TCA) is counter-productive.

90. Serotonin syndrome is caused by all except :

- a) Chlorpromazine
- b) Pentazocine
- c) Buspirone
- d) Meperidine

91. Hirsutism is seen with all of the following drugs except

- a) Danazol
- b) Phenytoin
- c) Norethisterone
- d) Flutamide

92. Which statement is false :

- a) Naloxone can be given orally, so used in Tt of constipation in opium addicts
- b) Naltrexone given orally in alcohol craving due to long  $t_{1/2}$
- c) Nalmefene has got less  $t_{1/2}$  than naloxone
- d) **Naltrexone acts faster than naloxone**

**Naloxone acts much faster than Naltrexone**, that's why Naloxone is used in opioid toxicity.

93. When linezolid is used for the treatment which of the following system is monitored :

- a) Renal
- b) GIT
- c) Audiometry
- d) **Platelet count**

94. Narrow therapeutic index is seen with

- a) Desipramine
- b) **Lithium**
- c) Penicillin
- d) Diazepam

95. MC congenital anomaly associated with lithium :

- a) **Cardiac malformation**
- b) NTD
- c) Renal anomaly
- d) Fetal hydantoin syndrome

Lithium causes **Ebstein's anomaly**.

96. Function of carbidopa in the treatment of parkinsonism

- a) **Inhibits peripheral conversion of levodopa**
- b) ↓ efficacy of levodopa
- c) ↑ dose of levodopa
- d) ↑ nausea

97. In equivalent concentration , corticosteroids are potent in which form :

- a) Gel
- b) Cream
- c) **Ointment**
- d) Lotion

**Ointment** → **Occlusive nature** → ↑ drug penetration → So more potent

# MICROBIOLOGY

98. Varicella Zoster remain latent in

- a) Trigeminal ganglion
- b) Microglia
- c) T cell
- d) B cell

99. Following causes biliary obstruction

- a) Giardia
- b) Strongyloides
- c) Ankylostoma
- d) Clonorchis

100. A 2 month child presented with sepsis . Bacteria isolated showed beta hemolysis on blood agar . Organism on culture is showing resistance to bacitracin . CAMP test is positive . The most probable Organism causing the condition is :

- a) S. pyogenes
- b) S. agalactiae
- c) Enterococcus
- d) Pneumococcus

Features are suggestive of Gr B streptococcal infection. They are  $\beta$ -hemolytic & are CAMP +ve.

101. A young woman complains of recurrent rhinitis , nasal discharge and bilateral nasal blockage since one year . She also had history of allergy and asthma . On examination , multiple polyps with mucosal thickening and impacted secretions are seen in nasal cavities . Biopsy was taken and the material on culture showed many hyphae with dichotomous branching typically at 45 degree . Which of the following is most likely organism responsible ?

- a) Rhizopus
- b) Aspergillus
- c) Mucor
- d) Candida

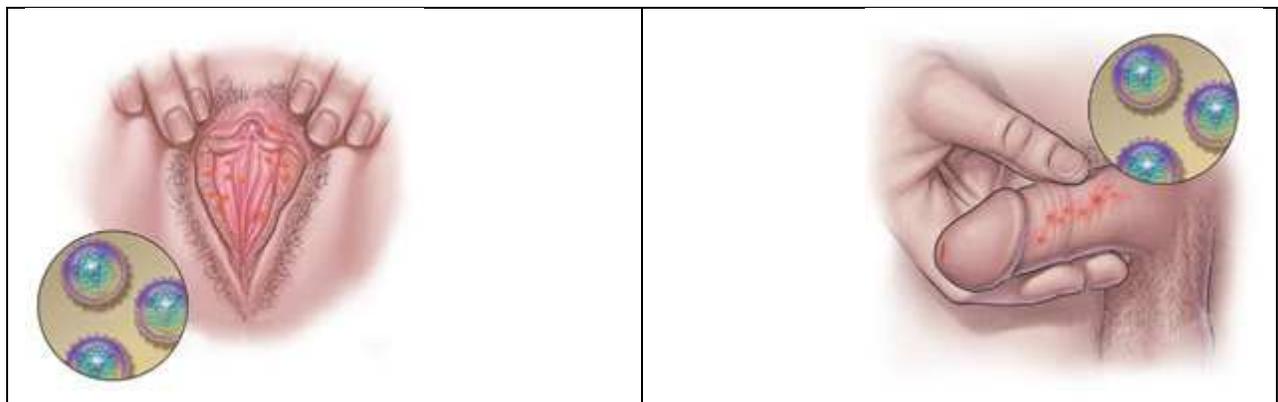
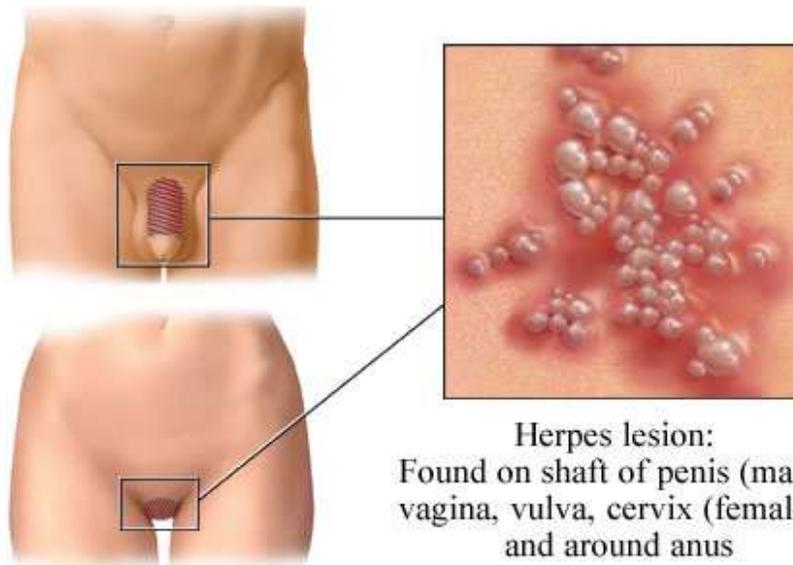


Mycelium with septate fungi branching at 45 degrees is characteristic of Aspergillus.

102. Most common cause of genital lesions in HIV patient is :

- a) Chlamydia
- b) Herpes
- c) Syphilis
- d) Candida

People with HIV-1 are commonly coinfecting with herpes simplex virus 2 (HSV-2), the usually cause of genital herpes, the investigators noted as background. HSV-2 is often reactivated as immune function declines, and prior research indicates that active herpes is associated with increased plasma and genital levels of HIV.



103. True statements about Staph aureus are all except :

- a) 30% are healthy carrier
- b) Methicillin resistance is chromosomally mediated

- c) Epidermolysin for TSS is a superantigen
- d) MC cause of infn is cross infection

Most individuals who develop Staph aureus infection are infected by their own colonizing strains.

104. Isolation of chlamydiae from tissue specimens can be done by :

- a) ELISA
- b) Yolk sac
- c) Direct immunofluorescence
- d) Amniotic cavity

The yolk sac inoculation procedure and several tissue culture procedures currently are used for chlamydial isolation. The only known host system that supports growth of all Chlamydia is the yolk sac of embryonated hen's egg.

105. A patient presents with dry cough for 2 weeks . On examination , there is seen a white patch on tongue .The probable diagnosis is :

- a) Invasive candidiasis
- b) Histoplasmosis
- c) Lichen planus
- d) Aspergillosis

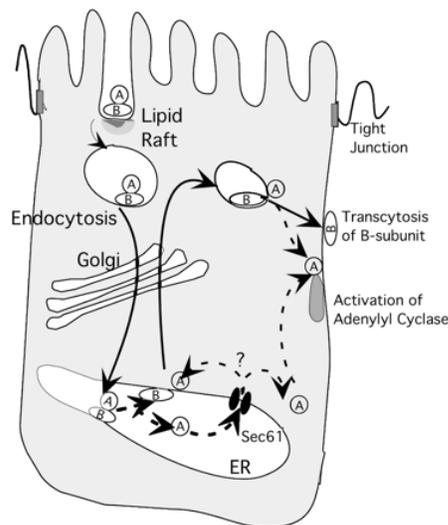
**Oral candidiasis** (also known as "thrush") is an infection of yeast fungi of the genus *Candida* on the mucous membranes of the mouth. It is frequently caused by *Candida albicans*, or less commonly by *Candida glabrata* or *Candida tropicalis*. Oral infections by *Candida* species usually appear as thick white or cream-colored deposits on mucosal membranes. The infected mucosa of the mouth may appear inflamed (red and possibly slightly raised). In babies the condition is termed thrush. Adults may experience discomfort or burning. When the cream-colored deposits are scraped, there is slight bleeding.



106. True about El Tor vibrio is all except :

- a) Acts on sites other than intestinal epithelial cells
- b) Humans are only reservoir
- c) Can survive in ice cold water for 2-4 weeks
- d) Killed by boiling for 30 secs

Cholera toxin (CT) produced by *Vibrio cholerae* is the virulence factor responsible for the massive secretory diarrhea seen in Asiatic cholera. To cause disease, CT enters the intestinal epithelial cell as a stably folded protein by co-opting a lipid-based membrane receptor, ganglioside  $G_{M1}$ .  $G_{M1}$  sorts the toxin into lipid rafts and a retrograde trafficking pathway to the endoplasmic reticulum, where the toxin unfolds and transfers its enzymatic subunit to the cytosol, probably by dislocation through the translocon  $sec61p$ . The molecular determinants that drive entry of CT into this pathway are encoded entirely within the structure of the protein toxin itself.



107. A diabetic patient with orbital swelling with pain and bloody nasal discharge . Culture of periorbital pus showed branching septate hyphae . Which is the most probable organism involved ?

- a) Fusarium
- b) Conidiobolus
- c) Aspergillus
- d) Rhizopus

Uncontrolled diabetes is a risk factor for aspergillosis. Aspergillus hyphae are hyaline, narrow & septate.

## FORENSIC

108. A factory worker presented with excessive salivation, blue line on gums, tremors, insomnia, loss of appetite, constipation and disturbed personality . The most probable type of poisoning is :
- Mercury
  - Lead
  - Arsenic
  - Phosphorus

### Mercury poisoning

- Chronic and intense acute exposure causes cutaneous and neurological symptoms. The classic triad found in chronic toxicity is tremors, gingivitis, and erethism (ie, a constellation of neuropsychiatric findings that includes insomnia, shyness, memory loss, emotional instability, depression, anorexia, vasomotor disturbance, uncontrolled perspiration, and blushing).
- Additional findings may include headache, visual disturbance (eg, tunnel vision), peripheral neuropathy, salivation, insomnia, and ataxia.

109. Aconite poisoning causes all except :
- Hypersalivation
  - Tingling & numbness
  - ↑BP
  - Chest pain

Marked symptoms may appear almost immediately, usually not later than one hour, and "with large doses death is almost instantaneous." Death usually occurs within 2 to 6 hours in fatal poisoning (20 to 40 mL of tincture may prove fatal). The initial signs are gastrointestinal including nausea, vomiting, and diarrhea. There is followed by a sensation of burning, tingling, and numbness in the mouth and face, and of burning in the abdomen. In severe poisonings pronounced motor weakness occurs and cutaneous sensations of tingling and numbness spread to the limbs. Cardiovascular features include hypotension, bradycardia, sinus tachycardia, and ventricular arrhythmias. Other features may include sweating, dizziness, difficulty in breathing, headache, and confusion. The main causes of death are ventricular arrhythmias and asystole, paralysis of the heart or of the respiratory center. The only post-mortem signs are those of asphyxia.

Treatment of poisoning is mainly supportive. All patients require close monitoring of blood pressure and cardiac rhythm. Gastrointestinal decontamination with activated charcoal can be used if given within 1 hour of ingestion. The major physiological antidote is atropine, which is used to treat bradycardia. Other drugs used for ventricular arrhythmia include lidocaine, amiodarone, bretylium, flecainide, procainamide, and mexiletine. Cardiopulmonary bypass is used if symptoms are refractory to treatment with these drugs. Successful use of charcoal hemoperfusion has been claimed in patients with severe aconite poisoning.

110. A woman died within 5 years of her marriage under suspicious circumstances. Her parent complained that her –in-laws used to have frequent demands of dowry . Under which Act , her autopsy will be done ?
- IPC 301
  - IPC 304
  - CrPC 174
  - CrPC 176

**CrPC Section 176. Inquiry by Magistrate into cause of death.**

(1) When any person dies while in the custody of the police or when the case is of the nature referred to in clause (i) or clause (ii) of sub-section (3) of section 174, the nearest Magistrate empowered to hold inquests shall, and in any other case mentioned in sub-section (1) of section 174, any Magistrate so empowered may hold an inquiry into the cause of death either instead of, or in addition to, the investigation held by the police officer; and if he does so, he shall have all the powers in conducting it which he would have in holding an inquiry into an offence.

(2) The Magistrate holding such inquiry shall record the evidence taken by him in connection therewith in any manner hereinafter prescribed according to the circumstances of the case.

(3) Whenever such Magistrate considers it expedient to make an examination of the dead body of any person who has been already interred, in order to discover the causes of his death, the Magistrate may cause the body to be disinterred and examined.

(4) Where an inquiry is to be held under this section, the Magistrate shall, wherever practicable, inform the relatives of the deceased whose names and addresses are known, and shall allow them to remain present at the inquiry.

111. Lightening flash can cause injury due to all except :
- Direct impact due to electric current
  - Burns due to superheated light
  - Compression of air in front of travelling wave
  - Expanded air around flash



A **compression wave** or shock wave is best described as a pressure wave. As the expansion takes place around the lightning bolt, (or as scientists call it the "channel" that the electric charge followed) **compression takes place just beyond the zone of expansion** and the pressure wave moves outward, away from the lightning bolt at the speed of sound.

112. Which of the following is commonly used in Narco – Analysis ?

- a) Atropine sulphate
- b) Opium compounds
- c) Phenobarbitone
- d) Scopolamine hydrobromide

In 1921, R. E. House, an obstetrician in Texas, observed in deliveries in which the mother had been given scopolamine that in a certain stage of anesthesia or sedation she might be talkative and reveal things she would not ordinarily discuss. He noted that after childbirth, the mother frequently forgot that she had suffered pain, that she had complained of it, and that she had spoken of personal matters. After the use of scopolamine, often with the addition of chloroform, had proved to have certain advantages in the obstetrical management of a woman delivering a baby, House persuaded himself to extend the use of scopolamine beyond its original purpose to the interrogation of criminal suspects. He gave many enthusiastic demonstrations throughout the United States. As a result, newspapers quickly applied the term "truth serum" to this sedative drug. House's enthusiasm about scopolamine as an adjunct in obstetrics led him to overenthusiastic statements about the value of the drug in interrogation. In 1931, on the basis of two cases, he stated that a person under scopolamine could not lie and that the drug could distinguish the innocent from the guilty.

Sedatives or hypnotics that alter higher cognitive function include ethanol, scopolamine, 3-quinuclidinyl benzilate, temazepam, and various barbiturates including sodium thiopental (commonly known as sodium pentathol) and sodium amytal (amobarbital).

India's Central Bureau of Investigation has also used intravenous barbiturates for interrogation. For many criminal cases the police use narco test which violates right against self incrimination. On May 5, 2010 the Supreme Court of India held that narco, polygraph and brain mapping test violate article 20(3) of the Constitution.

113. Primary impact injuries are seen on :

- a) Chest
- b) Abdomen
- c) Legs
- d) Head

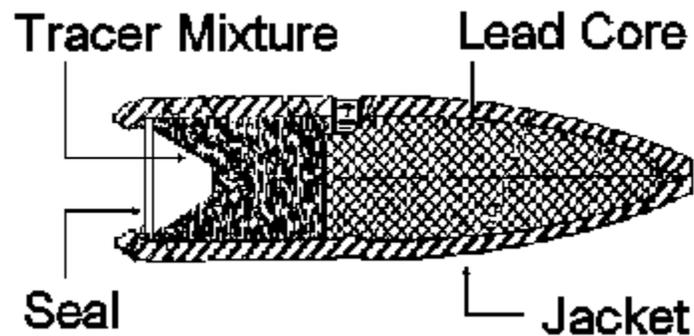


Primary impact is when a vehicle hits a pedestrian. This will injure the legs most commonly as when a car hits a person standing. Secondary injuries are when the person hits the ground or smashes into the windshield. Head injury is commonest secondary injury. Other parts of the body can have primary injuries if the vehicle is higher such as a truck or a bus.

114. Which bullet leaves a visible mark in its pathway so that a person can see it ?
- a) Tandem
  - b) **Tracer**
  - c) Dumdum
  - d) Incendiary

**Tracer ammunition (tracers)** are special bullets that are modified to accept a **small pyrotechnic charge in their base**. Ignited upon firing, the composition burns very brightly, **making the projectile visible to the naked eye**. This enables the shooter to follow the bullet trajectory relative to the target in order to make corrections to his or her aim.

When used, US tracers are usually loaded as every fifth round in machine gun belts, referred to as four-to-one tracer. Platoon and squad leaders will sometimes load their magazines entirely with tracers to mark targets for their soldiers to fire on. Tracers are also sometimes placed two or three rounds from the bottom of magazines to alert the shooter that his or her weapon is almost empty.



# ENT

115. Vestibular schwannoma arises from :

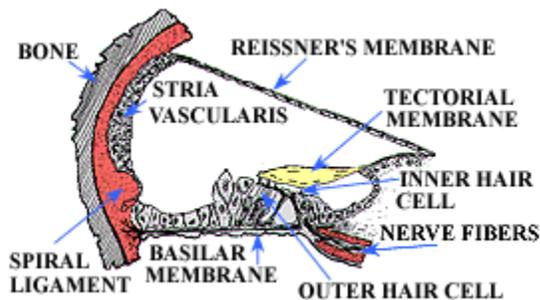
- a) Superior vestibular nerve
- b) **Inferior vestibular nerve**
- c) Facial nerve
- d) Abducent nerve

Schwannomas arise most commonly from the vestibular nerve (80%), occasionally from the cochlear (5 to 7%). The **inferior vestibular nerve is involved in 70%**, superior vestibular in 20% and cochlear nerve in 10%. The origin of the tumor is from junctional (Obersteiner Redlich) zone where the central and peripheral myelin meet. This zone is situated at the region of IAM or within the internal auditory canal.

116. Otoacoustic emissions arises from :

- a) Inner hair cells
- b) **Outer hair cells**
- c) Organ of Corti
- d) Both outer & inner hair cells

OAE or otoacoustic emission testing is the recording of sounds that the ear produces itself. Otoacoustic emissions were first reported by Kemp in 1978. They appear to be generated by motile elements in the cochlear **outer hair cells**. The mechanism underlying outer hair cell electromotility is thought to be the origin of spontaneous otoacoustic emissions.



117. Not seen on bronchoscopy is :

- a) Trachea
- b) Vocal cords
- c) **Subcarinal lymph nodes**
- d) 1<sup>st</sup> segmental division of bronchioles

118. All are true about Nasopharyngeal carcinoma except :

- a) Bimodal age distribution

- b) Nasopharyngectomy with B/L neck dissection is the primary Tt
- c) IgA antibody to EBV present
- d) SCC is the MC histological finding

Radiotherapy is the TOC.

119. True about Sodium Fluoride in otosclerosis is all except :

- a) Acts by inhibiting proteolytic enzymes at cochlea
- b) Inhibits osteoblast activity
- c) +ve schwartz sign is an indication
- d) NaF C/I in chronic nephritis

NaF reduces osteoclastic bone resorption & ↑osteoblastic bone formation.

120. Laser used in laryngeal operation is :

- a) NdYag
- b) CO<sub>2</sub>
- c) Argon
- d) Krypton

At present the CO<sub>2</sub>-laser is employed in almost every surgical specialism. If it is right to prefer laser treatment for special operations to conventional surgical methods, is not yet decided. However, the use of laser beam energy for laryngeal conditions offers undeniable advantages as: -- No direct instrumental contact. Involuntary movements which can occur when using long-reach instruments during conventional microlaryngeal surgery do not play a role. -- Little bleeding. The treating area stays all the time under good visual control. --Outmost precision by which an intended operation is to perform. Exact on the desired place with an adjust quantity of energy tissue can be vaporized as well with very limited amount as extensive, or be removed by cutting round. -- No postoperative edema. No or hardly any edema means that tracheotomy often needed for some conditions in another way of treatment usually is unnecessary.

121. Drug of Choice in Laryngeal Stenosis is :

- a) Cyclophosphamide
- b) Doxorubicin
- c) Adriamycin
- d) Mitomycin C

Topical application of Mitomycin C, a potent fibroblast inhibitor reduces granulation tissue formation and prevents recurrence.

122. A child presented with stridor, barking cough and difficulty in breathing since 2-3 days,. He also had high grade fever with increased leukocyte count. All of the following are true except ?

- a) On X-ray, subglottic stenosis & hypopharyngeal dilatation are seen

- b) Boys more affected than females
- c) Antibiotics are mainstay of Tt
- d) Involve subglottis MC

Croup is caused by Parainfluenza virus. Antibiotics are of no use.

123. All are extrinsic laryngeal membranes except

- a) Hyoepiglottic
- b) Cricothyroid
- c) Cricotracheal
- d) Cricotracheal

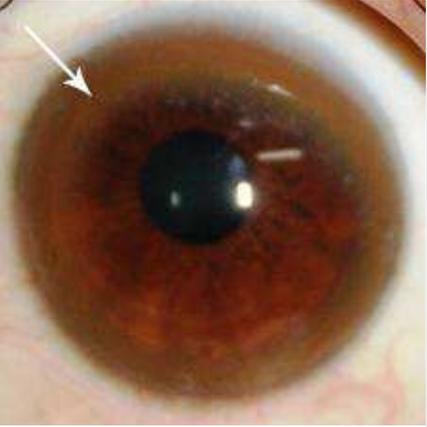
**Intrinsic ligaments & membranes**

- Quadrangular membrane
- Cricothyroid ligament
- Cricovocal membrane

# OPHTHALMOLOGY

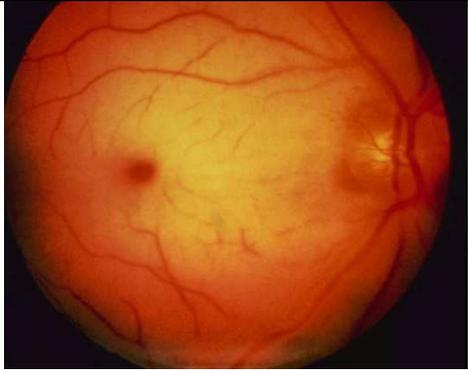
124. Kayser-Fleischer rings (KF rings ) are seen in

- Pterygium
- Hemochromatosis
- Wilson's disease**
- Traumatic cataract

<p style="text-align: center;"><b>Wilson's Disease</b></p> <ul style="list-style-type: none"> <li>• Autosomal recessive</li> <li>❖ Inadequate hepatic excretion of copper           <ul style="list-style-type: none"> <li>– Excess copper accumulates in liver and brain due to low concentration of copper binding enzyme ceruloplasmin in the serum</li> </ul> </li> <li>• In addition to cirrhosis it causes:           <ul style="list-style-type: none"> <li>• Degenerative changes in the lenticular nuclei of the brain               <ul style="list-style-type: none"> <li>• Psychiatric disorders</li> <li>• Movement disorders</li> <li>• Abnormal eye movements</li> </ul> </li> <li>❖ Kayser-Fleischer rings               <ul style="list-style-type: none"> <li>• Pathognomonic</li> <li>• Deposition of copper in the corneal limbus</li> </ul> </li> </ul> </li> </ul> <p style="text-align: right; font-size: small;">78 (c) 2007, Michael A. Kahn, DDS/Lynn W. Solomon, DDS</p>	
	<b>Kayser-Fleischer ring</b>

125. Cherry red spot is seen in all except

- Niemann Pick disease
- GM1 gangliosidosis
- Tay Sachs disease
- Gaucher's disease**

	<p><b>Cherry red spot in CRAO</b></p>	<p>Cherry red spots on retina seen in:</p> <ul style="list-style-type: none"> <li>• CRAO</li> <li>• Tay Sachs ds</li> <li>• GM1 gangliosidosis</li> <li>• Quinine amblyopia</li> <li>• Metachromatic leukodystrophy</li> <li>• Multiple sulfatase deficiency</li> <li>• Berlins ds</li> <li>• Sanhoffs ds</li> <li>• Niemann Pick ds</li> </ul>
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126. Arden index is related to

- a) ERG
- b) EOG
- c) VER
- d) Perimetry

### **THE ELECTRO OCULOGRAM**

The test records the slow, large change in the ocular resting potential which occurs as the retina passes from the light adapted to the dark adapted state.

The amplitude of the resting potential doubles in amplitude as the eye "light adapts". This is considered to be a function of the metabolism of the pigment epithelium influenced by light induced changes in the receptors.

The ratio of the light adapted potential to the dark adapted potential, expressed as a percentage, is the **ARDEN INDEX**. In a normal eye, this ratio is greater than 2.1. The Index is therefore greater than 200%.

Arden index =  $\frac{\text{light peak}}{\text{dark trough}} \times 100\%$

Normal > 200%

Equivocal 175 – 200%

Subnormal < 175%

Extinguished = 100%

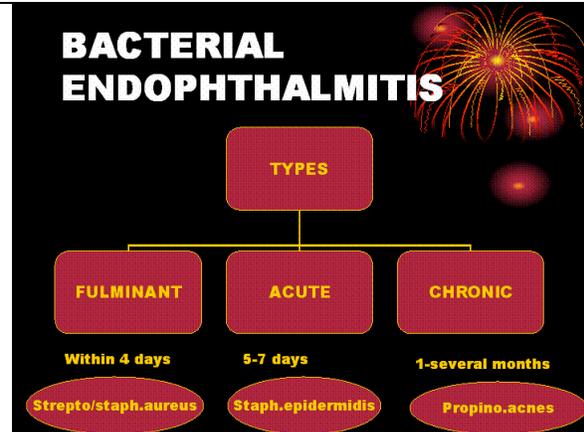
It is reduced in disorders of the pigment epithelium or the receptors – particularly the rods.

127. Endoophthalmitis include all except

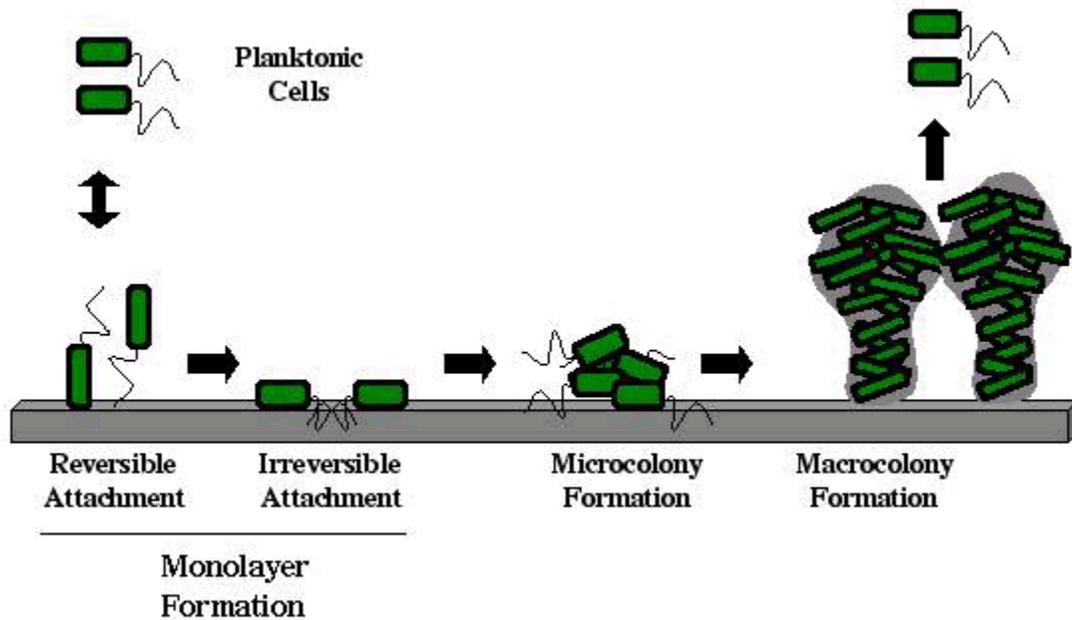
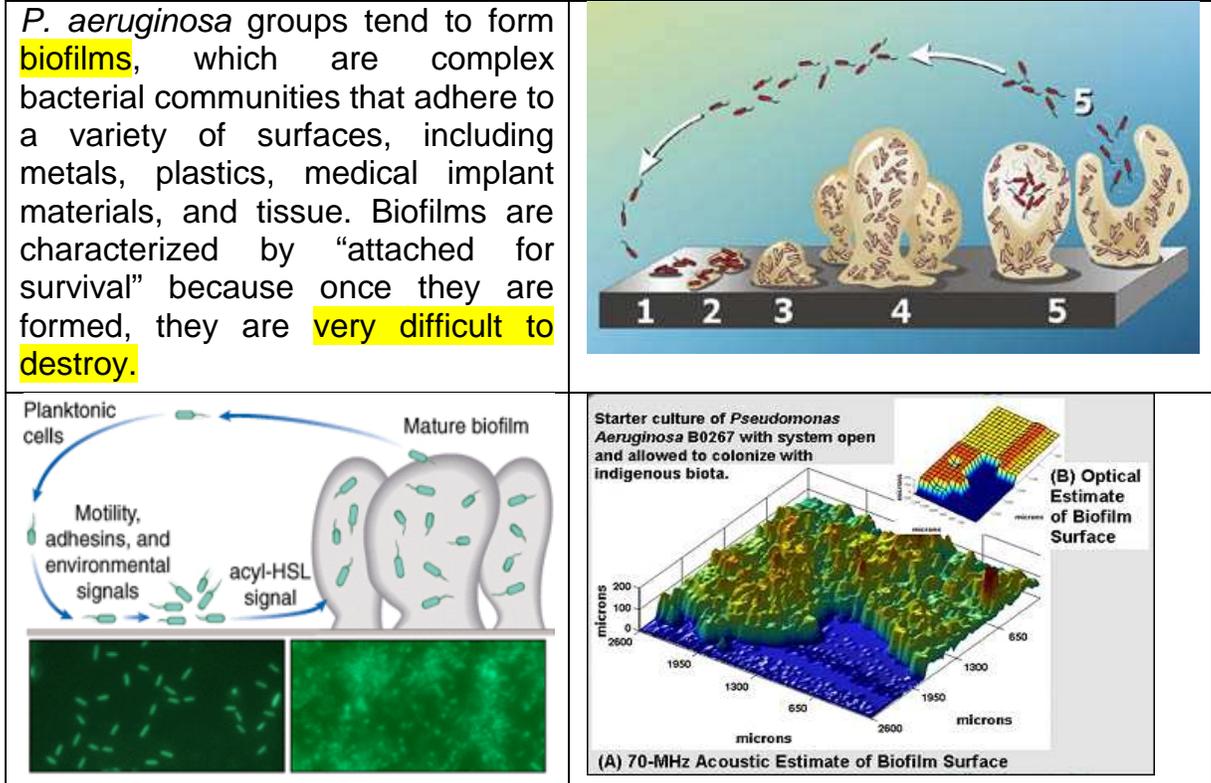
- a) Vitreous
- b) Sclera
- c) Uvea
- d) Retina

Endophthalmitis refers to intraocular inflammation predominantly involving the anterior chamber and the vitreous cavity of the eye. The term is usually associated with an infectious process or noninfectious stimulus such as retained lens material or toxic substance introduced into the eye during trauma or intraocular surgery.

*Ref: Medical Treatment of Endophthalmitis: Dr B. Vidyashankar, Dr Shweta Arora, Dr Rajat Singal, Dr Shahnawaz K, Dr Shanta S. Motwane, Journal of the Bombay Ophthalmological Association, Apr-Jun 2001*



128. A person who has been wearing contact lenses for 1-2 months comes with a H/O of irritation in left eye. On examination diagnosis of keratitis was made. Corneal scraping revealed presence of pseudomonas aeruginosa which was multidrug resistant. How did it attain the resistance?
- Transfer of resistance genes from normal conjunctival flora
  - Frequent instillation of antibiotics
  - Improper contact lens hygiene
  - Biofilm formation**



129. Regarding corneal transplant what is true
- a) Whole eye is preserved in tissue culture
  - b) Donor not accepted if age >60 yrs
  - c) **Specular microscopy analysis used to count the number of corneal endothelial cell**
  - d) HLA matching reqd

Since the **endothelial cells** are one of the most important structures in a donor cornea, their morphology and concentration must be carefully evaluated. This can be done at the slit lamp by a technique called specular reflection or with a **specular microscope**. Specular reflection refers to the viewing of objects that occurs when light is reflected from the interfaces of materials with different indices of refraction. This occurs in a mirror like fashion where the angle of incidence is equal to the angle of reflection. An endothelial cell is different in refractive index than the aqueous and also Descemet's. While most of the light goes through these two transparent layers, about 0.02% is reflected backward to form an image of these structures. The more regular and numerous the cells are, the better their function is thought to be. Tightly packed, hexagonal-shaped cells with little variation in shape and size are considered normal. Cell densities greater than 2000/mm are generally accepted as suitable for transplantation.



**Specular microscope**

130. A patient presenting with ptosis shows retraction on lids on chewing. What could be the possible cause ?
- Abducens paralysis
  - 3<sup>rd</sup> nerve palsy
  - Marcus Gunn phenomenon**
  - Aberrant regeneration of 3<sup>rd</sup> nerve

In 1883, Marcus Gunn described a 15-year-old girl with a peculiar type of **congenital ptosis** that included an **associated winking motion of the affected eyelid on the movement of the jaw**. This synkinetic jaw-winking phenomenon now bears his name.

Patients with Marcus Gunn jaw-winking syndrome have variable degrees of blepharoptosis in the resting, primary position. Although Marcus Gunn jaw-winking syndrome is **usually unilateral**, it can present bilaterally in rare cases.

The wink reflex consists of a momentary **upper eyelid retraction** or elevation to an equal or higher level than the normal fellow eyelid upon stimulation of the ipsilateral pterygoid muscle. This response is followed by a rapid return to a lower position. The amplitude of the wink tends to be worse in downgaze. This rapid, abnormal motion of the eyelid can be the most disturbing aspect of the jaw-winking syndrome.

The wink phenomenon may be elicited by opening the mouth, thrusting the jaw to the contralateral side, jaw protrusion, chewing, smiling, or sucking. This wink phenomenon is often discovered early, as the infant is bottle-feeding or breastfeeding.

## Pathophysiology

Marcus Gunn jaw-winking is thought to be a form of **synkinetic ptosis**. An **aberrant connection** appears to exist between the **motor branches of the trigeminal nerve (CN V3)** innervating the external pterygoid muscle and the fibers of the **superior division of the oculomotor nerve (CN III)** that innervate the levator superioris muscle of the upper eyelid.

Electromyographic studies demonstrate this synkinetic innervation by showing simultaneous contraction of the external pterygoid and levator muscle. In rare cases, synkinesis may be present between the internal pterygoid and levator muscles. In these cases, the eyelid elevates on closing the mouth and clenching the teeth.

A few authors have speculated that the jaw-winking is not due to a new aberrant pathway, but rather the disinhibition of preexisting phylogenetically more primitive mechanisms. This is thought to explain why individuals who are not affected will often open their mouths while attempting to widely open their eyes to place eye drops or to apply makeup.

Since jaw-winking ptosis is believed by most to be due to **abnormal innervation of the levator muscle and not secondary to myopathic changes**, it is not surprising that **most histopathologic studies have revealed normal striated muscle**.

One study found variable degrees of fibrosis within the affected levator muscle and to a lesser degree in the muscle of the normal, nonptotic eyelid.

131. Least common corneal dystrophy is

- a) **Macular dystrophy**
- b) Lattice type 1
- c) Lattice type 2
- d) Granular dystrophy

Macular dystrophy is an **autosomal recessive** condition, which is the **least common but the most severe** of the 3 major stromal corneal dystrophies. It is characterized by **multiple, gray-white opacities** that are present in the corneal stroma and that extend out into the peripheral cornea.



132. Relative afferent pupillary defect is seen in damage to
- Optic nerve
  - Optic tract
  - Visual cortex
  - Oculomotor nerve

### **General Points about the Relative Afferent Pupillary Defect**

- An RAPD generally occurs with **significant optic nerve or retinal disease**, when there is a difference in the disease process between the two eyes. If each eye has severe but equal disease, there will be no RAPD. Thus, a "bilateral" RAPD does not exist.
- Severe disease in one eye leading to an RAPD will not lead to anisocoria. The diseased eye's pupil will appear to be of equal size to the other eye due to the consensual light reaction (unless the iris itself is diseased or unreactive).
- Because of the consensual light reaction, only one functioning pupil is needed to determine the presence of an RAPD.
- The visual acuity does not necessarily correlate with an RAPD. Some conditions will lead to a marked reduction of visual acuity with an RAPD, while others spare the central vision. Often an extensive loss of peripheral vision correlates with an RAPD.

The "**swinging flashlight test**" is probably the best test for identifying an RAPD (as shown by Enyedi, L.B. et al in Ophthalmology 5/1998; 105:871-873, A Comparison of the Marcus Gunn and Alternating Light Tests for Afferent Pupillary Defects.) In this test, a strong, steady light is used. The light is shined into one eye, and then quickly switched to the other. This is repeated back and forth, until one of four conclusions is reached (listed below). Since light in one pupil causes both pupils to constrict, quickly switching from one eye to the other will give a "relative" indication of the functioning of each eye and optic nerve. If both eyes are equally dysfunctional, no "relative" defect would be found. The results of the test include:

- No Relative Afferent Pupillary Defect: Both pupils constrict equally without evidence of pupillary re-dilation with the "swinging flashlight test", except possibly for "hippus". Hippus refers to non-rhythmic fluctuations in pupillary size when there is a steady illumination.
- Mild Relative Afferent Pupillary Defect: The affected pupil shows a weak initial constriction, followed by dilation to a greater size.
- Moderate Relative Afferent Pupillary Defect: The affected pupil shows a stable or unchanged level of constriction, followed by dilation to a greater size.
- Severe Relative Afferent Pupillary Defect: The affected pupil shows an immediate dilation to a greater size.

## PSM

133. Movement in socioeconomic levels is

- a) Social equality
- b) Socioeconomic upliftment
- c) **Social mobility**
- d) Social insurance

“Social mobility is movement of people or classes of people along the social ladder based on knowledge & achievements.” – *Park 20e/600*

134. Full form is JSY is

- a) Jeevan swasthya yojana
- b) **Janani suraksha yojana**
- c) Jan shouchalaya yojana
- d) Jeevan suraksha yojana

The **Janani Suraksha Yojana (JSY)** is a centrally sponsored scheme aimed at reducing maternal and infant mortality rates and increasing institutional deliveries in below poverty line (BPL) families. The JSY, which falls under the overall umbrella of National Rural Health Mission (External website that opens in a new window), **covers all pregnant women belonging to households below the poverty line, above 19 years of age and up to two live births.**

The JSY modifies the existing National Maternity Benefit Scheme or NMBS. While the NMBS was connected with providing a better diet for pregnant women from below poverty line (BPL) families, the JSY integrates help in the form of cash with antenatal care during pregnancy period, institutional care during delivery as well as post-partum care (External website that opens in a new window). This is provided by field level health workers through a system of coordinated care and health centres.

1. Janani Suraksha Yojana (**JSY**) is a safe motherhood intervention under the National Rural Health Mission (NRHM) being implemented with the objective of reducing maternal and neo-natal mortality by promoting institutional delivery among the poor pregnant women. The Yojana, launched on **12<sup>th</sup> April 2005**, by the Hon’ble Prime Minister, is being implemented in all states and UTs with special focus on low performing states.

2. JSY is a **100 % centrally sponsored scheme** and it integrates cash assistance with delivery and post-delivery care. The success of the scheme would be determined by the increase in institutional delivery among the poor families

3. The Yojana has identified **ASHA**, the accredited social health activist as an effective link between the Government and the poor pregnant women in **10 low performing states, namely the 8 EAG states and Assam and J&K and the remaining NE States.** In other eligible states and UTs, wherever, AWW and

TBAs or ASHA like activist has been engaged in this purpose, she can be associated with this Yojana for providing the services.

135. Which is not true regarding maternal mortality rate ?

- a) Numerator includes total no of female deaths within 42 days of delivery
- b) Denominator includes still births & abortions
- c) It is a rate, not a ratio
- d) Expressed in per 1000

Denominator in MMR includes total no of live birth in the same area & year.

136. One of the following influenced by motivation & associated with emotional valence

- a) Attitude
- b) Belief
- c) Knowledge
- d) Practice

137. Not a synthetic pyrethroid compound is

- a) DDT
- b) Cypermethrin
- c) Permethrin
- d) Proparthin

DDT is a synthetic organo-chlorine compound.

138. IMNCI defers from IMCI by all except

- a) Malaria & anemia are included
- b) 0-7 days neonates are included
- c) Sick neonates are preferred over sick older child
- d) It is aimed at more than 1 disease at a time

“Proportion of time given to sick young infants & sick child is almost equal.” – Park 20e/387

139. Perinatal mortality rate includes

- a) Still births & deaths within 7 days of birth
- b) Neonate deaths within 30 days of birth
- c) Abortions & death within 7 days of birth
- d) Death betn 7-28 days of birth

140. Provision of primary health care was done by

- a) Bhole committee

- b) Alma Ata
- c) Srivastava committee
- d) National health policy

The International Conference on Primary Health Care was convened in Alma-Ata, Kazakhstan, in 1978, and was attended by virtually all the member nations of the World Health Organization (WHO) and UNICEF. The Alma-Ata Declaration of 1978 emerged as a major milestone of the twentieth century in the field of public health, and it identified primary health care (PHC) as the key to the attainment of the goal of Health for All (HFA).

141. Not an element of primary health care is
- a) Provision of essential drug
  - b) Cost effectiveness
  - c) Immunization against major infectious diseases
  - d) Health education
142. Latest trend in Health care
- a) Qualitative enquiry
  - b) Community participation
  - c) Equitable distribution
  - d) Primary health care
143. Highest level of community participation is measured by
- a) Based on felt need of people
  - b) They plan & decide their own action
  - c) Provide resources for programme
  - d) Co-operation with workers
144. The population of a community of 1<sup>st</sup> June was 165000. 22 new cases of tuberculosis were detected from 1<sup>st</sup> January to 31<sup>st</sup> June. Total registered cases of tuberculosis were 220. What is the incidence of TB per 10 lakh population ?
- a) 133
  - b) 220
  - c) 13
  - d) 22
145. Mass prophylaxis in endemic area is done in all except
- a) Yaws
  - b) Leprosy
  - c) Trachoma
  - d) Filaria

146. A case of acute flaccid paralysis must be observed for how many days for residual weakness
- 30 days
  - 42 days
  - 60 days
  - 90 days
147. Rural and urban differences in seen in all except
- Lung ca
  - TB
  - Mental illness
  - Chronic bronchitis
148. Health status of a child under 5 years of age will be adversely affected by all except
- Malnutrition
  - LBW
  - Maternal Hb < 11gm%
  - Infections
149. Resurgent malaria is due to all except
- Drug resistance in host
  - Drug resistance in parasite
  - Antigenic variation in parasite
  - Drug resistance in vectors
150. True about SCRUB typhus is all except
- Caused by tsutsugamushi
  - Tetracycline is TOC
  - Adult feeds on host
  - Mite is host

Larval mites (**chiggers**) feed on hosts.

151. NPCB strategy for screening of diabetic retinopathy is
- Opportunistic screening
  - High risk screening
  - Eye examination by PHC MO
  - Mass screening
152. ASHA is posted at
- Village level
  - PHC
  - CHC

d) Sub centre

153. Indian aims to eliminate which disease by 2015

- a) Malaria
- b) TB
- c) Kala azar
- d) **Filaria**

154. True about confidence limits is

- a) Smaller the confidence limit, wider the interval
- b) Lesser variable the data, wider the confidence interval
- c) Width of CI is independent of sample size
- d) **Confidence limit of 95% corresponds to 2 SE**

155. True about tuberculosis Annual Infection rate is all except

- a) Incidence in India is 1.7%
- b) **1% is equivalent with 75 cases**
- c) Shows recent trend affected by current control measures
- d) It is the no of new infn

1% of ARI is equivalent to **50 new cases** for 100,000 general population.

156. True about Z score is

- a) Used in binomial distribution
- b) **Normal distribution**
- c) Chi square test
- d) t test

157. True about Red cross emblem are all except

- a) Symbol contains 2 vertical & 2 horizontal crosses with equal length
- b) **Can be used by all UNO members**
- c) Its use without permission of GOI is punishable under Indian law
- d) Came into existence in Geneva

Can be used only by those belonging to **Red Cross Movement** & **Army Medical Services**.

158. A new test for diabetes was carried out. Out of the 80 people who were tested +ve, it was found that actually 40 had diabetes and out of 9920 people who were tested -ve, only 9840 do not have the disease in actual. Calculate the sensitivity of the test

- a) **33%**
- b) 50%
- c) 65%

d) 99%

		Disease		Total
		+	-	
Test	+	40	40	80
	-	80	9840	9920
Total		120	9880	

The best way to solve these problems is to make a table first.

$$\text{Sensitivity} = [TP/(TP+FN)] \times 100 = [40/120] \times 100 = 33\%$$

# MEDICINE

159. A patient presented with gait abnormality, urinary incontinence and dementia. What is the likely diagnosis ?
- NPH
  - Multiple system atrophy
  - Alzheimer's ds
  - Progressive supranuclear palsy

Normal pressure hydrocephalus (NPH) is a clinical symptom complex characterized by **abnormal gait, urinary incontinence, and dementia**. It is an important clinical diagnosis because it is a potentially **reversible cause of dementia**. First described by Hakim in 1965, NPH describes **hydrocephalus** in the absence of papilledema and with **normal cerebrospinal fluid (CSF) opening pressure** on lumbar puncture.

## Pathophysiology

NPH differs from other causes of adult hydrocephalus. **An increased subarachnoid space volume does not accompany increased ventricular volume**. Clinical symptoms result from distortion of the central portion of the corona radiata by the distended ventricles. This may also lead to interstitial edema of the white matter and impaired blood flow, as suggested in nuclear imaging studies. **The periventricular white matter anatomically includes the sacral motor fibers that innervate the legs and the bladder, thus explaining the abnormal gait and incontinence**. Compression of the brainstem structures (ie, pedunculo pontine nucleus) could also be responsible for gait dysfunction, particularly the freezing of gait that has been well described. **Dementia results from distortion of the periventricular limbic system**.

The term *normal pressure hydrocephalus* was based on the finding that all 3 patients reported by Hakim and Adams showed low CSF pressures at lumbar puncture, namely 150, 180, and 160 mm H<sub>2</sub> O. However, an isolated CSF pressure measurement by lumbar puncture clearly yields a poor estimation of the real intracranial pressure (ICP) in patients with NPH.

## History

Patients present with a gradually progressive disorder. As noted above, the **classic triad consists of abnormal gait, urinary incontinence, and dementia**. **The gait disturbance is typically the earliest feature noted and considered to be the most responsive to treatment**. The primary feature is thought to resemble an **apraxia of gait**. True weakness or ataxia is typically not observed.

The gait of NPH is characterized as **bradykinetic, broad based, magnetic, and shuffling**. The urinary symptoms of NPH can present as **urinary frequency, urgency, or frank incontinence**. While incontinence can result from gait disturbance and dementia, in a study by Sakakibara and colleagues, 95% of patients had urodynamic parameters consistent with detrusor overactivity.

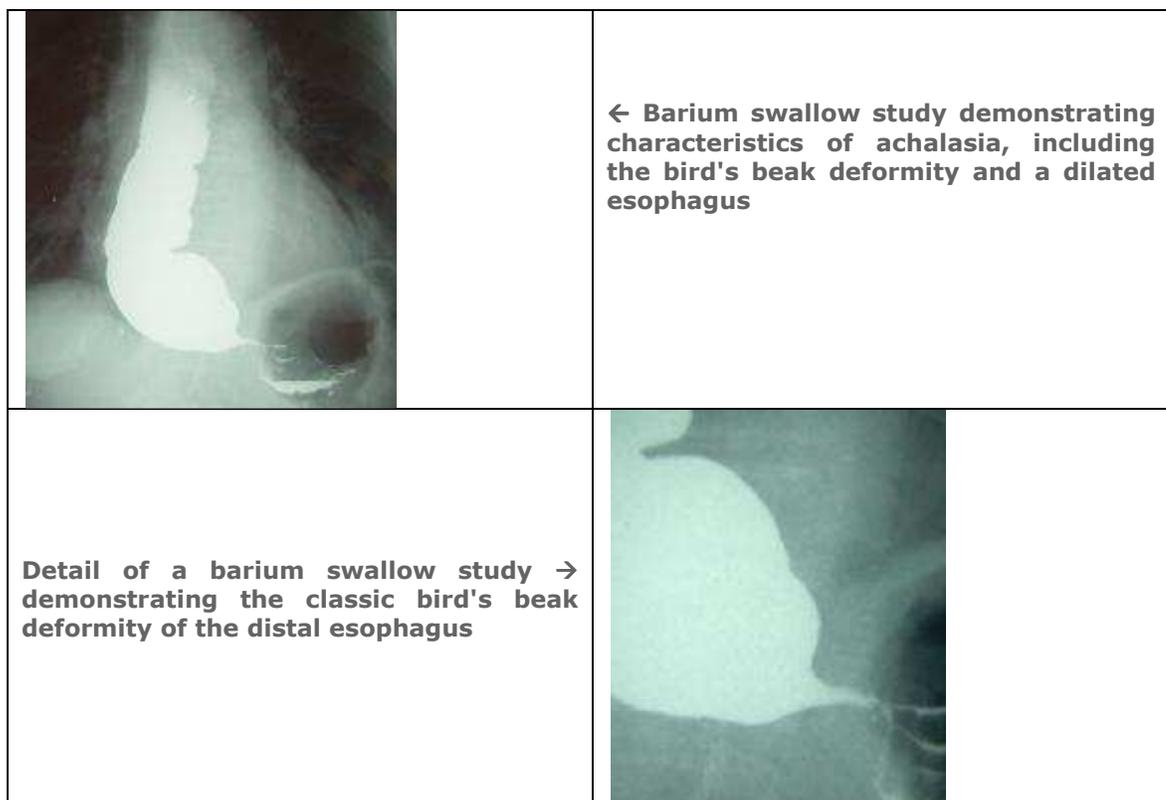
The **dementia** of NPH is characterized by **prominent memory loss and bradyphrenia**. Frontal and subcortical deficits are particularly pronounced. Such deficits include **forgetfulness, decreased attention,**

**inertia, and bradyphrenia.** The presence of cortical signs such as aphasia or agnosia should raise suspicion for an alternate pathology such as Alzheimer disease or vascular dementia. However, comorbid pathology is not uncommon with advancing age. In one study, more than 60% of patients with iNPH had cerebrovascular disease. In another similar study, more than 75% had AD pathology at the time of shunt surgery.

Patients commonly present with a **gait disorder and dementia**. On neurologic examination, pyramidal tract findings may be present in addition to the above findings.

160. A lady presented with non progressive dysphagia for solids since 2 months. Barium study showed proximal esophageal dilatation with distal tapering end. The most likely diagnosis is
- Peptic stricture
  - Ca esophagus
  - Achalasia cardia**
  - Lower esophageal ring

This pt presents with dysphagia for mainly solids which suggests mechanical obstruction. Classic barium study suggests achalasia cardia. Remember, dysphagia in achalasia cardia may be non-progressive.



161. A truck driver presented with non productive cough, fever, exertional dyspnea, breathlessness. He has lost 5 kg weight in a year. X-ray chest showed diffuse interstitial infiltrates. Likely diagnosis is
- Mycoplasma pneumonia
  - Pneumocystis carinii pneumonia**
  - Pulmonary TB
  - Disseminated candidiasis

## History

The symptoms of *P. carinii* pneumonia (PCP) are nonspecific. PCP in patients with HIV infection tends to run a more subacute indolent course and tends to present much later, often after several weeks of symptoms, compared with PCP associated with other immunocompromising conditions. Symptoms of PCP include the following:

- Progressive **exertional dyspnea** (95%)
- **Fever** (>80%)
- **Nonproductive cough** (95%)
- Chest discomfort
- **Weight loss**
- Chills
- Hemoptysis (rare)

## Physical

The physical examination findings of PCP are nonspecific and include the following:

- Tachypnea
- Fever
- Tachycardia
- Pulmonary symptoms: Pulmonary examination may reveal mild crackles and rhonchi but may yield normal findings in up to half of patients.
- Additional findings in children with severe disease
  - **Cyanosis**
  - Nasal flaring
  - Intercostal retractions
- Extrapulmonary manifestations: Although *Pneumocystis* infection rarely causes extrapulmonary manifestations, **they may be present in patients receiving aerosolized pentamidine for prophylaxis** or in patients with advanced HIV infection who are not taking any prophylaxis. They may also occur in the absence of lung involvement. Based on most well-documented findings, *Pneumocystis* infection may present in almost any organ system, as follows:
  - CNS
  - Bone marrow (may have necrosis with resultant pancytopenia)
  - Lymphadenopathy
  - Eyes (may have **retinal cotton-wool spots**)

- Thyroid (may present as a rapidly enlarging thyroid mass)
- GI tract

## Causes

PCP is caused by infection with *P jiroveci*. The following groups are at risk for PCP:

- Persons with HIV infection whose CD4<sup>+</sup> cells fall below 200/ $\mu$ L and who are not receiving PCP prophylaxis (In addition, in patients with HIV infection, findings of other opportunistic infections [eg, oral thrush] increases the risk of PCP, regardless of CD4<sup>+</sup> count.)
- Persons with primary immune deficiencies, including hypogammaglobulinemia and severe combined immunodeficiency (SCID).
- Persons receiving long-term immunosuppressive regimens for connective-tissue disorders, vasculitides, or solid-organ transplantation (eg, heart, lung, liver, kidney)
- Persons with hematologic and nonhematologic malignancies, including solid tumors and lymphomas
- Persons with severe malnutrition

162. Hyperglycemia is associated with

- a) Multiple myeloma
- b) Ewing sarcoma
- c) Osteosarcoma
- d) Chondrosarcoma

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare multisystemic disease that occurs in the setting of a plasma cell dyscrasia. The pathophysiologic link between the constellation of symptoms and the underlying disease is not well understood, but the link may be related to changes in the levels of a cytokine or a growth factor. POEMS syndrome was first described by Crow in 1956 and then by Fukase in 1968. The syndrome was termed Crow-Fukase syndrome (by which it is known in Japan) by Nakanishi in a study of 102 cases in Japan.

In 1980, the acronym POEMS was coined by Bardwick et al based on the 5 main features of the disease, namely, polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes.

No specific case definition exists for POEMS syndrome; however, most authors agree that patients with POEMS syndrome should have 3 or more of the 5 features. Some authors have proposed that the presence of 2 major criteria, including a monoclonal plasma-proliferative disorder and polyneuropathy, in addition to the existence of 1 minor criterion, is sufficient for diagnosis. The suggested minor criteria include sclerotic bone lesions, organomegaly, edema, endocrinopathy, papilledema, and skin changes. However, the findings of a retrospective analysis of 629 patients using these criteria suggest that this approach may be inadequate for excluding other disease processes that may account for symptoms and that atypical presentations of POEMS may be misdiagnosed.

The polyneuropathy associated with POEMS syndrome is a bilateral symmetric disturbance. It involves both motor and sensory nerves, begins distally, and has a progressive proximal spread. Associated cranial or autonomic nerves are not involved. Both demyelination and axonal degeneration are noted.

The liver, the lymph nodes, and the spleen are the organs most frequently involved. Enlargement of the lymph nodes and spleen is secondary to changes consistent with Castleman disease (giant angiofollicular hyperplasia, multicentric plasma cell variant) in most patients. Approximately 15% of patients with POEMS syndrome have concomitant evidence of Castleman disease. Hepatomegaly is not associated with any defined histologic or pathophysiologic changes.

Multiple endocrinopathies have been associated with POEMS syndrome, and most patients have more than 1 endocrine abnormality. Many of the abnormalities noted can be explained by elevations in estrogen levels. Impotence and gynecomastia are common among men. Amenorrhea is common among women. Diabetes mellitus and glucose intolerance are also noted in many patients. Other associated endocrinopathies include hypothyroidism, hyperprolactinemia, and hypoparathyroidism.

POEMS syndrome is seen in the setting of a plasma cell dyscrasia. Although many plasma cell disorders have been reported in patients with POEMS syndrome, most patients are seen with osteosclerotic myeloma or monoclonal gammopathy of unknown significance.

The M proteins most frequently found are the immunoglobulin A (IgA)-gamma and immunoglobulin G (IgG)-gamma light chains. In a case report of one patient with POEMS syndrome serum electrophoresis demonstrated an M-band with isolated IgA heavy chain but no abnormal light chain, which could suggest abnormal secretion of monoclonal protein or the rare possibility of coincidental heavy-chain disease in association with POEMS syndrome. A single case of POEMS syndrome in association with Waldenström macroglobulinemia, characterized by immunoglobulin M-kappa paraproteinemia, has been reported. Classic multiple myeloma has not been associated with the disease. The type of plasma cell disorder has not been shown to be correlated with the constellation of symptoms noted in patients with POEMS syndrome.

Multiple dermatologic changes have been associated with POEMS syndrome. The most common changes include hyperpigmentation, skin thickening, sclerodermoid changes, and hypertrichosis. Other skin changes, including whitening of the proximal nail (Terry nails), peripheral edema, hyperhidrosis, clubbing of the fingers, Raynaud phenomenon, and angiomas, have been observed.

Other signs and symptoms associated with POEMS syndrome include papilledema, anasarca, pleural effusions, ascites, fever, thrombosis, renal insufficiency, and diarrhea.

163. In motor neuron disease, the site of lesion is
- N-M jn
  - Peripheral nerve
  - Anterior horn cells
  - Dorsal root ganglion

“The pathologic hallmark of motor neuron disease is death of LMN (consisting of AHCs in spinal cord & their brainstem homologues innervating bulbar muscles).” – H17/2572

164. A patient presented with fever for 3 weeks. On examination, he had splenomegaly. On USG, there was a hypoechoic shadow in spleen near hilum. Blood culture isolated gram negative bacilli. What is the causative organism?

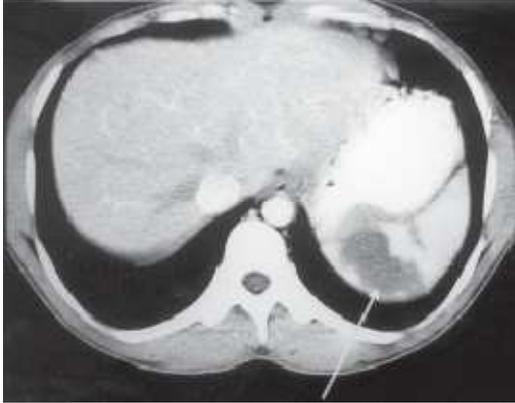
- a) CMV
- b) Toxoplasmosis
- c) **Salmonella**
- d) Lymphoma virus

*Enteric fever* is a misnomer, in that the hallmark features of this disease—fever and abdominal pain—are variable. While fever is documented at presentation in >75% of cases, abdominal pain is reported in only 30–40%. Thus, a high index of suspicion for this potentially fatal systemic illness is necessary when a person presents with fever and a history of recent travel to a developing country.

The incubation period for *S. Typhi* averages 10–14 days but ranges from 3 to 21 days, with the duration likely reflecting the inoculum size and the host's health and immune status. The most prominent symptom is prolonged fever (38.8°–40.5°C; 101.8°–104.9°F), which can continue for up to 4 weeks if untreated. *S. Paratyphi A* is thought to cause milder disease than *S. Typhi*, with predominantly gastrointestinal symptoms.

Early physical findings of enteric fever include rash ("rose spots"), hepatosplenomegaly (3–6%), epistaxis, and relative bradycardia at the peak of high fever. Rose spots (Fig. 146-2) make up a faint, salmon-colored, blanching, maculopapular rash located primarily on the trunk and chest. The rash is evident in ~30% of patients at the end of the first week and resolves without a trace after 2–5 days. Patients can have two or three crops of lesions, and *Salmonella* can be cultured from punch biopsies of these lesions. The faintness of the rash makes it difficult to detect in highly pigmented patients. – *H17/957*

**Splenic abscess** is one of the abdominal complications of untreated typhoid fever, developing frequently in the third or fourth week of infection. In a review of 173 cases of splenic abscesses, typhoid fever accounted for 2.9%. The incidence of splenic abscess in typhoid is reported between 0.29–2%. Published data from India is scarce. The diagnosis should be considered in a patient with fever, abdominal pain, non-specific chest findings and leucocytosis. Left upper quadrant tenderness and splenomegaly are frequently encountered signs on examination. Friction rub has been reported in 3.3% only. Leucocytosis has been reported in up to 75% of patients, however the level of leucocytosis is of no value in the estimation of severity of disease or prediction of outcome. Both our patients had normal counts, although leucocytosis was documented in peripheral smear, later in the course of illness in one patient. Abnormal findings have been reported in 82% of chest radiographs in the form of mass effect in left upper quadrant, left pleural effusion, elevated left hemi diaphragm and lower lobe infiltrate. USG has a sensitivity of 76%. It detects large abscesses easily, but may miss the small abscesses. The classic CT appearance is a hypodense lesion with a density range of 18–30 HU. CT Scan with a sensitivity of 96% and specificity of 90–95% remains the gold standard for the definitive diagnosis. It is also helpful in planning therapeutic strategies like percutaneous drainage. Differential diagnosis of splenic abscesses in CT and US images include splenic infarct, hematoma, neoplasm and even complicated cyst. – *Two cases of Salmonella Splenic Abscess, Piplani et al, MJAFI 2006; 62 : 77-78*



CT study of abdomen showing multiple splenic abscesses

165. A 7 year old girl presented with lethargy and easy fatiguability. Her Hb is 6gm/dl, MCV is 78fl, MCH is 22 pg and Red cell width is 28. What is the diagnosis?
- Fe deficiency anemia
  - Hereditary spherocytosis
  - Thalassemia minor
  - Sideroblastic anemia

Parameter	Normal	Pt in question	Inference
Hb	13-18 gm%	6 gm%	Anemia
MCV	86-98 fl	78 fl	Microcytic
MCH	28-33 pg/cell	22 pg	Hypochromia
RDW	11-14%	28	Raised

Conclusion: Microcytic hypochromic anemia with high RDW → Iron deficiency anemia

166. A patient presented with pCO<sub>2</sub> 30 mmHg, pO<sub>2</sub> 102 mmHg, pH 7.5. Which type of compensation it is ?
- Metabolic alkalosis
  - Metabolic acidosis
  - Resp alkalosis
  - Resp acidosis
167. WHO essential criteria for the diagnosis of polycythemia vera is
- Low EPO levels
  - Presence of JAK2(v617f) Stat mutation
  - High O<sub>2</sub> saturation
  - Erythrocytosis

Polycythemia vera (PV) is a stem cell disorder characterized as a panhyperplastic, malignant, and neoplastic marrow disorder. The most prominent feature of this disease is an **elevated absolute red blood cell mass because of uncontrolled red blood cell production**. This is accompanied by increased white blood cell (myeloid) and platelet (megakaryocytic) production, which is due to an abnormal clone of the hematopoietic stem cells with increased sensitivity to the different growth factors for maturation.

Several reasons suggest that a **mutation on the Janus kinase-2 gene (JAK2)** is the most likely candidate gene involved in polycythemia vera (PV) pathogenesis, as *JAK2* is directly involved in the intracellular signaling following exposure to cytokines to which polycythemia vera (PV) progenitor cells display hypersensitivity. A recurrent unique acquired clonal mutation in *JAK2* was found in most patients with polycythemia vera (PV) and other myeloproliferative diseases (MPDs) including essential thrombocythemia and idiopathic myelofibrosis.

A unique **valine to phenylalanine substitution at position 617 (V617F) in the pseudokinase JAK2 domain has been identified called JAK2V617F** that leads to a permanently turned on signaling at the affected cytokine receptors. How these mutations interact with the wild type kinase genes and how they manifest into different forms of MPDs need to be elucidated.

The **World Health Organization (WHO) criteria** for polycythemia vera diagnosis requires 2 components: reasonable elimination of apparent and secondary polycythemia and confirmation of polycythemia vera. However, the discovery of the *JAK2V617F* mutation have made these criteria insufficient. A proposed set of revised criteria have recently been published.

Diagnosis requires the presence of **both major criteria and one minor criterion** or the presence of the **first major criterion together with 2 minor criteria**.

#### Major criteria

- Hemoglobin level of more than 18.5 g/dL in men, more than 16.5 g/dL in women, or other evidence of increased red cell volume (hemoglobin or hematocrit levels >99<sup>th</sup> percentile of method-specific reference range for age, sex, altitude of residence; hemoglobin level >17 g/dL in men, >15 g/dL in women [if associated with a documented and sustained increase of at least 2 g/dL from the individual's baseline value that cannot be attributed to correction of iron deficiency], or elevated red cell mass >25% above mean normal value).
- Presence of **JAK2V617F** or other functionally similar mutation such as **JAK2 exon 12** mutation

#### Minor criteria

- Bone marrow biopsy showing hypercellularity for age, with trilineage growth (panmyelosis) with prominent erythroid, granulocytic, and megakaryocytic proliferation (not validated in prospective studies)
- Serum erythropoietin level below the reference range for normal
- Endogenous erythroid colony formation in vitro

168. A patient is found to have HBsAg+ve, other tests are unremarkably –ve. Liver enzymes are within normal range and patient is asymptomatic. What is the diagnosis ?
- Inactive HBV infn
  - Inactive HBV carrier
  - Acute HBV infn with active ds
  - Chronic HBV infn
169. True about Wilson disease is
- Low serum ceruloplasmin & low urinary copper
  - Low serum ceruloplasmin & high urinary copper
  - High serum ceruloplasmin & high urinary copper
  - High serum ceruloplasmin & low urinary copper

*Wilson disease is a very imp topic for PG & has been asked repeatedly in recent papers in AI & AIIMS. You need to know everything about this topic. Following are the High-Yield Facts on this topic, prepared from Harrison & Nelson. Read it carefully.*

## WILSON DISEASE

### INTRODUCTION

- Autosomal recessive
- Mutation in **ATP7B gene**, a P-type ATPase on **chromosome 13**
- Pathogenesis- ATP7B protein deficiency impairs biliary copper excretion, resulting in positive copper balance, hepatic copper accumulation, and copper toxicity from oxidant damage.

### CLINICAL PRESENTATION

#### (A) Hepatic

Wilson disease may present as **hepatitis**, **cirrhosis**, or as **hepatic decompensation**,

Hepatic decompensation is associated with

- elevated serum bilirubin
- reduced serum albumin and coagulation factors

- ascites
- peripheral edema
- hepatic encephalopathy

In severe hepatic failure, **hemolytic anemia** may occur because large amounts of copper derived from hepatocellular necrosis are released into the bloodstream. **The association of hemolysis and liver disease makes Wilson disease a likely diagnosis.**

## (B) Neurologic

The three main movement disorders include:

- dystonia
- incoordination
- tremor (**wing-beating tremor**)

Autonomic disturbances may include

- orthostatic hypotension
- sweating abnormalities
- bowel, bladder, and sexual dysfunction

## (C) Psychiatric

A history of **behavioral disturbances**, with onset in the five years before diagnosis, is present in half of patients with neurologic disease. The features are

- loss of emotional control (temper tantrums, crying bouts)
- depression
- hyperactivity
- loss of sexual inhibition

## (D) Others

- **Female** – Repeated abortions, amenorrhoea
- **Stones** – Cholelithiasis, Nephrolithiasis
- **Osteoarthritis** – Of knee
- **Microscopic hematuria**
- **Eye** – Sunflower cataracts, **K-F rings**

## DIAGNOSIS

**Table 354-1 Useful Tests for Wilson Disease**

Test	Usefulness <sup>a</sup>	Normal Value	Heterozygous Carriers	Wilson Disease
Serum ceruloplasmin	+	180–350 mg/L (18–35 mg/dL)	Low in 20%	Low in 90%
KF rings	++	Absent	Absent	Present in 99% + if neurologic or psychiatric symptoms present  Present in 30–50% in hepatic presentation and presymptomatic state
24-h urine Cu	+++	0.3–0.8 mol (20–50 g)	Normal to 1.3 mol (80 g)	>1.6 mol (>100 µg) in symptomatic patients  0.9 to >1.6 mol (60 to >100 g) in presymptomatic patients
Liver Cu	++++	0.3–0.8 mol/g (20–50 g) tissue	Normal to 2.0 mol (125 g)	>3.1 mol (>200 µg) (obstructive liver disease can cause false-positive results)
Haplotype analysis	++++ (Siblings only)	0 Matches	1 Match	2 Matches

- Symptomatic patients invariably have **urine copper levels > 1.6 mol (>100 µg) per 24 h.**
- The "**gold standard**" for diagnosis remains **liver biopsy with quantitative copper assays.** Affected patients have values >3.1 mol/g (>200 µg/g) dry weight of liver.

## **TREATMENT**

**Penicillamine** was previously the primary anticopper treatment but now plays a minor role because of its toxicity and because it often worsens existing neurologic disease if used as initial therapy.

**If penicillamine is given, it should always be accompanied by 25 mg/d of pyridoxine.**

**Trientine** is a less toxic chelator and is supplanting penicillamine when a chelator is indicated.

Table 354-2 Recommended Anticopper Drugs for Wilson Disease

Disease Status	First Choice	Second Choice
Initial hepatic		
Hepatitis or cirrhosis without decompensation	Zinc <sup>a</sup>	Trientine
Hepatic decompensation		
Mild	Trientine <sup>d</sup> and zinc	Penicillamine <sup>d</sup> and zinc
Moderate	Trientine and zinc	Hepatic transplantation
Severe	Hepatic transplantation	Trientine and zinc
Initial neurologic/psychiatric	Tetrathiomolybdate <sup>c</sup> and zinc	Zinc
Maintenance	Zinc	Trientine
Presymptomatic	Zinc	Trientine
Pediatric	Zinc	Trientine
Pregnant	Zinc	Trientine

<sup>a</sup>Zinc acetate is supplied as Galzin, manufactured by Gate Pharmaceutical. Recommended adult dose for all the above indications is 50 mg of elemental zinc three times daily, each dose separated from food and beverages other than water by at least 1 h, and separated from trientine or penicillamine doses by at least 1 h.

<sup>b</sup>Trientine is supplied as Syprine and penicillamine as Cuprimine, both manufactured by Merck. Recommended adult dosage for both drugs is 500 mg twice daily, each dose at least 1/2 h before or 2 h after meals.

<sup>c</sup>Tetrathiomolybdate is not yet commercially available but is expected to be marketed near the end of 2007.

For patients with hepatitis or cirrhosis, but without evidence of hepatic decompensation or neurologic/psychiatric symptoms, **zinc is the therapy of choice**, although some advocate therapy with trientine.

The first step in evaluating patients presenting with hepatic decompensation is to establish disease severity, which can be estimated using the **Nazer prognostic index** (Table 354-3). Patients with scores <7 can usually be managed with medical therapy. Patients with scores >9 should be considered immediately for liver transplantation, and those with scores between 7 and 9 require clinical judgment as to whether to recommend transplantation or medical therapy. A combination of trientine and zinc has been used to treat patients with Nazer scores as high as 9, but such patients should be watched carefully for indications of hepatic deterioration, which mandates transplantation.

Laboratory Measurement	Normal Value	Score (in Points)				
		0	1	2	3	4
<b>Serum bilirubin<sup>a</sup></b>	0.2–1.2 mg/dL	<5.8	5.8–8.8	8.8–11.7	11.7–17.5	>17.5
<b>Serum aspartate transferase (AST)</b>	10–35 IU/L	<100	100–150	151–200	201–300	>300
<b>Prolongation of prothrombin time (seconds)</b>	—	<4	4–8	9–12	13–20	>20

For initial medical therapy of patients with hepatic decompensation, a **chelator** (trientine is preferred) plus **zinc** is recommended (Table 354-2). Zinc should not, however, be ingested simultaneously with trientine, as it will chelate zinc and form therapeutically ineffective complexes; the two drugs should be separated by at least 1 h.

For initial neurologic therapy, **tetrathiomolybdate** is emerging as the drug of choice because of its rapid action, preservation of neurologic function, and low toxicity. Penicillamine and trientine should be avoided because they each have a high risk of worsening the neurologic condition.

Pregnant patients should be treated with **zinc** or **trientine** throughout pregnancy, but without tight copper control, because copper deficiency can be teratogenic.

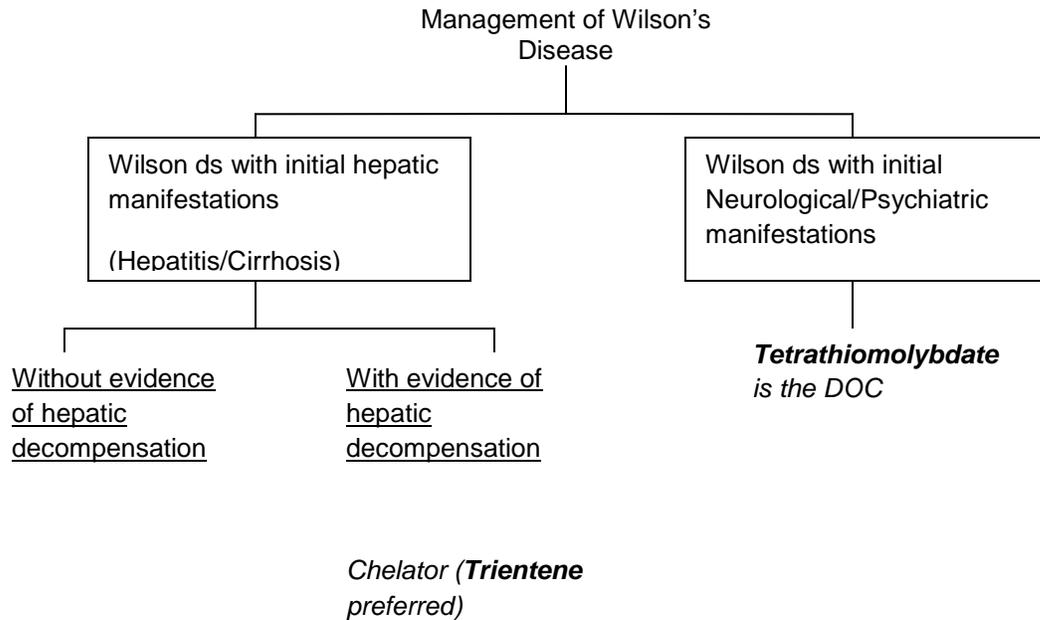
### **Monitoring Anticopper Therapy**

When first using trientine or penicillamine, it is necessary to monitor for drug toxicity, particularly **bone marrow suppression** and **proteinuria**. Complete blood counts, standard biochemical profiles, and a urinalysis should be performed at weekly intervals for a month, then at 2-weekly intervals for 2–3 months, then at monthly intervals for 3 or 4 months, and at 4- to 6-monthly intervals thereafter.

The anticopper effects of trientine and penicillamine can be monitored by following **24-h "free" serum copper**.

Zinc treatment does not require blood or urine monitoring for toxicity. Its only significant side effect is gastric burning or nausea in ~10% of patients, usually with the first morning dose. This can be mitigated by taking the first dose an hour after breakfast or taking the zinc with a small amount of protein.

## TREATMENT SYNOPSIS



## Review Questions

1. Following are true about Wilson disease EXCEPT- (AIIMS May 2009)
- Serum ceruloplasmin <20
  - Cu excretion <100 µg
  - Autosomal recessive
  - Zn used as mode of treatment

Answer:

1. B

170. Accelerated idioventricular rhythm is seen in
- Reperfusion therapy
  - Dilated cardiomyopathy
  - Digitalis toxicity
  - AV nodal ds

Both A & C appear to be correct. However, if we have to choose 1 option, I think A is better (A>C).

AIVR is currently defined as an enhanced ectopic ventricular rhythm with at least 3 consecutive ventricular beats, which is faster than normal intrinsic ventricular escape rhythm ( $\leq 40$  bpm), but slower than ventricular tachycardia (at least 100-120 bpm).

### AIVR in acute myocardial infarction

Clinically, AIVR has been best studied in patients with acute ST-elevation myocardial infarction (STEMI). In the thrombolysis era, AIVR was noted to be a marker of reperfusion. However, not all patients with reopened coronary artery have AIVR. In patients with acute myocardial infarction treated with primary percutaneous coronary intervention, the reported incidence of AIVR varied significantly, ranging from 15-50%, depending on methods of monitoring.

Recently, studies in patients with STEMI treated with primary percutaneous coronary intervention support that AIVR is a marker of occluded coronary artery reopening, but is not necessarily a marker for complete reperfusion. In fact, AIVR seems to be associated with more extensive myocardial damage and delayed microvascular reperfusion, although the mortality rates are similar in patients with and without AIVR.

Under certain conditions such as acute ischemia and digoxin toxicity, triggered activity has been suggested as the mechanism for AIVR

### Causes

The AIVR can occur in people with and without apparent heart diseases. The most common cause of AIVR is myocardial ischemia-reperfusion. Other causes include the following:

- Buerger disease
- Congenital heart disease
- Dilated cardiomyopathy
- Myocarditis
- Drugs: Digoxin toxicity, cocaine toxicity and various anesthesia agents
- Electrolyte abnormality
- Postresuscitation

171. Hypergonadotrophic hypogonadism in a males is associated with
- a) Viral orchitis
  - b) Klinefelter's syndrome
  - c) Kallmann syndrome
  - d) Noonan syndrome

In women with hypergonadotropic hypogonadism (ie, gonadal failure), the most common cause of hypogonadism is Turner syndrome, which has an incidence of 1 case per 2,500-10,000 live births. In men with hypergonadotropic hypogonadism, the most common cause is Klinefelter syndrome, which has an incidence of 1 case per 500-1000 live births. Hypergonadotropic hypogonadism is more rare.

Hypergonadotropic hypogonadism is more common in males than in females because the incidence of Klinefelter syndrome (the most common cause of primary hypogonadism in males) is higher than the incidence of Turner syndrome (the most common cause of hypogonadism in females). Incidence of hypogonadotropic hypogonadism is equal in males and females.

### **Causes of Hypergonadotropic hypogonadism**

- **Hypergonadotropic hypogonadism in males**
  - **Klinefelter syndrome**
  - Inactivating mutations
    - LH beta subunit
    - FSH beta subunit
    - LH receptor
    - FSH receptor
  - Other causes of primary testicular failure
    - Chemotherapy
    - Radiation therapy
    - Testicular biosynthetic defects
    - Sertoli-cell-only syndrome
    - LH resistance
    - Anorchism and cryptorchidism
  
- **Hypergonadotropic hypogonadism in females**
  - **Turner syndrome**
  - Inactivating mutations
    - LH beta subunit
    - FSH beta subunit
    - LH receptor
    - FSH receptor
  - XX and XY gonadal dysgenesis
    - Familial and sporadic XX gonadal dysgenesis and its variants
    - Familial and sporadic XY gonadal dysgenesis and its variants
  - Other causes of primary ovarian failure
    - Premature menopause
    - Radiation therapy
    - Chemotherapy
    - Autoimmune oophoritis
    - Resistant ovary
    - Galactosemia
    - Glycoprotein syndrome type 1
    - FSH-receptor gene mutations
    - LH/human chorionic gonadotropin (hCG) resistance
    - Polycystic ovarian disease
    - Noonan syndrome

## Klinefelter Syndrome: A Short Review

### History

**Infertility** and **gynecomastia** are the 2 most common symptoms that lead to diagnosis in patients with Klinefelter syndrome.

Other symptoms include fatigue, weakness, erectile dysfunction, osteoporosis, language impairment, academic difficulty, subnormal libido, poor self-esteem, and behavioral problems.

### Physical

#### Growth

- Infants and children achieve normal height, weight, and head circumference. About 25% have clinodactyly. Height velocity increases by age 5 years, and adults with Klinefelter syndrome are **usually taller than adults who do not have the syndrome**. Affected individuals also have **disproportionately long arms and legs**.
- Some individuals with **Klinefelter variant 49,XXXXY** have short stature .
- 



**G-banded 47,XXY karyotype.**

#### CNS

- Contrary to other genetic syndromes that arise from chromosomal trisomy (eg, Down syndrome, trisomy 18), the general cognitive ability of patients with Klinefelter syndrome is not typically in the intellectual disability range.
- Most males with the 47,XXY karyotype have **normal intelligence**. Family background influences intelligence quotient (IQ) score. Subnormal intelligence or mental retardation may be associated with the presence of a higher number of X chromosomes.
- About 70% of patients have **minor developmental and learning disabilities**. These may include academic difficulties, delayed speech and language acquisition, diminished short-term memory, decreased data-retrieval skills, reading difficulties, dyslexia, and attention deficit disorder.
- Patients may exhibit **behavioral problems and psychological distress**. This may be due to poor self-esteem and psychosocial development or a decreased ability to deal with stress.
- Psychiatric disorders involving **anxiety, depression, neurosis, and psychosis** are more common in this group than in the general population.

Dental: About 40% of patients have **taurodontism**, which is characterized by **enlargement of the molar teeth by an extension of the pulp**. The incidence rate is about 1% in healthy XY individuals.

#### Sexual characteristics

- Patients may **lack secondary sexual characteristics** because of a decrease in **androgen production**. This results in **sparse facial, body, or sexual hair; a high-pitched voice; and fat distribution as is observed in females**.
- By late puberty, 30-50% of boys with Klinefelter syndrome present with **gynecomastia**, which is secondary to elevated estradiol levels and an increased estradiol-to-testosterone ratio. The risk of developing **breast carcinoma** is at least 20 times higher than in healthy individuals.
- Postpubertal patients may have **testicular dysgenesis** (small firm testis; testis size, <10 mL).
- **Infertility, azoospermia**, or both may result from atrophy of the seminiferous tubules. Practically all individuals with a 47,XXY karyotype are **infertile**. Patients with Klinefelter syndrome mosaicism (46,XY/47,XXY) can be fertile. Guidelines for the assessment and treatment of people with fertility problems have been established.
- Patients may have an increased frequency of **extragonadal germ cell tumors** such as embryonal carcinoma, teratoma, and primary mediastinal germ cell tumor.
- Although **genital abnormalities are not commonly observed** in patients with Klinefelter syndrome, the association is important to note because Klinefelter syndrome is one of the causes of genital abnormality or ambiguity. The phenotype include complete sex reversal, true hermaphroditism (eg, ovotestes), testicular feminization, ambiguous genitalia/undervirilization (eg, hypospadias, micropenis, epispadias, female external genitalia), and mild genital abnormalities.

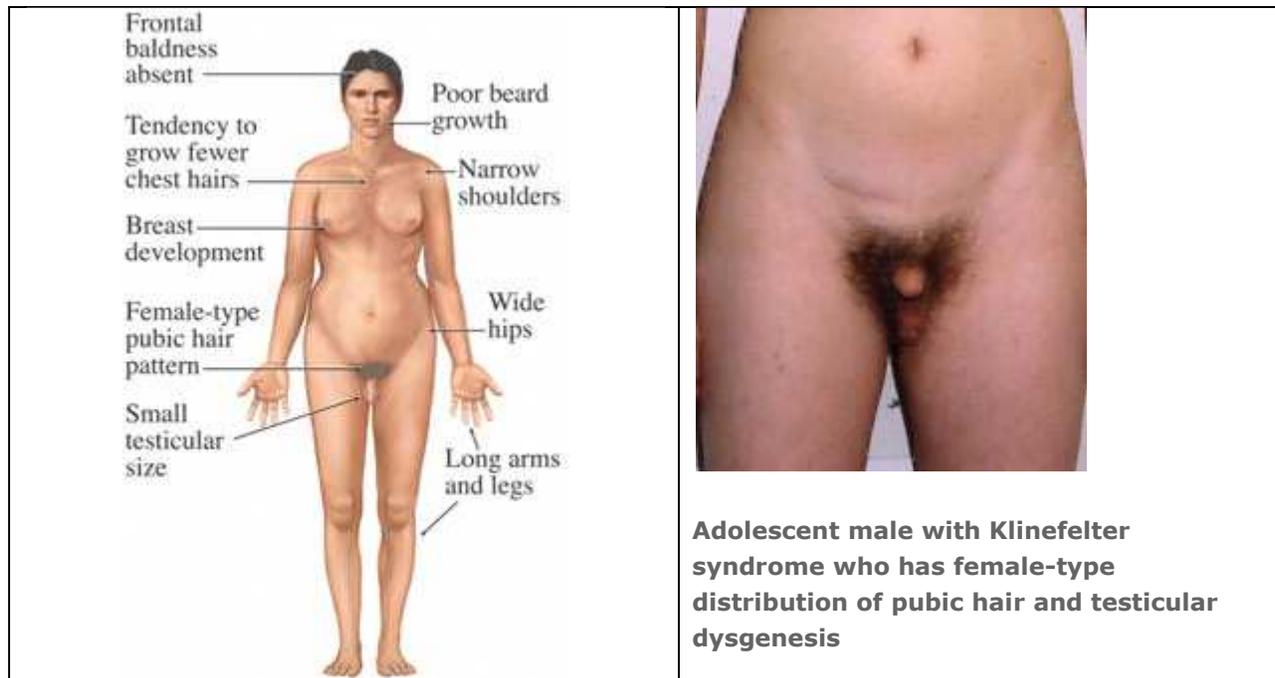
#### Cardiac and circulatory problems

- **Mitral valve prolapse** occurs in 55% of patients.
- **Varicose veins** occur in 20-40% of patients.
- The prevalence of **venous ulcers** is 10-20 times higher than in healthy individuals, and the risk of **deep vein thrombosis and pulmonary embolism** is increased.

#### Klinefelter variants

- **48,XXYY variant:** Patients typically have mild mental retardation; tall stature; eunuchoid body habitus; sparse body hair; gynecomastia; long, thin legs; hypergonadotropic hypogonadism; and small testes.
- **48,XXXYY variant:** Patients typically have mild-to-moderate mental retardation, speech delay, slow motor development, poor coordination, immature behavior, normal or tall stature, abnormal face (epicanthal folds, hypertelorism, protruding lips), hypogonadism, gynecomastia (33-50%), hypoplastic penis, infertility, clinodactyly, and radioulnar synostosis and benefit from testosterone therapy.
- **49,XXXYY variant:** Patients typically have moderate-to-severe mental retardation, passive but occasionally aggressive behavior and temper tantrums, tall stature, dysmorphic facial features, gynecomastia, and hypogonadism.
- **49,XXXXYY variant:** The **classic triad** is **mild-to-moderate mental retardation, radioulnar synostosis, and hypergonadotropic hypogonadism**. Other clinical features include severely

impaired language, behavioral problems, low birthweight, short stature in some individuals, abnormal face (round face in infancy, coarse features in older age, hypertelorism, epicanthal folds, prognathism), short or broad neck, gynecomastia (rare), congenital heart defects (patent ductus arteriosus is most common), skeletal anomalies (genu valgus, pes cavus, fifth finger clinodactyly), muscular hypotonia, hyperextensible joints, hypoplastic genitalia, and cryptorchidism. Pea-sized testes, micropenis, and infantile secondary sex characteristics are characteristic in patients with 49,XXXXY, whereas patients with 48,XXXXY exhibit milder hypogonadism similar to that found in patients with 47,XXY.

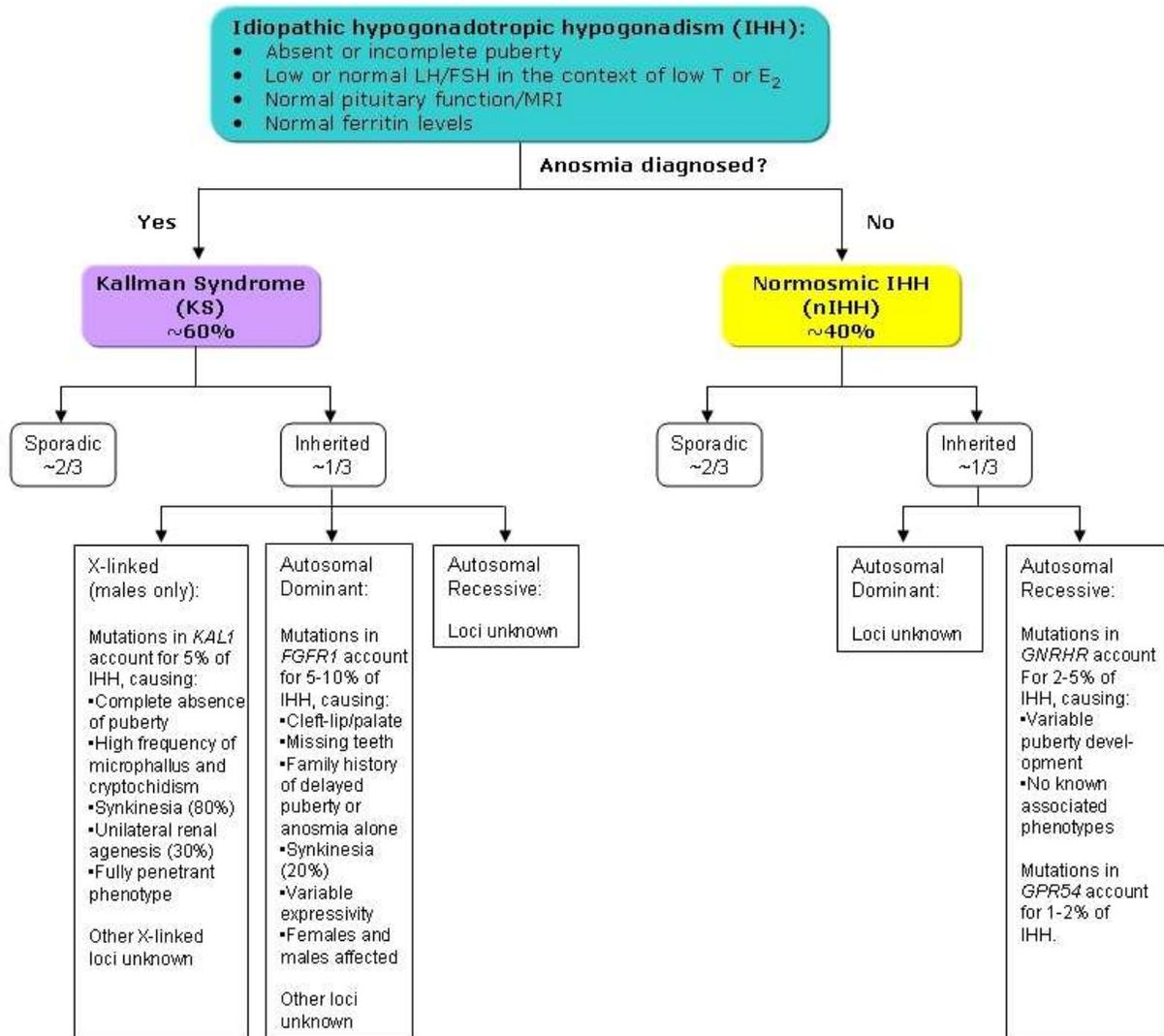


**Extra Edge:** Some facts about Hypogonadotropic hypogonadism

**Genetics of hypogonadotropic hypogonadism:** To date, numerous genes have been identified as causes of hypogonadotropic hypogonadism. The genes include the following:

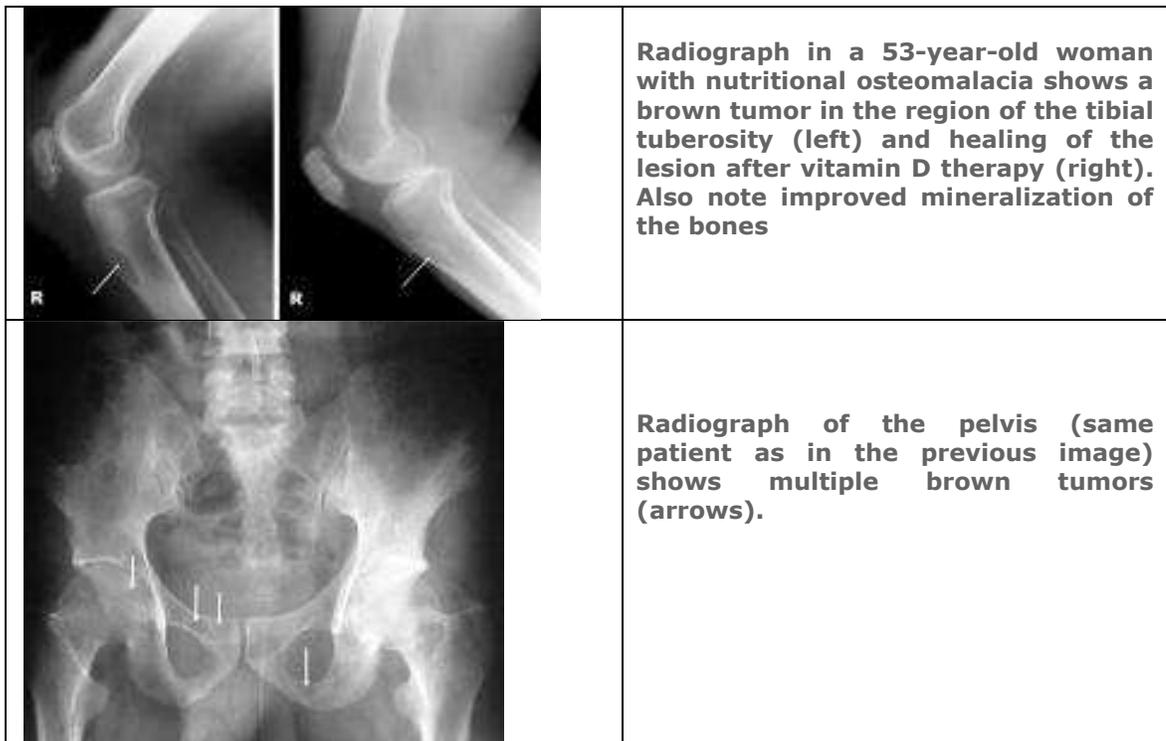
- **KAL** is located on the **X chromosome**, just below the pseudoautosomal region. An abnormality in this gene results in **Kallmann syndrome**, which is characterized by anosmia and hypogonadotropic hypogonadism. **FGFR1, FGF8, PROK2, and PROKR2** have also been associated with Kallmann syndrome. The relationship with Kallmann syndrome is thought to be due to the relation of these genes to the development and migration of gonadotropin-releasing hormone (GnRH) neurons.
- The **DAX1 gene** is associated with **X-linked adrenal hypoplasia congenita** (hypogonadotropic hypogonadism and adrenal insufficiency).
- **GNRHR** is the gene associated with the **GnRH (LHRH) receptor**.

- *GNRH1*, *KISS1R*, and *GNRHR* genes have been associated with normosmic (sense of smell is not disrupted) hypogonadotropic hypogonadism.
- *TAC3* and *TACR3* mutations have also been associated with normosmic hypogonadotropic hypogonadism, although their exact functions are unclear.
- *CHD7* mutation, which has been associated with **CHARGE syndrome**, has also been found in patients with both normosmic and anosmic hypogonadotropic hypogonadism.
- *PC1* is the gene for **prohormone convertase 1**. Abnormality of this gene causes hypogonadotropic hypogonadism and defects in prohormone processing.
- In addition, mutations in the *PROP1* gene have resulted in absence of several pituitary hormones, including growth hormone, thyroid-stimulating hormone, prolactin, and gonadotropins. *PROP1* encodes a protein expressed in the embryonic pituitary, which is necessary for function of *POU1F1* (formerly *PIT1*), which codes for a pituitary transcription factor.
- In addition, mutation of the gene *HESX1* has been associated with **septo-optic dysplasia**, which may include poor development of the pituitary.



172. Brown tumor is seen in
- Hypothyroidism
  - Hyperthyroidism
  - Hyperparathyroidism
  - Hypoparathyroidism

**Brown tumors** occur, but these are less common with secondary hyperparathyroidism than they are with **primary hyperparathyroidism**. However, as the life expectancy of patients with chronic renal disease has increased, brown tumors have increasingly been identified with **renal osteodystrophy**. Brown tumors may occur in the spine and form expansile masses, which can be complicated by paraplegia. Brown tumors of the sellar and/or parasellar regions and face may appear as destructive lesions.



173. Not true about primary gouty arthritis is
- 90% cases are due to overproduction than undersecretion
  - Uric acid level may be normal in acute attack of gouty arthritis
  - Seen more in males
  - Synovial fluid aspiration is the most confirmatory test

Gout develops in the setting of excessive stores of uric acid in the form of monosodium urate. Uric acid is an end-stage by-product of purine metabolism. Lacking uricase, humans remove uric acid primarily by renal excretion. When excretion is insufficient to maintain serum urate levels below the saturation level of 6.8 mg/dL (with some variability depending on temperature and pH), hyperuricemia may develop, and urate can crystallize and deposit in soft tissues. **Ninety percent of patients with gout develop excess urate stores due to an inability to excrete sufficient amounts of normally produced uric acid in the urine (underexcretion).** The remaining patients either overconsume purines or produce excessive amounts of uric acid endogenously (overproduction).

- In rare cases, overproduction of uric acid is primary, due to a genetic disorder. These disorders include hypoxanthine-guanine phosphoribosyltransferase deficiency (**Lesch-Nyhan syndrome**), glucose-6-phosphatase deficiency (**von Gierke disease**), fructose 1-phosphate aldolase deficiency, and PP-ribose-P synthetase variants.
- Overproduction of uric acid may also occur in disorders that cause high cell turnover with release of purines. These disorders **include myeloproliferative and lymphoproliferative disorders, psoriasis, chemotherapy (tissue lysis), hemolytic anemias, pernicious anemia, ineffective erythropoiesis (as in B-12 deficiency), excessive exercise, and obesity.**
- Overproduction of uric acid can occur from overconsumption of **foods high in purines.**
- Common causes of secondary gout due to underexcretion of uric acid include renal insufficiency, lead nephropathy (saturnine gout), starvation or dehydration, hypothyroidism, hyperparathyroidism, drugs (including diuretics and cyclosporine A), and chronic ethanol (especially beer and hard liquor) abuse. These disorders should be identified and corrected, if possible.

**Individual gout flares** are often triggered by acute increases or decreases in urate levels that may lead to the production, exposure, or shedding of crystals that are not coated with apo B or apo E. This can result from **acute alcohol ingestion, acute overindulgence in foods high in purines, rapid weight loss, starvation, trauma, or hemorrhage.** Medications that increase uric acid levels via effects on renal tubular transport include **diuretics** and low-dose **aspirin.** Gout flares can also result from agents that lower levels of uric acid, including the use of **radiocontrast dyes** and medications such as **allopurinol** or **uricosurics.**

174. Gout is a disorder of
- Purine metabolism**
  - Pyrimidine metabolism
  - ATP metabolism
  - Urea metabolism

Gout develops in the setting of excessive stores of uric acid in the form of monosodium urate. Uric acid is an end-stage by-product of **purine metabolism.**

175. A boy presented with seizure and absent femoral pulse. Blood pressure in upper limb was 200/140 mmHg. Most likely diagnosis is
- Takayasu aortoarteritis**
  - Renal parenchymal ds

- c) GTCS
- d) Fibromuscular dysplasia

Despite the term pulseless disease, which is a synonym for Takayasu arteritis, the predominant finding in individuals with Takayasu arteritis is **asymmetric pulse**. **Absent peripheral pulses occur late in the course of the disease**. Although 5-year survival rates exceed 90%, the disease has a high incidence of residual morbidity.

## History

- Systemic symptoms in Takayasu arteritis (TA) include the following:
  - Fever, night sweats
  - Fatigue
  - Weight loss
  - Myalgia and/or arthralgia and/or arthritis
  - Skin rash (eg, erythema nodosum, pyoderma gangrenosum)
  - Headaches and/or dizziness and/or syncope
  - Congestive heart failure, palpitations, angina
  - **Hypertension** (may be paroxysmal)
- Symptoms related to ischemia include the following:
  - Ischemic stroke and/or **transient ischemic attack**
  - Visual disturbances (eg, blurred vision, diplopia, amaurosis)
  - Carotidynia
  - Abdominal pain
  - Claudications (vary due to the development of collateral circulations; symptom is rare in children)

## Physical

- Blood pressure difference greater than 30 mm Hg between arms
- **Asymmetric pulses**
- **Diminished or absent pulses** (midaortic lesions found in children may not affect pulses)
- Asymmetric pulses (common) and absent pulses (rare), even in the later stages of the disease (awareness of this is critical)
- Poststenotic dilatations producing what appear to be bounding pulses (often present)
- Hypertension (may be paroxysmal): Because this typically results from renovascular compromise, this is a high-renin hypertension.
- Bruits, especially over subclavian arteries or aorta
- Funduscopic examination
  - Retinal hemorrhages
  - Cotton-wool exudates
  - Venous dilatation and beading
  - Microaneurysms of peripheral retina
  - Optic atrophy
  - Vitreous hemorrhage

- Classic wreathlike peripapillary arteriovenous anastomoses (extremely rare)
- Reported skin lesions including erythema nodosum–like lesions, pyoderma gangrenosum, leukocytoclastic vasculitis, and panniculitis

**Extra Edge:**

Takayasu arteritis has been reported in identical twins, leading to hypotheses of a hereditary basis for disease. In Japan and Korea, Takayasu arteritis is associated with human leukocyte antigens (HLAs)-A10, B5, Bw52, DR2, and DR4. These associations have not been confirmed in Western studies. TA is associated with HLA-B22 in the United States.

176. Beck's triad is seen in
- a) Cardiac tamponade
  - b) Constrictive pericarditis
  - c) LVMI
  - d) Restrictive CMP

**Beck's triad**

Beck's triad was described by the thoracic surgeon Calude S. Beck in 1935. It's components are:

1. Distended neck veins
2. Distant heart sounds
3. Hypotension

i.e. rising venous pressure, falling arterial pressure, and decreased heart sounds found in the presence of cardiac tamponade.

177. Treatment of choice for GIST is
- a) Sorafenib
  - b) Imatinib
  - c) Gefitinib
  - d) Erlotinib

Imatinib mesylate is the only available drug that has made a significant impact in the treatment of GISTs.

**Selective tyrosine kinase inhibitors**

Imatinib mesylate (STI 571) is a selective tyrosine kinase inhibitor with action against mutant c-Kit as occurs in association with GISTs. It represents a breakthrough in antineoplastic drug therapy because it is targeted against a specific molecular derangement.

### Multikinase inhibitors

Elicit actions via multiple tyrosine kinase inhibitors implicated in tumor growth, pathologic angiogenesis, and metastatic progression.

#### Sunitinib (Sutent)

**Multikinase inhibitor** that targets several tyrosine kinase inhibitors implicated in tumor growth, pathologic angiogenesis, and metastatic progression. Inhibits platelet-derived growth factor receptors (ie, PDGFR-alpha, PDGFR-beta), vascular endothelial growth factor receptors (ie, VEGFR1, VEGFR2, VEGFR3), stem cell factor receptor (KIT), Fms-like tyrosine kinase-3 (FLT3), colony-stimulating factor receptor type 1 (CSF-1R), and the glial cell-line-derived neurotrophic factor receptor (RET). Indicated for persons with gastrointestinal stromal tumors (GISTs) whose disease has progressed or who are unable to tolerate treatment with imatinib (Gleevec). Delays median time to tumor progression.

178. Bence jones proteinuria is seen in
- Alpha heavy chain disease
  - Gamma heavy chain disease
  - Mu heavy chain disease**
  - Epsilon heavy chain disease

Heavy chain diseases (HCDs) are rare B-cell proliferative disorders characterized by the **synthesis and secretion of incomplete immunoglobulin heavy chains**. These disorders initially were recognized as **gammopathies** due to the presence of monoclonal proteins in the patient's serum or urine. The disorders were defined in terms of the production of structurally aberrant immunoglobulin molecules.

Normal immunoglobulin molecules are symmetrical and are composed of 2 pairs of polypeptide chains designated the light and heavy chains, which are interconnected by disulfide bonds. The heavy chains are the larger polypeptide subunits; they are specific and distinctive structures that distinguish the major classes of immunoglobulins. Reductive cleavage of the immunoglobulin molecule by papain yields 2 Fab fragments (consisting of a light chain and an Fd fragment) and one Fc fragment (consisting of portions of the 2 heavy chains). **Plasma cell disorders characterized by an anomalous serum and urinary protein that is immunochemically related to the Fc fragment of the immunoglobulin molecule are known as HCDs. When the anomalous protein structurally resembles the heavy chain fragment of immunoglobulin M (IgM) molecule, it is designated as mu-HCD.** Ballard and colleagues first described this entity in 1970.

Urinary excretion of the mu fragment has been noted in only 2 patients; this presumably is because the polymers of the carboxy-terminal mu fragment are too large to be filtered by intact renal glomeruli. **Monoclonal light chains have been found in the urine in two thirds of cases.** Thus, **Bence Jones proteinuria is a common occurrence in patients with this disorder.** Nonetheless, renal complications are infrequent. Immunoglobulin light chains capable of producing amyloid are found in approximately 12% of cases, an incidence that is similar to that observed in patients with multiple myeloma.

Mu heavy chain disease was first described in 1969. The characteristic feature of HCD is the production of a monoclonal immunoglobulin molecule in which the heavy chain is truncated and the covalent attachment of light chains is absent. This may be due to lack of light chain production or the failure of heavy-light disulfide bond formation.

179. A male presented with HBsAg antigen positive and HBeAg antigen negative, HBV DNA copies 100 per ml, and SGOT and SGPT elevated 6 times the upper limit of normal value. What is the likely diagnosis
- HBV surface mutant
  - HBV pre-core mutant
  - Wild HBsAg
  - Unaffected HBV carrier

*Table . Diagnostic Criteria for HBeAg-Negative CHB*

HBsAg	Positive for at least 6 months
HBeAg	Negative for at least 6-12 months
Anti-HBe*	Positive for at least 6-12 months
ALT	> 2 x ULN on 1 occasion or > 1.5 x ULN on 2 monthly occasions
Serum HBV-DNA**	Detectable by any assay
Exclusion of other causes of liver diseases	
Alcohol use	< 20 g/day
Anti-HDV	Negative
Anti-HCV	Negative
Liver biopsy	Active necroinflammatory activity compatible with CHB

\*Anti-HBe may be negative in a minority of patients with HBeAg-negative CHB

\*\*Some recommendations suggest that serum HBV-DNA levels should be > 10<sup>5</sup> copies/mL

ULN: upper limit of normal; Anti-HDV: antibody to hepatitis D virus; Anti-HCV: antibody to hepatitis C virus.

The serologic profile of patients with HBeAg-negative chronic HBV infection includes (1) positive serum HBsAg for at least 6 months, to establish chronic HBV infection; and (2) negative serum HBeAg and usually positive anti-HBe antibody for at least 6, or preferably 12, months, to exclude patients who are still in the unstable phase of HBeAg seroconversion and thus who may revert to the HBeAg-positive CHB phase. In such patients, the diagnosis of HBeAg-negative CHB can be made when the following criteria are fulfilled

1. Increased serum ALT level (ie, ALT levels > 2 x upper limit of normal [ULN] on 1 occasion, or ALT > 1.5 x ULN on at least 2 monthly determinations), as a biochemical marker of ongoing hepatocellular damage;
2. Detectable serum HBV DNA levels, to establish active HBV replication and, presumably, HBV-induced hepatocellular damage;
3. Exclusion of other concomitant or superimposed causes of liver disease; and
4. Moderate-to-severe necroinflammation on liver histology, compatible with CHB.

180. In Guillain Barre syndrome all of the following are seen except

- a) Albuminocytological dissociation
- b) Ascending paralysis
- c) Sensory level
- d) Flaccidity

GBS manifests as rapidly evolving motor paralysis with or without sensory disturbance. The usual pattern is an ascending paralysis that is first noticed as rubbery legs. – H17/2667

181. Lesion in the lateral cerebellum causes all except

- a) Incoordination
- b) Intention tremors
- c) Resting tremors
- d) Ataxia

Damage to the cerebellum produces many characteristic abnormalities including hypotonia, ataxia & intention tremor.

Resting tremors are seen in Parkinsonism.

182. A young male presented with meningococcal meningitis and allergy to penicillin. What would you administer

- a) Chloramphenicol
- b) Meropenem
- c) Ciprofloxacin
- d) Teicoplanin

Penicillin G is the DOC for treating meningococcal disease. Either Chloramphenicol or a 3<sup>rd</sup> generation cephalosporin like cefotaxime or ceftriaxone is used in persons allergic to penicillin.  
– Jawetz Medical Microbiology, Chapter 21

183. Prenatal diagnosis of hemophilia is done by

- a) RT-PCR
- b) Linkage analysis
- c) Microarray
- d) Cytometry

## Brief Reports

INDIAN PEDIATRICS 577 VOLUME 42\_JUNE 17, 2005

### Cord Blood Analysis for Prenatal Diagnosis of Thalassemia major and Hemophilia A

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Manuscript received: April 28, 2004, Initial review completed: August 3, 2004;

Revision accepted: November 16, 2004.

Beta-thalassemia and Hemophilia A are common genetic disorders for which prenatal diagnosis (PND) is an accepted option. Our aim was to evaluate cord blood analysis as a method for PND of these disorders. Cord blood samples at 18-26 weeks gestation from nine mothers with previous thalassemia major child and five families with previous hemophilia A were studied. In the former; HbF, HbA<sub>2</sub> and HbF were determined by high performance liquid chromatography (HPLC) and in latter; Factor VIII and IX assays were done by one stage method. In HPLC studies for thalassemia, three out of nine fetuses were affected, five were carriers and one was normal. In hemophilia PND samples, 2 out of five fetuses were affected. Thus, HPLC and factor VIII assay in cord blood are feasible alternatives for PND in  $\alpha$ -thalassemia and hemophilia A respectively, especially when DNA analysis is uninformative or there are financial constraints.

**Keywords:** Cordocentesis, Factor VIII, HPLC, Linkage analysis, Thalassemia major.

**B**ETA-THALASSEMIA in its severest form, thalassemia major is transfusion dependent. The common five Indian  $\beta$ - thalassemia mutations are  $\beta^0$  or severe  $\beta^+$  and account for 90% of mutations. However, molecular diagnosis is not possible in families at-risk especially with non-informative DNA analysis. HbA ( $\alpha_2\beta_2$ tetramer) is normal adult hemoglobin. The fetus who is  $\beta^0/\beta^0$ , or  $\beta^0$  / severe  $\beta^+$  or severe  $\beta^+$  / severe  $\beta^+$  is likely to have no HbA or marked reduction in HbA synthesis. Normal fetuses have demonstrable low levels of HbA in mid-trimester of pregnancy. This is further reduced or absent in fetus who carries  $\beta^0$  or severe  $\beta^+$  on both chromosomes. Quantification of HbA in cord blood is advocated as another option for prenatal diagnosis (PND). Similarly, hemophilia A is common bleeding disorder manifesting in males. The usual method for PND is by linkage analysis in chorionic villi samples. In severe hemophilia families, analysis of Intron 22 and Intron 1 inversion mutations can be tried with approximately 50% results. However, in families where linkage is not informative and direct mutational analysis is not possible, cord blood factor VIII assay can be used to detect affected cases. In present communication, we present our experience on cord blood analysis for above two disorders.

184. A patient presented with hypoglycemia and hepatomegaly. His low blood sugar levels is not rising even after epinephrine administration. What is the likely diagnosis ?
- Gaucher's disease
  - Anderson's disease
  - Pompe's disease
  - Von Gierke's disease
- Initial symptoms of neonatal hypoglycemia occur shortly after birth in patients with glycogen-storage disease type I (GSD I), and patients do not respond to glucagon administration. Symptoms include the following:
    - Tremors
    - Irritability
    - Cyanosis
    - Seizures
    - Apnea
    - Coma
  - Older infants may present with the following:
    - Frequent lethargy
    - Difficult arousal from overnight sleep
    - Tremors
    - Overwhelming hunger
    - Poor growth
    - Apparent increase in abdominal girth, although extremities appear thin
    - A doll-like facial appearance caused by adipose tissue deposition in the cheeks
  - Young children with glycogen-storage disease type Ia may experience nosebleeds.
  - Young children with glycogen-storage disease Ib may have frequent otitides, gingivitis, and boils.
  - Symptoms of severe hypoglycemia in patients of all ages are likely to follow any illness that causes mild anorexia or fasting (eg, viral gastroenteritis).
  - In middle childhood, patients may manifest evidence of rickets and anemia.
  - Patients with glycogen-storage disease Ib at all ages may be affected by a Crohnlite ileocolitis (pseudocolitis). The severity of the primary disorder is not correlated with the intestinal symptoms.
185. Plasmapheresis is done for all except
- Acute polymyositis
  - Cholinergic crisis
  - AIDP
  - Myasthenia crisis

Plasmapheresis/leukapheresis is not effective in Polymyositis & Dermatositis.

### Indications of Plasmapheresis

Disease	Definitions	Quality	Strength
Guillain-Barré syndrome—severe	Established	Class I	Type A
Chronic inflammatory demyelinating polyneuropathy	Established	Class I	Type A
Polyneuropathy with monoclonal gammopathies of undetermined significance:			
IgG/IgA	Established	Class I	Type A
IgM	Investigational	Class I	
Myasthenia gravis—Pre-op preparation and crisis	Established	Class III	Type C
Lambert-Eaton myasthenic syndrome	Possibly useful	Class II/III	
Refsum's disease	Investigational	Class III	
Acquired neuromyotonia	Investigational	Class III	
Stiff-man syndrome	Investigational	Class III	
Cryoglobulinemic polyneuropathy	Investigational	Class III	
Central nervous system systemic lupus	Investigational	Class III	
Acute disseminated encephalomyelitis	Investigational	Class III	
Multiple sclerosis	Possibly useful	Class I/II	

**Ref: Neurology 1996;47:840-843**

Based on the review of the literature, therapeutic PP has a definite role in the treatment of patients with GBS, CIDP, polyneuropathies associated with MGUS, MG, and LEMS. PP may have a role in treating patients with Refsum's disease, acquired neuromyotonia, stiff-man syndrome, cryoglobulinemic polyneuropathy, CNS-SLE, ADEM, and MS, but these decisions should be made on a case-by-case basis. PP has no role in treating patients with ALS or paraneoplastic syndromes with circulating autoantibodies.

186. Correct for hyponatremia is all accept

- In pseudohyponatremia, plasma osmolality is low
- In hyperglycemia associated hyponatremia, plasma osmolality is high
- NSAIDs ↑ potency of vasopressin
- SIADH causes euvolemic hyponatremia

Pseudohyponatremia refers to ↓ plasma sodium in presence of normal or ↑ plasma osmolality.

187. A 29 year old anxious apprehensive lady presented with a history of progressive breathlessness for last 6 months. Her FVC is 90% FEV1/FVC is 86%. On exercise for oxygen ratio dropped from 92% to 86%. There is no history of cough and fever. What is the likely diagnosis?
- Alveolar hypoventilation
  - Primary pulmonary HTN
  - MS
  - Anxiety disorder

188. Unconjugated hyperbilirubinemia with increased urobilinogen excretion is seen in
- G6PD
  - Hemolytic anemia
  - Hereditary spherocytosis
  - Biliary cirrhosis

Hemolytic anemia - ↑unconjugated bilirubin, ↑LDH, ↑Urobilinogen in both stool & urine, ↓Haptoglobin

189. A 30 year old female presented with complain of progressive breathlessness since 6 months. There was a history of intermittent pain and palpitation. On examination, her pulse was 88/m and BP was 140/80 mmHg and a late systolic murmur with mid systolic click was found at cardiac apex. Pathological examination of the heart is most likely to show which of the following in the patient?
- Fatty tissue replacement of ventricle with thinning of RV free wall
  - Hooding of mitral valve leaflets with myxomatous degeneration
  - Ischemia of papillary muscles
  - Ruptured chordate tendineae

MVP shows interchordal ballooning(hooding) of the mitral leaflets.

190. 3<sup>rd</sup> heart sound is seen in
- ASD
  - Constrictive pericarditis
  - Rapid filling during ventricular systole
  - AS

An S3 that is earlier & higher pitched than normal often occurs in pts with constrictive pericarditis.- H17/1386

191. Urea/ creatinine ratio of 20:1 can be seen in all except
- Pre-renal failure
  - Rhabdomyolysis
  - Post partum RF
  - Ureteric stones

$U/Cr > 20:1$  seen in **Pre-renal failure**. Ureteric stones cause post-renal failure.

192. Pre-renal Azotemia will have all except

- $FE_{Na} < 1$
- Urinary osmolality  $> 500$
- Urinary Na excretion  $> 40$**
- Urine flow not  $\uparrow$  on giving diuretics

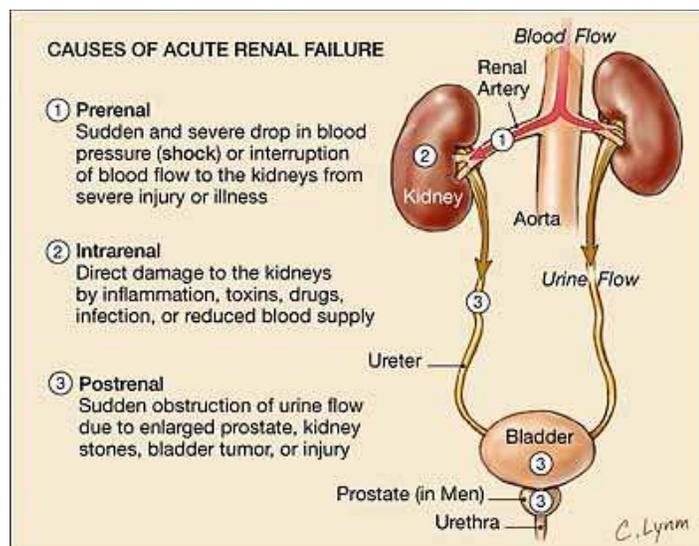
In pre-renal failure, **urinary Na is  $< 20$  mEq/L**.

**Table 1**

Guidelines for urinary indices whereby established ARF can be distinguished from renal vasoconstriction with intact tubular function (prerenal azotemia)

Laboratory test	Prerenal azotemia	ARF
Urine osmolality (mOsm/kg)	$> 500$	$< 400$
Urine sodium level (mEq/l)	$< 20$	$> 40$
Urine/plasma creatinine ratio	$> 40$	$< 20$
Fractional excretion of sodium (%)	$< 1$	$> 2$
Fractional excretion of urea (%)	$< 35$	$> 35$
Urinary sediment	Normal; occasional hyaline or fine granular casts	Renal tubular epithelial cells; granular and muddy brown casts

Osm, osmole; Eq, equivalent.



193. Not true about regarding Fanconi Anemia is
- Shows bone marrow atrophy
  - AD inheritance
  - Usually causes macrocytic anemia
  - Predisposes to malignancy

Fanconi anemia is an **autosomal recessive disease** in more than 99% of patients (FANCB is X-linked recessive); each patient with Fanconi anemia is homozygous or doubly heterozygous for mutations in one of the **13 genes** known to be responsible for FA. **The cloned genes are FANCA, B, C, D1, D2, E, F, G, I, J, L, M, and N.** Although most are unique genes, several were previously known, including *FANCD1* (*BRCA2*), *FANCG* (*XRCC9*), *FANCI* (*KIAA1794*), *FANCI* (*BRPI1/BACH1*), *FANCL* (*PHF9/POG*), *FANCM* (*Hef*), and *FANCN* (*PALB2*). Heterozygotes for *BRCA2* and possibly *BACH1* and *PALB2* are at increased risk of breast and other cancers.

About 75% of patients with Fanconi anemia **have birth defects, such as altered skin pigmentation and/or café au lait spots (>50%), short stature (50%), thumb or thumb and radial anomalies (40%), abnormal male gonads (30%), microcephaly (25%), eye anomalies (20%), structural renal defects (20%), low birth weight (10%), developmental delay (10%), and abnormal ears or hearing (10%).**

However, literature reports may be biased toward this association because the clinical diagnosis initially depended on the combination of **aplastic anemia** and physical anomalies; thus, the frequencies may be overestimated. Patients with biallelic mutations in *FANCD1/BRCA2* and *FANCI/PALB2* have a very severe phenotype, including features of the vertebral, anal, cardiac, tracheal, esophageal, and limb (VACTERL) association.

- Skin - Generalized hyperpigmentation on trunk, neck, and intertriginous areas; **café au lait spots**; hypopigmented areas
- Body - Short stature, delicate features
- Upper limbs
  - Thumbs - **Absent or hypoplastic**, supernumerary, bifid, rudimentary, short, low set, attached by a thread, triphalangeal, tubular, stiff, hyperextensible
  - Radii - **Absent or hypoplastic** (only with abnormal thumbs [ie, terminal defects]), absent or weak pulse
  - Hands - **Clinodactyly**, hypoplastic thenar eminence, 6 fingers, absent first metacarpal, enlarged abnormal fingers, short fingers
  - Ulnae - Dysplastic
- Gonads
  - Males - Hypogenitalia, undescended testes, hypospadias, **abnormal or absent testis**, atrophic testes, **azoospermia**, phimosis, abnormal urethra, micropenis, delayed development
  - Females - Hypogenitalia; bicornuate uterus; aplasia of uterus and vagina; **atresia of uterus, vagina, or ovary/ovaries**
- Other skeletal anomalies
  - Head and face - **Microcephaly**, hydrocephalus, micrognathia, peculiar face, **bird face**, flat head, frontal bossing, scaphocephaly, sloped forehead, choanal atresia
  - Neck - **Sprengel abnormality, short, low hairline, webbed**

- Spine - **Spina bifida** (thoracic, lumbar, cervical, occult sacral), scoliosis, abnormal ribs, sacroccocygeal sinus, Klippel-Feil syndrome, vertebral anomalies, extra vertebrae
- Feet - Toe syndactyly, abnormal toes, flat feet, short toes, **clubfoot, 6 toes**
- Legs - **Congenital hip dislocation**, Perthes disease, coxa vara, abnormal femur, thigh osteoma, abnormal legs
- Eyes - Small, strabismus, epicanthal folds, hypertelorism, ptosis, slanted, cataracts, astigmatism, blindness, epiphora, nystagmus, proptosis, small iris
- Ears - **Deaf** (usually conductive), abnormal shape, atresia, dysplasia, low-set, large, small, infections, abnormal middle ear, absent drum, dimples, rotated, canal stenosis
- Kidneys - Ectopic or pelvic, horseshoe, hypoplastic or dysplastic, absent, hydronephrosis or hydroureter, infections, duplicated, rotated, reflux, hyperplasia, no function, abnormal artery
- GI system - High-arch palate, atresia (eg, esophagus, duodenum, jejunum), imperforate anus, tracheoesophageal fistula, Meckel diverticulum, umbilical hernia, hypoplastic uvula, abnormal biliary ducts, megacolon, abdominal diastasis, Budd-Chiari syndrome
- Cardiopulmonary system - **Patent ductus arteriosus, ventricular septal defect**, peripheral pulmonic stenosis, aortic stenosis, coarctation, absent lung lobes, vascular malformation, aortic atheromas, atrial septal defect, tetralogy of Fallot, pseudotruncus, hypoplastic aorta, abnormal pulmonary drainage, double aortic arch, cardiomyopathy
- Other anomalies - Developmental delay, hyperreflexia, Bell palsy, CNS arterial malformation, stenosis of the internal carotid, small pituitary gland

### Causes

As described above, at least 13 genes are involved in the Fanconi anemia pathway. The exact link between mutations and phenotype is not clear, although patients who are homozygous for null mutations appear to have more severe Fanconi anemia than those with altered proteins. Various aspects of pathophysiologic research include the following:

- Fanconi anemia cells may be susceptible to damage by oxygen free radicals.
- Fanconi anemia cells have a defect in cell cycle regulation.
- The hematopoietic stem cell is defective in Fanconi anemia.
- A defect in the DNA-damage response pathway is present in Fanconi anemia.
- **Fanconi anemia is a premalignant disorder.**

### Laboratory Studies

- In Fanconi anemia (FA), CBC count may reveal trilineage pancytopenia or may only show **RBCs that are macrocytic for age**. Thrombocytopenia or leukopenia may precede full-blown aplasia.
- Chromosome breakage is usually examined in short-term cultures of peripheral blood T-cell mitogen-stimulated lymphocytes in the presence of DNA cross-linkers, such as DEB or MMC. These agents lead to increased numbers of breaks, gaps, rearrangements, and quadriradii in Fanconi anemia homozygote cells. Some patients may have hematopoietic somatic mosaicism, with correction of the Fanconi anemia defect in the blood. In these cases, skin fibroblasts may be needed for the chromosome breakage test.
- Flow cytometry of Fanconi anemia cells cultured with nitrogen mustard and other clastogens demonstrates an arrest in G2/M.
- Fetal hemoglobin (HbF) may be increased for age as a manifestation of stress erythropoiesis.

- Red cell adenosine deaminase (ADA) is increased in most patients with Diamond-Blackfan anemia (DBA) but appears to be normal in Fanconi anemia.
- Serum erythropoietin (Ep) levels are markedly increased and higher than expected for the degree of anemia, similar to that observed in DBA. However, levels may be low in patients with impaired renal function.
- **Bone marrow aspirate and biopsy may reveal hypocellularity**, loss of myeloid and erythroid precursors and megakaryocytes (with relative lymphocytosis), or full-blown aplasia with a fatty marrow. Signs of myelodysplastic syndrome include dyserythropoiesis (multinuclearity, ringed sideroblasts), dysmyelopoiesis (hyposegmentation, hypogranularity, hypergranularity), and hypolobulated or hyperlobulated megakaryocytes. Presence of a cytogenetic clone in a high and increasing proportion over time may suggest an evolution to leukemia, but this is currently unproven.

	<p>A 3-year-old patient with Fanconi anemia. Note the multiple birth defects, including short stature, microcephaly, microphthalmia, epicanthal folds, dangling thumbs, site of ureteral reimplantation, congenital dislocated hips, and rocker bottom feet.</p>
	<p>Thumbs attached by threads on a 3-year-old patient with Fanconi anemia</p>

**Physical Abnormalities in Fanconi's Anemia**

Abnormality	Percentage
Hyperpigmentation and, café au lait spots	77
Thumb anomalies	37
Other skeletal anomalies	29
Microsomy (small stature)	60
Low birth weight	56
Microcephaly	40
Renal anomalies	28
Hypogenitalism	24
Strabismus	22
Microophthalmia	16
Hyperreflexia	19
Mental deficiency	17
Ear anomalies and, or deafness	7
Congenital heart disease	6

194. A young adult male presented with focal seizures, fever and irritable mood. MRI showed frontal and temporal lobe enhancements. What is the likely diagnosis?
- Meningococcal meningitis
  - TB meningitis
  - Herpes simplex encephalitis
  - Japanese encephalitis

## History

Herpes simplex encephalitis (HSE) is an acute or subacute illness, causing both general and focal signs of cerebral dysfunction. It is sporadic and occurs without a seasonal pattern. Although the presence of fever, headache, behavioral changes, confusion, focal neurological findings, and abnormal CSF findings are suggestive of HSE, no pathognomonic clinical findings reliably distinguish HSE from other neurological disorders with similar presentations (eg, non-HSV encephalitis, brain abscess, tumor). Confirmation of the diagnosis depends on the identification of HSV in the CSF by means of a polymerase chain reaction (PCR) or on the identification of HSV in brain tissue by means of brain biopsy (see [Workup](#)).

Typical symptoms include the following:

- Fever (90%)
- Headache (81%)
- Psychiatric symptoms (71%)
- Seizures (67%)
- Vomiting (46%)
- Focal weakness (33%)
- Memory loss (24%)

## Physical

The most frequent findings on physical examination are fever and mental status abnormalities. Meningismus is uncommon.

Typical findings on presentation include the following:

- Alteration of consciousness (97%)
- Fever (92%)
- Dysphasia (76%)
- Ataxia (40%)
- Seizures (38%)
  - Focal (28%)
  - Generalized (10%)
- Hemiparesis (38%)
- Cranial nerve defects (32%)
- Visual field loss (14%)
- Papilledema (14%)

A causal or temporal relationship between peripheral lesions (eg, herpes labialis) and HSE does not exist. Also, many febrile diseases may precipitate herpes labialis. Therefore, the presence or absence of such lesions neither confirms nor excludes the diagnosis.

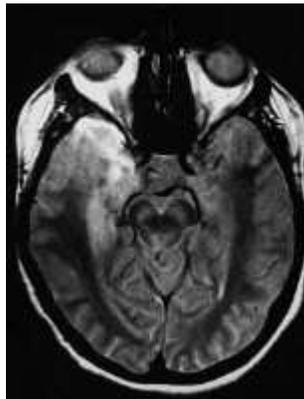
### Causes

- In children older than 3 months and in adults, **HSV-1** is responsible for virtually all cases.
- HSV-2 causes a small number of cases, particularly in the immunocompromised host.

### Imaging Studies

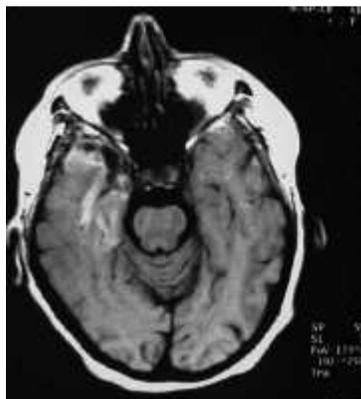
- Magnetic resonance imaging
  - **MRI of the brain is the preferred imaging study.** Abnormalities are found in 90% of patients with HSE; MRI may be normal early in the course of illness. The MRIs below show findings consistent with herpes simplex encephalitis.

○



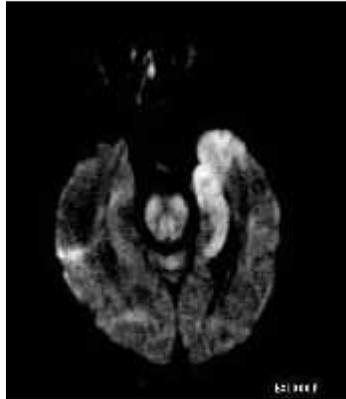
Axial proton density-weighted image in a 62-year-old woman with confusion and herpes encephalitis shows **T2 hyperintensity involving the right temporal lobe.**

○



Axial gadolinium-enhanced T1-weighted image reveals enhancement of the **right anterior temporal lobe** and parahippocampal gyrus. At the right anterior temporal tip is a hypointense, crescentic region surrounded by enhancement consistent with a small epidural abscess.

○



Axial diffusion-weighted image reveals **restricted diffusion in the left medial temporal lobe** consistent with herpes encephalitis. This patient also had a positive result on polymerase chain reaction assay for herpes simplex virus, which is both sensitive and specific. In addition, the patient had periodic lateralized epileptiform discharges on EEG, which supports the diagnosis of herpes encephalitis.

- **Findings of localized temporal abnormalities are highly suggestive of HSE**, but confirmation of the diagnosis depends on the identification of HSV by means of PCR or brain biopsy.
- MRI can noninvasively establish many of the potential alternative diagnoses of HSE.
- Computed tomography
  - Head CT may show changes in the temporal and/or frontal lobe, but CT is less sensitive than MRI.
  - Approximately one third of patients with HSE have normal CT findings on presentation.

195. Peripheral artery disease, CHD, stroke are associated with which hormone?

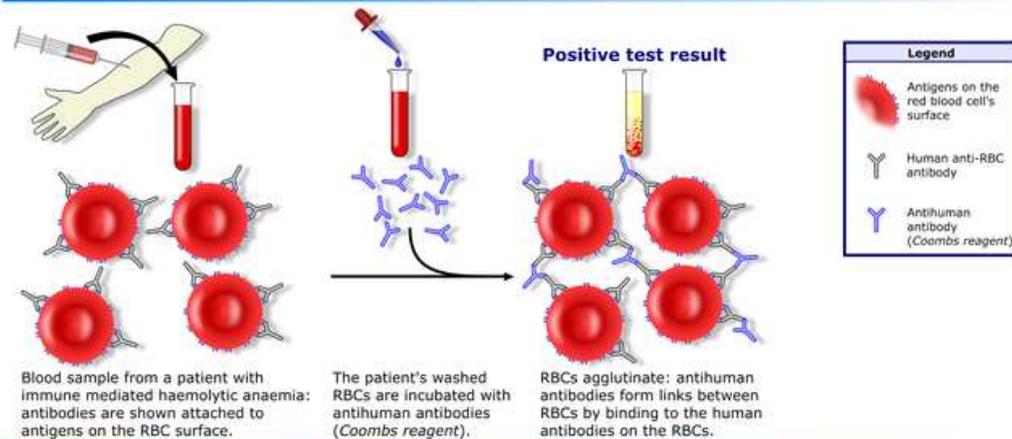
- a) **Insulin deficiency**
- b) Hyperestrogenemia
- c) Hypothyroidism
- d) Progesterone

Symptoms given in question are **chronic complications of DM**.

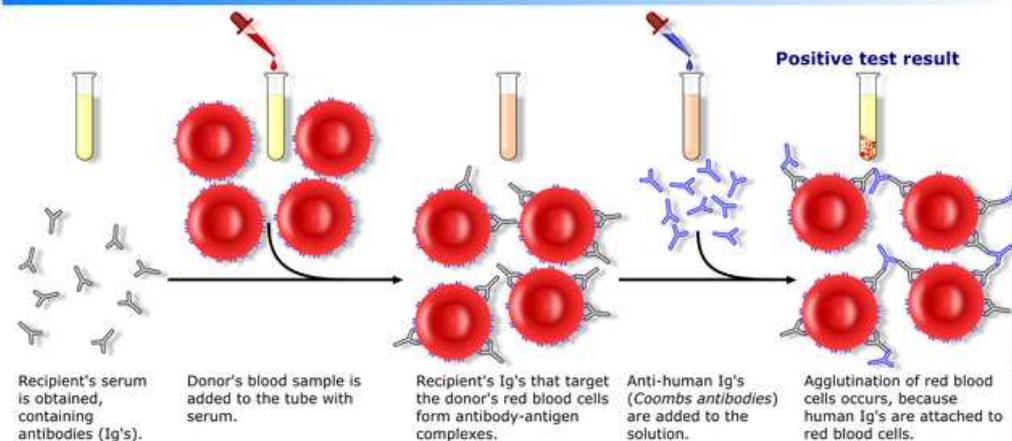
196. Not associated with Thymoma is
- SIADH
  - Myasthenia gravis
  - Red cell aplasia
  - Hypogammaglobulinemia
197. Non-matched blood transfusion is given, what immediate investigation should be done?
- Indirect Coombs test
  - Direct Coombs test
  - Antibody in pt serum
  - Antibody in donor serum

The direct Coombs test (also known as the **direct antiglobulin test** or DAT) is used to detect if **antibodies or complement system factors have bound to RBC surface antigens *in vivo***. The DAT is not currently required for pre-transfusion testing but may be included by some laboratories.

#### Direct Coombs test / Direct antiglobulin test



#### Indirect Coombs test / Indirect antiglobulin test



198. A 17 year old female presented with pain in abdomen and diarrhea at the time of menses and stress. There was mucus but no blood in stool, no weight loss and she remain predominantly constipated during rest of the time. Colonoscopy was normal. What is the likely diagnosis?
- Irritable bowel syndrome
  - Inflammatory bowel disease
  - Endometriosis
  - Giardiasis

Manning and associates established 6 criteria to distinguish irritable bowel syndrome from organic bowel disease. Although historically important, these criteria are insensitive (58%), nonspecific (74%), and less reliable in men. The Manning criteria to distinguish irritable bowel syndrome from organic disease are as follows:

- Onset of pain associated with more frequent bowel movements
- Onset of pain associated with looser bowel movements
- Pain relieved by defecation
- Visible abdominal bloating
- Subjective sensation of incomplete evacuation more than 25% of the time
- Mucorrhea more than 25% of the time

More recently, a consensus panel created and then updated the Rome criteria to provide a standardized diagnosis for research and clinical practice. The Rome III criteria (2006) for the diagnosis of irritable bowel syndrome require that patients must have recurrent abdominal pain or discomfort at least 3 days per month during the previous 3 months that is associated with 2 or more of the following:

- Relieved by defecation
- Onset associated with a change in stool frequency
- Onset associated with a change in stool form or appearance

Supporting symptoms include the following:

- Altered stool frequency
- Altered stool form
- Altered stool passage (straining and/or urgency)
- Mucorrhea
- Abdominal bloating or subjective distention

Four bowel patterns may be seen with irritable bowel syndrome. These patterns include IBS-D (diarrhea predominant), IBS-C (constipation predominant), IBS-M (mixed diarrhea and constipation), and IBS-A (alternating diarrhea and constipation). The usefulness of these subtypes is debatable. Notably, within 1 year, 75% of patients change subtypes, and 29% switch between constipation-predominant IBS and diarrhea-predominant IBS.

199. A patient with atrial fibrillation developed left upper limb paresis which slowly recovered after 2 months. What could be the cause ?
- Ischemic stroke
  - TIA
  - Hemorrhagic stroke
  - MI
200. Not a feature of APLA syndrome
- Single titre of anticardiolipin is diagnostic
  - Can present with recurrent fetal loss
  - Warfarin given as Tt
  - Rarely pulmonary HTN can develop

At least 2 +ve tests for APLA are required at least 12 wks apart.

Evaluate for lupus anticoagulant (LAC). At least 2 assays need to be performed, and at least one should contain a phospholipid-dependent step. If results are positive for LAC, a 4:1 or 3:1 (patient-to-normal) plasma mix test should be performed to correct for any coagulation factor deficiencies but not dilute out a low-titer antiphospholipid antibody.

- Dilute Russell Viper venom test (dRVVT)
- Hexagonal-phase LAC test
- Activated partial thromboplastin time (aPTT)
- Platelet neutralization procedure (PNP)
- Kaolin clotting time (KCT) or the Kaolin clot inhibition test
- Dilute prothrombin time (dPT)
- Textarin time (TT)
- Taipan snake venom time (TSVT)

CHART 4: Classification and detection of antiphospholipid antibodies

Antibody	Method of detection
Lupic anticoagulant antibody	<p><i>First stage:</i> prolonging of clotting in at least one <i>in vitro</i> phospholipid-dependent clotting test with the use of platelet-poor plasma. Tests can be subdivided according to coagulation cascade. Extrinsic pathway (diluted prothrombin time), intrinsic pathway (aPTT, diluted aPTT, silica-coloidal clotting time and kaolin clotting time), common on final pathway (dilute Russell's viper venom time, Taipan venom time, and textarin and ecarine time)</p> <p><i>Second stage:</i> failure in correcting prolonged clotting time by mixing normal patient plasma</p> <p><i>Third stage:</i> confirmation of presence of lupic anticoagulant antibody by shortening or correction of prolonging of clotting time after the addition of excess phospholipids or platelets.</p> <p><i>Fourth stage:</i> exclusion of coagulopathies by using assays for specific factors if confirming tests are negative or if a specific factor is suspected.</p>
Anticardiolipin antibody	<p>Solid-phase Immunoassay (generally ELISA) is formed by cardiolipin attached to wheels, generally in the presence of bovine serum <math>\beta_2</math>-glucoprotein.</p> <p>Anticardiolipin antibodies of patients with APS are <math>\beta_2</math>-glucoprotein I-dependent; antibodies of patients with infectious diseases are <math>\beta_2</math>-glucoprotein I-independent.</p>
Anti $\beta_2$ -glucoprotein I antibody	<p>Solid-phase Immunoassay (generally ELISA) is formed by human <math>\beta_2</math>-glucoprotein attached to wheels.</p>

Source: Levine JS et al.<sup>34</sup>

**Table – Revised classification criteria for the antiphospholipid antibody syndrome**

For antiphospholipid antibody syndrome to be diagnosed, at least 1 clinical and 1 laboratory criterion must be met.

**Clinical criteria**

*Vascular thrombosis:* One or more clinical episodes of arterial, venous, or small-vessel thrombosis in any tissue or organ, which must be validated by imaging studies or histopathology.

*Pregnancy morbidity:*

- One or more unexplained deaths of a morphologically normal fetus beyond the 10th week of gestation or
- One or more premature births of a morphologically normal neonate before the 34th week of gestation due to preeclampsia, eclampsia, or placental insufficiency or
- Three or more unexplained consecutive spontaneous abortions before the 10th week of gestation, with maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal causes excluded.

**Laboratory criteria**

(Each must be present on 2 or more occasions at least 12 weeks apart.)

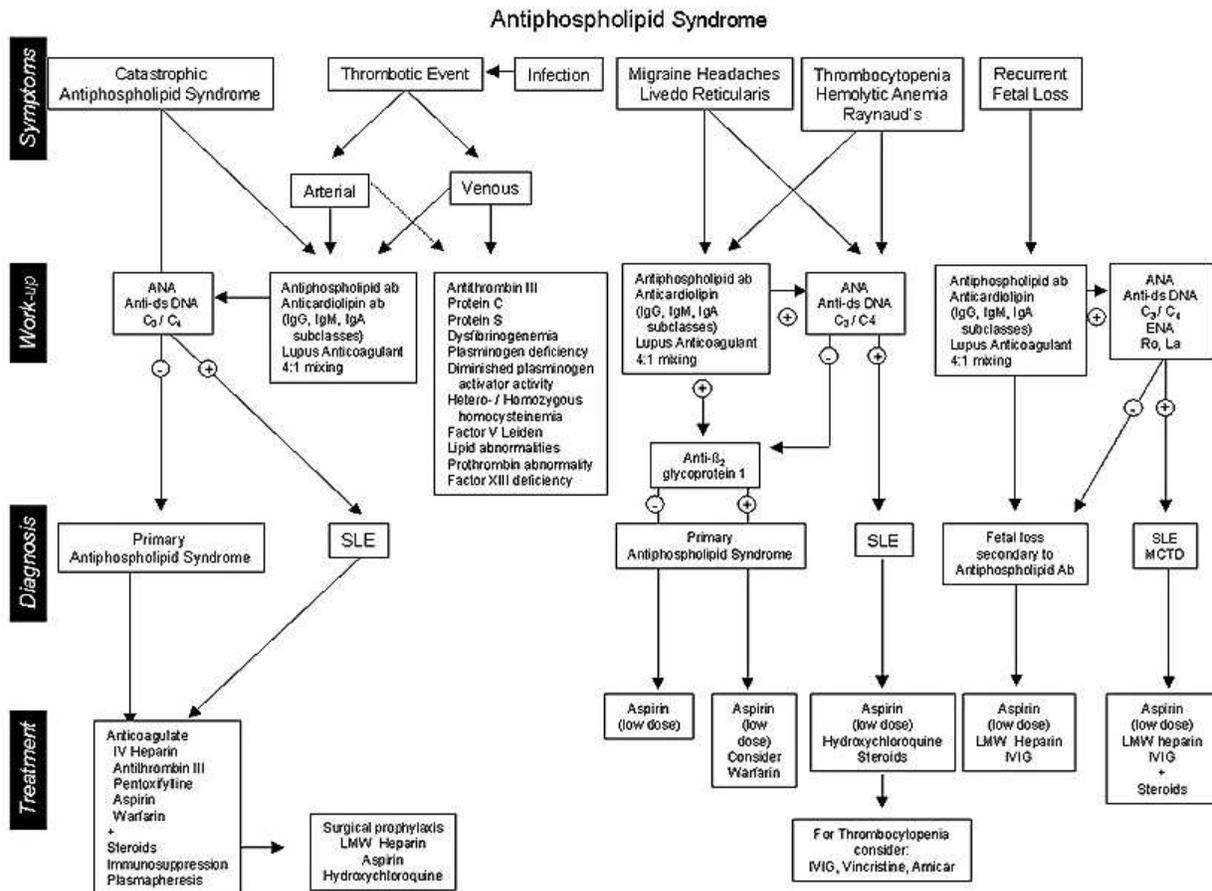
*Lupus anticoagulant:* Detected in plasma according to the guidelines of the International Society on Thrombosis and Hemostasis.

*Anticardiolipin antibody:* IgG or IgM isotype present in a medium or high titer (> 40 GPL or > 99th percentile), measured by ELISA.

*Anti-β<sub>2</sub> glycoprotein I antibody:* IgG or IgM isotype in high titer (> 99th percentile), measured by ELISA.

GPL, IgG phospholipid; ELISA, enzyme-linked immunosorbent assay.

Adapted from Miyakis S et al. *J Thromb Haemost.* 2006.<sup>4</sup>



201. A patient 50 year old comes with weakness and lethargy for 5-6 months. On examination he is found anemic and occult blood in stools is positive. What is the next best investigation for him?
- a) Colonoscopy
  - b) Ba meal
  - c) Ba enema
  - d) CT abdomen

Colonoscopy is the accurate & most widely used modality to evaluate the large bowel in pts with occult bleeding.

202. In a patient with head injury, damage to brain is aggravated by
- a) Hyperglycemia
  - b) Hypothermia
  - c) Hypercapnia
  - d) ↓osmolarity

Fever & hyperglycemia both worsen experimental ischemia & have been associated with worsened clinical outcome after stroke & head trauma. – H17/1722

# SURGERY

203. A patient presented with ITP and platelet count was 50,000 / cu.mm. Splenectomy was planned as treatment. What is the best to give platelet transfusion to patient?
- A day before surgery
  - Just before giving skin incision
  - After ligation of splenic artery
  - 24 hrs after surgery
204. Most common site of urethral carcinoma is
- Bulbomembranous urethra
  - Penile urethra
  - Prostatic urethra
  - External urethral meatus
205. Lord's placation is done for
- Inguinal hernia
  - Testicular cancer
  - Hydrocele
  - Testicular varices
206. Best time of surgery for undescended testis is
- 12 months
  - 24 months
  - 6 months
  - Just after birth
207. Stone hard to break
- Ca oxalate monohydrate
  - Ca oxalate dehydrate
  - Uric acid
  - Struvite stone
208. A 27 year old patient presented with left sided abdominal pain to ER 6 hrs after RTA. He was hemodynamically stable and FAST positive. CECT showed leak of contrast from spleen along with grade III laceration in it. What will be the most appropriate treatment
- Splenectomy
  - Splenorrhaphy
  - Splenic artery embolization
  - Conservative mgmt

*Both B & C appear to be correct. To choose one, C > B. Read the text below & decide yourself.*

### **Interventional radiology**

Splenic angioembolization is increasingly being used in both stable responders and transient responders for fluid resuscitation under constant supervision by a surgeon with an operating room on standby. Femoral artery access with embolization of the splenic artery or its branches can be accomplished with gel foam or metal coils. Such treatment requires intimate cooperation between the trauma surgeon and the interventional radiologist. Not all hospitals will have the proper facilities for such treatment, and any surgeon contemplating splenic angioembolization for a patient should first make sure the hospital interventional radiology suite and personnel are set up for rapid response at any hour of the day.

Failure of conservative treatment involves grade III, IV, or V injuries more often than grade I and II injuries. In many studies, SA embolization (SAE) has been described by using many different approaches. One primary point of discussion concerns the differences between main SAE, selective or superselective SAE, and embolization in a combination of sites. To the authors' knowledge, no studies have been performed to compare outcomes or complication rates based on the various levels of SAE. Theoretically, the infarction rate is expected to increase as embolization becomes more selective. Conversely, therapeutic failure is expected to increase with more proximal SAE. This presumption is based on collateral blood flow differences inherent with the particular level of embolization.

Sclafani et al and Hagiwara et al have described SAE techniques dependent on angiographic findings. The visualization of extrasplenic extravasation was treated with selective Gelfoam embolization or superselective gelatin sponge particle injection, respectively, followed by main SAE by means of coil occlusion. Main SAE alone was performed if intraparenchymal contrast-material extravasation was the only finding. Hagiwara et al also selected an additional group of patients whose angiograms demonstrated vascular disruption without extravasation. This group also was treated by using main SAE alone. SAE was not performed if the angiogram showed only avascular areas or evidence of subcapsular hematoma without extravasation. The overall success rate in the 2 studies was greater than 90%.

The authors contend that main SAE is effective because the decreased splenic blood pressure promotes hemostasis at the injury site. This effect is accomplished with low risk of infarction, because postembolization celiac angiograms demonstrated reconstitution of the SA distal to the coil occlusion via collateral flow in all patients. In fact, Sclafani et al reported continued contrast-agent extravasation in most patients after SAE. The persistence of extravasation immediately after SAE was not a poor prognostic indicator in the study, which differs from the data from the study of Hagiwara et al in which the single patient reported to have extravasation on the post-SAE angiogram was 1 of 2 patients in whom therapy failed and who eventually required splenectomy. The reason for this discrepancy is not clear.

### **Surgical Therapy**

Surgical therapy is usually reserved for patients with signs of ongoing bleeding or hemodynamic instability. In some institutions, CT scan-assessed grade V splenic injuries with stable vitals may be observed closely without operative intervention, but most patients with these injuries will undergo an exploratory laparotomy for more precise staging, repair, or removal. Adult surgeons may be more likely to operate in cases of splenic injury but less likely to transfuse than their pediatric surgical colleagues.

209. A lady presented in ER with stab injury to left side of abdomen. She was FAST positive CECT showed 2 cm left liver lobe laceration. She was hemodynamically stable. Laparoscopic surgery was planned. As soon as the pneumoperitoneum was created, the PO<sub>2</sub> of the patient suddenly dropped. What could be the possible cause?
- Air embolism through splenic artery
  - Rent in left lobe of diaphragm
  - IVC compression
  - Injury to colon

**Pts in favour of diaphragmatic rent:**

- H/O stab injury
  - Accentuated by pneumoperitoneum
  - Sudden drop in PO<sub>2</sub>
210. In orthotopic liver transplantation, which is the best way to get bile drainage in donor liver?
- Donor bile duct with recipient bile duct or Roux en Y choledochojejunostomy
  - Donor bile duct with duodenum of recipient
  - Donor bile duct with jejunum of recipient
  - External drainage for few days followed by choledochojejunostomy

Bile duct anastomoses are performed in succession, the last by end-to-end suturing of the donor & recipient CBDs or by choledochojejunostomy to a Roux-en-Y loop if the recipient CBD can't be used for reconstruction. – H17/1986

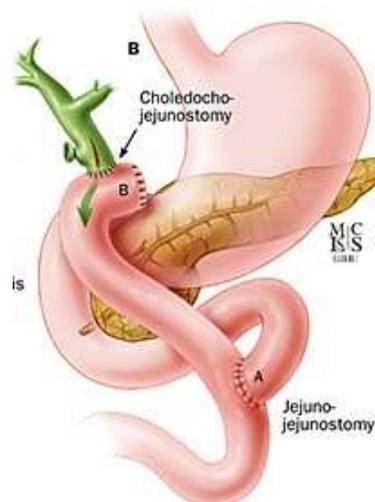
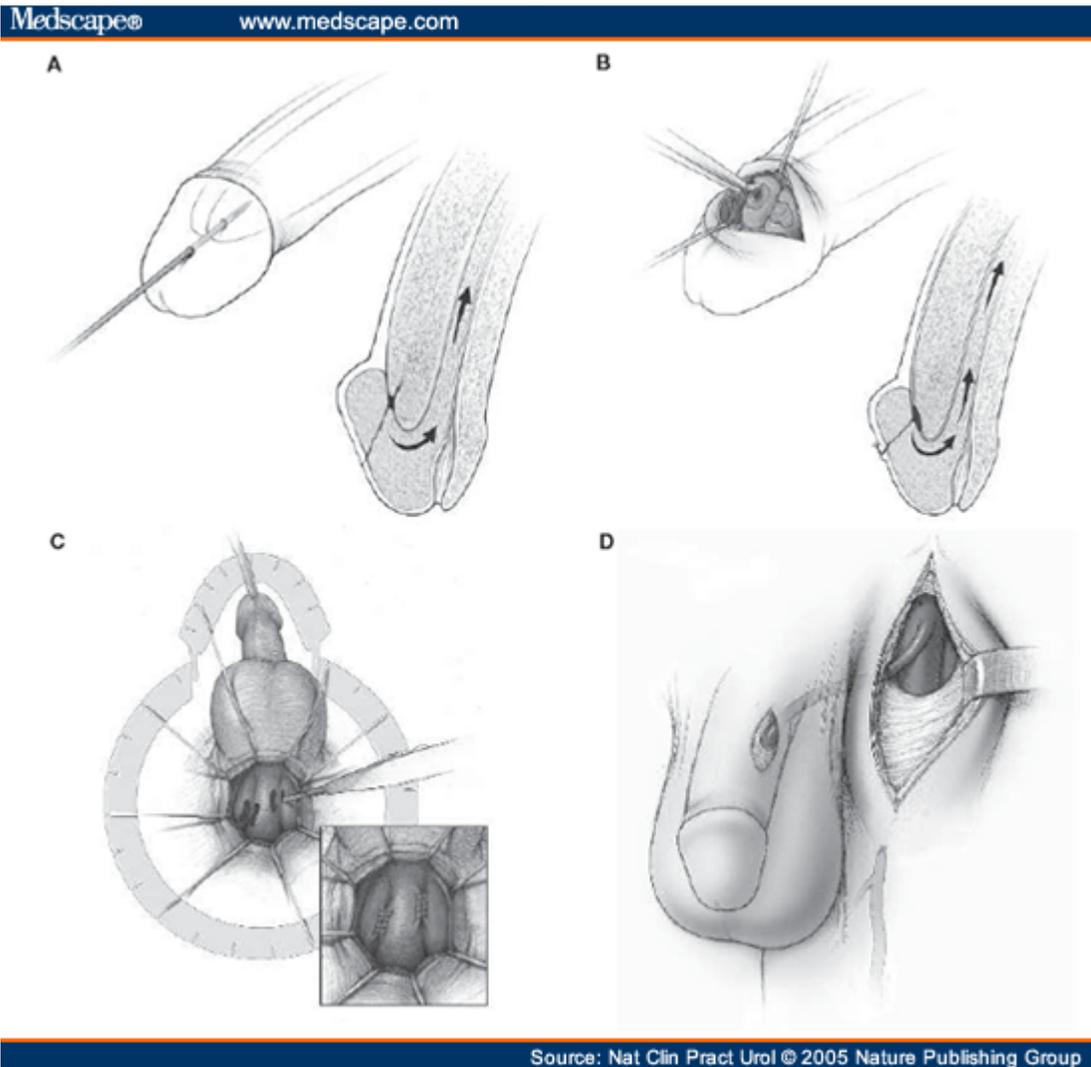


Fig. Choledochojejunostomy

211. Grayhack shunt is
- Between corpora cavernosa & saphenous vein
  - Between corpora cavernosa & corpora spongiosa
  - Between corpora cavernosa & dorsal vein
  - Between corpora cavernosa & glans penis

See the figure and the answer will be clear to you.



Source: Nat Clin Pract Urol © 2005 Nature Publishing Group

#### Surgical shunts for the treatment of priapism.

Distal cavernoglanular (corporoglanular) shunting is represented as (A) Winter and (B) El-Ghorab shunt procedures.

Proximal cavernospongiosal (corporospongiosal) shunting is represented as (C) Quackels/Sacher and (D) Grayhack shunt procedures.

**Surgery.** Surgical intervention is commonly applied at the discretion of the surgeon once it is apparent that intracavernous treatment has failed. Surgical shunting as a means for blood drainage from the corpora cavernosa involves either distal or proximal approaches (see figure above). Distal

cavernoglanular (corporoglanular) shunting is usually performed first because it is less invasive and carries a lower risk of complications than proximal shunting. Distal shunting techniques include placing a large biopsy needle (**Winter shunt**) or scalpel (**Ebbehoj shunt**) percutaneously through the glans, or excising the tunica albuginea at the tip of the corpus cavernosum (**El-Ghorab shunt**). If distal shunting fails, proximal cavernospongiosal (corporospongiosal) shunting can be used. This involves the creation of a window between the respective corporal bodies (**Quackels or Sacher shunt**), or an **anastomosis of a saphenous vein to one of the corpora cavernosa (Grayhack shunt)**.

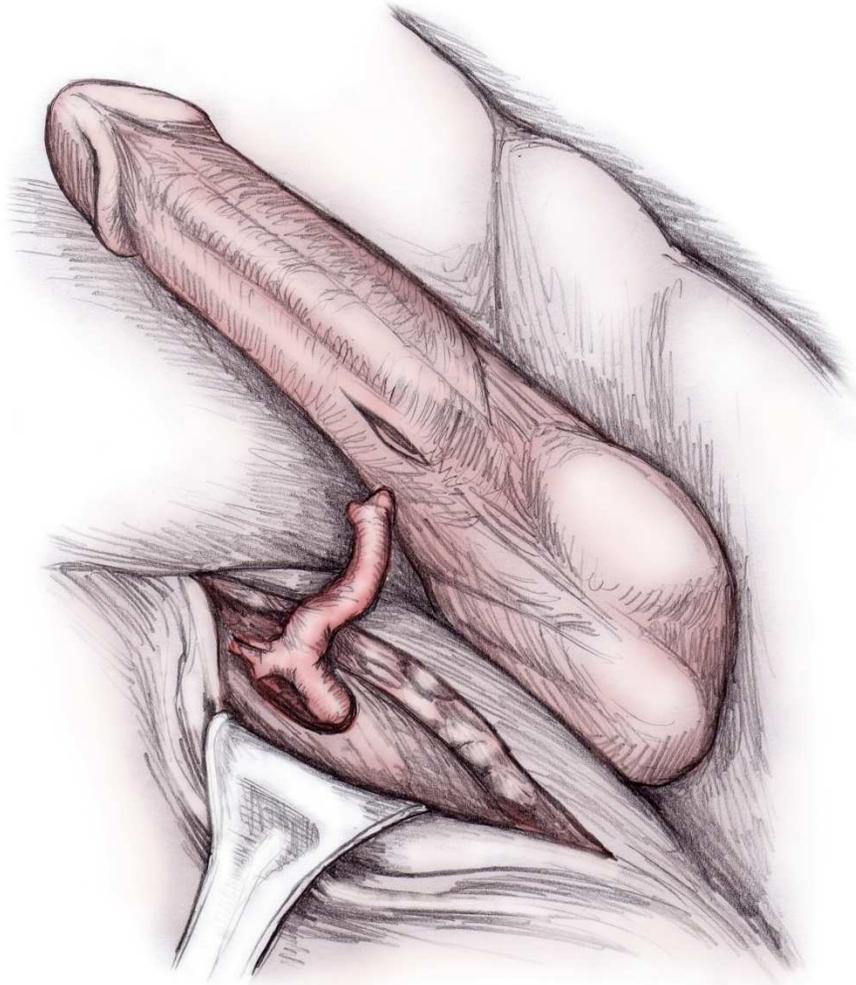


Fig. **Grayhacks shunt** - Proximal cavernosal-saphenous shunt (**Grayhack shunt**) surgically connects the proximal corpora cavernosum to the saphenous vein.

## Surgical Care

- A transglanular to corpus cavernosal scalpel or needle-core biopsy (**Ebbehoj or Winter technique**) is the first reasonable approach for refractory cases (see image below). A unilateral shunt is often effective. Bilateral shunts are used only if necessary (usually apparent after 10 min).

○



**Priapism. Winter shunt** placed by biopsy needle, usually under local anesthetic.

- The **El-Ghorab procedure** is a more aggressive open surgical cavernosal shunt and is indicated if the Winter shunt fails.
- **Quackel shunts** are cavernosal-spongiosum shunts (unilateral or bilateral) and are performed via a perineal approach (see image below). Such shunts are rarely effective if a more distal shunt has already failed (eg, El-Ghorab procedure) because thrombosis of the corpora is usually already present.

○



**Priapism. Proximal cavernosal-spongiosum shunt (Quackel shunt)** surgically connects the proximal corpora cavernosa to the corpora spongiosum.

- A **Grayhack shunt** is a **cavernosal-saphenous vein shunt** (rarely necessary or indicated; see image below).

○



**Priapism. Proximal cavernosal-saphenous shunt (Grayhack shunt) surgically connects the proximal corpora cavernosum to the saphenous vein.**

212. TRISS includes
- GCS
  - GCS+BP+RR
  - RTS+ISS+Age
  - RTS+Age

### Trauma Score - Injury Severity Score : TRISS

TRISS determines the probability of survival ( $P_s$ ) of a patient from the ISS and RTS using the following formulae:

$$P_s = 1 / (1 + e^{-b})$$

Where 'b' is calculated from:

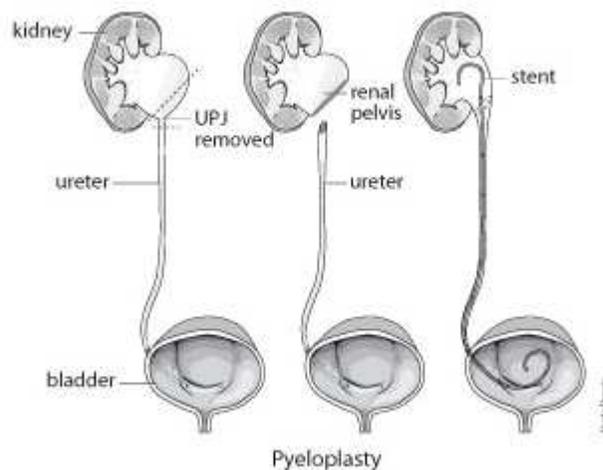
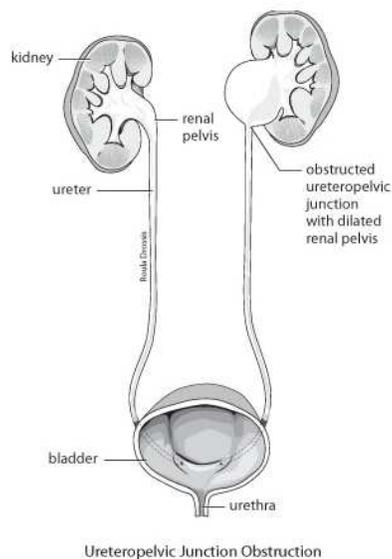
$$b = b_0 + b_1(RTS) + b_2(ISS) + b_3(AgeIndex)$$

The TRISS calculator determines the probability of survival from the ISS, RTS and patient's age. ISS and RTS scores can be inputted independently or calculated from their base parameters.

213. A child presented with pain in left flank. USG showed hydronephrosis in right kidney and 10 mm cortical thickness. Kidney differential fraction was 19%. Treatment of choice is
- Nephrectomy
  - Pyeloplasty
  - External drainage
  - Endopyelostomy

Open pyeloplasty remains the criterion standard for the treatment of UPJ obstruction. Although many variations exist in the methodology, this procedure typically involves the surgical excision of the narrowed segment of the UPJ and performance of a spatulated reanastomosis of the renal pelvis to the ureter. If significant dilation of the renal pelvis occurs, it is often reduced in size by trimming off redundant tissue, and then it is tailored in such a fashion that it funnels down towards the anastomosis. If an accessory or aberrant vessel exists near the UPJ, the anastomosis is positioned anterior to the vessel.

Traditionally, PUJ obstruction has been repaired with an operation called a **pyeloplasty**. Under general anaesthesia, an incision is made in the flank through which the renal pelvis and ureter are exposed. The narrow PUJ is cut out or cut open after which a wider connection is constructed. A temporary stent or nephrostomy tube may be placed. Patients may be in hospital for up to seven days and able to resume their usual activities within four to six weeks. This operation is successful in about 90% of cases.



214. A 50 year old female admitted for chronic abdominal pain with anuria. Radiological studies reveled bilateral impacted ureteric stones with hydronephrosis. Urine analysis showed RBCs with few pus cell in urine. Serum creatinine level was 16 mg/dl and urea level was 200 mmol/L. p H was 7.1. Which of the following should be immediate treatment
- Hemodialysis
  - J stent drainage
  - Lithotripsy
  - Ureteroscopic removal of stones

High urea & Cr along with low p H warrants immediate hemodialysis.

**Indications of immediate hemodialysis** (Mnemonic-AEIOU)

A-Acidosis

E-Electrolyte imbalance(e.g.hyperkalemia)

I-Intoxication

O-Overload(Fluid)

U-Uremia & its complications(e.g. pericarditis, encephalopathy,etc.)

215. What complication should one expect when PCNL is done through 11<sup>th</sup> intercostals space?
- Hydrothorax
  - Hematuria
  - Damage to colon
  - Remnant fragments

*BJU International*

(2002),

90,

809-813

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809

doi:10.1046/j.1464-4096.2002.03051.x

Accepted for publication 5 September 2002

## Prospective evaluation of safety and efficacy of the supracostal approach for percutaneous nephrolithotomy

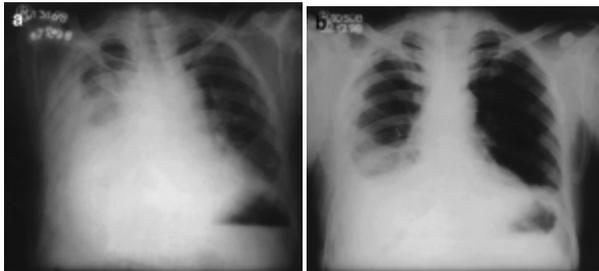
R. GUPTA, A. KUMAR, R. KAPOOR, A. SRIVASTAVA and A. MANDHANI

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The major complications of supracostal access are related to the potential for injury to the pleura and/or to the lung. Therefore a thorough knowledge of the anatomical relationships of the diaphragm, pleura

and lung is important to avoid this risk. The diaphragm is attached to the inferior border of the 12th rib, transverse process of the first lumbar vertebra and the anterior surfaces of the upper lumbar vertebra bodies. The parietal pleura is reflected to the level of 10th rib in the mid-axillary line and variable along the 12th rib posteriorly. Usually it crosses the 12th rib obliquely at its midpoint, such that the lateral half of this rib is inferolateral to the pleural limit. The visceral pleura never descends to the level of the midpoint the 12th rib except with forced ventilation. Therefore, all punctures that pass above the 12th rib pierce the diaphragm. Injury to the parietal pleura can be avoided by staying above the lateral half of the 12th. However, the possibility of injury to the lower lobe of lung or the parietal pleura needs to be considered. **Entry through the pleural space may lead to an accumulation of fluid, causing hydrothorax,** which occurred in 5% of the present patients and required insertion of a chest tube (Fig. 3). Others have reported the incidence of hydrothorax to be 0–12%. The cause of hydrothorax has been attributed to a failure to seal the tract with the working sheath during the procedure, or inadequate drainage of the kidney afterward. The second cause is the more probable explanation, as an Amplatz sheath was routinely used during PCNL. Thus a well-draining nephrostomy will minimize the leakage of urine into the pleural space.

Figure 3. Hydrothorax before ( a ) and after ( b ) inserting a chest tube.



216. According to Bismuth-Strasberg classification, cystic blow out comes under which type?
- Type A
  - Type B
  - Type C
  - Type D

### Staging

Several injury classification systems have been described for biliary tract trauma. Most of them are in the context of iatrogenic injuries during cholecystectomy and provide a recommended surgical approach for repair.

None of the classification systems is universally accepted, but the classification systems of Bismuth and

Strasberg are presently the most widely used.

Table 1. Bismuth's Classification (1982)

Type	Criteria
1	Low common hepatic duct stricture, with a length of the common hepatic duct stump of >2 cm
2	Proximal common hepatic duct stricture, with a hepatic stump length of <2 cm
3	Hilar stricture, no residual common hepatic duct, but the hepatic ductal confluence is preserved
4	Hilar stricture, with involvement of confluence and loss of communication between right and left hepatic duct
5	Involvement of aberrant right sectorial hepatic duct alone or with concomitant stricture of the common hepatic duct

Table 2. Strasberg's Classification (1995)

Type	Criteria
A	Cystic duct leaks or leaks from small ducts in the liver bed
B	Occlusion of a part of the biliary tree, almost invariably the aberrant right hepatic ducts
C	Transection without ligation of the aberrant right hepatic duct
D	Lateral injuries to major bile ducts
E	Subdivided as per Bismuth's classification into E1 to E5

Table 1. Bismuth's classification (1982)<sup>141</sup>

Type	Criteria
1	Low CHD stricture, with a length of the common hepatic duct stump of >2 cm
2	Proximal CHD stricture-hepatic duct stump <2 cm
3	Hilar stricture, no residual CHD, but the hepatic ductal confluence is preserved
4	Hilar stricture, with involvement of confluence and loss of communication between right and left hepatic duct
5	Involvement of aberrant right sectorial hepatic duct alone or with concomitant stricture of the CHD

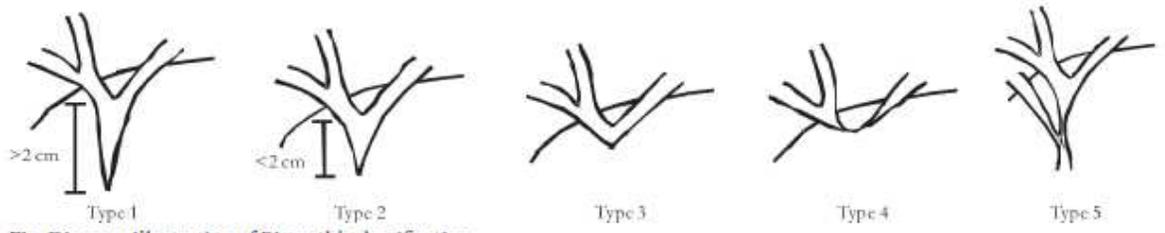
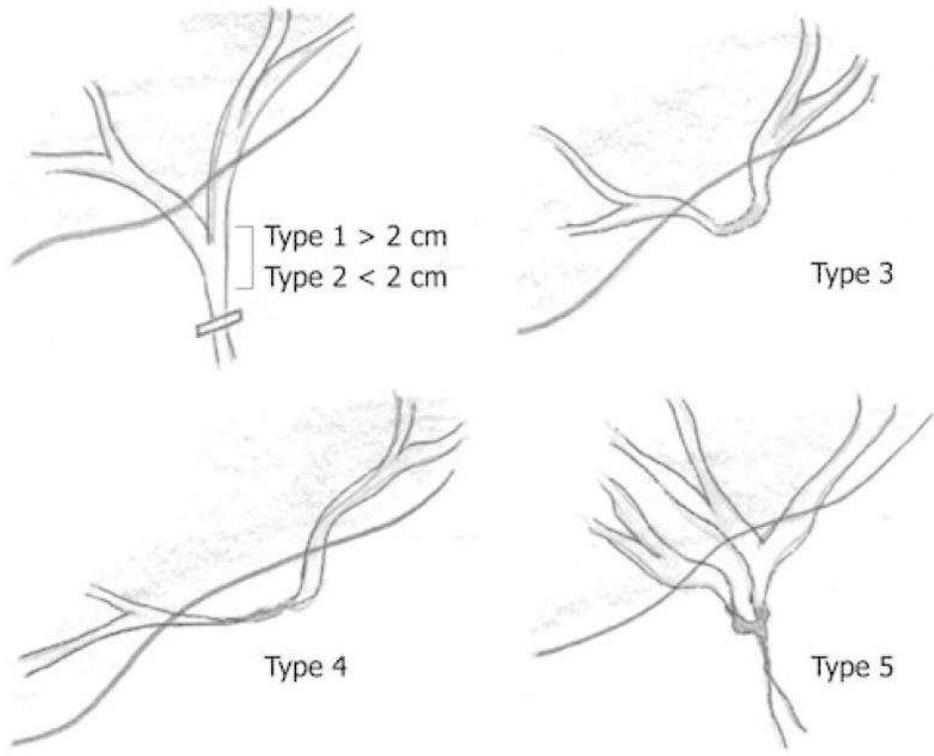


Fig. Diagram illustrating of Bismuth's classification.

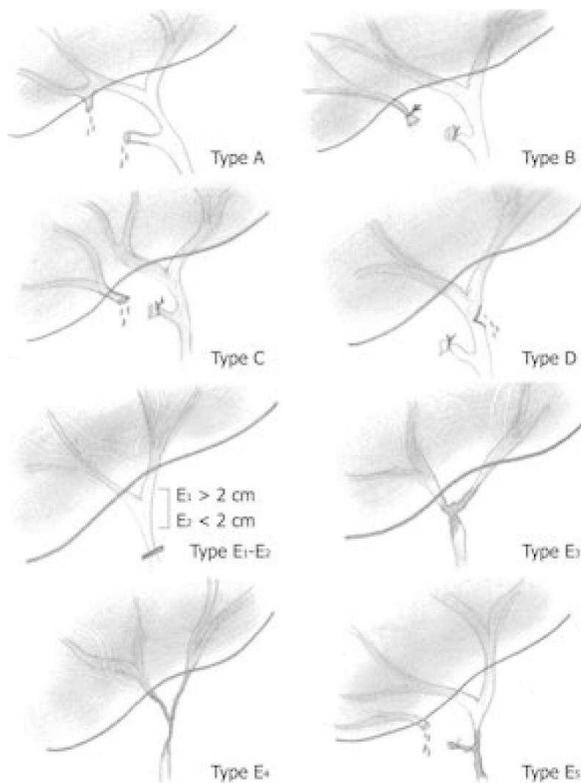


**Table 3.** Strasberg's classification (1995)<sup>[9]</sup>

Type	Criteria
A	Cystic duct leaks or leaks from small ducts in the liver bed
B	Occlusion of a part of the biliary tree, almost invariably the aberrant right hepatic ducts
C	Transection without ligation of the aberrant right hepatic ducts
D	Lateral injuries to major bile ducts
E	Subdivided as per Bismuth's classification into E1 to E5

**Table 4.** Amsterdam Academic Medical Center's classification (1996)<sup>[10]</sup>

Type	Criteria
A	Cystic duct leaks or leakage from aberrant or peripheral hepatic radicles
B	Major bile duct leaks with or without concomitant biliary strictures
C	Bile duct strictures without bile leakage
D	Complete transection of the duct with or without excision of some portion of the biliary tree



217. Best method to evaluate the prognosis of outcome of coma for the patient of subarachnoid hemorrhage is

- Glasgow coma scale
- Hess & Hunt scale

- c) Glasgow-Blatchford bleeding score
- d) Intracerebral hemorrhage score

## Hunt and Hess Stroke Scale

Hunt and Hess scale is used to classify the severity of **non-traumatic subarachnoid hemorrhage**.

1	Asymptomatic, mild headache, slight nuchal rigidity.
2	Moderate to severe headache, nuchal rigidity, no neurologic deficit other than cranial nerve palsy.
3	Drowsiness / confusion, mild focal neurologic deficit.
4	Stupor, moderate-severe hemiparesis.
5	Coma, decerebrate posturing.

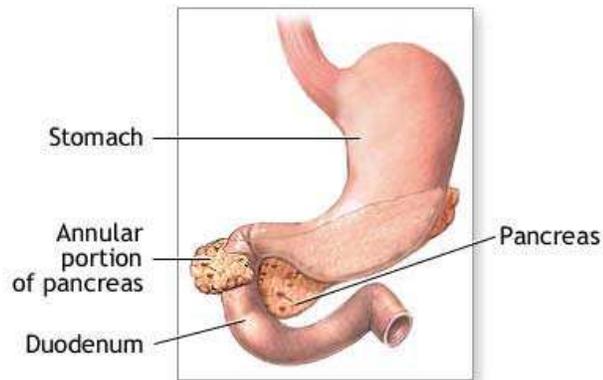
218. Treatment of annular pancreas is
- a) Division of pancreas
  - b) Duodenoduodenostomy
  - c) **Duodenojejunostomy**
  - d) Roux en Y loop

Annular pancreas refers to the presence of a **band of normal pancreatic tissue** that **partially or completely encircles the 2<sup>nd</sup> portion of the duodenum** & extends into the head of the pancreas. Treatment usually involves bypass, via **duodenojejunostomy**, rather than resection. – *Sabiston 17<sup>th</sup>/1646*

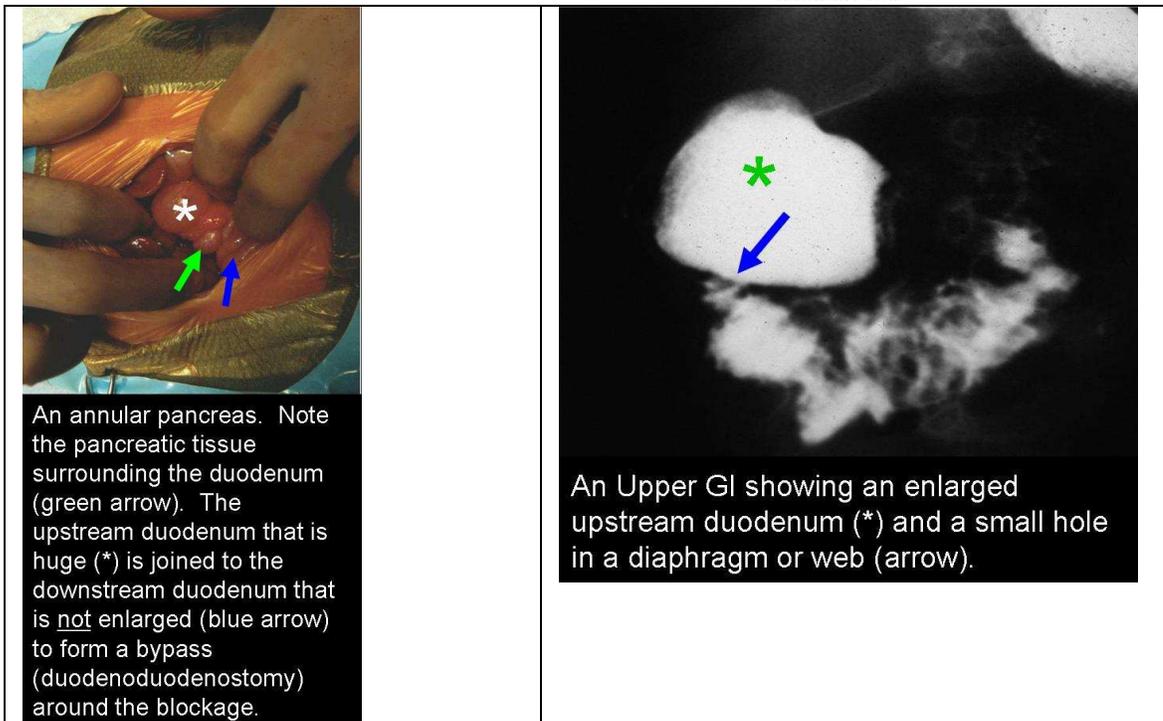
### Annular Pancreas

When the **ventral pancreatic anlage fails to migrate correctly** to make contact with the dorsal anlage, the result may be a ring of pancreatic tissue encircling the duodenum. Such an annular pancreas may cause **intestinal obstruction** in the neonate or the adult. Symptoms of postprandial fullness, epigastric

pain, nausea, and vomiting may be present for years before the diagnosis is entertained. The radiographic findings are **symmetric dilation of the proximal duodenum** with bulging of the recesses on either side of the annular band, effacement but not destruction of the duodenal mucosa, accentuation of the findings in the right anterior oblique position, and lack of change on repeated examinations. The differential diagnosis should include duodenal webs, tumors of the pancreas or duodenum, postbulbar peptic ulcer, regional enteritis, and adhesions. Patients with annular pancreas have an **increased incidence of pancreatitis and peptic ulcer**. Because of these and other potential complications, the treatment is surgical even if the condition has been present for years. **Retrocolic duodenojejunostomy is the procedure of choice**, although some surgeons advocate Billroth II gastrectomy, gastroenterostomy, and vagotomy. – Harrison 17<sup>th</sup>/2016



ADAM.



219. An adult presented with hematemesis and upper abdominal pain. Endoscopy revealed growth at the pyloric ntrum of the stomach. CT scan abdomen showed growth involving the pyloric antrum without infiltration or invasion into the surrounding structures. Laparotomy was planned which revealed neoplastic growth involving the posterior wall of the stomach and extending 5 cm up to tail of pancreas. What will be the most appropriate next step of management?
- Closure of abdomen
  - Antrectomy + partial gastrectomy + removal of tail of pancreas
  - Antrectomy & vagotomy
  - Antrectomy + distal pancreatectomy + splenectomy
220. A lady presented with recurrent attacks of giddiness and adnominal pain since 3 months. Endoscopy was normal. Her glucose level was found to be 40mg%. Ct abdomen shows a small well defined 8 mm enhancing lesion in the head of pancreas with no other abnormal finding. Diagnosis was clinched by her fasting blood glucose which was low and insulin level which was increased. What should be the treatment plan for this patient?
- Whipple's operation is the TOC
  - Enucleation is the curative Tt
  - Enucleation with RT will be more effective
  - A trial of Streptozocin is recommended before surgery

The majority (90%) of insulinomas are **benign & solitary**, & only 10% are malignant. They are typically cured by **simple enucleation**. – *Schwartz 8<sup>th</sup>/1275*

221. An adult male patient presented with recurrent attacks of hypoglycemia and seizures. During the attack of hypoglycemia episodes, his blood glucose level was 20-30% and insulin level was 71U/dl. The C-peptide level was 6 mmol/L. What is the most probable diagnosis ?
- Excess admn of insulin injection
  - Pancreatic ca
  - Insulinoma**
  - Sulphonylurea excess

Insulinomas are characterized clinically by the **Whipple triad**:

- Presence of symptoms of hypoglycemia
- Documented low blood sugar at the time symptoms are present
- Reversal of symptoms by glucose administration.

Most patients with insulinoma have normal physical examination findings.

A diagnostic **ratio of blood insulin : glucose of >0.4** or **C-peptide levels >2 nmol/L** has proved valuable.

222. A neonate presented with fever, lethargy, abdominal distention, vomiting and constipation. Clinically he was diagnosed as volvulus neonatorum with suspected perforation. Best investigation of choice is
- X-ray
  - Barium enema
  - Upper GI endoscopy
  - Ba meal follow through**

A plain X-ray of the abdomen is not helpful in the early stage of volvulus. **The most reliable radiological confirmation of malrotation is a barium meal with fluoroscopy**, which will show that the duodenojejunal flexure is located at a lower level than the pylorus. The contrast may show the spiral twist of the volvulus as well – *Jones' Clinical Pediatric Surgery: Diagnosis & Management by Peter G. Jones, John M. Hutson, Alan A Woodward 5<sup>th</sup>/40*

### Preferred examination

An upper GI series is the preferred diagnostic test for malrotation with midgut volvulus and must be performed, unless a delay in surgical treatment will compromise outcome (as in the case of a moribund child). **Upper GI series sensitivity is 85-95%**, with a higher specificity (false positives are rare).

In midgut volvulus, the **classic radiographic finding is a partial duodenal obstruction** (dilation of both the stomach and proximal duodenum, with a small amount of distal bowel gas). Complete obstruction of the duodenum may also be found. Less frequent, but more ominous, signs are a **gasless abdomen, ileus, or a distal small bowel obstruction with multiple dilated loops and air-fluid levels**. **A normal abdominal film does not exclude malrotation.**

The upper GI series is performed with barium administered either orally or through a nasogastric tube. The normal duodenojejunal junction (DJJ) lies to the left of the left-sided spinal pedicle at the level of the duodenal bulb on a true frontal view. The duodenal C-sweep courses posteriorly, inferiorly, anteriorly, and then superiorly.

The findings of a malrotation on upper GI series include the following:

- The DJJ is displaced downward and to the right on frontal view.
- The duodenum has an abnormal course on lateral view.
- Abnormal positioning of the jejunum (lying on right side of abdomen) should alert the physician to the possibility of a malrotation, but this finding should not be relied upon to either make or exclude the diagnosis.

In **malrotation with midgut volvulus**, the findings also include the following:

- A **dilated, fluid-filled duodenum**
- A proximal small bowel obstruction

- A "**corkscrew**" pattern (proximal jejunum spiraling downward in the right- or mid-upper abdomen in midgut volvulus, which is rare), as seen in the images below.
- Mural edema and thick folds



Upper GI series shows malrotation with midgut volvulus. An incomplete duodenal obstruction and dilation of the first and second portions are seen, as is the "**corkscrew sign**."



Upper GI series shows the "**corkscrew sign**" in a frontal view.

#### Extra Edge

Fetal midgut volvulus can be identified on prenatal sonography by – **Coffee bean sign**

223. True about carcinoid syndrome is all except
- 60% occur in ileum & appendix
  - Rectum spared**
  - 5 yr survival > 60%
  - Females more affected

In the GIT, more than 90% carcinoids are found in 3 sites:

- Appendix – 45%
- Ileum – 28%
- **Rectum – 16%**

224. Most common cyst of the spleen are
- Hydatid cyst
  - Dermoid cyst
  - Pseudocyst
  - Lymphangioma

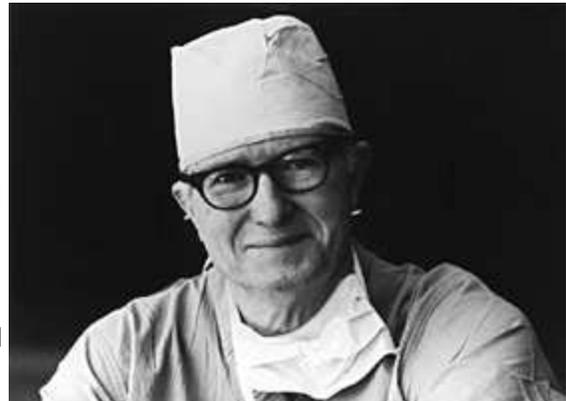
Parasitic infection is the MC cause of splenic cysts worldwide, & the majority are due to *Echinococcus* species. – Schwartz

225. First Autologous ureter transplant was done by
- Hardy
  - Kavosis
  - Higgins
  - Studor

Headlines around the world told the astounding news of a surgical team in Mississippi which had transplanted a human lung into another human.

The year was 1963, and the team was led by **Dr. James D. Hardy**, professor of surgery and chairman of the department at the University of Mississippi Medical Center.

But it was the following year when an event at the young Medical Center really had the world's press in a frenzy. Dr. Hardy and his team transplanted the heart of a chimpanzee--man's closest genetic relation--into the chest of a dying man. The world's first heart transplanted into man beat 90 minutes before it stopped.



In addition to the first lung and heart transplantations, Dr. Hardy's team also performed the first successful human kidney autotransplant and the first human adrenal autotransplant in the United States.

## Original Scientific Reports

### Renal Autotransplantation

Audun Flatmark, M.D., Dagfinn Albrechtsen, M.D., Gunnar Sjødal, M.D., Helge Bondevik, M.D., Arnt Jakobsen, Jr., M.D., and Inge B. Brekke, M.D.

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In the period, 1973–1985, a total of 305 renal autotransplantations (often with extracorporeal repair) were performed in 274 patients for fibromuscular (56) or atherosclerotic (98) renovascular disease, nephrolithiasis (97), renal tumor (15), or ureteral injury (8). One-third of the patients had reduced renal function, and approximately half of the patients with renovascular lesions presented with severe or malignant hypertension. The postoperative mortality rate of 4% was related to age over 60 years and reduced renal function. Postoperatively, 3.6% of the autografted kidneys were lost due to vascular thrombosis (7) or other causes (4). Twenty-five percent (69) of the patients had a solitary kidney. Two patients died postoperatively with functioning autografts. Long-term renal function remained stable in 60 patients, and deteriorated in 7.

We conclude that autotransplantation and extracorporeal repair provide safe and efficient treatment for selected difficult renal lesions.

Renal autotransplantation was first described by Hardy in 1963 [1]. A patient with high ureteral injury was treated with removal of the kidney, subsequent perfusion with cold glucose, and reimplantation in the iliac fossa.

The technique has later been adopted by many as an alternative to in situ operations for renovascular problems, stone disease, ureteral tumors, and renal carcinoma in a solitary kidney. The operation has also been named bench surgery, work-bench surgery, ex situ repair, ex vivo reconstruction, and extracorporeal renal surgery. The use of the technique has often been the only alternative to nephrectomy. Reconstructive surgery may be impossible in situ, but can safely be performed under direct vision after kidney removal and cold perfusion preservation.

Most reports on renal autotransplantation only deal with a single or few cases, nevertheless, 4 centers have reported more than 30 patients [2–6], the largest series being that of Gil-Vernet [4] comprising 92 patients with all types of renal problems.

In 1977, we reported on our preliminary experience with this technique [7]. The initial results were promising. The purpose of this article is to present our strategy and review our experience with special emphasis on patient mortality, kidney salvage, surgical complications, and long-term function of solitary kidneys. The long-term results of the different patient categories will be published separately.

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#### Material

A total of 305 renal autotransplantations were performed in 274 patients between April, 1973 and December, 1985 (Table 1). The largest group of patients (154) had hypertension and vascular lesions due to atherosclerosis or fibromuscular dysplasia. Patients with stone disease were either referred because of previous unsuccessful surgery, and/or judged to be inoperable with in situ techniques. Twenty-five percent (69) of all patients had a solitary functioning kidney.

There were 143 females and 131 males aged 2–76 years. Seventy-eight were less than 40 years old, 120 were between 40 and 59 years, and 76 were 60 years-old or older. Severe or malignant hypertension was present in 47% of patients with renovascular lesions. One-third of the patients (94/274) had reduced kidney function, i.e., serum creatinine exceeding 130  $\mu\text{mol/l}$ , and 44 of these had a serum creatinine of  $>200 \mu\text{mol/l}$ . In the group of patients with solitary kidney, 60% (42/69) had reduced kidney function.

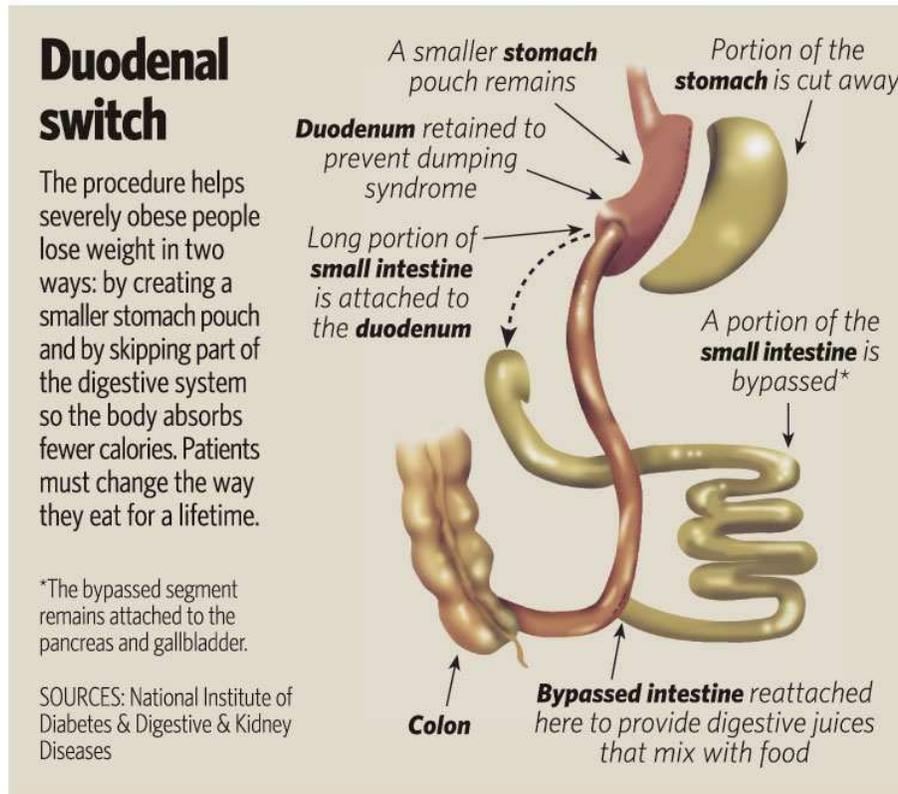
#### Methods

The standard procedure was to remove the kidney through a flank incision with reinsertion in the iliac fossa. In some cases, a long midline incision was used, permitting both removal and subsequent reimplantation. The ureter was always transected.

Most kidneys (292) were preserved with cold ( $4^{\circ}\text{C}$ ) perfusion and storage, originally using a low molecular weight dextran solution, but since 1978, Euro-Collins solution has been used [8]. Some kidneys with an occluded renal artery were machine-perfused in order to test their viability after ex vivo vascular repair. Thirteen kidneys had flow values of 100–270 ml/min on the machine, and were subsequently transplanted.

Additional surgery during hypothermia consisted of stone removal via nephrolithotomies with fluoroscopic confirmation of the removal of all stone material. Tumors were removed with histological examination of frozen sections to secure free margins. Renal vascular repair, including anastomosis of 2 or more renal arteries, and branched grafts as previously described [7], was necessary in 107 (35%) autografts. Aortoiliac thrombendarterectomy was required in 60 (22%) patients (Table 2).

226. All of the following are restrictive operation for morbid obesity except
- Vertical band gastroplasty
  - Switch duodenal operation
  - Roux en Y operation
  - Laparoscopic adjustable gastric banding



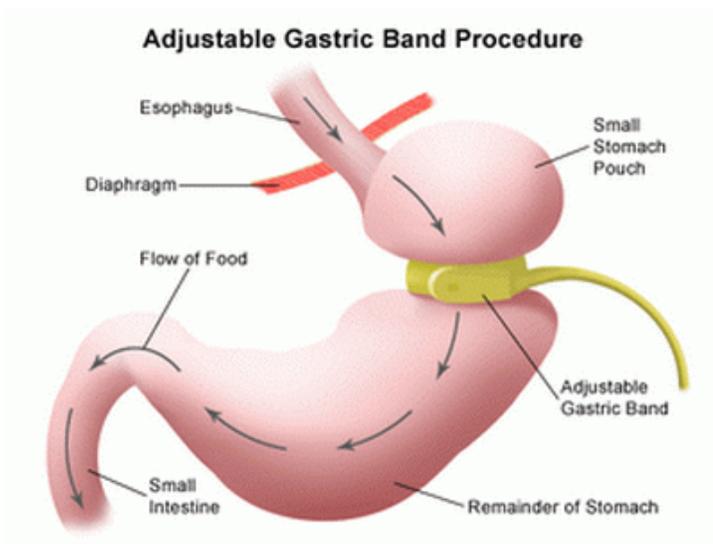
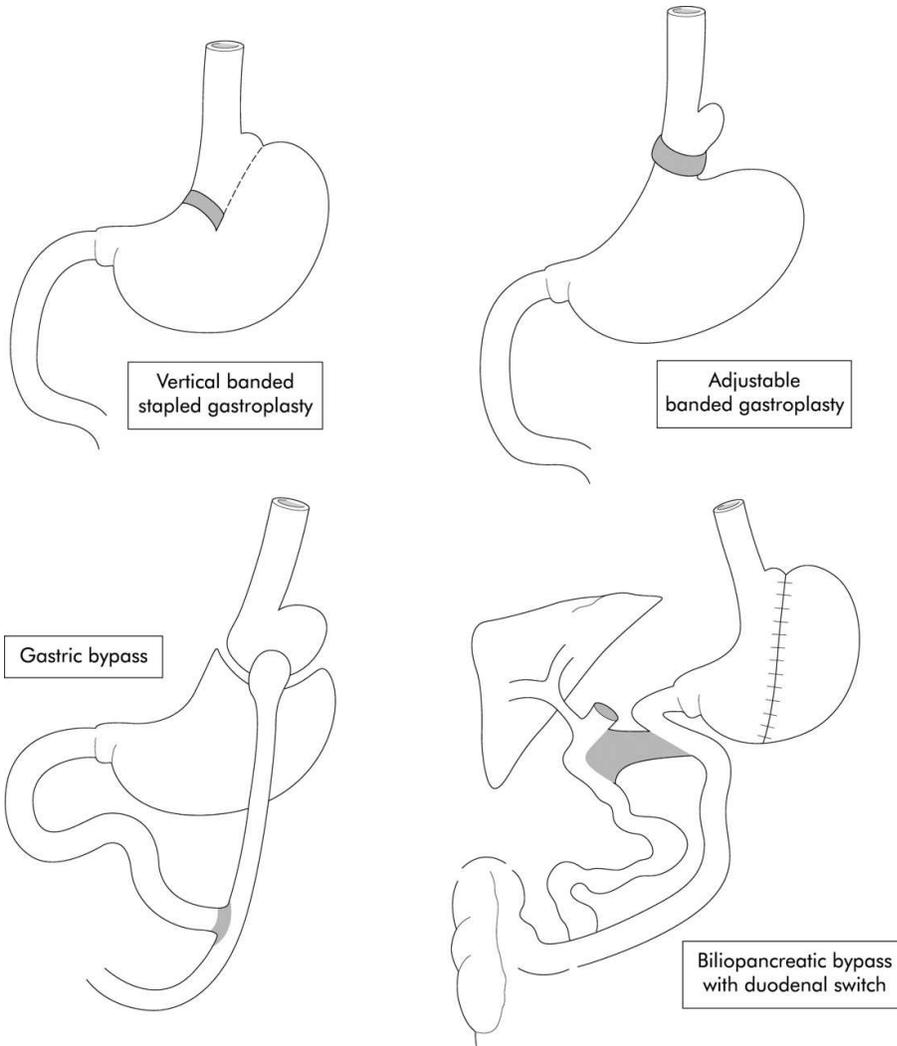
Staff graphic: Molly Quinn

## Surgical Therapy

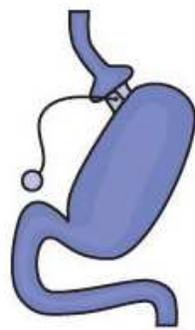
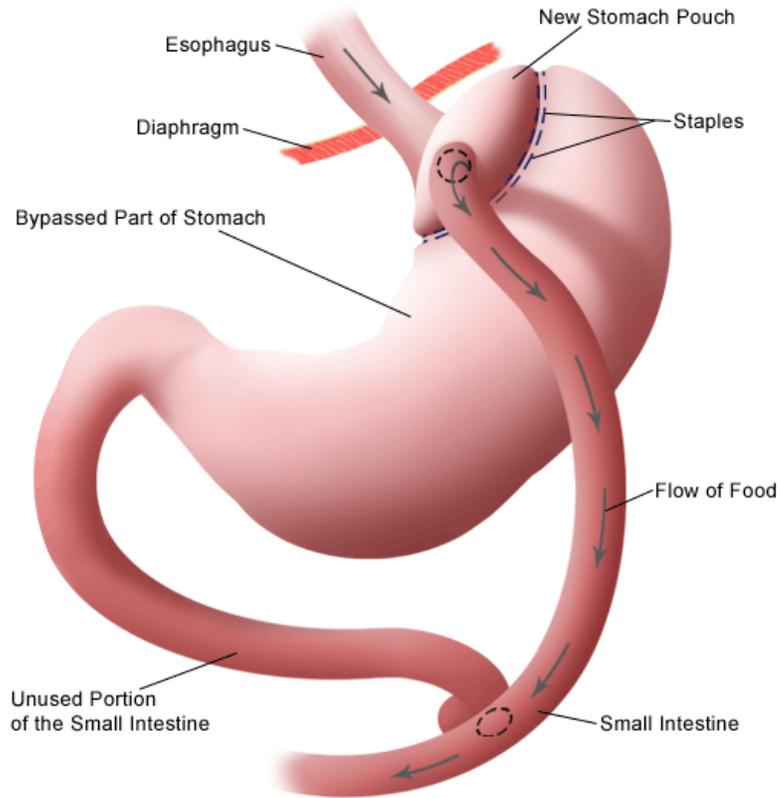
Types of bariatric surgery include the following:

- Restrictive procedures (eg, adjustable gastric banding, sleeve gastrectomy)
- Restrictive procedures with some malabsorption (eg, Roux-en-Y gastric bypass)
- Malabsorptive procedures with some restriction (eg, biliopancreatic diversion with duodenal switch)

Bariatric surgery can be performed by an open technique and by a laparoscopic technique. The laparoscopic approach has currently become the more popular approach.



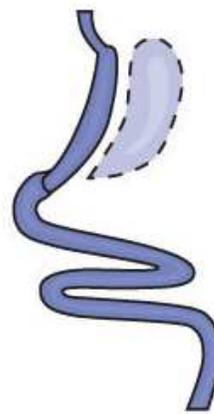
### Roux-en-Y Type of Gastric Bypass Procedure



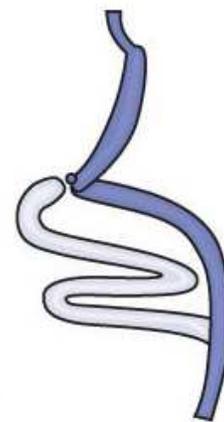
**Adjustable Gastric Band (AGB)**



**Roux-en-Y Gastric Bypass (RYGB)**



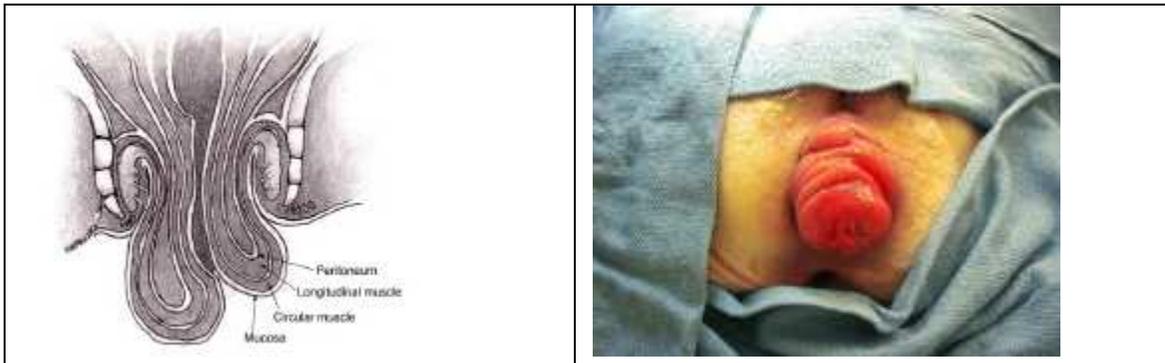
**Vertical Sleeve Gastrectomy (VSG)**



**Biliopancreatic Diversion With a Duodenal Switch (BPD-DS)**

227. A young male patient presented with rectal prolapse which was reducible. Surgery of choice will be
- Rectopexy
  - Delorme operation
  - Goodsall operation
  - Anterior resection

Rectal prolapse occurs when a mucosal or full-thickness layer of rectal tissue slides through the anal orifice. Problems with fecal incontinence, constipation, and rectal ulceration are common.



Whenever possible, an abdominal rectopexy is recommended. – Schwartz

Surgical treatment can be divided into 2 categories according to the approach used to repair the rectal prolapse: abdominal procedures and perineal procedures. The choice of abdominal versus perineal procedure is mainly dictated by the patient's age and comorbidities.

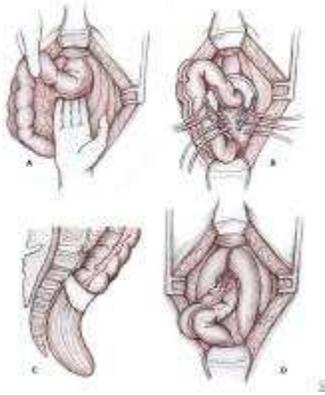
In general, the abdominal procedures have a lower recurrence rate but a higher morbidity rate. The converse is true of perineal procedures. In general, treat older, debilitated patients (whose life expectancy is shorter) with a perineal procedure, and treat younger, healthier patients with abdominal procedures, although surgeons with large experience and low recurrence rates also advocate perineal procedures for their younger, healthier patients.

### Abdominal procedures

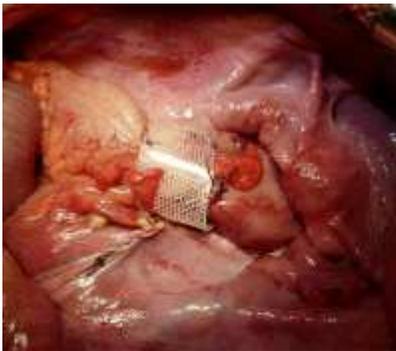
These procedures are typically performed in younger, healthier patients whose life expectancy is longer. For these patients, procedures with lower recurrence rates but higher morbidity rates are most appropriate. The choice of abdominal procedure is often dictated by the extent of the associated constipation and by the surgeon's preference.

- Abdominal rectopexy:** The rectum is mobilized and attached to the presacral fascia, using direct sutures or using a prosthetic material, a polypropylene mesh (Ripstein procedure; see the first 2 images below, or an Ivalon sponge (see the third image below). It has a high success rate for

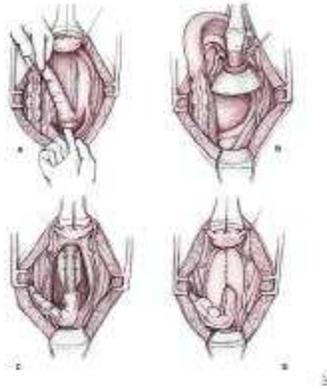
prolapse control, and incontinence is improved in 60% of patients, although as many as 60% of patients have constipation after this procedure.



The Ripstein procedure is designed to maintain the normal posterior rectal curvature by attaching it to the presacral fascia, thus avoiding a straight tube that intussuscepts during straining. (A) The rectum is mobilized down to the coccyx. (B) A Marlex mesh placed around the rectum while this is tensed upward and sutured with nonabsorbable material to the presacral fascia. The loop of mesh needs to be loose enough to prevent postoperative constipation. (C) Sagittal view shows the suspended rectum. (D) The peritoneum is closed with a running absorbable suture.



Intraoperative photograph of a 12-year-old girl with recurrent rectal prolapse and mucosal ulceration with profuse bleeding. She had a long sigmoid colon, which was resected; an end-to-end anastomosis was performed. This photograph depicts the anastomosed rectum fixed to the presacral fascia with mesh. Nonabsorbable sutures retain the mesh to the serosa. Note that the mesh is slightly loose to account for the child's growth. The uterus and its ligaments can be seen in front of the rectum. Iliac vessels remain intact on each side.



**Ivalon sponge procedure.** (A) The rectum is mobilized. Meticulous hemostasis is mandatory to prevent a hematoma that predisposes the patient to prosthetic material infection. (B) An Ivalon rectangular sponge made of polyvinyl alcohol is sutured to the sacral periosteum. (C) The rectum is retracted upward, and the sponge is wrapped around it and tied to the anterior surface. A portion of the anterior rectal wall is left free to prevent luminal obliteration. (D) The peritoneum is closed with a running absorbable suture.

#### Perineal procedures

- **Perineal resection:** Mikulicz first described this procedure in 1889.<sup>3</sup> Perineal rectosigmoidectomy with rectopexy, correction of the pelvic floor (plication of the puborectalis muscles), and coloanal anastomosis is promising and could be a good approach for pediatric patients with intractable prolapse and redundant sigmoid. It has been successfully performed using stapling devices for the resection and reconstruction of colonic continuity. This technique avoids the abdominal approach, with its obvious complications.
- **Delorme procedure:** The mucosa and part of the underlying rectal muscle are excised, and the rectum is then plicated with polydioxanone sutures towards the anal canal. Long-term results are not satisfactory, with a recurrence rate of 17%. It has been used in children with recurrent prolapse and has the advantage of not entering the abdomen. The observation that recurrence and complication rates may be lower in younger and medically fit patients suggests that the Delorme repair does not necessarily have to be restricted specifically to older, medically unfit patients.

228. Regarding CSF leak all are true except
- a) MC site is fovea ethmoidalis
  - b) B2 transferrin is specific
  - c) MRI Gd T1 is best for diagnosis
  - d) Fluorescein dye used intrathecally for diagnosis

Option C - A new test, injection of a small amount of gadolinium contrast into the lumbar spinal fluid followed by MRI is very promising (Jinkins et al, 2002). Gadolinium is non-toxic as is CT contrast dye, and

also while CT contrast could be confused with bony structures, gadolinium is not easily confused with other body parts. Of course, MRI can scan the entire neuroaxis if the site of leak is unclear.

*It is used for diagnosis but it is not the best test.*

Option D - A surer method is to inject radioactive label or a **fluorescent dye into the spinal fluid** and test for the label or dye in the fluid. Nasal pledgets can be left in the nose for extended periods, enabling detection of intermittent rhinorrhea. 0.5 ml of 5% fluorescein diluted in 9.5 cc of CSF is used (according to Hilinski et al, 2001). Smaller amounts are used in children. (Lue and Manolidis 2004). It is useful to use control pledgets under the upper lip for comparison. **The test is analyzed under ultraviolet light.** No complications were reported by Montgomery in more than 200 injections of fluorescein. However some authors have reported seizures from fluorescein can occur.

### Preferred examination

**A suggested algorithm for the diagnosis of a cerebrospinal fluid (CSF) fistula follows.**

**Confirm or exclude the presence of CSF in leaking fluid by means of an immunoelectrophoretic study of the fluid for beta-2 transferrin (B2Tr) or, where available, beta-trace protein.** For this specialized laboratory study, 0.5-1.0 mL of the fluid may be required. An absorptive sponge pad placed at or near the presumed site of fluid leak can facilitate the collection of the fluid.

Perform **high-resolution, thin-section axial and coronal cranial and facial computed tomography (CT)** scanning. Include all of the paranasal sinuses and petrous temporal bones in the scans.

Perform magnetic resonance **(MR) cisternography.** This study may also be useful for detecting inactive fistulas.

**CT cisternography** or **radionuclide cisternography** may be useful if CT and MR cisternography do not show the CSF fistula. **Radionuclide cisternography may be useful to detect an intermittently active CSF fistula.** Cisternography with an intrathecal injection of radioisotope or nonionic iodinated myelographic contrast medium or noninvasive magnetic resonance imaging (MRI) cisternography usually localizes the CSF leak.

**Brain and spinal MRI** is useful in demonstrating **meningocele and meningoencephalocele** when associated with CSF leak, as well as for examining patients with spontaneous intracranial hypotension syndrome.

On occasion, the methods listed above do not help in localizing the CSF fistula, and surgical exploration is necessary.

Fluid leaking from the nose or external auditory canal must first be positively identified as CSF. Drops of fluid from a CSF leak placed on absorbent filter paper may result in the **double-ring sign**, which is a **central circle of blood and an outer clear ring of CSF.** Results of glucose, chloride, and total protein tests of the fluid are not specific or conclusive for CSF.

All methods of **cisternography—radionuclide, CT, and MR**—provide improved or optimal **CSF fistula detection** when the fistula is active and when a Valsalva maneuver or jugular venous compression is added to the imaging protocol. CSF fistula can usually be demonstrated by using some method of

cisternography, but localization of the leak to the right or left nasal cavity may be difficult because of the tendency of the fluid to cross sides and flow from both nostrils. In a study of 4 patients who underwent radionuclide cisternography (RNC), as well as MRI and/or CT, for suspected CSF leaks, Thomas et al found that RNC accurately detected and localized the leaks in all patients. Each patient subsequently underwent a procedure for an epidural blood patch, and all patients experienced symptomatic relief.

Methods for detecting CSF fistulas with intrathecal injections of dye pose a risk of chemical meningitis. Methylene blue, indigo carmine, and phenolsulfonphthalein (PSP) dyes are no longer in use. Some otolaryngologists use a dilute solution of fluorescein to localize CSF fistulas both preoperatively and during surgery. Typically, 0.5 mL of a 10% fluorescein solution is injected into the lumbar subarachnoid space over more than 1 minute. Cotton pledgets are placed in the nose, as for radionuclide cisternography. The dye reaches the skull base in 6 hours and is present over the cerebral convexities in 24 hours. The pledgets are examined for green fluorescence in a dark room with ultraviolet light 6 hours after the intrathecal PSP injection.

## OBSTETRICS & GYNAECOLOGY

229. Treatment for a women suffering from Antiphospholipid antibody syndrome (APL) with past history of stillbirths and abortion is
- Aspirin only
  - Low dose Aspirin + LMW heparin
  - Start Low dose Aspirin + LMW heparin + Prednisolone
  - No treatment

Option B - Combined unfractionated heparin and low-dose aspirin regimens are thought to reduce the risk of spontaneous pregnancy loss by 54%, resulting in a live-birth rate of 70-80%. Evidence from small, controlled trials (25-50 patients) has suggested that patients with recurrent pregnancy loss associated with antiphospholipid antibodies and without prior thromboembolism benefit from treatment with low-dose unfractionated heparin at prophylactic doses of 5,000-10,000 IU twice daily, in addition to low-dose aspirin at 70-81 mg daily.

Option C - Prednisone and other immunomodulating therapies are seldom prescribed for pregnant women with APS, but prednisone is appropriate for clinically active SLE, if present. A small study of APS patients with and without SLE who were treated with 40 mg prednisone daily or heparin (10,000 IU twice daily at 6-8 weeks, reduced to 2000 IU twice daily to attain normal activated partial thromboplastin time at midtrimester), both with concomitant low-dose aspirin (81 mg), demonstrated equally high rates of live births in both treatment groups. Yet, maternal complications were greater in the prednisone-treated group.

230. Nuchal translucency at 14 weeks of gestation is increased in
- Turner syndrome
  - Down syndrome
  - Hydrocephalus
  - Skeletal dysplasia
231. Non-immune hydrops fetalis is caused by
- CMV
  - Parvovirus
  - HPV
  - HBV
232. Isolated marker of Neural Tube defects is
- Acetylcholinesterase
  - Pseudocholinesterase
  - AFP

- d) ↓hCG
233. Methyldopa is used in
- Parkinsonism
  - PIH
  - Hirsutism
  - Refractory HTN
234. Best method for female fertility assay during menstrual cycle is
- Fern test
  - Hormonal study
  - Vaginal cytology
  - Spinnbarkeit
235. What is not done in 3<sup>rd</sup> stage of labour ?
- Oxytocin injection
  - Gentle massage of uterus
  - Controlled cord traction
  - Ergometrine injection
236. In human which of the following is not associated with menstrual cycle
- Change in steroid level
  - Endometrial gland changes
  - Estrus cycle
  - Vaginal cytology
237. A 40 year old woman presented with CIN III on Pap smear. Treatment of choice is
- Hysterectomy
  - Colposcopy with LEEP
  - Trachelectomy
  - Conization
238. Which of the following is an absolute contraindication of pregnancy except
- Eisenmenger's syndrome
  - WPW syndrome
  - Coarctation of aorta
  - Primary pulmonary HTN
239. In a pregnant female with HIV all of the following measures reduces the risk of vertical transmission except
- Avoidance of ergometrine in 3<sup>rd</sup> stage of labour
  - Nevirapine intrapartum admn

- c) Elective Caesarian sction
- d) AZT 72 hrs before delivery

240. Primary amenorrhoea with normal ovaries normal external genitalia and normal breast is seen in

- a) Mayer Rokitansky Kuster Hauser syndrome
- b) Turner syndrome
- c) Noonan syndrome
- d) Androgen insensitivity syndrome

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome consists of vaginal aplasia with other müllerian (ie, paramesonephric) duct abnormalities. Its penetrance varies, as does the involvement of other organ systems. Type I Mayer-Rokitansky-Kuster-Hauser syndrome is characterized by an isolated absence of the proximal two thirds of the vagina, whereas type II is marked by other malformations; these include vertebral, cardiac, urologic (upper tract), and otologic anomalies.

The following may be observed in patients with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome:

- **Primary amenorrhea** and possible cyclic abdominal pain
  - These symptoms are common in individuals with Mayer-Rokitansky-Kuster-Hauser syndrome.
  - The patient undergoes puberty with normal thelarche and adrenarche; however, menses do not begin.
  - Patients may report cyclic abdominal pain due to cyclic endometrial shedding without a patent drainage pathway.
  - Because **ovarian function is normal**, patients experience all bodily changes associated with menstruation and puberty.
- **Infertility**
  - Patients who do not undergo evaluation for primary amenorrhea often seek clinical attention for infertility. However, patients rarely proceed to infertility evaluation without ever having had a menses due to Mayer-Rokitansky-Kuster-Hauser syndrome.
  - Although the ovaries function normally, the fallopian tubes may be closed, and the uterus is often anomalous.
- **Inability to have intercourse**
  - The degree of vaginal aplasia can vary from complete absence to a blind pouch.
  - The more shallow the canal, the greater the likelihood of the patient having dyspareunia.
- **Renal malformations**
  - Absence or ectopia of the kidneys is common. Diagnosis can lead to discovery of renal anomalies.
  - Some patients present with a history of voiding difficulties, urinary incontinence, or recurrent urinary tract infections (UTIs).

- **Vertebral anomalies**: Skeletal findings range in severity and clinical importance. Scoliosis is the most common of the skeletal anomalies.

241. A lady presented amenorrhoea and having a spontaneous abortion 6 months back. Her FSH level were 6 miu/ml. What is the most probable diagnosis ?
- Pituitary failure
  - Ovarian failure
  - Fresh pregnancy
  - Uterine synechiae**

Normal FSH level – **1-8 miU**

So here FSH is normal.

**Amenorrhoea with normal FSH** → **Uterine pathology**

242. Polyhydramnios is seen in
- Posterior urethral valve
  - Cleft palate**
  - Congenital diaphragmatic hernia
  - Exstrophy epispadias complex

Cleft palate → **Interferes with normal swallowing** → **Polyhydramnios**

243. Cause of leak per vaginum and meconium stained liquor in a 34 weeks gravid is
- Listeria**
  - CMV
  - Toxoplasma
  - Herpes

244. Most commonly associated with carcinoma cervix is
- HPV 16**
  - HPV 18
  - HPV 31
  - HPV 33

245. Sentinel biopsy most useful in which gynecological malignancy
- Ca endometrium
  - Ca cervix
  - Ca vulva**
  - Ca vagina

246. Most useful investigation in a case of VVF
- a) Triple swab test
  - b) Urine culture
  - c) Cystoscopy
  - d) IVP
247. Treatment of choice of intrahepatic cholestasis in pregnancy is
- a) Cholestyramine
  - b) Ursodiol
  - c) Steroid
  - d) Antihistamines

Ursodiol quickly relieves pruritus & lowers serum enzyme levels. - Williams

248. False about partial mole is
- a) Usually associated with triploidy
  - b) Rarely cause persistent GTD
  - c) Early diagnosis can be made by USG
  - d) May present as missed abortion

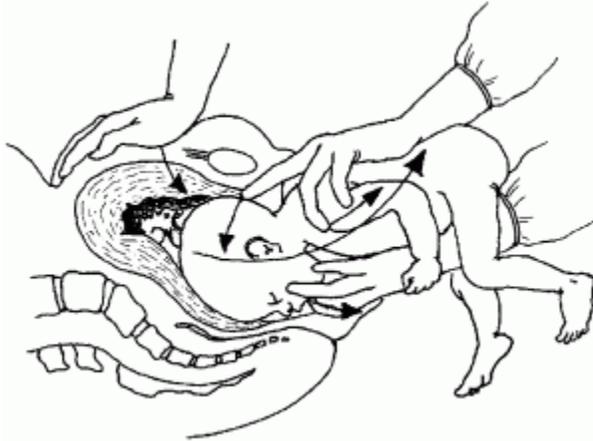
Sonography could lead to a false negative diagnosis if performed at very early gestational ages.

249. Both Mullerian and Wolffian duct structures occurring together is impossible in
- a) Anti-Mullerian hormone deficiency
  - b) FSH receptor mutation
  - c) Gonadal dysgenesis
  - d) Ovotesticular syndrome
250. Conversion of mole into choriocarcinoma is indicated by all except
- a) Plateau HCG level
  - b) Sub urethral nodule
  - c) Enlarged uterine size
  - d) Persistence of theca lutein cyst
251. Weight gain during pregnancy depends on all except
- a) Socioeconomic status
  - b) Pre-gestational weight
  - c) Smoking
  - d) Ethnicity

252. What is not done in shoulder dystocia

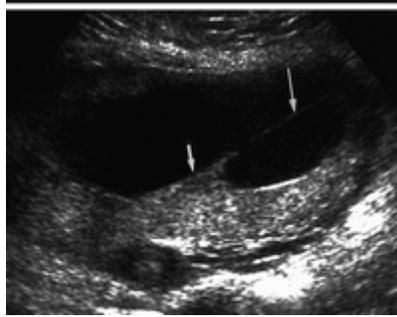
- a) Woods corkscrew maneuver
- b) McRoberts maneuver
- c) Suprapubic pressure
- d) Mauriceau-Smellie-Veit technique

Option D – It is used for delivery of the aftercoming head of breech (controlled delivery of breech fetal head). The fetal head should be maintained in a flexed position during delivery to allow passage of the smallest diameter of the head. The flexed position can be accomplished by using the Mauriceau Smellie Veit maneuver, in which the operator's index and middle fingers lift up on the fetal maxillary prominences, while the assistant applies suprapubic pressure.



253. Which of the following is true about multiple pregnancy
- a) Same sex rules out dichorionicity
  - b) Twin peak sign is seen in dichorionicity
  - c) Thick separating membrane is present in mono chorionic twins
  - d) Best detected after 16 weeks of gestation

The twin peak sign refers to a triangular projection of placental tissue extending from the placental surface, insinuating itself between the layers of the intertwin membrane, and seen on ultrasonographic (US) studies of multiple gestations. This placental tissue is widest at the placental surface and tapers gradually at a variable distance into the intertwin membrane (Fig 1) (1). This sign has also been referred to as the  $\lambda$  sign.

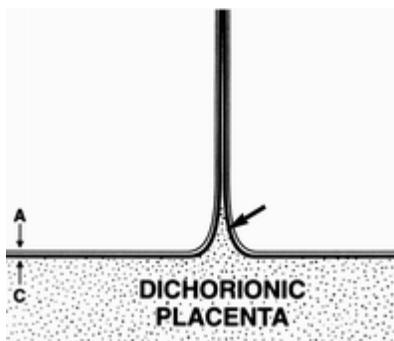


**Figure 1.**

Transverse image of the placenta of a twin pregnancy at 17 weeks gestational age reveals a triangular peak of villi (short arrow) extending from the placenta into the intertwin membrane (long arrow): the twin peak sign. Amniocentesis confirmed the pregnancy to be dizygotic, with male and female fetuses.

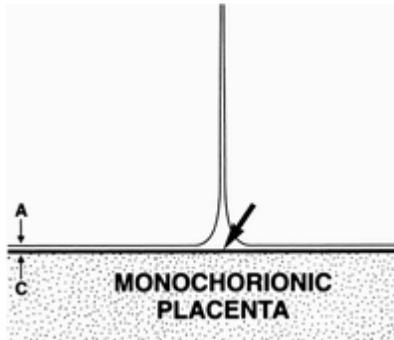
## EXPLANATION

The twin peak sign indicates the presence of a **dichorionic-diamniotic twin gestation**. The twin peak represents the extension of placental villi into the potential space that is formed from the reflection of apposed amniotic and chorionic layers from each fetus ([Fig 2](#)). It forms where two separate placentas grow contiguously and appear fused. The twin peak can be of variable size, and **only its presence is required to suggest that the pregnancy is dichorionic-diamniotic**. This sign is **most useful in assessing the chorionicity of pregnancies after 10 weeks**. Prior to this time, gestational sacs are readily recognizable and allow a rapid and accurate determination of chorionicity.



**Figure 2a.**

Drawing illustrates how the twin peak sign is reliable evidence of dichorionicity. **(a)** In a dichorionic pregnancy, both the amnions and the chorions reflect away from the placental surface, creating a potential space into which villi can grow. Large arrow shows the resulting twin peak sign. **(b)** Monochorionic diamniotic pregnancies have a single layer of continuous chorion (large arrow) limiting villous growth; the apposed amnions form a thin membrane separating the two amniotic cavities. *A* = amnion, *C* = chorion.



**Figure 2b.**

Drawing illustrates how the twin peak sign is reliable evidence of dichorionicity. **(a)** In a dichorionic pregnancy, both the amnions and the chorions reflect away from the placental surface, creating a potential space into which villi can grow. Large arrow shows the resulting twin peak sign. **(b)** Monochorionic diamniotic pregnancies have a single layer of continuous chorion (large arrow) limiting villous growth; the apposed amnions form a thin membrane separating the two amniotic cavities. *A* = amnion, *C* = chorion.

254. All of the following are associated with polycystic ovary syndrome except
- Ovarian ca
  - Endometrial ca
  - Insulin resistance
  - Osteoporosis
255. A lady presented with threatened abortion at 32 weeks of pregnancy. Which of the following is not useful
- Metronidazole for asymptomatic significant bacterial vaginosis
  - Dexamethasone for fetal lung maturity
  - Antibiotic prophylaxis even with unruptured membranes
  - Tocolytics for delayed labour

# PEDIATRICS

256. A mentally retarded child presented with history of infantile spasms and hypopigmented macule over the back. What is the likely diagnosis
- Tuberous sclerosis
  - Neurofibromatosis
  - Sturge-Weber syndrome
  - Linear epidermal nevus syndrome

*Tuberous sclerosis is a very imp topic. You should know everything about it.*

## SYNOPSIS : MUST-KNOW POINTS ABOUT TUBEROUS SCLEROSIS (Based on Nelson)

### Must know points

- It is an **Autosomal Dominant (AD)** disorder
- It results from inactivating mutations in either the **TSC1 gene encoding tuberin** or the **TSC2 gene encoding hamartin**

<u>Gene</u>	<u>Chromosome</u>	<u>Locus</u>	<u>Protein encoded</u>	<u>Function of protein</u>	<u>Incidence of mutation</u>	<u>Comments (if any)</u>
TSC1	9	9q34	Hamartin	Unknown	Less common	-
TSC2	16	16p13.3	Tuberin	Homology to GTPase activating protein	More common	Mnemonic : "TSC <u>T</u> wo encodes <u>T</u> uberin"

### **CLINICAL MANIFESTATIONS**

- SKIN LESIONS
- RETINAL AND BRAIN LESIONS
- LESIONS IN OTHER ORGANS

#### SKIN LESIONS

##### **Ash-leaf macules**

- Develop in infancy itself
- More than 90% of cases show the typical **hypomelanotic macules** that have been likened to an ash leaf on the trunk and extremities.

- Visualization of the hypomelanotic macule is enhanced by the use of a **Wood ultraviolet lamp**, particularly in the infant (see Chapter 553 ).
- **At least THREE hypomelanotic macules must be present**

#### Sebaceous adenomas (Adenoma sebaceum or Facial angiofibroma)

- develop **between 4 and 6 yr** of age
- increase with age
- they appear as tiny red nodules over the nose and cheeks and are sometimes confused with acne
- Later, they enlarge, coalesce, and assume a fleshy appearance

#### Shagreen patch

- is also characteristic of TS
- leathery thickenings in localized patches
- consists of a roughened, raised lesion with an orange-peel consistency
- located primarily in the **lumbosacral** region

#### Subungual or periungual fibromas

- arise from the **stratum lucidum** of the finger and toe in many patients with TS
- during adolescence

#### RETINAL AND BRAIN LESIONS.

##### Retinal lesions

It consist of two types:

- **mulberry tumors** that arise from the nerve head or round, flat gray lesions in the region of the disc
- **hamartoma** or depigmented areas

##### Brain lesions

The most common neurologic manifestations of TS consist of

- seizures
- cognitive impairment
- behavioral abnormalities including autism

The characteristic brain lesion is a **cortical tuber**.

Tubers are located in the convolutions of the cerebral hemispheres and are also present in the subependymal region, where they undergo calcification and project into the ventricular cavity, producing a  **Candle-dripping appearance**. ("candle-guttering.")

### The brain tumor most specific for TS – Subependymal astrocytoma

CT brain in TS shows the typical **CANDLE GUTTERING** appearance due to the projection of the tubers into the ventricular cavity.

TS may present during infancy with **infantile spasms** and a **hypsarrhythmic EEG pattern**. The seizures may be difficult to control and, at a later age, they may develop into myoclonic epilepsy

**Infantile spasms associated with TS are treated with vigabatrin**

### LESIONS IN OTHER ORGANS.

- **Rhabdomyomas** of the heart located at the **apex of the left ventricle**
- **Angiomyolipomas** are the **most common renal abnormality**
- Renal TS occurs in three forms: renal cysts, renal angiomyolipomas, and renal cell carcinoma

**Lymphangiomyomatosis** is the classical pulmonary lesion in TS

257. A month child presented with a history of running nose since 6 months, fever of 103-104F, conjunctival congestion. On blood test, Hb was normal, there was leucocytosis. No growth on culture was seen on conjunctival secretion. X-Ray showed opaque ethmoid sinus. What is next best investigation?
- Blood culture
  - CT scan orbit**
  - Repeat culture of conjunctival secretions
  - Urine culture

**Chronic rhinosinusitis** – **CT scan** is currently the method of choice for sinus imaging.

258. Which of the following predispose to develop epilepsy in later life after febrile convulsion EXCEPT
- Complex febrile seizure
  - Early age of onset**
  - Family history
  - Developmental history
259. Organism causing meningitis in patients of pediatric age group with recurrent CSF leaks is
- Meningococci
  - Streptococcus pneumoniae**
  - Hemophilus influenza

d) E coli

260. Bones in Scurvy are affected due to
- ↓ mineralization of bone
  - ↓ formation of osteoid matrix
  - Defective calcification of osteoid matrix
  - ↑ osteoclastic activity

In scurvy, the bony changes occur at the junction between the end of the diaphysis and growth cartilage. **Osteoblasts fail to form osteoid (bone matrix)**, resulting in **cessation of endochondral bone formation**. Calcification of the growth cartilage at the end of the long bones continues, leading to the thickening of the growth plate. The typical invasion of the growth cartilage by the capillaries does not occur. **Preexisting bone becomes brittle and undergoes resorption at a normal rate, resulting in microscopic fractures** of the spicules between the shaft and calcified cartilage. With these fractures, the periosteum becomes loosened, resulting in the classic subperiosteal hemorrhage at the ends of the long bones. Guidelines for the evaluation of fractures in infants and young children have been established. Intra-articular hemorrhage is rare because the periosteal attachment to the growth plate is very firm.

261. A boy presented with progressive weakness in lower limbs with hypertrophy of calf muscle and Gower's sign positive. His creatine phosphokinase (CPK) is 10,000 IU. The boy is suffering from
- Duchenne muscular dystrophy**
  - Spinal muscular atrophy
  - Myotonia congenital
  - Myotonia dystrophica

- Waddling gait**, manifesting in children aged 2-6 years, is **often the first symptom** in patients with Duchenne muscular dystrophy and is secondary to hip girdle muscle weakness.
- Sometimes a young boy may come to medical attention because of **elevated liver function enzymes (AST, ALT)**, and in such cases serum creatine kinases CK and GGT levels should be checked prior to considering liver biopsies. Occasionally a young boy may be referred for speech delay or learning issues, but he may turn out to harbor a dystrophin mutation. Most children with dystrophinopathy have IQs about one standard deviation lower than the general population.

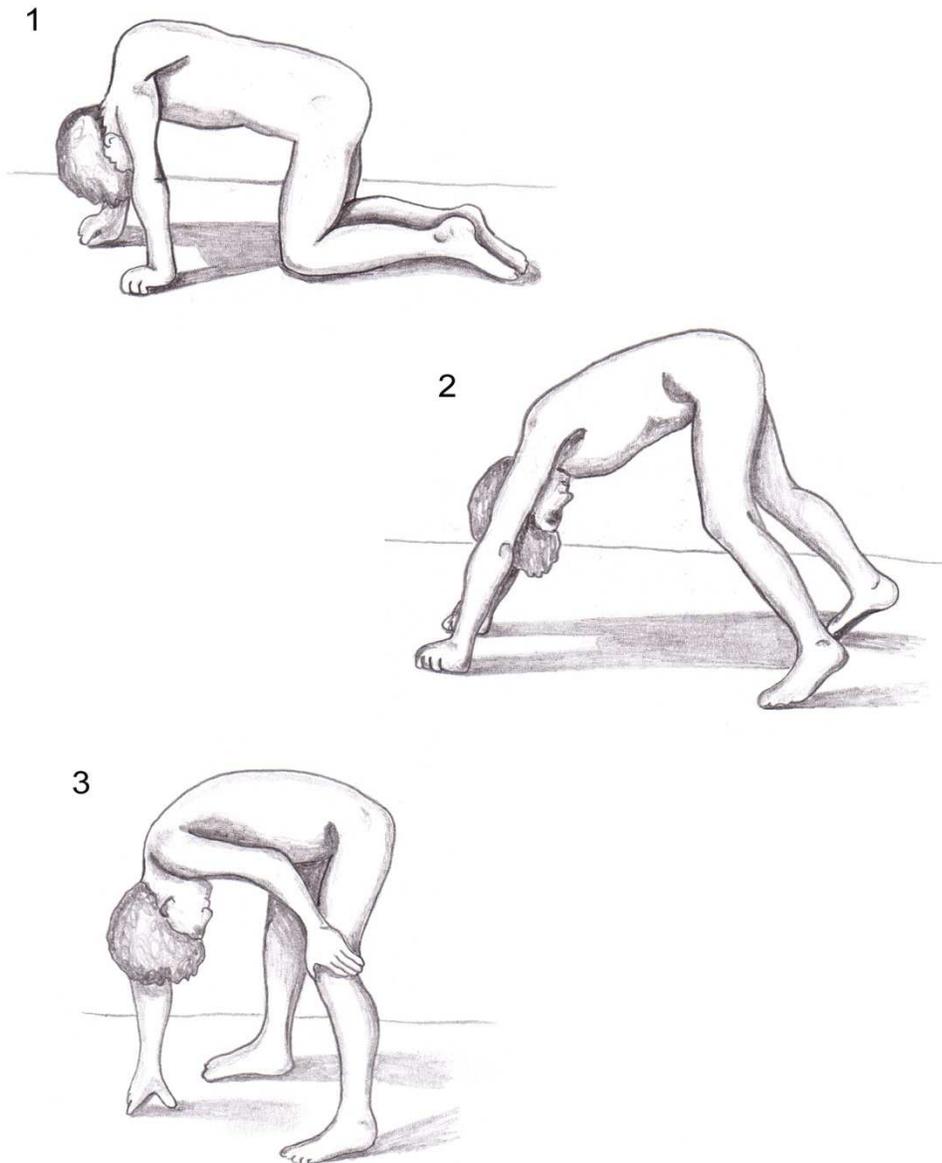
More children with Duchenne muscular dystrophy have **low intellectual skills** than children in the general population, but certainly plenty of exceptions exist. The low intellectual skills, such as cognitive issues (learning differences, attention deficit hyperactivity disorder, obsessive-compulsive disorder, pervasive developmental disorder, mental retardation), are seen in up to 30% of patients with dystrophinopathy. Children with Duchenne or Becker muscular dystrophy perform particularly poorly on tests of verbal skills and have challenges in processing complex verbal information.

- In some older boys or young men, **dilated cardiomyopathy** findings may lead to provincial diagnoses such as viral or idiopathic cardiomyopathy when in fact a dystrophin mutation may be the underlying reason.
- Because of **proximal lower back and extremity weakness**, parents often note that the **boy pushes on his knees in order to stand**; this is known as **Gowers sign**.



#### Gowers sign.

- The **calf enlargement** imparts the illusory appearance of strength, but, in fact, the enlarged calf muscles are caused by **fatty and fibrotic infiltration of degenerated muscles**. This is seen in conjunction with more prominent **toe-walking**. Sometimes an apparent **pseudohypertrophy is also seen in the forearms and tongue**. However, another explanation may relate to compensatory hypertrophy of the calves secondary to weak tibialis anterior muscles, which tend to be affected earlier and more prominently.
- **Contractures**
- Inexorable progressive weakness is seen in the proximal musculature, initially in the lower extremities, but later involving the neck flexors, shoulders, and arms.
- Cardiac surveillance should be implemented at time of diagnosis and should incorporate **echocardiography** plus ECG and pediatric cardiology expertise.
- Around the age of 8 years, most patients notice **difficulty with ascending stairs** and respiratory muscle strength begins a slow but steady decline.
- Approximately the time that independent ambulation is most challenged, the forced vital capacity begins to gradually wane, leading to **symptoms of nocturnal hypoxemia such as lethargy and early morning headaches**.
- Scoliosis may progress especially when more wheelchair dependent.
- If wheelchair bound and profoundly weak, patients develop terminal respiratory or cardiac failure, usually by the early 20s, if not sooner. Poor nutritional intake can also be a serious complication in individuals with severe end-stage Duchenne muscular dystrophy.
- Some families and individuals become socially withdrawn and may impact further on overall psychosocial health. Family, financial, school, community, and sibling issues can be significant.



### Mode of rising from the ground in pseudo-hypertrophic paralysis

In 1986, exactly 100 years after Gowers' keen observations, Kunkel identified the **Duchenne muscular dystrophy gene** located at **band Xp21** and provided molecular genetic confirmation of the **X-linked inheritance** pattern. The Duchenne muscular dystrophy gene was named **dystrophin**. **It is the largest recorded human gene** encoding a 427-kd protein, **dystrophin**. Dystrophin plays an integral role in sarcolemmal stability.

Duchenne and Becker muscular dystrophy **almost exclusively affect males** because of the X-linked inheritance pattern.

262. Most common cause of meningoencephalitis in children is
- Mumps
  - Arbovirus
  - Herpes
  - Enterovirus

Enteroviruses are the MC cause of viral meningoencephalitis. – Nelson 18<sup>th</sup>/2044

263. A child presented with vacant stare several times a day. Each episode lasts for about 20 seconds. The child is otherwise normal. What is the likely diagnosis?
- Grand mal seizures
  - Day dreaming
  - Absence seizures
  - ADHD
264. A 7 year girl is found to have asymptomatic elevated blood pressure on examinations. There is no other clinical sign. Urine examination is normal. What could the most probable cause?
- Essential HTN
  - PCKD
  - Chronic glomerulonephritis
  - Renal parenchymal ds

A review of literature revealed that 78% of 563 young patients with secondary hypertension had a renal parenchymal abnormality. In the remaining 22%, the cause of hypertension, in order of frequency, was renal artery stenosis, coarctation of the aorta, pheochromocytoma, and a variety of other conditions.

Table . Common Causes of Hypertension by Age

Infants	Children		Adolescents
	1-6 y	7-12 y	
Thrombosis of renal artery or vein	Renal artery stenosis	Renal parenchymal disease	Essential hypertension
Congenital renal anomalies	Renal parenchymal disease	Renovascular abnormalities	Renal parenchymal disease
Coarctation of the aorta	Wilms tumor	Endocrine causes	Endocrine causes
Bronchopulmonary dysplasia	Neuroblastoma	Essential hypertension	
	Coarctation of the aorta		

# ORTHOPAEDICS

265. One of the following doesn't occur in articular cartilage in old age
- Water content ↓
  - Proteoglycan content ↓
  - Proteoglycan synthesis ↓
  - Proteoglycan degradation ↑
266. In Klippel Feil triad, all are seen except
- Short neck
  - Low hair line
  - Limited neck movement
  - Elevated scapula
267. A patient presented after RTA with quadriparesis, sphincter incontinence, sensory loss up to the level of sternum, with RR 30/min and HR 110/min. What is the level of lesion?
- C1-C2
  - C4-C5
  - Medulla
  - T1-T2
268. Median nerve lesion at the wrist causes all except
- Thenar atrophy
  - Adductor pollicis paralysis
  - Lumbricals of 1<sup>st</sup> & 2<sup>nd</sup> finger affected
  - Flexor pollicis brevis affected
269. All of the following is true about synovial sarcoma except
- Originate from synovial lining
  - Seen in pts less than 60 yrs
  - Occurs more at extra-articular sites
  - Knee & foot are common sites involved

The origin of synovial cell sarcoma is unclear. In contrast to its name, synovial cell sarcoma is not associated with synovial joints. Because of the similarity between cells of this tumor and primitive synoviocytes, the term synovial cell sarcoma has been used.

A neurologic origin has been suggested. In fact, there is a histologic resemblance between neural cells of malignant peripheral nerve sheath tumor (MPNST) and synovial cell sarcoma. Typically, synovial cell sarcoma is associated with a history of a long-standing nodule, sometimes present for years, which increases rapidly in size over a few months; therefore, it is

sometimes overlooked. The tumor spreads along fascial planes and, thus, can be much more widespread than apparent on initial evaluation.

270. Lift off test is done for
- Supraspinatus
  - Infraspinatus
  - Teres minor
  - Subscapularis

Gerber and Krushall originally described the lift-off test for isolating a subscapularis injury. The test is performed by passively internally rotating the arm behind the back and off the patient's spine. A positive test occurs when the examiner releases the maximally internally rotated arm, and it falls to the patient's back. A weak test result is observed when the patient can maintain the position of maximal internal rotation but is unable to apply resistance to the examiner's hand.

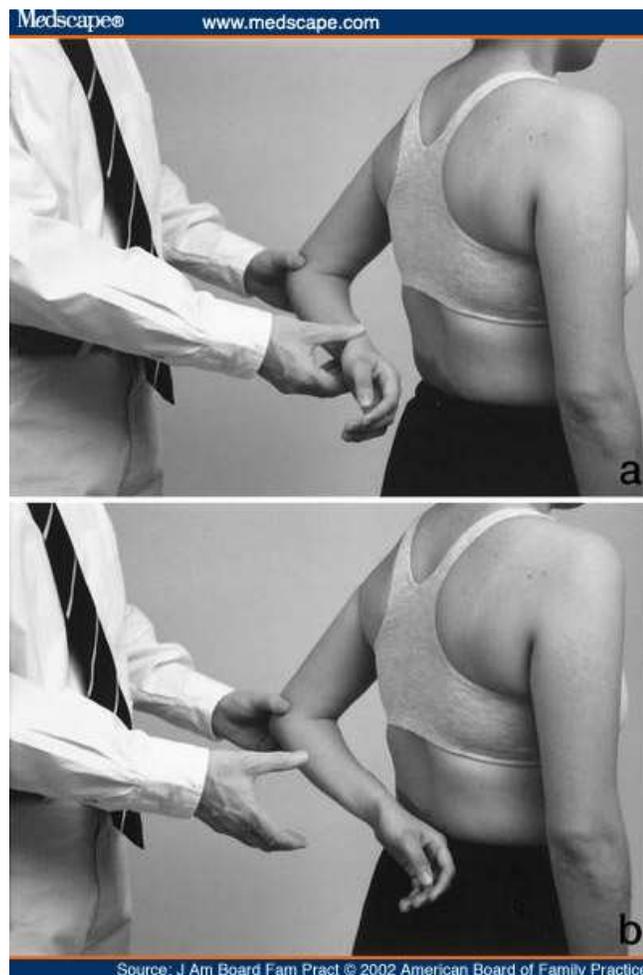


Fig. Positive lift-off test: (a) starting position, and (b) end position: Note that the patient is not capable of holding her forearm off her low back.

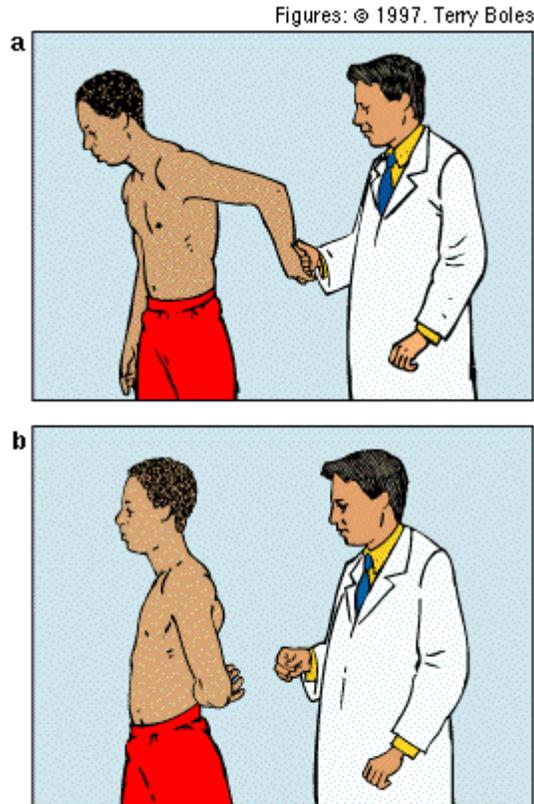


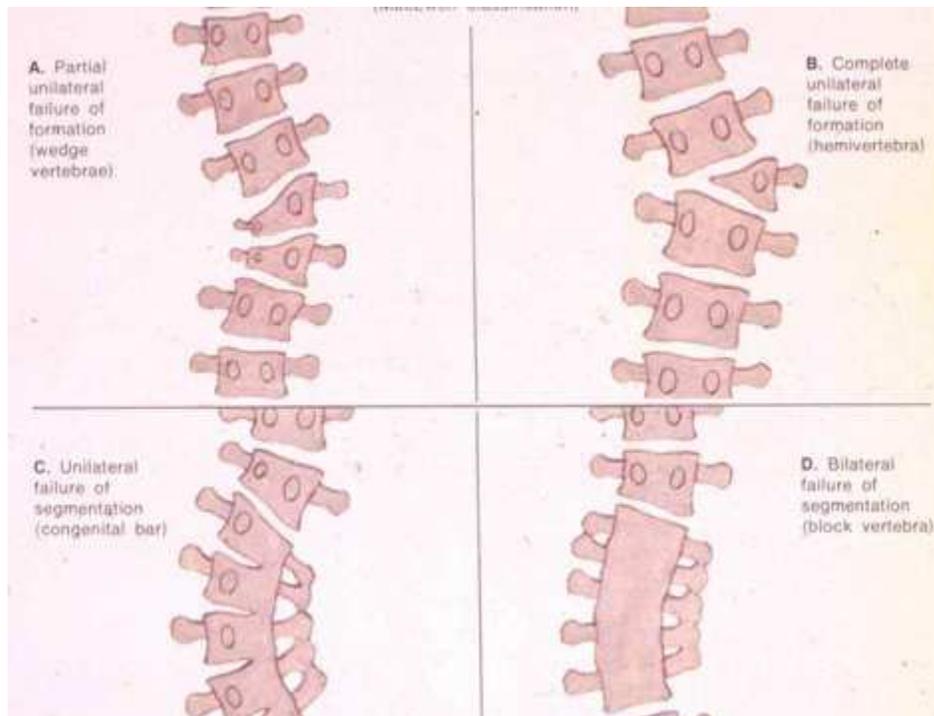
Figure 4. The subscapularis lift-off test is normal (a) when the patient can maintain the arm in a fully extended, maximally internally rotated position with the elbow flexed against resistance. It is abnormal (b) when the patient is unable to move the back of the hand away from the back because of subscapularis tear.

271. Congenital scoliosis progression is least likely to be seen with
- Hemi vertebrae
  - Wedge vertebrae
  - Block vertebrae
  - Unilateral unsegmented vertebrae with bar

- Type of vertebral anomaly - The type of anomaly that causes the most severe scoliosis is a unilateral unsegmented bar with contralateral hemivertebrae at the same level. Next in severity is a scoliosis caused by a unilateral unsegmented bar alone, followed by 2 unilateral fully segmented hemivertebrae, a single fully segmented hemivertebra, and a wedge vertebra. The least severe scoliosis is caused by a block vertebra. Congenital scoliosis caused by unclassifiable anomalies can be difficult to predict and requires careful monitoring. A poor prognosis is associated with a unilateral unsegmented bar with or without contralateral hemivertebra, which should be treated immediately, without a period of observation (see table below)..

**Table - Vertebral Anomalies Leading to Congenital Scoliosis**

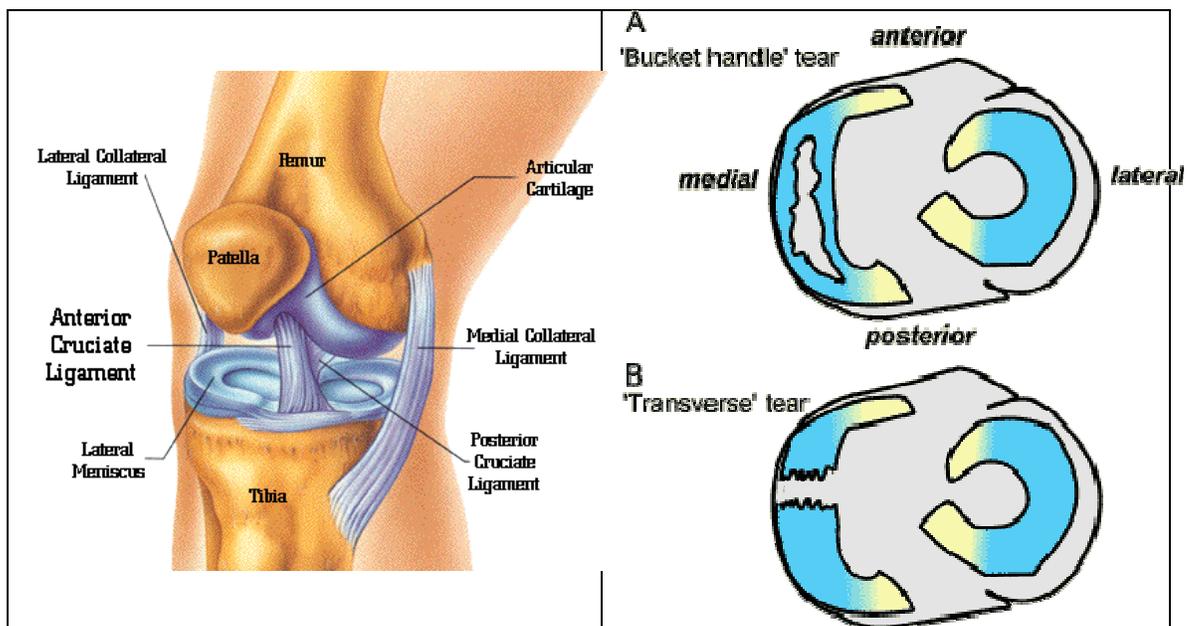
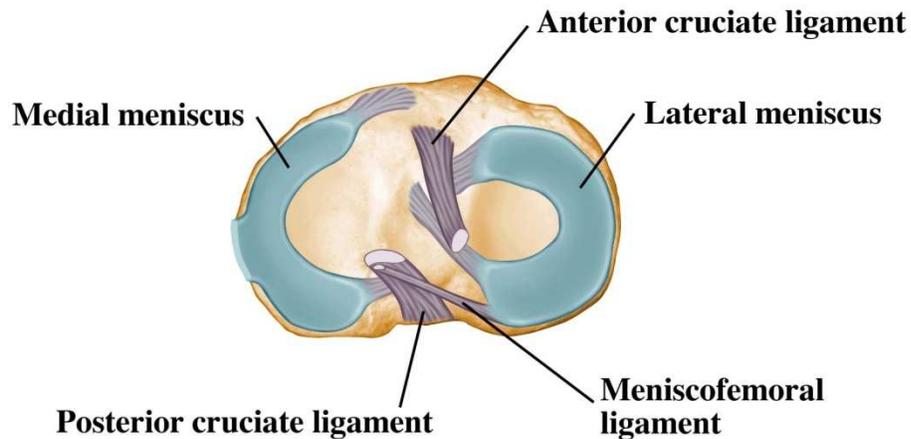
Risk of Progression (Highest to Lowest)	Curve Progression
Unilateral unsegmented bar with contralateral hemivertebrae	Rapid and relentless
Unilateral unsegmented bar	Rapid
Fully segmented hemivertebra	Steady
Partially segmented hemivertebra	Less rapid
Incarcerated hemivertebra	May slowly progress
<b>Unsegmented hemivertebra</b>	<b>Little progression</b>



272. After total hip replacement, on 2<sup>nd</sup> post operative day, a patient developed breathlessness and severe chest pain. Echo shows right ventricular dysfunction and TR. What is the most likely reason for the condition?
- MI
  - PE**
  - Hypotensive shock
  - Cardiac tamponade

273. Which of the following is not true about menisci
- Medial menisci is more mobile than lateral
  - Lateral menisci covers more tibial articular surface than medial
  - Menisci are made of Type I collagen
  - Medial meniscus is more commonly injured than lateral

Medial menisci is **less mobile** (and hence more prone to injury).



274. Metal on metal articulation is avoided in
- a) Primary failure
  - b) Young female
  - c) Inflammatory arthritis
  - d) Revision surgery

There are a lot of concerns with metal particles, so using metal-on-metal articulations in women younger than 40 years of age is contraindicated. Gender is probably the best known risk factor for pseudotumors following resurfacing hip replacement.

## SKIN

275. A young lady presented with white lacy lesions in the oral cavity and her proximal nail fold had extended onto the nail bed. What is the likely diagnosis?
- Psoriasis
  - Geographic tongue
  - Lichen planus
  - Candidiasis

Mucous membrane involvement is common in lichen planus and may be found without skin involvement. Lesions are most commonly found on the tongue and the buccal mucosa; they are characterized by white or gray streaks forming a linear or reticular pattern on a violaceous background (see the image below). Oral lesions are classified as reticular, plaque-like, atrophic, papular, erosive, and bullous. Ulcerated oral lesions may have a higher incidence of malignant transformation in men, but this observation may be confounded by other factors, such as smoking and chewing tobacco. Lesions may also be found on the conjunctivae, the larynx, the esophagus, the tonsils, the bladder, the vulva, and the vaginal vault; throughout the gastrointestinal tract; and around the anus.



276. Treatment of erythematous skin rash with multiple pus lakes in a pregnant woman is
- Corticosteroids
  - Isotretinoin
  - Methotrexate
  - Psoralen with PUVA

DOC for pustular psoriasis in pregnancy - Prednisolone

# ANAESTHESIA

277. A patient had serum bilirubin value of 8 mg/dl and serum creatinine 1mg/dl. What will be the muscle relaxant of choice in this patient ?
- Vecuronium
  - Pancuronium
  - Atracurium
  - Succinylcholine
278. A 27 year old female was brought to emergency department for acute abdominal pain following which she was shifted to operation theatre for laparotomy for surgical management of ileal perforation for which speedy intubation was done. Immediately after intubation her ETCO<sub>2</sub> raised and breath sounds were decreased on left side. What could be the possible cause?
- ETT blockage
  - Bronchospasm
  - Esophageal intubation
  - Endobronchial intubation on right side
- Atracurium undergoes Hoffmann elimination; hence can be used in renal failure.
279. A 25 year old overweight female was given fentanyl 1 pancuronium midazolam anesthesia for a surgery. After surgery she was extubated and on examination her chest wall motion was slow and she was unable to move her upper body. She was conscious and able to undertsnad but was lacking voluntary respiratory effort. Her blood pressure was normal and heart rate was also normal. What is the diagnosis ?
- Incomplete reversal of pancuronium
  - PE
  - Fentanyl induced chest wall rigidity
  - Respiratory depression
280. Not true about Succinylcholine is
- Train of Four phenomenon seen
  - TOF >0.4
  - Shows fade phenomenon
  - No post titanic facilitation

# RADIOLOGY

281. Which of the following is not a CT feature of adrenal adenoma
- a) Low attenuation
  - b) Homogenous density & well defined borders
  - c) Enhances rapidly, contrast stays in it for a long time & washes out late
  - d) Calcification is rare

CT and MRI are the modalities of choice in diagnosing adrenal cortical adenoma. On CT scans, adrenal cortical adenomas are **well-circumscribed mass lesions** that are **homogeneous** in their attenuation and enhancement patterns. The evaluation should be performed by using sections that are 5 mm or thinner to ensure that attenuation measurements are not affected by volume averaging.

The use of a sufficient milliampere-second (mAs) setting is important so that the measured attenuation values do not have a significant standard deviation. Heterogeneous enhancement or attenuation can be observed when a lipid-rich adenoma and a lipid-poor adenoma coexist. **A lesion that is poorly margined with heterogeneous enhancement is unlikely to be a simple benign adrenal cortical adenoma**, and other entities must be considered.



Homogeneous, well-defined, 7-HU ovoid mass is seen in the right adrenal gland; this finding is diagnostic of a benign adrenal adenoma.



Contrast-enhanced CT scan demonstrates a **homogeneously** enhancing ovoid mass in the left adrenal gland. As in this case, attenuation measurements of adrenal masses on contrast-enhanced CT scans are frequently nondiagnostic.



**Homogeneously enhancing** ovoid mass is seen in the left adrenal gland.



Dynamic and delayed contrast-enhanced CT scans demonstrate a homogeneously enhancing mass in the right adrenal gland. The degree to which enhancement diminishes over time is referred to as washout, which can be calculated by using the following formula:  $[1 - (\text{attenuation at 10 minutes}/\text{attenuation at 80 seconds})] \times 100$ , where the attenuations are in Hounsfield units. In this case, the washout equals  $[1 - (36/99)] \times 100$ , or 64%. Findings from a recent publication in a major journal suggests that **any washout greater than 50% is diagnostic of a benign adrenal adenoma**. Further studies are needed to confirm these promising results.

#### CT examination with intravenously administered contrast material

The initial enhancement patterns of adrenal cortical adenomas and adrenal metastases overlap substantially; therefore, simple attenuation measurements are not useful in distinguishing between the two. A delayed attenuation measurement (obtained 10 minutes after the injection) of 30 HU or less is diagnostic of benign adenoma, but only a small percentage of adrenal adenomas have this finding.

A calculation termed **contrast-agent washout** can be used to reliably determine if an adrenal mass is benign or malignant. Washout is calculated as follows:

1. Intravenous contrast agent is administered, and a scan is obtained after an 80-second delay.
2. A subsequent scan is obtained after a 10-minute delay.
3. A region of interest is drawn over the adrenal mass, and the attenuation is measured in Hounsfield units at 80 seconds and at 10 minutes.
4. The percentage of contrast agent washout is equal to  $[1 - (\text{attenuation at 10 minutes}/\text{attenuation at 80 seconds})] \times 100$ , where the attenuations are in Hounsfield units.

Washout is a measurement of the percentage decrease between the initial enhancement and the delayed enhancement. A large decrease is a high-percentage washout, and a small decrease is a low-percentage washout. If delayed enhancement is exactly half of the initial enhancement, the washout is exactly 50%.

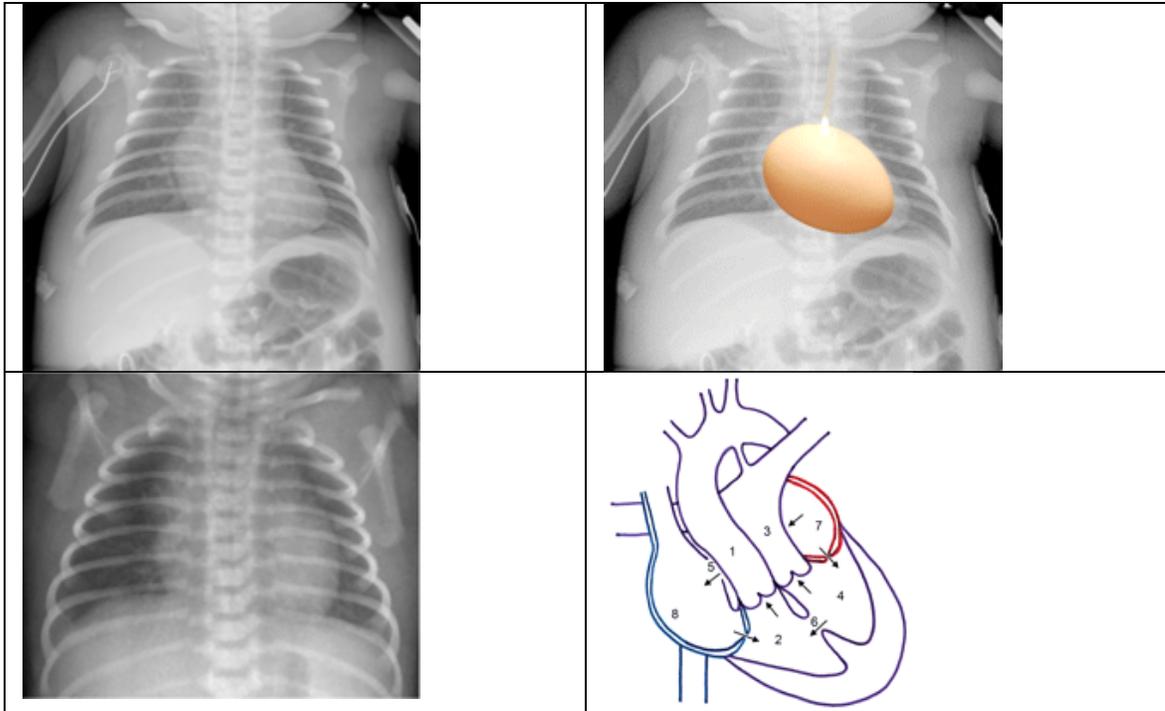
In a series of 101 adrenal masses, a **washout of greater than 50% was specific for benign adrenal adenoma**, and a **washout of less than 50% was specific for metastasis**. Interestingly, these findings are not correlated with the percentage of intracytoplasmic lipid, and the physiologic mechanism resulting in this distinction is not well understood. With a threshold of 50%, use of the washout value yields 98% sensitivity and 100% specificity.

282. Egg-on-side appearance is seen in
- Tricuspid atresia with VSD
  - TOF
  - TAPVC
  - TGA**

## Transposition of the Great Arteries and Egg-on-a-String Sign

Transposition of the great arteries, the **most common cyanotic congenital heart lesion found in neonates**, accounts for 5%–7% of congenital cardiac malformations. It is **most common in infants of diabetic mothers**. It is isolated in 90% of those affected and rarely is associated with a syndrome or an extracardiac malformation.

In the normal anatomy, the aorta is anterior to and at the right of the pulmonary artery; in transposition of the great arteries, the **pulmonary artery is situated to the right of its normal location and is obscured by the aorta on frontal chest radiographs**. This malposition, in association with stress-induced thymic atrophy and hyperinflated lungs, results in the **apparent narrowing of the superior mediastinum on radiographs**, the most consistent sign of transposition of the great arteries. The **cardiovascular silhouette** varies from normal in the first few days after birth to **enlarged and globular, with the classic appearance described as an egg on a string** (Fig 1). The right atrial border is abnormally convex, and the left atrium commonly is enlarged because of increased pulmonary blood flow. The appearance of the enlarged heart at chest radiography also has been likened to the profile of an **egg on its side** (1–3). The specific radiologic features are determined by the extent to which the great arteries are superposed in the plane of imaging, the size of the communication between the pulmonary and the systemic circulation, and the presence and severity of obstruction to pulmonary flow.



**Figure 1.** Transposition of the great arteries compared with normal anatomy. **(a)** Chest radiograph obtained in a neonate shows narrowing of the superior mediastinum, enlargement of the cardiac silhouette with abnormal convexity of the right atrial border, and increased vascular flow—typical features of transposition of the great arteries. **(b)** Same image as **a** with a superimposed drawing shows the characteristic cardiomeastinal silhouette: the **egg-on-a-string sign**. **(c)** Chest radiograph obtained in another neonate shows the normal appearance of the mediastinum, with a normal thymic shadow. **(d)** Drawing shows the pattern of blood flow (arrows) through the heart with transposition of the great arteries. The aorta (1) arises from the right ventricle (2), and the pulmonary artery (3) arises from the left ventricle (4). Communication between the systemic and the pulmonary circulation—an interatrial septal defect (5), an interventricular septal defect (6), or both—sustains life by allowing oxygenated blood from the left atrium (7) to mix with deoxygenated blood from the right atrium (8) before it flows via the right ventricle to the aorta and via the left ventricle to the pulmonary artery.

Transposition of the great arteries is produced by a **ventriculoarterial discordance** in which the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle (Fig 1d). To sustain life, a communication (eg, a patent foramen ovale, atrial septal defect, ventricular septal defect, or a combination of these) must be present between the systemic and the pulmonary circulation, in addition to systemic collateral arteries. The volume of the pulmonary flow may be normal in the first few days after birth, but it increases with closure of the ductus arteriosus. This increase may be mild to severe, depending on the size of the communication between the systemic and pulmonary vessels. In the presence of a large communication, the vessels are usually prominent. A large communication also leads to enlargement of the heart unless the shunt is balanced or impeded by an obstruction of the pulmonary artery.

## Imaging Findings in Congenital Heart Diseases

### Summary

TGA – Egg-on-a-String sign

TAPVR – Snowman sign

PAPVR – Scimitar sign

ECD – Gooseneck deformity

TOF – Boot-shaped heart

CoA – Figure-of-3 & Reverse Fig-of-3 sign

Ebstein anomaly – Box shaped heart

283. Most ionizing radiation is
- Alpha
  - Beta
  - X-ray
  - Gamma
284. Difference between X-ray and light is
- Energy
  - Mass of photon
  - Type of wave
  - Speed
285. A patient presented with acute renal failure and anuria. His USG was normal. Which of the following investigation will give best information regarding kidney function?
- IVP
  - Retrograde pyelography
  - Antegrade pyelography
  - DTPA
286. Dense nephrogram can be obtained by
- Dehydrate the pt
  - ↑dye concentration
  - Injecting the dye rapidly
  - Non-ionic media used

The quantity of contrast medium administered should be related to the weight of the patient (300 mg kg<sup>-1</sup> body weight). If the patient is well hydrated the dose may be increased to 600 mg kg<sup>-1</sup> body weight. The contrast medium should be **injected rapidly** so that a bolus reaches the **kidneys**. This **high concentration arriving at the glomeruli will produce a high concentration in**

the nephrons, and thus a denser nephrogram and subsequent pyelogram than would otherwise be the case. – Grainger & Allison's Diagnostic Radiology 5<sup>th</sup>/Ch 38

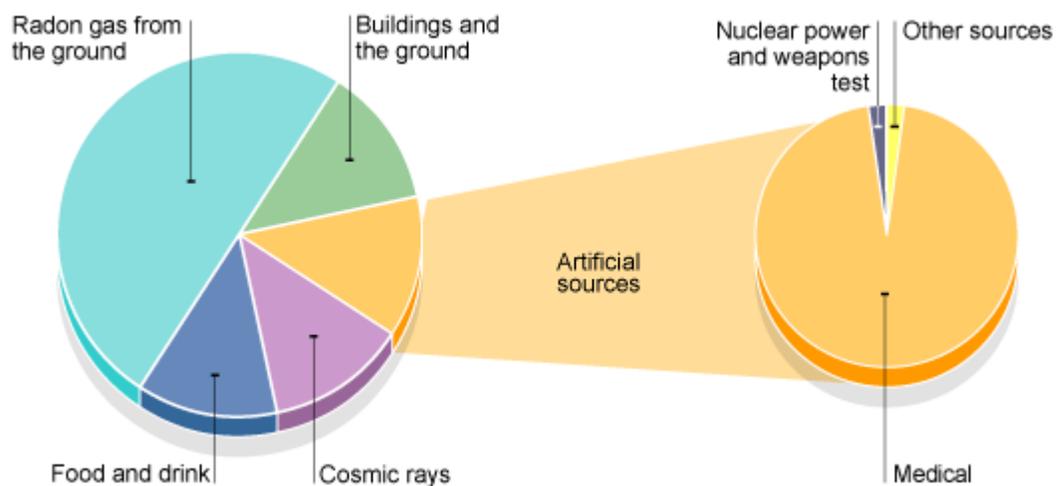
287. What is the background radiation
- Radiation after disaster
  - Radiation after atom bomb explosion
  - Radiation present constantly everywhere on earth
  - Radiation present in the background during radiological investigations

**Background radiation** is constantly present in the environment and is emitted from a variety of natural and artificial sources. Primary contributions come from:

- Sources in the earth. These include sources in food and water, which are incorporated in the body, and in building materials and other products that incorporate those radioactive sources;
- Sources from space, in the form of cosmic rays;
- Sources in the atmosphere. One significant contribution comes from the radon gas that is released from the Earth's crust and subsequently decays into radioactive atoms that become attached to airborne dust and particulates. Another contribution arises from the radioactive atoms produced in the bombardment of atoms in the upper atmosphere by high-energy cosmic rays.

Naturally occurring sources are responsible for the vast majority of radiation exposure. However, about 3% of background radiation comes from man-made sources such as:

- Self-luminous dials and signs
- Global radioactive contamination due to historical nuclear weapons testing
- Nuclear power station or nuclear fuel reprocessing accidents
- Normal operation of facilities used for nuclear power and scientific research
- Emissions from burning fossil fuels, such as coal fired power plants
- Emissions from nuclear medicine facilities and patients



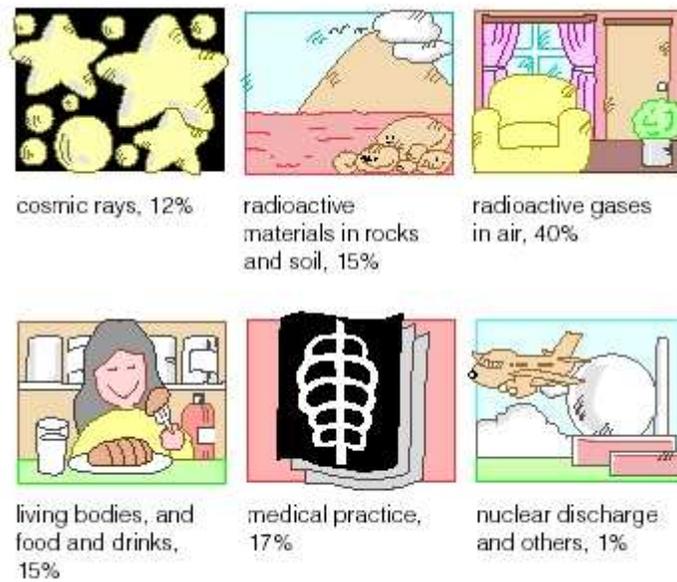


Fig. Sources of Background Radiation

288. CT scan room doors are made up of
- Glass
  - Tungsten
  - Steel
  - Lead
289. The total of radiation for which a particular organ is exposed is called as
- Equivalent dose
  - Absorbed dose
  - Exposure dose
  - Effective dose

The **equivalent dose** ( $H_T$ ) is a **measure of the radiation dose to tissue** where an attempt has been made to allow for the **different relative biological effects of different types of ionizing radiation**. Equivalent dose is therefore a less fundamental quantity than radiation absorbed dose, but is **more biologically significant**. Equivalent dose has units of **sieverts**. Another unit, Röntgen equivalent man (REM or rem), is still in common use in the US, although regulatory and advisory bodies are encouraging transition to sieverts (100 Röntgen equivalent man = 100 REM = 1 sievert.)

Equivalent dose ( $H_T$ ) is calculated by **multiplying the absorbed dose to the organ or tissue ( $D_T$ ) with the radiation weighting factor,  $w_R$** . This factor is selected for the type and energy of the radiation incident on the body, or in the case of sources within the body, emitted by the source. The value of  $w_R$  is 1 for x-rays, gamma rays and beta particles, but higher for protons, neutrons, alpha particles etc.

$$H_{T,R} = w_R \times D_{T,R}$$

Where  $H_{T,R}$  = equivalent dose to tissue  $T$  from radiation  $R$

$D_{T,R}$  = absorbed dose  $D$  (in grays) to tissue  $T$  from radiation  $R$

290. True about Stochastic effects is
- Causes erythema & cataract
  - Probability of occurrence is a function of dose
  - Has a threshold
  - Cancer chances  $\uparrow$  with dose

## Stochastic effects

Effects that occur by chance and which may occur without a threshold level of dose, whose probability is proportional to the dose and whose severity is independent of the dose. In the context of radiation protection, the main stochastic effect is cancer.

Stochastic effects are those that occur by chance and consist primarily of cancer and genetic effects. Stochastic effects often show up years after exposure. As the dose to an individual increases, the probability that cancer or a genetic effect will occur also increases. However, at no time, even for high doses, is it certain that cancer or genetic damage will result. Similarly, for stochastic effects, there is no threshold dose below which it is relatively certain that an adverse effect cannot occur. In addition, because stochastic effects can occur in individuals that have not been exposed to radiation above background levels, it can never be determined for certain that an occurrence of cancer or genetic damage was due to a specific exposure.

# PSYCHIATRY

291. Autistic disorder has all except
- a) Visual impairment
  - b) Lack of social impairment
  - c) Delayed development of speech
  - d) Stereotypic movements
292. Alcoholic paranoia is associated with
- a) Fixed delusions
  - b) Hallucinations
  - c) Drowsiness
  - d) Impulse agitation

## Pathology caused by alcohol drinking

**Delirium tremors:** This results from the long continued action of the poison on the brain. It occurs in chronic alcoholics due to:

(1) Temporary excess (2) sudden withdrawal (3) shock after receiving an injury, such as fracture of a bone, or (4) from acute infection, such as pneumonia, influenza, erysipelas etc

It typically begins 72 to 96 hours after the last drink. There's an acute attack of insanity in which the main symptoms are coarse muscular tremors of the face, tongue and hands, insomnia, restlessness, loss of memory, agitation, confusion, disorientation, uncontrollable fear and has tendency to commit suicide, homicide or violent assault or to cause damage to property. Other symptoms are diarrhea, dilated pupils, fever, tachycardia, tachypnoea and hypertension. There is disorientation as to time and place and a peculiar kind of delirium of horrors owing to hallucination of the sight and hearing. The patient imagines that insects are crawling under the skin, or snakes are crawling on his bed it is considered unsoundness of mind and not intoxication. To control agitation diazepam should be given

**Alcoholic Polyneuritis and Korsakoff's Psychosis:** The symptoms of Polyneuritis are weakness; pain in the extremities, wrist and foot drop, unsteady gait, loss of deep reflexes and tenderness of muscles of arms and legs

**Alcoholic paranoia:** In this there are fixed delusions but no hallucinations. The person becomes deeply suspicious of the motives and actions of those he meets and of his family members.

**Acute Alcoholic hallucinations:** Persistent hallucinations develop within 48 hours after cessation of alcohol intake. The hallucinations may be auditory or visual and their content is usually unpleasant and disturbing. The disorder may last several weeks or months

**Alcoholic epilepsy:** seizures occur after a day or more of the terminations of a drinking session. Sometimes the attack may occur while the patient is actually drinking

**Wernicke Encephalopathy:** This results from a brain or spinal cord lesion due to heavy drinking, Vitamin

B1 deficiency occurs

Symptoms include disturbance of consciousness, drowsiness, amnesia, peripheral neuropathy, external ocular palsies and stupor. It has a high mortality and can cause death in 24 hours. If untreated it can progress to a more chronic condition called Korsakoff psychosis, in which impairment of short term memory with inability to learn new information and confabulation (recitation of imaginary experiences to fill gaps in the memory) are seen

**Cardiac dysrhythmias:** In alcohol withdrawal tachyarrhythmias are common probably because of high adrenergic nervous system activity, which may cause sudden death.

**Marchiafava Syndrome:** Degeneration of the corpus callosum may occur in alcoholics.

**Mallory-Weiss Syndrome:** Ruptured esophagus with mediastinitis occurs

Other pathologies include **Malnutrition, Gastric and peptic ulcer, Cirrhosis, Myocarditis pancreatitis and Mental illness**

293. A 30 year old man since 2 months suspects that his wife is having affair with his boss. He thinks his friend is also involved from abroad and is providing technology support. He thinks people talk ill about him. His friends tried to convince him but he is not convinced at all. Otherwise he is normal, he doesn't have any thought disorder or any other inappropriate behavior. It is case of
- Paranoid personality disorder
  - Persistent delusion disorder
  - Schizophrenia
  - Acute & transient psychosis

Delusional disorder is an illness characterized by the presence of **nonbizarre delusions** in the absence of other mood or psychotic symptoms, according to the *Diagnostic Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR)*. It defines delusions as false beliefs based on incorrect inference about external reality that persist despite the evidence to the contrary and these beliefs are not ordinarily accepted by other members of the person's culture or subculture.

- **Jealous type**
  - Related terms include conjugal paranoia, **Othello syndrome**, and pathological or morbid jealousy.
  - The main theme of the delusions is that **her or his spouse or lover is unfaithful**. Some degree of infidelity may occur; however, patients with delusional jealousy support their accusation with delusional interpretation of "evidence" (eg, disarrayed clothing, spots on the sheets).

- Patients may attempt to confront their spouses and intervene in imagined infidelity. Jealousy may evoke anger and empower the jealous individual with a sense of righteousness to justify their acts of aggression. Both the intimate partner and the (perceived) lover may be the targets of aggression and violence. This disorder can sometimes lead to acts of violence, including suicide and homicide.
- Easton et al indicate that *DSM-IV-TR* criteria are not inclusive enough to diagnose this subtype. They looked at a database of 398 patients with a jealousy disorder and found that only 4% met diagnostic criteria for delusional disorder-jealous type.
- **Persecutory type**
  - Most common type of delusional disorder.
  - Most commonly associated with comorbid Axis 1 disorders.
  - Patients believe that they are being persecuted and harmed.
  - In contrast to persecutory delusions of schizophrenia, the delusions are systematized, coherent, and defended with clear logic. No deterioration in social functioning and personality is observed.
  - Patients are often involved in formal litigation against their perceived persecutors. Munro refers to an article by Freckelton who identifies the following characteristics of deluded litigants: determination to succeed against all odds, tendency to identify the barriers as conspiracies, endless drive to right a wrong, quarrelsome behaviors, and "saturating the field" with multiple complaints and suspiciousness.
  - Patients often experience some degree of emotional distress such as irritability, anger, and resentment. In extreme situations, they may resort to violence against those who they believe are hurting them.
  - The distinction between normality, overvalued ideas, and delusions is difficult to make in some of the cases.

294. Not a cognitive dysfunction is
- a) Catastrophic thinking
  - b) Arbitrary inference
  - c) Overgeneralization
  - d) Thought block

Based on the work of Aaron Beck and others, in *Feeling Good: The New Mood Therapy*, David Burns outlines 10 common mistakes in thinking, which he calls cognitive distortions.

- 1) All or nothing thinking
- 2) Overgeneralization
- 3) Mental filter
- 4) Disqualifying the positive
- 5) Jumping to conclusions
- 6) Magnification & minimization
- 7) Emotional reasoning
- 8) Shoulding
- 9) Labeling & mislabeling

## 10) Personalisation &amp; blame

295. All are parts of cognitive behavior change techniques except
- Precontemplation
  - Consolidation**
  - Action
  - Contemplation

**Stages of habit change**

- Precontemplation
- Contemplation
- Preparation
- Action
- Maintenance
- Termination

296. A 60 year old male is brought by his wife. He thinks that he had committed sins all through his life. He is very much depressed and has considered committing suicide but has not thought how do go about it. He had also attended sessions with a spiritual guru. He is not convinced by his wife that he has lead a pious life. He does not want to hear anything on the contrary. How will you treat him?
- Antipsychotic + Antidepressant**
  - Antidepressant with cognitive behavioral therapy
  - Guidance & recounselling with guru + Antidepressant
  - Antidepressant alone
297. All of the following are done in behavior therapy to increase a behavior except
- Punishment**
  - Operant conditioning
  - Negative reinforcement
  - Modeling

**Punishment** leads to **↓ in a behavior**.

298. A 25 yr old lady presented with sadness, occasional palpitation, loss of appetite & insomnia. There is no complaint of hopelessness, suicidal thoughts & there is no past H/O any precipitating event. She is remarkably well in other areas of life. She is doing her office job normally & her social life is also normal. What is the probable diagnosis in this case?
- GAD
  - Mixed anxiety depression**
  - Adjustment disorder
  - Mild depressive episode

**Mixed anxiety-depressive disorder** is a diagnostic category defining patients who suffer from both anxiety and depressive symptoms of limited and equal intensity accompanied by at least some autonomic features. The World Health Organization's ICD-10 describes *Mixed anxiety and depressive disorder*: "...when symptoms of anxiety and depression are both present, but neither is clearly predominant, and neither type of symptom is present to the extent that justifies a diagnosis if considered separately. When both anxiety and depressive symptoms are present and severe enough to justify individual diagnoses, both diagnoses should be recorded and this category should not be used."

### Diagnostic features

- Low or sad mood.
- Loss of interest or pleasure.
- Prominent anxiety or worry.
- Multiple associated symptoms for example:
  - disturbed sleep
  - disturbed appetite
  - tremor
  - suicidal thoughts or self-harm
  - fatigue or loss of energy
  - dry mouth
  - palpitations
  - tension and restlessness
  - poor concentration
  - irritability
  - dizziness
  - sexual dysfunction.

### Extra Edge

The **Hamilton Anxiety Scale** is a commonly used instrument to assess anxiety disorders in general. The Generalized Anxiety Disorder Questionnaire for DSM-IV (GAD-Q-IV) is a more recent diagnostic tool, and is specific to GAD.

299. A pt with self-harm is prone to suicidal tendency. In which of the following condition it does not warrant a specialist referral?
- a) Formal thought disorder
  - b) Alcohol & drug abuse
  - c) Current physical illness
  - d) Social isolation
300. A 60 yr old man had undergone cardiac bypass surgery 2 days back. Now he started forgetting things & was not able to recall names & phone numbers of his relatives. What is the probable diagnosis?
- a) Depression

- b) PTSD
- c) Cognitive dysfunction
- d) Alzheimer's disease

**Postoperative cognitive dysfunction** (POCD) represents a mild form of **surgery induced encephalopathy**. Cases with postoperative encephalopathy of moderate severity are characterized by **acute transient confusion (postoperative delirium) during first days after surgery** and cognitive deficits at delayed follow-up. Severe forms of surgery induced encephalopathy include fatal diffuse brain damage or permanent stroke.

Most studies of postoperative cognitive dysfunction have focused on **cardiac surgery**.



**Post-operative delirium!**

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