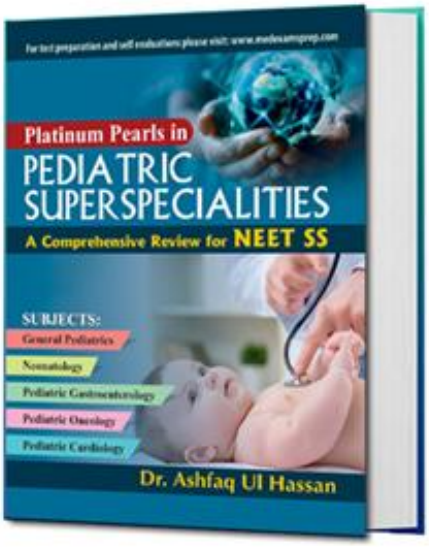
Platinum Pearls in Pediatric Superspecialities: A Comprehensive Review for NEET SS



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NOTES

Q 18. Microdeletions of specific DNA sequences from chromosomes 22q11.2 is found in young patients who also develop tetany. This is because of failure to develop:

A. Pitutary and thymus
B. Pineal and Thymus
C. Heart and thymus
D. Parathyroid and thymus

Ans.: D. Parathyroid and thymus

DiGeorge syndrome, or thymic hypoplasia, results from Microdeletions of specific DNA sequences from chromosome 22q11.2 and defective third and forth pharyngeal pouch development causing hypoplasia or aplasia of the thymus and parathyroid glands. Other structure forming at the same gestational age may also be affected. Therefore, some other features of children with DiGeorage Syndrome may include congenital heart disease, hypertelorism, esophageal atresia, a bifid uvula, and micrognathia. The diagnosis is often made in the newborn period after the patient presents with a hypocalcemic seizure.

Q 19. Persistent Truncus Arteriosus is thought to result from:

A. Failure of the conus arteriosus to develop normally
B. Failure of the ligamentum arteriosum to develop normally
C. Failure of the truncal ridges and aorticopulmonary septum to develop normally
D. Failure of the ductus venosus to develop normally.

Ans.: C. Failure of the truncal ridges and aorticopulmonary septum to develop normally

Persistent Truncus Arteriosus: Persistent TA results from failure of the truncal ridges and aorticopulmonary septum to develop normally and divide the TA into the aorta and pulmonary trunk. In this defect, a single arterial trunk, the TA, arises from the heart and supplies the systemic, pulmonary, and coronary circulations.

Q 20. Ostium Secundum Defect: Females out number males by Ratio

A. 3:1 B. 30:1
C. 300:1 D. 3000:1

Ans.: A. 3:1

This defect, in the region of the fossa ovalis, is the most common form of ASD and is associated with normal atrioventricular valves. The defects may be single or multiple, and in symptomatic older children openings of 2 cm or more in diameter are not unusual. Large defects may extend inferiorly toward the inferior vena cava and ostium of the coronary sinus, superiorly toward the superior vena cava, or posteriorly. Females outnumber males 3:1.

Q 21. "Tet" spells are seen in:

A. ASD B. TGA
C. TOF D. All of Above

Ans.: C. TOF

- Dyspnea occurs on exertion. Characteristically, children assume a squatting position for the relief of dyspnea due to physical effort; the child is usually able to resume physical activity within a few minutes.
- Paroxysmal hypercyanotic attacks (hypoxic, "blue," or "tet" spells) are a particular problem during the first 2 yr of life. The infant becomes hyperpneic and restless, cyanosis increases, gasping respirations ensue, and syncope may follow. The spell occurs most frequently in the morning upon first awakening or following episodes of vigorous crying.

Q 22. L. Cor Triloculare Biatriatum means:

A. 2 chambered heart B. 3 chambered heart
C. 1 chambered heart D. 5 chambered heart

Ans.: B. 3 chambered heart

CHAPTER

5

PEDIATRIC CARDIOLOGY

Q 1. In Pediatric age group, Cardiac involvement of Pulmonary Stenosis and Hypertrophic Cardiomyopathy is commonest in:

A. Laurence-Moon-Bardet- Biedl Syndrome
B. Carpenter Syndrome
C. Frohlich Syndrome
D. Noonan Syndrome

Ans.: D. Noonn Syndrome

CVS features of Noonan Syndrome are:

+ Pulmonary Stenosis
+ Hyperyrophic Cardiomyopathy
+ CHF in Infancy

Q 2. Triangle of Koch is bounded by the following structures, except:

A. Torus aorticus
B. Base of septal cusp of tricuspid valve
C. Margin of coronary sinus opening
D. Tendon of Todaro

Ans.: A. Torus aorticus

Triangle of Koch is bounded by Tricuspid valve, Margin of coronary sinus opening and Tendon of Todaro. It is a part of fibrous skeleton of the heart.

Q 3. In CHARGE Syndrome Cardiac Manifestation is denoted by letter:

A. C B. H
C. E D. G

Ans.: B. H

CHARGE Syndrome is:

+ Coloboma
+ Heart anomalies
+ Atresia (Chonal)
+ Retardation of growth
+ Genitourinary anomalies
+ Ear Anomalies

Because of the fact that newborn infants can breathe through their mouths, the obstruction does not produce the same symptoms in every infant. When unilateral, the infant usually does not have severe symptoms at birth and may be asymptomatic for a long period. Infants with bilateral choanal atresia who have difficulty with mouth breathing will make vigorous attempts to inspire, often suck in their lips, and will develop cyanosis. Persistent mouth breathing and cyanosis when the mouth is closed (which is relieved when the infant cries) are additional manifestations.

acetylcholine receptor on the motor endplates within the synaptic junction. The deficiency of acetylcholine receptor results in episodic muscle weakness and fatigue. Lab investigations reveal Anti – Nicotinic acetyl choline receptor antibodies in these patients. Mediastinal masses especially Q Thymomas are a frequent association.

Q 47. Main mechanism of cell killing by radiation is:

A. Damage to cell membrane
B. Damage to DNA
C. Damage to mitochondria
D. Changes in cellular metabolism

Ans.: B. Damage to DNA

Mechanism of Action of Radiation:

- Penetration of radiation wave into the protoplasm and production of ion-pair (ionization).
- Primary Radio-chemical reaction: Here occurs release of electrons of high kinetic energy. So Free-ions as H and OH are formed.
- Here high energy electrons damage adjacent molecules including DNA via oxygen dependent mechanism.
- The DNA damage is mostly repaired by enzymes in a matter of hours. Certain DNA damages are irreparable and it causes chromosomal abnormalities.

Q 48. Onion peel appearance is seen in:

A. Ewing's sarcoma
B. Osteosarcoma
C. Osteoclastoma
D. Synovial sarcoma

Ans.: A. Ewing's sarcoma

Ewing's sarcoma is believed to arise from endothelial cells in bone marrow.

It appears predominantly in mid diaphysis

Onion peel appearance is seen.

Prognosis is poor and surgery alone is not of much help. Radiotherapy has a dramatic effect on the tumor but overall survival is not much enhanced.

Chemotherapy is much more effective. The best results are seen by combination therapy.

Q 49. Central stellate scar on CT scans are seen in:

A. Renal haemangiomas
B. Renal oncocytomas
C. Wilm's tumors
D. Papillomas

Ans.: B. Renal oncocytomas

Renal cell carcinomas arise from the proximal renal tubular epithelium. Although most are solitary, 7% are multicentric. The histopathologic subtypes of renal cell carcinoma are the clear cell, the granular cell, and the spindle cell or sarcomatoid variant. The clear cell subtype is the most common form (75% of cases), and the less frequent sarcomatoid variety (1 to 6% of cases) is associated with a poorer prognosis.

Oncocytomas are rare variants of renal cell carcinoma thought to arise from the distal tubule. These are well differentiated tumors of low malignant potential. In contrast to the common forms of renal cell carcinoma, 3p deletions are not found in oncocytomas.

Oncocytomas are tubular adenomas with specific histological appearance characterised by the Oncocyte.

They can occur in any age group and vary in size from 1 to 20 cm in diameter.

They are demonstrated in USG as solid masses with internal echoes with stellate hypoechoic centre.

They are demonstrated in CT as solid masses with low attenuation centre scar.

Q 50. Brown Tumors are seen in:

A. Hypothyroidism
B. Hyperparathyroidism
C. Myeloma
D. Eosinophilic granuloma

CHAPTER

4

PEDIATRIC ONCOLOGY

Q 1. In case of Children, Limitless Replicative Potential in case of Tumor Cells is by virtue of:

A. A DNA polymerase
B. Reverse transcriptase
C. Helicase
D. Isomerase

Ans.: A. A DNA polymerase

Telomerase is a DNA polymerase. Limitless Replicative Potential in case of Tumor Cells is By virtue of telomerase. It is not active in normal somatic cells. Cancer cells reactivate Telomerase.

Q 2. Shagreen patches and renal hematomas and involvement of Chromosome 9 with Epilepsy are a feature of:

A. Tuberous Sclerosis
B. NF1
C. NF2
D. Sturge Weber Syndrome

Ans.: A. Tuberous Sclerosis

Tuberous sclerosis is inherited as an autosomal dominant condition and linked with chromosomes 9 and 11. It is characterized by epilepsy, mental retardation, ash leaf macules, adenoma sebaceum, peri ungual fibromas, shagreen patches, and renal hamartomas.

Q 3. Metastatic Potential in case of Tumor Cells is great. In the Body the Metastasis Suppressor Gene is:

A. TWIST
B. TGF Beta Gene
C. SNAIL
D. KA1-1

Ans.: D. KA1-1

Tumor Cells have potential to induce Metastasis and Vascular Proliferation. Certain Genes Oppose these Actions. NM 23 and KA1-1 and miR-226 are important Metastasis suppressor Genes.

Q 4. In a Child Combination of Mental retardation, Ash leaf macules, Adenoma sebaceum, renal hamartomas are a feature of:

A. NF1
B. NF2
C. Sturge Weber Syndrome
D. Tuberous Sclerosis

Ans.: D. Tuberous Sclerosis

Ans.: B. 4:1

Hirschsprung disease is common in males. The male:female ratio of children with Hirschsprung disease (Short Segment disease) is 4:1. It should be suspected in any full-term infant (the disease is unusual in preterm infants) with delayed passage of stool.

Q 22. Proximal half of Duodenum develops from:

A. The foregut
B. The midgut
C. The hindgut
D. Cloaca

Ans.: A. The foregut

- The foregut endoderm gives rise to the esophagus, stomach, proximal half of the duodenum, liver and pancreas.
- The midgut endoderm gives rise to the distal half of the duodenum, jejunum, ileum, cecum, appendix, ascending colon and the right 3/4 of the transverse colon.
- The hindgut endoderm develops into the left 3/4 of the transverse colon, the descending colon, sigmoid colon and the rectum down to the anorectal line (the endoderm-ectoderm junction).

Q 23. Clotting factor not affected in Liver Disease is:

A. Factor II
B. Factor V
C. Factor VII
D. Factor VIII

Ans.: D Factor VIII

Synthesis of fibrinogen, prothrombin, and Factors II, V, IX, X, XI, XII, and XIII occurs in the liver. Synthesis of prothrombin and Factors VII, IX, and X depends on an adequate supply of vitamin K, which activates certain hepatic polypeptides by stimulating the synthesis of the calcium-binding residue, g-carboxyglutamic acid. An abnormal prothrombin time is commonly caused by vitamin K deficiency, liver disease, or both and may rarely be seen with inherited abnormalities. Vitamin K, a fat soluble vitamin that is found in many foods, is also produced by intestinal bacteria

The PT is the most common laboratory test for monitoring oral anticoagulant therapy because it is sensitive to alterations in prothrombin and Factors X and VII, three of the vitamin K–dependent coagulation factors.

Q 24. Rapid administration of glucose in a patient suffering from thiamine deficiency can lead to:

A. Nonketotic hyperosmolar coma
B. Hyperchloremic metabolic alkalosis
C. Lactic acidosis
D. Ketoacidosis

Ans.: A. Nonketotic hyperosmolar Coma

- Thiamine should always be replenished when refeeding a patient with alcoholism, as carbohydrate repletion without adequate thiamine can precipitate acute thiamine deficiency.
- Parenteral thiamine should be given prophylactically to all chronic alcoholic patients in the emergency room, or as soon as they are admitted, to prevent precipitation of thiamine deficiency after the provision of glucose-containing solutions.
- A number of conditions, particularly disorders related to acute and chronic alcoholism, can mimic the clinical features of hepatic encephalopathy.
- These include acute alcohol intoxication, sedative overdose, delirium tremens, Wernicke's encephalopathy, and Korsakoff's psychosi.

Q 25. Which of the following lesions is rare in liver?

A. Portal vein obstruction
B. Microsteatosis
C. Liver infarct
D. Portal tract fibrosis

CHAPTER

3

PEDIATRIC GASTROENTEROLOGY

Q 1. Currarino Triad in neonates includes all except:

A. Anorectal malformations
B. Sacral anomalies
C. Presacral anomalies
D. Esophageal duplications

Ans.: D. Esophageal duplications

Currarino Triad in neonates includes:

- Anorectal malformations (Anal stenosis, atresia).
- Sacral anomalies (Hypolasia).
- Presacral anomalies (anterior meningoceles, cysts, teratoma).

Q 2. Not a Member of Vitamin B Complex group is:

A. Thiamine
B. Tocopherol
C. Niacin
D. Pantothenic acid

Ans.: B. Tocopherol

Various Members of Vitamin B complex group are:

- Thiamine (Vitamin B1).
- Riboflavin (Vitamin B2).
- Niacin (Vitamin B3) (Nicotinic acid or Nicotinamide).
- Pantothenic acid (Vitamin B5).
- Pyridoxine (Vitamin B6) (Pyridoxal or Pyridoxamine).
- Cobalamine (Vitamin B12) Biotin.
- Folic acid (Pteroylglutamic acid).

Q 3. The Pancreas develops in embryonic life:

A. From the hindgut in the 4th week
B. From the neural crest in the 4th week
C. From the midgut
D. From the Ectodermal bud

Ans.: C. From the midgut

Pancreas: The pancreas develops between the layers of the mesentery from dorsal and ventral pancreatic buds of endodermal cells, which arise from the caudal or dorsal part of the foregut. Most of the pancreas is derived from the dorsal pancreatic bud. The larger dorsal pancreatic bud appears first and develops a slight distance cranial to the ventral bud.

Q 66. A survey was conducted for Abdominal Masses in neonates. Which one of the following is the most common cause of abdominal mass in neonates?

A. Neuroblastoma
B. Wilms' tumour
C. Multicystic dysplastic kidneys
D. Renal Cell Cancer

Ans.: C. Multicystic dysplastic kidneys

It has been found that Multicystic dysplastic kidneys are the most common cause of abdominal mass in neonates. All other causes mentioned are not as common as this entity.

Q 67. A Neonate has apnea cyanosis and feeding problems related to the macroglossia. He also develops seizures and hypoglycemia. Most likely cause is:

A. Amyloidosis
B. Beckwith–Wiedemann syndrome
C. Sturge Weber Syndrome
D. Tuberous Sclerosis

Ans.: B. Beckwith–Wiedemann syndrome

Beckwith–Wiedemann syndrome in neonates presents with apnea and cyanosis and in severe cases with feeding problems related to the macroglossia. Neonates may also have seizures and hypoglycemia. Patients are at higher risk for Neonatal polycythemia and Wilms Tumor.

Q 68. In Blooms Syndrome, the defect is seen in:

A. Recombination Repair B. MicroRNA
C. Helicase D. BRCA 2 gene

Ans.: C. Helicase

Helicase is normally involved in DNA Repair by Homologous Recombination. Mutated Helicase cannot perform this Function leading to Defects in DNA. The Patients with Blooms Syndrome have a high propensity to Develop Cancers as a result.

Q 69. A neonate is extremely pale. He has PBF and Lab Values showing Anemia of Type Macrocytic anemia. Bone marrow aspiration shows Reduced Red Cell Precursors. Serum Erythropoietin levels are Elevated. Most Likely cause is:

A. Alpha Thalessmia
B. Myelofibrosis
C. Diamond Blackfan syndrome
D. Siderroblastic Anemia

Ans.: C. Diamond Blackfan syndrome

Pure red cell Aplasia is also Known as Diamond Blackfan syndrome. It may have a familial or genetic basis.It may present as severe pallor right from birth or may manifest slowly in few months. Typically the Bone marrow aspiration shows reduced Red cell precursors. Serum and urine erythropoietin levels are invariably elevated.

Q 70. All of the following are features of Down's syndrome, except:

A. Commonest Monosomy
B. Absent nasal bone
C. Increased free beta HCG levels
D. Associated with Development of Leukemia

Ans.: A. Commonest Monosomy

Down's syndrome is a Common Trisomy NOT Monosomy.

- Features are varied and one should try to remember as many as possible features.
- Head circumference is small brachycephalic skull. The neck is short and thick. Hypotonicity is present. Brushfield's spots (whitish spots scattered round the periphery of the iris).Nuchal fold thickeness. The tongue appears large and may protrude because the mouth is relatively small.

CHAPTER

2

NEONATOLOGY

Q 1. Translocations involving PAX5 are a feature of neonatal:

A. Retinoblastoma
B. Thymoma
C. Lymphoma
D. All of Above

Ans.: C. Lymphoma

- Translocations involving PAX3 and PAX7 are seen in alveolar rhabdomyosarcomas.
- Translocations involving PAX5 are seen in subsets of lymphomas.
- Translocations involving PAX8 are seen in thyroid cancers.
- FMR gene: It is involved in fragile X- Syndrome
- PTEN gene: Located to chromosome 10q is associated with endometrial cancers and glioblastoma. (Phosphatase and tensin homologue).

Q 2. Tetraploidy is a condition whereby cells contain:

A. 36 Chromosomes
B. 92 Chromosomes
C. 12 Chromosomes
D. 48 Chromosomes

Ans.: B. 92 Chromosomes

Tetraploidy is a rare condition. Here cells contain 92 chromosomes. It results in spontaneous abortion of the conceptus. Tetraploidy occurs as a result of failure of the first cleavage division.

Q 3. In Classical congenital Galactosemia the deficient enzyme is:

A. Glucokinase
B. Galactokinase
C. Isomerase
D. Uridyl transferase

Ans.: D. Uridyl transferase

Classical galactosemia is due to deficiency of uridyltransferase.

Three enzyme deficiencies implicated:

- GPUT Deficiency (Galactose 1 Po_4 uridyl transferase) most common.
- UDP galactose 4 epimerase deficiency.
- Galactokinase deficiency.

Q 4. In Humans Mutations of HOX D 13 genes was detected. From a Genetic Point this results predominantly in defects of:

A. Lung Development
B. Liver Development
C. Digits of Limb Development
D. Heart Development

Ans.: C. Digits of Limb Development

regulation of glomerular permeability, and acts as a linker between the plasma membrane and the cytoskeleton. Defects in this gene are the cause of autosomal recessive steroid-resistant nephrotic syndrome (SRN).

Q 91. The Trait in which the parents are clinically normal and only siblings are affected but males and females are affected in equal proportions is:

A. AD
B. AR
C. XLD
D. XLR

Ans.: B. AR

Autosomal recessive Traits: Autosomal recessive conditions are clinically apparent only in the homozygous state, i.e., when both alleles at a particular genetic locus are mutant alleles. The following features are characteristic:

- The parents are clinically normal;
- only siblings are affected;
- males and females are affected in equal proportions;
- if an affected individual marries a homozygous normal person, none of the children is affected but all are heterozygous carriers;
- if an affected individual marries a heterozygous carrier, one half of the children are affected, and the pedigree pattern superficially suggests a dominant trait;
- if two individuals who are homozygous for the same mutant gene marry, all of their children are affected;
- if both parents are heterozygous at the same genetic locus, one fourth of their children are homozygous affected, on average one fourth are homozygous normal, and one half are heterozygous carriers of the same mutant gene; and
- the less frequent the mutant gene is in the population, the greater the likelihood that the affected individual is the product of consanguineous parents.

Q 92. Most common cause of Heridetary Spherocytosis is:

A. Spectrin
B. Glycophyrin
C. Ankyrin
D. B and 4

Ans.: C. Ankyrin

Hereditary Spherocytosis (HS): Hereditary spherocytosis is an inherited hemolytic anemia characterized by dense, osmotically fragile, partially spherical red cells that are selectively trapped by the spleen. "Ankyrin defects are the most frequent" (= 30 to 60% of cases). These occur in both dominant and recessive HS and are characterized by combined deficiency of ankyrin and spectrin (which binds to ankyrin) in roughly equivalent proportions.

Q 93. Gaucher's disease is:

A. Inherited as AR
B. Inherited as AD
C. Inherited as XLR
D. Inherited as XLD

Ans.: A. Inherited as AR

Gaucher's disease is a "lipid storage disease" resulting from "deficient activity of a lysosomal hydrolase" characterized by "deposition of glucocerebroside" in cells of the macrophage-monocyte system. Inherited is Autosomal recessive disease. Gaucher's disease should be considered in the differential diagnosis of patients with unexplained organomegaly, easy bruisability, and/or bone pain. The pathologic hallmark is the presence of the "Gaucher cell" in the macrophage-monocyte system, particularly in the bone marrow. These cells, which are 20 to 100 mm in diameter, have a characteristic "wrinkled-paper" appearance resulting from intracytoplasmic substrate deposition. These cells stain "strongly positive with periodic acid–Schiff", and their presence in bone marrow and/or other tissues suggests the diagnosis

CHAPTER

1

GENERAL PEDIATRICS

Q 1. A young Child has Blue Sclera with Retinal Detachment and Corneal Rupture. Orthopedic Exam Revealed Kyphoscoliosis. Most likely he is having:

A. Marfans syndrome
B. Fragile X syndrome
C. Ehler Danhlos syndrome
D. Down syndrome

Ans.: C. Ehler Danhlos syndrome

Features of Ehler Danhlos Syndrome Type VI are Blue Sclera with Retinal Detachment and Corneal Rupture. Children have Detectable Mutations in genes Controlling Collagen Synthesis.

Q 2. In A Child with Beta-thalassemia major what is seen:

A. A defect in the alpha subunit of hemoglobin
B. Precipitation of beta chains
C. Hemolytic red blood cell destruction
D. Effective erythropoiesis

Ans.: C. Hemolytic red blood cell destruction

Beta Thalassmia is characterized by:

+ A defect in the beta subunit of hemoglobin
+ Precipitation of alpha chains
+ Hemolytic red blood cell destruction
+ Ineffective erythropoiesis
+ Hyper plastic bone marrow, liver and spleen.

Q 3. The Classic Clinical Condition occurring as a result of deletion of regions on paternal Chromosome 15 is:

A. Sjogren's syndrome B. Marfans Syndrome
C. Prader Willi Syndrome D. Leighs Syndrome

Ans.: C. Prader Willi syndrome

Prader Willi Syndrome is a Clinical Condition occurring as a result of deletion of regions on paternal Chromosome 15. It is an Example of Genomic Imprinting Disorder. Loss of functions of SNORP Gene is a feature.

Q 4. A 5 year old male has a genetic defect of combination of Diabetes Insipidus, Exophthalmos and Bony Skull defects. Most Likely cause is:

A. Jobs Syndrome

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Platinum Pearls in

PEDIATRIC SUPERSPECIALITIES

A Comprehensive Review for NEET SS

Dr Ashfaq Ul Hassan

Neomed Education Private Limited