1. **A 3.5 kg male infant born at term after an uncomplicated pregnancy and delivery develops respiratory distress shortly after birth and requires mechanical ventilation. The chest radiograph reveals a normal cardiothymic silhouette but a diffuse ground glass appearance to the lung fields. Surfactant replacement fails to improve gas exchange. Over the first week life, the hypoxemia worsens. Results of routine culture and echocardiographic findings are negative. A term female sibling died at 1 mo of age with "respiratory distress". Which of the following is the most likely diagnosis?**

a) Total anomalous pulmonary venous return

b) Meconium aspiration

c) Neonatal pulmonary alveolar proteinosis

d) Disseminated herpes simplex infection

Correct Answer - C
Neonatal pulmonary alveolar proteinosis [Ref: Nelson 18/e p. 18211 "Respiratory distress in an infant along with a positive family history of similarly affected newborn infants strongly suggests, pulmonary

alveolar proteinosis"

- Pulmonary alveolar proteinosis is *a* disorder characterized by the *intraalveolar accumulation of pulmonary surfactant.*
- Two clinically distinct forms of pulmonary alveolar proteinosis are seen *:- Fatal form -Presenting shortly after birth (congenital PAP)*
  - Gradually progressive form Presenting in older infants and children.

Pathology

- *Although the mechanisms that lead to alveolar proteinosis are undefined, histological findings suggests that they result in a disruption of pulmonary surfactant metabolism.*
- *The main surface tension lowering agent in surfactant is phospholipids i.e. primary dipahnityol phosphatidyl choline.*
- *However DPCC needs "surfactant protein" (in endogenous and natural surfactants) for efficient dispersion which enables the formation of a phospholipid monolayer on the alveolar surface.*
- *There are two surfactant proteins present in the body i.e., protein A and protein B.*
- *"In pulmonary alveolar proteinosis, there is absence of protein B".*
  - In the absence of protein B, the rapid spread and absorption of the phospholipid (DPCC) does not take place so they cannot form a phospholipid monolayer on the alveolar surface.
  - This in turn leads to failure of expansion of alveoli leading to poor cardiorespiratoty adaptation at birth.

Clinical manifestation:-

- *This disorder is inzmediately apparent in the newborn period a rapidly leads to respiratory failure.*
- *Congenital pulmonary alveolar proteinosis is clinically and radiographically indistinguishable from more common disorders of the newborn that lead to respiratory failure including pneumonia, generalized bacterial infection, respiratory distress syndrome and total anomalous pulmonary venous return with obstruction.*

Diagnosis

- *Histopathological examination of lung biopsy specimen is the gold standard for diagnosis.*

- *On histopathological examination distal air spaces are filled with a granular, eosinophillic material that stains positively with periodic-acid schiff reagent and is diastase resistant.*

Treatment

- *Untreated, alveolar proteinosis in newborns is rapidly fatal and no successful medical therapy has been developed.*
- *Repeated bronchoalveolar lavage is a temporizing measure.*
- *Lung transplantation is the only therapeutic option but its use is limited by concerns about disease recurrence.*

2. Ductus dependent blood flow is required for all of these congenital heart diseases except

a) Persistent truncus arteriosus

b) Hypoplastic left heart syndrome

c) Pulmonary stenosis

d) TGA with intact ventricular septum

Correct Answer - A
Persistent truncus arteriosus [ReP V. Mohan Reddy. Cardiac Surgery for Premature and Low Birth Weight Neonates, Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery 2003; 4; 271-76]

- Congenital heart disease in the newborn can be broadly categorized by the relationship between the patient's cardiac defect and the patent ductus arteriosus and this categorization yields four distinct groups.
- First are newborns dependent on a patent ductus arteriosus (PDA) for pulmonary blood flow.
- Second are newborns dependent on the PDA for systemic blood flow.
- Third are those dependent on the patent ductus for proper mixing of oxygenated blood
- Fourthly are neonates with a non- ductal dependent circulation.
- For the ductal dependent group, "intravenous prostaglandin" (E-1) is used as necessary to maintain ductal patency and is the single most important step in supporting these patients to diagnosis and definitive therapy.

- Nevertheless, all newborns ductal dependent for systemic or pulmonary blood flow require an intervention, surgical or cardiologic to eliminate ductal dependence prior to discharge.

1. Pulmonary flow ductal dependence

- Newborns with congenital heart disease who are dependent on the patency of their ductus for pubnonary blood .flow present with varying degrees of cyanosis.
- Critical Pulmonary Valve Stenosis with Intact Ventricular Septum
- Tricuspid AtresiaQ
- Tetralogy of Fallote

2. Systemic flow ductal dependence

- These newborns are dependent on their ducats arteriosus for systemic blood flow and unlike the babies who are pulmonary flow dependent, these newborns present with severely decreased cardiac output.
- This decreased systemic flow is characterized by pallor, diminished peripheral pulses, low urine output, cool extremities and varying degrees of metabolic acidosis.
- Included are newborns born with left ventricular outflow tract obstruction at various levels.
  Congenital Valvular Aortic Stenosis
  Coarctation of the Aorta
- Interruptued Aortic Arch
  Hypoplastic Left Heart Syndrome (HLHS)

3. Oxygenation ductal dependence

- These are babies who are born with congenital heart disease that requires a patent ductus for adequate mixing of saturated and desaturated blood.
- Transposition of great arteries

4. Ductal independence

- These are neonates who are not dependent on a patent ductus but still require urgent operation
- Total anomalous pulmonary venous return (TAPVR)
- Truncus Arteriosus
- Anomalous origin of the left main coronary artery from the pulmonary artery.

3. A 3 week old patient presenting with vomiting and failure to thrive is found to have pyloric stenosis. What should be the next step of management?

a) its emergency so do pyloromyotomy immediately

b) fluid resuscitation may be delayed

c) correction of electrolyte disturbances

d) Cardiopulmonary resuscitation

Correct Answer - C
Correction of electrolyte disturbances *[Ref. Schwartz 9/e p1425 (8/e, p1486; 7/e, p1728); Rudolph's Pediatrics, 21st Edition Chapter 17; Bailey & love 25/e p78 (24/e p1033; 23/e, p 899)]*
Pyloric stenosis is *never a surgical emergency* although dehydration and electrolyte abnormalities may present a medical emergency
Fluid resuscitation and correction of electrolyte abnormalities and metabolic alkalosis is essential before surgery. Surgery done is *Fredet-Ranzstedt pyloromyotomy. (In it the pyloric mass is split without cutting the mucosa)*
More about Hypertrophic Pyloric Stenosis (HPS)

- The musculature of the pylorus and adjacent antrum is grossly hypertrophied, the hypertrophy being maximum in the pylorus itself.
- It occurs in approximately 3 in 1000 live birth and is the *most common surgical cause of vomitting in infancy.*
- M > F (4 I)*
- Characterstically the first born male child is affected.
- The conditon is *most commonly seen at 4 wks after birth* ranging from the 3rd wk to on rare occasion, the 7th. However, 20% of infants

are symptomatic from birth, and most are symptomatic within the first 2 months after birth.

- Non-bilious vomiting, becoming increasingly projectile, occurs over several days to weeks. Eventually the infant will develop a nearly complete obstruction by the second to fourth week of life and will not be able to hold down even clear liquids. This invariaby proceeds to severe dehydration if not tlt.
- These infants develop a metabolic alkalosis with severe depletion of potassium and chloride ions.
- *" The diagnosis of pyloric stenosis usually can he made on physical examination by palpation of the typical 'olive' in the right upper quadrant and the presence of visible gastric waves on the abdomen.*
- *When the olive cannot be palpated, ultrasound in experienced hands will diagnose the condition accurately in 95 percent of pts." - Schwartz.*
- Also know
- *Administration of erythromycin in early infancy has been linked to the subsequent development of HPS.*
- *Jaundice may be seen in HPS, although cause is not clear.*

4. A 32 wk new born baby with RR-86/min, grunting present with no nasal flaring, abdomen behind in movement than chest, minimum interocostals retraction & no xiphisternal retraction. What is the Silverman scoring -

a) 1

b) 4

c) 3

d) 6

Correct Answer - B

4 *[Ref: Maternity and Pediatric Nursing By Susan Scott Ricci, Terri Kyle, P. 729-30;Silverman WC, Anderson DR. Controlled clinical trial on effects* of *water mist on obsrtructive respiratory signs, death rate and necropsy findings among premature infants. Pediatrics 1956;17:1-4; OP Ghai. 7/e, p 143]*

- *Silverman- Anderson Score is used to assess the degree of respiratory distress in newborn (usually with Respiratory Distress Syndrome).*

-The index involves observation of five features, each of which is scored as 0, 1 or 2.

- The higher the score, the greater the respiratory distress.

- A score over 7 sueeests severe res[p]irator[y] distress.

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Silverman — Anderson Index

Upper Chest Lower Xiphoid

Score	Retraction	Chest Retraction	Retraction	Nares flaring	Grunt
0	Chest and abdomen rise together	Nostals Interco retractions	No Xiphoid retractions	No nasal Flaring	No expiratory Bunt
1.	Lag or minimal sinking of upper chest as abdomen rises	Minimal intercostals retractions	Minimal xiphoid retractions	Minimal nasal fairing	Expiratory grunt heard with stethoscope
7	Upper chest and abdomen move as a see-saw"	Marked intercostals retractions	Marked xiphoid retractions	Marked nasal flaring	Audible expiratory grunt

Now look at the values in the question

• Upper chest	Lag on inspiration present	—>	Score 1
• Lower chest	—> Slight retraction present		Score 1
• Xiphoid retraction	Not present	--->	Score 0
• Nares flaring	—> Not present	—>	Score 0
• Grunting	-4 Audible grunting		Score 2

Total = Score 4

- *The Downes' score is more comprehensive and can be applied to any gestational age and condition*

Score	Respiratory rate	Cyanosis	Air entry	Grunt	Retraction
0	< 60/min	Nil	Normal	None	Nil

i	60-80	In room air	Mild	Auscult with stethoscope	Mild
2	>80/min	In 40% 0,	Marked	Audible with naked air	Moderate

5. Most common cause of Neonatal sepsis in hospital in India is

a) Escherichia coli

b) Klebsiella

c) Staph aureus

d) Listeria monocytogen

Correct Answer - B

Ans: B. Klebsiella [Ref Ghai 6th/e p 161, Meherbaan Singh OW p 209 (Repeat from AIIMS Nov 09)]

6. A term neonate with unconjugated hyperbilirubinemia of 18 mg/dl on 20th day. All are common causes except :

a) Breast milk jaundice

b) Congenital cholangiopathy

c) G 6PD deficiency

d) Cotrimoxazole

Correct Answer - B

Congenital cholangiopathy [*Ref Ghai G^h/e p 171-174 Nelson 17^th/e p 594-595] "Congenital cholangiopathv will cause conjugated hyperbilirubinemia"*

Physiological jaundice

- Most neonates develop visible jaundice due to elevation of *unconjugated bilirubin* concentration during their first week
- This common condition is called physiological jaundice.
- This pattern of hyperbilirubinemia has been classified into two functionally distinct periods.

Phase one

- Last for *five days in term infants* and about *seven days* in *pretenn infants,* when there is a rapid rise is serum bilirubin levels to 12 and 15 mg/dl respectively.

Phase two

- There is decline to about 2 mg/dl, which lasts for two weeks after which adult values are attained. Breastfeeding jaundice and Breastmilk jaundice
- There is *strong association* between *exclusive breastfeeding* and neonatal jaundice.

- A few babies who remain on exclusive breast feed develop jaundice in the *second week of life* and continue well into the *third month.* This is called *breastmilk jaundice.*
- A bilirubin level of over 20 mg/dl may be attained. *(It is presumed to be due to inhibitory substances in the breastmilk that interfere with bilirubin conjugation e.g. pregananediol and free fatty acids).*
- *Temporary interruption* of breastmilk feeds will *dramatically reduce* the serum levels of bilirubin and there may be slight increase in bilirubin when breast feeding is resumed, but it never reaches the previous levels. Hypothyroidism
- *Persistent elevation* of indirect bilirubin is the *first sign* of congenital hypothyroidism in neonates.
- This is due to *decreased activity of UDPGT* for weeks or months after birth.
- At least 10% of all infants with congenital hypothyroidism will have jaundice as the presenting symptom.
- Treatment with thyroxine promptly *alleviates* jaundice.

Cotrimoxazole taken in 3rd trimester may cause significant increase in the serum unconjugated bilirubin level.

Differential diagnosis of Breast milk jaundice

Differential diagnosis of unconjugated hyperbilirubinemia :

Increased production

- Fetomaternal blood group incompatibility Rh, ABO.
- Hereditary spherocytosis
- *Non spherocytic hemolytic anemia,* (G6PD) deficiency, pyruvate kinase deficiency, alpha thalassemia, vitamin K induced hemolysis.
- Sepsis
- Increased enterohepatic circulation, pyloric stenosis or large bowel obstruction.

Decreased clearance

- Inborn errors of metabolism. Criggler Najar Syndrome type I & II.
- Drugs and hormones, hypothyroidism, Breast milk jaundice.

7.

Which among the following is an important fatty acid present in breast milk essential for growth?

a) Docosahexaenoic acid

b) Palmitic acid

c) Linoleic acid

d) Linolenic acid

Correct Answer - A

Docosahexaenoic acid and eicosapentaenoic acid are omega 3 fatty acids which are important constituents of the communication membranes of the brain, and they are necessary for normal brain development. They are also active in the retina of the eye.

Docosahexaenoic acid is formed from linolenic acid.
Linolenic acid is an essential fatty acid present in the breast milk.
It is the starting material for the synthesis of eicosapentaenoic acid and Docosahexaenoic acid.

Biochemically important omega 3 and 6 fatty acids:

Omega 3 acid	**Omega 6 acid**
Linolenic acid	Linoleic acid
Eicosapentaenoic acid	Arachidonic acid
Docosahexaenoic acid	

Ref: Organic and Biological Chemistry By H. Stephen Stoker page 301

8. **Correct about respiratory distress syndrome is:**

a) Seen after 6 hours of birth

b) Over production of surfactant.

c) More commonly seen in term neonates

d) Air bronchograms seen on X-ray chest

Correct Answer - D

Respiratory distress usually occurs within the first 6 hours of life. It is mainly seen in preterm neonates.
Premature infants with respiratory distress syndrome present with progressively worsening retractions, tachypnea, and oxygen requirements because their lungs are too immature to synthesize surfactant.
This disease has a characteristic radiographic pattern that includes "ground glass" opacities in the lung parenchyma and prominent air bronchograms.

Initial therapy for respiratory distress syndrome involves using CPAP to stent open the airways, thereby reducing the collapse of the alveoli and limiting further damage.

Term infants can have respiratory complications, although these are much less frequent than in those born preterm.
Common causes in term infants include sepsis, especially from group B streptococcal disease and intrauterine-acquired pneumonia; persistent pulmonary hypertension of the newborn; meconium aspiration syndrome; and pulmonary hemorrhage.

Ref: Wood K.S., Gordon P.V. (2011). Chapter 4. Neonatal and Pediatric Transport. In J.E. Tintinalli, J.S. Stapczynski, D.M. Cline, O.J. Ma, R.K. Cydulka, G.D. Meckler (Eds), Tintinalli's Emergency Medicine: A Comprehensive Study Guide, 7e.

9. Which of the following is not a feature of Juvenile Idiopathic Arthritis?

a) Rheumatoid nodules

b) Spikes of high fever

c) Uveitis

d) Raynaud's phenomenon

Correct Answer - D

Raynaud's Phenomenon is not mentioned in association with Juvenile Idiopathic Arthritis (JIA).

Ref: Current Diagnosis and Treatment in Rheumatology, 2nd Edition, Pages 196-197; Nelson’s Textbook of Pediatrics, 18th Edition, Page 1003; Primer on The Rheumatic Diseases By John H. Klippel, Page 145

10. **A newborn baby presented with profuse bleeding from the umbilical stump after birth. Other physical examination are normal. PT and APTT are within normal limits. What is the most probable diagnosis you would consider?**

a) Factor X deficiency

b) Glanzmann thrombasthenia

c) Von Willebrand disease

d) Bernard Soulier syndrome

Correct Answer - A

Bleeding from umbilical stump is an early manifestation of factor X deficiency. Congenital factor X deficiency is an autosomal recessive disease. Homozygous patients shows markedly prolonged PT and APTT, and heterozygous individuals show mildly prolonged or normal PT and APTT. Since this child is presenting with bleeding from umbilical stump with normal PT and PTT, most likely diagnosis is Factor X deficiency.

- **In Von Willibrand disease:** PT is normal, PTT is increased.
- **In factor XIII deficiency:** PT and PTT are normal, minimally prolonged thrombin time, increased fibrin degradation products. Diagnosis is established by increased clot solubility in 5M urea, dilute monochloroactic acid or acetic acid.
- **In Bernard soulier sydrome:** abnormally large platelets, moderate thrombocytopenia and prolonged bleeding time. Platelet aggregation studies shows defective response to ristocetin and normal response to other agonists. Addition of normal platelets corrects abnormal aggregation.
- **In Glanzmann's thrombasthenia:** Platelet aggregation studies shows marked impairment of aggregation in response to stimulation with typical agonists.

11. **A previously healthy 6 week old female infant, is found unresponsive in on the bed. In the emergency department, she is noted to be well developed and well nourished with normal blood pressure and appearance of the genitalia but with increased pigmentation of her skin. Her blood glucose level is 30 mg/dL. What is the most likely diagnosis?**

a) CAH due to 21 alpha hydroxylase deficiency

b) Familial glucocorticoid deficiency

c) Insulinoma

d) Cushing syndrome

Correct Answer - B

Infant is showing features of **familial glucocorticoid deficiency** a rare autosomal recessive disorder characterized by primary adrenal insufficiency without mineralocorticoid deficiency. In this condition there is adrenocortical resistance to ACTH.
Patients presents during early infancy with hypoglycemia, hyperpigmentation, low plasma cortisol and failure to respond to ACTH stimulation with increased steroidogenesis. Zona fasciculate and reticularis in these patients are reduced to a narrow band of fibrous tissue whereas the zona glomerulosa is well preserved.

Patients are treated with replacement doses of hydrocortisone or equivalent doses of other glucocorticoids. They do not require mineralocorticoid supplementation as their zona glomerulosa is normal.

12. A 5-yr-old boy presents with pubic hair development. He is tall and has increased pigmentation of his genitalia and phallic enlargement. Blood pressure is 130/90 mm Hg. Measurement of which of the following hormones would be most likely to be diagnostic?

a) Testosterone

b) 17-Hydroxyprogesterone

c) Increase aldosterone

d) Increase 11 deoxycortisol

Correct Answer - D
This male has a non-salt-losing form of congenital adrenal hyperplasia due to 11-hydroxylase deficiencies. The metabolic defect produces salt-retaining steroids (deoxycorticosteroids) and is thus associated with hypertension and usually presents later in life than is typical for salt-wasting forms of 21-hydroxylase deficiencies. (See Chapter 570in Nelson Textbook of Pediatrics, 17thed.)

13. **Bacterial meningitis in children (2 months-12 years of age) is usually due to the following organisms except:**

a) Streptococcus pneumoniae

b) Neisseria meningitidis

c) Hemophilus influenzae type B

d) Listera monocytogenes

Correct Answer - D

Listeria monocytogenes cause meningitis < 2months and > 55yrs.

Age	Causative agent
	Listeria monocytogenes
2-3 months	Pneumococcus
3 months- 2 years	**World:** Pneumococcus **India:** H. influenzae
2-12 years	**World:** Pneumococcus India: N. meningitidis
>12 years	Pneumococcus

In infants who have not received routine immunizations, common causes of bacterial meningitis include-

S. pneumoniae (many serotypes; particularly in infants with no record of S. pneumoniae conjugate vaccination)

Neisseria meningitidis (especially serogroup B, but occasionally groups A, C, Y, or W135)

H. influenzae type b (particularly in infants with no record of H. influenzae type b conjugate vaccination)

Ref: Text Book of Pediatrics By Nelson, 17th Edition, Page 2038

14.

Which of the following infection aquired during pregnancy result in hypoplasia of limbs and scarring in the fetus?

a) Varicella

b) Herpes simplex

c) Rubella

d) Toxoplasma

Correct Answer - A

Women who acquire VZV infection before 20 weeks of gestation have 2% risk of delivering an infant with fetal varicella syndrome. It is unlikely in women who have chickenpox during the last half of pregnancy.

Clinical manifestations of congenital varicella syndrome includes:

- IUGR
- Growth retardation
- Limb hypoplasia
- Chorioretinitis
- Cataract
- Microphthalmia
- Cutaneous scarring
- Neurological abnormalities (microcephaly, hydrocephalus, horners syndrome, cranial neuropathies)

15. **Which is the best screening test to evaluate hearing in a neonate?**

a) Pure Tone Audiometry

b) Stapedial Reflex

c) Otoacoustic Emissions

d) Brainstem evoked auditory response

Correct Answer - C
Otoacoustic emissions (OAEs) are objective, noninvasive, and rapid measures (typically less than 2 minutes) used to determine cochlear outer hair cell function.
OAE testing is commonly used in newborn hearing screening because of its speed and noninvasive nature.

It is also used in confirming pure-tone test results obtained from young children, in patients for whom a functional hearing loss is suspected, for audiometric configuration confirmation, for ototoxic drug monitoring, and in hearing aid candidacy.

More recently, OAEs, in conjunction with ABR, can be used in identifying individuals with auditory neuropathy, also termed auditory dyssynchrony.

Ref: Sweetow R.W., Sabes J.H. (2012). Chapter 45. Audiologic Testing. In A.K. Lalwani (Ed), CURRENT Diagnosis & Treatment in Otolaryngology—Head & Neck Surgery, 3e.

16. **A child presents with seborrheic dermatitis, lytic skull lesions, ear discharge and hepatosplenomegaly; likely diagnosis:**

a) Leukemia

b) Lymphoma

c) Histiocytosis X

d) Multiple myeloma

Correct Answer - C
LCH provokes a non-specific inflammatory response, which includes fever, lethargy, and weight loss. Organ involvement can also cause more specific symptoms.
Bone: The most-frequently seen symptom in both unifocal and multifocal disease is painful bone swelling. The skull is most frequently affected, followed by the long bones of the upper extremities and flat bones. Infiltration in hands and feet is unusual. Osteolytic lesions can lead to pathological fractures.
Skin: Commonly seen are a rash which varies from scaly erythematous lesions to red papules pronounced in intertriginous areas. Up to 80% of LCH patients have extensive eruptions on the scalp.
Bone marrow: Pancytopenia with superadded infection usually implies a poor prognosis. Anemia can be due to a number of factors and does not necessarily imply bone marrow infiltration.
Lymph node: Enlargement of the liver in 20%, spleen in 30% and lymph nodes in 50% of histiocytosis cases.
Endocrine glands: Hypothalamic pituitary axis commonly involved. Diabetes insipidus is most common. Anterior pituitary hormone deficiency is usually permanent.
Lungs: Some patients are asymptomatic, diagnosed incidentally because of lung nodules on radiographs; others suffer from chronic

cough and shortness of breath.
Less frequently gastrointestinal tract and central nervous system.

Ref: Kliegman, Behrman, Jenson, Stanton (2008), Chapter 507, “Histiocytosis Syndrome of Childhood”, In the book, “NELSON TEXTBOOK OF PEDIATRICS”, Volume 2, 18th Edition, New Delhi, Page 2159

17. **A 2 month old infant is presented with failure to thrive, recurrent emesis, hepatosplenomegaly, and adrenal insufficiency. Adrenal calcification is noted radiologically. What is the most likely diagnosis?**

a) Adrenal hemorrhage

b) Wolman's disease

c) Pheochromocytoma

d) Addison's disease

Correct Answer - B

This child is showing features of Wolman disease.

Wolman disease appear in the first few weeks of life and presents with persistent vomiting and diarrhea, hepatosplenomegaly, xanthomatosis and adrenal calcification. Patients have a complete absence of enzyme A of lysosome acid lipase.

Investigations: shows liver enzyme abnormalities, decreased adrenal responsiveness to ACTH stimulation, normal or decreased plasma lipids. Cholesterol ester and triglyceride deposition occur in the lysosomes of liver parenchymal and Kuppfer cells, and in macrophages of adrenal gland, lymph node, intestinal mucosa etc.

18. A 3 year old boy with normal developmental milestones with delayed speech and difficulty in communication and concentration. He is not making friends. Most probable diagnosis is *?*

a) Autism

b) ADHD

c) Mental retardation

d) Specific learning disability

Correct Answer - A
Ans. is 'A' i.e., Autism
Delayed speech, difficulty in communication and concentration in a 3 year old child suggests the diagnosis of autism.
Autism is characterized by impaired social interaction and communication, and by restricted and repetitive behavior. These signs all begin before a child is three years old.
Autism affects information processing in the brain by altering how nerve cells and their synapses connect and organize
It is one of three recognized disorders in the autism spectrum, the other two being Asperger syndrome, which lacks delays in cognitive development and language, and pervasive developmental Disorder-not otherwise specified (commonly abbreviated as PDD-NOS)

19. Which of the following is not true about newborn assessment -

a) APGAR at 7 min indicates neonatal mortality depression

b) APGAR at 1 min, indicators for neonatal resuscitation

c) Fetus can rapidly washout CO_2 through placenta

d) Anaerobic metabolism causes acidemia

Correct Answer - A

Ans is 'a' i.e., APGAR at 7 min indicates neonatal mortality depression

- Later times APGAR score (after 5 minutes) indicates about long term neurological damage (not neonatal mortality)

Interpretation of APGAR Score

- *The test is generally done at one and five minutes after birth,* and may be repeated later if the score is and remains low. Scores 3 and below are generally regarded as critically low, 4 to 6 fairly low, and 7 to I 0 generally normal.

* *A low score on the one-minute test may show that the neonate requires medical attention (e.g. resuscitation)* but is not necessarily an indication that there will be long-term problems, particularly if there is an improvement by the stage of the five-minute test. *If the Apgar score remains below 3 at later times such as 10, 15 or 30 mintues, there is a risk that the child will suffer longer-term neurological damage.* There is also a small but significant increase of the risk of cerebral palsy. However, the purpose of the Apgar test is to determine quickly whether a newborn needs immediate medical care; it was *not* designed to make long-term predicitions on a child's health.

CO_2 transport across placenta

* CO_2 is cleared by placenta by simple diffusion. CO_2 is produced abundantly in the fetus, and the PCO2 of fetal blood is higher than maternal blood. CO_2 therefore diffuses from fetal blood, through the placenta, into the maternal circulation, and is disposed by expiration from mother's lung.

Anaerobic metabolism causes acidemia due to lactate (lactic acid) production

* Anoxic perfusion causes an increase in glucose consumption which is more than two fold higher than that seen in the oxygenated perfusion, resulting finally in placental uptake of glucose not only from the maternal but also from the fetal circulation.

* *Lactate production is increased during the anoxic perfusion,* while the final tissue energy value lies between the values observed for fresh tissue and for the oxygenated perfusion. The shift to anerobic metabolism shown by placental tissue in anoxic conditions enables continued functioning of the tissue over the 2-h perfusion period but it appear that under anoxic conditions the tissue may incur an energy debt not observed in oxygenated perfusions.

20. A newborn with respiratory distress with RR 86/ min, nasal flaring, audible grunting, abdomen lagged behind chest respiratory movement, no lower chest or xiphoid retraction. What is silverman's score *?*

a) 1

b) 3

c) 5

d) 6

Correct Answer - C

Ans. is `c' i.e., 5

Assessment of respiratory depression

The severity of respiratory distress is assessed by Silverman-Anderson score and Downes' score. While the Silverman Anderson Retraction Score is more suited for preterms with HMD, the Downes' score is more comprehensive and can be applied to any gestational age and condition.

Silverman Anderson retraction score

Score	Upper chest retraction	Low chest retraction	Xiphoid retraction	Nasal flaring	Grunt
	Synchronized	None	None	None	None
1	Lag on inspiration	Just visible	Just visible	Minimal	Stethoscope only
2	See- saw	Marked	Marked	Marked	Naked ear

A score of >6 is indicative of impending respiratory failure o

Now analyzing our question data :-

Upper chest - Lag on inspiration present → score 1
Lower chest - No retraction → score 0
Xiphoid - No retraction → score 0
Nasal flaring - Present → score 2
Grunting - Present → score 2 → So, total score is 5.

Downers's score

Score	Respiratory rate	Cyanosis	Air entry	Grunt	Retraction
0	< 60/min	Nil	Normal	None	Nil
1	60-80/train	In room air	Mild?	Ausc with stethoscope	Mild
2	>80/ min	En >40% 0_2	Marked?	Audible with naked ear	Moderate

A score of > 6 is indicative of impending respiratory failure.

21. A 5-years old male child presents with episodic anaemia and jaundice since birth. He is least likely to have which of the following

a) Hereditary spherocytosis

b) Sickle cell anemia

c) PNH

d) G-6-PD deficiency

Correct Answer - C
Ans is `c' i.e., PNH
* Causes of Jaundice since birth are:
(i) Rh incompatibility (erythroblastosis fetalis)
(ii) ABO incompatibility
(iii) Congenital infections (TORCH)
(iv) Sepsis
(v) Concealed hemorrhage
(vi) Red cell membrane defect (hereditary spherocytosis)
(vii) Red cell enzyme defect (G6PD deficiency)

- So, option a & d can cause jaundice since birth.

* In sickle cell anemia, affected infants do not develop symptoms in the first few months of life because the hemoglobin produced by the developing fetus (fetal hemoglobin) protects the red blood cells from sickling. This fetal hemoglobin disappears after 5 month of age so that by 5 months of age, the sickling of the red blood cells is prominent and symptoms begin.
* PNH is manfested in adults.

- So, both PNH and sickle cell anemia does not cause jaundice since

birth.

* But among these two I would prefer PNH as the answer because it is manifested in adulthood while the patient in question is a 5-years old child.

- Sickle cell anemia symptoms develop at the age of 5 months and it is one of the cause of jaundice (en.wikipedia.org).

22. A 50-hour old full-term breast-fed newborn boy weighing 3100g presents with clinically evident jaundice. Physical examination is otherwise normal. The total bilirubin is 8.0 mg/dl with a direct bilirubin of 0.4 mg/dl. What would be the correct treatment -

a) Continue breast feeds and review after 48 hours

b) Stop breast feeds and review after 24 hours

c) Continue breast feeds and start blue-light phototherapy

d) Arrange for a double-volume exchange transfusion

Correct Answer - C
Ans. is 'c' i.e., Continue breast feeds and start blue-light phototherapy
o This is a case of breast milk jaundice.
Preferred treatment option for a 4 days old baby with bilirubin of 8 mg/dL is to continue breastfeeding and phototherapy.
Breast milk jaundice is a different disorder that causes persistently high indirect bilirubin in a thriving healthy baby that become evident later than breastfeeding jaundice, but which generally declines in the 2"^{d} to 3'^{d} week of life.
Infants with severe or persistent jaundice should be evaluated for problems such as galactosemia, hypothyroidism, urinary tract infection, and hemolysis before ascribing the jaundice to breast milk that might contain inhibitors of glucuronyl transferase or enhanced absorption of bilirubin from the gut.

Persistently high bilirubin can require changing from breast milk to infant formula for 24-48 hr and/or phototherapy without cessation of breastfeeding.
Breastfeeding should resume after the decline in serum bilirubin.
Parents should be reassured and encouraged to continue collecting breast milk during the period when the infant is taking formula

23. A6 month old child with Tetralogy of Fallot develops cyanotic spell initiated by crying. Which one of the following drugs you would like to avoid-

a) Sodium bicarbonate

b) Propranolol

c) Phenylephrine

d) Isoprenaline

Correct Answer - D
Ans. is 'd' i.e., Isoprenaline
In Tetralogy of Fallot there is right outflow obstruction due to pulmonary stenosis with supravalvular pulmonary artery obstruction. In case of severe obstruction, the right ventricular pressure becomes greater than the left ventricular presence and the deoxygenated blood starts moving to the left ventricle resulting in severe cyanosis and erythrocytosis.

24. A child is admitted on 7 days of life with severe respiratory distress and shock. He was discharged 2 days back healthy. What could be the probable diagnosis -

a) VSD large

b) Hypoplastic left heart syndrome

c) Ebstein anomaly

d) AP window defect

Correct Answer - B
Ans is 'b' i.e., Hypoplastic left heart syndrome
<u>Hvpolastic left heart syndrome</u>

- Hypoplastic left heart syndrome occurs when parts of the left side of the heart (mitral valve, left ventricle, aortic valve, and aorta) do not develop completely. *The condition is present at birth (congenital).*
- Hypoplastic left heart is a rare type of congenital heart disease. It is more common in males than in females. As with most congenital heart defects, there is no known cause. About 10 % of patients with hypoplastic left heart syndrome also have other birth defects.
- The problem develops before birth when the left ventricle and other structures do not grow properly, including the: (i) Aorta-the blood vessel that carries oxygen-rich blood from the left ventricle to the entire body Entrace and exit of the ventricle Mitral and aortic valves
- This causes the left ventricle and aorta to be poorly developed, or hypoplastic. In most cases, the left ventricle and aorta are much smaller than normal.
- In patients with this condition, the left side of the heart is unable to send enough blood to the body. As a result, the right side of the

heart must maintain the circulation for both the lungs and the body. The right ventricle can support the circulation to both the lungs and the body for a while, but this extra workoad eventually causes the right side of the heart to fail.

- The only possibility of survival is a connection between the right and left side of the heart, or between the systemic arteries and pumonary arteries (the blood vessels that carry blodd to the lungs). Babies are normally born with two of these connections:
  (i) Foramen ovale (a hole between the right and left atrium)
  Ductus arteriosus (a small vessel that connects the aorta to the pulmonary artery)
- Both of these connections normally close on their own a few days after birth.
- In babies with hypoplastic left heart syndrome, blood from the right side of the heart travels through the ductus arteriosus. This is the only way for blood to get to the body. if the ductus arteriosus is allowed to close in a baby with hypoplastic left heart syndrome, the patient may quickly die because no blood will be pumped to the body.
- Babies with lcnown hypoplatic left heart syndrome are usually started on a medicine to keep the ductus arteriosusopen.
- Because there is little or no flow out of the left heart, blood reaming to the heart from the lungs needs to pass through the foramen ovale or an atrial septa! defect (a hole connecting the collecting chambers on the left and right sides of the heart) back to the right side of the heart. If there is no foramen ovale, or if it is too small, the baby could die. Patients with this problem have the hole between their atria opened, either with surgery or using a thin, flexible tube (heart catheterization).

Symptoms

- At first, a newborn with hypoplastic left heart may apper normal. *Symptoms usually occur in the first few hours of life, although it may take up to a few days to develop symptoms.* These symptoms may include:

  Bluish (cyanosis) or poor skin color
  Cold hands and feed (extremities)
  Lethargy

Poor pulse
Poor suckling and feeding
Pounding heart
Rapid breathing
Shortness of breath.

Since the systemic circulation is dependent on the patent ductus arteriosus *the closure of ductus arterious leads to shock.*

Signs of heart failure usually appears within the first few days or weeks of life and include dyspnoea, hepatomegaly and low cardiac output. When PDA closes suddenly shock occurs all the peripheral pulses may be weak or absent.

Ventricular septal defect

o *These patients with VSD 's become symptomatic around 6-10 weeks of age.*

o *They usually present with congestive cardiac failure.*

Ebstein's anomaly

o *Ebstein's anomaly consists of downward displacement of an abnormal tricuspid value into the right ventricle.* o *These patients usually present in teenage/adolescent years.*

o *They may also present in infancy but they usually do not present with shock or severe hypoperfusion.*

Aorticopulmonary window defect

o *An Aorticopulmanry window defect consists of a communication between the ascending Aorta and the main pulmonary artery.*

o *In these cases minimal cyanosis is present and they may present with heart failure in infancy.*

25. Loud S1 in Mitral stenosis is seen in-

a) Prolonged flow through mitral valve

b) 1st degree heart block

c) Calcification of the valve

d) Immobilization of the valve

Correct Answer - A

Ans. is 'a' i.e., Prolonged flow through mitral valve

- S_i is louder in Mitral stenosis because AV flow is prolonged.
- The loud S_i in mitral stenosis usually signifies that the mitral valve is pliable and that it remains open at the onset of isovolumetric contraction because of the elevated left atrial pressure.

S1 can be soft even in the presence of mitral stenosis, when the anterior mitral leaflet is immobile and rigid due to calcification.

26. 4 year old boy presented with recurrent chest infections. Sweat chloride test was done, showed values of 36 and 42. What is the next best investigation to confirm the diagnosis?

a) 72 hour fecal fat estimation

b) CT chest

c) Transepithelial nasal potential difference

d) DNA analysis of delta F 508 mutation

Correct Answer - C

Ans. is 'c' i.e., Transepithelial nasal potential difference

Diagnosis of cystic fibrosis

Sweat chloride testing

o The sweat test is the standard approach to diagnosis.

o The diagnosis is made by elevated sodium and chloride level in the sweat > 60 meq/l.

o Two test on different days are required for accurate diagnosis.

o A normal sweat chloride dose not exclude the diagnosis.

Genotyping and other tests such as measurement of nasal membrane potential difference, pancreatic function should be done if there is high clinical suspicion of cystic fibrosis.

Nasal potential difference

o Measurement of nasal transepithelial potential difference in vivo can be useful adjunct in the diagnosis of cystic fibrosis.

o Individuals with cystic fibrosis demonstrate a significantly more negative baseline nasal potential difference, with the topical application of amiloride there is loss of this potential difference.

Genetic analysis

o Cystic fibrosis is an autosomal recessive disorder.
o It is caused due to defect in CFTR *(Cystic fibrosis transmembrane conductance regulator) protein.*
o Cystic fibrosis is associated with large number of mutations.
o More than 1500 CFTR polymorphisms are associated with cystic fibrosis syndrome.
o The most prevalent mutation of CFTR is the deletion of single phenylalanine residue at amino acid A508. o This mutation is responsible for high incidence of cystic fibrosis in northern European populations.
o Approximately 50% of individuals with CF who are of northern European ancestry are homozygous for A508 and >70% carry at least one A508 gene. The remainder of patients has an extensive array of mutation, none of which has prevalance of more than several percent.
o Testing for cystic fibrosis mutation was not possible because of the large no. of mutations associated with the disease.
o Now days commercial laboratories test for 30-80 of the most common CFTR mutations.
o This testing identifies >90% individuals who carry 2CF mutations.
o No where it is mentioned in the texts that testing only for A508 is enough for diagnosis. Detection of atleast 2 CF mutations are necessary for making the diagnosis of cystic fibrosis.

27. A child present with recurrent sinusitis and recurrent chest infections. Chest X-ray reveals dextrocardia and situs invertus. The diagnosis is *?*

a) Kartagener's syndrome

b) Good-pasture's syndrome

c) Ehlers-Danlos syndrome

d) William Campbell syndrome

Correct Answer - A
Ans is 'a' i.e., Kartagener's syndrome
* Kartagener's syndrome is a subgroup of primary ciliary dyskinesia.
Primary ciliary dyskinesia:
* It is an autosomal recessive syndrome.
* It is characterized by poorly functioning cilia. There is absence or shortening of Dynein arms that are responsible for the coordinated bending of cilia.
* Approximately half of the patients with primary ciliary dyskinesia have kartagener's syndrome.

28. A neonate is suspected to be suffering from necrotizing enterocolitis (NEC). On further examination and investigation, he is diagnosed to be Bell's stage I NEC. The management of choice would be-

a) Laparotomy and proceed

b) Insertion of bilateral pelvic drains

c) Conservative management with IV fluids and antibiotics

d) Initial conservative management and laparotomy after 24 hours

Correct Answer - C
Ans. is 'c' i.e., Conservative management with IV fluids and antibiotics

29. A child comes with steroid resistant nephrotic syndrome secondary to FSGS, not responsive .to methylprednisolone. What next should be given *?*

a) Oral cyclophosphamide

b) Oral cyclosporine

c) Oral mycophenolate

d) IV cyclophosphamide

Correct Answer - B
Ans is 'b' i.e., Oral cyclosporine
o The treatment options for steroid resistant nephrotic syndrome are:
(i) Calcineurin inhibitors (cyclosporine, Tacrolimus)
(ii) IV or oral cyclophosphamide
(iii) Levamisol
(iv) Mycophenolate
(v) Pulse corticosteroid
o All above immunosuppressants are used along with corticosteriods (Prednisolone or methylprednisolone).
o *Cyclosporine and cyclophosphamide are most commonly used.*
o Despite these options, there is lack of consensus on first line appropriate therapy for steroid resistant nephrotic syndrome.
o According to Indian Journal of pediatric (vol. 46, Jan 17, 2009) the efficacy of these durgs are (in decreasing order): *Tacrolimus + Prednisolone > cyclosporine + Prednisolone > IV cyclophosphamide + Prednisolone > Pulse Corticosteroids (IV dexamethasone + oral cyclophosphamide + Prednisolne) > oral cyclophosphamide + PrednisoIon e.*

- *o* So, amongst the given options, best answer is cyclosporine.

30. A male child with Fanconi syndrome with nephrocalcinosis has a variant of dent disease. All are true except -

a) Hypercalciuria

b) Proteinuria

c) Similar presentation in father

d) Rickets

Correct Answer - C
Ans is `c' i.e., Similar presentation in father
Dent's disease
o Dent's disease, a *familial proximal tubular syndrome,* is an *X-linked recessive* disorder of proximal tubules characterized by:
(i) Hypercalciuria and nephrocalcinosis and nephrolithiasis
(ii) Low-molecular-weight proteinuria
(iii) Metabolic bone disease/Rickets
(iv) Progressive renal failure
(v) Marked male predominance
o In addition to above features, other renal proximal tubular defect can cause:
(i) Fanconi syndrome (Aminoaciduria, proteinuria, phosphaturia) Glycosuria, uricosuria and kalliuresis
(iii) Impaired urinary acidification
o Dent's disease occurs due to mutations that inactivate a voltage-gated chloride transporter, *CLC-5.* In some cases, it is associated with mutations in the *OCRL-1 gene* that is also mutated in the oculocerebral syndrome of Lowe. Coming back to question
o *Dent's disease is an X-linked renal tubular disorder.*

o *X-linked disease cannot be transmitted from male to male i.e., from father to son.*

31. A 7-day old infant has a leaky meningomyelocele. The most useful test for diagnosis and management of the condition is -

a) Blood-culture and sensitivity

b) Urine-culture and sensitivity

c) Rectal swab-culture and sensitivity

d) Wound swab-culture and sensitivity

Correct Answer - A
Ans is 'a' i.e., Blood-culture and sensitivity
* Most cases of meningocele are recommended for treatment as soon after birth as possible.
* In the case of a sac which is leaking fluid, the treatment is most urgent.
* Generally, treatment includes a surgical procedure to close and remove the soft tissue covering of back.
* As there are chances of bacterial meningitis and sepsis due to CSF leak, *CSF and blood sample should be send for culture and sensitivity* and peri-operative antibiotics (broad spectrum) can be changed to the specific antibiotics after the blood and CCF culture and sensitivity report.

32. An infant has hepatorenomegaly, hypoglycemia, hyperlipidemia, acidosis and normal structured glycogen deposition in liver. What is the diagnosis ?

a) Her's disease

b) Von Gierke's disease

c) Cori's disease

d) Anderson's disease

Correct Answer - B
Ans. is 'b' i.e., Von Gierke's disease
Von-Gierke disease (Type I glvcogenosis)
o It is an *autosomal recessive* disorder.
o It is due to absent or deficient activity of *glucose-6-phosphatase* in *Liven Kidney, Intestinal mucosa* **o** It can be divided into two subtypes *?*
i) *Type Ia* glucose - 6 - phosphatase is defective
ii) *Type Ib* Translocase is defective (translocase transports glucose-6-phosphatase across microsomal membrane).

33. NESTROFT test is a screeing test for-

a) β -thalassemia

b) Hereditary spherocytosis

c) Autoimmune hemolytic anemia

d) Megaloblastic anemia

Correct Answer - A
Ans. is 'a' i.e., Thalassemia
* NESTROFT (Naked Eye Single Tube Red Cell Osmotic Fragility Test) is a screening test for detection of beta thalassemia trait.
Thalassemia screening
* Widespead prevlence of thalessemia has led to a pressing demand for community screening.
* Various methods for screening of thalassemia trait are available which *include peripheral smear examination, RBC indices, Meintzer's fraction, discriminant functions, NESTROFT etc.*
* NESTROFT TEST is used in many centres in India for screening of thalassemia trait.
* If mother is NESTROFT positive, the confirmatory test for HbA2 is done for mother and the father is subjected to screening by NESTROFT. If father is also NESTROFT positive, confirmatory test for HbA2 is done for father.
* If both the parents have thalessemia trait, there is 1:4 chance of fetus having thalassemia major.
* Therefore, prenatal diagnosis is done by chorionic villus sampling (CVS) in first trimester.
* If CVS confirms the fetus to be having thalasemia major, termination of pregnancy is indicated after counseling the parents.
* If the fetus has thalassemia minor (trait) pregnancy is continued as

such and baby will have normal lifespan.
* *It is worth noting here that a positive NESTROFT test is seen in other conditions beside beta thalassemia trait.*
These are iron deficiency anemia, alpha thalassemia trait, homozygous and heterozygous HbE, HbS as well as hereditary persistence of fetal hemoglobin.
Therefore, a positive NESTROFT test should always be followed by a confirmatory test for HBA2 (eg. serum electrophoresis).

34. Which one of the following drugs is used for fetal therapy of congenital adrenal hyperplasia ?

a) Hydrocortisone

b) Prednisolone

c) Fludrocortisone

d) Dexamethasone

Correct Answer - D
Ans. is 'd' i.e., Dexamethasone
`*Recommendations for pregnancy at risk consists of adinistration of dexamethasome a steroid that readily crosses the plecenta'*

35. Short child with low T4 and raised TSH and swelling of pituitary, what is the diangosis?

a) Primary hypothyrodism

b) Pituitary tumor

c) TSH Secreting pituitary adenoma

d) TSH resistance

Correct Answer - A
Ans is 'a' i.e., Primary hypothyrodism
* Normally, T4 and T3 exert negative feedback on TSH secretion by pituitary in two ways:
(i) Block the secretion of TSH by pituitary directly major action
(ii) Block the secretion of TRH by hypothalamus (TRH stimulates the secretion of TSH by pituitary) o In primary hypothyrodism, T4 and T3 are not produced or produced in low concentration by thyroid. This results in
abolition of negative feedback on TSH secretion. Thus, there is elevated TSH and pituitary can become hyperplastic
to produce more TSH.
* In pituitary cause of hypothyroidism (secondary hypothyroidism) both TSH and Thyroid hormone (T4, T3) are low as pituitary does not secrete TSH and TSH is the major stimulation for production of T4 and T3.
About option d
* TSH resistance can also produce same picture i.e. raised TSH and low T4 with pituitary swelling.
* There is thyroid insensitivity to TSH which results in hypothyroidism (1T4 and T3). Because of reduced thyroid hormone

hypothyroidism (↓T4 and ↑TSH). Because of reduced thyroid hormone feedback, TSH is markedly elevated.
* However, Amongst the given options best option is primary hypothyrodism because TSH resistance is a very rare condition and further T4 levels are normal in Mild and moderate (Partial) TSH resistance.

36. All of the following statements about congenital rubella are true except -

a) IgG persists for more than 6 months

b) IgM antibody present at birth

c) Most common anomalies are hearing and heart defects

d) Increased risk of congenital malformation if infection occur after 16 weeks.

Correct Answer - D
Ans. is 'd' i.e., Increased risk of congenital malformation if infection occurafter 16 weeks

- Maternal infection in early pregnancy can lead to fetal infection, with resultant congenital rubella. The classic signs of congenital rubella are cataract, heart disease, deafness, and myriad other defects.
  o Congenital infection is considered to have occurred if the infant has IgM rubella antibodies shortly after birth or IgG antibodies persists for more than six months by which time maternally derived antibodies would have disappeared.
  o The most important factor in the pathogenicity of rubella virus for the fetus is gestational age at the time of infection. Maternal infection during the first trimester leads to fetal infection in – 50% of cases; maternal infection early in the second trimester leads to fetal infection in about one-third of cases. Fetal malformations not only are more common after maternal infection in the first trimester but also tend to be more severe and to involve more organ systems. Infection in the second trimester may cause deafness, but those infected after 16 weeks suffer no major abnormality.

37. Which of the following does not establish a diagnosis of congenital CMV infection in a neonate *?*

a) Urine culture of CMV

b) IgG CMV antibodies in blood

c) Intra-nuclear inclusion bodies in hepatocytes

d) CMV viral DNA in blood by polymerase chain reaction

Correct Answer - B

Ans. is `b' i.e. IgG CMV antibodies in blood

" Ig G antibody test is of little diagnostic value as positive results also reflects maternal antibodies."

38. A study under Australian collaborative trial on steroids use in neonates was done. Which of the following is true -

a) No difference between placebo & corticosteroid

b) Corticosteroid to children causes behavioural worsening

c) Corticosteroid to children causes reduction in head circumference

d) Corticosteroid to children causes neuro sensitivity degradation

Correct Answer - B
Ans is 'b' i.e., Corticosteroid to children causes behavioural worsening
o The Australian Collaborative Trial of Repeat Dose of Steroids (ACTORDS) conducted as multicentric RCT, in Australia and New Zealand by Crowtheriv, v and colleagues enrolled 982 pregnant women less than 32 weeks to receive either single dose of intramuscular Betamethasone or salined placebo weekly till 32 weeks of gestation or delivery which ever was earlier. This study showed some short term benefits in the repeat corticosteroid group with significant reduction in RDS (33% vs. 41%, RR = 0. 82[95% C10.71-0.91)]. There was also shorter duration of mechanical ventilation, lesser need for oxygen therapy and less severe lung disease. The mean weight, length and head circumference were not different. However the weight and head circumference Z scores were lower in the repeat steroid dose group. In the long term follow up these infants at 18-22 months corrected age there was no difference in growth, major disability or lung disease between the two groups. There was increased occurrence of attention problems

in repeat steroid group.

39. A neurosurgeon dropped his kid to the school then there he saw a child with uncontrollable laughing and precocious puberty. When he again went to the school in capital parents teachers meeting, he talked to the father of that boy and advised him to get an In1R1 done and the diagnosis was confirmed. What is the most probable diagnosis

a) Hypothalamic hamartoma

b) Pineal germinoma

c) Pituitary adenoma

d) Craniopharyngioma

Correct Answer - A

Ans. a. Hypothalamic hamartoma

- Uncontrollable laughing and precocious puberty are suggestive of hypothalamic hamartoma.

Hypothalamic Hamartoma

- Central precocious puberty starting before the age 3 years is often due to hypothalamic hamartoma[Q]
- . Seizures, especially laughing spells (gelastic seizures)° are seen in children with hypothalamic hamartoma.

Hypothalamic Hamartoma

- Central precocious puberty starting before the age 3 years is often due to hypothalamic hamartoma[Q]

- Hypothalamic hamartoma picked up by MRI[Q]
- Seizures, especially laughing spells (gelastic seizures)[Q]are seen in children with hypothalamic hamartoma

Precocious Puberty

- Central precocious puberty is a.lso known as true precocious puberty[Q], peripheral precocious puberty is called pseudo-precocious puberty[Q].
- McCune-Albright syndrome causes pseudo-precocious puberty (Peripheral precocious puberty).

40. A child presented to the casualty with seizures. On examination an oval hypo-pigmented macules were noted on the trunk, along with stib-norto:il [In] Probable diagnosis of the child is:

a) Neurofibromatosis

b) Sturge Weber

c) Tuberous sclerosis

d) Incontinenta pigmenti

Correct Answer - C

Ans. c. Tuberous sclerosis

- Most probable diagnosis in a child with seizures, oval hypo-pigmented macules on the trunk, and sub-normal IQ is Tuberous sclerosis.

41. A child is able to dress herself, knows her gender, feeds without spilling. What is her age

a) 2 years

b) 3 years

c) 4 years

d) 5 years

Correct Answer - B
Ans. b. 3 years
Development Milestones in a child of 3 years:

- Motor: Goes up stairs with one foot on each step, riding tricycle.
- Fine motor: Makes tower of ten cubes, draw a circle.
- Social: Dry by night, knows gender.
- Linguistic: Know age, gender and name

42. Which of the following in the natural course of disease has no reversal of the shunt

a) ASD

b) VSD

c) TOF

d) PDA

Correct Answer - C
Ans. c. TOF

- Infants with acyanotic gradually become cyanotic as a result of worsening condition of the infundinular stenosis a.nd polycythemia
- Polycythemia develop secondary to cyanosis
- Hypoxic spells may develop in infants
- Growth retardation may be present if cyanosis is severe Brain abscess and cerebrovascular accidents rarely occur
- SABE is occasionally a complication
- Some patients, particularly those with severe TOF develop AR
- Coagulopathy is a late complication of longstanding cyanosis

Eisenmenger syndrome

- Eisenmenger syndrome was so named by Dr. Paul Wood after Dr. Victor Eisenmenger, who first described the condition in 1897.

Congenital heart defects causing Eisenmenger syndrome

- Atrial septal defects
- Ventricular septal defects
- Patent ductus arteriosus
- More complex types of acyanotic heart disease

43. A preterm infant with poor respiration at birth starts throwing seizures at 10 hours after birth. Anti- epileptic of choice shall be:

a) Leveteracetam

b) Phenytoin

c) Phenobarbitone

d) Lorazepam

Correct Answer - C
Ans. c. Phenobarbitone
Phenobarbital:

- Drug of first choice in neonatal seizures
- Loading dose: 20 mg/Kg
- If this dose is not effective, then additional doses of 5-10 mg/kg can be given until a dose of 40 mg/kg is reached.
- After 24 hours of loading dose, maintenance dosing can be started at 3-6 mg/Kg/day, usually administered in two separate doses

Treatment of Neonatal Seizures

- A mainstay in the therapy of neonatal seizures is the diagnosis and treatment of underlying etiology[Q].
- Etiologies: Hypoglycemia[0], hypocalcemia[Q], meningitis[Q], drug withdrawal and trauma0

Drugs used in Neonatal Seizures
Phenobarbital

- Drug of first choice in neonatal seizures[Q]
- Loading dose: 20 mg/Kg
- If this dose is not effective, then additional doses of 5-10 mg/kg can

be given until a dose of 40 mg/kg is reached.

- After 24 hours of loading dose, maintenance dosing can be started at 3-6 mg/Kg/day, usually administered in two separate doses.
- Metabolized by liver, excreted by kidney

Drugs used in Neonatal Seizures

Phenytoin/Fosphenytoin

- If a total loading dose of 40 mg/kg of Phenobarbital was not effective, then a loading dose of 15-20 mg/Kg of Phenytoin or Fosphenytoin can be administered IV.
- Rate should not exceed 0.5-1 mg/Kg/min in order to prevent cardiac problem.
- Fosphenytoin, which is phosphate ester prodrug is preferred. It is highly soluble in water and can be administered very safely IV or IM, without causing injury to tissues

Lorazepam

- The initial drug used to control acute seizure is usually Lorazepam
- Can be use either as the initial drug or as a 2"^{d} line treatment in a newborn who does not respond to the treatment with Phenobarbital and Phenytoin.
- Lorazepam is distributed to brain very quickly and exerts its anticonvulsants action in minutes.

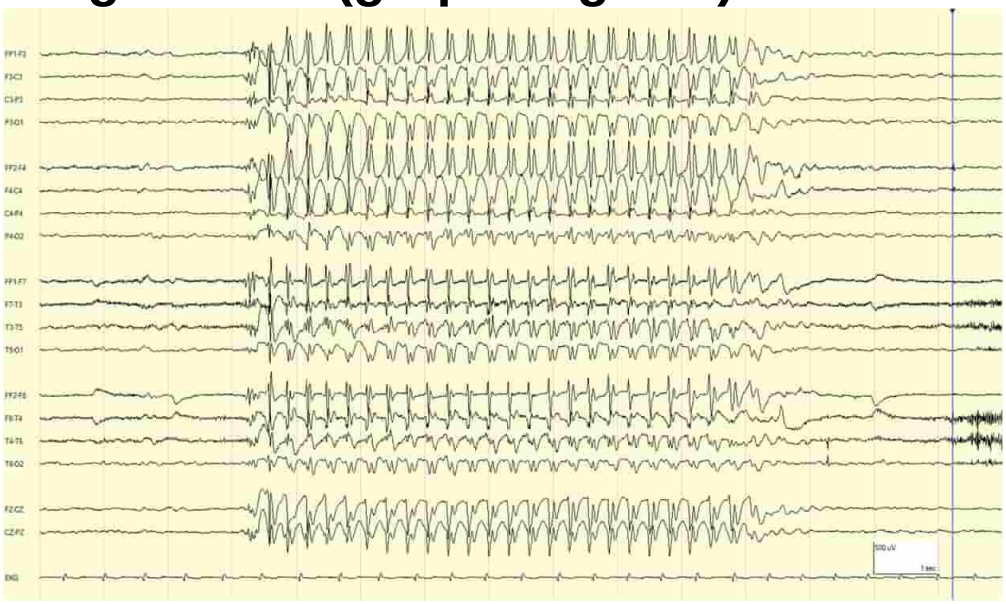
Diazepam and Midazolam

- Highly lipophilic, so it distributes very rapidly into the brain and then is cleared very quickly out, carrying the risk of recurrence of seizures
- Carries the risk of apnea and hypotension
- However, the IV preparation contains sodium benzoate and benzoic acid, it is currently not recommended as a first line agent.

Leveteracetam

- Dose: 10-30 mg/Kg/day
- Drug of 2"^{d} or 3rd choice

44. A 7 years old girl with falling grades and complaints by teacher that she is inattentive in class to her parents and has bad school performance. On hyperventilation her symptoms increased and showed the following EEG findings. Diagnosis is (graph is given):



a) Myoclonic epilepsy

b) Myoclonus

c) Absence seizure

d) Juvenile myoclonic epilepsy

Correct Answer - C
Ans. c. Absence seizures

Absence Seizures: **-**
Start at 5-8 years of age
Do not have an aura
Usually last for only a few seconds
Accompanied by flutter or upward rolling of eye
Hyperventilation for 3-5 minutes can **precipitate the seizure**

45. One of the parents has a balanced translocation between chromosome 15 and 21. What advice will you provide to the couple to prevent a child being born with Down's syndronu

a) Prenatal diagnosis and advice abortion

b) Artificial insemination with donor's sperm

c) Adoption

d) No need to worry as there is no increased risk

Correct Answer - A
Ans. a. Prenatal diagnosis and advice abortion
abortion *Ref- Nelson 19/e p 02, 4054)*
www.ncbi.nlm.nih.gov/pubmed/21910218
Nelson says "Translocation (21;21) carriers have a 100% recurrence risk for a chromosomalIN abnormal child."
If the mother is a 'balanced translocation' carrier to another chromosome (usually 13, 14, 15, 22) then the recurrence risk is about 1 in 8.
In the question, one of the parents has a balanced translocation between chromosome 15 and 21. Prenatal diagnosis and abortion will prevent a child being born with Down's syndrome

46. About trisomy 13, which of the following is a true statement

a) Bilateral mircophthalmia

b) Neurofibroma

c) Adenoma

d) Dermoid cyst

Correct Answer - A
Ans. a. Bilateral mircophthalmia
Nervous system
Intellectual disability and motor disorder
Microcephaly
Holoprosencephaly (failure of the forebrain to divide properly).
Structural eye defects, including microphthalmia, Peters' anomaly, cataract, iris or fundus (coloboma), retinal dysplasia or retinal detachment, sensory nystagmus, cortical visual loss, and optic nerve hypoplasia
Meningomyelocele (a spinal defect)
Musculoskeletal and cutaneous
Polydactyly (extra digits)
Cyclopia
Proboscis
Congenital trigger digits
Low-set ears
Prominent heel
Deformed feet known as rocker-bottom feet
Omphalocele (abdominal defect)
Abnormal palm pattern
Overlapping of fingers over thumb

Cutis aplasia (missing portion of the skin/hair)
Cleft palate

47. 152. A child with hypoglycemia is not able to utilize glucose from glycogenolysis or gluconeogenesis. Which of the following enzyme is deficient in the child?

a) Fructokinase

b) Glucokinase

c) Glucose 6-phosphatase

d) Transketolase

Correct Answer - C
Ans. c. Glucose 6-phosphatase
Von Gierk's disease (AR)

- Inherited as autosomal recessive°
- Caused by glucose-6-phosphatase deficiency in liver, kidney, and intestinal mucosa.
- Skeletal muscle is deficient in glucose-6 phosphatase, so muscles are not affected°.
- Structure of glycogen is normal° but not available metabolically

Two subtypes of GSD I

Type la	Type lb
Defective enzyme:Glucose-6- phosphatase	Defective enzyme: Translocase

- The defects in both subtypes lead to inadequate conversion in the liver of glucose-6-phosphate to glucose and thus make affected individuals susceptible to fasting hypoglycemia

Von Gierk's disease (AR)

- Clinical Features
- Clinical features are hypoglycemia, lactic academia, ketosis, hyperlipidemia, increased uric acid and splenomegaly[Q]
- Liver cells and renal tubule cells loaded with glycogen°.
- Organs affected are liver, intestine and kidney°

Long-Term Complications

- Gout, polycystic ovaries, hepatic adenomas
- Increased risk of pancreatitis, cardiovascular disease.
- Frequent fractures (osteopenia/osteoporosis)
- Renal disease (proteinuria, hypertension, kidney stones, nephrocalcinosis, and altered creatinine clearance)
- Pulmonary hypertension (rare)

Diagnosis:

- **C**linical presentation and abnormal plasma lactate and lipid values suggest that a patient may have GSD I
- Gene-based mutation analysis provides a noninvasive means of reaching a definitive diagnosis for most patients of GSD I
- Definitive diagnosis required a liver biopsy to demonstrate a deficiency.

Treatment of Type I GSD:

- Maintenance of normal blood glucose levels through continuous infusion of glucose via feeding tube or ora administration of uncooked cornstarch (slow-release form of glucose)
- Fructose and galactose cannot be converted to free glucose, their dietin take should be restricted
- Dietary supplements of multivitamins, calcium and Vitamin D
- Allopurinol
- Use of medium chain triglycerides, fish oil and lipid-lowering drugs such as statins and fibric acids
- ACE inhibitors, Citrate supplementation
- Orthotropic liver transplantation: Reserved for GSD I patients with liver malignancy, multiple liver adenomas, metabolic derangements refractory to medical management, and/or liver failure.

48. In a child, CSF examination is not used in diagnosis of:

a) ALL

b) Hodgkin's lymphoma

c) Non- Hodgkin's lymphoma

d) AML

Correct Answer - B

Ans. b. Hodgkin's lymphoma

- Acute Lymphoblastic Leukemia (ALL), Non-Hodgkin's lymphoma and Acute Myeloid Leukemia disseminate via CSF. So, CSF examination can be used in the diagnosis. Prophylactic craniospinal irradiation is used in the treatment
- In Hodgkin s lymphoma, there is no CSF spread, so CSF examination is not used in the diagnosis

Prophylactic Craniospinal Irradiation

- Prophylactic craniospinal irradiation is useful in CNS malignancy which disseminate via CSF or any malignancy with high risk of CNS spread[Q]

Indications of Prophylactic Craniospinal Irradiation

- Medulloblastoma[Q]
- Glioblastoma[Q]
- Germinoma[Q]
- Small cell Ca of lung
- AL[LQ]
- Non-Hodgkin's lymphoma[Q]
- Leptomeningeal
- Rhabdomyosarcoma[Q]

49. Which of the following is not a sign of PDA in a preterm baby?

a) Apnea

b) Tachycardia

c) Necrotizing enterocolitis

d) Narrow pulse pressure

Correct Answer - D
Ans. d. Narrow pulse pressure

- "Premature newborns cannot tolerate PDA, so it results in heart failure, respiratory distress or necrotizing enterocolitis. So they require prompt management"- Ghai 7/e p406

50. All of the following are side effects of growth hormone (supplements) therapy except

a) Slipped capital femoral epiphysis

b) Gynaecomastia

c) Hypoglycemia

d) Pseudotumour cerebri

Correct Answer - C
Ans. c. Hypoglycemia
Ref Goodman and Gilman's 12/e p1116; Harrison 19/e p2260, 18/e p2894) ***Hypoglycemia is not a side effect of growth hormone (supplements) therapy***

51. A 10-year old male child was presented to the paediatrician for evaluation of a seizure disorder. On examination a vascular plaque was found along the ophthalmic and maxillary divisions of the trigeminal nerve. The mother informed the paediatrician that the lesion was present since birth and there was no change in morphology. The most likely possibility is

a) Sturge-Weber syndrome

b) Infantile hemangioma

c) Congenital hemangioma

d) Proteus syndrome

Correct Answer - A

Ans. a. Sturge-Weber syndrome

History of seizure disorder with a vascular plaque along the ophthalmic and maxillary divisions of the trigeminal nerve present since birth without any change in morphology is highly suggestive of Sturge-Weber syndrome

Sturge-Weber syndrome / Encephalotrigeminal syndrome

- Usually sporadic, characterized by:
- Large unilateral cutaneous angioma^{Q} (port-wine stain)
- Angiomas in brain involving ipsilateral cerebral hemisphere and meninges
- Focal seizures° typically occurs opposite to the side of lesion°

Adrenal pheochromocytoma

- - Cerebral angiomas lead to cortical atrophy[Q]
- Angiomas are visible radiologically as Tram-track or rail track calcification mainly in occipital region[Q]

52. What is the pathology of edema in nephrotic syndrome

a) Reduced plasma protein

b) Sodium and water retention

c) Increased venous pressure

d) Hyperlipidemia

Correct Answer - B
Ans. b. Sodium and water retention
Sodium and water retention is the pathology of edema in nephrotic syndrome.
The nephrotic syndrome is characterized by proteinuria, edema, and hypoalbuminemia. Renal sodium retention and changes in variables of the Starling equation are fundamental to the pathophysiology of nephrotic edema. There is evidence for both intravascular volume expansion (overfilling) and intravascular volume depletion (under filling) in patients with nephrosis. Microvascular fluid exchange is described using a formulation of the Starling driving forces (DP and Dp) and it is through this equation that nephrotic edema is conceptualized. Previous theories have focused on abnormalities in DP and Dp to explain nephrotic edema. Studies have shown that hypoalbuminemia (and thus Dp) is not a likely cause of edema formation in most nephrotic patients owing to a parallel decrease in interstitial fluid albumin and an increase in interstitial fluid pressure, both of which serve to maintain edema driving forces constant. There is limited evidence suggesting that abnormalities in vascular permeability (Kf and s) may contribute to edema formation. A major advance in our understanding of the pathophysiologic basis of edema formation in the nephrotic syndrome is the discovery that

proteinuria can cause primary renal sodium retention through ENaC activation. This mechanism is likely active in all patients with nephrotic syndrome, regardless of their intravascular volume status. Other causes of primary renal sodium retention include increased renal efferent sympathetic nerve activity, ANPase, and in the expression and activity of the Ne—le in the collecting duct in animal models. Furthermore, excess serum vasopressin levels have been found to contribute to free water retention in some patients with the nephrotic syndrome."- Eric Siddall and Jai Radhakrishnan. The pathophysiology of edema formation in the nephrotic syndrome

Nephrotic Syndrome

Manifestations of Nephrotic Syndrome

- **1.** Massive proteinuria, with the daily loss of 3.5 gm or more of protein[Q]
- 2. Hypoalbuminemia, with plasma albumin levelsQ
- 3. Generalized edema[Q]
- 4. Hyperlipidemia and lipiduria[Q]

Pathophysiology:

- Renal sodium retention and changes in variables of the Starling equation are fundamental to the pathophysiology of nephrotic edema.
- There is evidence for both intravascular volume expansion (overfilling) and intravascular volume depletion (under filling) in patients with nephrosis.

Nephrotic Syndrome

Mechanisms of Sodium Retention in the Nephrotic Syndrome

- Increased angiotensin 11-independent afferent and efferent arteriolar tone because of increased efferent sympathetic nerve activity.
- Tubular resistance to atrial natriuretic peptide (ANP).
- Increased number of open epithelial sodium channel (ENaC) channels in the cortical collecting duct due to proteolytic activation of ENaC by plasmin.
- Increased number and activity of cortical collecting duct Na/K ATPase channels

Most important facts about Nephrotic syndrome

- The lipid appears in the urine either as free fat or as oval fat bodies,

representing lipoprotein resorbed by tubular epithelial cells and then shed along with the degenerated cells.

- Most proteins are decreased in nephrotic syndrome except Fibrinogen and lipoproteins, due to increased synthesis.

Proteins decreased	**Consequence**
Albumin	*Edema due to hypoalbuminemia*[Q]
Transferrin	*Iron resistant microcytic anemia*[Q]
Cholecalciferol binding proteins	*Hypocalcemia*[Q]
Thyroxin binding globulin	*Decreased thyroxin (Hypothyroid state)*[Q]
IgG	*Increased susceptibility of infections*[Q]

- Renal vein thrombosis is particularly common (up to 40%) in patients with nephrotic syndrome due to membranous glomerulopathy, memranoproliferative glomerulonephritis, and AmyloidosisQ.
- As a consequence of hypercoagulability and changes in proteins, patients can develop spontaneous peripheral arterial or venous thrombosis, renal vein thrombosis, and pulmonary embolismQ

53. Which of the following findings in a newborn suggests RDS

a) Onset after 6 hours of birth

b) Antenatal cortocosteroid therpay in mother

c) Air bronchogram on chest X-ray

d) Term gestation

Correct Answer - C
Ans. c. Air bronchogram on chest X-ray
Ref Nelson 18/e p731-735) Air bronchogram on chest X-ray in a newborn suggests RDS.

- RDS is common in pre-term infants
- Overall incidence is 10-15%, but can be as high as 80% in neonates
- Surfactant production starts around 20 weeks of life and peaks at 35 weeks of gestation.

Approximately 80% of neonates born at 26-28 weeks gestation develop RDS, whereas less than 30% of premature neonates born at 30-31 weeks gestation develop RDS

54. On exposure to cold, a neonate shows all of the following mechanisms except

a) Shivering

b) Crying and flexion of body like fetus position

c) Cutaneous vasoconstriction

d) Increased production of noradrenaline for breakdown of brown fat in adipose tissue

Correct Answer - A
Ans. a. Shivering
On exposure to cold and wet environment, the neonate tries to generate heat by increased activity (crying with agitated movement) and a sympathetic surge that causes vasoconstriction and non-shivering thermogenesis in the brown fat. Babies attempt to conserve heat by peripheral vasoconstriction."
"Brown fat is a well-vascularized, sympathetically innervated lipid collection located in the axillae, groin, nape of the neck, interscapular area and perinea! area. Cold stress causes the release of norepinephrine that uncouples beta-oxidation in fat with resultant heat generation. Preterm and small-for-gestational age infants have immature thermogenic response because of scanty brown fat stores."
Role of Brown Fat in Thermogenesis

- A newborn baby is more prone to develop hypothermia because of large surface area per unit of body weight.
- In infants, brown fat is an important site of thermogenesis. It results in the so-called non-shivering thermogenesis.
- Brown fat is located around the adrenal glands, kidney, nape of neck, interscapular and axillary region.

- Metabolism of brown fat leads to heat production.
- Blood flowing through the brown fat becomes warm and through circulation, transfers heat to other parts of the body. This mechanism oh heat production is known as non-shivering thermogenesise.Q

55. For a neonate at 48 hours of birth with a history of non-passage of meconium, next step in evaluation will be:

a) Sweat chloride level

b) CFTR mutation analysis

c) Lower GI study

d) Manometry

Correct Answer - C
Ans. c. Lower GI study
For a neonate at 48 hours of birth with a history of non-passage of meconium, next step in evaluation will be lower GI
study.
'Timely passage of the first stool is a hallmark of the well-being of the newborn infant. Failure of a fall-term newborn
to poss meconium in thefirst 2 4 hours may signal intestinal obstruction. Lower intestinal obstruction may be associated
with disorders such as Hirschsprung's disease, anorectal malformations, meconium plug syndrome, smctll left cobn
st'ndrome, hypoganglionosis, neunsnal intestinal dysplasia and megacystis-microcolon-intestinal hypoperistalsis syndrome.
Radiologic studies are usually required to make the diagnosis

56. Which of the following vasculitis is not seen in adults?

a) Takayasu arteritis

b) Susac syndrome

c) Henoch-Schonlein purpura

d) Kawasaki disease

Correct Answer - D
Ans. d. Kawasaki disease
Main visceral arteries and their branches
Kawasaki disease
- Arteritis with mucocutaneous lymph node syndrome; usually occurs in children.
Coronary arteries can be involved with aneurysm formation and/or thrombosis.

57. A 32-week, 1400 g neonate is born to a primigravida. The baby did not require resuscitation and showed stable vitals. The baby was transferred to the NICU. How will you manage the feeding of the patient?

a) Start total enteral feeding and IV feeding not required

b) Start IV feeding with minimal enteral feeding

c) Start IV feeding and introduce feeding on 2nd day of life

d) Start parenteral feeding and institute oral feeding on 2nd day of life

Correct Answer - A
Ans. a. Start total enteral feeding and IV feeding not required
The optimal time to introduce enteral feeding to a sick premature or LBW infant is controversial.
. Trophic feeding is the practice of feeding very small amounts of enteral nourishment to VLBW preterm infants
to stimulate development of the immature gastrointestinal tract.
. The benefits of trophic feeding include enhanced gut motility, improved
growth, decreased need for parenteral nutrition, fewer episodes of sepsis,
and shortened hospital stay.
. Once the infant is stable, small-volume feedings are given in addition to
intravenous fl uids/n utrition.
. Feeding is gradually advanced and parenteral nutrition

. Feeding is gradually advanced and parenteral nutrition decreased.
. This approach may reduce the incidence ofnecrotizing enterocolitis.

58. Which of the following is not a component of Kangaroo mother care (KMC)?

a) Skin to skin contact

b) Supplementary nutrition

c) Exclusive breast feeding

d) Early discharge and follow-up

Correct Answer - B

Ans. b. Supplementary nutrition

Kangaroo position

The kangaroo position consists of skin-to-skin contact (SSC) between the mother and the infant in a strictly vertical position, between the mother's breasts and under her clothes.

SSC should be started as early as possible after birth and can be of two types depending upon the duration: continuous or intermittent.

Kangaroo nutrition

This can be exclusive breastfeeding/fortification.

- Kangaroo nutrition is the delivery of nutrition to "kangarooed" infants as soon as oral feeding is possible.
- It is based on exclusive breastfeeding by direct sucking, whenever possible.
- Goal is to provide exclusive or nearly exclusive breastfeeding with fortification, if needed.
- Breastfeeding is an integral component of KMC and it might contribute to significant gains in neurological development and IQ

59. Asymmetric Moro's reflex at birth is indicative of:

a) HIE

b) Brain damage

c) Erb's palsy

d) Kernicterus

Correct Answer - C
Ans. c. Erb's palsy
Moro's Refex:
Obtained by placing the infant in a semi-upright position. The head is momentarily allowed to fall backward, with immediate resupport by the examiner's hand.
The child symmetrically abducts and extends the arms and flexes the thumbs, followed by flexion and adduction of the upper extremities.
An absent or exaggerated response in a newbom is ominous suggesting significant dysfunction of the CNS
An asymmetrical response may signify a fracture clavicle, brachial plexus injury or hemiparesis

60. A very preterm baby on 30 mL/kg of enteral feeding developed sudden severe abdominal distension with visible bowel loops on day 6 of life. The baby also showed temperature instability and lethargy. s-ray of the abdomen showed portal venous gas. The staging of NEC is:

a) lb

b) 2a

c) 2b

d) 3a

Correct Answer - C
Ans. c. 2b
IIB- Proven NEC, moderately ill
Same as lA + mild metabolic acidosis + mild thrombocytopenia
Same as lA + absent bowel sounds, definite tenderness, abdominal cellulitis, right lower quadrantmass
Same as llA + portal venous gasa + ascites
NPO, Antibiotics x 14 days

61. . The most common fungal infection in the neonates transmitted by caregiver's hand is:

a) *Candida albicans*

b) *Candida glabrata*

c) *Candida tropicalis*

d) *Candida parapsilosis*

Correct Answer - D
Ans. d. Candida parapsilosis
"C. parapsilosis infections are esptecially associated with hyperalimentation solutions, prosthetic devices, and indwelling catheters, as well as the nosoeomial sprcad of disease throagh the hands of health care workers

62. 'Bull-neck' is seen in severe cases of which of the following?

a) Diphtheria

b) Tubercular lymphadenitis

c) Mumps

d) Goitre

Correct Answer - A
Ans. a. Diphtheria
Respiratory Diphtheria
Clinical Features:

- The clinical diagnosis of diphtheria is based on:
- Constellation of sore throat[Q]
- Adherent tonsillar, pharyngeal, or nasal pseudomembranous lesions[Q]
- Low-grade fever[Q]
- Occasionally, weakness, dysphagia, headache, and voice change are the initial manifestations[Q]
- Neck edema and difficulty breathing are seen in more advanced cases and carry a poor prognosis[Q].
- The systemic manifestations of diphtheria stem from the effects of diphtheria toxin and include weakness as a result of neurotoxicity and cardiac arrhythmias or congestive heart failure due to myocarditis[Q]
- The pseudomembranous lesion is most often located in the tonsillopharyngeal region[Q]
- The diphtheritic pseudomembrane is gray or whitish and sharply demarcated[Q]Pseudomembrane in diphtheria is tightly adherent to

the underlying tissues[Q]

- Less commonly, the lesions are detected in the larynx, [p]ares, and trachea or bronchial passages. Large pseudomembranes are associated with severe disease and a poor prognosis[Q].
- A few patients develop swelling of the tonsils and present with "bull-neck" diphtheria, which results from massive edema of the submandibular and paratracheal region and is further characterized by foul breath, thick speech, and stridorous breathing[Q]
- Unlike the exudative lesion associated with streptococcal pharyngitis, the pseudomembrane in diphtheria is tightly adherent to the underlying tissues

Diagnosis:

- Attempts to dislodge the membrane may cause bleeding.
- Hoarsensess suggests laryngeal diphtheria, in which laryngoscopy may be diagnostically helpful.
- In addition, diagnosis requires the isolation of C. diphtheria or the histopathologic isolation of compatible gram-positive organisms[Q].

63. A child presents with albinism. He should be evaluated for:

a) ENT consultation

b) Eye consultation

c) Electrocardiography

d) Neurosurgery

Correct Answer - B
Ans. b. Eye consultation
uoculocutaneans albinism We 1 (OCA I) is characterized by great reductinn in or absence of erosinase acttvily, OCAIA, the most severcform, is characterized by a tack of visible pigment in hair, skir4 and eyes. This manifesi as photophobia, nystagmas, defective visual acui4t, white hair, and white skin OCAIB, or yellow mutant albinism, manifests at birrh as white haiti pink skin, and gray e"ves. OCA2 ranges from nearly normal to closely resembling type I albinism. This is the most commonform of albinism seen worldwidc. Progressive improvement in visual ocuity and nystagmus occurs with aging

64. A 1.5 year old female is brought to the clinic with complaints of excessive enlargement of head, intolerance to feeds and severe malnourishment. MRI imaging was suggestive of a medulloblastoma causing obstructive hydrocephalus. Which of the following is an example of irrational management-the patient?

a) Craniotomy and sub-total excision of the tumour. Surgeon leaves the layer of the tumour adherent with colliculus

b) First ventirculoperitoneal shunt was done

c) CCNU and vincristine were given as chemotherapy

d) Radiotherapy 35-40 Gy was given to the whole craniospinal axis

Correct Answer - D
Ans. d. Radiotherapy 35-40 Gy was given to the whole craniospinal axis
Highly malignant tumor found in cerebellum and infratentorial location
. Occur predominantly in children (peak inciden ce at 34 years)
. Medulloblastoma is most radiosensitive brain tumor
. MC site: Vermis (75%)
. MC site in adults: Lateral ceretrellar hemisphere
Clinical Characteristics:
. Child usually presents with features ofincreased intracranial

tension.
. Adults present with ataxia and unilateral dysmetria as lateral origin is more common.
Metastasis:
. Dissemination through CSF is common leading to drop metastasis.
. Metastasis outside CNSa affects bone, lymph node and liver.
. Tumor dissemination is most important prognostic factor.
Treatment:
. Despite of extreme radiosensitivity, it should be surgically excised.
. Surgical excision should be followed by radiotherapy and chemotherapy.
. BCNU and vincristine are primarily used for recurrences. in poor-risk patients, and in children <3 years to avoid radiation therapy.

65. True about SLE is?

a) Autoimmune disease

b) Childhood SLE had poor prognosis than adult SLE

c) Presence of ANA

d) All are true

Correct Answer - D
Ans. is 'd' i.e., All are true
SLE (sytemic lupus Erythematosus)

- Autoimmune disorder
- Inflammation of blood vessel
- Childhood SLE had poor pnognosis than adult SLE

Hall mark of SLE is presence of antinuclean antibody (ANA)

- More common in female.
- Malar rash in pathognomic of SLE
- Non - erosive arthritis
- Nephritis
- Encephalopathy
- Pleuritis / Pericarditis
- Cytopenia

66. Kartagener syndrome all true except?

a) Bronchitis

b) Sinusitis

c) Bronchiectasis

d) Infertility

Correct Answer - A

Ans. is 'a' i.e., Bronchitis

- Kartagener syndrome is a subset of primary ciliary dyskinesia, an autosomal recessive condition characterized by abnormal ciliary structure and/or function leading impaired mucociliary clearance.
- Kartagener is a primary ciliary disorder and as such the sperms have abnormal/reduced motility (Asthenozoospermia).
- Blockage of epididymis is a feature of Young syndrome (But there is no Situs inversus like Kartagener syndrome and patients tends to have normal sperm motility)

Clinical presentation

- Kartagener syndrome is characterised by the clinical triad of 1
- Situs inversus
- Chronic sinusitis and/or nasal polyposis
- Bronchiectasis

Other features include

- Telecanthus: widened interpupillary distance by a nasal polyp
- Infertility in males
- Subfertility in females

67. Hypergonadotropic hypogonadism ?

a) Decrease FSH and LH

b) Decrease FSH and increase LH

c) Increase FSH increase LH

d) Increase FSH decrease LH

Correct Answer - C
Ans. is ‘-c' i.e., Increase FSH increase LH
Hypergonadotropic hypogonadism
* Also K/a primary or peripheral hypogonadism.
* Characterised by hypogonadism due to an impaired response of the gonads to the gonadotropin, FSH and LH.
* In turn a lack of sex steroid production and elevated gonadotropin level(FSH and LH).
Causes : Chromosomal abnormalitis
* Turner's syndrome
* Klinefelter syndrome
* Swyer's syndrome
Enzyme defect
* 17 , hydroxylase
* 17, 20 lyase deficiency

68. APGAR score - include A/E

a) Heart rate

b) Respiratory rate

c) Muscle tone

d) Color

Correct Answer - B
Ans. is 'b' i.e., Respiratory rate
The Apgar test is-
Breathing effort
Heart rate
Muscle tone
Reflexes
Skin color

69. A 7-year-old child with steroid dependent nephrotic syndrome has developed corticosteroid toxicity and posterior subcapsular cataracts. Which of the following is the best alternative for the treatment of the patient?

a) Levamisole

b) Cyclophosphamide

c) Mycophenolate

d) Cyclosporine

Correct Answer - B
Ans. b. Cyclophosphamide

- If a child with nephrotic syndrome on steroid develops features of corticosteroid toxicity (cushingoid features, hypertension, cataract and growth failure) then cyclophosphamide should be used.

70. Which of the following is least likely in PDA?

a) CO, wash out

b) Necrotizing enterocolitis

c) Bounding pulse

d) Pulmonary hemorrhage

Correct Answer - A
Ans. a. CO, wash out

- Infants and children with a small PDA are generally asymptomatic; infants with a large PDA present with signs of heart failure.
- 'Premature newborns can't tolerate PDA, so it results in heart failure, respiratory distress or necrotizing enterocolitis.
- Premature infants may present with respiratory distress, apnea, worsening mechanical ventilation requirement or other serious complications (e.g. necrotizing enterocolitis)
- Signs of heart failure occur earlier in premature o infants than in full-term infants and may be more severe.
- A large ductal shunt in a premature infant often is a major contributor to the severity of the lung disease of prematurity

71. All of the following are considered development delay except:

a) Pincer grasp not at 9 months

b) Not able to sit at 9 months

c) Not able to go up and down stairs at 2.5 years of age

d) 2 words phrase at 18 months of age.

Correct Answer - D

Ans. d.2 words phrase at 18 months of age (Ref. Ghai 7/e p3t)

- Spontaneous 2-word phrases (pivotal speech), consisting of the flexible characteristic of 2 years old and reflects the emergence of grammatical.
- Development Milestones in a child of 3 years:
- Motor: Goes up stairs with one foot on each step, riding tricycle.
- Fine motor: Makes tower of ten cubes draw a circle.
- Social: Dry by night, knows gender.
- Linguistic: Know age, gender and name

72. A 2-day-old neonate in the neonatal ICU develops seizures. Which of the following would be the next best investigation for the child?

a) Transcranial ultrasound

b) CT Head

c) MRI brain

d) X-ray

Correct Answer - A
Answer- A (Transcranial ultrasound)

- `Cranial ultrasonouraphy is the main imagine modality of premature neonates and well suited for the study of neonates in general.

73. A 2-year-old male child presents with a lump in the right side of the abdomen. Ultrasound revealed it to be a solid mass. On examination, his right arm and leg were found to be longer. The most likely diagnosis is:

a) Wilm's tumor

b) Neuroblastoma

c) Nephroblastoma

d) Angiomyolipoma

Correct Answer - A
Answer- A (Wilm’s tumour)
Wilm's tumor: MC primary renal tumor of childhood (2-5 years)

- Wilm's tumor: 2nd MC malignant abdominal tumor in children (MC is neuroblastoma).
- Arise from kidney, composed of three elements- blastema, epithelium and strome.
- MC presenting feature is asymptomatic abdominal mass or swelling.
- Mostly unilateral.
- Characterized by triad of abdominal mass, fever and microscopic hematuria.
- Fever typically resolves after tumor resection

74. A 2-year-old child was brought to the emergency department at 3 AM. The child presented with fever and cough. On examination, respiratory rate was 36/ min, temperature was 39°C and saturation on pulse oximetry was 96%. The child had barking cough and stridor only on crying. Otherwise, the child was hydrated, able to drink and consolable. The nest step in management would be:

a) Racemic epinephrine nebulization

b) Single dose of dexamethasone

c) Complete blood count and culture

d) Nasal washings for Respiratory Syncytial Virus (RSV) and influenza

Correct Answer - B

Answer- B (Single dose of dexamethasone)

- Corticosteroids decrease the edema in the laryngeal mucosa through their anti-inflammatory action.
- Oral steroids are beneficial, even in mild croup, as measured by reduced hospitalization, shorter duration of hospitalization, and reduced need for subsequent intimations such as epinephrine administration.

75. A 4 years old child presented in emergency with respiratory difficulty and noisy breathing with drooling of saliva. Emergency X-ray showed thumb sign. Diagnosis is:

a) Croup

b) Epiglottitis

c) Foreign body aspiration

d) Retropharyngeal abscess

Correct Answer - B
Answer- B (Epiglottis)
Clinical Features:

- Onset is sudden
- Symptoms: Fever, dysphagia, drooling of saliva, muffled voice, inspiratory retractions, cyanosis and soft stridor
- Patients often sit in sniffing dog position

Imaging:

- On laryngoscopy: 'Cherry Red' swollen epiglottis
- X-ray:'Thumb print' sign

76. A 12 years old boy develops sore throat of 4 day s duration. On examination, yellow grayish patch seen over both the tonsils and foul smell coming from his mouth. Which of the following non-suppurative complication is of concern?

a) Acute rheumatic fever

b) Acute glomerulonephritis

c) Both acute rheumatic fever and acute glomerulonephritis

d) Scarlet fever

Correct Answer - C
Answer- C (Both acute rheumatic fever and acute glomerulonephritis)
In streptococcal infections, both acute rheumatic fever and acute glomerulonephritis are non-suppurative complication and is of concern.

77. 8-year-old boy complains of increasing muscle weakness. On examination, his calves are bulky and Muscle tightening. His serum creatine kinase levels are increasing with age. Which of the following is likely diagnosis

a) Hereditary sensorimotor neuropathy

b) Myelin deficiency

c) Dystrophin deficiency

d) Congenital myopathy

Correct Answer - C
Answer- C (Dystrophin deficiency)

- Most likely diagnosis is Duchenne Muscular Dystrophy due to Dystrophin deficiency.

78. A baby can breathe and suck at the same time. This is due to:

a) Highly placed larynx

b) Wide short tongue

c) Short soft palate

d) Short pharynx

Correct Answer - A
Answer- A (Highly placed larynx)

- A baby can suck milk into mouth and because of its palate in mouth is separated from its nasal cavity so while baby is sucking in milk can also breath through nose.
- When the infant has to swallow, the soft palate rapidly moves upward to close off the back of the back of nasal air tube'.

79. Presented edema, oliguria and frothy urine. He has no past history of similar complaints. On examination, his urine was positive for 3+ proteinuria, no RBCs/WBCs and no casts. His serum albumin was 2.5 gm/L and serum creatinine was 0.5 mg/dL. The most likely diagnosis is:

a) Minimal change disease

b) IgA nephropathy

c) Interstitial nephritis

d) Membranous nephropathy

Correct Answer - A
Ans: A.Minimal change disease
(Ref Harrison 19/e p184, 18/2345)

- **Most likely diagnosis - Minimal change disease.**
  Minimal Change Disease:
- **Peak age of onset =** Between 6-8 years of age (usually <10 years)
- **Type of onset** = Insidious
- **Typical presentation =** Nephrotic syndrome
- **Hallmark of Nephrotic syndrome =**Peripheral edema.
- Occurs when serum albumin levels < 3 gm/dl
- **Laboratory findings =** Proteinuria & hypoalbuminemia.

80. Initial fluid of choice for diarrhea in an infant is:

a) Salt water

b) Sugar water

c) ORS

d) Dextrose

Correct Answer - C
Ans: C. ORS
(Ref Ghai 8/e p293-294)

- Cornerstone of acute diarrhea management = Rehydration by using oral rehydration solutions (ORS).
- Initial fluid of choice for diarrhea in an infant = ORS.

81. A male child with coarse facial features, macroglossia, thick lips presents with copious mucous discharge from nose at 10 months of age. The child was absolutely normal at birth. On examination he was found to have enlarged Liver and Spleen. Diagnosis is:

a) Hurler's syndrome

b) Beckwith-Weidman syndrome

c) Hypothyroidism

d) Proteus syndrome

Correct Answer - A
Ans: A. Hurler's syndrome
(Ref Nelson 20/e p739)

- Likely diagnosis = Hurler's syndrome.

Hurler's syndrome:

- Type 1 mucopolysacharidoses.
- AR disorder.
- Characterized by alpha L-iduronidase deficiency resulting in accumulation of dermatan > heparin-sulfate.

Clinical features:

- Presents with Gortesque gargoyle facies:
- Coarse and heavy face.
- Enlarged head - Due to hydrocephalus caused by meningeal deposits.
- Low forehead & ears.

- Eyes wide set.
- Wide nose
- Poorly formed & widespread teeth.
- Open mouth, enlarged tongue & everted lips.
- Hepatosplenomegaly.
- Short neck.
- Thoracolumbur kyphosis.
- Gibbus deformity with motor delays.
- Flexion contracture of joints.
- Short stature.
- Genu valgum.
- Fiat feet.
- Broad-short hand, radially curved little finger.
- Carpal tunnel syndrome in children.

82. A 1-month-old child presented with conjugated bilirubinemia and intrahepatic cholestasis. On Liver biopsy staining with PAS, red colored granules were seen inside the hepatocytes. Probable diagnosis is:

a) Congenital hepatic fibrosis

b) Wilson's disease

c) Alpha-1 antitrypsin deficiency

d) Hemochromatosis

Correct Answer - C

Ans: C. Alpha-1 antitrypsin deficiency

(Ref Harrison 19/e 367-e2; Nelson 20/e p2052; Robbins. 9/e p815)

- Most probable diagnosis = Alpha-I Antitrypsin deficiency.

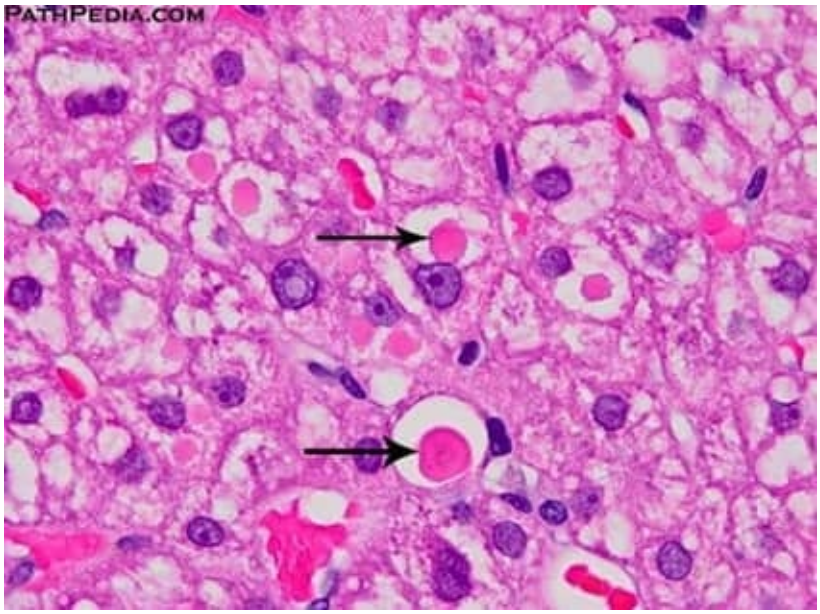
Alpha-I Antitrypsin deficiency:

Diagnosis:

- Confirmed by blood tests.
- Reduced levels of serum AAT & accompanied by Pi determinations.

Liver biopsy:

- Determines stage of hepatic fibrosis.
- Shows characteristic PAS-positive & diastase-resistant round- to-oval cytoplasmic globular inclusions inside hepatocytes of lobule periphery.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83. An infant at 7 months of age presented with history of vomiting and failure to thrive. Patient improved with administration of intravenous glucose and came out of coma within 24 hours. After one month he returned with similar complaints. On evaluation he is found to have raised blood ammonia and No ketones. Also, he has high urinary glutamine, alanine and uracil. Which is the likely enzyme defect is this patient?

a) Ornithine transcarbamoylase

b) CPS1

c) Arginase

d) Argininosuccinate lyase

Correct Answer - A
Ans: A. Ornithine transcarbamoylase
(Ref: Nelson 20/e p670, Harper 30/e p295. 356. 30/e p295, 356, 28/e p242-246)

- Most probable diagnosis - Urea cycle defect.
- Most common urea cycle defect - Due to deficiency of enzyme Ornithine transcarbamoylase.

Ornithine Transcarbamoylase (OTC) Deficiency:

- X-linked partially dominant disorder

- Hemiazygous males more severely affected than heterozygous females.
- MC form of all urea cycle disorders.
- OTC Gene mapped to the X chromosome (Xp21.1)0

Clinical Features:

Mild forms:

- Characteristically have episodic manifestations.
- Occur at any age (usually after infancy).
- Episodes of hyperammonemia - Manifested by vomiting & neurologic abnormalities (ataxia, mental confusion, agitation, combativeness & frank psychosis).
- Separated by periods of wellness.
- Usually occur after high-protein diet / due to catabolic state (infection).
- Hyperammonemic coma, cerebral edema & death may result.
- Gallstones seen survivors - Unclear mechanisms.

Diagnosis:

- Major laboratory finding during acute attack = **Hyperammonemia, increased plasma glutamine & alanine concentrations & lowered citrulline & arginine** levels.
- Decreased blood urea level.
- Increased urinary excretion of Orotic acid.
- Differentiates from Carbamoyl Phosphate Synthase deficiency.
- Orotate precipitates in urine as a pink colored gravel or stones.

84. In a child, surgery was done for biliary stricture with hepatojejunal anastomosis. Postoperative bilirubin level after 2 weeks was 6 mg/dL from a preoperative level 12mg/dL. The reason for this could be:

a) Normal lowering of bilirubin takes time

b) Anastomotic stricture

c) Delta bilirubin

d) Mistake in lab technique

Correct Answer - C
Ans: C. Delta bilirubin
(Ref Harrison 19/e p280, 18/e p325)
Delta Bilirubin:

- Albumin-linked bilirubin fraction (delta fraction/biliprotein)
- Is part of direct-reacting bilirubin fraction including conjugated bilirubin, covalently linked to albumin.
- Represents an important fraction of total serum bilirubin in patients with cholestasis & hepatobiliary disorders.
- Clearance rate of albumin-bound bilirubin from serum approximates half-life of albumin (12-14 days) rather than short half-life of bilirubin (4 hours).
- Due to tight albumin bonding.

85. A 7 years old child come with fever and tibabiabswelling exhibits on X-ray exhibits periosteal reaction Laboratory results shows raised ESR and TLC. Wath is the next step in diagnosis of the patient.

a) MRI

b) Pus culture

c) Bone biopsy

d) Blood culture

Correct Answer - A
Ans. a. MRI
(Ref Apleys 8/e pl'',().- Robbins 9/e p 8/e p475)

- **MRI** is essential to **elucidate soft-tissue involvement in Ewing's sarcoma, because the** tumor has **low signal intensity on Tl-weighted images** compared with the normal high signal intensity of the bone marrow.
- It is considered the **most useful imaging technique to evaluate suspected osteomyelitis** because of its **ability to demonstrate changes in the water content of bone marrow with an excellent structural definition and spatial resolution.**

86. Delayed neuronal migration and organization leads to certain disorders. Which of the following is the least likely possibility?

a) Lissencephaly

b) Schizencephaly

c) Polymicrogyria

d) Focal cortical dyplasia without balloon cells

Correct Answer - D
Ans. d. Focal cortical dyplasia without balloon cells (Ref http://enm kipedia.org/wiki/Neuronal migration_ disorder)

- Delayed neuronal migration and organization leads to certain disorders. Least likely possibility is focal cortical dyplasia without balloon cells

87. A 5 year-old child presents with non-utatiening macules and pappules on the skin. Skin biopsy revealed a perivascular IgA deposition. Which of the following is the most likely diagnosis?

a) Henoch Shonlein purpura

b) Wegener's granulomatosis

c) Kawasaki disease

d) Drug-induced vasculitis

Correct Answer - A
Ans. a. Henoch-Shonlein purpura (Ref Harrison 19/e p2100, 18/e p2797)

- Presence of non-blanching palpable purpura (vasculitis of skin) and colicky abdominal pain (vasculitis of gastrointestinal tract) in a five-year-old child together with evidence of IgA deposition in immune complexes suggests the diagnosis of Henoch Scholein Purpura

88. Which of the following conditions is worsened by prostaglandin E infusion?

a) Pulmonary atresia without VSD

b) Hypoplastic left heart syndrome

c) Obstructive TAPVC

d) Aortic arch interruption

Correct Answer - C
Ans. c.'Obstructive TAPVC { Rc/: Ll lllohttn Retlth,. Cunliut. Srr ('unliut surgen''4,,uar oJ rht' se,tinurs in Trnrucic and Cardiovascular Surgery 2003;4:271-276)
* Obstructive TAPVC is worsened by prostaglandin E infusion
* ln infants with or who have a clinical suspicion for a ductal dependent congenital heart defect, prostaglandin E, should be administered until a definitive diagnosis or treatment is established.
TAPVC
* Total anomalous pulmonary venous connection (TAPVC) is characterized by abnormal drainage of pulmonary veins into the right heart either by direct connection into the right atrium or into its tributaries.
* According to the site or level of connection of the pulmonary veins to the systemic venous system TAPVC has been classified into four types :-Infracardiac type of TAPVC is always obstructive whereas cardiac and supracardiac type may be obstructive or nonobstructive.
- Type I (most common: 45%) : Anomalous connection at supracardiac level (PV drains into left innominate vein or SVC)
- Type II (25%) : Anomalous connection at cardiac level (PV joins the coronary sinus or enter right atrium directly).

- Type III (25%) : Anomalous connection at infracardiac level (PV drain into portal vein).
- Type IV (5%) : Anomalous connection at multiple levels.

X-ray findings of TAPVC

- Cardiomegaly
- Plethoric lung fields
- Snowman or figure of '8' configuration - In supracardiac TAPVC.
- Ground glass appearance of lung - In obstructive TAPVC.

Clinical manifestations of TAPVC

1. Nonobstructive TAPVC - Patients presents with mild cyanosis and CHF at 6-8 weeks.
2. Obstructive TAPVC - Paients presents with severe cyanosis and CHF within first week.

- In supracardiac TAPVC the pulmonary veins join to form a single trunk (common pulmonary vein) which than drain through anomalous connection

89. A 2-year-old child with fever and barking cough for last 2 days presented to the pediatric emergency at 2.30 am. On examination, respiratory rate is 36/ min, temperature of 39 °C and stridor heard only on crying. No other abnormality is found. What is the next best step in management?

a) High-dose dexamethasone

b) Racemic epinephrine nebulization

c) Reassurance

d) Intravenous antibiotics

Correct Answer - A

Ans: A. High-dose dexamethasone

(Ref Ghai 8/e p376, 398: Nelson 20/e p2032-2034)

- Diagnostic of laryngotracheobronchitis or croup of mild severity.
- Hence, high-dose dexamethasone will be the treatment of choice.

Treatment:

Cornerstone of Treatment: Glucocorticoids & nebulized epinephrine°

Glucocorticoids:

- Useful in mild, moderate & severe croup.
- Dexamethasone is most effective corticosteroid.

Nebulized epinephrine:

- Useful in moderate to severe distress.

- By adrenergic stimulation causes,
- Constriction of precapillary arterioles.
- Decreases capillary hydrostatic pressure leading to fluid resorption from interstitium.
- Improvement in laryngeal mucosal edema.
- Antibiotics are not indicated
- Heliox (mixture of oxygen & helium) has low viscosity & low specific gravity.
- Allows for greater laminar airflow through respiratory tract.
- Considered in treatment of children with severe croup.

90. A 1-year old infant presents with 10-12 episodes of watery stools per day for the last 9 days. Along with zinc supplementation, what else should be prescribed to the child?

a) ORS with antibiotics

b) ORS only

c) ORS with low-lactose diet

d) ORS with low-lactose diet and probiotics

Correct Answer - B

Ans: B. ORS only

- The infant is suffering from acute diarrhea and treatment includes oral rehydration therapy, zinc supplementation and continued breastfeeding.
- Low lactose diet is required in management of persistent diarrhea.
- Antibiotics are required in management of dysentery, i.e. blood in stools.
- Routine use to probiotics in acute diarrhea is not recommended.

91. Milk is deficient in:

a) Iron and vitamin C

b) Iron and vitamin A

c) Phosphorus and vitamin A

d) Saturated fats

Correct Answer - A

Ans: A. Iron and vitamin C

- Human breast milk has enough of all nutrients except Vitamin D and Vitamin K. It is also slightly deficient in Vitamin C and iron.

Human vs Cow milk – nutritional value comparison:

- Human milk is richer in carbohydrate (lactose), iron & water content.
- Cow's milk is richer in fat, protein, calcium & energy content.

Human milk proteins:

- More cystine & taurine; less methionine; better digested than cow's milk proteins.

Human milk fats:

- Higher levels of PUFAs, esp., linoleic acid & linoleic acid; better digested and absorbed; low calcium content but better absorbed than cow's milk.
- Human milk is richer in Vitamin A & C; richer in copper, cobalt & selenium; richer in iron & higher bioavailability; high calcium/phosphorus ratio.
- Human milk has lesser sodium.

92. All of these are criteria for severe acute malnutrition in a 6-month-old child except:

a) Mid-upper arm circumference

b) Symmetrical edema

c) Weight for height

d) Height for age

Correct Answer - D

Ans: D. Height for age

- Height for age is not a criterion for severe acute malnutrition in a 6-month old child.

Severe acute malnutrition (SAM):

-]Among children 6-59 months of age is defined by World Health Organization (WHO) and UNICEF as any of the following:
- Weight-for-height below –3 standard deviation (SD or Z scores) of the median WI-10 growth reference
- Visible severe wastinge
- Presence of bipedal edema
- Mid-upper arm circumference below 11.5 cm
- This classification is used to identify children at high risk of death.

93. How is under-nutrition defined?

a) Weight for age < –2 SD

b) Weight for height < –2 SD

c) Weight for age < –3 SD

d) Weight for height < –3 SD

Correct Answer - B

Ans: B. Weight for height < –2 SD

Under nutrition is defined in terms of Weight for Height, i.e.

WHO Classification of Malnutrition

	Moderate malnutrition	S
Symmetrical edema	No	Yes (edematous malnutrition)
Weight-for-height	SD score from –2 to –3	SD score < –3 (severe wasting)
Height-for-age	SD score from –2 to –3	SD score < –3 (severe stunting)

94. A 3.5 kg male infant born at term after an uncomplicated pregnancy and delivery develops respiratory distress shortly after birth and requires mechanical ventilation. The chest radiograph reveals a normal cardiothymic silhouette but a diffuse ground glass appearance to the lung fields. Surfactant replacement fails to improve gas exchange. Over the first week life, the hypoxemia worsens. Results of routing culture and echocardiographic findings are negative. A term female sibling died at 1 month of age with respiratory distress. Which of the following is the most likely diagnosis?

a) Neonatal pulmonary alveolar proteinosis

b) Meconium aspiration

c) Total anomalous pulmonary venous return

d) Disseminated herpes simplex infection

Correct Answer - A
Ans: A. Neonatal pulmonary alveolar proteinosis
(Ref. Nelson 20/e p852, 2119)

- Suggestive of neonatal pulmonary alveolar proteinosis.

Pulmonary alveolar proteinosis:

- Disorder characterized by intra-alveolar accumulation of pulmonary surfactant.

Two clinically distinct forms of pulmonary alveolar proteinosis are seen:

- Fatal form: Presenting shortly after birth (congenital PAP)
- Gradually progressive form: Presenting in older infants & children.

Clinical manifestation:

- Immediately apparent in the newborn period & rapidly leads to respiratory failure.
- Clinically and radiographically indistinguishable from more common disorders of the newborn that lead to respiratory failure including pneumonia, generalized bacterial infection, respiratory distress syndrome and total anomalous pulmonary venous return with obstruction.

95. A 4 years old girl presented with abdominal lump. Bone scan is needed in:

a) Wilm's tumor

b) Neuroblastoma

c) Rhabdomyosarcoma

d) PNET

Correct Answer - B

Answer- B. Neuroblastoma

Bone scan is needed in Neuroblastoma as metaslasis is present in 60-70% of patients at the time of diagnosiso and MC site of metastasis in older children are bones.

MC presentation: Fixed, lobular mass extending from the flank toward the midlinea of the abdomen.

Most (80%) cases present before 4 years and peak incidence is 2 years of age.

Metastasis is present in 60-700/o of patients at the time of diagnosis MC site of metastasis in older children are bones (long bones >facial bones, skull particularly sphenoid), bone marrow and LN.

96. Children with germline retinoblastoma are more likely to develop other primary malignancies in their later lifetime course. Which of the following malignancy can occur in such patients?

a) Osteosarcoma of lower limbs

b) Thyroid carcinoma

c) Seminoma

d) Renal cell carcinoma

Correct Answer - A
Answer- A. Osteosarcoma of lower limbs
Children with germline retinoblastoma are more likely to develop other primary malignancies in their later lifetime course. Osteosarcoma of lower limbs can occur in such patients.
Retinoblastoma Syndrome (Primary site malignancy)- Familial Retinoblastoma

97. Children with germline retinoblastoma are more likely to develop other primary malignancies in their later lifetime course. Which of the following malignancy can occur in such patients?

a) Osteosarcoma

b) Renal cell carcinoma

c) Pinealoblastoma

d) Chondrosarcoma

Correct Answer - A
Answer- A. Osteosarcoma
Children with germline retinoblastoma are more likely to develop other primary malignancies in their later lifetime course, Osteosarcoma can occur in such patients.

98. A 3 days old baby is admitted with intraventricular hemorrhage. Baby develops abdominal distention. The X-ray abdomen showed pneumatosis portalis. Stage the necrotizing enterocolitis:

a) lb

b) 2a

c) 2b

d) 3a

Correct Answer - C
Answer- C. 2b
The severity, radiology and management of Necrotizing enterocolitis is best exemplified by the 'Modified Bell's staging
crileria'- According to 'Modified Bell's staging criteria'pneumatosis portalis (Presinie of gas in pofiai vein) is suggestive
of stage 2b.

99. To establish the diagnosis of H-type trachea-esophageal fistula, which if the following is required?

a) Chest X-ray

b) Tracheo-bronchoscopy

c) CT scan

d) Esophagoscopy

Correct Answer - B

Answer- B. Tracheo-bronchoscopy

Isolated tracheoesophageal fistula (TEF) (H-type fistula):

- Congenital isolated TEF (H-type) is a rare disorder posing diagnostic and management problems.
- H-type TEF is more frequent than H-type, owing to the oblique angle of the fistula from the trachea (carina or main bronchi) to the oesophagus, anatomically at the level of the neck root (c7-T1).
- Pressure changes between both structures can cause entry of air into the oesophagus, or entry of oesophageal content into the trachea.

100. Efficacy of phototherapy is not affected by-

a) Skin pigmentation

b) Type of light used

c) Spectral irradiation by incident light

d) Initial concentration of bilirubin

Correct Answer - A
Answer- A. Skin pigmentation
'Efficacy ofphototherapy depends upon irradiance, surface area exposed, distance from phototherapy unit, initial serum total bilirubin and adequacy of breastfeeding.'- Ghai 7/e pl50

101. A 6 weeks old baby presents with cough and cold for the last three days. Respiratory rate is 48/min. Patient is febrile, there are no chest retractions but wheezing is present. Which of the following statement is not true?

a) Antibiotics are not required

b) Child is suffering from pneumonia

c) Treat only wheezing

d) Treat only fever

Correct Answer - B

Answer- B. Child is suffering from pneumonia

Pneumonia:

- The World Health Organization has defined pneumonia in children clinically based on either a cough or difficulty breathing and a rapid respiratory rate, chest indrawing, or a decreased level of consciousness.
- A rapid respiratory rate is defined as >60 breaths/minute in children <2 months old, 50 breaths/minute in children 2 months to 1 year old, or >40 breaths/minute in children 1 to 5 years old.

102. A new born who has frothing of mouth. Cyanosis is present on day one. The most probable diagnosis is:

a) Lung hypoplasia

b) Lung cyst

c) Diaphragmatic hernia

d) Esophageal atresia

Correct Answer - D

Answer- D. Esophageal atresia

Esophageal atresia:

- The diagnosis of esophageal atresia is entertained in an infant with excessive salivation along with coughing or choking during the first oral feeding.

103. All of the following syndromes are associated with uniparental disomy except-

a) Prader-Willi syndrome

b) Russell-Silver syndrome

c) Bloom syndrome

d) Angelman syndrome

Correct Answer - C

Answer- C. Bloom syndrome

Bloom syndrome is not associated wilh uniparental disomy. Uniparental disomy is the term used when both chromosome of a pair of chromosome in a person with normal number of chromosome, have been inherited from only one parent.

104. A pediatrician in a district hospital with specialization neonatal care unit calls an ophthalmologist for consultation for which of the following?

a) A newborn with respiratory distress

b) A baby born at 28 weeks of gestation

c) Newborn with jaundice

d) A newborn with birth weight 2300 grams

Correct Answer - B
Answer- B. A baby born at 28 weeks of gestation
Premature baby needs to be screenedfor Retinopathy of prematurity.
All babies weighing <1500 gm or having a gestational period <32 weeks should be screened with indirect ophthalmoscopy between 32-36 weeks postconception.

105. A 2 year old premature neonate develops GTCS. What is the best investigation done to diagnose the pathology?

a) Transcranial ultrasound

b) CT Head

c) MRI brain

d) X-ray

Correct Answer - A

Answer- A. Transcranial ultrasound

Diagnostic Procedures:

- Polygraphic video-EEG recording ofsuspected events is probably mandatory for an incontrovertible seizure diagnosis.
- It is performed at the bedside and provides effective assessment of ventricular size and other fluid-containing lesions as well as effective viewing of haemorrhagic and ischaemic lesions and their evolution.
- CT brain scan is often of secondary or adjunctive importance to ultrasound. Last-generation CT brain scan images
- are of high resolution, can be generated within seconds and can accurately detect haemorrhage, infarction, gross
- malformations and ventricular and other pathological conditions.
- MRI is much superior for abnormalities of cortical development used for the detection of structural abnormalities
- such as malformations of cortical development, intracranial haemorrhage, hydrocephalus and cerebral infarction.

106. An 8 year old boy complaints of increasing muscle weakness. On examination, his calves are bulky and show muscle tightening. His serum creatine kinase levels are increasing with age. Which of the following is the most likely diagnosis?

a) Hereditary sensorimotor neuropathy

b) Myelin deficiency

c) Dystrophin deficiency

d) Congenital myopathy

Correct Answer - C
Answer- C. Dystrophin deficiency
Most likely diagnosis is duchenne Muscular Dystrophy due to Dystrophin deficiency.

107. A 6-week baby presents with cough and cold for the past 3 days. Respiratory rate is 48/min. On examination, patient is febrile with wheezing but no chest indrawing. Which of the following is not true?

a) Child has pneumonia

b) Antibiotics are not required

c) Wheezing to be treated

d) Fever to be treated

Correct Answer - A

Answer- A. Child has pneumonia

Respiratory rate is 48/min Patient is febrile, there are no chest retractions but wheezing is present. This child is not suffering from pneumonia, as respiratory rate is <60/ minute without chest indrawing. The child is most probably suffering from bronchiolitis.

108. A 6-year old girl presents with fever tor the past 5 days, generalized erythematous rash, strawberry tongue and cervical lymphadenopathy. The most likely diagnosis is: (Asked twice)

a) Kimura disease

b) Kawasaki disease

c) Scarlet fever

d) Rosie-Dorfman syndrome

Correct Answer - B
Answer- B. Kawasaki disease
The most likely diagnosis is Kawasaki disease.

109. A 6-year old girl presents with fever tor the past 5 days, generalized erythematous rash, strawberry tongue and cervical lymphadenopathy. The most likely diagnosis is:

a) Kimura disease

b) Kawasaki disease

c) Scarlet fever

d) Rosie-Dorfman syndrome

Correct Answer - B
Answer- B. Kawasaki disease
The most likely diagnosis is Kawasaki disease.

110. Which of the following is not an example of a syndrome caused by uniparental disomy?

a) Prader-Willi syndrome

b) Angelman syndrome

c) Russell-Silver syndrome

d) Bloom syndrome

Correct Answer - D

Answer- D. Bloom syndrome

- Uniparental disomy is the term used when both chromosome of a pair of chromosome is a person with normal number of chromosome, have been inherited from only one parent (Normally one chromosome of a pair is inherited from only one parent).
- The two chromosomes inherited from the same parent maybe identical (Uniparental isodisomy) or different (Uniparental heterodisomy).
- In Uniparental isodisomy, both chromosomes in the pair are identical; consequently the genes on both chromosomes are also identical.
- Angelman syndrome, Prader-Willi syndrome and Russell-Silver syndrome are ossociated with uniparental disomy.

111. A video of sick intubated neonate having bilateral jerks of both right and upper limbs with some occasional twitching of neck as well. Likely seizures:

a) **Focal** clonic

b) Multifocal clonic

c) Multifocal tonic clonic

d) Focal tonic

Correct Answer - B

Ans. b. Multifocal clonic

Morphology of Neonatal Seizures

- Subtle seizures: They are called subtle because the clinical manifestations are mild and are often missed. They are the commonest type, constituting about 50% of all seizures. Common examples of subtle seizures include:
- Ocular: Tonic horizontal deviation of eyes or sustained eye opening with ocular fixation or cycled fluttering
- Oral-facial-lingual movements: Chewing, tongue-thrusting, lip-smacking, etc.
- **Limb movements:** Cycling, paddling, boxing-jabs, etc.
- **Autonomic phenomena:** Tachycardia or bradycardia
  Apnea
- **Focal clonic seizures: They are rhythmic jerks of one arm, or one leg, or one arm and one leg on the same side of the body. They have both fast and slow components, occur with a frequency of 1-3 jerks per second, and are commonly associated with EEG changes.**

- **Multifocal clonic seizures:** Characterized by jerks in limbs on both sides of the body. May even involve neck or facial muscles in some cases. May show migration from one side to another or simultaneously occur at multiple sites. Often associated with significant asphyxia and may be seen in sick babies.
- **Tonic seizures:** This type refers to a sustained flexion or extension of axial or appendicular muscle groups. Not associated with EEG abnormalities and are rare.
- **Tonic-Clonic seizures are** rare to absent in neonates.

112. A child presented at 2 years of age with delayed motor development, mental retardation and finger biting. He was normal at birth. He subsequently develops cerebral palsy arthritis and dies due to renal failure at age of 25 years. What is the likely enzyme deficiency implicated?

a) Hexosaminidase deficiency

b) Adenosine deaminase deficiency

c) HGPRT deficiency

d) Ornithine transcarbamoylase deficiency

Correct Answer - C

Ans: C. HGPRT deficiency

(Ref: Nelson 20/e p746-747)

Lesch-Nyhan syndrome:

- Complete HGPRT deficiency characterized by hyperuricemia, self-mutilative behavior, choreoathetosis, spasticity & mental retardation.

HPRT gene:

- Located on X chromosome.
- Affected males - Hemizygous for mutant gene.
- Carrier females - Asymptomatic.

Partial HPRT deficiency:

- Kelley¬Seegmiller syndrome associated with hyperuricemia without CNS manifestations.

- Hyperuricemia - Due to urate overproduction à Causing uric acid crystalluria, nephrolithiasis, obstructive uropathy & gouty arthritis.
- Early diagnosis and appropriate therapy with allopurinol can prevent or eliminate all the problems attributable to hyperuricemia without affecting behavioral or neurologic abnormalities.

113. A neonate on routine examination at birth was found to have hepatomegaly. Rest of the examination was essentially unremarkable. On investigations, Anti-HCMV antibodies were found to be positive. What sequelae in later life is the child at risk of?

a) Renal failure

b) Mental retardation

c) Hepatic fibrosis

d) Sensorineural hearing loss

Correct Answer - D

Ans: D. Sensorineural hearing loss

(Ref: (Ghai 8/e p272 ; Nelson 20/e p 592-1594)

- Positive human cytomegalovirus (HCMV) antibodies at birth - Suggestive of congenital asymptomatic CMV infection.
- Symptomatic child at increased risk à Develops mental retardation.
- Asymptomatic child is at as high as 7% risk à Develop sensorineural hearing loss.

Congenital CMV infection:

Characteristic signs & symptoms:

- Clinically manifested infections - Intrauterine growth restriction, prematurity, hepatosplenomegaly, jaundice, blueberry muffin—like rash, thrombocytopenia and purpura, microcephaly & intracranial calcifications.

Neurologic problems:

- Chorioretinitis, sensorineural hearing loss & mild increases in cerebrospinal fluid protein.

114. Palivizumab is a humanized monoclonal antibody. For which of the following conditions has it been approved for?

a) Avian influenzae

b) Avian influenzae

c) Respiratory syncytial virus

d) Coxsackie virus

Correct Answer - C

Ans: C. Respiratory syncytial virus

(Ref: Nelson 20/e p1608)

Palivizumab:

- Monoclonal antibody.
- Used in prevention & treatment of respiratory syncytial virus (RSV) infections.
- Recommended for high-risk infants 24 months of age.
- Due to prematurity or other medical problems like congenital heart disease.

115. A 2 months old child was brought to the subcenter by his mother with complaints of fever for two days. Weight of the child is 2 kg. On examination, the child is restless and irritable, skin pinch went back in 2 seconds, oral mucosa is dry and eyes were sunken. There were ten pustules on his forehead. What should be done at the subcenter?

a) Refer to higher center with mother giving frequent sips of ORS.

b) Immediately admit the child, give IV fluids and then refer to higher center.

c) Give first dose of antibiotic and refer to higher center in an ambulance with sips of ORS along the way.

d) Send child home with few packets of ORS and call after 3 days.

Correct Answer - C

Ans: C. Give first dose of antibiotic and refer to higher center in an ambulance with sips of ORS along the way.

(Ref .Ghai 8th/e p752-753: IMNCI Guidelines).

- Multiple signs of possible serious bacterial infection (10 skin pustules, diarrhea with severe dehydration & weight = 2 kg which is <-3SD).
- Hence, child should be immediately given first dose of injectable antibiotic, kept warm & referred to hospital with sips of ORS along the way

116. A 6 days old neonate weighing 2800 gm (birth weight 3200 gm) was brought with the complaints of fever, poor feeding and poor activity. There was no history of vomiting or diarrhea. Axillary temperature was 39°C with depressed fontenalle, sunken eyes, decreased urine output and decreased skin turgor. Her mother has the history of decreased milk production. What is your diagnosis?

a) Neonatal sepsis

b) Galactosemia

c) Fever & dehydration

d) Acute renal failure

Correct Answer - A

Ans: A. Neonatal sepsis

Initial Signs & Symptoms of Infection in Newborn Infants

General

- Fever, temperature instability°
- Not doing well, poor feeding°
- Edema°

Cardiovascular System

- Pallor, mottling, cold clammy skin°
- Hypotension, tachycardia°
- Bradycardia

Gastrointestinal System

Central Nervous System

- Abdominal distention°
- Vomiting, diarrhea°
- Hepatomegaly

Respiratory System

- **Apnea, dyspnea, tachypnea°**
- **Retractions, flaring, grunting°**
- **Cyanosis°**

- Irritability, lethargy, high pitched cry
- Tremors, seizures°
- Hyporeflexia, hypotonia, abnormal Moro's reflex°

Hematological System

- **Pallor, jaundice, splenomegaly°**
- Bleeding
- Petechiae, purpura

117. A neonate presented with cicatrizing skin lesions all over the body with hypoplasia of all limbs. An MRI of the brain revealed diffuse cerebral atrophy. An ophthalmologic evaluation reveals chorioretinitis. Which of these tests is most likely to show a positive result in this patient?

a) Anti-HCMV antibodies

b) Anti-toxoplasma antibodies

c) Anti-VZV antibody

d) Anti-rubella antibody

Correct Answer - C

<u>Ans: C. Anti-VZV antibody</u>

(Ref Ghai 8/e p2215).

- Suggestive of congenital varicella infections detected using Anti-VZV antibodies.

Congenital varicella syndrome:

- Characterized by cicatricial skin scarring in a zoster-like distribution, limb hypoplasia, and neurologic (e.g., microcephaly, cortical atrophy, seizures, and mental retardation), eye (e.g., chorioretinitis, microphthalmia, and cataracts), renal (e.g., hydroureter and hydrottephrosis) and autonomic nervous system abnormalities (neurogenic bladder, swallowing dysfunction, and aspiration pneumonia).

118. What should be the ideal temperature in delivery room for the neonates to be kept in warmer?

a) 22-26°C

b) 28-30°C

c) 30-35°C

d) 37°C

Correct Answer - A

Ans: A. 22-26°C

(Ref Ghai 8/e p133).

Nursery temperature:

- Ideal temperature in delivery room for neonates in warmer state - 22-26°C (72-78"F).
- Should be free from drill of air.
- Ventilation system for each delivery & resuscitation room designed to control ambient temperature between 72-78 degrees Fahrenheit (22-26 degrees Centigrade) during delivery, resuscitation & stabilization of newborn.

119. Ponderal index is:

a) Square root of height in feet by weight in grams

b) Weight in kilograms by cube of height in meters

c) Mid-upper arm circumference to head circumference ratio

d) Head circumference to abdominal circumference ratio

Correct Answer - B

Ans: B. Weight in kilograms by cube of height in meters

(Ref Dafiary Manual of Obstetrics 3/e p199)

Ponderal Index:

- Calculated by multiplying weight in grams by hundred and then dividing by cube of length in cm.
- Ponderal index = Birth weight (gm)/ Length (cm)3 x 100

Interpretation:

- Usually less than 2 in asymmetric growth retardation.
- 2 or more in babies with normal growth or symmetric growth retardation.

120. Ideal route of drug delivery in neonatal resuscitation is:

a) Intraosseous

b) Through umbilical vein

c) Through peripheral vein

d) Through umbilical artery

Correct Answer - B

Ans: B. Through umbilical vein

*(*Ref: Ghai 8/e p132).

- Umbilical vein - Preferred route for drug delivery during resuscitation.
- Due to ease of approach.
- Veins in scalp or extremities are difficult to access during resuscitation.
- For umbilical vein catheterization, 3.5 Fr or 5 Fr umbilical catheter inserted into umbilical vein such that its tip is just inside the skin surface and there is free flow of blood.
- Direct injection into umbilical cord is not desirable.
- No intracardiac injection recommended.

121. A neonate presented with jaundice on first day of life. His mother's blood group is `0' positive. How will you manage this patient?

a) Observe only as it is mostly physiological jaundice

b) Exchange transfusion

c) Liver function tests and liver biopsy as it is mostly due to cholestasis

d) Phototherapy

Correct Answer - D

Ans: D. Phototherapy

(Ref: Nelson 20/e p873)

- Jaundice on day 1 of life - Due to some hemolytic disease or congenital infections.
- Treated using phototherapy.
- Possibility of Rh incompatibility ruled out as mother blood group is 0+.
- Ideally basic investigations for hemolytic anemia should be sent simultaneously (not given in the option).
- Best option would be phototherapy.

122. What is the recommended dose of steroids for attaining fetal lung maturity?

a) Inj. betamethasone 12 mg for 2 doses 12 hours apart

b) Inj. betamethasone 12 mg for 2 doses 24 hours apart

c) Inj. dexamethasone 6 mg for 4 doses 24 hours apart

d) Inj. dexamethasone 12 mg for 2 doses 12 hours apart

Correct Answer - B

Ans: B. Inj. betamethasone 12 mg for 2 doses 24 hours apart

(Ref: Goodman Gillman 12Ic p1231).

- Betamethasone (12 mg intramuscularly every 24 hours for two doses) dexamethasone (6 mg intramuscularly every 12 hours for four doses) administered to women with definitive signs of premature labor between 26 and 34 weeks of gestation.
- For attaining lung maturity.

Antenatal glucocorticoids:

- Used frequently in premature labor setting à Decreases respiratory distress syndrome incidence, intraventricular hemorrhage & death in infants delivered prematurely.

123. In a child with tetralogy of Fallot with fever and diarrhea, which of the following is the surest sign of a cyanotic spell?

a) Hepatomegaly

b) Absence of murmur

c) S3 gallop rhythm

d) Arterial oxygen saturation of less than 75%

Correct Answer - B

Ans: B: Absence of murmur

Explanation:

(Ref: Nelson 20Ie p2212; Ghai 8/e p422)

- During cyanotic spell, temporary disappearance or a decrease in intensity of the systolic murmur is usual as flow across the right ventricular outflow tract diminishes.
- Paroxysmal hypercyanotic attacks (hypoxic, "blue," or "tet" spells) are a particular problem during the 1st 2 years of life.
- The infant becomes hyperpneic and restless, cyanosis increases, gasping respirations ensue, and syncope may follow.
- Most frequently in morning on initially awakening or after episodes of vigorous crying.

124. A mother comes with her 3 months' child asking the physician if she can give cereals to her child. What problems can this lead to her child?

a) Allergy due to the food content

b) Risk of gastrointestinal infection

c) Retarded oro-motor development

d) Contaminated food leading to reflux

Correct Answer - A

Ans: A: Allergy due to the food content

Explanation:

(Ref: Nelson 20/e p1139; Ghni 8/e p90)

- Babies should receive only breast milk or infant formula for the first 6 months of life.
- Most important reason for this is allergy due to the food content.
- Exclusive breastfeeding for the first 4-6 months of life may reduce allergic disorders in the first few years of life. Potentially allergenic foods (eggs, milk, wheat, soy, peanut and fish) should be introduced after this period of exclusive breastfeeding to decrease chances of food allergy.

125. A child who was normal at birth develops chronic liver failure and muscle weakness at 3 months of age. On investigations, serum glucose is low, along with ketoacidosis and decreased pH. ALT and AST are raised. Blood lactate and uric acid levels are normal. Intravenous glucagon given after meals raises the blood glucose levels, but does not raise glucose when given after an overnight fast. Liver biopsy shows increased glycogen in liver. Which is the enzyme likely to be defective in this child?

a) Glucose-6-phosphatase

b) Muscle phosphorylase

c) Branching enzyme

d) Debranching enzyme

Correct Answer - D
Ans: D: Debranching enzyme
(Ref: Nelson 20/e p717-720; Harrison 19/e p433 e-2, 18Ie p3200, 3201)
In this child, a combination of liver and muscle involvement with

ketoacidosis and raised liver enzymes points towards Type III glycogen storage disease, i.e. Cori's disease caused by deficiency of debranching enzyme.

Type IIIa Glycogen Storage Disease or Cori's Disease or Forbes Disease:

- Due to deficiency ofglycogen debranching enzyme activity.
- Debranching enzyme, together with phosphorylase, is responsible for complete degradation of glycogen. When debranching enzyme is defective, glycogen breakdown is incomplete and an abnormal glycogen with short outer branch chains and resembling limit dextrin accumulates.
- Deficiency of glycogen debranching enzyme causes hepatomegaly, hypoglycemia, short stature, variable skeletal myopathy & variable eardiomyopathy.

126. A 5 years old child presented with continuous fever and features of sepsis with a BP of 90/60 mm Hg, Pulse rate 144/min and respiratory rate of 30/min. What is the initial fluid of choice for management?

a) 10 mL/kg of 10% dextrose

b) 10 mL/kg of hydroxyethyl starch

c) 20 mL/kg of 0.45% normal saline

d) 20 mL/kg of 0.9% normal saline

Correct Answer - D

Ans: D : 20 mL/kg of 0.9% normal saline

Explanation:

(Ref: Ghai 8Ie p718)

- In hypovolemic or septicemic shock, replacement of intravascular volume by isotonic fluids is the main stay of treatment.
- Hence, normal saline resuscitation with 20 ml/kg boluses.

127. A 6 years male child comes with complaints of bedwetting. The child is continent during the day and problem is only at night. Growth and development of the child were normal. Urine microscopy is normal and urine specific gravity was 1.020. How will you manage?

a) Reassure the parents and follow up after 6 months

b) Refer to psychiatrist

c) Complete blood counts

d) Ultrasound-KUB

Correct Answer - A

Ans: A: Reassure the parents and follow up after 6 months

Explanation:

(Ref: Nelson 20Ie p2585; Ghai 8/e p504)

- Bed-wetting is normal till 5 years of age.
- In a child with only night-time bed wetting, when urinalysis (to rule out infections) and urine osmolality (to rule out diabetes) are normal, only regular follow up is required.

Treatment:

- To reassure the child and parents that the condition is self-limited and to avoid punitive measures that can affect the child's psychologic development adversely.
- Fluid intake should be restricted to 2 oz after 6 or 7 pm.
- Parents should be certain that the child voids at bedtime.
- Avoiding extraneous sugar and caffeine after 4 pm also is beneficial.

- If the child snores and the adenoids are enlarged, referral to an otolarvngologist should be considered, because adenoidectomv can cure the enuresis.

128. All of the following are sequelae of fetal alcohol syndrome except:

a) Macrocephaly

b) Holoprosencephaly

c) Microcephaly

d) Thinning of corpus callosum

Correct Answer - A
Answer--A. Macrocephaly
Fetal alcohol syntlrome is ussociated with microcephuly rather lhan macntcephaly. Holoprosencephaly can occur in extreme forms of fetal alcohol syndrome along with midline hypoplasia.
"Fetal Alcohol Syndrome (FAS) No single malformation or characteristic malformation complex has been described in few cases of FAS in wihch neuropathology has been reported. The neuropathologic changes are varied and nonspecitic and include microcephaly, hydrocephalus, leptomeningeal, white matter and periventricalar neuroglial heterotopias, agenesis of corpas callosum and the cerebellar vermis, incomplete holoprosencephaly, and neural tube defects

129. Which of the following is true regarding congenital CMV infection?

a) Diagnosed only by persistent presence of IgM antibody after 6 months

b) It is the most common cause on nonsyndromic sensory neural hearing loss

c) All babies born are symptomatic

d) Mothers of developing countries who transmit the virus are usually symptomatic

Correct Answer - B

Answer- B. It is the most common cause on nonsyndromic sensory neural hearing loss

Asymptomatic congenital CMV infection is likely a leading cause of sensorineural hearing loss, which occurs in approximately 7-10% of oll infants with congenital CMV infection, whether symptomatic at birth or not. The definitive method for diagnosis is virus isolalion or PCR and not IgM antibody detection, which is non-specific and may be due to acquired CMV infection after birth.

130. A 5 years old child brought to the hospital with history of loose stools but no history of fever or blood in stools. Mother says he is irritable and drinks water hastily when given. On examination eyes are sunken and in skin pinch test, the skin retracted within two seconds but not immediately. What is the treatment for this child?

a) Administer the first dose of IV antibiotic and immediately refer to hither center

b) Give oral fluids and ask the mother to continue the same and visit again next day

c) Consider severe dehydration, start IV fluids, IV antibiotics and refer to higher center

d) Give Zinc supplementation and oral rehydration solution only and ask mother to come back if some danger signs develop

Correct Answer - D

Answer- D. Give Zinc supplementation and oral rehydration solution only and ask mother to come back if some danger signs develop

The child in this given scenario is having some dehydration, as the child is restless and irritable, drink water readily
and skin pinch goes back slowly (< 2 seconds) with sunken eyes.
Treatment includes oral rehydration therapy, zinc
supplementation and continued breastfeeding according to the WHO

supplementation and continued breastfeeding according to the WHO IMNCI protocol plan B.

131. An 18 months child weighing 11.5 kg comes to the PHC with fever and respiratory difficulty. On examination, the child is lethargic, with a respiratory rate of 46 bpm and no chest retractions. What is the most appropriate management of this child?

a) Prescribe oral antibiotics, warn of danger signs and send home

b) Intravenous fluids alone

c) Intravenous antibiotics and observation

d) Give intravenous antibiotics and refer to a higher center

Correct Answer - A

Answer- A. Prescribe oral antibiotics, warn of danger signs and send home

This child is having fast breathing (respiratory rate >46/minute) without danger signs like lower chest wall indrawing or stridor. Hence, the child will be classified to have pneumonia (non-severe). So, the child should be prescribed appropriate antibiotic and advise mother about supportive measures and when to return for follow-up.

132. A child presents to the emergency with a history of ingestion of button battery, on X-ray it was found in the stomach or duodenum. What is the next step?

a) Endoscopic removal of battery

b) Wait and watch

c) Repeat X-ray after 5 days

d) Immediate laparotomy

Correct Answer - B
Ans: B. Wait and watch
Ref: NBIH Button bsttery ingestion triage and treatment guideline.

- The management of a Button battery ingestion depends upon the anatomical site of the impacted battery.
- Button batteries that have cleared the stomach usually pass through the gastrointestinal tract within one week without complications.
- Follow-up radiographs should be performed in asymptomatic patients who have not passed the battery by 10 to l4 days, regardless ofsize or earlier if patient becomes symptomatic.

133. Absence of which of the following milestone in 3 yr old chitd is called delayed development?

a) Hopping on one leg

b) Drawing a square

c) Feeding by spoon

d) Passing a ball to someone

Correct Answer - C
Ans: C. Feeding by spoon
Ref: Ghai Essentiul Pediatrics, 8''' ed., pg. 49

- Hopping on one leg - Should be attained by 4 years ofage
- Drawing a square - Should be attained by 4Il2 years of age
- Feeding by spoon Should be attained by l8 months of age
- Passing a ball to someone- Should be attained by 3 years

134. After the delivery of an infant of diabetic mother, blood glucose of the infant was 60 mg/dt. Which other investigation docs the sister expects that the physician would ask her to do?

a) Serum potassium

b) CBC

c) Serum calcium

d) Serum chloride

Correct Answer - C
Ans: C. Serum calcium
Ref Ghai Essential Pedistrics 8,h ed, pg. 181 und Nelson Textbook of Pediatrics 20h ed" pg. 897
Infants of a diabetic mother are ata higher risk of metabolic complications as compared to normal infants.
These complications include:
* Hypoglycemia
* Hypocalcemia
* Hypomagnesemia
* Hence the infant needs to be checked for these as soon as possible.

135. An un-immunized 13 months old child comes to you in OPD, according to the latest immunizations schedule, what vaccines will you advise??

a) OPV 3 doses, I lPV 3 Pentavalent and I measles

b) BCG, OPV 3 doses, 3 lPV, 3 Pentavalent and I measles

c) OPV 3 doses, I IPV 3 Pentavalent and 2 measles

d) OPV 3 doses, 3 IPV 3 DPI : Hep-B

Correct Answer - D
Ans: D. OPV 3 doses, 3 IPV 3 DPI : Hep-B
Ref:https://mohfw.gouin/sites/default/files/245453521061489663573.pd ip. o rg/file s/I A P-imm unizution-sc he du le- 2 0 I 6- I P- 2 0 I 6-E p u b. p df
* The latest schedule of immunization under NIS is as follows:
* vNational Immunization Schedule (NIS) for Infants, children and Presnant women –
- OPV 3 doses, 3 IPV 3 DPI : Hep-B

136. Correct order ofsuctioning during neonatal resuscitation is?

a) Mouth-Nose

b) Nose-Mouth

c) Mouth-Nose-Trachea

d) Trachea-Nose-Mouth

Correct Answer - A
Ans: A. Mouth-Nose
Ref: Ghai Essential Pediatrics, S't' ed., pg. 127

- At the time of birth, if the baby is:
- Not of term gestatton
- Not breathing or crying
- Not having good tone.
- Then the baby requires resuscitation

137. How long should a child be isolated after being diagnosed with bacterial meningitis to prevent further transmission?

a) Till 24 hours after starting antibiotics

b) Till cultures become negative

c) Till antibiotic course is complete

d) Till l2hrs alter admission

Correct Answer - A
Ans: A. Till 24 hours after starting antibiotics
Ref: Ghai Essentisl Pediatrics, 8't' ed,, pg. 565' *https://www.cdc.gov*
Prevention of transmission:

- Droplet precautions for the first 24 hours of antimicrobial therapy is sufficient

138. You have been called to declare a brain dead l2-year-ord chitd in pICU, all ofthe given are signs of brain death except?

a) Normal BP without pharmacological support

b) Positive spinal reflexes on stimulation

c) Sweating and tachycardia

d) Decorticate and decerebrate posturing

Correct Answer - D
Ans: D. Decorticate and decerebrate posturing
Ref Dhanwate AD. Brainstem death: A comprehensive review in Indian perspective' [ndian Journal o/ Critical Care Medicine : Peer-reviev,ed, o.fficiat pul.tlicalion of Indian Society of Critical Care Medicine. 2014;18(9):596-605. doi:10.4103/0972-5229.140151. Goila AK, Pawar M. The diagnosis of brain death. Indian Jrurnal of Critical Care Medicine; Peer-reviewed, o.fficial publication of Indian Society of Critical Corc Madicine. 2009;I 3(I):7-11. doi: I 0.4103/0972-5229.53 I0t.
Diagnosis of brain death in India:
Who should diagnose:

- Team of four medical experts including
- Medical Administrator In charge of the hospital.
- AuthorizedSpecialist
- Authorized Neurologist/Neuro-Surgeon
- Medical Officer treating the patient.

139. A 2-year-old child with history of fall one year back with parietal bone fracture now presented with painful and growing parietal swelling.

a) Growing scalp hematoma

b) Growing fracture

c) Subdural hygroma

d) Chronic abscess

Correct Answer - B
Ans: B. Growing fracture
Ref: Growing skull fractures: classiJication and management. Naim-UFRahman etsl. Br J Neurosurg. (1994)
Growing Fractures:

- Also known as traumatic encephaloceles or leptomeningeal cysts
- Skull fractures associated with an underlying dural tear may fail to heal properly.

140. Craniopagus is defined as fusion of:

a) Head and spine

b) Head only

c) Thorax and spine

d) Thorax only

Correct Answer - B
Ans: B. Head only
Ref: Coran Pediatric Surgery 7u ed., pg. 1728
Craniopagus:

- **Extent of union:**Cranial neuropore
- Skull venous sinus &meninges l00%
- Cerebral cortex 37%

141. An adolescent school girl complaints of dropping objects from hands, it gets precipitated during morning and during exams. There is no history of loss of consciousness and her cousin sister has been diagnosed with epilepsy. EEG was done and was suggestive of epileptic spikes. What is the diagnosis?

a) Juvenile myoclonic epilepsy

b) Atypical absence

c) Choreo-athetoid epilepsy

d) Centrotemporal spikes

Correct Answer - A
Ans: A. Juvenile myoclonic epilepsy
Ref: Nelson Textbook of Pediatrics, 20' ed., pg. 2836
Juvenile myoclonic epilepsy (Janz syndrome):

- Starts in early adolescence with 1 or more of the following manifestations:
- Myoclonic jerks in the morning, often causing the patient to drop things;
- Generalized tonic–clonic or clonic–tonic–clonic seizures upon awakening; and
- Juvenile absences.
- Sleep deprivation, alcohol (in older patients), and photic stimulation, or, rarely, certain cognitive activities (such as exams) can act as

precipitants.

142. A child can walk upstairs one step at a time, can ride cycle but can't jump can also speak sentences, can tell his/her name gender but finds difficult to narrate the story.What is her actual developmental age

a) 1 yrs

b) 2

c) 3

d) 4

Correct Answer - B

Ans. b) 2years

GROSS MOTOR MILESTONES:

Age	**Milestone**
3 months	Neck holding
5 months	Rolls over
6months	Sitting supported
8months	Sitting without support
9months	Stands with support
12months	Stands without support, Walks but falls
15months	Walks alone, Creeps upstairs
18months	Runs, explores drawers
2 years	Walks upstairs (baby steps), Jumps
3 years	Walks upstairs (alternate feet), rides tricycle
4 years	Hops on one foot, walks downstairs (alternate feet)

4 years	Hops on one foot, walks downstairs (alternate feet)

143. Video based question -
AIIMS NICU - neonate shown on O2 with na
intubated, sister shown inserting a tube thr
camera zooms in, 18 (calibration) mark see
end as of now not connected to anything, v
the procedure that is being done?
VIDEO LINK:
https://media.giphy.com/media/ln0aVCWqP

a) Oral suction

b) Oropharyngeal suction

c) Nasogastric tube insert

d) Orogastric tube insertion

Correct Answer - D
Ans. D. Orogastric tube insertion

144. Pediatric patient presented with 45 mins h/o continuous convulsions.
CASE 1: SR told to give iv lorazepam but J.R cant secured iv line. Then what he has given?

a) Rectal diazepam

b) Inhalation Phenobarbital

c) IV carbamazepine

d) Subcutaneous midazolam

Correct Answer - A
Answer A. Rectal diazepam

- Benzodiazepines are Drug of Choice and in pediatric patients rectal route should be preffered.

Reference - https://www.uspharmacist.com/article/emergent-treatment-of-status-epilepticus-in-children

145. CASE -2 SR visit again but the condition is not improved but this time IV cannula was set. What drug should be given now?

a) Midazolam

b) I/V Phenobarbital

c) Oral valproate

d) IV carbamazepine

Correct Answer - B
Ans. In this case the First Line has failed . So for second line therapy I/V Phenobarbital is preffered
Reference - https://www.uspharmacist.com/article/emergent-treatment-of-status-epilepticus-in-children

146. Sequential arrangement of fetal scans -

a) Growth scan

b) Triple marker

c) Anomalous Scan and NT scan

d) All

Correct Answer - D
Ans: D. All
The **NT scan** must be **done** between 11 and 14 **weeks** pregnant, because this is when the base of baby's neck is still transparent. (The last day **for scan is** 13 **weeks** and 6 days pregnant.)
Triple marker test is performed in pregnant women at the end of first trimester and the beginning of the second trimester.
The **anomaly scan**, also sometimes called the anatomy scan, 20 week ultrasound, or level 2 ultrasound, is a pregnancy ultrasound performed between 18–22 weeks
A **growth scan** is an ultrasound **scan** that determines whether your baby's **growth** is normal. Doctors typically recommend it for women during the third trimester of pregnancy; one of the reasons it is also a fetal **growth scan between 28 weeks and 32 weeks of pregnancy.**

147. 4yr boy absence of right testes, diagnostic laparoscopy done, a blind end vessel seen what to do next?

a) Open laprotomy

b) Nothing to be done

c) Scrotal approach

d) Inguinal approach

Correct Answer - D

Ans. D. Inguinal approach

- Cryptorchidism is the most common congenital abnormality of the genitourinary tract.
- Cryptorchidism means hidden testis.
- An absent testis may be due to agenesis or atrophy secondary to intrauterine vascular compromise also known as the "vanishing testis syndrome".
- Bilaterally absent testes is anorchia which is 10% cases.
- More common on Right Side.
- Complications of undescended testes
- Torsion can be seen in incomplete testicular descent
- Sterility is seen in bilateral cases (especially intra-abdominal testes)
- Incomplete testicular descent predisposes to malignant disease; cancer is more common in an incompletely descended testes-orchidopexy may or may not diminish the risk.
- Atrophy of an inguinal testes before puberty may possibly be caused by recurrent minor trauma.
- In patients with a unilateral nonpalpable testis, a descended testis that is larger than expected suggests an atrophic undescended

testis; confirmation requires surgical intervention typically via diagnostic laparoscopy to seek an intra-abdominal testis or confirm testicular agenesis. However, scrotal or inguinal exploration is sometimes done if a testicular remnant distal to the internal inguinal ring is suspected.

148. Arrange the following in the sequence of closure :

a) Umbilical Artery

b) Umbilical Vein

c) Ductus venosus and Foramen ovale

d) All

Correct Answer - D

Ans. D. All

Changes After Birth: Closing of Shunts

Shunt	Functional closure	Anatomical closure	Remnant
Ductus arteriosus	10 – 96 hrs after birth	2 – 3 wks after birth	Ligamentum arteriosum
Formamen ovale	Within several mins after birth	One year after birth	Fossa ovalis
Ductus venosus	Within several mins after birth	3 – 7 days after birth	Ligamentum venosum

Umbilical arteries → Umbilical ligaments

Umbilical vein → Ligamentum teres

149. Which of the following is considered as high risk infant?

a) Working mother

b) Antenatal preeclampsia

c) Third child

d) Twins

Correct Answer - A
Ans. A. Working mother
Identification of 'At Risk' Infants
* Birth weight < 2.5 Kg
* Birth order 5 or more
* Artificial feeding
* Weight below 70% of expected weight (i.e. grade II & III malnutrition)
* Failure to gain weight during 3 successive months
* Children with PEM, Diarrhea.
* Working mother, one parent.
* Spacing of less than 1 year.

150. In CPR for Infants-

1. The sequence followed is Compression – Breathing- Airway
2. 30 chest compression is given
3. 1 breathe every 15 compressions
4. Chest Compression should be 1 ½ inches in infants
5. In infants, the brachial pulse should be assessed.

a) True False False True False

b) True False False True True

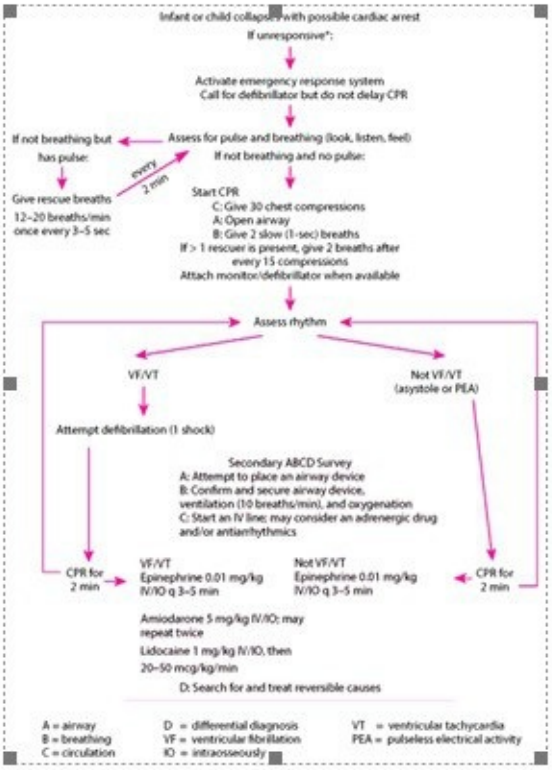
c) False True False True False

d) False True False True True

Correct Answer - D
Ans. D. False True False True True
Emergency care providers should check the victim's pulse for at least 5 seconds but no longer than 10 seconds. For children aged one to adolescence, the pulse should be checked at the carotid artery. In infants, the brachial pulse should be assessed.
If there is a palpable pulse within 10 seconds, then a rescue breath should be given every 3 seconds. Breaths should last one second and the chest should be observed for visible rise. If the victim has an advanced airway, then the provider should administer breaths 10-12 times per minute.
If the pulse is less than 60/minute, or if the victim has signs of poor

perfusion after adequate ventilation and oxygenation, the provider should begin chest compressions. In the absence of a pulse, a lone rescuer should begin CPR with 30 high quality compressions followed by two breaths. If two healthcare providers are available, the cycle of compressions to breaths should be 15:2 (pediatrics). High quality compressions in CPR should be a minimum of 1/3 the AP diameter of the chest, or approximately 1 ½ inches in infants (4 cm) and 2” in children from age one to adolescence. The rate of compressions should be 100-120 per minute. Chest recoil should be complete between compressions.

Pediatric compression is performed with the head of one hand over the lower ½ of the sternum, between the nipples. In infants, use two fingers, or use the thumb encircling technique if multiple providers are available



151. 6 yr old child H/O fever 5 days back which is now afebrile with normal muscle mass, tone and reflex, no neurological deficit but pain on palpating muscles and CPK levels 2000, diagnosis is

a) GBS

b) Dermatomyositis

c) DMD

d) Acute viral myositis

Correct Answer - D

Ans.D. Acute viral myositis

The clinical picture suggests the most probable diagnosis is Viral myositis. The specific history of URI suggests influenza myositis. Infectious myositis has a male predominance and are typically seen in young adults.

The typical presentation of the childhood form includes fever, malaise, and rhinorrhea which is usually followed in 1-7 days by severe pain, especially in the calves. The muscle pain is usually worse with movement and the symptoms last for about a week. Muscle weakness, tenderness, and swelling are usually seen with more severity in adults and the Proximal muscles are affected predominantly.

In children, toe walking and wide-based gait may be seen because of the involvement of the gastrocnemius-soleus muscles.

Lab features of influenza myositis include

Elevated CK may be as high as 500 times normal

Urine myoglobin is usually positive