Drugs

- Platelet function defect (thrombathenia)
    - Hereditary
    - Acquired (Drugs e.g. aspirin)
    - renal failure

- What are the complications of immune thrombocytopenic purpura?
- What are the complications of treatment lines?
- What are the treatment lines?
- What are the types of ITP?
-WHAT are the testes of assessment of platelets?

Abdominal cases

History

- Personal history

Age

Veno-occlusive disease (1-4 years)
Metabolic liver disease (below 1 yr)
Bilharziasis usually in late childhood

Residence

Bilharziasis is endemic in certain areas

- Complaint

- Abdominal distention (usually)
- Hematemesis
- Jaundice
- Abdominal pain

- Present history

1- Complaint analysis (onset- course-duration)

If Hematemesis (denotes portal hypertension)

Number of attacks
Amount of bleeding
Color of blood.....
Bleeding from other sites

Need for blood transfusion

Jaundice (denotes chronic hepatitis or liver cell failure)

Color of urine and stool

Abdominal distention (denotes organomegaly or ascites)

Onset, course and duration

If abdominal pain (denotes organomegaly)

Site of pain, nature, what increase, what decrease and radiation

Other Q;

Bilharziasis

- Bleeding per rectum or terminal hematuria
- Contact with channel water
- Intake of antibilharzial treatment
- Residence (area endemic for Bilharziasis)

Cirrhosis

- Jaundice - color of urine and stool
- History of previous blood transfusion
- Bleeding tendency and encephalopathy

Veno-occlusive disease

- Course of abdominal distention (rapidly filling ascites)

Metabolic liver disease

- Infancy

History SUGESSTIVE of Metabolic disease

- Convulsions, vomiting,abnormal odour, of liver disease

Other abdominal symptoms → see before

Other symptoms of other systems.....

.....

Past history

a) Perinatal history:

-Prenatal

-Natal (History of umbilical vein catheterization or umbilical sepsis)

-Postnatal

b) Developmental history

Motor development - Mental development

c) Nutritional history

d) Vaccination history

e) Previous infections, significant illness

Family history

Similar condition in the family

- History of similar conditions in the family

Examination

1- General

Observation: - ***Level of consciousness (LOC) and activity:*** Conscious, lethargic or comatose

- ***Appearance:*** Pallor, cyanosis, jaundice
- **Abnormal** features (e.g. mongolism).

2. Vital signs:

a) Temperature: 37.0 - 37.5°C.

b) Heart rate:

c) Respiratory rate

d) Blood pressure

- **Head:**

Jaundice - pallor and signs of vitamin deficiency

- Neck: lymph nodes all groups
- Trunk: purpuric eruptions - tender sternum
- Limbs: upper limbs: floppy tremors- palmer erythema Lower limbs: edema

2- Systems

Abdominal examination

Inspection

- Shape and contour → Generalized distention if huge spleen and liver
- sub costal angle is wide
- Free respiratory movement
- No visible peristalsis.
- Divercation of recti.

Umbilicus: Site: Normal: midway between xiphisternum and symphsis pupis Shifted downward in distention

Shape: everted (increased intra-abdominal pressure)

- Normal genitalia.
- Back →no mass or spina bifida.
- No striae or pigmentation.
- Skin: stretched-may be striae from distension
- Scars: especially that of splenectomy
- No hernia.....
- Subcutaneous tissues: visible veins

Palpation

Superficial: tenderness rigidity and superficial masses

Deep: aim to detect organ enlargement

Hepatomegaly or splenomegaly or hepatosplenomegaly according to the case

Hepatomegaly

Lower border of the liver is felt

In midline ..... cm below costal margin

In MCL ..... cm below costal margin

It is not tender firm in consistency with rounded border

Splenomegaly

Spleen is enlarged, its lower border is felt.... cm below left costal margin, it is not tender , firm in consistency with rounded, border a notch my be felt

Kidney

- **No renal masses could be felt**

Percussion

- **Upper border of the liver: - 5th intercostal space (4th space in cirrhosis)**
- **For ascites**

Mild: - knee chest position

Moderate amount: - shifting dullness

Huge amount: - transmitted thrill (in veno-occlusive disease)

Auscultation

- **Intestinal sounds.**
- **Venous hum: paraumiblical vein (midway between the sternum and umbilicus**

What is your diagnosis?

- Tense ascites, hepatomegaly →Veno-occlusive disease

-A case of hepatomegaly +/-portal hypertension (+/-liver cell failure)→ Bilharzial

-A case of splenomegaly Hematemesis, ascites with shrunken liver most probably post- hepatitic cirrhosis with portal hypertension.

- Manifestations of liver cell failure →jaundice, palmer erythema, spider naevi - hepatic precoma or coma-bleeding-failure to thrive-fever-feter hepaticum)

-infant +hepatomegaly only +hypoglycemia most probably metabolic liver disease

- hepatosplenomegaly +anemi+ bleeding + G lymphadenopathy most probably leukemia

Why Bilharzial?

- Age usually above 7 years
- Contact with water channels
- Terminal hematuria or bleeding per rectum
- Intake of antibilharzial treatment

Why veno-oclusive disease ?

- Young age (1 -4 years)
- Tense rapidly filling ascites
- Dilated veins on the abdominal wall
- Hepatomegaly

Why metabolic liver disease?

- Young age
- History of similar conditions on the family
- Hypoglycemic symptoms (glycogen storage) or neurological symptoms
- Huge hepatomegaly or hepatosplenomegaly

What are the Causes of hepatomegaly?

What are the Causes of splenomegaly?

What are the metabolic liver diseases you know?

1- Inborn errors of carbohydrate metabolism

- **Galactosemia (hepatomegaly-cataract and mental retardation)**
- **Glycogen storage disease (hepatomegaly- hypoglycemic episodes)**

2- Inborn errors of lipid metabolism (lipidosis)

- **Gaucher disease (hepatosplenomegaly mainly splenomegaly and neurological manifestations)**
- **Neimann Pick disease (hepatosplenomegaly mainly hepatomegaly and neurological manifestations)**

3- Inborn error of cupper metabolism

- **Wilson disease: -**

  Start after 15 years of age

What are the manifestations of portal hypertension?

What are the treatment lines of portal hypertension?

What are the investigations to be done in this case?

1- Laboratory

- Liver function tests
  - Plasma protein
  - Prothrombin time and prothrombin concentration
  - Enzymes: ALT, AST and alkaline phosphatase
  - Bilirubin
- Complete blood picture
- Stool and urine for Bilharziasis.
- Hepatitis markers
- Study of the ascetic fluid tap ifascites is present
- Other laboratory tests if metabolic liver disease is suspected :
  - Serum cupper and ceruloplasmin (Wilson)
  - Enzyme assay in cultured skin fibroblast

2- Imaging

- Sonar
  - Size of liver and spleen
  - Special echo pattern in liver cirrhosis
  - Portal and splenic vein diameter in portal hypertension
  - Ascites: amount
- Barium swallow for varices
- Splenoportography to detect size of portal vein obstruction

- Doppler study in splenic or portal vein obstruction

3- Endoscopy

- Upper endoscopy for varices
- Sigmoidoscopy and rectal snip for Bilharzial polypi

4- Biopsy

- Liver biopsy
    - Metabolic liver disease
    - Chronic hepatitis

What is the treatment of bilharziasis ?

Praziquantel 40 mg/kg single dose

Cardiology

History

Personal history

Age: CHD →MAY< 3 years

Rheumatic above 5 years

Complaint

- One of the cardiac symptoms :

The commonest is shortness of breath – generalized body swelling

- Arthritis

Present history:

I- Complaint analysis :**O C D**

II- Cardiac symptoms (symptoms of left and right side failure)

I- Left side heart failure

Pulmonary congestive symptoms

Dyspnea→ GRADES I.....

II.....

III.....

→ paroxysmal nocturnal dyspnea→ orthopnea

Onset, duration and course

Related to exertion

Character: dry (due to congestion) or productive (infection)

Cough of cardiac diseases is usually dry exertional and follow dyspnea

Hemoptysis not common in pediatric age group

Low cardiac output symptoms

Syncope

Easy fatigue

Coldness of extremities

2- Right side heart failure (systemic congestive symptoms)

- Edema

  Onset and course

  Site of appearance.....

  Pain on the right hypochondrium... ... ...

  ... ...

- GIT congestion: e.g. vomiting

3- Others

- Palpitation: related to exertion
- Chest pain (pericarditis)
- (fever)

Other symptoms of same system.....

.....

Other symptoms of other systems.....

.....

Past history

a) Perinatal history: *-Prenatal -Natal -Postnatal*

b) Developmental history

Motor development $..... - Mental development

c) Nutritional history

d) Vaccination history

e) Previous infections → **Recurrent attacks of tonsillitis- previous attacks of rheumatic fever -Treatment e.g. long acting penicillin**

Family history

Similar condition in the family

Examination

General

1- Vital signs

- Pulse
  - Rate.....
  - Rhythm: Irregular in.....
  - Character →water hammer pulse in aortic regurgitation
  - Volume: →large volume in aortic regurgitation
  - Equality on both sides
  - Equality in both UL & LL
- Temperature: fever in cardiac patient →infective endocarditis
- Blood pressure: big pulse pressure in aortic regurgitation.
- Respiratory rate.....

2- Head examination

- Eye → jaundice
- Eye lid : puffy or not
- Mouth
  - Lips: pallor
  - Tongue: cyanosis

3- Neck

Carotid

- Inspection

  Exaggerated carotid pulsation

  → Aortic regurgitation

  → Hyperdynamic circulation
- Palpation: Thrill over the arteries (in aortic regurgitation)

Jugular veins

- **Neck veins are not congested shows systolic collapse**
- If congested measure the amount of congestion in cm (while the patient in semisitting) by distance between level of congestion and sternal angle- if more than 2 cm it is significant

4- Upper limbs

- Clubbing.....
- Splinter hemorrhage.....
- Oslar nodules.....

5- Lower limbs

- Pulsation →Dorsalis pedis artery
- Edema
- Clubbing

Peripheral signs of aortic regurgitation

- **Head and neck**
  - **Prominent carotid pulsation**
  - **Head nodding**
  - **Suprasternal pulsation**
- **Upper limbs**
  - **Capillary pulsation**
  - **Water hammer pulse**
  - **Increased pulse pressure**
- **Lower limbs**
  - **Pistol shot (over femoral)**
  - **Duroiezier sign (systolic and diastolic murmur over femoral artery)**
  - **Hill sign (blood pressure in lower limb more than 50mm Hg than upper limb) normally it is 20mmHg**

Heart examination

1- Combined inspection and palpation
2- Percussion
3- Auscultation

I- Combined inspection and palpation

- Inspection	- Palpation
Pericordial bulge In long standing cardiomegaly **Pulsation** 1- Apex - Site Lower most, outer most area of pulsation. Normal apex : Up to 4 years: on the 4th intercostal space just outside mid-clavicular line Above 4 years : 5th intercostals space - Area (localized =2cm)	confirm - Area - Site Character • Hyperdynamic in aortic regurg.and mitral regurg. • Slapping in mitral stenosis Thrill • Systolic in mitral regurg. • Diastolic in mitral stenosis
2- Left parasternal (3,4,5 space) Pulsation	- Uplift - Thrill in VSD
3- Epigastric Pulsation	Origin of pulsation
4- Pulmonary area (2nd Lt. Space) Pulsation	Palpable second sound
5- Aortic area (2nd Rt. Space) Free	

There is no Pericordial bulge; apex is seen on the 5th left intercostals space inside MCL, localized.

There is no left parasternal pulsation no pulsation in the epigastric pulmonary or aortic area and no pulsation is seen elsewhere

Confirm Apex - thrills- parasternal pulsation- epigastric pulsation- Aortic and pulmonary pulsation

Interpretation of the results of inspection, palpation and percussion

Lt. ventricular dilatation → Apex is deviated out. &down

→ Apex is localized Hyper-dynamic

Rt. ventricular hypertrophy → Apex is deviated outward

→ Apex is diffuse

→ LT. Parasternal pulsation.

→ Epigastric pulsation.

4- Auscultation

Sounds

- First heart sound: - due to closure of mitral and tricuspid valve, heard best over mitral and tricuspid area
- Second heart sound: - due to closure of pulmonary and aortic valves, heard best over pulmonary and aortic area

Timing of heart sounds by carotid pulse

First heart sound: systole (with the pulsation)

- Apex : First heart sound
  - →Mitral regurgitation : muffled
  - →Mitral stenosis : accentuated
- Pulmonary area: second heart
  - →Pulmonary hypertension: accentuated second sound

Murmurs

I- Mitral area

1- Mitral regurgitation

- Site: apex
- Area of propagation : axilla
- Character :soft
- Timing: pansystolic

Heard best by

- Lying in the left lateral position
- Diaphragm of the stethoscope

2- Mitral stenosis

- Site :apex
- Area of propagation: localized
- Character: rumbling
- Timing: mid diastolic presystolic

Heard best by

- Lying in the left lateral position
- The cone

II- Aortic area

Aortic regurgitation

- Site : 2nd aortic area (3rd left space)
- Area of propagation: apex
- Character: soft
- Timing: early diastolic (decrescendo)

III- Pulmonary area

Pulmonary hypertension

Soft ejection systolic murmur (functional)

Diagnosis of a cardiac case

1- Etiological diagnosis →CHD- rheumatic

2- VALVE AFFECTED:

Mitral regurgitation

Mitral stenosis

Aortic regurgitation.

&ventricle enlarged

Left ventricle .....

Right ventricle.....

Biventricular.....

3- Compensated -OR-Heart failure

4- Complications

Infective endocarditis - Activity (arthritis, carditis, chorea) - Chest infection

The condition started 2 month ago by acute onset of fever, arthritis affecting the right ankle and knee joint, the affected joints were swollen, tender and the child was unable to move. The condition was stationary for 2 days then arthritis migrated to affect the left elbow joint.5 days later the child complained of dyspnea associated with orthopnea and paroxysmal nocturnal dyspnea and cough that was dry and related to exertion The condition was also associated with chest pain and palpitation. There was no history of lower limb edema or ascites, no history fainting .The patient was admitted to hospital and arranged for laboratory assessment & received treatment with gradual improvement There is past history of recurrent tonsillitis.

What are the signs of pulmonary hypertension?

What are the causes of fever in cardiac patient?

What are the major criteria of rheumatic fever?

What are the minor criteria of rheumatic fever?

What are the other causes of arthritis you know?

What are the pattern & the distribution of rheumatic arthritis?

What is the etiological theory of RF?

What are the investigations to be done in this case?

- Investigations to detect activity
    - Complete blood picture
    - Erythrocyte sedimentation rate
    - C reactive protein
    - ASOT

* Investigations to evaluate cardiac condition
    - X-ray
    - ECG
    - Echocardiography

What is the management of this case:

1- If no activity:

Benzathine penicillin 1,200.000 IM every 3 weeks as prophylaxis for life

2- If there is rheumatic activity

- Adequate treatment of streptococcal infections :
  - Single injection of Benzathine penicillin 1,200.000 IM or oral course of penicillin for at least 10 days
  - Erythromycin 50 mg/kg/day in those allergic to penicillin
- Supportive →Rest: especially in carditis (absolute bed rest for at least 4 weeks.
- Specific treatment

1- Arthritis (or carditis without cardiomegaly)

Salicylates

100 mg /kg/ for 2 weeks then 75 mg/ kg for 4-6 weeks.

2- Carditis with cardiomegaly →Prednisone 2 mg/kg/ day for 3 weeks, then taper

Salicylates 75 mg /kg/ day during tapering and for 1 month after Stopping Prednisone to avoid rebound phenomenon

3- Chorea

Phenobarbitone 3-5 mg/kg/day

Haloperidol 0.02-0.1 mg/kg/day (in patients over 12 years)

3- If heart failure is present:

1- Supportive: - rest, 0_2, low salt diet, small frequent meal.

2- Specific

- Preload reducing agents: Diuretics: Furosmide 2 mg/kg / day
- Inotropes →Digoxins :Digitalizing dose 0.02-0.04mg/ kg
  Maintenance dose 0.005:0.01 mg /kg/ day
- After load reducing agents (Vasodilator) e.g. Captopril

Chorea

History

Personal

More common in girls

Age: 5-15 years

Complaint

- Involuntary movements with repeated falling of objects from her hands

- Weakness and inability to perform fine movement as writing
- Emotional instability

Present history

- **Onset (insidious), course (regressive in rheumatic chorea) and duration.**
- **Exclusion of other causes of chorea.**
  - **History of drug intake (major tranquilizer)**
  - **History of fever, convulsions (encephalitis)**
  - **Neurological symptoms: weakness, increased tension, and convulsions So, it is rheumatic chorea by exclusion of other causes of chorea**

- Exclude associated carditis (ask about all cardiac symptoms)

Past history

a) Perinatal history:

-Prenatal -Natal

-Postnatal →jaundice (post kernicteric chorea).

b) Developmental history

Motor development - Mental development

c) Nutritional history

d) Vaccination history

- ***e) Previous infections*** →Repeated tonsillitis

Family history

Similar condition in the family

History of similar conditions in the family

Examination

- Level of consciousness (LOC) and activity: Conscious, lethargic or comatosed

- ***Appearance:*** Pallor, cyanosis, jaundice
- **Abnormal** features (e.g. mongolism).

2. Vital signs:

a) Temperature: 37.0 - 37.5°C.

b) Heart rate: Normal heart rate varies

c) Respiratory rate

d) Blood pressure

Regional examination →*Head and neck &UL & LL& CHEST & ABDOMEN* →→→Cardiac

→→→Neurological

1- Choreic movements

- Affect tongue, face, trunk and extremities
- The movements are
  - pseudopurposive Involuntary, static, dysrhythmic, sudden jerky
  - increased by →emotional stress
    →FATIGUE
    →Sleep deprivation
    - absent during sleep
- **Pronation** occurs in the elevated arms
- **protrusion** of the Tongue →can not be maintained protruded except by holding
- **Buttoning** test: patient can not button his shirt easily

2- Hypotonia can be demonstrated by:

- Milker sign: - ask the patient to squeeze the examiner **hand** , fluctuation of the hand grip is observed
- Boat shaped **hand**s: on arm extension, flexion of the wrist and hyperextension of the fingers
- Pendular **knee** jerk

3- Emotional lability

Sudden laughing or crying

What is your diagnosis?

A case of rheumatic chorea, the usual type

Why chorea?

History

A girl, 6 years old presenting with involuntary movement in the form of

Examination

- Involuntary movements
- Hypotonia
- Emotional lability

What are the clinical varieties of rheumatic chorea ?

- Usual type

- Chorea mollies: paralytic chorea due to severe hypotonia
- Chorea gravies: choreic movements interfere with speech and sleep
- Hemichorea: chorea affects one side

What are the causes of chorea?

- The most important is rheumatic chorea
- Other causes are :
    - Hereditary
    - Post kernicteric chorea
    - Post encephalitic chorea
    - Degenerative: Huntington chorea
    - Drugs: chlorpromazine

Why you diagnose your case as rheumatic chorea?

- Other causes of chorea are excluded as there is
    - No history of preceding fever, convulsions (encephalitis)
    - No history of drug intake
    - Age of onset (after 5years) exclude hereditary and post-kernicteric chorea
    - Regressive course, exclude neurodegenerative causes of chorea
- Valve lesion, if present support the diagnosis of rheumatic chorea

What is the site of damage in chorea?

What is the differential diagnosis of chorea ?

From other types of abnormal movements

Chorea	Athetosis	Dystonia	Tremors	Tics
- Irregular - Sudden jerky - Pseudopurposive - Proximal - Hypotonia	- Irregular - Slow (Snake like) - Purposeless - Distal - Hypertonia	- Irregular - Very slow (Twisting) Purposeless - Proximal Hypertonia	- Regular - Rhythmic - Purposeless - Distal	- Regular - Purposeless - The same muscles

What are the investigations to be done in this case?

What is the treatment of rheumatic chorea ?

- Haloperidol 0.05 mg/kg/day

Duchenne dystrophy

History

- Personal

Age: after the age 3 years

Sex: males (X linked recessive)

- Complaint

- Loss of the ability to (walk- stand- ascend stairs).
- Difficulty in getting up stairs, getting up from sitting position slowly

- Present history

- Complaint analysis

Onset, course (progressive), duration. Insidious onset and slowly progressive course up to difficulty in waking

Developmental history

Neurological symptoms delayed- *Motor development*

-Mental development

No symptoms suggestive of cranial nerve affection, sensory affection

Associated symptoms-investigations -treatment

Past history

a) Perinatal history:

Prenatal- Natal-Postnatal

b) Developmental history

Motor development: ……

……

Mental development: ……

c) Nutritional history

d) Vaccination history

e) Previous infections, significant illness

Family history

Parents: consanguinity

Similar condition →May be positive from the mother side

Examination

- Neurological examination

Motor system

Inspection: Pseudohypertrophy of calf muscles the (commonest site) followed by muscles of the forearm

Lower limbs: Calf muscles- Qudriceps group-Gluteal muscles

Upper limbs: Deltoid - Supraspinatus - Infraspinatus

- **Power**
  - Weakness proximal more than distal
  - The following signs will demonstrate muscle weakness :
- Gower sign : the patient climb himself when asked to stand from sitting position
- Waddling gait
- Exaggerated lumbar lordosis on standing

- **Tone**

Decrease on the affected muscles

Reflexes: - intact or hyporeflexia depending on the stage of the disease

Sensations and cranial nerves: - are intact

2- **Heart**: cardiomyopathy (muffled heart sounds)

3- **Chest:** chest infection from respiratory muscle weakness

What is your diagnosis

A case muscle dystrophy most probably Duchenne

→With or without chest infection

→ With or without chest infection

Why

→Male patient

→gradual onset , slowly progressive course motor weakness.

→No symptoms suggestive of sensory, cranial nerve or sphincter affection.

What are the Investigations?

- Laboratory

CK (creatine kinase): marked elevation even at birth

Elevated also in female carriers

- EMG: myopathic pattern
- Echocardiography
- DNA studies (probe): deletion on dystrophin gene)
- Muscle biopsy: diagnostic (absent dystrophin)

What are the treatment lines ?

- Good nutritional support
- Physiotherapy
- Treatment of complications e.g. chest infection
- Myoblast transfer
- Gene therapy

Is there any method for antenatal diagnosis?

Amniocentesis (amniotic fluid fibroblast culture) then chromosomal and gene study

What are the causes of death in Duchenne dystrophy?

Why males are only affected in Duchenne dystrophy?

What are the situations in which females are affected in Duchenne ?

- Turner syndrome (XO)
- Lyonization

DD of myopathy?

I- Pelvic girdle type

1- Duchenne

Early onset and rapid course

Dystrophin is absent

2- Beker

Delayed onset, slower course

Dystrophin is present but reduced in amount

Both are sex linked recessive

II- Limb girdle type

- Autosomal recessive (girls and boys are affected)

- Normal muscle dystrophin

III- Shoulder girdle type

1- Scapulohumoral

- Sex linked recessive
- Delayed onset and slower course
- Muscle wasting, no hypertrophy

2- Facio Scapulohumoral

- Autosomal dominant
- Present late in childhood
- Muscle wasting in facio Scapulohumorai distribution

→ Inability to close the eye tightly

→ Winging of the scapula

What is the pathogenesis of Duchenne?

How to differentiate myopathy & neuropathy?