Cyanotic Congenital Heart Disease

Occurs when blood is shunted from the venous to the arterial system as a result of abnormal communication bet. the two systems (deoxy to oxy. R to L shunts)

Decreased pulmonary blood flow (Right to left shunt)

Tetralogy of Fallot (Cyanotic)

Localized narrowing of the aorta, the most common cardiac malformation responsible for cyanosis in a child over 1 year. **4 major defects:**

Pulmonary stenosis	Ventricular septal defect
Aorta overriding VSD	Right ventricular hypertrophy

Pathophysiology

•PS impedes the flow of blood to lungs

•increases pressure in the right ventricle (hypertrophy)

•Forces deoxy blood through the septal defect to the left ventricle

•Overriding aorta receives blood from both right and left ventricles.

Signs & Symptoms

•Clubbing of the fingers, murmur

•Severe dyspnea, hypercyanotic spells & acidosis

•Polycythemia & clot formation

•Failure to thrive and growth retardation.

Treatments

Tetralogy of Fallot (Cyanotic) (cont)

•If O2 level ex low, prostaglandins (IV) to keep PDA open.

•Complete repair when 6 mos. of age.

•Closure of the VSD with dacron patch

•The narrowed pulmonary valve is enlarged

•Coronary arteries will be repaired

•Hypertrophy of right heart should remodel when pressure in right side is reduced.

Acute Rheumatic Fever (RF)

Streptococcal infections causes damage to the heart muscle and valves. All heart layers are affected.

•Pancarditis (all layers)	•Pericarditis
•Myocarditis	•Endocarditis

Pathogenesis

•An acute attack of streptococcal pharyngitis by group A beta-hemolytic streptococci

•Within 2-4 weeks after this attack anti-streptococcal antibodies are formed and attack the heart and the extra cardiac sites.

Diagnostic test

•Throat swab, blood test, ECG heart test

Tricuspid Atresia (Cyanotic)

Tricuspid valve fails to develop causing complete closure of the tricuspid valve that results in mixed blood flow.

Signs & Symptoms

•Cyanosis in NB, Chronic hypoxia, clubbing

•No blood flow form the right A to the right V

•Heart failure, chronic hypoxemia

•Failure to thrive and growth retardation.

Diagnosis

•Listening with a steth for changes in heart sounds

•O2 sat to see how much O2 is getting into blood

•Chest x-ray to see the size & position of the heart

•ECG to check the electrical activity

•Ultrasound scan (echocardiography)

Treatment

•A glenn | Cavopulmonary | HemiFontan shunt

making a passage between the superior vena cava to the right lung (pulmonary) artery

•A fontan | Total Cavo-Pulmonary Connection

This operation joins the inferior vena cava to the pulmonary arteries.

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Erythema Marginatum

Nonpruritic, painless erythematous rash on trunk and/or proximal extremities.

•Macular lesions with raised margins & central clearing

•May last from weeks to months

Jones Criteria for Diagnosis of RF

Minor Manifestations

Clinical: Fever, arthralgia, history of RF

Lab test: Acute phase reactants, ECG changes

Major Manifestations

• Carditis: friction rub, murmur, cardiomegaly, CHF

•Arthritis: migratory polyarthritis, swollen, red, tender

•Chorea, subcutaneous nodules, Erythema marginatum

Sydenham's Chorea

A neurologic disorder with muscular weakness, emotional lability and involuntary, uncoordinated, purposeless movement

•Disappear during sleep, lasts 2-4 months

•Mainly occurs in hands, feet and face, sensation intact

Acquired Heart Defects

• An autoimmune disease affecting the heart and extra cardiac sites (joints, brain, skin)

•Occurs 10 days to 6 weeks after pharyngitis

•The disease affects children and young adults (5-15yrs)

•The disease follows upper respi infection (tonsillitis)

Cardiac Manifestations

•Pericardial friction rubs, weak heart sounds

•Tachycardia, arrythmias

•Extracardiac: fever, polyarthritis, arthralgia, skin lesions, chorea

•Pharyngeal culture may be negative

• Anti streptolysin O (ASO) titer will be high

Arthralgias and Arthritis

•May be migratory. last for 2 - 3 weeks

•Warm, swollen, tender joints, inflamed membrane

•Usually involves the knees, ankles, elbows, and wrists

•Eroded cartilage and narrowed joint space

Subcutaneous Nodules

Usually associated with severe carditis and occur several weeks after

•Firm, painless nodules (up to 2cm) found over bony surfaces and tendons. persist for 1-2 weeks.

•Occur near elbows, knees, wrists, achilles tendon, vertebral joints

Kawasaki Disease

Idiopathic multisystem disease. vasculitis of small & medium blood vessels & arteries. Usually <5 yrs old.

Mucocutaneous Ocular Syndrome (MCOS) the original name of kawasaki disease. The cause is unknown.

Diagnostic Criteria

•Changes in extremities: edema, erythema, desquam.

•Polymorphous exanthem, usually truncal

•Conjunctival injection, fissuring of lips & oral cavity

•Cervical lymphadenopathy

Symptoms

•Red eyes, changes in lips, tongue & mouth

•Redness, cracking on lips, red strawberry tongue

•Hands & feet: red, swollen, hands & soles.

•Body rash : 1st appears w/fever. bumpy to touch

•Swelling of glands in the neck, soft, painless nodes

Phases

Acute: 1-2 weeks from onset, febrile, irritable, oral changes, rash, edema/erythema of feet

•Subacute: 2-8 weeks, desquamation, persistent arthritis or arthralgias, gradual improvement.

• Convalescent (months to years later)

Treatment

•No single medicine can treatment or cure

• Meds : Aspirin and gamma globulin

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