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TRANSPOSITION OF GREAT ARTERIES

DEFECTS OF CARDIO VASCULAR SYSTEM (CYANOTIC AND ACYANOTIC)

- The heart is developed completely in the first 8 weeks of intrauterine life. One or several anamalies may result from maldevelopment of heart or great blood vessels leading to and from the heart, producing congenital heart diseases.

DEFINITION

Congenital heart disease is defined as the structural, functional or positional defect of the heart in combination, present from birth, but may manifest at any time after birth or may not manifest at all.

CAUSES

- Such defects may be due to hereditary, vitamin deficiency and viral infections such as rubella occurring during first trimester of pregnancy. Some of other includes:
- Hereditary.
- Consanguineous marriage
- Intrauterine viral infections and rubella
- Teratogenic effects of drugs like phenytoin and thalidomide.

Contd...

- Alcohol intake
- Radiation during I trimesters of pregnancy.
- Maternal IDDM.
- High altitude.
- Fetal hypoxia.
- Birth asphyxia.

CLASSIFICATION

CHD (Congenital Heart Disease) can be classified into two groups based on defects in cardiac structures.

1. **Cyanotic Heart Disease:** Infant shows varying degrees of cyanosis because mixing will occur.
2. **Acyanotic heart disease:** No cyanosis because there is no mixing of deoxygenated blood in the systemic circulation**.**

CHD (CONGENITAL HEART DISEASE)

1. Cyanotic heart Disease

- Tetralogy of Fallot (TOF)
- Transposition of great arteries (TGA)
- Tricuspid Atresia (TA**)**

- **2. Acyanotic heart disease**

Left – right shunts

- Ventricular Septal Defect (SD)
- Atrial Septal Defect (ASD)
- Patent DuctusArteriosus (PDA)

Outflow obstruction

1. Pulmonary Stenosis
2. Aortic Stenosis
3. Coarctation of aorta

TSV
(TRANSPOSITION OF GREAT VESSELS)

DEFINITION:

Transposition of great arteries (TGA) occurs when the pulmonary artery originates from the left ventricle and the aorta originates from the right ventricle.

Incidence

- lt accounts 5 to 10 per cent of all CHDs.
- It occurs predominantly in males. Incidence significantly high in the history of diabetes in grandparents and the babies having large birth weight.

Pathophysiology

- In TGA , there are two independent circuits of circulation. Life can only be maintained post-natally, if some communication between systemic and pulmonary circulation exists.
- Aorta carries deoxygenated blood to the systemic circulation
- Pulmonary artery carries oxygenated blood back to lungs

Systemic Circulation:

- Deoxygenated blood from body organs

- Blood enters into the right atrium

- Blood enters into the right ventricle

- Enter into the aorta

- Deoxygenated blood enter in systemic circulation

Pulmonary Circulation:

Lungs (oxygenated blood)

(Pulmonary veins)

Enter into the left atrium

Enter into the left ventricle

Enter the pulmonary artery

Again oxygenated blood goes to lungs

CLINICAL MANIFESTATIONS

Clinical features depend on size and associated defects:

- Cyanosis at birth.
- Dyspnoea.
- Metabolic acidosis.
- Severe hypoxia.
- Cardiomegaly.
- Clubbing may occur in few months.
- Cool and clammy skin.
- Symptoms of congestive heart failure.
- Growth failure.
- Tachycardia

DIAGNOSIS

- **History collection** about occurrence of disease.
- **Physical examination** to detect heart murmurs.
- **Arterial Blood Gas analysis** reveals hypoxemia and acidosis.
- **Chest X-ray** may reveal the classic findings of a narrow upper mediastinum, with an "egg on side" appearance of the cardiac shadow.
- **ECG** is rarely helpful in establishing the diagnosis as it is usually normal.
- **Echocardiography** is essential to demonstrate the abnormal arterial connections and associated abnormalities

MANAGEMENT

Medical Management:

- Administration of intravenous prostaglandin E which may be initiated to temporarily increase blood mixing, if systemic and pulmonary mixing is inadequate to provide an oxygen saturation of 15% or to maintain cardiac output (Digoxin, Diuretics and Iron therapy).

SURGICAL MANAGEMENT:

- **Arterial switch procedure:** This is the procedure of choice performed in first week of life. It involves transecting the great arteries and anastmosing the main pulmonary artery to the proximal aorta just above the aortic valve) and anastomosing the ascending aorta to the proximal pulmonary artery.

- **Intra atrial baffle repairs:** It is rarely performed. An intra atrial baffle is created to divert venous blood to the mitral valve and pulmonary venous blood to the tricuspid valve using the patient's atrial septum (senning procedure) or a prosthetic material (mustard procedure) performed at first year of life.

- **Rastelli procedure:** It is choiceful operation in infants with TGA, VSD, and severe pulmonic stenosis (PS). It involves closure of the VSD with a baffle, directing left ventricular blood through the VSD into the aorta.

COMPLICATIONS

- Dysrhythmias
- Right ventricular dysfunction.
- Baffle obstruction.
- Sudden death (2% to 10%).