1. TETRALOGY OF FALLOT (TOF).

Definition

Tetralogy of fallot is the most complex congenital heart defect with decreased pulmonary blood flow. It includes a combination of 4 defects:

i) Ventricular Septal defect.
ii) Overriding of aorta.

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iii) Pulmonary stenosis
iv) Right ventricular hypertrophy.

Incidence
TOF accounts for about 6-10% of all Congenital heart defects

Pathophysiology
In TOF blood returns normally from systemic circulation into the right atrium & then reaches the right ventricle. The outflow of blood from the right ventricle to lungs is restricted due to pulmonary stenosis, leading to right ventricular hypertrophy. As pressure in the right ventricle increased due to severe pulmonary stenosis blood starts shunting from right to left, through ventricular septal defect. Unoxygenated blood mixes with oxygenated blood. Because of pulmonary stenosis, blood flow to the lungs is reduced so amount of unoxygenated blood reaching the systemic circulation (through overriding aorta) increases, leading to cyanosis.

The body attempts to compensate for unoxygenated blood by developing polycythemia. The resulting increased viscosity of blood causes slowing of circulation & possibly thrombophlebitis, emboli & cerebrovascular disease.

Clinical Features :-
- The most significant symptom is cyanosis. The degree of cyanosis depends on severity

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of pulmonary stenosis. The infants with mild pulmonary stenosis may be pink at rest & become cyanosed on crying or during activity. In severe obstructions cyanosis occur even at rest.
• In severely affected infants, skin becomes dusky or bluish in color.
• Clubbing of finger & toe nails occur by 1-2 years of age.
• Exercise causes dyspnea. Children often assume 'squatting position' is between play, as this position relieves dyspnea.
• Paroxysmal dyspneic attacks (anoxic blue spells known as "tet spells") may occur during first 24 months of life & may last from few minutes to hours. These dyspneic & cyanotic spells result in hypoxia & metabolic acidosis.
• The children are small & their nutritional status is poor.

Diagnostic Evaluation.
(i) Cardiac Examination:- A harsh systolic murmur is heard along left sternal border.
ii) Electrocardiogram:- Right ventricular hypertrophy is almost always present on ECG.
iii) Chest Radiograph:- The characteristic "boot-shaped" heart is seen due to right ventricular hypertrophy. The

pulmonary artery is small & aorta is larger than normal. The hilar areas of lung appear clear due to reduced or diminished pulmonary blood flow.

iv) Echocardiography: With color flow 2D echo & continuous wave doppler, the VSD, overriding aorta & pulmonary stenosis can be visualized.

v) Cardiac Catheterization: Cardiac catheterization shows systolic hypertension in right ventricle with rapid fall in pressure. pressure as the catheter goes into the pulmonary artery.

Therapeutic Management:

* Medical Management

- Place the infants or the child in knee chest position. This position enhances the systemic venous return, which helps to dilate the right ventricle thereby decreasing right ventricular pressure leading to right to left shunting.
- Propranolol is usually administered in a dose of 1 mg/kg body weight, upto four times in a day, to reduce the pulmonary artery * valve spasm.
- IV prostaglandin E, therapy is given to neonates with tetralogy of fallot. This helps in keeping ductus arteriosus patent, thereby increasing pulmonary blood flow & improving systemic arterial blood oxygenation.

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* Surgical Management:
Infants who are severely cyanotic or have TETT spells require either a palliative procedure to increase the blood flow to lungs or a total repair.

a) Palliative Surgery
These procedure are done if the infant is too young for a full repair & include -

- Blalock-Taussig shunt :- In older infants & children, an artificial ductus is created by connecting right or left subclavian artery to the pulmonary artery of same side. This allows increased blood flow to the lungs. The procedure is performed through a lateral thoracotomy incision.
- Potts procedure : The upper descending aorta is anastomosed with left pulmonary artery.
- Waterston shunt :- It involves side to side anastomosis of ascending aorta with right pulmonary artery.
- Brock's procedure :- In this surgery, pulmonary valvotomy is done to correct pulmonary stenosis. This surgery increases pulmonary blood flow. but does not correct VSD.

b) Corrective Surgery
It includes a patch closure of VSD & pulmonary valvotomy for relief of

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pulmonary stenosis. If the pulmonary stenosis is severe, a homograft is done to correct. Conduit is placed from right ventricle to the pulmonary artery, to bypass the obstruction.

* Postoperative Complications
- Conduction abnormalities (heart block)
- Residual ventricular septal defect.
- Residual pulmonary stenosis.
- Pulmonary valve regurgitation.

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Transposition of Great Arteries

The transposition of great arteries is ventriculo arterial discordance, in which the aorta arises from the morphologic right ventricle & the pulmonary artery arises from the morphologic left ventricle.

Etiology

- Etiology is unknown & is presumed to be multifactorial
- This defect is more common in infants of diabetic mothers

Pathophysiology

(The pulmonary & systemic circulations functions in parallel, rather in series)

Causes

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Transposition of great arteries

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Oxygenated pulmonary venous blood returns to the left atrium & left ventricle

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Recirculated to the pulmonary vascular bed via the abnormal pulmonary arterial connection to the left ventricle

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Deoxygenated systemic venous blood returns to the right atrium & right

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ventricle
↓
Pumped to the systemic circulation, effectively bypassing the lungs
↓
Deficient oxygen supply to the tissues & an excessive right & left ventricular workload
↓
It is incompatible with prolonged survival unless mixing of oxygenated and deoxygenated blood occurs at some Anatomic level like ASD, VSD, PDA

Clinical Manifestations

- Prominent & progressive cyanosis within the first 24 hours of life is the usual finding in infants.
- Tachypnea
- Tachycardia
- Diaphoresis
- Failure to gain weight

Diagnosis

- History collection
- Physical examination
- Echocardiography

→ Cardiac examination: A singly or narrowed split, diminished second heart sound heard. Systolic ejection murmur may be present.

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- Chest radiography demonstrate the classic "egg on a string" appearance in approximately one third of patients.

Management

- Initial treatment consists of maintaining ductal patency with continuous intravenous (IV) prostaglandin E_1 infusion to promote pulmonary blood flow
- Antibiotic prophylactic Regimens for Endocarditis.
- Fluid replacement.
- Bicarbonate administration - Acidosis
- Mechanical ventilation

Surgical Management

- Rastelli-type procedure - ligate the main pulmonary artery & place an aortopulmonary shunt during the newborn period to restrict pulmonary blood flow.

- Arterial switch procedure

Complications

- Congestive heart failure
- Arrhythmia
- Eisenmenger syndrome (irreversible and progressive pulmonary vascular obstructive

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disease).

Total Anomalous Pulmonary Venous Connection (TAPVC).

TAPVC is defined as the anomaly in which the pulmonary veins have no connection with the left atrium. Rather, the pulmonary veins connect directly to one of the systemic veins

Cause

Unknown cause.

- It occurs because of abnormal development of the heart's pulmonary veins during early fetal growth.

Signs & symptoms

- A bluish tint to the skin & lips
- Trouble breathing
- Rapid breathing
- Poor feeding
- Poor growth
- Pulmonary hypertension
- Tachycardia

Diagnosis

- Chest X-ray
  Normal or mild cardiomegaly
  Varying degrees of pulmonary edema

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- ECG
  Right Ventricular volume overload.
- Echocardiography
- Cardiac Catheterization: To visualise the abnormal connection of pulmonary vein particularly if an obstruction is present.

Management

It requires open-heart surgery in all cases. Critically ill newborns will have surgery immediately. If the child is not critically ill, doctors may wait upto two months to perform surgery.

In TAPVC pulmonary veins despite their abnormal connections to other veins, all end in a collection (called a confluence) at the back of the left atrium. The surgeon opens the confluence so that the veins can drain into the left atrium. Then ties off all the abnormal connections between the pulmonary veins & other veins, so that blood can follow only the path to the left atrium.

Nursing Management

Preoperatively note history of cyanosis, tiring easily & difficulty in feeding

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Observe the chest for prominence of right ventricular impulse & retractions with tachypnoea. Auscultate the heart, noting fixed splitting of the second heart sound & a murmur. Palpate the abdomen for hepatomegaly.

Postoperatively, monitor for dysrhythmias, heart block & persistent heart failure