Cyanotic Congenital Heart Defects

Most cyanotic congenital heart defects (CHDs) manifest during the neonatal period, requiring a correct diagnosis for appropriate management. Detection of cyanosis has been made much easier in recent years by routine use of pulse oximetry .in asymptomatic newborns

Cardiac Causes of Cyanosis

According to age

a. Newborn (1st week)—TGA, hypoplastic right heart syndrome (tricuspid atresia, pulmonary atresia with intact ventricular septum), Ebstein's anomaly, TAPVC

b. Late newborn (upto 4 weeks)—TOF, severe PS with ASD, TGA with VSD and PS, single ventricle with PS, truncus with hypoplastic .pulmonary artery

c. Infants and toddlers—The common causes are two-As and five-Ts: Two A's are Atresia of aorta (aortic atresia) and Atresia of pulmonary artery (pulmonary atresia) and five- T's are: Tetralogy of Fallot, TAPVC, TGA, Tricuspid atresia, Truncus arteriosus

:Objectives

.Define TGA and pathology.1

Recognise clinical manifestations and physical signs of.2
.TGA

.Describe the management of TGA.3

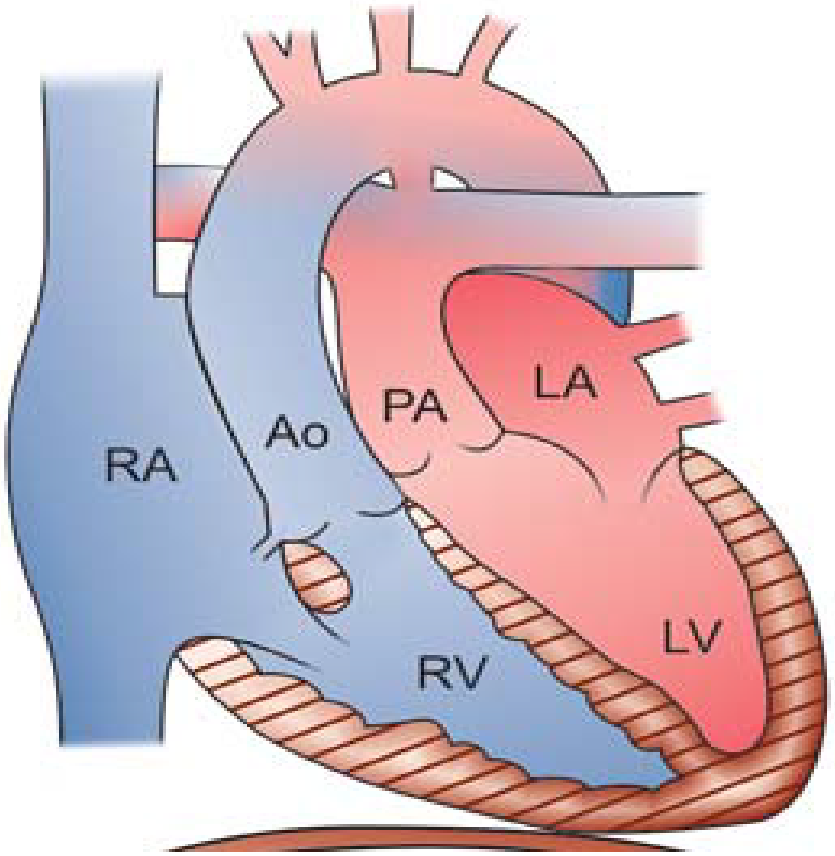
Complete Transposition of the Great Arteries

Prevalence

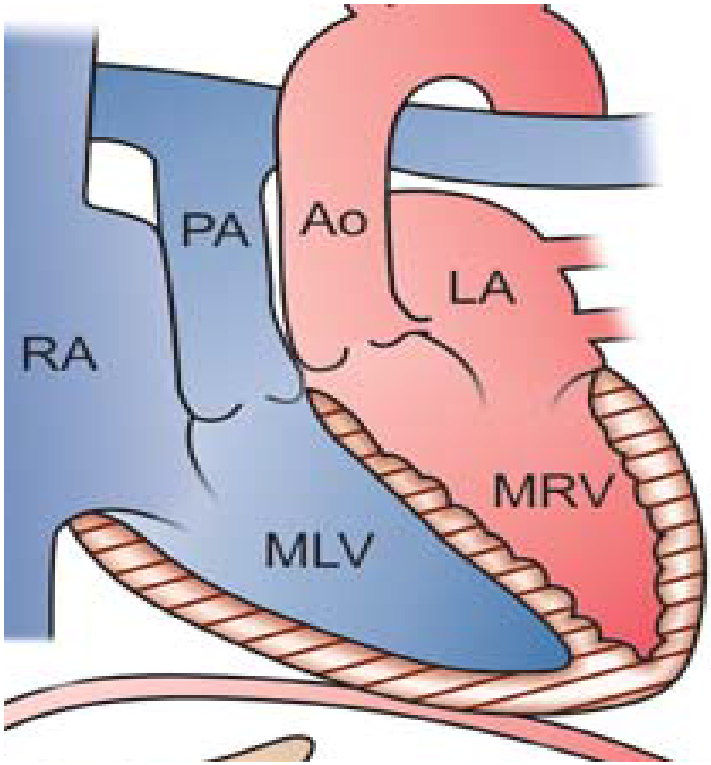
Complete transposition of the great arteries (TGA) occurs in about 5% to 7% of all CHDs. It is more common in males than in females .(male-to-female ratio of 3:1)

Pathophysiology

In complete TGA, the aorta arises anteriorly from the RV. 1 carrying
desaturated blood to the body, and the pulmonary artery (PA) arises posteriorly from the LV carrying oxygenated blood back to the lungs. In the classic complete TGA, the aorta is located anteriorly and to the right (dextro) of the PA. This is why the prefix *D* is used and thus the condition is called *Dtransposition* (D-TGA). (When the transposed aorta is located to the left of the PA, it is .called *L-transposition*



Schematic diagram of complete
.transposition of great arteries



Schematic diagram of L-TGA

In about 5% of the patients, left ventricular outflow tract (LVOT)
obstruction (or subpulmonary stenosis) occurs. The obstruction may be dynamic or fixed. VSD is present in 30% to 40% of patients with D-TGA and may be located anywhere in the ventricular septum

Clinical Manifestations

History

.History of cyanosis from birth is always present. 1

Signs of congestive heart failure (CHF) with dyspnea and feeding. 2

.difficulties develop during the newborn period

Physical Examination

Moderate to severe cyanosis is present, especially in large male. 1

newborns.Such an infant is tachypneic but without retraction unless

.CHF supervenes

The S2 is single and loud. No heart murmur is heard in infants with. 2

an intact ventricular septum. An early or holosystolic murmur of VSD

may be audible in less cyanotic infants with associated VSD. A soft

.midsystolic murmur of PS (LVOT obstruction) may be audible

.If CHF supervenes, hepatomegaly and dyspnea develop. 3

Laboratory Studies

Severe arterial hypoxemia usually with acidosis is present.. 1

.Hypoxemia does not respond to oxygen inhalation

.Hypoglycemia and hypocalcemia are occasionally present. 2

Electrocardiography

Right ventricular hypertrophy (RVH) is usually present after. 1 the first few days of life. The QRS voltages and the QRS axis may be normal in many newborns with the defect. After 3 days of life, an upright T wave in V1 may be the only abnormality suggestive .of RVH

Biventricular hypertrophy (BVH) may be present in infants. 2 with large VSD, PDA, or pulmonary vascular obstructive disease because all of these conditions produce an additional left .ventricular hypertrophy (LVH)

.Occasionally, right atrial hypertrophy (RAH) is present. 3

Radiography

Cardiomegaly with increased pulmonary. 1
.vascularity is typically present

An egg-shaped cardiac silhouette with a. 2
narrow, superior mediastinum is
characteristic

Management

Medical

a. Arterial blood gases and pH should be obtained, and metabolic. 1 acidosis should be corrected. Hypoglycemia and hypocalcemia, if .present, should be treated

b. PGE1 infusion should be started to improve arterial oxygen .saturation by reopening the ductus

This should be continued throughout the cardiac catheterization or .until the time of surgery

c. Oxygen should be administered for severe hypoxia. Oxygen may help lower pulmonary vascular resistance (PVR) and increase pulmonary blood flow (PBF), which in turn increases systemic arterial oxygen .saturation

Before surgery, cardiac catheterization and a balloon atrial. 2 .septostomy

are often carried out to have some flexibility in planning surgery. If adequate interatrial communication exists and the anatomic diagnosis of TGA is clear by echocardiographic examination, the patient may go to surgery without cardiac catheterization or the balloon atrial septostomy. The need for the balloon septostomy may be determined by inadequate atrial mixing through the PFO (evidenced with a high Doppler flow velocity of .>1 m/sec) or a lack of readiness for surgical intervention

CHF may be treated with diuretics (and **Surgical**. 3

Palliative Procedure

No palliative procedure is performed unless an arterial switch operation

.cannot be performed early in life) ASO(

Definitive Repair

Historically, definitive surgeries performed for TGA were procedures that

switched right- and left-sided blood at three levels: the atrial level (intraatrial repair surgeries such as the Senning or Mustard operation), the ventricular level (i.e., Rastelli operation), and the great artery level(ASO)

Procedures

Atrial baffle operations (Mustard and Senning. 1 operations)

These procedures reroute pulmonary and systemic venous returns at the atrial level with resulting physiologic correction. The pulmonary venous blood eventually goes to the aorta, and the systemic venous blood goes to the PA

Arterial switch operation. 2

The ASO is now firmly established as the procedure of choice. There are almost no situations which would justify the performance of a Senning or Mustard procedure for D-TGA. The coronary arteries are transplanted to the PA, and the proximal great arteries are connected to the distal end of the other great artery, resulting in an anatomic .correction

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