Crimea State Medical University, Simferopol

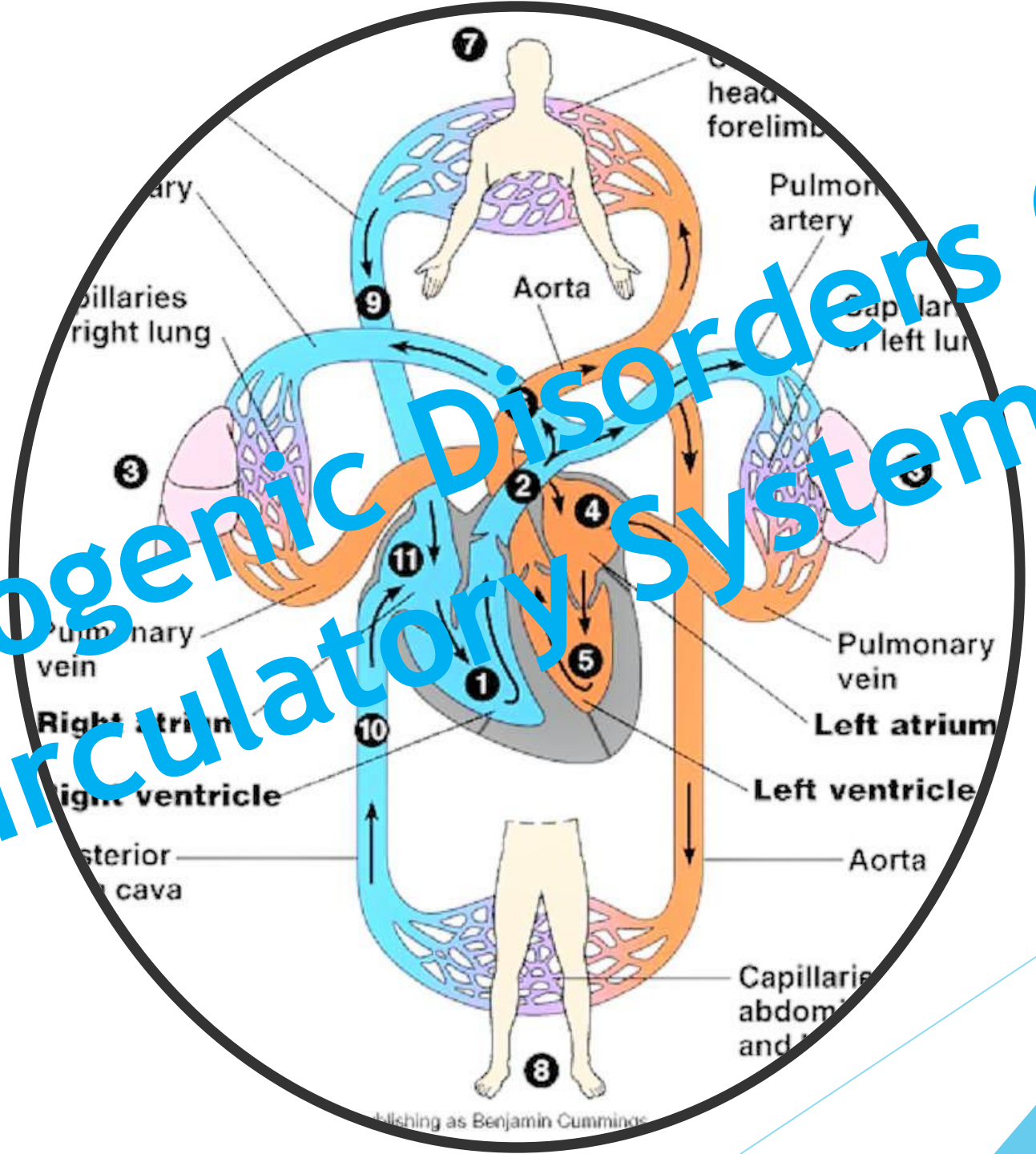
Biology Project

Teacher – Anna Zhukova

Made By - Mohammad Imran Sheikh

LA- 194 A

Phylogenic Disorders Of Circulatory System…



Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome is a complex and rare heart defect present at birth (congenital). The left side of the heart is critically underdeveloped in hypoplastic left heart syndrome.

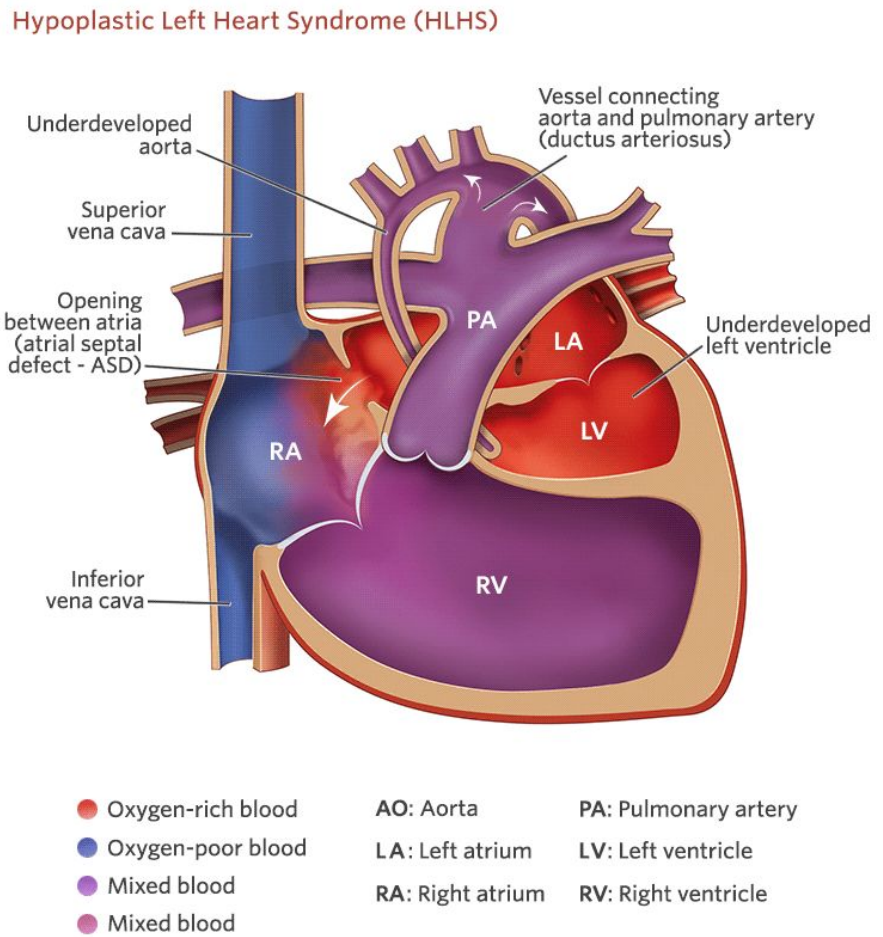
If your baby is born with hypoplastic left heart syndrome, the left side of the heart can't effectively pump blood to the body. Instead, the right side of the heart must pump blood to the lungs and to the rest of the body.

Medication to prevent closure of the connection (ductus arteriosus) between the right and left sides, followed by either surgery or a heart transplant, is necessary to treat hypoplastic left heart syndrome. With advances in care, the outlook for babies born with hypoplastic left heart syndrome is better now than in the past.

Symptoms

Babies born with hypoplastic left heart syndrome usually are seriously ill soon after birth. Hypoplastic left heart syndrome symptoms include:

Grayish-blue skin color (cyanosis)
Rapid, difficult breathing
Poor feeding
Cold hands and feet
Weak pulse
Being unusually drowsy or inactive



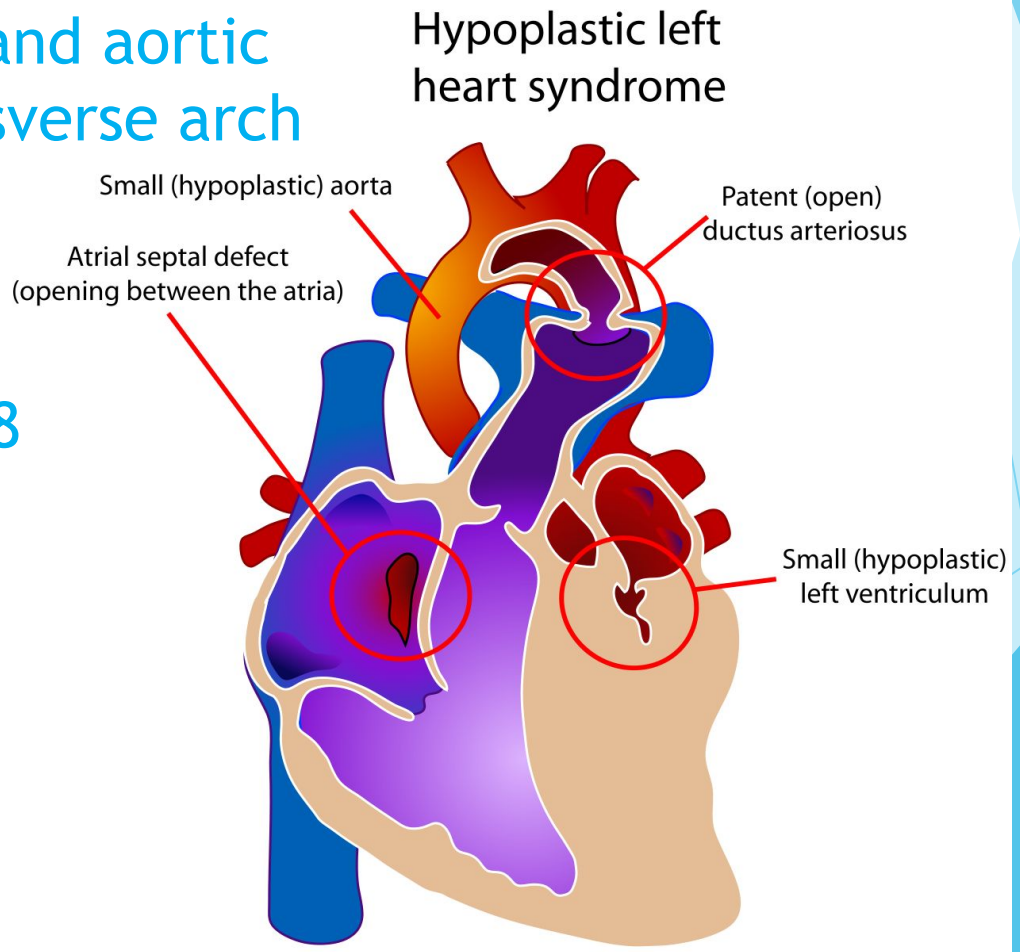
Pathophysiology

At birth, the ductus arteriosus is still open, and there is higher than normal resistance to blood flow in the lungs. This allows for adequate oxygenation via mixing between the atria and a normal appearance at birth. When the ductus begins to close and pulmonary vascular resistance decreases, blood flow through the ductus is restricted and flow to the lungs is increased.

In typical anatomy, the left side of the heart receives oxygen-rich blood from the lungs and pumps it to the rest of the body. In people with HLHS, the aorta and left ventricle are underdeveloped (beginning *in utero*), and the aortic and mitral valves are either too small to allow sufficient blood flow or are atretic (closed) altogether. As blood returns from the lungs to the left atrium, it cannot be pumped to the rest of the body by the left ventricle. The neonate is reliant on blood flowing through an atria septal defect to mix oxygenated and deoxygenated blood, and on a patent ductus arteriosus to allow blood to reach the aorta and the systemic circulation via the right ventricle. This is what defines HLHS as a "single ventricle" defect.

Diagnosis

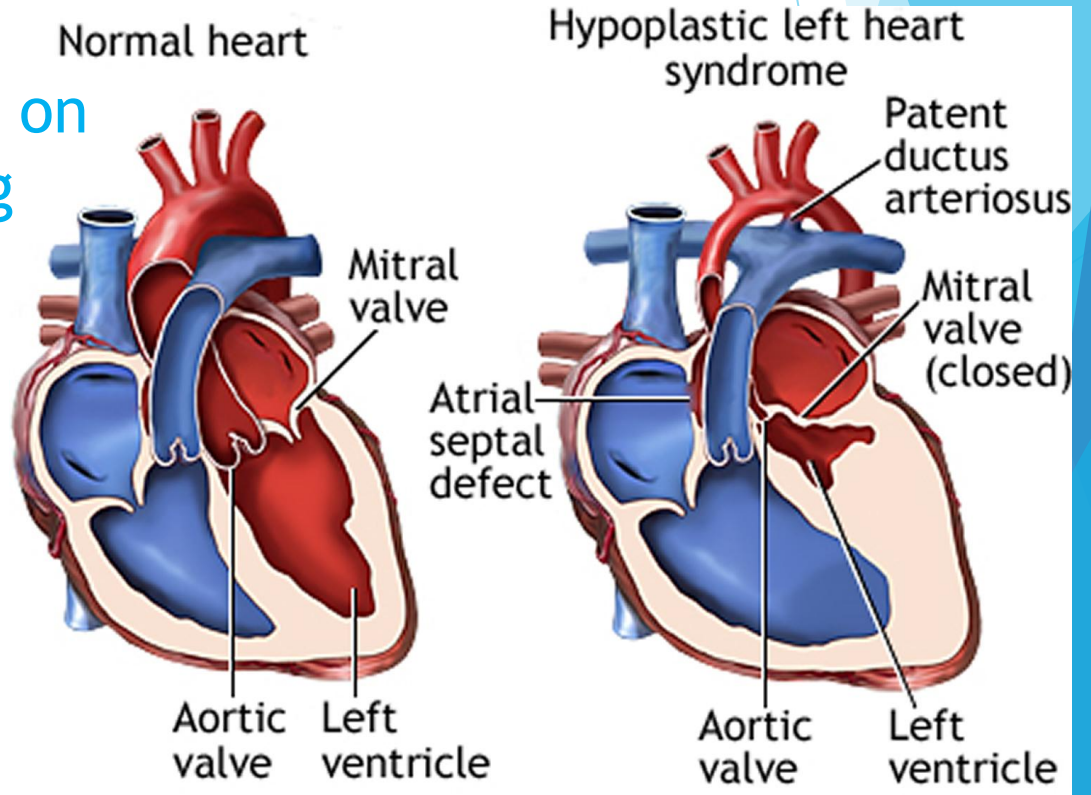
Hypoplastic left heart syndrome can be diagnosed prenatally or after birth via echocardiography. Typical findings include a small left ventricle and aorta, abnormalities of the mitral and aortic valves, retrograde flow in the transverse arch of the aorta, and left-to-right flow between the atria. It is often recognized during the second Trimester of pregnancy, between 18 and 24 weeks' gestation.



Prognosis

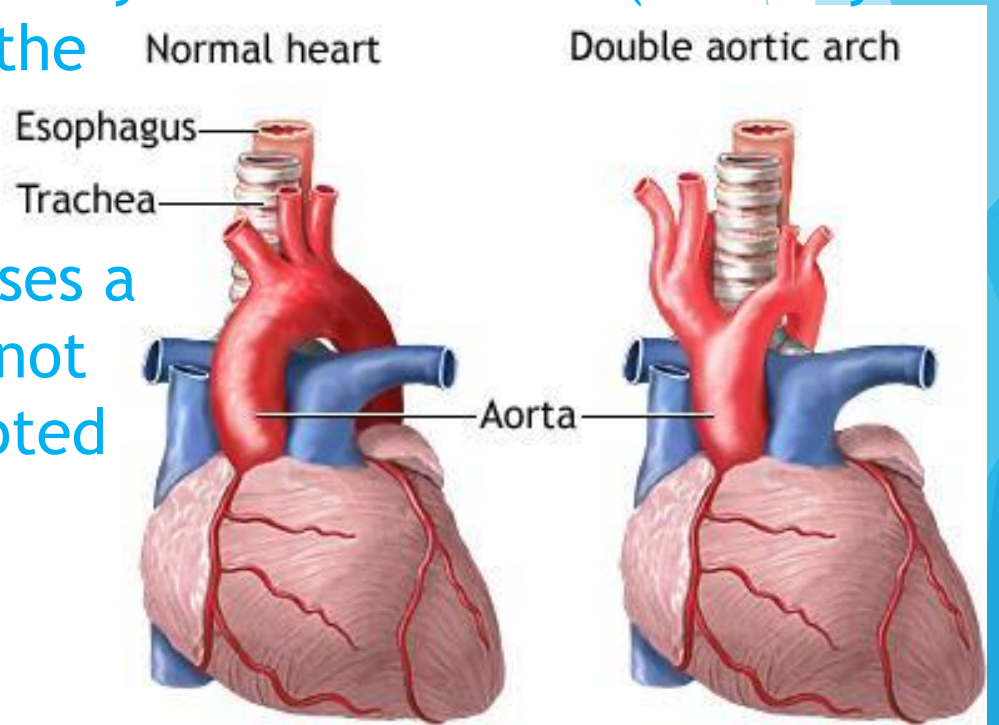
95% of untreated infants with HLHS die in the first weeks of life. Early survival has improved since the introduction of the Norwood procedure. Since there are no long-term studies of HLHS adults, statistics are usually derived from post-Fontan patients; it is estimated that 70% of HLHS patients may reach adulthood.

Prognosis is dependent upon the health of the child, as there is an increased demand on respiratory and heart rate in infants during common childhood illnesses. This fragile population has little cardiac reserve to accommodate these demands and provide hemodynamic stability during illnesses.



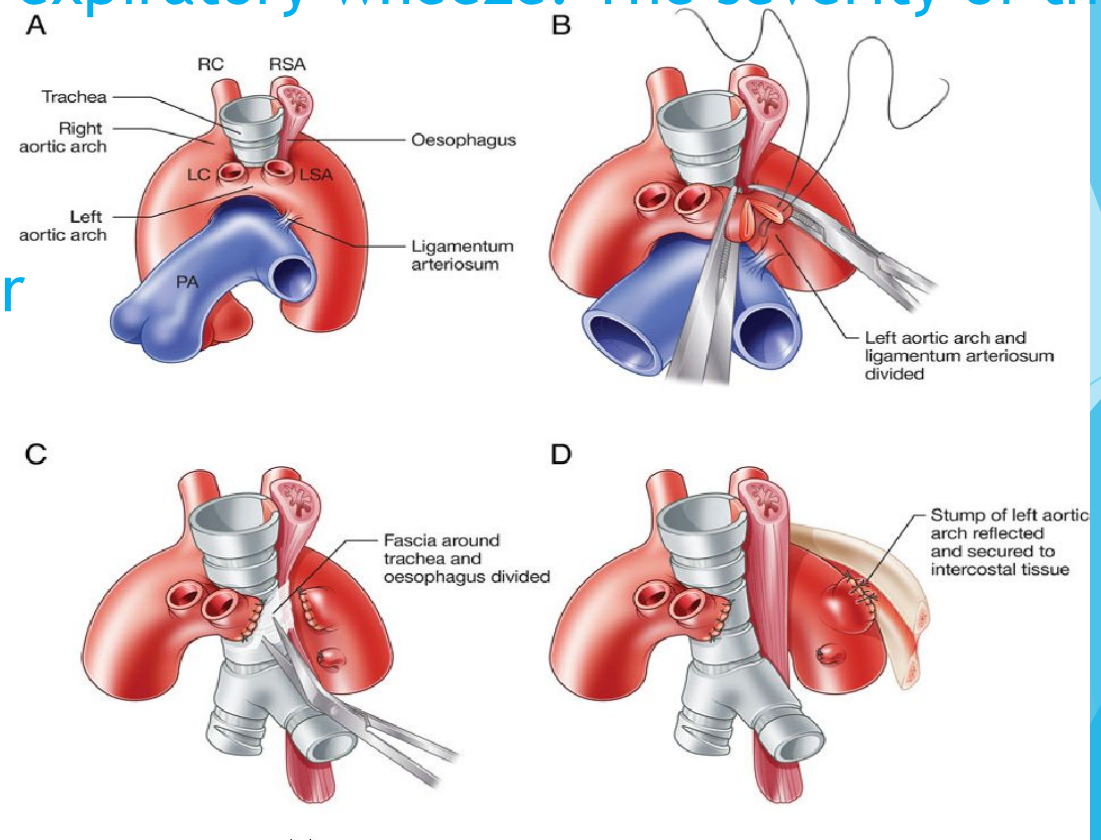
Double Aortic Arch

Double aortic arch is a relatively rare congenital cardiovascular malformation. DAA is an anomaly of the aortic arch in which two aortic arches form a complete vascular ring that can compress the trachea and/or esophagus. Most commonly there is a larger (dominant) right arch behind and a smaller (hypoplastic) left aortic arch in front of the trachea/esophagus. The two arches join to form the descending aorta which is usually on the left side (but may be right-sided or in the midline). In some cases the end of the smaller left aortic arch closes (left atretic arch) and the vascular tissue becomes a fibrous cord. Although in these cases a complete ring of two patent aortic arches is not present, the term ‘vascular ring’ is the accepted generic term even in these anomalies.



Symptoms

Symptoms are caused by vascular compression of the airway, esophagus or both. Presentation is often within the first month (neonatal period) and usually within the first 6 months of life. Starting at birth an inspiratory and expiratory stridor (high pitch noise from turbulent airflow in trachea) may be present often in combination with an expiratory wheeze. The severity of the stridor may depend on the patient's body position. It can be worse when the baby is lying on their back rather than their side. Sometimes the stridor can be relieved by extending the neck (lifting the chin up).



Causes

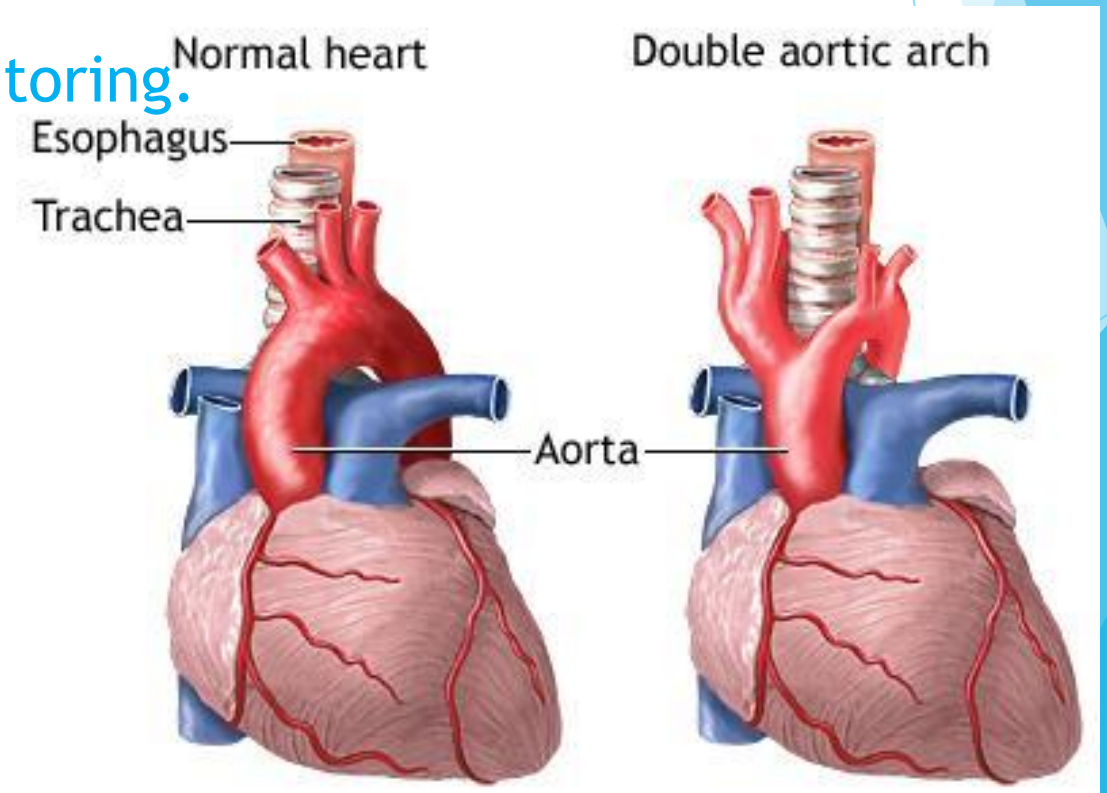
Myocardial ischemia occurs when the blood flow through one or more of your coronary arteries is decreased. The low blood flow decreases the amount of oxygen your heart muscle receives.
Myocardial ischemia can develop slowly as arteries become blocked over time. Or it can occur quickly when an artery becomes blocked suddenly.
Conditions that can cause myocardial ischemia include:

1. Coronary artery disease (atherosclerosis). Plaques made up mostly of cholesterol build up on your artery walls and restrict blood flow. Atherosclerosis is the most common cause of myocardial ischemia.

Treatment

Surgical correction is indicated in all double aortic arch patients with obstructive symptoms (stridor, wheezing, pulmonary infections, poor feeding with choking). If symptoms are absent a conservative approach (watchful waiting) can be reasonable. Children with very mild symptoms may outgrow their symptoms but need regular follow-up.

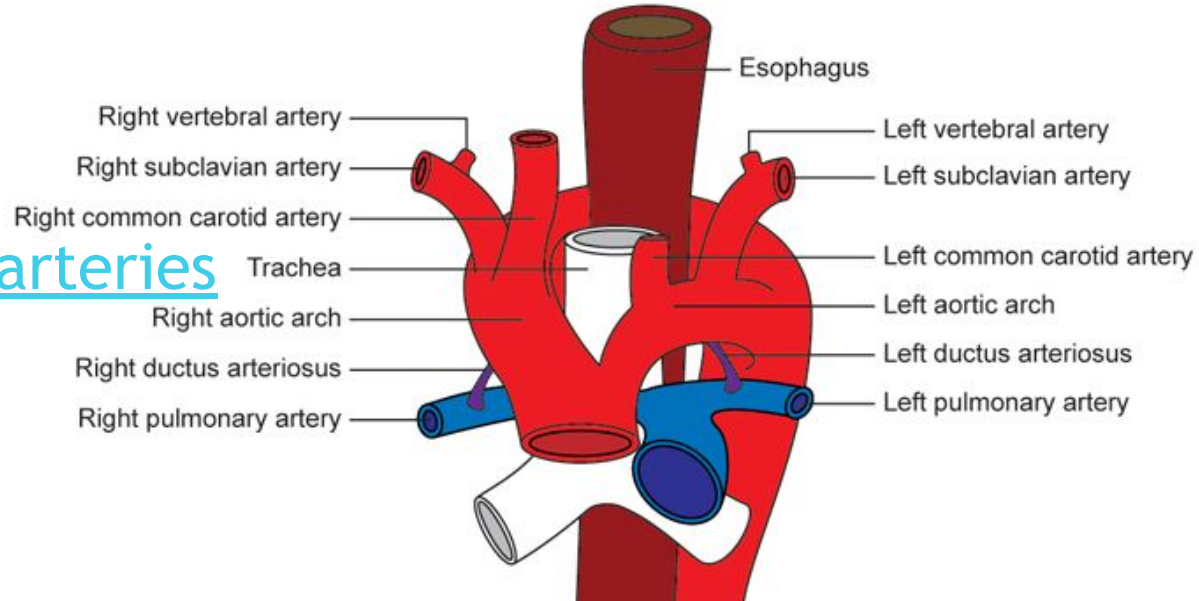
1. Anesthesia and intraoperative monitoring.
2. Open division of vascular ring.
3. Postoperative care.



Epidemiology

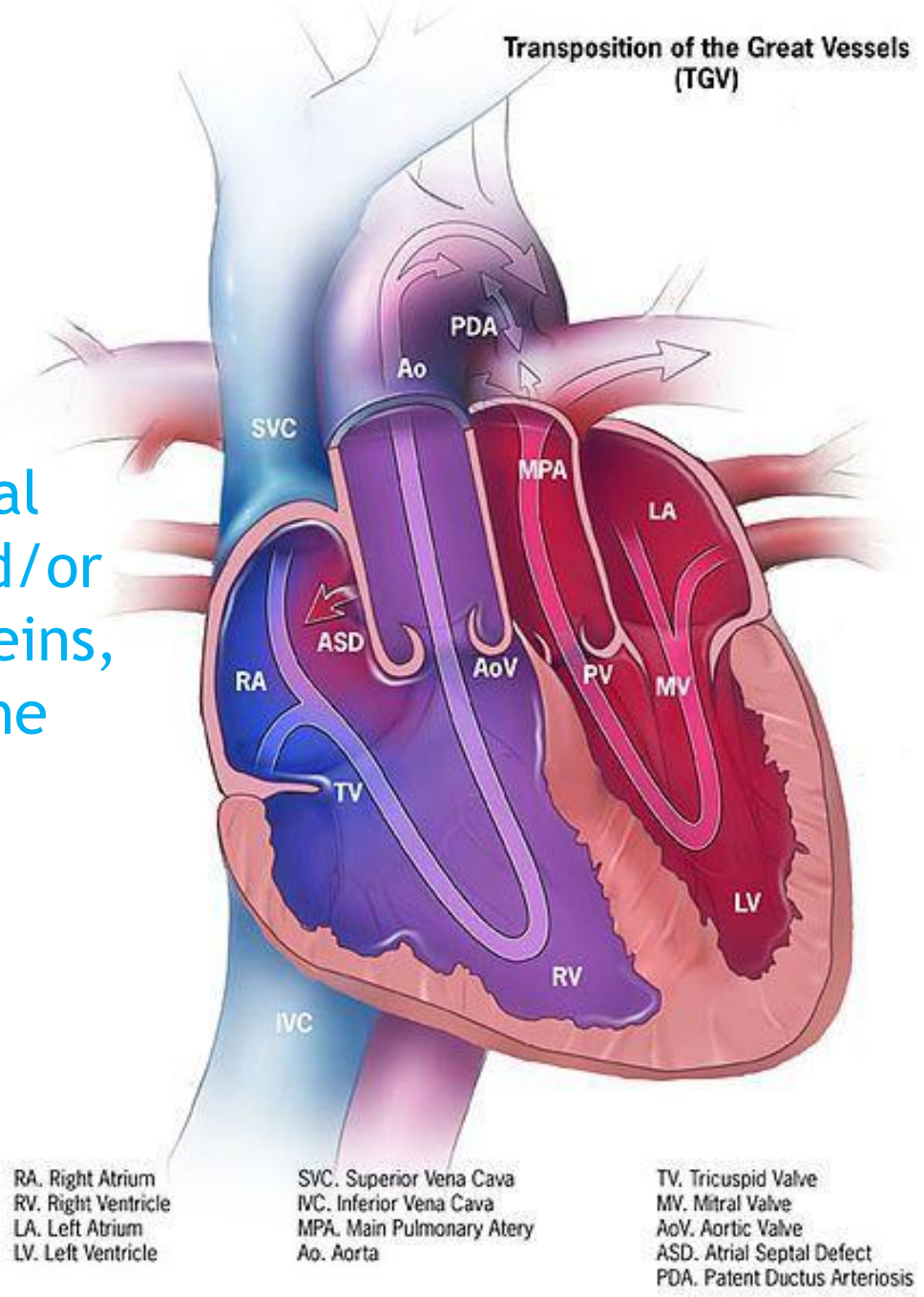
Complete vascular rings represent about 0.5-1% of all congenital cardiovascular malformations. The majority of these are double aortic arches. There is no known gender preference, i.e. males and females are about equally affected. There is also no known ethnic or geographic disposition.
Associated cardiovascular anomalies are found in 10-15% of patients. These include:

Atrial septal defect
Ventricular septal defect
Patent ductus arteriosus
Tetralogy of Fallot
Transposition of the great arteries



Transposition Of The Great Vessels

Transposition of the great vessels (TGV) is a group of Congenital heart defects involving an abnormal spatial arrangement of any of the great vessels: superior and/or inferior venae cavae, pulmonary artery, pulmonary veins, and aorta. Congenital heart diseases involving only the primary arteries (pulmonary artery and aorta) belong to a sub-group called transposition of the great arteries.

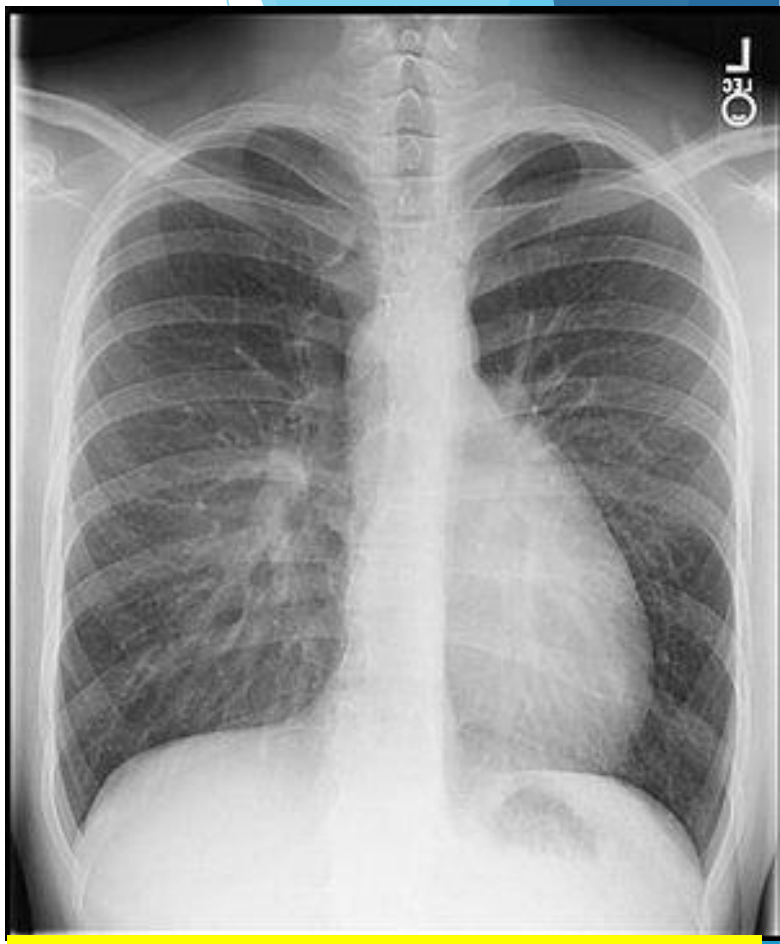


Types

Transposed vessels can present a large variety of atrio venous, ventriculoarterial and/or arteriovenous discordance. The effects may range from a change in blood pressure to an interruption in circulation, depending on the nature and degree of the misplacement and which vessels are involved. Although "transposed" literally means "swapped", many types of TGV involve vessels that are in abnormal positions, while not actually being swapped with each other. The terms TGV and TGA are most commonly used in reference to dextro-TGA – in which the arteries *are* in swapped positions; however, both terms are also commonly used, though to a slightly lesser extent, in reference to levo-TGA – in which both the arteries and the ventricles are swapped; while other defects in this category are almost never referred to by either of these terms.

Diagnosis

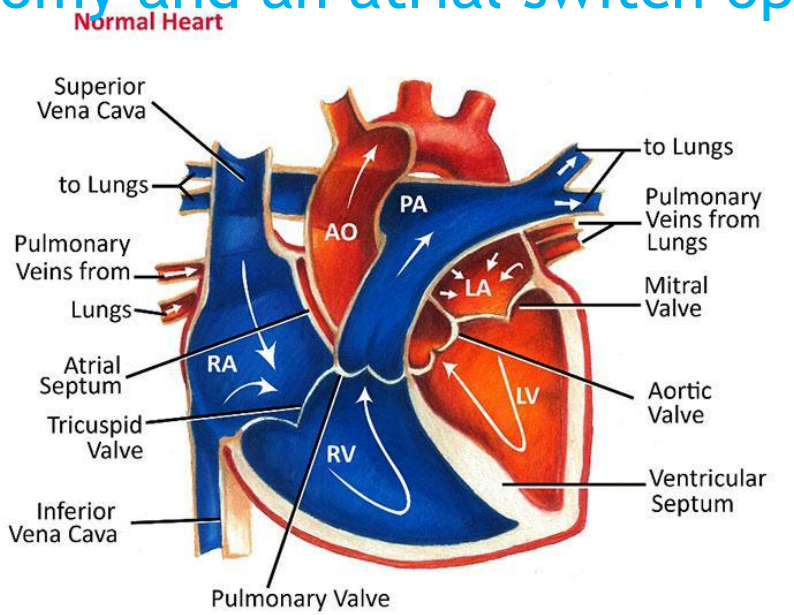
On chest X-ray, transposition of the great vessels typically shows a cardio-mediastinal silhouette appearing as an "*egg on a string*", wherein in which the enlarged heart represents an egg on its side and the narrowed, atrophic thymus of the superior mediastinum represents the string.

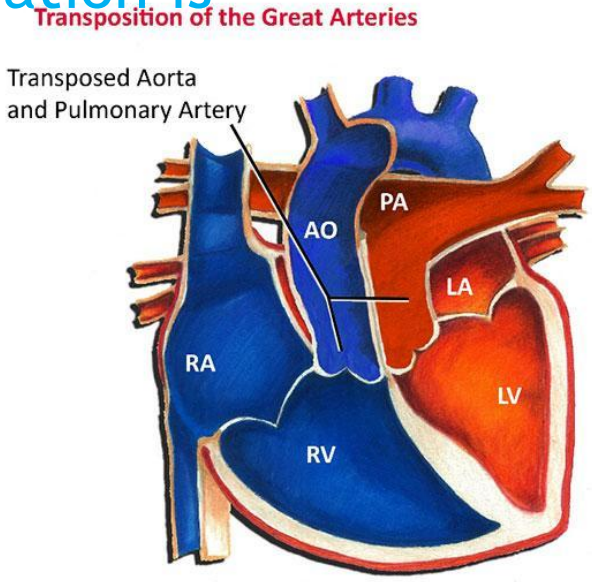


X-ray showing characteristic finding in case of Transposition of the great vessels which is called egg on side sign.

Treatment

For newborns with transposition, prostaglandins can be given to keep the ductus arteriosus open which allows mixing of the otherwise isolated pulmonary and systemic circuits. Thus oxygenated blood that recirculates back to the lungs can mix with blood that circulates throughout the body. The arterial switch operation is the definitive treatment for dextro-transposition. Rarely the arterial switch is not feasible due to particular coronary artery anatomy and an atrial switch operation is preferred.






Thank You