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PULMONARY VALVE ATRESIA AND STENOSIS

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PULMONARY VALVE ATRESIA AND STENOSIS

Condition and Description

Pulmonary valve atresia/stenosis is a rare congenital heart defect where the pulmonary artery that leaves the right ventricle is not open and in addition the right ventricle is very small. The tricuspid valve is typically abnormal and there is a right to left shunt through an opening between the upper two chambers (atria). Blood flows to the lungs through a vessel called a patent ductus arteriosus. There is no known cause for this abnormality that occurs because of a failure of tissues to properly form in the early weeks of fetal life.

Babies with pulmonary valve atresia and stenosis are usually born at term and appear healthy at birth because the heart is not required to provide oxygen to the baby prior to birth (the placenta serves this function before birth). Shortly after birth the ductus arteriosus closes, which is a life-threatening condition for the baby since there is then no way for blood to travel to the lungs to be refreshed with oxygen. The baby will begin to breathe quickly, have a rapid heart beat and be bluish-grey in color (due to cyanosis) because of low levels of oxygen in the blood in the body. Lack of oxygen in the heart muscle and brain is extremely life-threatening, and life-saving measures will be necessary for the baby.

Prevalence

Pulmonary valve atresia is a rare condition, occurring in 7-8 children per 100,000 live births. It makes up about 3% of the congenital heart defects.

Common Associated Conditions

Conditions associated with pulmonary valve atresia include septal defects, tricuspid atresia, proximal pulmonary artery atresia, right atrial dilatation, aortic stenosis, and Ebstein anomaly of the tricuspid leaflets.

Short-term Treatment and Outcomes

Babies will be given a drug (prostaglandin) into the blood stream to keep the ductus arteriosus open so that blood can go through this fetal vessel to the lungs to receive oxygen. Chest x-ray, cardiac catheterization, angiography and other tests will be done to determine the extent of the condition.

Children with pulmonary valve atresia should receive maximum calories so that they will grow as much as possible to help the heart to increase in size before surgery and to ensure that they will be in the best possible health at that time.

Surgery will depend on the nature of the defect, which is unique in each child. Also critical is the structure of the coronary arteries that are often also abnormal; the coronary arteries feed the heart muscle with oxygen-rich blood, essential to feed the heart muscle to keep it functioning well.

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During surgery, the pulmonary valve will be repaired as much as possible, and a stent (a mess tube) may be placed in the ductus arteriosus in order to keep it open. Second stage surgery will typically be necessary, and if the heart grows well during infancy the repair in the second stage will be more successful. If the valve and ventricle do not grow the baby might receive a Fontan surgical procedure (creating a single ventricle to pump the blood). In some cases heart transplantation will be the best option for the child.

Long-term Treatment and Outcomes

Individuals who have had these reconstructive heart surgeries are at risk for later complications of many types (see below). The risk of complications later in life will vary in each situation depending on many individual factors.

Individuals with congenital heart defects will typically require antibiotics when they have dental work because the bacteria in the mouth can circulate through the blood and cause infection in the heart structures (endocarditis). They will need to be followed long-term by cardiologists to be sure that any complications or new conditions are quickly detected and treated. The surgery typically has a 90% survival rate, and many children do very well post-operatively, depending on how many complications and associated defects they have.

Common Complications

Complications are many and include ischemia of the heart muscle (lack of oxygen to that essential tissue), fibrosis of that muscle (replacement of the muscle with scar tissue, also called endocardial fibroelastosis), or infarction (destruction of the tissue of the heart from lack of blood supply), among others.

Implications for Children's Development

The effect of pulmonary valve atresia/stenosis on a child's life can vary greatly depending on the nature and severity of the defects and symptoms. There may be minimal effects and very successful surgery with totally normal development. Other children have considerable difficulty with symptoms that affect growth, energy, resistance to infection and overall physical development. Social and emotional development can be affected by frequent illnesses and hospitalizations.

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