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TRICUSPID VALVE ATRESIA AND STENOSIS

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TRICUSPID VALVE ATRESIA AND STENOSIS

Condition and Description

Tricuspid valve atresia/stenosis is a defect of the heart that occurs in the early weeks of fetal life for an unknown reason. The tricuspid valve fails to develop so blood is unable to enter the right ventricle and that ventricle does not develop normally. The right ventricle is very small. The small right ventricle is supplied with blood through a hole in the heart's wall from the left ventricle (called a ventricular septal defect, VSD). The baby will also have a hole between the upper pumping chambers of the heart (an atrial septal defect, or ASD). Babies will also have an under-developed pulmonary artery and valve within that artery (pulmonary valve). Blood will typically return to the lungs through a pulmonary artery that comes from the aorta (truncus arteriosus) or through another vessel such as the ductus arteriosus. There are several types of tricuspic atresia, depending on the anatomy of the heart and the large vessels. There is a right to left shunt through the ASD, resulting in blood that lacks oxygen going out to the body and to the brain. The baby will have a grayish-blue color from the low oxygen (called cyanosis).

Prevalence

Tricuspid valve atresia/stenosis occurs in 1 of 15,000 live births, and is found in equal numbers in girls and boys.

Common Associated Conditions

Typically, the child will have a number of other heart-related defects in addition to the absence of the tricuspid valve and small ventricle. These might include pulmonary atresia (very small pulmonary artery), pulmonary stenosis (narrowing of the pulmonary artery), ventricular septal defect (VSD, a hole between the 2 lower pumping chambers, the ventricles), or subaortic stenosis (narrowing of the aorta as it leaves the heart chamber).

Short-term Treatment and Outcomes

Diagnosis will be made with echocardiograms, cardiac catheterization and angiograms as well as chest x-rays. Multistage surgery is necessary to redirect blood to the lungs, and once oxygenated, to the body and back to the heart. The surgical plan will be unique to each child since the defects will be somewhat different in each child. The child will be given maximum nutrition so that the heart will grow, and in particular the right ventricle will gradually increase in size so that in subsequent surgeries it can eventually help to pump blood to the lungs. Several new surgical procedures have recently been developed with variable success to redirect blood flow and repair valves. The goal will be to keep the baby's brain and body well supplied with oxygen while protecting the delicate lung tissue from damage from overload of blood with heart failure that causes pulmonary hypertension.

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Long-term Treatment and Outcomes

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

Children with tricuspid atresia can have emboli (clotting of the blood) in various parts of the body, a very serious complication. They can have pulmonary hypertension that damages the tissues in the lungs, which can be permanent. Heart failure can cause slow growth, frequent respiratory infections, fatigue and rapid breathing.

Implications for Children's Development

The effect of tricuspid atresia defect on a child's life can vary greatly depending on how severe the symptoms are. There may be minimal effect and very successful surgery with totally normal development. Other children have considerable difficulty with symptoms that affects 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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