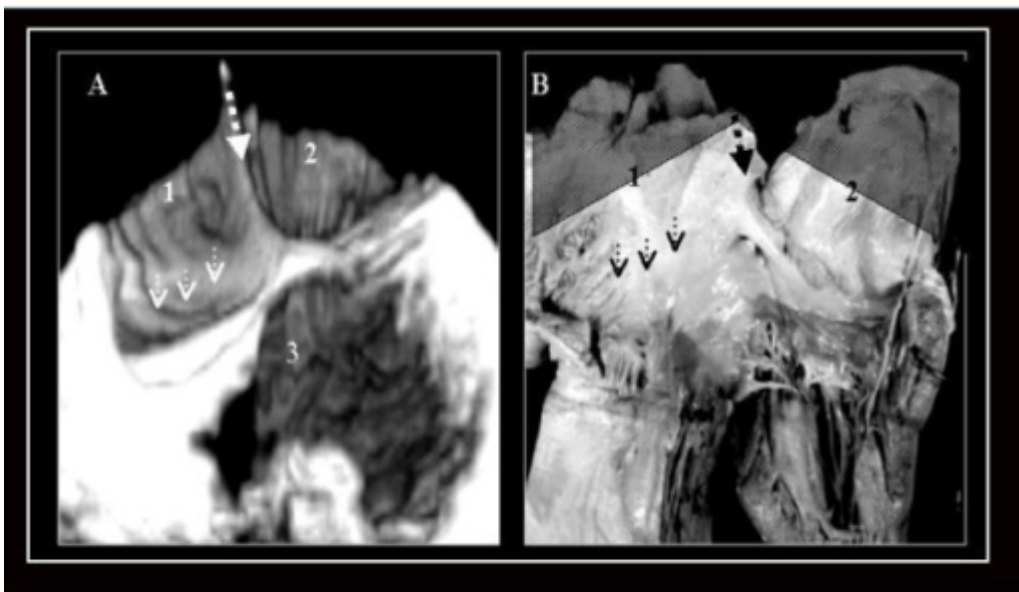


Tricuspid Atresia — Pathophysiology and Treatment

[See online here](#)

Tricuspid atresia is the third most common type of the cyanotic congenital heart diseases, the other two being transposition of the vessels and tetralogy of Fallot. Patients usually present with cyanosis, respiratory distress, syncope, and confusion. Patients with headaches or seizures might have brain abscesses due to the embolism of an infected mural thrombus from the heart to the brain. Polycythemia is commonly seen in patients with tricuspid atresia and is attributed to cyanosis. Echocardiography is the imaging modality of choice for the confirmation of the diagnosis and the exclusion of other cardiovascular abnormalities. Medical treatment of tricuspid atresia includes the administration of prostaglandin E to maintain the patency of the ductus arteriosus. Surgical intervention is indicated in patients with severe cyanosis or with congestive heart failure.



Overview

Tricuspid atresia is one common form of **congenital heart disease that is characterized by the lack of formation of the tricuspid valve**. The absence of the tricuspid valve is associated with the absence of direct communication between the right atrium and ventricle.

Epidemiology of Tricuspid Atresia

Tricuspid atresia is considered the third most common type of cyanotic congenital heart disease and accounts for approximately 3% of all congenital heart disease cases.

Patients with tricuspid atresia are usually symptomatic since birth. Both sexes appear to be affected equally without any significant racial differences.

Mortality of tricuspid atresia is largely dependent on the co-presence of other cardiac or systemic congenital abnormalities. The condition is usually fatal if left untreated.

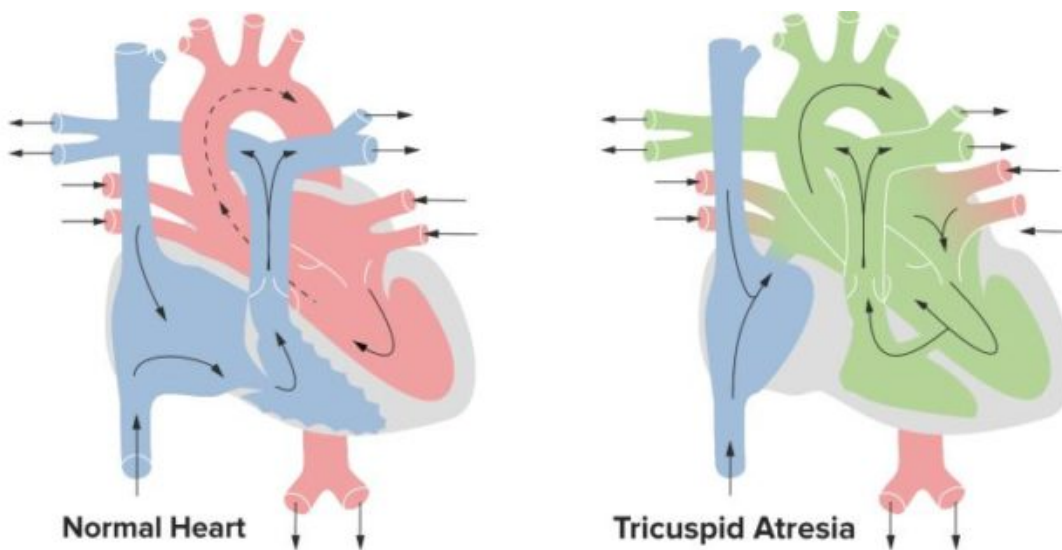
Pathophysiology of Tricuspid Atresia

The exact etiology of tricuspid atresia is unknown, but the condition is **commonly seen in children with other cardiovascular anomalies**. A possible gene that might be indicated with tricuspid atresia is the FOG2 gene. Patients might have ventricular septal defects, pulmonary valvular pathology, or transposition of the great arteries.

Patients with tricuspid atresia do not have any communication between the right atrium and right ventricle. Venous return to the right atrium must escape to the left atrium via an atrial septal defect. Because of the mixing of blood within the left atrium, the oxygen saturation in the systemic circulation is usually diminished.

Volume overload ensues within the left ventricle because the left ventricle receives all the venous return in addition to the oxygenated blood. Because of this, ventricular fibrosis, ventricular dilatation, and mitral valve insufficiency usually complicate the picture.

Cyanosis in patients with tricuspid atresia is depending on the presence or absence of pulmonary artery abnormalities. Patients, who have a normal pulmonary artery and valve usually have adequate oxygenation of the blood and are less likely to be cyanotic. Patients with pulmonary artery atresia, or pulmonary valve stenosis, appear cyanotic.



"Tricuspid Atresia" Image created by Lecturio

Clinical Presentation of Tricuspid Atresia

Patients with tricuspid atresia are usually cyanotic early in life. Almost 50% of patients present clinically on the first day of life. Two third of patients present at the end of the first week and 80% of patients present by the end of the first month. Poor weight gain is a common finding in cyanotic congenital heart diseases and tricuspid atresia is not an exception. Due to volume overload, systolic left ventricular dysfunction is common and patients might develop congestive heart failure.

Affected infants might be cyanotic or appear pale to the parents. Poor feeding habits are also commonly seen. Patients might present with fever, lethargy, neurological deficits or seizures which are suggestive of a brain abscess. Infective endocarditis is also common in patients with tricuspid atresia.

Digital clubbing is commonly seen in cyanotic heart disease, including tricuspid atresia. A left ventricular impulse can be felt on the chest examination. Patients with congestive heart failure might have a pulsatile enlarged liver and a distended jugular vein.

Diagnostic Workup for Tricuspid Atresia

A complete blood count is indicated in patients presenting with cyanosis. Polycythemia is commonly seen. Prothrombin and activated partial thromboplastin time might be abnormal in patients with tricuspid atresia and polycythemia.

A chest X-ray usually demonstrates cardiomegaly. In most patients with tricuspid atresia, pulmonary blood flow is decreased; therefore, pulmonary vascular markings are usually diminished.

The imaging modality of choice for establishing the diagnosis of tricuspid atresia is echocardiography. Echocardiography can help in the assessment of the presence or absence of the tricuspid valve in a cyanotic patient, identify other cardiovascular abnormalities and help with the assessment of the left ventricle systolic function. Other investigations include ECG, pulse oximetry to observe oxygen saturation, radioisotopic scans, and cardiac catheterization.

Treatment of Tricuspid Atresia

The treatment of tricuspid atresia is largely dependent on the degree of cyanosis and hypoxemia in the patient. Infants who are cyanotic and have severe hypoxemia should receive prostaglandin E infusions to maintain the patency of the ductus arteriosus. The importance of maintaining a patent ductus arteriosus in patients with tricuspid atresia is further emphasized by the finding that cyanosis usually worsens once the ductus arteriosus starts closing 48 hours after birth.

Patients with severe cyanosis and hypoxemia also have acidosis. Correction of metabolic acidosis might be needed as it has been linked with increased mortality.

Patients with tricuspid atresia that is also complicated by transposition of the great arteries and a large ventricular septal defect might develop overt congestive heart failure. In this group of patients, digoxin and diuretic therapy for congestive heart failure is indicated. Early surgical intervention is recommended in this group of patients.

Surgical intervention for tricuspid atresia depends on the goals of the surgeon. When the goal is to help with cyanosis and decreased pulmonary blood flow, a shunt procedure to

allow for a passive venous return to the pulmonary circulation is recommended. The shunt can connect the subclavian artery to the pulmonary artery. Patients with severe congestive heart failure due to increased pulmonary blood flow need a surgical procedure to restrict the pulmonary arterial blood flow. The procedure of choice in this cohort of patients is pulmonary artery banding.

The risk of recurrence of cyanosis after these procedures is high. Patients with progressive polycythemia decreased exercise tolerance and recurrent cyanosis are possible candidates for re-operation. A Fontan procedure is recommended at this stage with the aim to connect the right atrium directly to the pulmonary artery while completely bypassing the right ventricle. Unfortunately, this option is not readily available for all patients with tricuspid atresia.

The Fontan operation should be performed only on children older than 4 years of age, who have a normal right atrium volume, who have a normal sinus rhythm, and who have a pulmonary artery-to-aorta diameter that is above 75%. The mean pulmonary arterial pressure should be less than 15 mm Hg.

Unfortunately, most procedures that have been described for tricuspid atresia do not eliminate the risk of progressive polycythemia, brain abscesses, and embolic disease. Fortunately, though, patients nowadays can survive until their early adulthood.

Patients with congestive heart failure and volume overload should be put on a low-salt diet. Older children with tricuspid atresia are very likely to have congestive heart failure and significantly decreased exercise tolerance. Despite this, exercise should always be encouraged even though the child might seem tired and out of breath.

Prognosis

Untreated cases have a poor prognosis. Only 10-20% of cases remain alive until the first year of life.

Surgical palliation increases the chances of survival. The Fontan procedure improves the prognosis. Adults who had undergone this procedure initially show high mortality and morbidity due to re-operation, arrhythmias, and thromboembolic events. Patients with total cavo-pulmonary connection show less morbidity and a good survival rate.

References

Mancini M. Tricuspid Atresia. Background, Pathophysiology, Epidemiology. <http://emedicine.medscape.com/article/158359>. Published January 6, 2017. Accessed March 14, 2017.

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