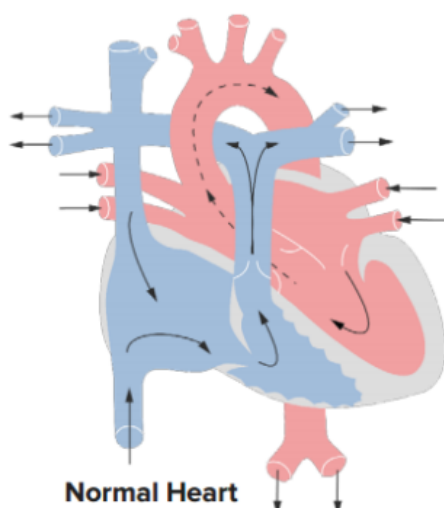


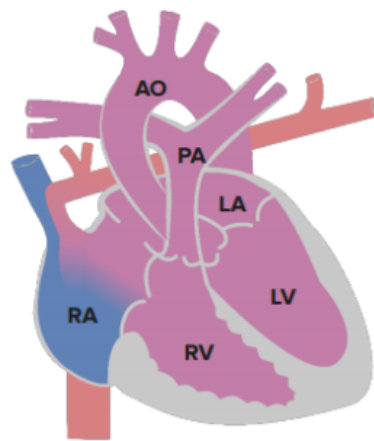
Total Anomalous Pulmonary Venous Connection (TAPVR) — Pathophysiology and Surgery

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Patients with a total anomalous pulmonary venous connection (TAPVC) have abnormally draining pulmonary veins that drain into the right atrium or the systemic venous circulation. Patients are usually symptomatic early in life. If left untreated, the condition is usually fatal. Echocardiography and magnetic resonance imaging are helpful in the confirmation of the diagnosis. Surgical correction for redirection of the pulmonary veins drainage to the left atrium is the mainstay of treatment.



Normal Heart



Total anomalous pulmonary venous return

Definition

Total Anomalous Pulmonary Venous Connection (TAPVC) is a congenital heart abnormality that is characterized by abnormal blood flow, where the pulmonary veins drain into the right atrium or the systemic venous circulation.

Overview of the heart circulation

Blood from the peripheral circulation enters the heart through the vena cava into the right atrium and then the right ventricle. Later, the heart pumps blood through the pulmonary circulation into the lungs for oxygenation. Oxygenated blood travels through the pulmonary vein into the left atrium and left ventricle, where it is pumped to the peripheral circulation via the aorta.

In TAPVC, the oxygenated blood either empties into the right atrium or the pulmonary artery. Thus, there are two separate circulation systems, leading to poor vascularization

of peripheral tissues and cyanosis.

For the patient to survive during the fetal and the neonatal period, the foramen ovale between the two atria must be patent to allow for surgical correction of the defect.

Epidemiology of Total Anomalous Pulmonary Venous Connection

The prevalence of TAPVC is estimated to be around 6.8 per 100,000 in live births. The incidence of TAPVC in patients with congenital heart defects is estimated at 1.5%.

Sex differences are still under debate. While the Baltimore-Washington Infant Study showed a very small female predominance, other studies have shown a significant male predominance of up to 3:1 male to female ratio.

Pathophysiology of Total Anomalous Pulmonary Venous Connection

Patients with TAPVC have a volume overload on the right atrium and right ventricle. When the foramen ovale is restrictive, right atrial pressure increases. Patients might develop venous systemic and pulmonary congestion. Additionally, blood flow to the pulmonary circulation usually increases, and patients are at risk of developing pulmonary arterial hypertension.

The blood flow to the left atrium and left ventricle depends on the foramen ovale's patency and restrictiveness. Therefore, the left side of the heart receives a lower than normal blood volume, and the cardiac index is usually reduced.

Patients with TAPVC might have normal blood oxygenation by increasing the pulmonary blood flow by three to four times.

Clinical Presentation of Total Anomalous Pulmonary Venous Connection

The presentation of TAPVC is dependent on whether the patient has pulmonary vein obstruction or not. Patients who have the site of drainage of the pulmonary veins below the diaphragm (infracardiac) or above the heart (supracardiac) are very likely to have some degree of pulmonary venous obstruction. These patients are usually symptomatic during the first days of life. They might develop tachypnea, tachycardia, and cyanosis.

Pulmonary hypertension, which is characterized by a worsening of cyanosis, can occur very early in these patients. If left untreated, the neonate usually dies within the first month of life. A physical examination usually reveals a prominent right ventricular impulse. The heart is usually not enlarged, and the apex beat is not displaced. Liver enlargement is common in this group of patients. Finally, patients might have an increased pulmonary component of the second heart sound.

Patients who do not have pulmonary vein obstruction usually have milder symptoms. Failure to thrive and recurrent respiratory tract infections, similar to large atrial septal defects presentation, are common. Patients might also have difficulty feeding.

A physical examination usually reveals a wide split in the second heart sound but with a normal pulmonary component. Patients might be cyanotic, but this is often less common than it would be in the previously described cohort of patients.

If the foramen ovale becomes more restrictive to blood flow, pulmonary hypertension might develop. In that case, the patient might develop signs and symptoms suggestive of right-sided heart failure, i.e., systemic and pulmonary venous congestion.

Diagnostic Workup for Total Anomalous Pulmonary Venous Connection

Determining the peripheral blood oxygenation level is essential in the diagnostic workup of the neonate or infant who presents due to TAPVC. Patients are also at risk of developing acid-base imbalances, and their arterial blood gases should be examined.

The main finding of TAPVC on electrocardiography is that of right ventricular hypertrophy. A chest X-ray might show a normal heart with diffuse reticular patterns from the hilum. Patients with an unobstructed pulmonary venous return to the right heart might have cardiomegaly on their chest X-ray due to right atrial and right ventricular enlargement. Patients with pulmonary congestion might have pulmonary edema.

Echocardiography usually provides definitive evidence for TAPVC diagnosis. Color Doppler studies are usually indicated to evaluate the pulmonary veins and to see if they drain within the left atrium. Additionally, echocardiography can help delineate the restrictiveness of the patent foramen ovale. Fetal echocardiography can also be used to confirm the diagnosis in a high-risk fetus, i.e., a positive family history of congenital heart disease in general or TAPVC in particular.

Magnetic resonance imaging is superior to echocardiography in that it is not operator-dependent and can also help evaluate associated lung diseases, such as lymphatic congestion.

Patients with multiple intracardiac and extracardiac sites of pulmonary venous return might benefit from cardiac catheterization.

Treatment of Total Anomalous Pulmonary Venous Connection

If early surgical correction is not possible, an atrial septostomy might be performed, especially if the patient has a restricted foramen ovale. Patients with severe obstruction to pulmonary venous return might benefit from stent placement until a definitive surgical correction is possible.

The main goal of surgical treatment for TAPVC is to restore the normal relationship between the pulmonary veins and the left atrium. Patients with extracardiac TAPVC should have their common pulmonary vein opened and then connected, side-to-side, to the left atrium.

Patients who have intracardiac TAPVC, i.e., draining into the right atrium or the coronary sinus, should undergo a partial resection of the atrial septum. After the resection of the atrial septum, the patients should get a new atrial septum that is the site of the entrance of the pulmonary vein, i.e., to direct the pulmonary veins to the left atrium. In other

words, the left atrium now consists of the normal left atrium in addition to the part of the right atrium, where the pulmonary veins are draining.

After directing the pulmonary veins to the left atrium, the foramen ovale should be closed. Patients with multiple extracardiac drainage sites should undergo a side-to-side procedure to connect the left atrium with the common pulmonary artery, followed by ligation of the other pulmonary veins.

Postoperatively, the patient might develop episodic pulmonary hypertension, which might harm cardiac output. In that case, nitric oxide should be used for its pulmonary dilator effects. Extracorporeal membrane oxygenation might be needed in acutely ill patients with poor blood oxygenation.

Newborns who are too ill to undergo surgical intervention should receive diuretics to relieve pulmonary edema, and surgical relief of the obstructed TAPVC should be offered as soon as possible.

References

Wilson A. Total Anomalous Pulmonary Venous Connection. Background, Pathophysiology, Epidemiology. <http://emedicine.medscape.com/article/899491>. Published January 6, 2017. Accessed March 14, 2017.

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