

Pulmonary venous return anomaly

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Abstract

Pulmonary venous return anomaly consists of partial ([PAPVC](#)) and total ([TAPVC](#)) anomalous pulmonary venous connections. These are partial and total forms of pulmonary venous drainage into the systemic venous system. PAPVC account for 0.5% of the congenital cardiac defects and are commonly associated with atrial septal defects (ASD). Clinical symptoms of PAPVC are similar to those triggered by ASDs with normal PV connection. Depending on the shunt volume, children are either asymptomatic or show failure to thrive, increased number of respiratory infections and limitation of physical exercise. Diagnosis can be usually made by transthoracic or transoesophageal echocardiography. Elective surgery is favoured in preschool age with patch closure of the ASD and redirection of the abnormally draining PV into the left atrium. Long-term results are excellent. TAPVC accounts for 1-2% of all congenital cardiac malformations. In that case, all PV usually drain into a common PV sinus. Clinical course depends on the patency of the interatrial communication and on the presence of an obstruction of the pulmonary venous return (PVO). Patients without PVO mostly present within the first weeks of life with signs of cardiac insufficiency as tachypnea, dyspnea, hepatomegaly and discrete but increasing cyanosis. Diagnosis is usually made on echocardiography also. Both PAPVC and TAPVC are due to a morphological malformation. A familial form with autosomal dominant inheritance has been reported. Curative surgery in neonatal age or early infancy is required and should provide a wide anastomosis between PV sinus and left atrium. Late results are excellent in most cases.

Keywords

Congenital cardiac defect, ventricular septal defect, cardiac insufficiency, anomalous pulmonary venous, connections failure to thrive, respiratory infections

Definition

Pulmonary venous return anomaly consists of partial (PAPVC) and total (TAPVC) anomalous

pulmonary venous connections. These are partial and total forms of pulmonary venous drainage into the systemic venous system.

Since frequency, clinical signs, diagnosis, treatment are different depending on the type of anomalous pulmonary venous connection, the 2 entities will be dealt with separately.

Partial anomalous pulmonary venous connection (PAPVC)

Frequency

PAPVC account for about 0.5% of the congenital cardiac defects and are commonly associated with atrial septal defects (ASD) which comprise 7-10% of cardiac malformations. About 90% of sinus venous defects and about 25% of ostium secundum defects present with PAPVC. Anomalous insertion of one or more pulmonary vein(s) (PV), predominantly of the upper right PV into the superior caval vein, contributes to left-to-right shunt resulting in an elevated ratio of pulmonary to systemic flow. The scimitar syndrome is a rare entity associating a PAPVC of the right PV into the inferior caval vein with hypoplasia of the right lung.

Clinical signs

Clinical symptoms of PAPVC are similar to those triggered by ASDs with normal PV connection. Depending on the shunt volume, children are either asymptomatic or show failure to thrive, increased number of respiratory infections and limitation of physical exercise. In cases of hemodynamically relevant left-to-right shunt, that is usually caused by the complete abnormal return of all veins of one lung, widely and fixedly split 2nd heart sound and a systolic murmur due to relative pulmonary stenosis and diastolic murmur due to relative tricuspid stenosis are typical symptoms.

Etiology

PAPVC is due to a morphological malformation in early embryological development with partial disconnection of the canalizing pulmonary vein and the pulmonary venous plexus. Anastomoses between unconnected pulmonary segments and the systemic venous plexus persist and develop. Most cases occur sporadically.

Diagnosis

Electrocardiography may show right axis deviation and right ventricular hypertrophy. Chest X-rays demonstrate cardiomegaly with prominent right atrium, right ventricle and dilated pulmonary vessels. Diagnosis can be usually made by transthoracic or transoesophageal echocardiography with colour flow mapping. Cardiac catheterization with angiocardiography is only required if the exact site of the PAPVC has not been determined.

Treatment

Elective surgery is favoured in preschool age with patch closure of the ASD and redirection of the abnormally draining PV into the left atrium. Long-term results are excellent. Increased risk of atrial arrhythmias must be considered.

Total anomalous pulmonary venous connection (TAPVC)

Frequency

TAPVC accounts for 1-2% of all congenital cardiac malformations. In that case, all PV usually drain into a common PV sinus. Depending on the site of drainage of the PV sinus, 3 types of TAPVC have been delineated: the supracardiac type (about 50% of TAPVC) draining either into the left brachiocephalic vein, into the right superior caval vein, into the azygos system of veins, or into the left superior caval vein; the cardiac type (about 25% of TAPVC) draining into the right atrium; and the infracardiac or infradiaphragmatic type (about 25% of TAPVC) draining into the inferior caval vein. Mixed types exist (3-5%).

An ASD or a patent foramen ovale allowing a right-to-left shunt is necessary for survival.

Clinical signs

Clinical course depends on the patency of the interatrial communication and on the presence of an obstruction of the pulmonary venous return (PVO). Patients without PVO mostly present within the first weeks of life with signs of cardiac insufficiency as tachypnea, dyspnea, hepatomegaly and discrete but increasing cyanosis.

Functional PVO is present in about 33% of TAPVC. Stenosis may occur at any anatomical site between the common PV sinus and the venous connection, or be due to a restrictive ASD. PVO are more frequent in the infracardiac and supracardiac types than in the cardiac type. PVO causes severe pulmonary venous and pulmonary arterial hypertension leading to right ventricular failure, hypoxemia and metabolic acidosis.

Neonates with TAPVC and PVO present with severe cyanosis and respiratory distress, while those without PVO usually have mild cyanosis without respiratory signs. The natural course of TAPVC with PVO is fatal.

Etiology

TAPVC is due to a morphological malformation in early embryological development with disconnection of the 4 PV to the left atrium, caused by either agenesis, involution, or atresia of the embryological common PV and persistent connections between pulmonary and systemic veins. Supracardiac and cardiac forms are

predominant in males, whereas infracardiac forms affect males and females equally. A familial form with autosomal dominant inheritance has been described.

Diagnosis

Electrocardiography shows right atrial and right ventricular hypertrophy. Chest X-rays show, in cases with PVO, a small cardiac silhouette with reticular appearance or ground-glass opacification of both lungs. In cases without PVO, cardiomegaly and a prominent pulmonary trunk are present. Diagnosis is usually made on echocardiography with colour flow mapping. Cardiac catheterization and angiocardiography might be necessary in complex cases associating mixed forms.

Treatment

Curative surgery in neonatal age or early infancy is required and should provide a wide

anastomosis between PV sinus and left atrium. Late results are excellent, except for occasional late PVO manifesting after 4-6 weeks in about 10% of the cases. These patients require re-operation or intervention with an increased risk of recurrence or mortality.

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