

ANOMALOUS PV CONNECTIONS, AV MALFORMATIONS, CORONARY AND LV ABNORMALITIES

Total Anomalous Pulmonary Venous Connection (TAPVC)

In total anomalous pulmonary venous connection (TAPVC) all 4 pulmonary veins drain into systemic veins or the right atrium with or without pulmonary venous obstruction. Systemic and pulmonary venous blood mix in the right atrium. A VSD or ASD allows flow to the left atrium and a PDA may be present. Diagnosis of most patients occurs in early infancy.

Connections have been classified to *supracardiac* (the 4 pulmonary veins drain via a common vein into the right superior vena cava, left superior vena cava, or their tributaries), *cardiac* (the pulmonary veins connect directly to the right heart eg coronary sinus or directly to the right atrium), *infradiaphragmatic* (the common pulmonary vein is anterior to the esophagus through the diaphragm to connect to the portal venous system) and *mixed* (the right and left pulmonary veins drain to different sites eg, left pulmonary veins into the left vertical vein to the left innominate, right pulmonary veins directly into the right atrium or coronary sinus). The degree of cyanosis in the neonate depends on the degree of venous obstruction and the size of the ASD.

Without surgery up to 90% of patients die within the first year of life. Surgical mortality in experienced centers is <10% and three-year survival exceeds 85%.¹ Most patients survive surgery without late cardiovascular problem manifestations. An increased incidence of neurodevelopmental difficulties has been reported, and there is a risk for recurrent pulmonary vein stenosis or anastomosis at the anastomosis site between the common pulmonary vein and the left atrium. Postoperative pulmonary vein obstruction may occur in patients with hypoplastic/stenotic pulmonary veins.¹ The sutureless technique has gained favor as a prophylactic method recently.²

Partial Anomalous Pulmonary Venous Connection (PAPVC)

Part or all of one lung drains into systemic veins or the right atrium. Sinus venosus defects have PAPVC from the right upper and middle lobe pulmonary veins to the SVC. PAPVC to the IVC (*scimitar syndrome*) may be associated with a hypoplastic right lung, pulmonary sequestration and ASD. Signs and symptoms resemble these of a secundum ASD (Table 1). Surgical correction is indicated in the presence of RV overload (Table 2). Follow-up in Table 3.

Pulmonary Arteriovenous Malformations

They are abnormal connections between branches of the pulmonary arterial and pulmonary venous system. They are usually part of the hereditary hemorrhagic telangiectasia syndrome, an autosomal-dominant disorder, but may also occur as isolated or multiple defects secondary to trauma or infection.³ They may create a pulmonary right-to-left shunt because the desaturated blood bypasses the oxygenation mechanisms at the alveolar level. In the presence of clinical cyanosis, transcatheter embolization is the treatment of choice. Surgical lung resections are rarely required.

Aneurysms of the Pulmonary Artery

The upper limit of the main PA in adults is 29 mm, and the upper interlobar PA is 17 mm. Diameters exceeding those are accepted as indicating aneurysms, but a number of 40 mm has also been used.⁴ They are rare abnormalities usually seen in congenital heart disease and left to right shunts such as PDA, VSD and ASD. Infectious causes such as syphilis and tuberculosis, vasculitis, pulmonary hypertension and chronic embolism, and neoplasms are other causes, whereas idiopathic formation is rare. Aneurysms are usually asymptomatic. Dyspnea, chest pain, hoarseness, cough and hemoptysis may be seen with aneurysms exceeding 70 mm. A systolic murmur is usually heard and on chest X ray reveals a hilar enlargement, or pulmonary mass. Diagnosis is established by contrast-enhanced CT. Surgery is indicated with a diameter ≥ 55 mm or increase in the diameter ≥ 5 mm in 6 months.⁴

Congenital Coronary Anomalies

Congenital anomalous origin of the coronary arteries may occur in 1-5% of all coronary angiograms performed, with 0.15% of them having the highest-risk lesions of the left coronary artery arising from the right sinus of Valsalva.^{3,5} Interarterial (ie between aorta and pulmonary artery) anomalous origin of the left coronary artery is much rarer than interarterial anomalous origin of the right, but carries a risk of sudden cardiac death.⁶ In 80% of autopsies in athletes with sudden cardiac death and anomalous coronary artery origins, the affected coronary artery coursed between the aorta and the pulmonary artery.³ An anomalous pulmonary origin of coronary arteries is generally considered malignant.⁵ However, in a recent study on middle-aged individuals with newly diagnosed anomalous origin of a coronary artery from the opposite sinus, mid-term outcome was not statistically different to a matched control cohort without coronary artery anomalies, regardless of whether interarterial course variants were present.⁷ Anomalous origin without an interarterial or interseptal course (ie anterior or posterior to aorta) definitely does not indicate adverse prognosis (Tables 4-9 and Figure 1).⁸ Restriction from competitive sports is recommended in patients with

interarterial anomalous left coronary artery.⁶ A history of aborted sudden cardiac death, documented ischemia, exertional syncope or chest pain are ominous prognostic signs, and surgical treatment is necessary. Myocardial bridges (intramyocardial course of the coronary with systolic compression) (Table 9) occur in 1% of coronary angiograms and requires treatment when causing ischemia.³ CABG is the treatment of choice although stents have also been successfully tried.

Coronary Fistulas

Communications between the coronary arteries and the cardiac chambers (coronary-cameral fistulas) or low-pressure veins (coronary arteriovenous malformations) are most often congenital in nature (usually to RV, RA or PAs).⁹ They also may be acquired secondary to trauma or from invasive cardiac procedures such as pacemaker implantation, endomyocardial biopsy, coronary artery bypass grafting, or coronary angiography. With larger fistulas, a diastolic runoff may occur, drawing blood away from the normal coronary pathway with a widened pulse pressure and a coronary steal, thus creating a left to right or left to left shunt. A continuous murmur may be audible at the left lower sternal border. Most coronary artery fistulas are small, and patients are asymptomatic. Small coronary fistulas in children tend to grow with age and may cause clinical symptoms such as angina or congestive heart failure and rarely endocarditis or fistula rupture in up to 60% of older patients. Large, hemodynamically significant fistulas can be closed electively either by surgery on a beating heart from the epicardial surface or through transcatheter occlusion techniques.

Vascular Rings

Vascular rings are rare and are typically diagnosed in infancy or early childhood, but they may be first identified in adulthood. Adult presentation is typically after years of unexplained respiratory and/or gastrointestinal symptoms, or the condition may be found incidentally in an asymptomatic adult.¹⁰ Symptoms can include dyspnea, wheezing, stridor, and dysphagia. Recommendations are provided in Tables 10 and 11.

Left Ventricular Protrusions

Congenital left ventricular outpouchings include diverticula, aneurysms, and hernias and can be seen in up to 0.8% of patients undergoing cardiac catheterization and found to have normal coronary arteries.¹¹ Diverticula have a thick wall made up of histological normal heart wall tissue and are connected to the main left ventricular chamber through a narrow neck, whereas aneurysms are thin walled, fibrotic, noncontractile, and have a wide communication with the main chamber. Cardiac hernias are defined as myocardium protruding through a pericardial defect. Left ventricular protrusions can be a source of embolic stroke.

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