
Anatomy of Total Anomalous Pulmonary Venous Connection

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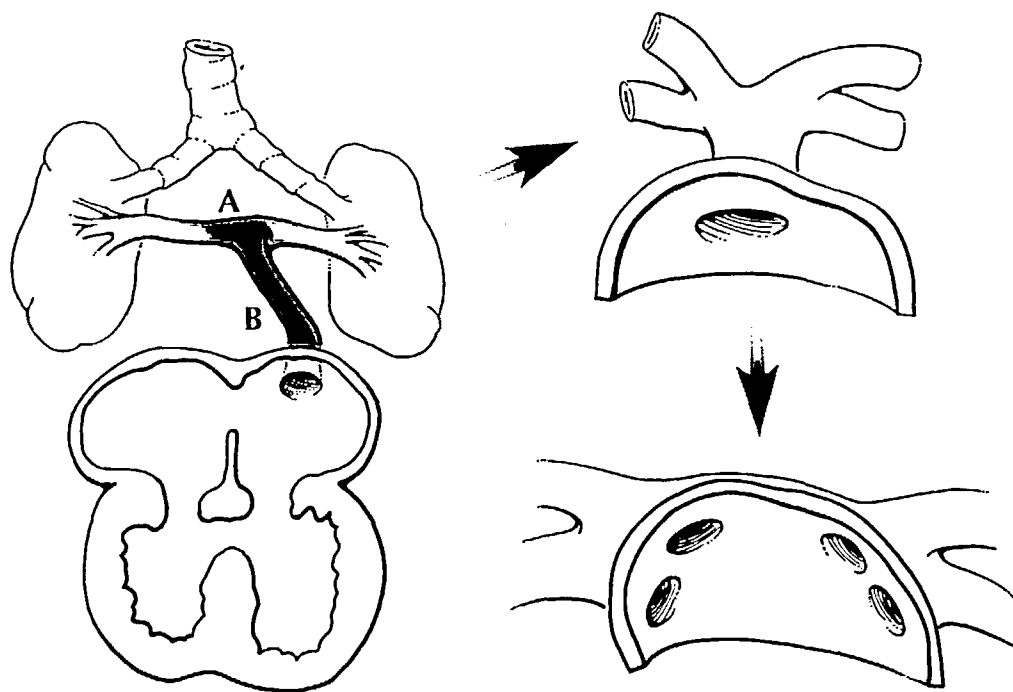
Total anomalous pulmonary venous connection (TAPVC) constitutes about 1% of all congenital heart disease.¹ It is not synonymous with total anomalous pulmonary venous *return* (TAPVR), in which the pulmonary veins proper connect appropriately to the left atrium but drain anomalously to the right side through an interatrial communication secondary to the presence of a left-sided structural atresia at some level. TAPVC, on the other hand, originates from nonfusion of the confluence of the pulmonary venous drainage and the left atrium. Through various routes, the oxygenated pulmonary venous blood is directed to the systemic venous side of the heart and pumped once again to the lungs. Some of the mixed venous and arterial blood in the right atrium crosses the atrial septum to enter the left atrium and ventricle for circulation to the systemic circulation. Because of the limited flow during embryonic and fetal development, the left atrium is usually small and the left ventricle is in the lower range of normal. Although pulmonary blood flow is usually more than adequate and there is no intrinsic failure of oxygenation at the alveolar level, TAPVC is associated with significant hypoxemia unresponsive to increased F_iO_2 . This hypoxemia occurs because of the obligatory mixture of pulmonary venous return with systemic venous return before partially oxygenated blood is delivered to the systemic circulation, in addition to mixing at the ductal level. TAPVC is generally unimproved by the addition of prostaglandins to maintain ductal patency. Obstruction of the venous drainage can occur, particularly with types III and IV (see below). This is the lone remaining pediatric cardiac surgical emergency, because the infant cannot be palliated with prostaglandins or Rashkind balloon septostomy. Many infants have an associated patent ductus arteriosus, usually associated with pulmonary hypertension. Although other congenital heart defects have been reported in association with TAPVC, including tetralogy of Fallot and double-outlet right ventricle,² TAPVC is usually an isolated lesion, except in conjunction with the heterotaxy syndromes, in which multiple complex anomalies are common.^{3,4}

History

Although the basic anatomy of this congenital defect has been known for more than 200 years, the specific embryology and anatomy has been well defined and understood only within the last few decades. The first review of the anatomic characteristics of TAPVC was published by Brody in 1942,⁵ with additional information provided by Darling and colleagues in 1957.⁶ The first antemortem diagnosis was not made until 1950,⁷ and the first successful complete repair was performed by Lewis in 1956.⁸ Modern technology with cardiac catheterization subsequently refined the preoperative anatomic definition, and more recently improved imaging quality with echocardiography has decreased the need for catheterization, except in the more complex cases such as those associated with heterotaxy syndromes or with type IV TAPVC.

Embryology

By the first 4 weeks of development, the embryonic lung buds have invaginated off the foregut, taking with them their own blood supply and venous drainage. This venous drainage is originally in communication with the splanchnic venous system via the cardinal and umbilicovittelline veins.⁹ When the embryo is at the 15mm stage, lung development proceeds and the four major pulmonary veins meet in the midline posterior to the developing heart to form the common pulmonary vein.¹⁰ Simultaneously the common atrium has undergone its formation and septation, and the confluence of the common pulmonary vein fuses with the posterior wall of the left atrium, creating the normal pulmonary venous connection (Fig 1). After this connection is established, communications with the systemic splanchnic veins normally involute. Partial failure of this fusion and dissolution of the intervening wall leads to cor triatriatum, whereas complete failure creates the various forms of TAPVC.^{11,12} Absence of drainage into the left atrium obligates the persistence of some other connection to the venous system through one or more



I Formation of the pulmonary vein confluence (A) and the common pulmonary vein (B), and normal fusion with the left atrium.

portions of the cardinal or umbilicovitelline systems. The communication that remains intact determines the type of TAPVC.

Classification

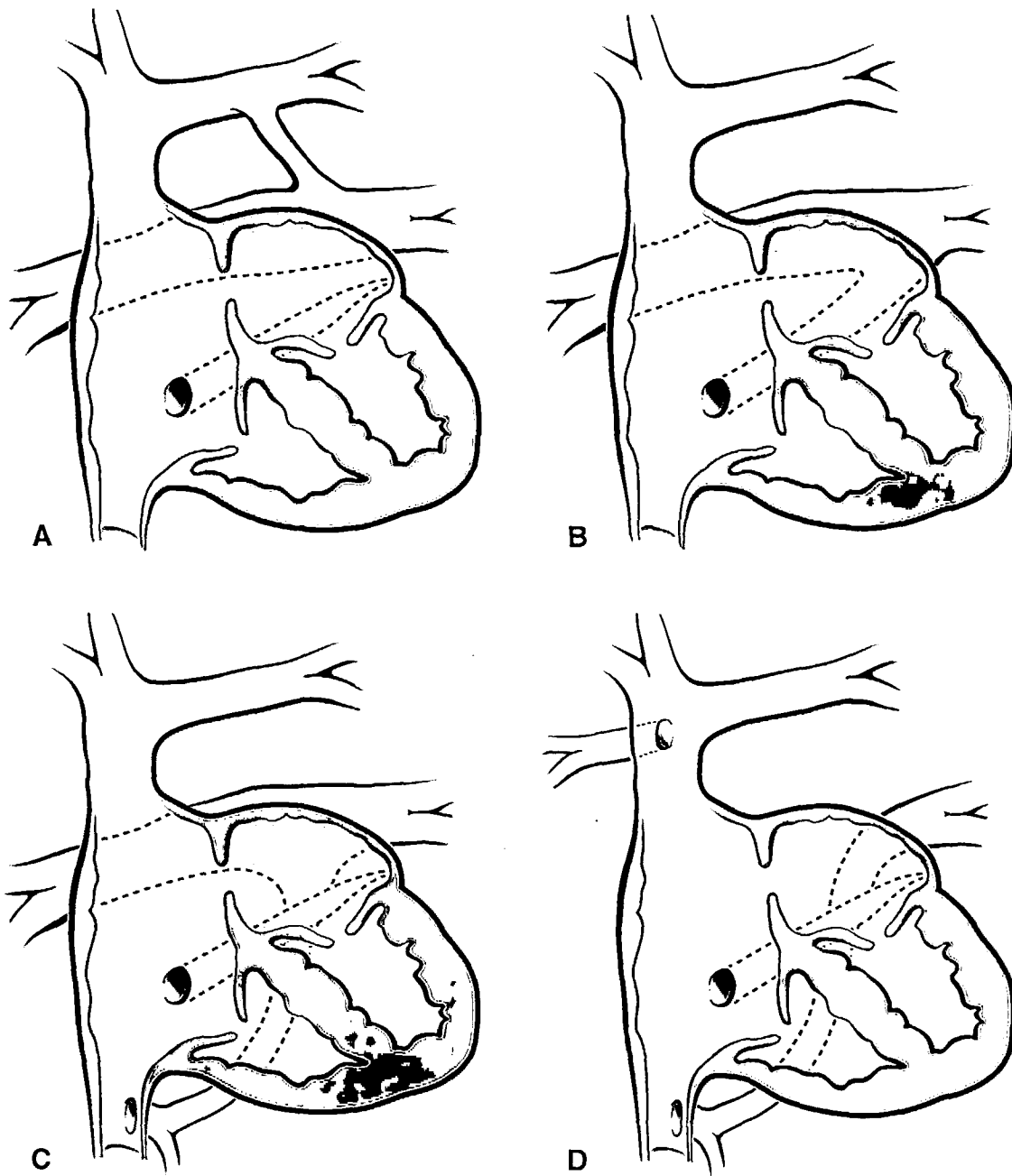
All forms of TAPVC involve complete drainage of pulmonary venous blood to the systemic venous side of the heart. TAPVC is generally classified into four major types as originally described by Darling⁶ based on anatomic location of the pulmonary venous drainage in relation to the heart: supracardiac, cardiac, infracardiac, and mixed (Fig 2).

Type I: Supracardiac

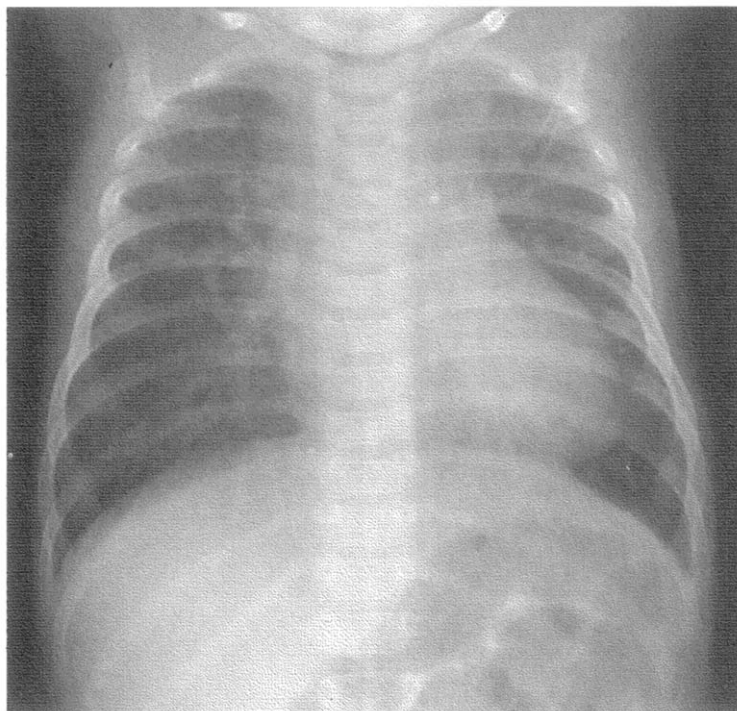
Supracardiac drainage is the most common form of TAPVC, constituting about 50% of cases in most series.^{1,13-16} Drainage of pulmonary venous blood coalesces in the midline, forming a confluence that is usually transverse and lies just behind and slightly above a small left atrium. The confluence most commonly

drains leftward and superiorly into the remnant of the left cardinal vein, into the left innominate vein, across the midline into the superior vena cava, and into the right atrium. Obstruction is not common but can occur, usually at the level of the pericardial reflection in the cardinal vein remnant. In fewer than one-third of supracardiac cases, the confluence drains to the right cardinal vein and enters the superior vena cava directly, usually near the level of the azygous vein insertion.

Because the pathophysiology of unobstructed supracardiac TAPVC is very similar to that of a large atrial septal defect, lesions not detected in infancy may present later with evidence of right ventricular volume overload, including right atrial and right ventricular dilation and main and branch pulmonary artery enlargement from the significantly increased pulmonary blood flow. In addition, the vertical vein and innominate vein may be dilated, producing a characteristic chest x-ray appearance with widening of the superior mediastinum (Fig 3).



2 Examples of the most common forms of TAPVC: (A) type I (supracardiac) with drainage into the left cardinal vein, into the left innominate vein; (B) type II (cardiac) with drainage into the coronary sinus; (C) type III (infracardiac) with drainage into the ductus venosus, into the liver; (D) type IV (mixed) here showing right supracardiac connection into the superior vena cava and left infracardiac connection into the liver.



3 Chest radiograph of a child with unobstructed type I (supracardiac) TAPVR, showing the prominent left upper mediastinal shadow from a dilated left vertical (cardinal) vein.

Type II: Cardiac

Cardiac or *intracardiac* drainage of TAPVC constitutes about 25% of total cases in most series.^{1,13-16} Almost all of these cases involve direct communication of the transverse pulmonary vein confluence with the coronary sinus. A persistent left superior vena cava may or may not be present. The coronary sinus is usually very large, and rarely does type II TAPVC cause obstruction. As in all types, there is an obligatory atrial level communication, ranging from a stretched patent foramen to complete absence of the atrial septum. In rare cases the common pulmonary vein is fused to the posterior atrium, but on the rightward side of the septum. In this form of type II TAPVC, the pulmonary veins enter a sinus that drains directly into the right atrial superior-posterior wall.

Type III: Infracardiac

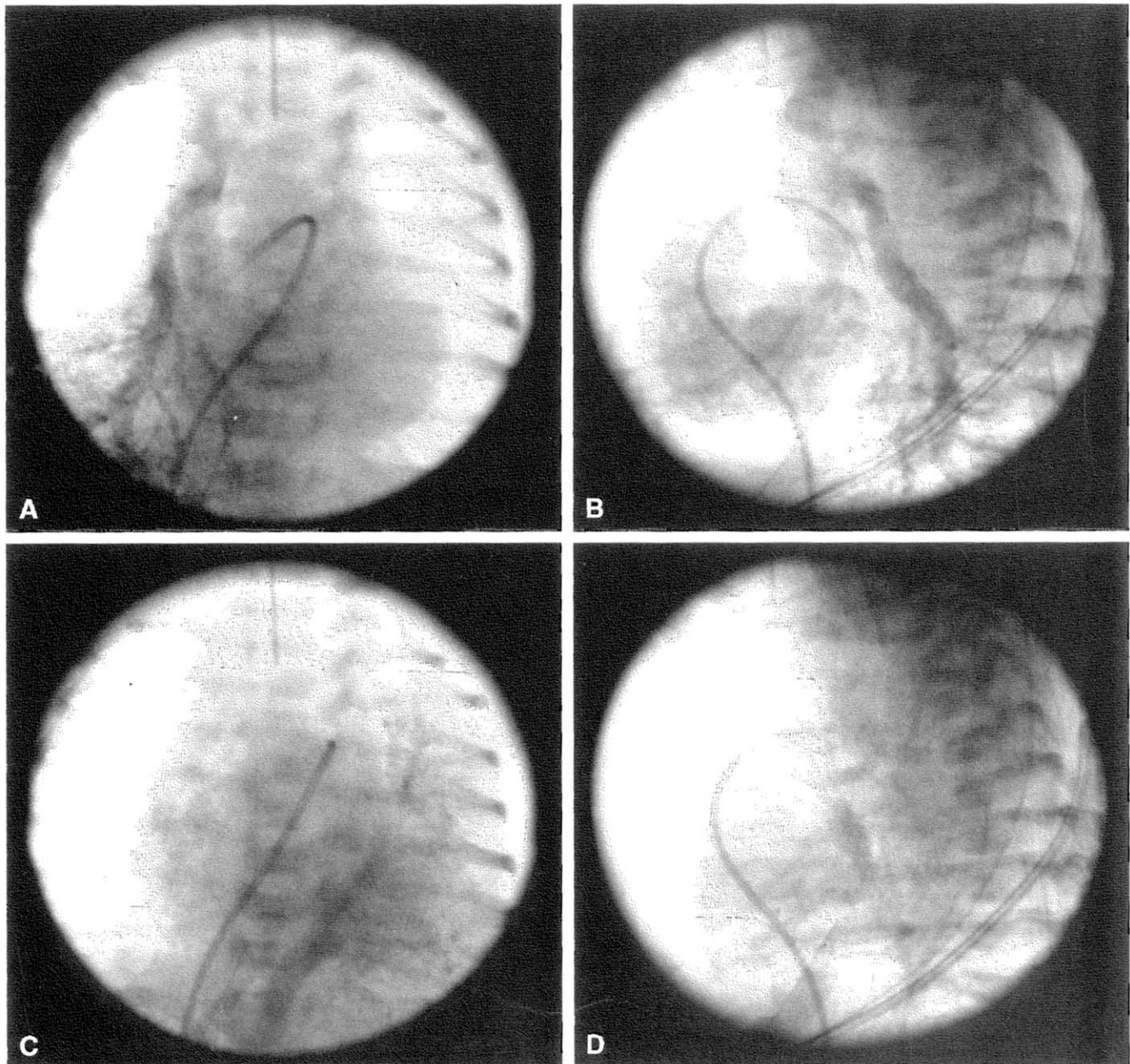
Slightly fewer than 25% of infants with TAPVC present with *infracardiac* or *subdiaphragmatic* connection.^{1,13-16} In these infants, the pulmonary confluence is usually more longitudinal than transverse and

drains into one of several remnants of the umbilico-vitelline vein. The communication runs inferiorly, traversing the diaphragm anterior to the esophagus and entering the splanchnic veins through the ductus venosus, through a hepatic vein, or directly into the inferior vena cava. The shape of this pulmonary venous confluence and connecting communication has been described as an "inverted Christmas tree," with the individual pulmonary veins sloping outward to the lungs hila as the branches of the tree. All of these cases essentially involve obstruction of the venous drainage at the level of the diaphragm or the ductus venosus, and thus present with profound hypoxemia and preoperative and postoperative pulmonary hypertension. The magnitude of pulmonary hypertension is related to the level and severity of obstruction. If the obstruction is at the level of a membrane within the vertical vein or due to a closing ductus venosus, then the clinical course is predictably acute and severe. Connections to the portal venous system, which obligate pulmonary venous flow to traverse the hepatic capillary bed, are usually associated with more moderate obstruction and a less severe clinical course.

Type IV: Mixed Drainage

Type IV TAPVC is relatively rare, constituting only about 5% of all cases.^{1,13-16} Usually there is no midline pulmonary confluence, and the right and left lungs have separate and different venous connections. This can involve almost any combination of drainage into the superior vena cava, the innominate veins, the cor-

onary sinus, the right atrium, the azygous vein, or the hepatic veins and inferior vena cava (Fig 4). Occasionally the entire drainage of one lung and part of the drainage of the other lung will join to form a confluence posteriorly, with the remaining vein draining separately into another splanchnic bed. In some cases these are very small accessory vessels, and reimplantation is not required.



4 Angiography of a child with type IV TAPVC. Contrast demonstrates that the right pulmonary veins connect to the right superior vena cava [(A) anteroposterior projection; (B) lateral projection], and the left pulmonary veins enter the liver below the diaphragm [(C) anteroposterior and (D) lateral].

Summary

Total anomalous pulmonary venous drainage is an unusual congenital heart defect classified as supracardiac, cardiac, infracardiac, or mixed based on the site of pulmonary venous connection to the systemic venous drainage. When the connection is obstructed, as commonly occurs with the infracardiac type, it is usually associated with severe pulmonary hypertension, profound hypoxia, and an acute clinical picture. Because prostaglandin-induced ductal patency and Rashkind balloon septostomy are not useful in palliating infants with obstructed TAPVC, this anatomic lesion is the one remaining congenital cardiac surgical emergency.

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