CASE REPORT

Maldevelopment of conotruncal and aorto-pulmonary septum with absent left central pulmonary artery: anatomical and clinical implications

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Abstract

In a patient with pulmonary valve atresia with hypoplastic main pulmonary artery selective angiography showed absence of the central left pulmonary artery and a right pulmonary artery originating from the ascending aorta close to the left coronary artery. This unusual anatomical

arrangement complicates interventional and surgical treatment.

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In patients with pulmonary valve atresia blood flows to the lung either through the ductus arteriosus or major aorto-pulmonary collateral arteries.¹ These collaterals usually arise from the descending aorta, but they can also originate from bronchial arteries, the subclavian artery, or other major branches of the aorta.² Coronary fistulas feeding central or peripheral pulmonary or collateral arteries have also been described³ and can complicate surgical repair.⁴ An abnormal left or right pulmonary artery originating directly from the ascending aorta can cause unilateral or bilateral pulmonary artery hypertension with ensuing pulmonary vascular obstructive disease.⁵

The occurrence of pulmonary valve atresia together with a right pulmonary artery with an abnormal origin and absence of the left central pulmonary artery has to the best of our knowledge not been reported before.

Case report

The patient was admitted to our institution when he was 13 days old with the diagnosis of pulmonary valve atresia with ventricular septal defect with major aorto-pulmonary collateral arteries to the lung. At operation (age 16 days) no central communication was found between the left and right pulmonary artery and no ductal ligament or patent ductus was seen. A modified Blalock-Taussig anastomosis was established on the left side by connecting a 5 mm Gore-tex prothesis end-to-side with the left subclavian artery and end-to-end with the left pulmonary artery. The procedure was well tolerated.

Postoperative re-catheterisation when the patient was 8 months old showed a major aorto-pulmonary collateral artery from the upper descending aorta supplying blood flow to the right upper lung (fig 1A). The shunt was connected to the distal part of the left pulmonary artery and the proximal part was missing (fig 1B).

Figure 1 Angiograms in anterior-posterior views showing (A) the main aortopulmonary collateral artery arising from the upper descending aorta and supplying the circulation for the right upper lung lobe and (B) a modified left Blalock-Taussig anastomosis connected with the distal part of the left pulmonary artery.

Figure 2 (A) Angiogram in laid back view, showing selective angiography into the left dorsal sinus of Valsalva. Separate orifices are visible for the left coronary artery (LCA) and the right pulmonary artery (RPA), and the RPA connects with the distal end of the hypoplastic main pulmonary artery (MPA). (B) A drawing to illustrate the arrangement seen in (A).

Additional selective angiograms showed an abnormal vessel (diameter 2.5 mm) originating 3 mm above the left coronary artery orifice and connecting with the distal end of a hypoplastic pulmonary artery stem. This vessel (diameter 7 mm) provided the pulmonary blood flow to the right middle and lower lung and was thus considered to be the right pulmonary artery. The left coronary artery originated normally from the left dorsal sinus of Valsalva of the aortic root and its orifice lay just below the orifice of the right pulmonary artery (fig 2A and B). No communication between the coronary artery and the right pulmonary artery was shown by repeated selective injections.

Further simultaneous selective angiograms into the right pulmonary artery and into the infundibulum of the right ventricle confirmed the existence of a hypoplastic main pulmonary artery (fig 3A). The atretic pulmonary valve membrane was perforated with a specially designed perforation needle (0.014

inch). Repeated selective angiograms below the pulmonary valve then showed the anatomical continuity of the hypoplastic right ventricular outflow tract with the main pulmonary artery (fig 3B). Because the pulmonary artery, the left coronary artery, and the abnormal origin of the right pulmonary artery lay so close together further attempts to open the atretic pulmonary valve were abandoned.

Discussion

This was an unusual case of pulmonary valve atresia with an hypoplastic main pulmonary artery, an abnormal origin of the right pulmonary artery from the ascending aorta, and absent left central pulmonary artery.

The anatomical continuity from the right ventricle to the hypoplastic main pulmonary artery and the atretic membrane indicate completed aorto-pulmonary septation in the valve area and exclude a truncus arteriosus.^{6,7} Conotruncal malformations (truncus arteriosus, tetralogy of Fallot, or pulmonary atresia) cannot occur together, but each can coexist with malformations of the pulmonary arterial system because the pulmonary artery system and the conotruncal part of the heart develop independently and at different times during embryogenesis.^{8,9} In some forms of tetralogy of Fallot both abnormal origin and absence or atresia of the pulmonary arteries have been described.^{2,10-12} When there is a right aortic arch the left pulmonary artery is generally abnormally connected to the ascending aorta, and vice versa.^{5,13}

The origin of the right pulmonary artery from the aorta could indicate that the complicated formation of the bifurcation was disturbed. The anomalous origin of a pulmonary artery from the ascending aorta has a typical morphology¹³ and is explained by failure of the right sixth aortic arch to migrate within the wall of the embryonic truncus from right to left towards the left sixth aortic arch and the main pulmonary artery during embryonic formation of the bifurcation.⁸ The fact that in our case the right pulmonary artery originated from the left side of the ascending aorta suggests that migration had occurred and was complete. The left pulmonary artery, however, was missing. This suggests persistence of the origin of the right pulmonary artery from the truncus and, after aortopulmonary septation, from the aorta. Unlike the right side, the left central pulmonary artery is formed by nonarch respiratory vessels from the lung buds. The proximal part of the left sixth aortic arch connects these pulmonary vessels to the right sixth aortic arch and later largely disappears.⁸ The extent to which the proximal part is involved in the formation of the bifurcation is uncertain. The distal part of the left sixth aortic arch becomes the ductus arteriosus. In our case, both parts of the left sixth aortic arch—the proximal part as the left central pulmonary artery with connection to the bifurcation and the distal part as ductus arteriosus—could not be found either during

Figure 3 Angiograms in the lateral view showing the continuity of the hypoplastic right ventricular infundibulum, perforated atretic pulmonary valve, and hypoplastic main pulmonary artery stem shown by selective angiography into the orifice of the right pulmonary artery (A) and into the right ventricular infundibulum after needle perforation of the valve (B).

operation or at catheterisation. This evidence suggests congenital absence of the left sixth aortic arch.

We think that our case represents an extreme form of tetralogy of Fallot with pulmonary valve atresia and a hypoplastic main pulmonary artery that is associated with a complex pulmonary artery malformation. This unusual anatomical arrangement, with the origin of the abnormal right pulmonary artery lying between where the aorta is cross clamped and the aortic valve, means that at operation cardioplegia solution will enter the

coronary arteries and the pulmonary arterial bed. Continuing collateral arterial perfusion of the lungs during cardiopulmonary bypass will maintain significant pulmonary venous return.

As techniques for selective and supraseductive angiography together with new methods for treatment, such as embolisation of collateral vessels or interventional transcatheter opening of pulmonary valve atresia¹⁴ develop the precise anatomical delineation of complex cardiac malformations will become more relevant. In our patient further interventional transcatheter opening of the atretic pulmonary valve was impossible, however, because the valve lay close to the left coronary artery and because the connection with the high pressure in the aorta would have led to pulmonary regurgitation with unpredictable short and long term results.

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