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Brief Report

Critical pulmonary stenosis with an aortopulmonary window

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Abstract In neonates with severe aortic or pulmonary stenosis, the obstructed circulation is usually supported through patency of the arterial duct. We describe a neonate in whom an aortopulmonary window supported a critically obstructed pulmonary circulation.

Keywords: Aortopulmonary fenestration; pulmonary valve

In neonates with critically obstructed systemic or pulmonary blood flow, the presence of a communication between the systemic and pulmonary circulations is necessary for survival. In most patients, patency of the arterial duct provides this communication, although in some, particularly those with tetralogy of Fallot and pulmonary atresia, aortopulmonary collateral arteries may subserve this role. Although an aortopulmonary window usually exists in isolation, and is rare, it, too, can support a critically obstructed circulation. Thus, there have been two case reports describing neonates with aortic atresia where an aortopulmonary window supported the systemic circulation.[1,2] As far as we are aware, however, no cases are reported in which an aortopulmonary window supported pulmonary flow in the setting of critical pulmonary stenosis. We describe such a case.

Case report

A 22-hour-old baby, born at term weighing 2.2 kg, was referred because of circulatory collapse. He was resuscitated and started on an intravenous infusion of Prostaglandin E1, before being transferred to our centre.

On examination, he was mechanically ventilated, and had good peripheral pulses, with no brachio-femoral delay. The arterial saturation of oxygen, in the presence of an inspired concentration of oxygen of 30%, was 94%. The second heart sound was single, and there was a soft systolic murmur in the pulmonary area. A chest radiograph revealed cardiomegaly with increased pulmonary vascular markings.

The definitive diagnosis was obtained using cross-sectional echocardiography. The atrial arrangement was usual, and the atrioventricular and ventricular-arterial connections were concordant. There was left-to-right shunting across a patent oval foramen, while the interventricular septum was intact. The pulmonary valve was thickened and stenotic, with very little antegrade flow (Fig. 1). Instead, most of the flow of blood to the lungs was from the aorta,

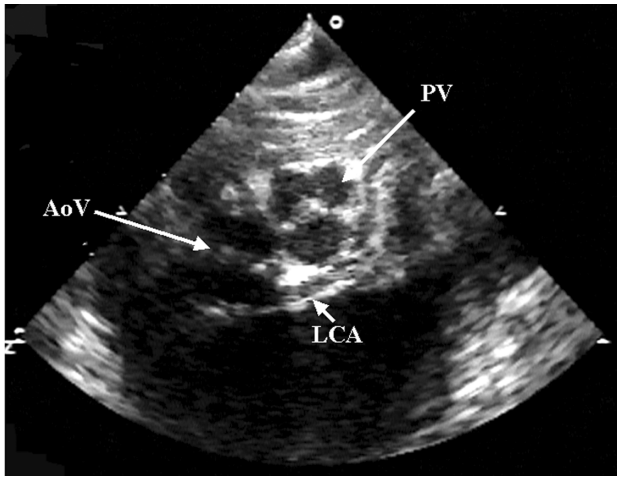

Figure 1.
Parasternal short axis view showing the thickened pulmonay valve. PV: pulmonary valve; AoV: aortic valve; LCA: left coronary artery.

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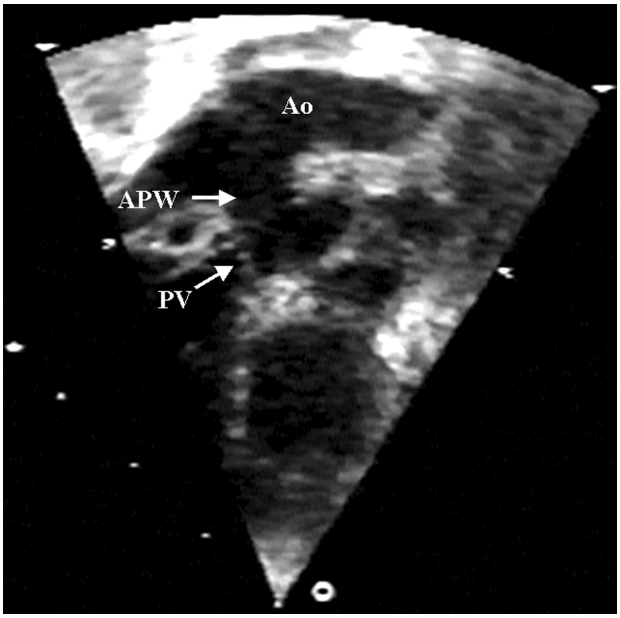

Figure 2.
Subcostal short-axis view showing a large aortopulmonary window. Ao: aortic arch; APW: aortopulmonary window; PV: pulmonary valve.

through a large aortopulmonary window (Fig. 2). There was no arterial duct. Bilateral superior caval veins were noted, with the left one draining into the right atrium via an enlarged coronary sinus. The infusion of prostaglandin was stopped, and the patient was referred for surgery.

He underwent surgical repair of the aortopulmonary window and pulmonary valvotomy on the third day of life. At surgery, the heart was moderately enlarged. There was a proximal aortopulmonary window, measuring 3 to 4 mm in diameter. The pulmonary valve was trifoliate, thickened, and dysplastic, its aperture accepting only a 1 mm dilator. The right and left pulmonary arteries were of adequate size. The presence of the left superior caval vein was confirmed, with the coronary sinus being dilated and opening normally to the right atrium. The post-operative course was complicated by a low cardiac output state requiring use of a left ventricular assist device. He was successfully weaned from extracorporeal support on the tenth post-operative day, but he died 13 days later as a result of sepsis. Permission for postmortem was refused.

Discussion

Neonates with critical pulmonary stenosis usually present with cyanosis and become rapidly symptomatic with the closure of arterial duct. Infusions of Prostaglandin E1 are now used to maintain the patency of the arterial duct, until a definitive procedure can be performed on the pulmonary valve. In our patient, a large aortopulmonary window provided unrestricted pulmonary blood flow in the presence of critical pulmonary stenosis. The unrestricted left-to-right shunt at arterial level was responsible for the rapid onset of congestive failure soon after birth.

There have been reports of neonates with a critically obstructed systemic circulation, due to aortic atresia in which systemic flow of blood was maintained by an aortopulmonary window.[1,2] To our knowledge, ours is the first patient described in whom the flow of blood to the lungs was almost exclusively provided by an aortopulmonary window.

In our patient, a comprehensive diagnosis was made with cross-sectional echocardiography, and cardiac catheterisation was not required. Cardiac catheterisation in the sick neonate carries a small but definite risk, which can be, avoided by using non-invasive imaging. Taken separately, both pulmonary stenosis and aortopulmonary window can be accurately diagnosed by cross-sectional echocardiography.[3–5] It should come as no surprise, therefore, that the combination of these anomalies was correctly diagnosed in our patient.

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