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Brief Report

Hypoplasia of the left heart with discordant ventriculoarterial connections

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Abstract We report the rare combination of a severely hypoplastic left ventricle and discordant ventriculoarterial connections, with associated subpulmonary obstruction of the left ventricular outflow tract. The long tortuous arterial duct originated at an acute angle from the aorta, suggesting that the subpulmonary obstruction developed early in fetal life. Residual flow via the hypoplastic left ventricle to the pulmonary circulation after insertion of an aortopulmonary shunt resulted in haemodynamic deterioration, which was instantly reversed by closing the pulmonary trunk.

Keywords: Hypoplastic left heart syndrome; transposition of the great arteries; functionally univentricular heart

SEVERE HYPOPLASIA OF THE LEFT VENTRICLE sufficient to render a patient unsuitable for biventricular repair is exceedingly rare when the ventriculoarterial connections are discordant, with only seven cases thus far reported, as far as we know, in the literature.[1–5] Premature constriction of the arterial duct and oval foramen, with concomitant decrease in flow through the left side of the heart, have been suggested as potential pathogenetic factors for the left-sided hypoplasia.[1] Due to the fact that, because of the discordant ventriculoarterial connections, the combination produces the haemodynamics of pulmonary atresia, it is necessary to create an aortopulmonary shunt should palliation be required in the neonatal period.[4] We report another case of this rare malformation, in which we encountered unexpected haemodynamic complications after construction of an aortopulmonary shunt.

Case report

The male infant, measuring 52 centimetres in length, and weighing 3.2 kilograms, was born at term. He presented at the age of 2 days, with desaturation of oxygen, at around 60%, the level improving to 85% subsequent to treatment with prostaglandins. Echocardiography revealed usual atrial arrangement with concordant atrioventricular but discordant ventriculoarterial connections. The great arteries were positioned with the aorta anterior to the pulmonary trunk. The aortic root, measuring 9.3 millimetres in diameter (Fig. 1a), originated from an enlarged right ventricle, which had an end-diastolic diameter of 15.5 millimetres. The hypoplastic pulmonary trunk, measuring only 4.2 millimetres in diameter (Fig. 1b), originated from the left ventricle. The right and left pulmonary arteries were slightly hypoplastic, being calculated to give a Nakata index[6] of 138 square millimetres per square metre of body surface area. The right atrium was moderately enlarged, and the left atrium was of normal size, but there was marked hypoplasia of the mitral valve, the left ventricle, calculated to have an end-diastolic volume of only 1 cubic millilitre, and the mitral valvar annulus, measured at 2 millimetres diameter. Colour Doppler revealed a small subpulmonary ventricular septal defect of 2 millimetres dimensions. Subvalvar and valvar obstruction of the left ventricular outflow tract produced a Doppler gradient of 57 millimetres of mercury. There was antegrade flow, nonetheless, across the mitral and pulmonary valves, the latter valve having

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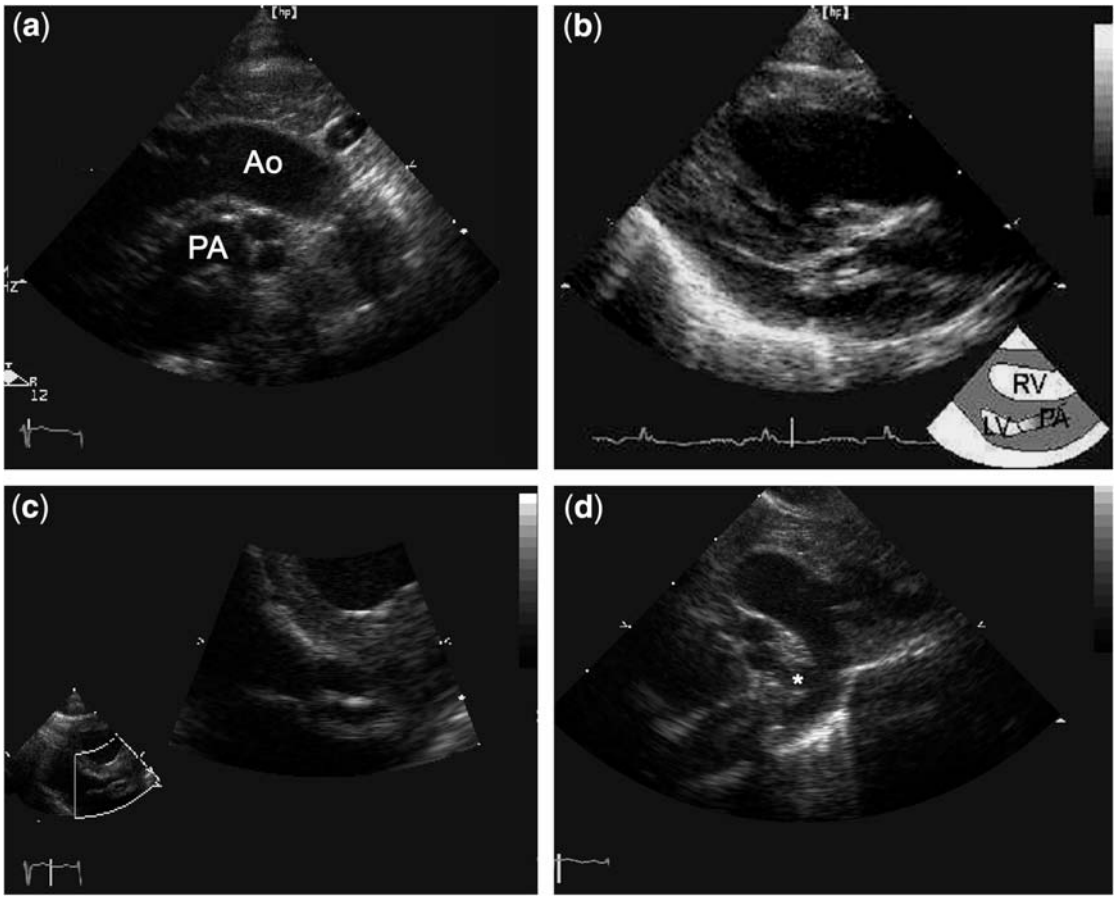

Figure 1.
*The cross-sectional echocardiogram shows (**a**) an anterior-posterior position of the great arteries, the aorta being of normal size (Ao) and (**b**) the small pulmonary trunk (PA). The parasternal long axis view (**c**) reveals the enlarged right ventricle (RV), with the pulmonary trunk originating from the severely hypoplastic left ventricle. Note the hypoplastic pulmonary valve, which has two leaflets. (**d**) shows the acute angle of origin of the arterial duct (*).*

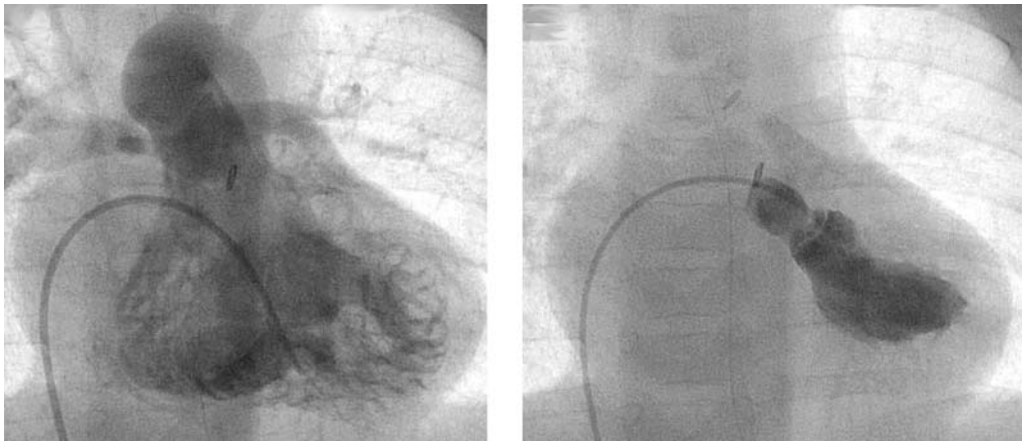

Figure 2.
The postoperative angiocardiograms, taken at the age of 10 months, show (left) the dilated right ventricle feeding the aorta and pulmonary arteries, and (right) the hypoplastic left ventricle. Note that the ventricular septal defect has closed spontaneously, and that the pulmonary trunk is now blind-ending subsequent to the operative clipping.

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two leaflets (Fig. 1c). The oval foramen was mildly restrictive, permitting a maximum velocity of flow of 1.2 metres per second during atrial contraction, with left-to-right shunting. Whilst receiving prostaglandins there was shunting from the aorta to the pulmonary arteries across the arterial duct, with a gradient of 18 millimetres of mercury. The duct originated at an acute angle from the aorta (Fig. 1d). Catheterisation and angiography were not performed in the neonatal period. The child underwent an atrial septectomy, implantation of a central aortopulmonary shunt of 5 millimetres diameter, and closure of the arterial duct, all achieved at the age of four days. Within 12 hours of the procedure, the child developed metabolic acidosis, and we suspected pulmonary hyperperfusion. Secondary closure of the pulmonary trunk by placement of a surgical clip produced an instant improvement of the haemodynamic condition, and the further course was uneventful. Catheterisation (Fig. 2) at the age of ten months revealed good function of the systemic morphologically right ventricle. The ventricular septal defect had closed spontaneously, and the pressures were suprasystemic in the hypoplastic left ventricle. The patient successfully underwent a bidirectional Glenn anastomosis at the age of ten months, and a modified extracardiac Fontan operation at the age of 26 months. He is now doing well at the age of 4 years.

Discussion

The development of left ventricular hypoplasia in combination with discordant ventriculoarterial connections probably requires two fundamentally different causative mechanisms. The factors contributing to the impaired growth of the left-sided structures could be the result of diminished prenatal perfusion of the left heart, such as produced by restriction of the oval foramen,[1] obstruction of the left ventricular outflow tract, and possibly abnormal function of the developing ventricle during the prenatal period. The causes for discordant ventriculoarterial connections are still unclear, and a matter of ongoing research, but since the outflow tract is topographically apart from the cardiac inflow, we suspect different developmental factors. It is, perhaps, the requirement for two independent pathogenetic mechanisms that accounts for the paucity of this combination. Discussion concerning the case might centre on whether the combination should be defined as "discordant ventriculoarterial connections with left heart hypoplasia", or if the term "hypoplasia of the left heart with discordant ventriculoarterial connections" would be more apposite. With regard to the sequence of events during early embryonic stages, we can derive clues from the morphology of the arterial duct. Postnatally, the duct originated at an acute angle from the aortic arch, and ran a long and tortuous course, resembling the arrangement seen in pulmonary atresia with intact septum, or with a ventricular septal defect, rather than the arrangement typically seen in transposition.[7] The morphology, angle and the site of origin of the arterial duct seem to depend on the timing of the embryological insult responsible for the pulmonary obstruction.[8] An acute angle between the arterial duct and the aortic arch is frequently associated with a long and tortuous course. This morphology points towards a malformation starting earlier in the developing heart[9] than disorders associated with an obtusely angled arch. Taking the ductal morphology into account, therefore, we opted to label the combination as "hypoplasia of the left heart with discordant ventriculoarterial connections".

The postoperative haemodynamics in our patient differed markedly from those encountered in neonates with classical hypoplastic left heart syndrome. Residual patency of the left ventricular outflow tract and aortic valve is usually well tolerated in these patients following the first stage of the Norwood sequence. Because of the severely hypoplastic left ventricle, and the markedly obstructed left ventricular outflow tract, we chose not to close the connection between the left ventricle and the pulmonary arteries at the time of constructing the aortopulmonary shunt. The hypoplastic left ventricle, nonetheless, was able to contribute significantly to the flow of blood to the lungs, despite the marked obstruction of the left ventricular outflow tract. This can be explained by the fact that the left ventricle was connected to the low resistance pulmonary vascular bed. The haemodynamics improved immediately following occlusion of the pulmonary trunk. The communication between the left ventricle and the pulmonary arteries, therefore, may need to be closed in these patients at the time of creating a palliative aortopulmonary shunt.

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