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Images in Congenital Heart Disease

Proximal origin and hypoplasia of the left pulmonary artery in association with chromosome 22q11 deletion, right aortic arch, and persistently patent right-sided arterial duct

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Two neonates, diagnosed prenatally with chromosome 22q11 deletion, were both found to have a right aortic arch, a persistently patent right-sided arterial duct, and proximal origin of a hypoplastic left pulmonary artery. In the first, there was interruption of the aortic arch between the right common carotid and right subclavian arteries, with aberrant retrooesophageal origin of the left subclavian artery. Postnatal echocardiography in the parasternal long axis view (Fig. 1) also demonstrated a doubly committed juxta-arterial ventricular septal defect (VSD), dysplastic aortic valve, and hypoplastic ascending aorta (AAO). A hypoplastic and stenotic left pulmonary artery (LPA), originated anteriorly from the proximal pulmonary trunk (PA), and was best seen in the high parasternal cut, along with the patent arterial duct (PDA) and descending aorta (DAO) (Fig. 2). This neonate underwent repair by directing the left ventricle to the pulmonary trunk as the systemic outflow, reconstruction of the aortic arch, and implantation of an extracardiac conduit from the right ventricle to the pulmonary arteries at 2 weeks of age. The left pulmonary artery was resected from the pulmonary trunk, reconstructed, and anastomosed to the right pulmonary artery (RPA). This patient made an uneventful recovery.

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In the second patient, an isolated, perimembraneous ventricular septal defect was closed uneventfully at two months of age. The small left pulmonary artery, arising proximally from the anterior aspect of the pulmonary trunk, as well as the patent arterial duct and right-sided descending aorta, (Figs. 3 and 4) were clearly demonstrated in the parasternal short axis view, but did not require surgical intervention.

Most patients with chromosome 22q11 deletion have malformations of the ventricular outflow tract, along with the derivatives of the fourth aortic arch.

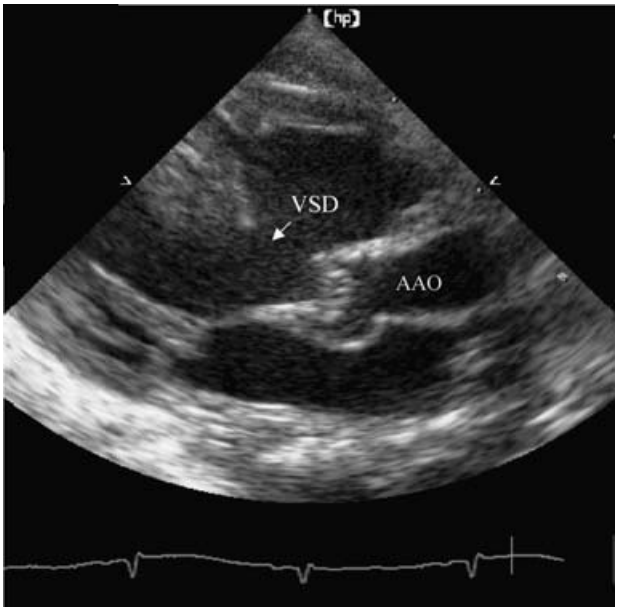

Figure 1.

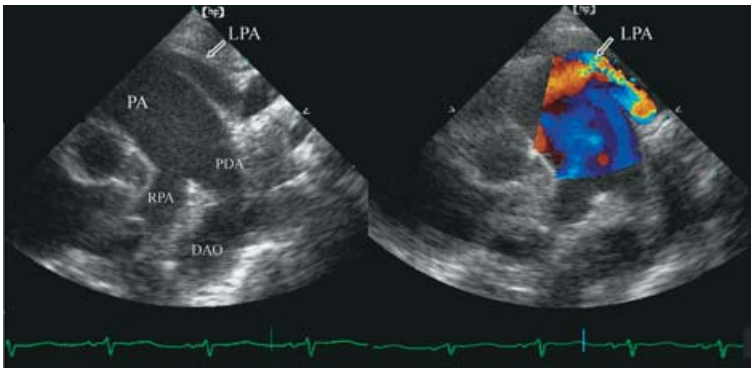

Figure 2.

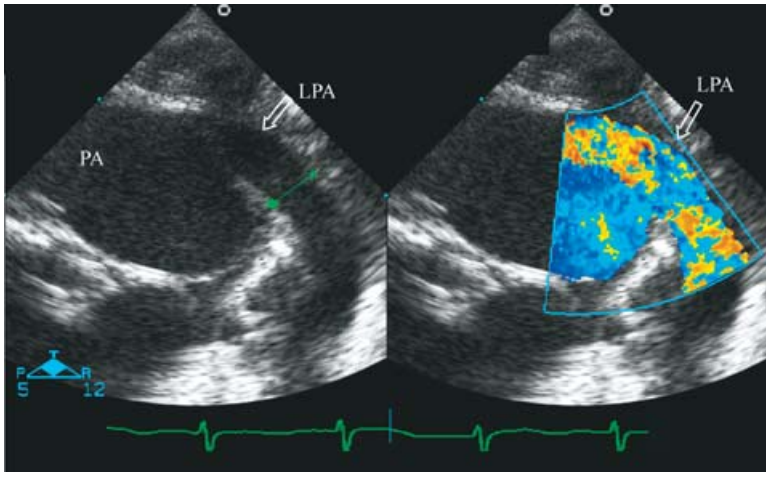

Figure 3.

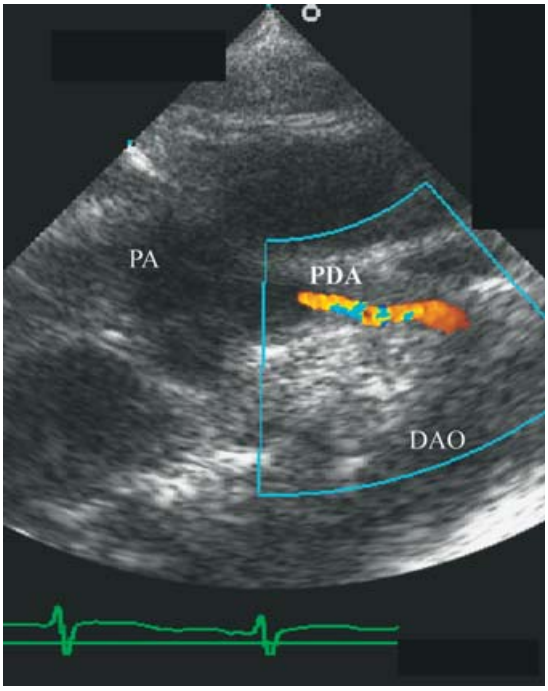

Figure 4.

While it is not clear whether proximal origin of a hypoplastic left pulmonary artery is a manifestation of maldevelopment of the sixth arch in the setting of chromosome 22q11 deletion, or a consequence of the right aortic arch with right-sided arterial duct, the combination of right aortic arch and chromosome 22q11 deletion should raise the possibility of this anomaly. Preoperative recognition of this anomaly is mandatory, since proximal origin of the left pulmonary artery excludes the possibility of banding the pulmonary trunk, and may require enlargement with a patch or reanastomosis of the stenotic vessel.

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