Cardiology in the Young

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Brief Report

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Echocardiography: conotruncal anomaly: a case of common arterial trunk with intact ventricular septum and hypoplastic left heart complex with unbalanced pulmonary stenoses

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Abstract

We describe the echocardiographic findings of a common arterial trunk with intact ventricular septum, mitral and left ventricular hypoplasia, atretic left ventricular outlet and bilateral, and unbalanced pulmonary artery stenoses.

Conotruncal anomalies comprise a large variety of CHDs and are usually associated with balanced ventricles and, if found, defects of the outlet ventricular septum. Common arterial trunks develop due to an incomplete or absent rotation of the outflow tract by an abnormal migration of cardiac neural crest cells impairing the elongation of the outflow tract from the second anterior heart field.[1] Trunks with hypoplastic ventricles have been described previously, but were mostly associated with ventricular septal defects.[2]

Case report

We report on a child with a prenatally known cardiac defect classified as hypoplastic left heart complex with mitral and aortic atresia. Antenatal chromosomal testing was normal. Delivery was uneventful after 39 weeks, the child, being small for gestational age, adapted well (birth weight 2900 g, length 46 cm, and APGAR 8/9/9).

The initial echocardiography showed a single outlet vessel from the right ventricle in posterior orientation. The outlet valve was bicuspid with neither stenosis nor regurgitation. A small protuberance from the proximal trunk to the right was considered as an abortive paleo-aorta. The mitral valve and the left ventricle were severely hypoplastic without any forward or backward flow via the mitral valve and without any outlet of the left ventricle. The ventricular septum was intact. The pulmonary arteries originated from the single outlet vessel with separate orifices and showed unbalanced stenoses. An unobstructed left aortic arch and multiple atrial septal perforations were present. The malformation was therefore addressed as a common arterial trunk type A2 (van Praagh's classification) with intact ventricular septum, hypoplastic left heart complex and bilateral, unbalanced pulmonary stenoses with small, and non-hypoplastic pulmonary arteries. Morphologic features are shown in Figure 1a–f and in the supplementary video S1.

Due to the single ventricle, a univentricular palliation was mandatory. A bilateral pulmonary banding was performed to allow further growth of the child and prevent pulmonary hypertension. A Glenn-anastomosis (superior caval vein to right pulmonary artery) with augmentation of the central pulmonary arteries and total cavopulmonary connection are planned in the future.

Discussion

We describe a rare case of a common arterial trunk with intact ventricular septum and hypoplastic left heart complex. To our knowledge, this anatomy has been so far exclusively described in post mortem specimens. Echocardiography showed the typical morphologic features. Rotational anomalies might not be the only pathomechanism in the development of truncal malformations Therefore, an "outlet-less" hypoplastic left ventricle may also be present in common arterial trunks.[2] Univentricular palliation may pose a great challenge, especially considering the elevated risk of pulmonary hypertension during lifetime and an unpredictable growth capacity of the central pulmonary arteries.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951123003554.

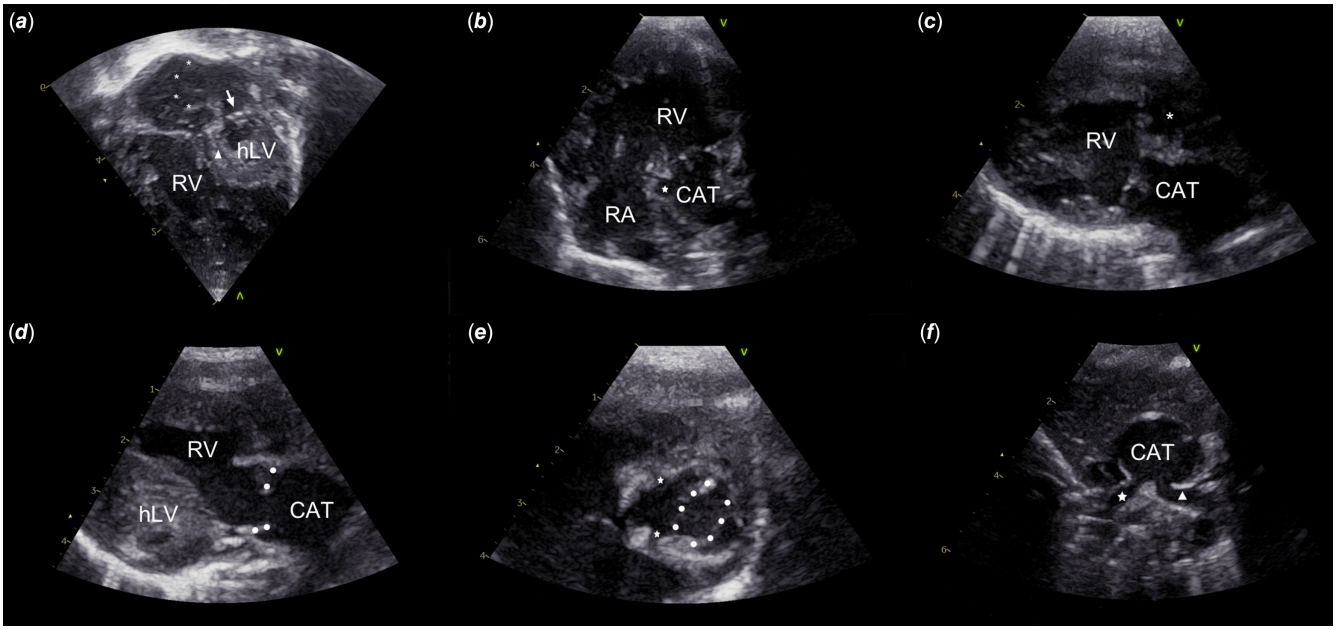

Figure 1. Echocardiographic findings, still images. ***a***: Apical 4-chamber view: hypoplastic left ventricle (hLV), normally developed right ventricle (RV), atrial septum (asterisks) aneurysmatically protruding into the right atrium; mitral atresia (arrow); intact ventricular septum (triangle). ***b***: Apical long axis, modified: inlet–outlet of RV; protuberance (asterisk) from the CAT to the right as an abortive 'paleo-aortic remnant'; right atrium (RA). ***c***: Parasternal long-axis view: the solitary outlet (CAT) from the right ventricle is positioned more posteriorly, the infundibulum of the RV (asterisk) ends. ***d*** and ***e***: Parasternal long (d) and short (e) axis: hypoplastic left ventricle (hLV) without outlet; slightly dysplastic, opened truncal valve (dots depicting the commissures); coronary arteries from the right sinus (asterisks). ***f***: Parasternal short axis: CAT with origins of the pulmonary arteries; right pulmonary artery (asterisk) with stenosis, left pulmonary artery unobstructed (triangle).

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Ethical standards. The parents consented to the use of the echocardiographic loops and images in an anonymous publication.

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