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Brief Report

Total anomalous systemic venous drainage in left heterotaxy syndrome

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Abstract Total anomalous systemic venous drainage is an extremely rare congenital heart defect. In this study we describe an 11-year-old girl who presented with a history of fatigue and central cyanosis that she had had since early childhood with unremarkable precordial examination results. Investigations revealed left heterotaxy with all systemic venous drainage to the left-sided atrium with non-compaction of the left ventricle.

Keywords: Cyanosis; systemic venous anomaly; non-compaction

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Systemic venous anomalies are rare congenital heart defects and are seen either as an isolated anomaly or more commonly in association with complex congenital heart disease. Total anomalous systemic venous drainage to the left atrium is an extremely rare systemic venous anomaly. In this anomaly, all systemic venous flow, including from the hepatic veins and the coronary sinus, drains abnormally into the heart. Here, we report an unusual case of total anomalous systemic venous drainage in the absence of other severe intra-cardiac defects in a patient with left heterotaxy syndrome and left ventricular non-compaction who underwent a successful surgical correction.

Case report

An 11-year-old girl presented with a history of fatigue and central cyanosis that she had had since early childhood. On examination, her heart rate was 60 per minute and blood pressure was 110/70 millimetres of mercury. Jugular venous pressure was normal with irregular cannon waves. Her precordial examination was unremarkable. Biochemical parameters were within normal limits, except for haemoglobin 18.9 grams per decilitre and packed cell volume 57.10. Oxygen saturation with pulse oxymetry in room air was 74%.

The 12-lead electrocardiogram revealed intermittent isorhythmic atrioventricular dissociation. Two-dimensional cross-sectional echocardiographic and Doppler examination revealed an interrupted inferior caval vein with hepatic veins draining directly into the left atrium. There was ostium secundum atrial septal defect with left-to-right shunt and non-compaction of the left ventricle myocardium with deep intra-trabecular recesses (Fig 1). The right superior caval vein was dilated. Contrast echocardiography revealed systemic veins from the upper and lower limbs draining directly into the left atrium.

Cardiac catheterisation and angiography study showed an interrupted inferior caval vein draining through the azygous system into the dilated right superior caval vein and into left atrium (Supplementary Video 1). Oxymetry revealed an average saturation of 82% in all four cardiac chambers. Pulmonary artery pressure was normal. There was no evidence of a persistent left superior caval vein. Coronary sinus drainage was to the left atrium. Pulmonary venous drainage was normal. Both left and right ventriculography revealed adequately sized and normally functioning ventricles. Computerised tomographic angiography, apart from confirming our findings, also revealed polysplenia, a bilateral similar left bronchial pattern. Hepatic

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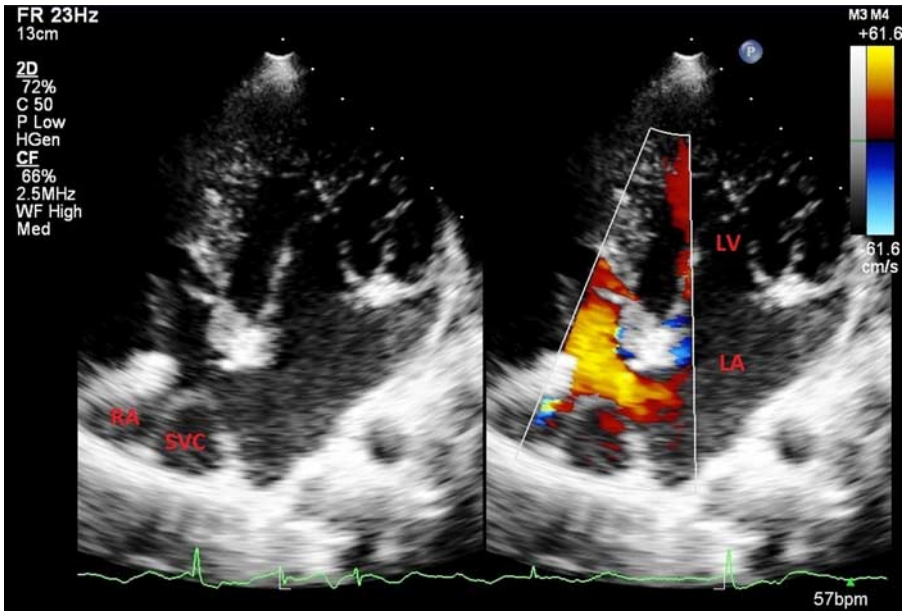

Figure 1.
Echocardiographic apical four-chamber view showing ostium secundum atrial septal defect L→R shunt; dilated left atrium and left ventricle; non-compaction of left ventricle; dilated superior caval vein in cross-section. LA = left atrium; LV = left ventricle; RA = right atrium; SVC = superior vena cava (superior caval vein).

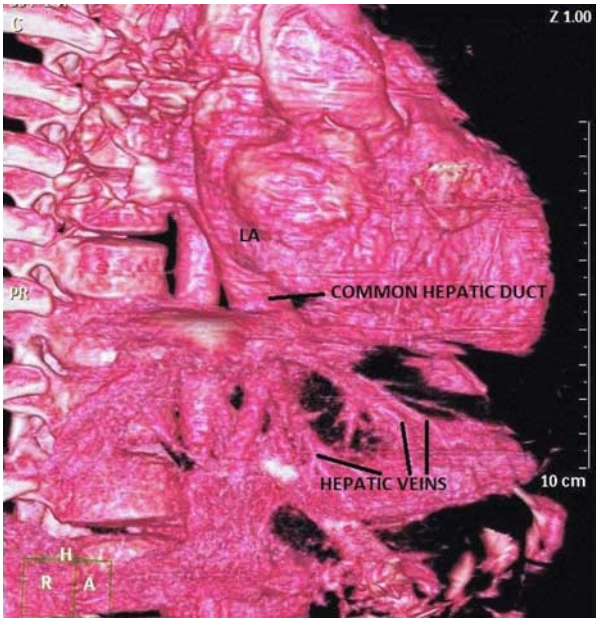

Figure 2.
Computed tomography angiography showing hepatic veins draining through the common hepatic duct to the dilated left-sided atrium. LA = left atrium.

veins were draining through a common channel into the left atrium (Fig 2).

The patient underwent successful surgical correction. After opening the right atrium, the limbus was incised and sutured to the left atrium roof in such a manner that the superior caval vein now drained into the right atrium. The remaining defect was closed with a pericardial patch baffle keeping the hepatic and coronary sinus drainage to the right atrium. Post-operatively the patient recovered without any complication, and saturation was 98% in room air. Holter monitoring revealed atrioventricular dissociation with no significant sinus pauses, and hence pacemaker implantation was deferred. At 8-month follow-up, contrast echocardiography revealed systemic veins from the upper and lower limbs draining into the right atrium and an intact inter-atrial septum (Supplementary Video 2).

Discussion

Total drainage of systemic venous blood into the left atrium is an exceptional finding in the absence of severe intra-cardiac malformation. Usually, total anomalous systemic venous drainage is associated with other complex congenital heart defects in the form of atrioventricular septal defect or ventricular septal defect or patent ductus arteriosus to allow the systemic venous return to reach the pulmonary circulation. There exist very few case reports in the literature on total anomalous systemic venous drainage to the left atrium, and nine of them were successfully corrected surgically and associated with other congenital defects.[1–9] Total anomalous systemic venous drainage to the coronary sinus has also been reported in the literature and one such case was diagnosed prenatally.[10] Of the nine surgically

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corrected case reports of total anomalous systemic venous drainage to the left atrium, interrupted inferior caval vein was seen in five[1–4,9] and bilateral superior caval vein in two.[4,8] All of them had a persistent left superior caval vein. In our case, an interrupted inferior caval vein was draining through the azygous system to the dilated right-sided superior caval vein and there was no persistent left superior caval vein. There was no other associated complex intra-cardiac defect except for an atrial septal defect.

Embryologically, it has been hypothesised that total anomalous systemic venous drainage may occur either because of failure of regression of the right valve of the systemic venous sinus ("sinus venosus") or because of the systemic venous sinus being incorporated into the left atrium.

The clinical importance of these systemic venous anomalies is threefold. First, they are the rare cause of cyanotic congenital heart disease with unremarkable precordial examination. Second, total anomalous systemic venous drainage is another cause of equal oxygen saturation in all four cardiac chambers, other than in patients with total anomalous pulmonary venous drainage, with the only difference being that the mixing in total anomalous systemic venous drainage takes place in the left atrium. Finally, the drainage of the hepatic veins to the left atrium if left unrepaired during surgery may be a cause of cyanosis and pulmonary arteriovenous fistula seen during late follow-up. This may probably be because of an unknown hepatic factor needed for prevention of pulmonary arteriovenous fistula.

Conclusion

Total anomalous systemic venous drainage is a rare congenital cardiac anomaly and is usually associated with various other congenital heart defects. It is rare to have total anomalous systemic venous drainage in the patient with left heterotaxy syndrome without evidence of complex intra-cardiac defects.

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Supplementary material

For supplementary material referred to in this article, please visit http://dx.doi.org/doi:10.1017/S1047951112000819

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