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Brief Report

Unusual totally anomalous pulmonary venous connection in right isomerism and functionally univentricular heart

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Abstract We report a rare variation in the pattern of totally anomalous pulmonary venous connection in that two vertical veins drained into the left brachiocephalic vein from a common pulmonary venous confluence. The child had associated right isomerism and functionally univentricular heart. Awareness of this possibility may avoid a reoperation or even death.

Keywords: Double inlet ventricle; asplenia; visceral heterotaxy

THE ACCURATE DIAGNOSIS OF THE ANOMALOUS pulmonary venous connection is vital for the operative management of patients with congenital heart disease.[1–6] In patients with reduced pulmonary blood flow, the diagnosis of totally anomalous pulmonary venous connection may be missed preoperatively, and the anomaly may be recognised only intraoperatively.[2] We report a case of complex congenital heart disease where an unusual pattern of pulmonary venous drainage was only partly recognised intraoperatively.

Case report

An 8-month-old boy, weighing 4.2 kg, presented to us with recurrent cyanotic spells. He was born after a full term normal pregnancy, and had been cyanosed since birth. On examination, a grade II/VI ejection systolic murmur was heard, but there was no evidence of cardiomegaly or congestive cardiac failure. The non-invasive oxygen saturation at rest was 70 per cent as determined by pulse oxymetry. Chest radiography revealed normal cardiac size and pulmonary oligemia. Cross-sectional echocardiography established the diagnosis of double outlet of the right ventricle, hypoplastic left ventricle, non-committed ventricular septal defect, and pulmonary stenosis. A gradient of 80 mmHg was present across the right ventricular outflow tract to the pulmonary arteries. The angiocardiogram showed confluent pulmonary arteries. The pulmonary venous return was delayed, but the precise arrangement was not clear. The child was referred for consideration of a bidirectional Glenn anastomosis.

Totally anomalous pulmonary venous connection draining to the right superior caval vein via the left brachiocephalic vein was diagnosed during surgery. At operation, one vertical vein was identified. The pulmonary arteries were less than 50 per cent of the anticipated confidence limits for age and body surface area, as measured intraoperatively using Hegar dilators, and bilateral right atrial appendages were identified. The common pulmonary vein was anastomosed to the posterior left atrial wall, a bidirectional Glenn was performed, and the pulmonary trunk was transected (Fig. 1). Despite this, there was persistent desaturation and high mean pulmonary arterial pressures at 30 mmHg were noted in the postoperative period. A repeat study revealed an additional vein running parallel to left vertical vein on the lateral aspect. This was ligated in continuity at second operation (Fig. 2). The mean pulmonary arterial pressure fell to 16–18 mmHg. The child died on the 12th postoperative day due to septicemia. Permission for autopsy was denied.

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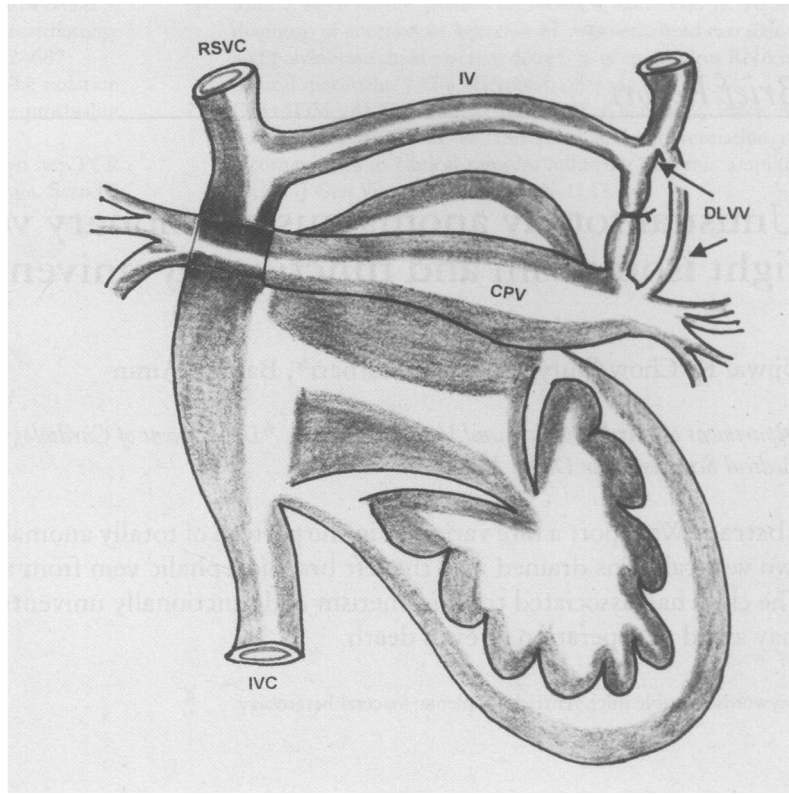

Figure 1.
Surgical technique. Totally anomalous pulmonary venous connection (TAPVC) to the right superior caval vein (RSVC) via double left vertical veins (DLVV). The common pulmonary vein (CPV) was anastomosed to the posterior left atrial wall (not shown). A bidirectional Glenn anastomosis was performed (not shown). The double left vertical veins were ligated on two occasions. Abbreviations: IVC. Inferior caval vein. IV. Brachiocephalic vein.

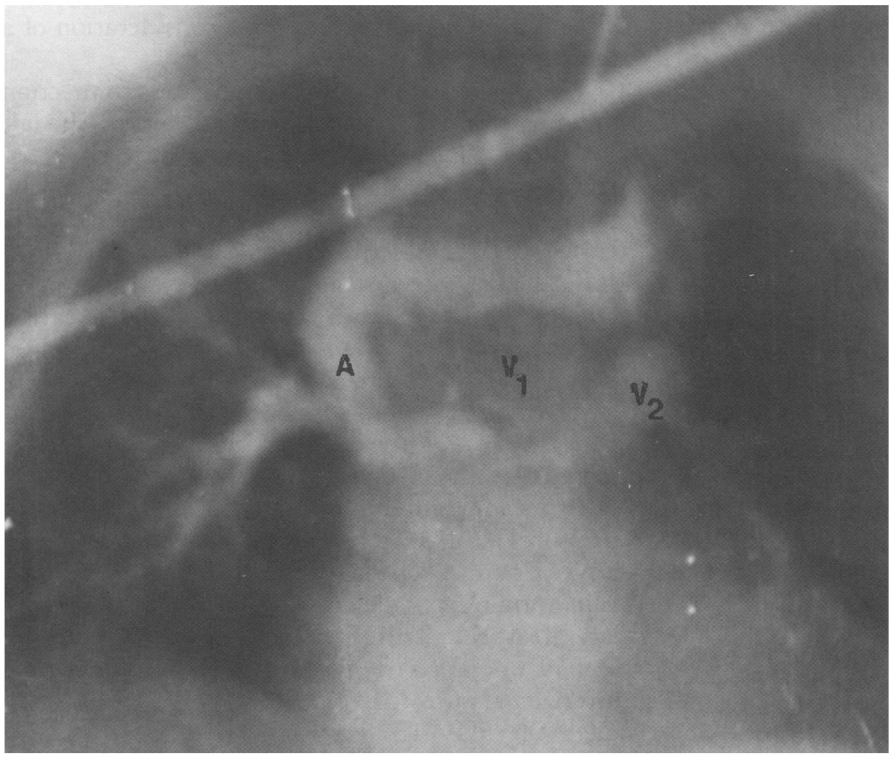

Figure 2.
Totally anomalous pulmonary venous connection to the right superior caval vein via double left vertical veins. The postoperative angiocardiogram shows a functioning bidirectional Glenn anastomosis (A), a previously ligated left vertical vein (V1), and an additional left vertical vein (V2) on the lateral aspect. This was ligated during the second operative procedure.

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Discussion

The variations in the anatomy of totally anomalous pulmonary venous connection are well known, and 'mixed drainage' occurs infrequently.[1–3,5–7] Two emissary veins leading to different sites are well described[3] but, to the best of our knowledge, two vertical veins from a single pulmonary venous confluence draining to the left brachiocephalic vein has not previously been described. Because of this lack of awareness, we ligated only one vertical vein, missing the other during the initial construction of the bidirectional Glenn anastomosis.. The embryological basis of these dual left vertical veins is not clear. More recently, Benassi and Edwards suggested the term superior emissary veins for the vertical veins.[8]

Identification of totally anomalous pulmonary venous connection in a patient undergoing a Glenn operation is of practical importance.[1,2,5,6] In our experience, during the last 11 years, 20 out of 600 patients with functionally univentricular heart had totally anomalous pulmonary venous connection (3.3%), 6 of whom died, including all 4 in whom the pulmonary venous abnormality was not recognised preoperatively or during surgery. Presently, it is our policy to identify the pulmonary veins routinely in all cases at the time of a Fontan operation.

In a collective review, Kanjuh et al.[3] identified the mixed type of totally anomalous pulmonary venous connections most commonly at supracardiac and cardiac levels, less commonly at supra and infracardiac levels, and rarely at cardiac and infracardiac levels. Cooley et al.[7] suggested that correction of totally anomalous pulmonary venous connection is of greater operative risk when multiple terminations are present than when there is single termination.

The association of functionally univentricular heart, right isomerism and totally anomalous pulmonary venous connection is well recognised. The great majority are obstructed and require urgent surgical intervention in infancy.[2,6,9,10] In our centre, the youngest patient with this combination was 6 months old, and only 15% of children had associated right isomerism. Possibly, this points to natural selection. In addition, they have shown themselves to be somewhat better candidates by surviving to 6 months of age.

The low supracardiac anomalous pulmonary venous connection constitutes a significant portion of the patterns encountered in patients with both visceral heterotaxy syndrome and non-heterotaxic anomalous pulmonary venous connection.[4–6,10] Various ingenious techniques have been described to manage this difficult subset of patients.[4–6,10] McElhinney, Reddy and Hanley described a technique of implanting the anomalous left pulmonary vein to the atrial stump of left superior caval vein, and anastomosis of the right pulmonary vein to the cardiac stump of the right superior caval vein, effectively conserving the suture line in two consecutive patients.[6] Another technique is transection of the superior caval vein sufficiently superior to the entrance of the pulmonary venous conduit to avoid possible obstruction.[6]

Bilateral superior caval veins are often of small size and, in the presence of low supracardiac anomalous pulmonary venous connection, an alternative may be to make an anastomosis between the pulmonary venous chamber and the left-sided atrium, ligate the vertical vein draining into the superior caval vein, and utilise the detached lower end of the superior caval vein for the cavopulmonary anastomosis. This would avoid a restrictive anastomosis which might result if the superior caval vein was transected superior to its junction with the pulmonary venous confluence. The prerequisite of all these techniques is the presence of a common pulmonary vein behind the left-sided atrium to allow construction of an unrestricted anastomosis. The absence of a posterior venous collector necessitates an alternative surgical technique. Extracardiac total cavopulmonary connection is an alternative in these selected cases

In conclusion, totally anomalous pulmonary venous connection may rarely take the form of two vertical or superior emissary veins originating from a common pulmonary venous confluence and extending to a supracardiac termination. Awareness of this pattern may be life-saving in some cases. Exclusion of the diagnosis of anomalous pulmonary venous connection is imperative in all patients with functionally univentricular hearts, either preoperatively or during the operative procedure.

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