Anomalous cross-sectional patient drainage probably produced tricuspid regurgitation, acute right ventricular distention, increased tension of the papillary muscles, and chordae tendineae and tear of the latter structure. Probably the moderate pulmonary hypertension still present in this case increased further the stress on the subvalvular apparatus, contributing to rupture of the septal leaflet chordae tendineae and to the severity of right ventricular failure following this rupture.

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Cross-sectional Echocardiographic Imaging of Supracardiac Total Anomalous Pulmonary Venous Drainage to a Vertical Vein in a Patient With Holt-Oram Syndrome:

David J. Sahn, M.D.; Stanley J. Goldberg, M.D.; Hugh D. Allen, M.D.; and Jesus M. Canale, M.D.

We report the unusual association of total anomalous pulmonary venous drainage in a three-year-old with Holt-Oram syndrome and a new technique for imaging.

Vertical vein drainage of anomalously draining pulmonary veins by suprasternal cross-sectional echocardiography.

Total anomalous pulmonary venous return represents a clinical spectrum, which in its minimally cyanotic (unobstructed) forms is occasionally found in a patient clinically suspected of having a large atrial septal defect.1,2 M-mode echocardiographic criteria exist for the diagnosis of total anomalous pulmonary venous return.3,4 Nonetheless, a recent study from our laboratory5 suggests that the common venous is often located above or below the heart and is overlooked on M-mode.6 In that paper, we outlined two-dimensional techniques for imaging anomalous venous chambers, emphasizing apex and subxiphoid four-chamber views. This report adds the utility of performing suprasternal notch echocardiography to image the superior vena cava and brachiocephalic venous systems to those techniques for diagnosing anomalous pulmonary venous return. This technique recently has allowed us to image a dilated vertical vein draining the pulmonary veins and entering into the dilated brachiocephalic trunk in a patient with total anomalous pulmonary venous return. The patient, incidentally, also has Holt-Oram syndrome6,7 and a familial history of atrial septal defect, an uncommon association.1

CASE REPORT

A three-year-old, 10.9-kg child was admitted for elective catheterization. She had been observed since infancy, with clinical diagnosis of large atrial septal defect without congestive heart failure, a diagnosis associated with the familial history of Holt-Oram syndrome. As an adult her father had been diagnosed as having secundum atrial septal defect associated with Holt-Oram syndrome. His thumbs were completely absent, with mild hypoplasia of the radius and a moderately large secundum atrial septal defect. During surgical closure of the atrial septal defect at age 31 years, he suffered a stroke while on cardiopulmonary bypass, resulting in significant, persistent loss of left upper extremity function. His daughter, who also had no thumbs, was followed up until three years of age with a clinical diagnosis of atrial septal defect by chest x-ray examination, ECG, and M-mode echocardiography. After much persuasion, the family allowed a hemodynamic evaluation.

On admission to the hospital, the child weighed 10.6 kg, visually was acyanotic, and had an exaggerated right ventricular precardial impulse. She had a harsh 3/6 systolic ejection murmur and a wide, fixedly split second heart sound with an increased pulmonary component. A grade 1-2/6 diastolic rumble was heard over the tricuspid area. Peripheral pulses were normal. Hemoglobin was 13.5 g/dl; hematocrit was 39.6 percent. An ECG showed moderate right ventricular hypertrophy of the rSr' wave volume overload type. A chest x-ray film showed increased pulmonary vascularity, right ventricular enlargement, and some widening of the upper mediastinum without a classic "snowman" pattern. The M-mode echocardiogram displayed features of right ventricular volume overload with flattened septal motion and a right ventricular dimension of 2.1 cm, without suggestions of pulmonary hypertension.
Cross-sectional echocardiogram was performed with a 3.5-MHz, 84° mechanical sector scanner (EkoSector, Smith Kline Instruments). Examination disclosed findings of severe right ventricular enlargement and flattening of the left ventricle. A large atrial septal defect was visualized on subxiphoid views and was estimated to be approximately 2 cm and in the secundum position. Pulmonary veins were identified on apex four-chamber view (Fig 1) and on subxiphoid view entering a leftward, posteriorly located chamber superior to the left atrium. The true left atrium was small. A suprasternal notch cross-sectional echocardiogram was performed in a short axis suprasternal plane, with a transducer scanning a coronal plane parallel to a line between the patient’s shoulders. The main pulmonary artery and aorta were identified in the center of the image, and a dilated superior vena cava was identified on the right side of the image (Fig 2). The brachiocephalic trunk was also dilated. Just distal to the entry of the left subclavian vein into the brachiocephalic trunk, a large, dilated, vertically oriented structure was visualized joining the brachiocephalic trunk. This structure could be traced to the common venous chamber and was imaged to receive the pulmonary veins. An echocardiographic diagnosis of total anomalous pulmonary venous drainage to the superior vena cava via vertical vein could be made from this invasive study.

At cardiac catheterization, the dilated vertical vein and brachiocephalic system could be immediately identified fluoroscopically on the left heart border. Arterial blood gas determinations revealed a PO₂ of 50 mm Hg, saturation of 89 percent with a pH of 7.45. Saturation was 88 percent in the left atrium and 87 percent in the left ventricle. Saturation in the left brachiocephalic trunk was 96 percent. Using inferior vena cval saturations of 74 and 76 percent and pulmonary artery saturation of 88 percent, a QP:QS ratio of 4:3:1 was determined. Right ventricular pressure was 40/12 mm Hg, and pulmonary artery pressure was 30/15 mm Hg with a mean of 20 mm Hg. Left ventricular pressure was 85/12 mm Hg, and there was no gradient across the atrial septum. Pullback from the pulmonary venous chamber to the superior vena cava to the right atrium showed consistent mean pressures of 17 mm Hg, with no suggestion of a significant gradient.

The left ventriculogram showed a small but normally functioning left ventricle with minimal, probably catheter-induced, mitral regurgitation. Pulmonary arteriography disclosed a dilated pulmonary artery system, and on the levophase, drainage of all four pulmonary veins to a common venous chamber draining via a vertical vein to the superior vena cava was demonstrated (Fig 2). At surgery, the vertical vein was ligated, and the common venous chamber was marsupialized to the left atrium, and patch closure of the atrial septum was accomplished. The pulmonary artery dimension was 1½ times that of the aorta, the atrial septal defect was 2.5 cm in diameter, and the vertical vein was described as being the same size as the pulmonary artery.

The patient has done well in the postoperative period. Repeated two-dimensional echocardiography, performed...
FIGURE 3. Postoperative still frame of a two-dimensional examination from the suprasternal notch shows reduction in size of the vertical vein and brachiocephalic vein as well as the position of ligation of the vertical vein structure. Abbreviations as in Fig 2.

from the suprasternal notch two weeks after surgery, imaged the ligated vertical vein structure (Fig 3). The pulmonary veins were imaged marsupialized into the left atrium.

DISCUSSION

Our own experience suggests that it is not uncommon to miss the M-mode echocardiographic diagnosis of total anomalous pulmonary venous drainage. In the case just cited, the family history with the presence of Holt-Oram syndrome suggested atrial septal defect. While some patients with Holt-Oram syndrome have been reported to have patent ductus arteriosus, coarctation, ventricular septal defect, transposition, single coronary artery or mitral valve prolapse, total anomalous pulmonary venous return, as far as we know, has been reported only twice in the English literature in association with Holt-Oram syndrome. The single patient reported by Gathman and Nadas also had anomalous drainage to the left superior vena cava system.

In this patient, M-mode echocardiography did not suggest the diagnosis, but the cross-sectional imaging study suggested the diagnosis and the site of anomalously draining pulmonary veins. The short axis or coronal suprasternal notch examination that we used differs from the suprasternal notch examination for cross-sectional echocardiography that initially was described for examining the aortic arch. Tajik has described the use of this coronal plane as a technique for imaging the superior vena cava. In our case, this plane of examination was used to further define the drainage of the pulmonary venous chamber imaged from subxiphoid. The images from the suprasternal notch allowed noninvasive imaging of the vertical vein.

This case report emphasizes the uncommon association of anomalous pulmonary venous drainage in patients with cardiac limb syndrome (Holt-Oram) and presents a technique for suprasternal notch cross-sectional imaging designed to image anomalous vertical vein (left superior vena cava) drainage of the pulmonary veins.

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