recognized causes of bronchial arterial enlargement, namely pulmonary hypertension and pulmonary arterial constriction. Experimental pulmonary venous obstruction alone has not resulted in enlarged bronchial arteries, although Nasrallah et al. noted bronchial arterial enlargement in their case. It may be that bronchial arteries enlarge whenever pulmonary blood flow falls.

Unequal distribution of pulmonary blood flow in children with transposition of the great arteries has been a subject of concern to several authors. Muster et al. described increased right pulmonary flow in many children with transposition of the great arteries and considered left venous obstruction a possible explanation; however, the distribution of flow was similar before and after surgery, suggesting that venous obstruction by the baffle was not a factor.

Our case has implications for other children surviving Mustard's operation. First, a lack of clinical symptoms can occur with severe or even complete unilateral pulmonary venous obstruction, despite previous experience. Secondly, although the patient is asymptomatic, the left lung does not participate in the exchange of gases, and this child's eventual prognosis is not known. Finally, it is likely that the left pulmonary veins have regressed in size over six years, and relief of the obstruction might have been possible had the diagnosis been made at the first postoperative catheterization. We would recommend, therefore, that after Mustard's operation the distribution of pulmonary blood flow should be evaluated by chest x-ray film and perfusion lung scan and that pulmonary venous anatomy should be investigated, regardless of symptoms, in any patient with gross inequality in the distribution of pulmonary blood flow.

REFERENCES

Echocardiographic Diagnosis of Atrial Septal Aneurysm in an Infant with Hypoplastic Right Heart Syndrome*

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Echocardiographic findings are described in a patient with hypoplastic right heart syndrome (pulmonary atresia type with intact ventricular septum and small right ventricular cavity) who had an associated atrial septal aneurysm. An unusual appearance of echoes behind the aorta bulging into the left atrium in diastole on both the M-mode and cross-sectional echo suggested this diagnosis prior to cardiac catheterization. The angiographic findings confirmed the diagnosis of right ventricular hypoplasia, pulmonary atresia and the large atrial septal aneurysm. The infant died after surgery and the atrial septal aneurysm was observed at autopsy. The importance of the diagnosis of the atrial septal aneurysm and its association with restriction of right-to-left atrial shunting prompts this report.

Infants born with significant obstruction to right ventricular inflow or outflow often require patency of the atrial septum in the form of an atrial septal defect or patent foramen ovale for their survival. Thus, emergency therapy for infants with forms of hypoplastic right heart syndrome (pulmonary atresia, tricuspid atresia) includes palliative systemic-pulmonary anastomosis and balloon atrial septostomy or operative septectomy with or without pulmonary valvulotomy. The pathologic findings of aneurysmal dilation of the atrial septum have previously been described. Additionally, the angiographic diagnosis during life of an atrial septal aneurysm in an infant with tricuspid atresia has recently been reported. In this latter report, the association of this finding with a restriction of right-to-left atrial shunting was emphasized. We wish to report the echocardiographic findings in an infant with hypoplastic right heart syndrome (pulmonary atresia, tricuspid atresia, hypoplastic right ventricle, small aorta, patent foramen ovale) whose atrial septum was aneurysmal.

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monary atresia type 1) in whom the diagnosis of an atrial septal aneurysm was made echocardiographically prior to catheterization and in whom the angiogram and autopsy confirmed this unusual associated malformation of the atrial septum.

**Figure 1.** This M-mode echocardiogram shows a sweep from the normal left ventricular cavity and mitral valve complex (AMVL = anterior mitral leaflet; PMVL = posterior mitral valve leaflet) to the aorta. A thickened right ventricular wall (W) and a small right ventricular cavity (RVC) are evident. The abnormal echoes are visualized behind the aorta. The walls of the aneurysm separate most in late diastole and early systole. LVPW = left ventricular posterior wall.

**CASE REPORT**

The patient was a 2,050-gram girl of 42 weeks' gestation born to a gravida 4, para 1 mother after an apparently uncomplicated delivery. Apgar score was 8 at 5 minutes with some acrocyanosis. At four hours of age, the patient was

**Figure 2.** Polaroid still frame and line drawing from the real-time cross-sectional echocardiogram demonstrates atrioventricular anatomy in the transverse plane. Image orientation is shown on the compass. The right ventricular cavity (RV) is small and the tricuspid valve (TV) is seen with it. The right atrial cavity (RA) appears dilated and the extra abnormal echoes are in the position of the interatrial septum (dotted lines) between the right and left atria (LA). In real-time, these echoes moved toward the left in diastole and toward the right in systole. An M-mode pattern similar to the abnormal echoes in Figure 1 could be derived by recording individual element output from this area. The origin and course of the aorta (AO) are superimposed on the line drawing. S = septum; LV = left ventricle; MV = mitral valve.
noted to be cyanotic and a chest x-ray film revealed a large heart with normal pulmonary vascularity.

The infant was transported to the University of Arizona Health Sciences Center for Cardiac evaluation. Upon arrival, she was mottled and cyanotic. Blood pressure was 80 mm Hg systolic in the right arm, left arm and left leg. Heart rate was 150/minute, respirations 45-55/minute with deep but unlabored breathing. Cardiac examination revealed an active precordium with no thrills and a grade 4/6 long systolic ejection murmur at the left upper sternal border transmitted widely over the chest. The second heart sound appeared to be single and a prominent third heart sound was heard. The liver was firm with a rounded edge 4 cm below the right costal margin. The electrocardiogram revealed left axis deviation and a paucity of right ventricular forces. Echocardiogram revealed a small right ventricular cavity and unusual echoes behind the aorta thought to be within the left atrium (Fig 1).

The pulmonary valve could not be demonstrated.

Because of confusion as to the source of echoes in the left atrial cavity, a real-time cross-sectional echocardiogram was performed with a 7 MHz linear multiple-crystal system. In the transverse plane, the cross-sectional echocardiogram showed that the echoes arose in the position of the atrial septum behind the aorta, rather than within the left atrial cavity (Fig 2). In real-time, these echoes moved toward the left in diastole and toward the right in systole. The pulmonary valve could not be demonstrated using the cross-sectional system, but the diminutive size of the right ventricular cavity was confirmed.

At cardiac catheterization, left atrial blood gas levels (100 percent O₂) revealed a pH of 7.27, Pco₂ of 42 mm Hg, Po₂ of 38 mm Hg and a bicarbonate level of 19. Pressure determinations revealed an elevated right ventricular pressure of 60/35 mm Hg in the small right ventricular cavity. Right atrial mean pressure was 18 mm Hg with an early diastolic gradient of approximately 6 mm Hg across an apparently stenotic tricuspid valve. The left atrium was entered from the right atrium through the foramen ovale. Mean left atrial pressure was approximately 12 mm Hg and the mean gradient was 8 mm Hg (right atrium > left atrium) across the atrial septum on pullback. An angiogram performed in the right atrium revealed a dilated right atrial cavity. The tricuspid valve appeared to have a stenotic annulus and regurgitation was noted. The left atrium opacified as did the small right ventricle. The great vessels were normally oriented and the pulmonary artery filled through a left-to-right shunting patent ductus arteriosus. A prominent atrial septal aneurysm bulging toward the left into the left atrium was seen early on the right atrial angiogram. Right ventriculogram demonstrated a hypoplastic right ventricular chamber with a hypoplastic and atretic pulmonary valve. A balloon atrial septostomy was performed with apparent success and the infant was taken to surgery where a pulmonary valvulotomy and tricuspid valve dilation were performed under inflow occlusion. A Waterston anastomosis was then performed. Despite an improvement in oxygenation, the infant was anuric in the postoperative period and developed a diffuse bleeding tendency. She died 12 hours after operation. At autopsy, pulmonary atresia, type 1 (pulmonary valve surgically opened), normally formed but small tricuspid valve leaflights and the atrial septal aneurysm were demonstrated (Fig 3).

**DISCUSSION**

Bulging of the interatrial septum toward the left atrium in diastole (during atrial systole), in conditions where the right ventricle is hypoplastic, obstructed or both, or when tricuspid atresia exists, has important hemodynamic implications. It suggests that there is obstruction to right-to-left flow across the atrial septum and probable elevation of right atrial pressure. Initial confusion arose in this case as to the source of the echoes in the left atrium and there was some concern that this might be a left or right atrial tumor in a newborn. Such confusion has previously been reported regarding the source of a right atrial filling defect on angiography in an adult who had a similar aneurysm. The echoes in this case were quite mobile and differed significantly from those normally seen within the left atrial cavity or those reported in association with total anomalous pulmonary venous return. Moreover, the echoes of the aneurysm were not confused with the mitral valve, as on sweep they appeared more superiorly behind the aorta and were separated in early systole (Fig 1). Identical M-mode patterns were recorded using the multi-element array from the area of the atrial septum, shown in Figure 2 (dotted lines). Finally, in support of the reproducibility of this finding, we have recently been shown a nearly identical echo tracing derived from an infant who at autopsy had tricuspid atresia and an atrial septal aneurysm.

![Figure 3. Pathology photograph and drawing shows the position of the atrial septal aneurysm as viewed from the left atrium (LA) and expanded by the insertion of a scalpel handle. The arrow shows the position of the small interatrial defect and the tear attributed to the balloon atrial septostomy. LV = left ventricular wall; MVO = mitral valve orifice.](image-url)
Reconstruction of the Superior Vena Cava in a Patient with a Thymoma*

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Surgical treatment was attempted in a 71-year-old man who had superior vena cava syndrome caused by incomplete obstruction of the superior vena cava with a thymoma. Using a temporary internal shunt, the superior vena cava was almost entirely resected together with the tumor and reconstructed by autogenous venous grafts. Patency of the reconstructed vein was proved by angiography one year after the operation. He has been asymptomatic for 15 months after surgery.

Surgical treatment has been rarely indicated in malignant tumor involving the superior vena cava, not only because of extreme extension of the tumor, but also because of poor results obtained from the resection and reconstruction of the superior vena cava.

We attempted to resect and reconstruct the superior vena cava of a patient in whom the superior vena cava was involved by a thymoma. This report describes the technique and results of the reconstruction of the superior vena cava in this patient.

Case Report

A 71-year-old man was admitted to Nakano National Chest Hospital on May 6, 1975, because of progressive dyspnea, swelling of the face, and hoarseness during the four months prior to admission. Physical examination revealed a swelling and venous distension in the head, neck and upper extremities.

Laboratory examinations, liver function tests and blood chemistry levels were within normal limits. Roentgenograms of the chest showed a mass in the upper anterior mediastinum. Superior vena cavaograms showed complete blockage of the left innominate vein and marked stenosis of the right innominate vein and superior vena cava as well (Fig 1). The venous pressure of the right innominate vein was 400 mm of saline solution. Preoperative electrocardiograms were normal. On May 27, 1975, he was operated upon. The mediastinum was explored through a median sternotomy. A tumor, which measured approximately 10 cm in diameter, involved the left innominate vein and occluded it completely. The tumor also involved the right innominate vein and superior vena cava, covering 6 cm in length, producing marked stenosis. After administering heparin, a No. 28F caval catheter (Bardic) with two side-holes at the proximal portion, was introduced from the right auricular appendage into the lumen of the intact right innominate vein, passing through the lumen of the involved portion of the superior vena cava and right innominate vein. Under this temporary internal shunt, the involved area of the wall of the superior vena cava and right innominate vein, three-fourths of the circumference in width and 7 cm in length, was resected together with the tumor. Therefore, only a quarter of the circumference of the wall of the veins was left intact. During the procedure, the left innominate vein and azygos vein were ligated and the uninvolved portion was divided. Prior to removal of the tumor, the saphenous and external jugular veins had been resected and opened longitudinally for making grafts. The superior vena cava was reconstructed by joining three long strips of these venous grafts, using the running over-and-over suture technique with 6-0 Prolene (Fig 2).

Histologically, the resected tumor was a thymoma of

Figure 1. A superior vena cavo gram before operation showing complete blockage of the left innominate vein and marked stenosis of the right innominate vein and superior vena cava.

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