Partial Anomalous Pulmonary Venous Connection
Associated with Coarctation of the Aorta:
Total Surgical Correction at One Operation*

Report of a Case

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The concomitant appearance of congenital lesions is a well known phenomenon which is especially true of the cardiovascular system. The recognition of multiple abnormalities allows total correction during one operation.1,2 Of greater importance is the fact that if a second lesion is not recognized, it may cause either serious complications or even death when extracorporeal circulation is subsequently utilized for repair. The subject of this report presented two congenital cardiovascular abnormalities, recognition of which at surgery afforded us the opportunity to correct totally the patient’s anomalies without recourse to a second procedure.

Case Report

This 45-year-old white woman was referred to the City of Hope Medical Center for investigation of a heart murmur of many years duration and hypertension first discovered three years prior to her admission. Her primary symptomatology consisted of headaches and easy fatigability. On physical examination, the blood pressure in her right arm was 150/75 mm Hg and the blood pressure in the lower extremities was unobtainable. There was a Grade II to III precardial systolic murmur heard best at the left sternal border radiating into the neck. The radial pulses bilaterally were graded as IV plus; abdominal aortic pulsation was palpable and graded as II plus. Femoral pulses both right and left were graded as II plus. The popliteal, dorsalis pedis and posterior tibial pulses were absent bilaterally. The patient’s liver was palpable 1 cm. below the right costal margin; the edge was smooth and non-tender.

Laboratory data were essentially normal. Electrocardiogram was within normal limits, although right ventricular hypertrophy was suggested. Vectorcardiogram clearly indicated mild right ventricular hypertrophy. Chest x-ray films were thought to be normal for age, and the roentgenologist made a specific comment that there was no increase in pulmonary vascularity.

In November, 1960, right and left heart catheterization was carried out. This revealed a coarctation of the aorta at the usual site and arterialization of blood high in the right atrium or in the low superior vena cava indicating a left-to-right shunt of approximately 35 per cent (Fig. 1). The presence of the coarctation was confirmed by angiography; dye dilution curves confirmed the presence of the left-to-right shunt. In addition to a coarctation of the aorta, it was believed the patient had an atrial septal defect possibly associated with anomalous pulmonary venous drainage. The anticipated course of treatment for this patient was to be initial correction of the coarctation to be followed in a few months by correction of the shunt utilizing extracorporeal circulation.

Accordingly, she was re-admitted to the hospital in June, 1961 for surgery. Left posterolateral thoracotomy was performed entering the pleural cavity in the bed of the non-resected fifth rib. A moderate amount of collateral circulation was encountered on opening the chest wall. On exploring the pleural cavity, typical coarctation of the aorta distal to the left subclavion artery was found. Further examination revealed a large vein approximately 1 cm. in diameter lying along the mediastinum running from the upper aspect of the left hilum towards the apex of the chest (Fig. 2A). On subsequent examination with the pericardium open it was apparent that this was an anomalously connected left superior pulmonary vein, draining the left upper lobe and emptying into the left innominate vein. Examination of the fold of Marshall confirmed the fact that there was no left superior vena cava.

Because the gradient across the coarctation as measured at the time of catheterization was not great, the procedure was carried out with moderate hypothermia so that at the time of the cross clamping of the aorta the patient’s rectal temperature was 33°C. The coarctation was resected and aortic continuity restored without a graft. The anomalous pulmonary vein was then dissected out in its entirety, ligated and divided high in the chest. The left atrial appendage had been prepared previously and an anastomosis was carried out utilizing a continuous suture of 6-0 arterial silk approximating the endocardium to the...
intima of the vein as described by Gerbode and Hultgren (Fig. 2B). The pulmonary artery to the left lung was not occluded during this procedure. At completion, the anastomosis was patent and there was no congestion of the left upper lobe. The patient's postoperative course was uncomplicated except for a short febrile episode due to a urinary tract infection. At no time did she develop hemothysis or evidence of congestion of the left upper lobe clinically or roentgenographically. At the time of her discharge from the hospital, 17 days after surgery,
the blood pressure in her upper extremities was 120/70 mm.Hg and in her lower extremities 130/80 mm.Hg. All of her pulses were graded as IV plus.

The patient returned in October, 1961 for re-study. In the interim she had done quite well, engaging in normal activity without symptoms. Physical examination revealed blood pressure of 125/80 mm.Hg in both arms. There was a Grade II systolic murmur at the apex. Cardiac catheterization was carried out and revealed no evidence of cardiovascular shunts (Fig. 1). Dye dilution curves with injections of cardiogreen into several sites were normal.

DISCUSSION

Anomalous connection of part or all of the left pulmonary veins has been previously reported.1,4,10 Various authors describe connection to either a persistent left superior vena cava or left innominate vein. Developmentally, this originates as drainage into the left common cardinal vein which may persist as a left superior vena cava or left innominate vein.1,10 In our case, the fold of Marshall revealed no vessel indicating that the flow was cephalad to the innominate vein. Other lesions previously described with this anomaly include coarctation of the aorta,5 mitral stenosis1,4,10 and atrial septal defect.4,3,4

Anomalous venous connection of all or part of one lung can mimic atrial septal defect both clinically and at cardiac catheterization.1,10 Evidence of increased pulmonary blood flow and a pulmonic systolic murmur are common features of both defects. At cardiac catheterization, arterIALIZATION at atrial level is also found with both defects. Differentiation can be definitively made by indicator dye dilution curves after separate injections into the right main pulmonary arteries as described by Swan and his associates.11 This technique is worthy of consideration whenever atypical features suggest something other than the usual type of atrial septal defect. Thus, in the case just described, increase in oxygen saturation in the low SVC or high right atrium in retrospect suggests the anomaly eventually found at surgery. Angiography may be the clearest way of gaining an accurate diagnosis preoperatively. Finally, however, it is the obligation of the operating surgical team to assess carefully the anatomic and physiologic lesions that present themselves in the chest at the time of repair of a lesion. If the vein in this case had not been noted in the operating room an atrial septal defect or anomalous venous drainage on the right would have seemed a certainty. At a subsequent operation, even more serious complications than a negative right thoracotomy could have occurred particularly if extracorporeal circulation were utilized.

REFERENCES


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