Left Subclavian Steal, Interrupted Aortic Arch, Complete Transposition of the Great Vessels and Single Left Ventricle*

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Congenital subclavian steal consists of atresia of the proximal end of the subclavian artery, while the blood supply to the distal portion of the subclavian artery originates from the circle of Willis. It should be suspected in complicated malformations of the heart associated with marked hypoplasia or interruption of the aortic arch. Minimal differences in blood pressure determinations may be an important clue in the diagnosis of this anomaly. The diagnosis may be confirmed by angiography with careful attention to late arterial phase films.

Among the 27 cases of congenital subclavian steals reviewed by Pieroni and co-workers1 in 1972, nine had some form of interruption or marked coarctation of the aorta involving the origin of the subclavian artery. Various malformations of the heart with this complicating anomaly have been described, eg, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, supravalvar aortic stenosis, etc, but to our knowledge none with single ventricle and complete transposition of the great vessels. The rarity of such a combination prompts us to report it.

CASE REPORT

A five-week-old white girl was referred to Jackson Memorial Hospital in Miami, Florida because of heart failure secondary to suspected congenital heart disease. Family his-

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Table 1—Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Site</th>
<th>Saturation</th>
<th>Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td>02%</td>
<td>4.5 mm Hg</td>
</tr>
<tr>
<td>RA</td>
<td>42</td>
<td>4 (m)</td>
</tr>
<tr>
<td>LPV</td>
<td>81</td>
<td>—</td>
</tr>
<tr>
<td>LA</td>
<td>84</td>
<td>13 (m)</td>
</tr>
<tr>
<td>LV apex</td>
<td>84.5</td>
<td>95/8</td>
</tr>
<tr>
<td>Hypoplastic</td>
<td>76.5</td>
<td>95/8</td>
</tr>
<tr>
<td>Chamber</td>
<td>72</td>
<td></td>
</tr>
<tr>
<td>Ace Aorta</td>
<td>75</td>
<td>90/50</td>
</tr>
<tr>
<td>R Fem Art*</td>
<td>76</td>
<td>70/56</td>
</tr>
<tr>
<td>Hypoplastic</td>
<td>72</td>
<td></td>
</tr>
<tr>
<td>Chamber*</td>
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*Taken simultaneously.

tory was irrelevant. She was the product of a normal pregnancy and delivery, with a birth-weight of 3.2 kg (7 lb). On admission she showed questioned peripheral cyanosis and moderate grayish discoloration of the skin. She was in moderate respiratory distress with a respiratory rate of 90 per minute. Rales were present in the left lower lobe. The cardiac apex was felt at the fifth left intercostal space 1 cm outside the midclavicular line, with no thrill. The first heart sound was increased and there was a grade 2/6 systolic ejection murmur over the second and third left intercostal spaces. The second sound was single and loud over the pulmonary area. Pulses were rather weak in the four extremities. Blood pressures were determined by cuff method: right upper extremity 130 mm Hg, left upper extremity 125 mm Hg, lower extremities 120 mm Hg. The liver was enlarged 3 cm below the right costal margin at the midsclavicular line and the spleen was palpable. The electrocardiogram (Fig 1) showed left ventricular and left atrial enlargement. Vectorcardiogram indicated wide posterior counterclockwise loop. Chest x-ray examinations showed marked left ventricular and left atrial enlargement with increased pulmonary markings. Barium swallow had no retroesophageal impression. She was treated with digoxin and diuretics with significant improvement in her cardiac status. On the third day after admission cardiac catheterization and angiography were performed.

Cardiac catheterization revealed a single ventricle which was entered from either the right or left atria. The catheter was advanced through a ventricular septal defect into a rudimentary anterior chamber and then into the ascending aorta. The pulmonary artery was not entered. Hemodynamic data are shown in Table 1.

Biplane ventricular angiography (Fig 2, 3) re-

Figure 1. Electrocardiogram showing signs of left ventricle enlargement.
Figure 2. AP angiocardiogram into left ventricle. Arrow tip points to the diverticulum-like structure in right subclavian artery, and its tail to the pulmonary artery pouch described in the text. AO=aorta, PA=pulmonary artery, and LV=single ventricle.

Figure 3. Lateral left ventricular angiocardiogram. No aortic arch is seen. RV=rudimentary right ventricle, VSD=ventricular septal defect, DSC=descending aorta. Arrow points to patent ductus arteriosus.

Figure 4. AP aortic root injection. Arrow points to diverticulum in right subclavian artery.

Figure 5. Late film of aortic root angio, showing retrograde filling to left vertebral and subclavian arteries.

The patient tolerated the procedure well and was discharged on the seventh day after the study. Despite use of extensive cardiotonic drugs, she remains in chronic cardiac failure at 16 months of age.

Discussion

Becker and associates,² in a review of congenital subclavian steal, described all the anatomic potentials for the right lung. There was interruption of the aorta beyond the left carotid artery. The descending aorta, located to the left of the spine, filled from the pulmonary artery through a left-sided ductus arteriosus. A second injection in the root of the aorta (Fig 4, 5) showed a competent bicuspid valve. The ascending aorta gave rise to the right innominate artery, which branched into the right subclavian and carotid arteries. There is complete interruption of the aorta beyond the left carotid artery and on the late films retrograde flow through the left vertebral artery into the distal portion of the left subclavian produced the opacification of this vessel. Despite use of extensive cardiotonic drugs, she remains in chronic cardiac failure at 16 months of age.
this phenomenon. A rare case of congenital pulmonary and subclavian arteries steal syndrome has been reported by Shaher and co-workers.

The present case has been classified as having a left-sided aortic arch, no retroesophageal compression and low pressure in the left subclavian artery due to anatomic obstruction at the ostium of this vessel. We assume the latter had to arise from the descending aorta near the isthmus. However, no connection is seen angiographically due to tight stenosis or atresia of its proximal segment.

The diverticulum-like structure in the right subclavian artery and the small pouch at the junction of the right pulmonary artery and the main peaked trunk (Fig 2) are probably remnants of a right ductus arteriosus.

The single ventricle is a left one as shown by the angiogram (Fig 2, 3). The sinus of the malformed right ventricle forms a common cavity with the left ventricle, while its conal portion, isolated from the sinus, connected with the main cavity of the single ventricle through a ventricular septal defect. The ventricular chamber delivers blood into the lungs and descending aorta through the pulmonary artery and to the transposed aorta through the ventricular septal defect.

The difference of 20 mm Hg in systolic pressure between the ascending aorta and the femoral artery is probably caused by the resistance and pressure drop at the level of the ductus arteriosus supplying the descending aorta. Blood pressures taken by cuff differed by 25 to 30 mm Hg from those obtained during catheterization. This may be due to the fact that the baby was well sedated during the latter, while during physical examination on admission, she was restless and crying. Retrospective review of the physical findings in this case revealed no definite clue to the existence of a left subclavian steal, except perhaps the 5 mm Hg difference in systolic blood pressure between the right and left arm. When minimal differences in blood pressure are present clinically with the use of cuff method, they should be verified by methods which have a higher degree of accuracy (Doppler). However, if these pressure differences are present, regardless of how minimal, a high index of suspicion and angiography may discover subsequent congenital subclavian steal syndrome.

REFERENCES

Strangulation of the Left Atrial Appendage through a Congenital Partial Pericardial Defect*

Erwin Robin, M.D., F.C.C.P.,** Sunilendu N. Ganguly, M.D.,** and Marcia S. Fowler, M.D.†

The first case of strangulation of a left atrial appendage through a partial congenital pericardial defect is presented. Surgery consisted of a left atrial appendectomy and closure of the defect.

In recent years the diagnosis of congenital pericardial defects has been made with increasing frequency. Sudden death has been attributed to cardiac strangulation, or associated lesions such as congenital heart disease. This report describes the first case of strangulation of left atrial appendage through a congenital partial pericardial defect. Emergency surgery was effective in correcting this situation.

CASE REPORT

The patient, a 28-year-old man, was admitted because of an abnormal cardiac silhouette was noted on a routine chest x-ray examination. He was asymptomatic, and his past medical and family histories were unremarkable. The physical examination was unremarkable except for a grade 2/6 mid-systolic murmur best heard over the pulmonic area.

On admission, routine laboratory data were within normal limits. A chest x-ray film showed a mediastinal mass in the area of the main pulmonary artery (Fig 1). During fluoroscopy, this mass appeared to pulsate paradoxically with the pulmonary artery.

Cardiac catheterization revealed normal pressures, oxygen saturations, and cardiac output. Herniation of the atrial appendage through a congenital partial defect was confirmed by induction of a left pneumothorax with 300 ml of air (Fig 2).

Because of an uneventful hospital course the patient was released. Two months later he was readmitted because of the sudden onset of crushing chest pain. Physical examination revealed an acutely ill patient. Blood pressure was 70/50 mm Hg, heart rate 130 per minute and regular, respiratory rate 34 per minute and rectal temperature 38.5°C. White blood cell count was 14,000/mm³ with 82 percent neutrophils. An ECG revealed sinus tachycardia. The chest x-ray examination was similar to the one obtained on the first admission.

Because of the possibility of strangulation of the left atrial appendage through the pericardial defect, emergency surgery was performed. Left thoracotomy was performed through the fifth intercostal space. A defect measuring 3 by 4 cm was found at the base of the left side of the heart. The appendage was fixed to the pericardium by multiple adhesions. Its surface had a dusky appearance with many small focal hemorrhages. The edges of the defect formed a tight ring near the junction of the appendage and atrium. The

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