A Case of Aortic Atresia Associated with Right Aortic Arch

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SUMMARY
A case of aortic atresia associated with right aortic arch is described. This is the second case in the literature. The diagnosis of aortic atresia was made prior to death by right heart catheterization with angiocardiography and this was confirmed at autopsy. The necessity of an early and accurate diagnosis of this disease is discussed for providing more opportunity of surgical treatment.

Additional Indexing Words:
Aortic atresia Right aortic arch Angiocardiographic finding

Aortic atresia which is invariably combined with hypoplasia of the left side of the heart is relatively uncommon. To our knowledge, less than 180 cases have been reported in the literature, the majority of them having been found at autopsy. Consequently, only a few reports are available about the cardiac catheterization with angiocardiography of patients with this defect. In addition, a combination of aortic atresia and right aortic arch is very rare, only one report having been made in the literature. We therefore wish to report a case of aortic atresia associated with right aortic arch because of its very low incidence. In this report we also describe main findings of the diagnostic cardiac catheterization with angiocardiography.

CASE REPORT

A 11-day-old boy was transferred to Hokkaido University Hospital because of tachycardia, tachypnea, and hepatomegaly. He was the first child of a 23-year-old healthy mother. Birth was full term, delivery normal and the pregnancy uncomplicated. The birth weight was 2,700 Gm. From the time of birth, tachypnea and tachycardia were observed. On the fourth day of life he received a digitalis therapy along with oxygen administration because of the appearance of dyspnea.
Table I. Catheterization Data

<table>
<thead>
<tr>
<th>Site</th>
<th>$O_2$ Saturation (%)</th>
<th>$PO_2$ (mmHg)</th>
<th>$PCO_2$ (mmHg)</th>
<th>Pressure (mmHg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>54</td>
<td>30</td>
<td>57.5</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>63</td>
<td>34</td>
<td>58</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>79</td>
<td>44</td>
<td>54</td>
<td>12/3 (5)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>83.3</td>
<td>49</td>
<td>52.5</td>
<td>90/0 (75)</td>
</tr>
</tbody>
</table>

Mean pressure is indicated in parentheses. Right ventricle showed high pressure almost equal to systemic pressure.

and edema in the extremities. At the time of admission, slight cyanosis existed around the mouth. The respiratory rate was 72 per minute with sternal and costal retractions. The peripheral pulse could be weakly palpable. The lung was clear to auscultation. The liver was palpable 4.5 cm below the right costal margin. The spleen was not palpable. A grade 2 to 3 of 6 systolic ejection murmur was audible along with the left sternal border. A chest X-ray film showed marked cardiomegaly and increased pulmonary vascularity. The electrocardiogram revealed right axis deviation of plus 145° and right ventricular hypertrophy.

Laboratory data were following: red cell count, \(469 \times 10^4/mm^3\); white cell count, \(17,600/mm^3\); platelet count, \(188,000/mm^3\); hematocrit, 49%; hemoglobin, 14.6 Gm/100 ml; serum electrolyte: Na, 135.2 mEq/L; K, 5.9 mEq/L; Cl, 92.2 mEq/L; Ca, 4.7 mEq/L; P, 3.9 mg/100 ml.

On the fourth day of life serum electrolytes were examined at the maternity hospital and serum Na and K were 126 mEq/L and 6.1 mEq/L, respectively. Such abnormal values in serum electrolytes reached within normal limits after transfer to Hokkaido University Hospital.

Right heart catheterization with angiocardiography was performed on the second day in our hospital. Hemodynamic data obtained by the catheterization are shown in Table I. The high oxygen content in right atrium and ventricle was certainly produced by the flow of left atrial blood through the patent foramen ovale.

A right ventricular angiogram obviously revealed large patent ductus arteriosus, dilated main pulmonary artery and right aortic arch (Fig. 1). The hypoplastic ascending aorta was also revealed radiographically in a retrograde fashion, although the left ventricle was not seen (Fig. 2). On the basis of these findings, the diagnosis of aortic atresia was made. From the time of admission, vigorous digitalis therapy was continued and diuretic was injected twice a day. The patient was kept in an incubator with high oxygen concentration and the intake of water was strictly controlled. In response to this therapy, tachycardia, tachypnea, and hepatomegaly were gradually reduced and edema in the extremities disappeared. General conditions of the patient appeared to improve. On the 31st day of life, despite such intensive therapy, he suddenly died prior to surgical treatment.

At autopsy, hypoplastic ascending aorta, aortic atresia, and right aortic arch were confirmed. Furthermore, there existed severe mitral stenosis, severe hypoplastic left ventricle, dilated main pulmonary artery, large patent ductus arteriosus, patent foramen ovale, hypertrophied and dilated right atrium, and right ventricle. The ventricular septum was intact.
Fig. 1. Angiocardiogram with right ventricular injection of contrast medium, anteroposterior view: The dilated main pulmonary artery and right aortic arch are clearly demonstrated.

Fig. 2. Lateral view: The hypoplastic ascending aorta is faintly revealed in a retrograde fashion (arrow) and also large patent ductus arteriosus is demonstrated. These findings suggest strongly the presence of aortic atresia.

This is the second case in the literature of aortic atresia associated with right aortic arch.

DISCUSSION

Congenital cardiovascular anomalies are one of the major causes of death in newborn infants, and aortic atresia is generally considered to be rare. This defect has been intractable and the prognosis hopeless. But, 2 cases in the literature are of special interest. One is a patient who survived long with hypoplastic left heart syndrome, including aortic atresia. This may reveal a desirable hemodynamics contributing to long life and give a suggestion to surgical approach. The other is a case of successful surgical palliation for this syndrome. In our case, the patient lived for 31 days, a life-span longer than the mean survival of 3 to 5 days reported by the previous authors.

This may mainly be due both to the patent foramen ovale of appropriate
size and to large patent ductus arteriosus. Thus it appears adequate that the proper creation of an atrial septal defect to improve the left-to-right shunt is essential and palliative for longer survival. This procedure is possible by balloon atrial septostomy at the time of cardiac catheterization. For the effective surgical treatment, it is necessary to differentiate the aortic atresia from other cardiac defects. However, there have been a few cases in which accurate diagnosis of this defect was accomplished by means of angiography. In our case, the diagnosis of aortic atresia was made by right heart angiocardiography and this was confirmed at autopsy. This is one of the rare cases in which accurate diagnosis was obtained prior to death, mainly owing to successful demonstration of hypoplastic ascending aorta by means of right-side catheterization.

Watson et al.7) insisted that cardiac catheterization with venous angiography is not very useful and retrograde aortography is most informative. However, the retrograde aortography would not always be necessary, even though it appears most helpful for more accurate visualization of hypoplastic ascending aorta. We emphasize here that an earlier and accurate diagnosis of congenital cardiac defects, including aortic atresia, would provide more opportunities of early surgical correction and longer survival.

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References