INTERRUPTION OF THE AORTIC ARCH WITH PATENT DUCTUS ARTERIOSUS AND VENTRICULAR SEPTAL DEFECT IN AN ADULT

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A 21-year-old female was diagnosed as having a triad of interruption of the aortic arch, patent ductus arteriosus and ventricular septal defect. The patient died from bacterial endocarditis associated with acute peritonitis and perirenal abscess at the age of 31. This is the third longest survival of a case affected with the above triad, as far as we know.

INTERRUPTION of the aortic arch is a rare congenital anomaly with an unfavorable prognosis. The patients usually die in infancy. Up to now there have been only 18 cases, including 8 Japanese, who survived over the age of 15. The following is a report of a female with this anomaly associated with patent ductus arteriosus and ventricular septal defect and survived to the age of 31.

Case Report

First Admission
A 21-year-old female was admitted to our hospital for evaluation of heart disease. She was born prematurely as one of twin sisters. Her birth weight was 1,600 g and later development was fair. She was discovered cyanotic at the age of 3 when she saw a doctor for respiratory infection. The patient complained of shortness of breath from childhood. Bloody sputum appeared 5 months before admission and she suffered from a hemoptysis and a syncopal attack 10 days before admission.

Key Words:
Interruption of the aortic arch
Patent ductus arteriosus
Ventricular septal defect
Pulmonary hypertension

Fig.1. Chest x-ray film shows marked dilatation of proximal pulmonary arteries and clear peripheral lung field.

(Received January 20, 1981; accepted March 1, 1982)
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Her family history was not contributory. Her twin sister is doing well.

On admission, the patient was markedly cyanotic. Clubbed fingers and toes were remarkable. Her blood pressure was 112/74 mmHg in the right arm and 100/58 mmHg in the right leg. A grade 2/6 systolic murmur and 2/6 diastolic blowing murmur were heard at the pulmonary area. The lungs were clear on auscultation. No abnormal findings were found in the abdomen.

Her hemoglobin was 22.0 g/dl, erythrocytes $7.4 \times 10^6$/mm$^3$ and serum uric acid 13.4 mg/dl. Other routine hematological and biochemical examinations were within normal limits. Pulmonary function tests were normal. Analysis of the arterial blood while breathing room air showed a reduction of partial pressure of oxygen to 56 mmHg.

A chest x-ray revealed an enlarged cardiac shadow and marked dilatations of the proximal pulmonary arteries. Peripheral lung fields were clear (Fig. 1). An electrocardiogram showed right ventricular hypertrophy (Fig. 2).

A right heart catheterization revealed a right ventricular pressure of 110/2 mmHg, showing virtually identical systolic pressure to the arterial one. The oxygen saturation of blood samples taken from the superior vena cava, the right atrium, the right ventricle and the femoral artery was 67.5%, 68.1%, 70.0% and 79.6%, respective-

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Fig. 2. Electrocardiogram recorded at the first admission shows a classical pattern of right ventricular hypertrophy.

Fig. 3. Pulmonary arteriogram. The catheter was introduced to the pulmonary artery via the right femoral artery, the descending aorta and patent ductus arteriosus.
ly. On angiocardio graphic study, the contrast material injected into the right ventricle opacified the descending aorta via the pulmonary artery and the patent ductus arteriosus, but neither the aortic arch nor the carotid and subclavian arteries could be shown. By retrograde left heart catheterization from the left femoral artery, the catheter could not reach the aortic arch, but was introduced to the pulmonary artery from the descending aorta through the patent ductus arteriosus (Fig. 3). Moreover, a catheter was introduced to the ascending aorta from the right axillary artery and complete interruption of the aortic arch was clearly demonstrated by ascending aortography (Fig. 4).

Her hospital course was uneventful and the patient was discharged without consideration of surgical repair.

Second Admission
After discharge from our hospital, the patient was admitted 5 times to another hospital because of recurrent syncopal attacks. Five days before the second admission, the patient suffered from a sharp abdominal pain followed by chills and fever and was admitted to our hospital 10 years after her first admission at the age of 31.

The patient was markedly cyanotic. A diastolic thrill was palpable and a grade 4/6 harsh diastolic blowing murmur was heard along the left sternal border. The lungs were clear on auscultation. Although liver, kidney and spleen were not palpable, there was a tenderness in the left upper quadrant.

Her hemoglobin was 20.4 g/dl, erythrocyte count was $7.78 \times 10^6$/mm$^3$ and leukocyte count was $11,200$/mm$^3$. Chest x-ray films and an electrocardiogram revealed similar findings to those of the previous admission.

The abdominal pain continued and obvious signs of acute peritonitis appeared on 16th hospital day despite vigorous antibiotic treatment. A laparotomy was performed, but the patient died on 17th hospital day.

Autopsy Findings
The right ventricle was markedly hypertrophied with the free wall being 9 mm in thickness. There was a large ventricular septal defect, 1.5 x
TABLE I  SALIENT FEATURES OF REPORTED CASES OF INTERRUPTION OF THE AORTIC ARCH SURVIVED OVER THE AGE OF 15

<table>
<thead>
<tr>
<th>No.</th>
<th>Reporters</th>
<th>Age/Sex</th>
<th>VSD</th>
<th>PDA</th>
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<td>(-)</td>
<td>(-)</td>
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<tr>
<td>2</td>
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<td>Morgan 5 (1970)</td>
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<td>(-)</td>
<td>(-)</td>
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<tr>
<td>5</td>
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<td>16/F</td>
<td>(-)</td>
<td>A-P window</td>
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<td>6</td>
<td>Le Page 7 (1971)</td>
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<td>13</td>
<td>Judez 13 (1974)</td>
<td>18/F</td>
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<td>14</td>
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<td>Present case</td>
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F: female, M: male, VSD: ventricular septal defect, PDA: patent ductus arteriosus

2.5 cm in size. The main pulmonary arterial trunk and both proximal pulmonary arteries were markedly dilated and sclerotic. A large thrombus obliterated almost totally the proximal portion of the right pulmonary artery.

The aortic arch was completely interrupted just distal to the branching of the left subclavian artery (Fig. 5). Not even a rudimentary connection was demonstrated between the aortic arch and the descending aorta which was connected to the pulmonary artery by the large patent ductus arteriosus.

There were 4 relatively hard verrucae: 3 attached to the aortic valve and one to the pulmonary valve. Histological studies of the verruca attached to the pulmonary valve revealed a granuloma formation surrounding bacterial growth.

The lungs were congested. The pulmonary arterioles were narrowed by marked intimal proliferations. Two small abscess foci in liver, each 2 cm in diameter, left perirenal abscess and peritonitis were demonstrated. Histological studies of the kidneys revealed subacute diffuse glomerulonephritis.

**DISCUSSION**

This patient was diagnosed as having interruption of the aortic arch with patent ductus arteriosus at the age of 21. She died from bacterial endocarditis associated with acute peritonitis, perirenal abscess and subacute glomerulonephritis at the age of 31. At autopsy, complete interruption of the aortic arch, type A of Celoria's classification, associated with patent ductus arteriosus and a large ventricular septal defect was demonstrated.

The patients with interruption of the aortic arch rarely survive to the adult age. Only 19 cases, including the present case, were found to survive over the age of 15 in the Japanese and English literatures (Table I). Although this anomaly is known to be usually associated with patent ductus arteriosus and ventricular septal defect, this triad was present in only 7 of the above-mentioned 19 cases. Curiously, 6 of 8 Japanese cases who reached adult age had this triad. The relatively benign course in patients with solitary interruption of the aortic

arch, not associated with other congenital cardiovascular malformations, was pointed out by Dische and his coworkers. They considered that these patients might benefit from the formation of effective collateral circulation during fetal and early postnatal life. Lev thought that interruption of the aortic arch with and without patent ductus arteriosus represent two separate entities, while Perloff described that the clinical course was very different when not associated with supra- or intracardiac shunts.

Among the adult cases with the triad of interruption of the aortic arch, patent ductus arteriosus and ventricular septal defect, those reported by Takashina and his coworkers and Sunakawa and his coworkers were 32 years old and were still alive at the time of their report. Thus the present case is the third longest survivor as far as we know. The presence of a large ventricular septal defect and the increased pulmonary vascular resistance might be responsible for the longevity in the present case. Her large ventricular septal defect perhaps allowed a good mixing of the arterial and venous blood, while the increased pulmonary vascular resistance might restrict the left to right shunt, preventing excessive overload to the left ventricle.

REFERENCES


Japanese Circulation Journal Vol. 46, July 1982