Coarctation of the Aorta in Adult: Report of three Cases and Review of the Literature

Jung Ho Lee, M.D., Hoe Sung Yu*, M.D., Soo Woong Yoo, M.D., Hak Choong Lee, M.D.**

Coarctation of the aorta is an important congenital cardiovascular defect, which occurs in a significant number of persons.

The basic anatomic defect is a localized deformity of the media, manifested by two types of strictures in the aorta: "true" coarctation and tubular hypoplasia.

The zone of coarctation is characteristically located distal to the origin of left subclavian artery at or just beyond the insertion of the ligamentum arteriosum.

It shortens life if untreated, but it can be corrected to render the patient functionally normal.

Here we have a 2-year operative experiences with 3 Cases of the aortic coarctation, two or hypoplastic type and one of post ductal type, at age of 17 to 19 year old Korean, operated in 1965 and 1968 at the National Medical Center. The purposes of this report are to describe the immediate and late effect of surgery, histopathologic bases and the rarity of this lesions in Korea.

Abstract

Coarctation of the aorta is a congenital cardiovascular defect that affects a significant number of individuals.

The basic anatomic defect is a localized deformity of the media, manifested by two types of strictures in the aorta: "true" coarctation and tubular hypoplasia.

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Here we have a 2-year operative experience with 3 cases of aortic coarctation, two of hypoplastic type and one of post ductal type, at age of 17 to 19 year old Korean, operated in 1965 and 1968 at the National Medical Center. The purposes of this report are to describe the immediate and late effect of surgery, histopathologic bases and the rarity of this lesion in Korea.
the N.M.C. on May 3, 1965 with an one year history of mild chest discomfort, dyspnea on exertion, progressive visual disturbances and frequent episodes of nasal bleeding. However, 4 years prior to this admission to the N.M.C. he suffered a massive nasal bleeding without any known causes, and was informed that he had a congenital heart disease. Otherwise his past history and the family history were not contributory.

Physical examination revealed a normally developed and well nourished patient with a relatively prominent development of the trunk compared to that of the lower extremities (Height 176cm Weight 63.3kg). The blood pressure was significantly higher in the upper (230/120 mmHg) than in the lower (120mmHg systolic by palpation) extremities. The pulse was more powerful in both upper arms and diminished and delayed in the lower, rate around 72/min. A systolic thrill was noted on palpation along the left sternal border. On percussion, the heart dullness was wider than usual, but no abnormal pulmonary alteration was found. A systolic murmur Gr. Ⅱ/Ⅲ was audible over the entire precordium, with the maximum point at the 3 ICS on LSB.

The pulsation from possible collateral blood vessels was noted on the chest wall, shoulder, neck and in the interscapular region. No hepatosplenomegaly or other abdominal organomegaly was demonstrable but a faint systolic murmur on the paraumbilical region was audible.

The ECG showed left ventricular hypertrophy (Fig.1) and episodes of arrhythmia with some nodal escape beats. Serial chest and heart X-ray film showed cardiomegaly especially a left ventricular enlargement, multiple rib notchings in both sides and an indented left upper mediastinal shadow as the "3 sign", 6505251 Im K.

caused by the dilated proximal segments and the left subclavian artery. On aortography, a marked coarctation of the aorta was found just distal to the origin of the left subclavian artery and the extensive collateral circulation was observed through the intercostal arteries and especially both

Fig. 1. May 12, 1965 Interpretation: Left Ventricular Hypertrophy Op. diagnosis: Coarctation of Aorta.

Fig. 2a. Case I. Im K. A picture of plain chest X-Ray film showing left ventricular enlargement and indentation at the beginning of the descending aorta, "3" sign. The arrow point to classical notches in the ribs.

amarian arteries which were widened and tortuous. The proximal part of subclavian artery was markedly dilated.
Fig. 2b. Case 1 Im K. Early stage of aortography reveals abrupt narrowing and complete stenosis of aorta at junction of arch and descending aorta. From the aorta above the stenosis a big internal mammary mammary artery is visualized along the course of the vertebral column, with non-visualization of descending aorta below the coarctation.

Fig. 3a. 6505816 Kim, Y. M. Jan. 14, 1965 Interpretation: Supraventricular Tachycardia Nov. 25, 1966 Interpretation: Left Ventricular Hypertrophy Op. diagnosis: Coarctation of Aorta

On May 19, 1965, the involved part of the aorta was resected through the posterior-lateral thoracotomy. Following the aortic resection, a transplant of a "Teflon graft" 10cm long was made and an appropriate anastomosis performed. (Table 5.) The histopathology of the resected aorta revealed organized thrombus formation in the arterial wall in which hemorrhage and recanalization was observed. The adventitial tissue showed marked hemorrhage but no signs of inflammatory or
Table 1. Pre- and Post-operative Changes in Blood Pressure (mmHg) Above and Below the Coarctation of Aorta

<table>
<thead>
<tr>
<th>Extremities</th>
<th>Before Operation</th>
<th>After Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Syst.</td>
<td>Diast.</td>
</tr>
<tr>
<td>Rt. Uppe</td>
<td>230</td>
<td>120</td>
</tr>
<tr>
<td>Lt. Upper</td>
<td>230</td>
<td>120</td>
</tr>
<tr>
<td>Rt. Lower</td>
<td>120</td>
<td>—</td>
</tr>
<tr>
<td>Lt. Lower</td>
<td>120</td>
<td>—</td>
</tr>
<tr>
<td>Difference</td>
<td>110</td>
<td>—</td>
</tr>
</tbody>
</table>

Fig 3b. 6505816 Kim, Y.M. Jan. 12, 1967 Interpretation: Left Ventricular Hypertrophy with strain
Op. diagnosis: Coarctation of Aorta
sclerotic changes of arterial of arterial wall were noted. The postoperative course involved complications in that evidence of nasal bleeding, pneumothorax and pleural fluid in the left hemithorax remained temporarily. The hypertension noted

Fig 4. Case J. Kim Y.M. Late phase of aortography shows vimalization of the descending aorta through collateral arteries. The arrow points to the stenotic part of aorta.

Fig 5a Im. K. 6505251 J
Fig. 5a  Im. K. 6505251

Fig. 5b. Kim. M.M. 6505816. Stenotic area:

Fig. 5b. Kim. Y.H. 6050816. Sudan Stain.

were in good and strong. He has no subjective difficulties in ordinary life except for a slight tingling sensation over both arms.

Case 1. Kim Y.M. Male 6505816

On March 29, 1965, a 17-year-old Korean male was referred to the N.M.C. from the Galvary Clinic for the investigation and elucidation of the cause of his hypertension with the possibility of surgical correction. Actually, he had been suffering from palpitation, severe headache, dizziness and general malaise for the last 4 years, when he was diagnosed for the first time as having a hypertension.

The initial onset of symptoms occurred at the age of 10 when he suffered from headache and palpitation after physical exertion. The past and the family histories were non-contributory. Upon admission, May 12, 1965, physical examination revealed the patient to be well nourished and of normal general development (Height 167 cm, Weight 54.5kg).

The blood pressure was remarkably high in both upper extremities (Rt. 190/130 mmHg, Lt. 180/110mmHg) and showed a definite pressure gradient externally compared to the lower extremities (Rt. 150/120mmHg, Lt. 140/120mmHg). The pulse was regular and around 80/min at rest. It was more powerful in both upper extremities, while weak and delayed in the lower extremities. The abnormal pulsation was demonstrable on the anterior chest wall around the sternum, below the clavicular region and on the back. The heart was enlarged with increased cardiac dullness. A grade I/II systolic murmur was audible on the cardiac apex and 3 ICS on LSB. The ictus was on the 6th intercostal space to the left midclavicular line without thrill. Normal findings were obtained upon fundus examination. Abdominal examination showed no demonstrable organomegaly and neurological findings were within normal limits. ECG showed left ventricular hypertrophy. Serial chest and heart x-ray films disclosed left ventricular enlargement with prominent ascending aorta and multiple rib
Table 2 ***Case I, Kim, Y.M.*** Pre-and Post- operative Changes in Blood Pressure(mmHg) Above and Below the Coarctation of Aorta

<table>
<thead>
<tr>
<th>Extremities</th>
<th>Syst.</th>
<th>Diast.</th>
<th>Pulse Pressure</th>
<th>2 months</th>
<th>Syst.</th>
<th>Diast.</th>
<th>Pulse Pressure</th>
<th>2 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rt. Upper</td>
<td>190</td>
<td>130</td>
<td>160</td>
<td>140</td>
<td>110</td>
<td>30</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lt. Upper</td>
<td>180</td>
<td>110</td>
<td>70</td>
<td>140</td>
<td>100</td>
<td>40</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rt. Lower</td>
<td>150</td>
<td>120</td>
<td>30</td>
<td>150</td>
<td>120</td>
<td>30</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lt. Lower</td>
<td>140</td>
<td>120</td>
<td>20</td>
<td>150</td>
<td>110</td>
<td>40</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Difference</td>
<td>40</td>
<td>-10</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

notchings.

The typical indentation of the left upper mediastinal shadow was present. On aortography, the characteristic narrowing of the aorta was found just distal to the origin of the left subclavian artery. Enlarged internal mammary arteries, large tortuous intercostal arteries and other collateral circulation were demonstrable. On June 14, 1965 the narrowing of the aorta was resected through a left posterior-lateral thoracotomy and an end-to-end anastomosis was made without graft. (Table 5.)

The histopathology of the resected aorta showed severe intimal thickening composed of fibrous tissue with ingrowth of blood vessels in most of the stenotic area. The elastic tissue in this part of the artery was somewhat irregular. (fig 5b)

The postoperative course was quite smooth, without complications and the preoperative high blood pressure in the upper extremities declined to normal; a relatively increased blood pressure in the lower extremities now being present. (Table 2.)

On Jan. 11, 1967 this patient was readmitted because of exertional dyspnea and numbness on both the hands and feet and for aortography. The blood pressure showed no pressure difference between upper and lower extremities (Lt. upper 140/100 mmHg, Lt. lower 140/100 mmHg) and the femoral pulse was good in both. The heart showed an audible systolic murmur Gr. II on apex and Lt. sternal border. Compared to the previous ECG trace, the present trace showed no interval changes with left ventricular strain. (fig 3b) On the 2nd aortography, there were some narrowing “wasting” of the aorta but the caliber was still relatively good. But there was some dilation for the distally in the descending aorta.

Case II Park Y.H. Female 6804814

This 17-year-old female was admitted to the N.M.C. on March 25, 1968, because of palpitation, exertional dyspnea cough with recurrent U.I.R. and nasal bleeding which had become worsened for 2 years. Cardiomegaly discovered at her age of 3 during an episodes of common cold while at productive cough, nasal bleeding and dyspnea occurred.

Physical examination revealed emaciated and retarded physical development with height 158cm and weight 38kg. The blood pressure was high and strong in thearms (220/90mmHg) and faint to absent in the legs (120/80 mmHg). Heart examination disclosed pansystolic murmur Gr. II on the entire precordium and continuous murmur audible at the left 2nd and 3rd intercostal spaces and thrill palpable. Liver was enlarged to 3FB along the right costal margin and spleen tip palpable without ascites.

ECG showed suggestive biventricular hypertrophy with left ventricular preponderance (fig 6). Chest and heart x-ray film (86319) showed pancardio-megaly and increased vascularity of lung suggesting left to right shunt. Aortogram demonstrated patent ductus with possible coarctation of aorta and renal angiogram showed a little smaller left kidney

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without vascular abnormalities and splenomegaly also noted. Cardiac catheterization revealed markedly elevated main pul. artery (109.5/74.5 (Mean 87) mm Hg) and no pressure gradient across the pul. valve. (R.V. 109.5/0/5 mmHg)

There was a marked oxygen step-up in main pul. artery (11.5 Vol%) over the right ventricle (8.4 Vol%). Cardiac output was 5.35 L/min and pul. blood flow 10.4 L/min. Calculated Lt to Rt. shunt was 48.6% of total pul. blood flow (table 3.)

Aortogram showed patent ductus arteriosus with possible coarctation of aorta and no distinct collaterals.

**Table 3. Cardiac Catheterization Report**

<table>
<thead>
<tr>
<th>Cath. Position</th>
<th>$O_2$ Sat. (vol %)</th>
<th>Pressure mmHg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main pul. Artery</td>
<td>11.52%</td>
<td>109.5/74.5 (Mean 87.0)</td>
</tr>
<tr>
<td>Lt. pul. Artery</td>
<td>11.53%</td>
<td></td>
</tr>
<tr>
<td>Right ventricle, Mid.</td>
<td>8.40</td>
<td>109.5/0/5.33</td>
</tr>
<tr>
<td>Right Atrium high Mid.</td>
<td>8.86</td>
<td>23.1/2.67(M10.66)</td>
</tr>
<tr>
<td>Brachial Artery</td>
<td>14.89</td>
<td>240.0/83.5(M 128.0)</td>
</tr>
<tr>
<td>Arterial $O_2$ Sat.</td>
<td>95. 5%</td>
<td></td>
</tr>
<tr>
<td>Cardiac Output</td>
<td>5.35 L/min</td>
<td></td>
</tr>
<tr>
<td>Pul. Blood Flow</td>
<td>10.40 L/min</td>
<td></td>
</tr>
<tr>
<td>Lt. to Rt. Shunt</td>
<td>48. 6% of total pul. blood flow</td>
<td></td>
</tr>
</tbody>
</table>

Under the diagnosis of PDA or in combination with coarctation of aorta, on July 19, 1968 left standard thoracotomy was made.

No remarkable collateral vessels noted and just beyond the origin of subclavian artery marked coarctation of aorta was disclosed. Proximal part of aorta measured 2.5cm in diameter and distal part 4.5cm in diameter as poststenotic dilatation. Lt. subclavian artery was also dilated to 1.5cm in diameter. The PDA was measured 1.5cm in length and 1cm in diameter. To rule out other possible cardiac anomalies pericardium was opened and

**Fig 6**. 6804814 Park, Y.H. F Mar. 26, 1968

Intretation: Left Ventricular Hypertrophy with strain

Clinical diagnosis: PDA
Table 4. *Case II, Park, Y.H*** Pre- and Post-operative Changes in Blood Pressure (mmHg)
Above and Below the Coarctation of Aorta

<table>
<thead>
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<tr>
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<td>Diast.</td>
</tr>
<tr>
<td></td>
<td>Pressure</td>
<td></td>
</tr>
<tr>
<td>Rt. Upper</td>
<td>200</td>
<td>80</td>
</tr>
<tr>
<td>Lt. Upper</td>
<td>220</td>
<td>90</td>
</tr>
<tr>
<td>Rt. Lower</td>
<td>120</td>
<td>80</td>
</tr>
<tr>
<td>Lt. Lower</td>
<td>110</td>
<td></td>
</tr>
<tr>
<td>Difference (M)</td>
<td>95</td>
<td></td>
</tr>
</tbody>
</table>

Table 5. Sketches of Pathoanatomical Situation in 3 cases of Coarctation of Aorta

Case I: "Post-ligamental" The dotted line is shown the Teflon graft applied after resection of involved aorta.

Case II: "Post-ligamental" The dotted line is shown a part of resected area of aorta and end to end anastomosis was possible without application of graft.

Case III: "Juxta-ductal" type The dotted line is shown a part of resected area of aorta and end to end anastomosis was possible without application of graft ligation of PDA.

The heart was checked to be anatomically normal. The PDA was ligated Pott's vascular clamps were applied proximal and distal parts of coarctation. During this procedure, blood pressure was elevated upto 280 mmHg in systole which was controlled with Alfonade infusion down to 220 mmHg.

The coarctation of aorta was resected in 3cm length combined with aortic part of PDA and end-to-end anastomosis of aorta was made (table 5.) Also collaterals to distal part of aorta was not so remarkable. After operation, blood pressure was 180/110 mmHg on the upper extremities and 130/110 mmHg on lower extremities.(table 4) Resected aorta for histopathology disclosed a detachment of endothelial lining cells and fragmentation of elastic fibers in association with collagenization of media (fig 5c).

The postoperative course was uneventful with good femoral pulsations but residual pressure gradient between upper and lower extremities.
考案

大動脈을 따르며 그 내경의 침소(tubular hypoplasia) 또는 속쓰어의 침소(constriction)가 가해진 것으로 대동맥의 침소
양성 및 그조직의 생지학적 과학으로 생겨, Turner과 침소형 증가자에서 특히 최고 생기는 하나, 일반적으로 여자
보다는 남자에서 더 많다(M: F = 2:1) Abott(1928), Reifenstein, Levine과 Gross(1947)는 대동맥의 침소
환자들의 내경의 60~70%가 40세 이전에 발생한다고 하였다. 32세 이전에 50%가 발생하며 평균생명은 34세였다 한다.

5.2.3.1. 靜脈閉塞는 속쓰어의 침소 및 동맥의 혈류 제한으로 인해 형성되는 작은 양의 혈관이 생성된다. 
2.1.3.1. 患者의 1/3에서 볼 수 있는 다른 형성학적 병변과 함께 발생한다. 혈액을 공급하는 혈관은 Bicuspid aortic valve, PDA, small VSD, Congenital aortic stenosis, Intracranial A-V malformation 등이다. 

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