Interruption of the Aortic Arch Without a Patent Ductus Arteriosus


SUMMARY
Complete interruption of the aortic arch without a patent ductus arteriosus is compatible with survival for many years. This is demonstrated by the case reported herein and two others reported in the literature. With no connection between the ascending and the descending aorta and with both subclavian arteries coming off the descending aorta, blood supply to the entire body, except for the head, was by retrograde flow through the vertebral arteries and left cervical collaterals. Bounding carotid pulsations with diminished arm and leg pulses are apparent in this anomaly. Surgical restoration of the continuity between the ascending and the descending aorta was successful in this case and the two previously reported cases. The surgical results of all cases of interruption or atresia of the aortic arch, including those with a patent ductus arteriosus, are reviewed, and the appearance of the ascending aorta on angiography is suggested as a possible differential point between interruption and atresia of the aortic arch.

Additional Indexing Words:
Atresia of aortic arch  Aberrant right subclavian artery  Cardiac catheterization
Subclavian steal syndrome  Angiocardiography  Anatomic findings
Embryology  Prosthetic grafts

Prior to 1964, interruption of the aortic arch as an isolated anomaly was thought to be incompatible with life. In that year Pillsbury and associates reported the first case without an associated patent ductus arteriosus. The second case was described by Zetterqvist in 1967. The purpose of the present paper is to describe the third case of interruption of the aortic arch without a patent ductus arteriosus and to review the surgical results with this anomaly.

Report of Case
C. H., a 19-year-old, white male, active duty sailor, was referred to the Naval Hospital, San Diego, for evaluation of a possible carotid aneurysm. He had been aware of prominent bilateral neck pulsations all of his life. He was entirely asymptomatic and could exercise strenuously without leg or arm claudication, headaches, or symptoms of cerebrovascular insufficiency. The family history was negative.

Physical examination revealed a well-developed, muscular male with no cyanosis or clubbing. Cuff blood pressure in both arms and legs was 95/70 mm Hg. Both carotid pulses were bounding, whereas both radial, brachial, and femoral pulses could not be felt. No precordial lift or thrill was palpable. The first and second heart sounds were normal, and no extra sounds were audible. A grade II/VI, systolic ejection murmur was present in the aortic area; this murmur became less intense along the left sternal border.
Figure 1

Preoperative thoracic roentgenogram. Absence of an aortic knob; no descending aorta or rib notching is visible.

and apex. Prominent systolic carotid bruits were present: grade III/VI on the right, grade II/VI on the left. The most prominent bruit was heard over the left posterior occipital area, where it was grade IV/VI behind the left ear and grade II/VI behind the right ear. A grade II/VI systolic bruit was heard over the upper thoracic spine, where it spilled briefly past the second heart sound. The remainder of the examination was negative.

The electrocardiogram was normal.

A thoracic roentgenogram (fig. 1) revealed a normal-sized heart with no visible aortic knob, descending aorta, or rib notching. A barium swallow showed, in the lateral projection, a posterior indentation of the esophagus compatible with an aberrant right subclavian artery.

The clinical diagnosis was coarctation of the aorta involving the origin of the left subclavian artery with an aberrant right subclavian artery, with or without a cervical aorta or other anomaly of the aortic arch.

Catheterization was attempted from the right groin, plus the right and left brachial arteries. The right heart pressures were normal, that is, pressure in the right atrium was 6 mm Hg mean, pressure in the right ventricle was 25/6 mm Hg, and pulmonary artery pressure was 25/15 mm Hg. Normal oxygen saturation data, indocyanine-green dye curves, and a hydrogen inhalation study excluded shunts. A Brockenbrough transseptal catheter accidentally entered the ascending aorta instead of the left atrium; withdrawal of the catheter with surgical standby was uneventful. A damped pressure in the ascending aorta was 130/80 mm Hg. Pressure in the descending aorta, femoral artery, and right subclavian artery was 95/70 mm Hg. The catheter from the femoral artery could not be passed around the aortic arch; it always went into the left side of the neck.

Figure 2

Pulmonary artery angiograms. (A) Posteroanterior view 4 sec after injection. Large bilateral common carotid arteries seen, without an aortic arch or descending aorta. Collateral vessels just starting to fill in left neck region. (B) Oblique view 4 sec after injection. Descending aorta is faintly visualized, but no definite connection is seen with ascending aorta. (C) Later film. Demonstrates many large collateral vessels in the left neck region.
Injection of contrast material showed the direction of blood flow to be from the neck vessels to the descending aorta. Catheters from both the right brachial and left brachial arteries would only go down the descending aorta. A pulmonary artery angiogram, made by using a single plane Sanchez-Perez film changer, demonstrated huge right and left common carotid arteries arising from the ascending aorta (fig. 2A and B), but no definite connection was seen between the ascending and descending aorta. Both subclavian arteries and the descending aorta filled late, by retrograde flow through the vertebrals, after a large number of collateral vessels were seen in the left neck region (fig. 2C).

Surgical Procedure. A left anterolateral thoracotomy with extension across the sternum was performed. There was complete anatomic separation of the arch of the aorta between the left common carotid and left subclavian artery. The descending aorta arose at the junction of the aberrant right subclavian and left subclavian arteries and was physically separated from the ascending aorta by about 10 cm. A prominent ligamentum arteriosum was present between the pulmonary artery and the descending aorta at the left of the left subclavian artery. The left recurrent laryngeal nerve looped around the ligamentum arteriosum in normal fashion. The descending aorta was thinned and considerably more friable than normal. A 10-mm knitted Dacron bypass graft was inserted between the ascending and descending aortas, utilizing partial occluding clamps. Cardiopulmonary bypass was unnecessary. Initially the graft was inserted anterior to the phrenic nerve, but kinking required revision with transfer of the graft to a position posterior to the phrenic nerve. The postoperative course was uneventful. Foot pulses were noted immediately after surgery.

Two weeks after surgery, the left arm blood pressure was 115/90 mm Hg. Results of cardiac examination were unchanged, except that the systolic carotid and cranial bruits had decreased in intensity. Strong left radial and bilateral dorsalis pedis pulses were present, but the right radial and both femoral pulses continued to be damped. The postoperative thoracic roentgenogram now showed a prominent aortic knob.

Discussion

In 95% of the reported cases of interruption of the aortic arch, a ventricular septal defect has been present.4,5 Except for the cases reported by Zetterqvist,3 and Pillsbury and associates2 and the present case, a patent ductus arteriosus has always been present. In atresia of the aortic arch, which is included in

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**Figure 3**

Schematic drawings of the three cases without patent ductus arteriosus. (A) Present case: Interruption beyond the left common carotid artery with an aberrant right subclavian artery. (B) Case of Zetterqvist (1967)3: Similar to the present case, except that the right common carotid artery is absent. (C) Case of Pillsbury and associates (1964)2: Interruption between the right and left common carotid arteries.

Abbreviations: ALSA and ARSA = anomalous left and right subclavian arteries; LVA and RVA = left and right vertebral arteries; LAA and RAA = left and right axillary arteries; LCCA and RCCA = left and right common carotid arteries; LSA and RSA = left and right subclavian arteries; ASC.AO = ascending aorta; DESC.AO = descending aorta.
most series with interruption of the aortic arch, a patent ductus arteriosus has always been present, except in the case described by Evans at necropsy.

In the present case of a 19-year-old youth, there was interruption of the aortic arch after the left common carotid artery and an aberrant right subclavian artery was present (fig. 3A). There was also a well-developed collateral system in the left neck region.

Zetterqvist described an 8-year-old boy with interruption of the aortic arch beyond the left common carotid artery (fig. 3B). The right common carotid artery was absent, but an aberrant right subclavian artery was present. No remarkable collateral system was visualized in the neck or thorax. The ascending and descending aortas were approximated by side-to-end anastomosis without need of a prosthetic graft. This boy was doing well 1 year postoperatively.

In the first published case by Pillsbury and associates, a 16-year-old girl had interruption of the aortic arch between the right and left carotid arteries (fig. 3C). Massive collaterals were present in the right side of the neck and chest with associated rib notching, but no collaterals were present in the left chest. There was a normal right subclavian artery; therefore, this case was different from the other two, in that an entry was present from the right brachial artery into the aorta and left ventricle. A Dacron graft was inserted between the ascending and the descending aorta. The patient is now 22 years old, is married, and recently had a baby (Shumway NE: Personal communication to authors).

In two of the three cases of interruption of the aortic arch without a patent ductus arteriosus an aberrant right subclavian artery has been present with weak pulses in both arms and legs and with at least one bounding carotid pulse. This combination of findings should suggest the possibility of interruption of the aortic arch.

The embryologic background of the present case is presumed to be bilateral regression of the fourth aortic arches with bilateral persistence of the dorsal aortas. The right fourth aortic arch normally develops into the proximal part of the right subclavian artery; regression is responsible for the aberrant right subclavian artery. The left fourth aortic arch forms the segment of the aortic arch between the left common carotid and the left subclavian artery; regression is responsible for the interruption of the aortic arch following the left common carotid artery.

Most reports in the literature combine cases of interruption of the aortic arch (complete absence of aortic arch) with cases of atresia of the aortic arch (connection of the ascending aorta and descending aorta by an atretic segment resembling a fibrous band). Although surgical treatment at the present time is essentially the same in both conditions,

Table 1
Review of Reported Cases of Interrupted or Atretic Aortic Arch

<table>
<thead>
<tr>
<th>Author</th>
<th>No. of cases</th>
<th>Surgery</th>
<th>Survival surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>T</td>
</tr>
<tr>
<td>Previous cases</td>
<td>111</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Tawes, et al.7</td>
<td>21</td>
<td>-</td>
<td>9*</td>
</tr>
<tr>
<td>McNamara &amp; Rosenberg6</td>
<td>17</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Rochette et al.9</td>
<td>7</td>
<td>5</td>
<td>-</td>
</tr>
<tr>
<td>Rainer10</td>
<td>1</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>Zetterqvist3</td>
<td>1</td>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>Pilapil et al.11</td>
<td>1</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Present case</td>
<td>1</td>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>Total</td>
<td>160</td>
<td>15</td>
<td>14</td>
</tr>
</tbody>
</table>

*Two patients died before definitive procedure was performed.
†Two late deaths.
Abbreviations: I = interrupted arch; A = atretic arch.
they probably should be differentiated for reasons of classification. Differentiation may be difficult except by surgical exploration or necropsy. The complete lack of an aortic knob on the chest roentgenogram favors interruption of the aortic arch. When a patent ductus arteriosus is present, right heart angiograms will reveal a similar pulmonary artery-to-ductus-to-descending aorta appearance in the usual case of either atresia or interruption. However, angiography of the ascending aorta in atresia of the aortic arch may give some indication of the beginning curve of an aortic arch. In cases of complete interruption of the aortic arch, there should be no indication at all of the beginning curve of an aortic arch; the direction of the ascending aorta is straight superiorly.

A total of 160 cases of interruption and atresia of the aortic arch (including those with a patent ductus arteriosus) have now been reported, with over half of these cases having complete interruption of the aortic arch (table 1). Most of these cases have been diagnosed at necropsy, with only 24 cases of interruption of the aortic arch and 12 cases of atresia of the aortic arch diagnosed during life. Surgery has been attempted on a total of 15 patients with interruption of the aortic arch, with six patients surviving surgery; 14 patients with atresia of the aortic arch have been operated with nine patients surviving surgery. Of the total of 15 survivors of surgery for both interruption and atresia, four have been 8 years of age or older (the three cases of interruption of the aortic arch without a patent ductus arteriosus and the one case of atresia of the aortic arch with a patent ductus arteriosus). The operative risk in the younger age group in either interruption or atresia of the aortic arch is still extremely high.

References


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JACOB R. MORGAN, CDR, ALAN D. FORKER, LCDR, RICHARD G. FOSBURG, CAPT, M. KEN NEUGEBAUER, CDR, ALBERT K. ROGERS, CDR and CARL R. BEMILLER, CDR

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