fibrous subpulmonic stenosis) may require a Rastelli\textsuperscript{11} procedure and corrective surgery therefore is recommended at a later age.\textsuperscript{12} For patients with TGA and subpulmonic obstruction due to AMS the Mustard type of repair can be done early in life.

References

RADIOLOGY

Complete Interruption of the Aortic Arch

2. Characteristic Angiographic Features with Emphasis on Collateral Circulation to the Descending Aorta

RICHARD B. JAFFE, M.D.

SUMMARY The angiograms of 17 patients with aortic arch interruption are reviewed to emphasize the variations in arch interruption and origin of the brachiocephalic vessels, and collateral circulation to the descending aorta. Depending on the anatomical type and subtype of arch interruption, collateral flow to the descending aorta in the presence of a stenotic or closed ductus will be dependent on the development of intercostal collaterals and/or the presence of retrograde flow in all brachiocephalic vessels arising from the descending aorta. Familiarity with the potential pathways for collateral circulation may permit differentiation into types and subtypes on chest radiograph. Patients with Type I interruption may have bilateral rib notching if the right subclavian artery originates normally from the innominate artery, but will have rib notching confined to the left side if the origin of the right subclavian artery is aberrant. Type II or Type III interruption patients will have rib notching confined to the right side if the right subclavian has a normal origin, but no rib notching if the origin of the right subclavian artery is aberrant.

\(\square\) COMPLETE INTERRUPTION of the aortic arch is characterized by discontinuity of the arch between the proximal ascending aorta and the distal descending thoracic aorta. The pathologic anatomy of the three different types and three subtypes of aortic interruption, and characteristic radiographic findings in 21 patients have been discussed in Part 1.\textsuperscript{1} This section will review the angiograms of 17 of these patients and others reported in the literature to enhance the recognition of the variations in arch interruption and origin of the brachiocephalic vessels, and collateral circulation to the descending thoracic aorta.

The site of arch interruption and origin of the brachiocephalic vessels are best delineated in patients without transposition following left atrial,\textsuperscript{2} left ventricular, or ascending aortic angiography. The ascending aorta is characteristically hypoplastic and ascends almost directly vertically into the neck in both frontal and lateral projections (fig. 1A, B). As will be discussed, the origin of the brachiocephalic vessels in relation to the site of arch interruption determines the potential for collateral circulation to the descending thoracic aorta.

The descending thoracic aorta originates from the pulmonary artery through a patent ductus arteriosus. Evans\textsuperscript{8} has introduced the term “pulmonary-ductus-descending aorta trunk” to include the pulmonary artery, patent ductus arteriosus, and descending thoracic aorta. In patients with Type II interruption, the left subclavian artery, and in Type III interruption, the left subclavian and left common carotid arteries arise from this trunk. If the right subclavian artery has an aberrant origin it also may be from this location. Typically the main pulmonary artery is moderately to
markedly enlarged and continuous with a large patent ductus arteriosus approximately the same size as the proximal descending thoracic aorta (fig. 1C, D). Occasionally, coarctation of the proximal descending thoracic aorta is present.\(^8\) \(^4\) Except for two patients,\(^6\) the descending thoracic aorta always arose from a left ductus arteriosus.

Occasionally, the ductus arteriosus is narrowed\(^6\) -\(^14\) or closed,\(^4\) \(^7\) \(^15\) -\(^20\) and in these patients collateral circulation must develop to the descending thoracic aorta, either through the brachiocephalic vessels arising from the proximal descending thoracic aorta and/or the intercostal arteries. The origin and direction of blood flow in the brachiocephalic vessels then becomes the determining factor in the radiographic and angiographic findings. Review of previously reported patients with aortic arch interruption and a stenotic or closed ductus arteriosus indicates that retrograde flow will be seen angiographically in all the brachiocephalic vessels arising from the proximal descending thoracic aorta. Of particular interest is that while collateral circulation through the intercostal arteries also may be seen to the descending thoracic aorta, it occurs only if antegrade flow is present in the proximal subclavian artery on that side.

In Type I interruption, the ascending aorta is discontinuous distal to the origin of the left subclavian artery. Most commonly each brachiocephalic vessel arises from the hypoplastic ascending aorta. The absence of the transverse arch distal to the left subclavian artery, and the close approximation of this vessel to the left common carotid artery often results in the angiographic appearance of both right and left innominate arteries which may appear as the letter "Y" on frontal angiograms (fig. 1A).

In the most common pattern of Type I interruption, with all brachiocephalic vessels arising normally from the ascending aorta (Subtype I-A), collateral circulation to the descending aorta in the presence of a stenotic or closed ductus is dependent on branches of the right and left subclavian arteries, and in particular the intercostal arteries. As expected with antegrade flow in both right and left subclavian arteries, these intercostal collaterals develop on both right and left sides as illustrated by the patient of Ruiz Villalobos et al.,\(^14\) with bilateral rib notching on chest radiographs and bilaterally dilated intercostal arteries at operation, and angiographically by Castellanos et al.,\(^7\) Sissman,\(^20\) and Tyson et al.\(^13\)

In three reported patients\(^17\) -\(^21\) and one patient in this series with Type I interruption, the right subclavian artery had an aberrant origin from the descending thoracic aorta (Subtype I-B). In these patients, the right subclavian artery is potentially available as a large collateral channel to the descending aorta. Collateral circulation to the descending aorta can occur in these patients from a right vertebral-subclavian "steal" with normal antegrade flow in the right and left common carotid and left vertebral arteries and retrograde flow down the right vertebral artery into the proximal right subclavian artery, and then into the descending thoracic aorta. The presence of a right vertebral-subclavian "steal" may be seen angiographically in these patients following injection of contrast media into the main pulmonary artery, ductus arteriosus, or proximal descending thoracic aorta.\(^12\)

When the ductus becomes stenotic or closed, collateral circulation to the descending thoracic aorta will be dependent in part on the aberrant right subclavian artery. The anatomy is such that retrograde flow will be present in the proximal right subclavian artery (right vertebral-subclavian "steal"), and although the internal mammary and lateral thoracic branches of the right subclavian and axillary artery conceivably could provide collateral circulation to the descending aorta through the right intercostal arteries, the retrograde flow in the proximal right subclavian artery most likely will prevent the development of intercostal collaterals on that side. Thus, in patients with Type I interruption, aberrant origin of the right subclavian artery, and a stenotic or closed ductus, collateral circulation through the intercostal arteries may occur only on the left side where normal antegrade flow is present. If the patient develops to the age where intercostal collaterals result in rib notching on chest radiograph, this notching may be expected to be present only on the left side. Angiographically, retrograde flow will be present in the proximal right subclavian artery, while additional collateral flow through the intercostal arteries, if present, will be seen only on the left side. Such patients will be similar to those reported with aortic coarctation and an aberrant right subclavian artery arising distal to the coarctation where rib notching and collateral intercostal circulation are seen only on the left side, and retrograde flow is observed in the aberrant right subclavian artery.\(^23\)

In Type II interruption, the aortic arch is discontinuous distal to the origin of the left common carotid artery. The left subclavian artery arises from the descending aorta near the continuance of the ductus arteriosus into the descending thoracic aorta, and is available as a potential pathway for collateral circulation to the descending aorta (left vertebral-subclavian "steal").

Retrograde flow in the left subclavian artery has been recognized angiographically in several patients with Type II interruption, normal origin of the right subclavian artery (Subtype II-A), and wide patency of the ductus arteriosus,\(^12\) \(^24\) and was seen in two patients in this series (fig. 2). Occasionally antegrade flow may be present in the left subclavian artery if the ductus arteriosus is widely patent and equilibration of ventricular pressures can occur through the ventricular septal defect. Antegrade flow was demonstrated angiographically in the left subclavian artery in one patient in this series, and has been reported in three other patients with Type II interruption and normal origin of the right subclavian artery.\(^25\) \(^26\) If the ductus arteriosus becomes stenotic or closed in patients with Type II interruption and normal origin of the right subclavian artery, collateral circulation to the descending aorta will be dependent on retrograde flow in the left subclavian artery arising from the proximal descending thoracic aorta, and the development of right intercostal collaterals.\(^15\)

Two patients in this series with Type II interruption, normal origin of the right subclavian artery, and a stenotic ductus demonstrated similar radiographic findings to that noted in a 12-year-old patient with similar anatomy reported by Asano et al.\(^15\) (fig. 3). Both had rib notching on chest radiograph confined to the right side, and demonstrated retrograde blood flow angiographically in the left sub-
clavian artery. Antegrade flow was demonstrated in the normally arising right subclavian artery in these three patients, and the intercostal collaterals to the descending aorta were confined in angiography to the right side as expected.

It is unlikely that collateral circulation through the left intercostal arteries will be seen in patients with Type II interruption and a stenotic or closed ductus arteriosus, as retrograde flow invariably will be present in the proximal left subclavian artery into the descending thoracic aorta. This probably will prevent the development of the intercostal collaterals on that side. However, a unique case of aortic interruption, Type II-A, was reported by Kauff et al. with bilateral rib notching. Origin of the left vertebral artery from the left common carotid artery arising from the ascending aorta was present in this patient (personal communication), and this anatomy prevented a left vertebral-subclavian "steal" into the descending aorta. Instead, antegrade flow was present in the proximal left subclavian artery as it arose from the descending thoracic aorta and resulted in the development of left-sided intercostal
collateral vessels.

In Type II interruption, the right subclavian artery most commonly arises normally from the innominate artery (Subtype II-A), but frequently it arises aberrantly from the descending aorta just distal to the left subclavian artery (Subtype II-B).2, 4, 6, 8, 12, 16, 18, 20-35 In these patients an additional vessel is potentially available for collateral circulation to the descending aorta (left and right vertebral-subclavian “steal”). Antegrade flow may be seen angiographically in both the left and aberrant right subclavian arteries if the ductus arteriosus is widely patent and equilibration of ventricular pressures can occur through the ventricular septal defect.5, 8, 20, 24 Interestingly, one infant in this series with Type II-B interruption and a widely patent ductus had retrograde flow angiographically in both the left subclavian and aberrant right subclavian arteries (fig. 4).

Gokcebay et al.,16 Morgan et al.,18 and Zetterqvist,4 have each reported a patient with Type II interruption, aberrant origin of the right subclavian artery from the proximal descending thoracic aorta, and closed ductus arteriosus. Angiographic studies in the latter two patients demonstrated retrograde flow in both the aberrant right and left subclavian arteries (right and left vertebral-subclavian “steal”) into the descending thoracic aorta. Although the patient described by Morgan et al.18 had extensive neck collaterals in addition, neither of these patients (ages 19 and 7½ years) had collateral flow through the intercostal arteries by chest radiograph or angiographically. Intravenous angiographic studies were performed in the 16-year-old patient described by Gokcebay et al.,16 but the direction of flow in the subclavian arteries was not recognized. However, the chest radiograph revealed no evidence of rib notching, nor were large intercostal arteries seen at operation.

The two patients in this series with Type II-B interruption and a stenotic ductus arteriosus demonstrated similar radiologic findings. Neither had rib notching on chest films, or angiographic evidence of intercostal collaterals. Both had bilateral vertebral-subclavian “steal” with retrograde flow into the descending aorta. In one patient the left subclavian was stenotic at its origin16 and prevented retrograde flow from entering the descending aorta.

Review of these five patients indicates that retrograde flow in both subclavian arteries in patients with aortic interruption, Type II-B, seemingly prevents the development of intercostal collaterals on either right or left side.

Six patients have been reported in addition to the one patient in this series with Type II interruption and aberrant origin of the right subclavian artery from the right pulmonary artery through persistence of the right ductus arteriosus (Subtype II-C).5, 8, 29, 37-39 Another patient had the mirror image of this pattern39 (interrupted right aortic arch; right subclavian artery arising from the descending aorta; left subclavian artery arising from the left pulmonary artery through persistence of the left ductus arteriosus). These eight patients may be considered as having persistence of both right and left ductus arteriosus. Usually the right ductus arteriosus is narrowed, creating an area of obstruction between the right pulmonary artery and the right subclavian

FIGURE 2. Four-day-old infant with aortic arch interruption, Type II-A, and patent ductus arteriosus. A) Ascending aortic cineangiography following catheterization from the right axillary artery demonstrates aortic arch interruption distal to the left common carotid artery. B) Later frame demonstrates delayed opacification of the descending aorta from a left vertebral-subclavian "steal."
As little shunt flow occurs in either direction, the right subclavian artery has been designated as being “isolated” from main arterial channels, and is dependent on collateral circulation from either the left subclavian and possibly the left common carotid arteries (right vertebral-subclavian “steal”) or right carotid artery collaterals (right external carotid artery to thyrocervical and costocervical branches of the right subclavian artery).

Angiographic studies have been reported in one patient with Type II interruption and aberrant origin of the right subclavian artery from the right pulmonary artery. Antegrade flow was demonstrated in both the left subclavian artery and the aberrant right subclavian artery arising from the right pulmonary artery following a right ventricular injection. However, angiographic studies in one patient with isolation of the right subclavian artery in association with coarctation, hypoplasia of the aorta, atrial and ventricular septal defects, and bilateral patent ductus arteriosi, clearly demonstrated the presence of retrograde flow into the right vertebral artery, the right subclavian artery, and stenotic persistent right ductus arteriosus. Identical findings have been demonstrated angiographically by Sunderland et al. on the left side in a patient with a right aortic arch and origin of the left subclavian artery from the left pulmonary artery. In two patients reported by Shuford et al. with presumed origin of the left subclavian artery from the left pulmonary artery, one had retrograde filling of the left vertebral artery with opacification of the “isolated” left subclavian artery, and the other had collateral channels from the left carotid artery to the left subclavian artery (occipital branch of the left external carotid to thyrocervical and costocervical branches of the left subclavian artery). Angiographic studies in future patients with Type II interruption and “isolation” of the right subclavian artery can be expected to show similar findings on the right side, but it is conjectural whether antegrade or retrograde flow will be observed in the left subclavian artery arising from descending aorta.

In Type III interruption, the rarest form of aortic arch interruption with only seven previously reported cases, the aortic arch is discontinuous distal to the innominate artery, or distal to the right common carotid artery if the right subclavian artery has an aberrant origin. The hypoplastic ascending aorta continues into the neck as either the right innominate or right common carotid artery. As both the left common carotid and left subclavian arteries arise from the descending thoracic aorta, two large pathways are potentially available for collateral circulation to the descending aorta. In the case of Type III interruption reported by Pillsbury et al. with an intact ventricular septum and closed ductus arteriosus, retrograde flow in both these vessels (left carotid and left vertebral-subclavian “steal”) was inferred from angiographic studies. Interestingly, the retrograde flow in the left subclavian and left common carotid arteries in this patient was not sufficient and additional collaterals developed through the intercostal arteries, but only on the right side. Rib notching was confined to the right side on chest radiograph. At angiography antegrade flow was demonstrated in the normally arising right subclavian artery and collateral intercostal circulation was seen only on the right side.

One infant in this series had autopsy-proved Type III interruption and aberrant origin of the right subclavian artery.

Figure 3 Five-month-old boy with aortic arch interruption. Type II-A, and stenotic ductus arteriosus. Upper panel A) Close-up of right rib cage demonstrates rib notching involving the fifth, sixth, and seventh ribs. B) Injection of contrast material into the right axillary artery demonstrates aortic arch interruption distal to the left common artery. C) Later frame demonstrates delayed opacification of the descending aorta from a left vertebral-subclavian “steal” and right-sided intercostal arteries (arrows). LSA = left subclavian artery.
from the descending thoracic aorta. This infant, the first reported case of aortic arch interruption, Type III-B, had an additional potential channel for collateral circulation to the descending aorta (left carotid, right and left vertebral-subclavian "steal").

An unusual case of Type III interruption with aberrant origin of the right subclavian artery from the right pulmonary artery (Subtype III-C) has been reported recently by Tawes et al. It was not mentioned if angiographic studies were performed, but theoretically in similar patients one may visualize angiographically the presence not only of a left carotid and left vertebral-subclavian "steal," but also a right vertebral-subclavian "steal" with flow into the right pulmonary artery.

In summary, review of angiocardiograms of 17 patients in this series and those previously reported with aortic arch interruption indicates that patients with Type I interruption, normal origin of the right subclavian artery, and a stenotic or closed ductus arteriosus will have the potential for bilateral rib notching and collateral flow through both right and left intercostal arteries. If the right subclavian artery has an aberrant origin from the proximal descending thoracic aorta in a patient with Type I interruption and a stenotic or closed ductus arteriosus, angiography will show retrograde flow in the proximal right subclavian artery, and if additional collateral flow through intercostal arteries is present, it will be only on the left side. Patients with Type II or Type III interruption, normal origin of the right subclavian artery, and a stenotic or closed ductus arteriosus can be expected to show angiographically retrograde flow in the brachiocephalic vessels arising from the proximal descending thoracic aorta, and if additional collateral flow through the intercostal arteries is present, it will be only on the right side. If the right subclavian artery arises aberrantly from the proximal descending thoracic aorta in a patient with Type II or Type III interruption and a stenotic or closed ductus arteriosus, collateral flow through the intercostal vessels in all likelihood will not be present. Rather retrograde flow will be noted angiographically in both the right and left subclavian arteries, and in patients with Type III interruption, the left common carotid artery, and this retrograde flow in the subclavian arteries seemingly will prevent the development of intercostal collaterals. Obviously rib notching will not be present in these patients.

**Discussion**

Differentiation of aortic arch interruption into types and subtypes is useful since many radiographic and angio-
graphic findings are dependent on the site of arch interruption and origin of the brachiocephalic vessels. Likewise the surgical approach and ease of operation may be influenced by the location of arch interruption and origin of the brachiocephalic vessels. Arch reconstruction by direct anastomosis may be easier in patients with Type I than Type II interruption because more of the aortic arch is present in Type I than Type II. Occasionally, in patients with Type I interruption, the left innominate artery, or the left subclavian artery has been ligated distally and used for anastomosis to the descending thoracic aorta. Similarly, the left common carotid artery has been used for anastomosis in patients with Type II interruption.

In the plain film evaluation of patients with aortic arch interruption, discussed in Part I, attention should be directed to the presence or absence of a retroesophageal right subclavian artery on barium swallow radiographs, and rib notching. If rib notching is detected in the presence of other characteristic radiographic findings of aortic arch interruption it implies a stenotic or closed ductus arteriosus with collateral intercostal circulation to the descending thoracic aorta. The location of the rib notching, bilateral or unilateral, right or left side, is dependent on the site of arch interruption and origin of the subclavian arteries, and therefore, may permit differentiation into types and subtypes on plain radiograph (table I).

Patients with bilateral rib notching almost invariably will have Type I interruption with normal origin of the right subclavian artery (Subtype I-A) and a stenotic or closed ductus arteriosus. If rib notching is present only on the left side, one may expect the presence of Type I interruption with aberrant origin of the right subclavian artery and a stenotic or closed ductus arteriosus. One should see a retroesophageal right subclavian artery on barium swallow in these patients if this vessel arises from the proximal descending thoracic aorta (Subtype I-B). If one is confident that rib notching is present only on the left side, but the barium swallow fails to reveal any evidence of a retroesophageal right subclavian artery, one may suggest that, perhaps, the right subclavian artery has an aberrant origin from the right pulmonary artery (Subtype I-C). If rib notching is visualized only on the right side in association with the other findings, one may diagnose Type II or Type III interruption with normal origin of the right subclavian artery (Subtype II-A, or III-A) and a stenotic or closed ductus arteriosus. Patients with the clinical finding described by Rochette et al. of absent humoral and weak femoral pulses in association with bounding carotid pulses can be expected to have Type II interruption with an aberrant right subclavian, but will show no evidence of rib notching on chest radiograph if the ductus arteriosus becomes stenotic or closed, as retrograde flow in both the proximal left subclavian and aberrant right subclavian artery will prevent the formation of intercostal collaterals.

When the ductus arteriosus is stenotic or closed in patients with aortic arch interruption, the descending aorta is dependent on collateral circulation for its blood supply. The diminished pressure in the descending aorta results in retrograde flow in all brachiocephalic vessels arising from the proximal descending thoracic aorta. Most frequently this retrograde collateral flow is observed angiographically in the right and left subclavian arteries in the more commonly occurring Types I-B, II-A, and II-B, and may be present in Types III-A and III-B. In patients with aortic arch interruption Types III-A and III-B, retrograde flow also may be present in the left carotid artery.

If the ductus arteriosus is widely patent and equilibration of ventricular pressures can occur through the ventricular septal defect, pressures may be similar in both the ascending and descending thoracic aorta. In these instances antegrade flow may be observed angiographically in the brachiocephalic vessel arising from the proximal descending thoracic aorta.

During angiography, the duration of filming should be extended to evaluate potential collateral flow in the brachiocephalic vessels or intercostal arteries with the proximal portions of these vessels included in the field during filming. The recognition of collateral flow is important, as patients with aortic arch interruption and well-developed collateral flow into the descending thoracic aorta usually have had the longest survival without operation and are the best operative candidates for total correction. The frequent presence of a vertebral-subclavian "steal" and rarely a carotid "steal," as well as intercostal collateral flow insures adequate circulation to the descending thoracic aorta in patients with a stenotic or closed ductus arteriosus or failing right ventricle.

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References


Table 1. Location of Intercostal Collaterals in Patients with Interruption of the Aortic Arch and a Stenotic or Closed Ductus Arteriosus

<table>
<thead>
<tr>
<th>Aortic arch interruption type</th>
<th>Origin of right subclavian artery</th>
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<tbody>
<tr>
<td></td>
<td>Normal (Subtype A)</td>
</tr>
<tr>
<td>I</td>
<td>Bilateral</td>
</tr>
<tr>
<td>II</td>
<td>Right side only</td>
</tr>
<tr>
<td>III</td>
<td>Right side only</td>
</tr>
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*When only left-sided rib notching is seen in patients with suspected aortic arch interruption, the aberrant origin of the right subclavian artery can be determined by barium swallow. If this demonstrates a retroesophageal right subclavian artery, then this artery arises aberrantly from the proximal descending thoracic aorta (Subtype B); if, however, the barium swallow shows no evidence of a retroesophageal vessel, then the right subclavian artery may arise aberrantly from the right pulmonary artery (Subtype C).


27. Sundararajan V, Molthan ME: Truncus arteriosus (Type II) associated with interruption of the aortic arch (Type B). Am J Dis Child 123: 494, 1972


Correction

In the first part of this paper (Circulation 52: 714 1975) the relevant portion of figure 6 was not reproduced. The correct figure is printed here.

![Figure 6](http://circ.ahajournals.org/) 22-year-old female with operatively repaired aortic arch interruption, Type II-B. Left anterior oblique barium swallow demonstrates a large esophageal right subclavian artery arising from the descending aorta.
Complete interruption of the aortic arch. 2. Characteristic angiographic features with emphasis on collateral circulation to the descending aorta.

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