CLINICOPATHOLOGIC CORRELATIONS

Right Aortic Arch
Types and Associated Cardiac Anomalies

By LAURA KNIGHT, M.D. AND JESSE E. EDWARDS, M.D.

SUMMARY
An analysis was made of 78 pathologic specimens in which a right aortic arch was present. In four, the right arch was part of a double aortic arch. In 74 cases the right aortic arch was the only arch. The cases of right aortic arch could be subdivided into two groups as follows: (1) that with a retroesophageal aortic segment (three cases) and (2) that without a retroesophageal segment (71 cases).

The latter cases could be subdivided according to the nature of the origin of the branches of the arch as follows: (1) mirror image branching (60 cases), (2) aberrant left subclavian artery (ten cases) and (3) isolation of left subclavian artery (one case).

Congenital heart disease, of which the tetralogy of Fallot was the most common, was observed in each of the cases of right arch without retroesophageal aortic segment. Such an association was seen in one of three cases of right aortic arch with retroesophageal aortic segment and in three of the four cases of double aortic arch.

The potential for significant tracheal and esophageal obstruction is present in double aortic arch, in right aortic arch with retroesophageal segment and in right aortic arch without retroesophageal segment but with an aberrant left subclavian artery. In the latter condition, significant esophageal and tracheal compression depends upon the presence of a left-sided ductus arteriosus. This state was observed in five of ten cases with right aortic arch and aberrant left subclavian artery.

Additional Indexing Words:
Vascular rings
Double aortic arch
Congenital heart disease

WHEN A RIGHT AORTIC ARCH is present, it may be part of a double aortic arch or it may be the only channel at that level of the aorta. Moreover, when there is a single arch in the form of a right arch, variations occur with respect (1) to its course and branching and (2) to the state of the ductus arteriosus, as well as (3) association with intracardiac anomalies. In some patterns, the posterior wall of the esophagus is compressed either by a segment of the aorta or by the left subclavian artery, yielding the potential for a vascular ring. In other patterns, this does not occur.

In order to identify some of these variations, we studied the 78 specimens available to us. In 74 only a right arch was present and in four specimens there was a double aortic arch. The presence or absence of associated intracardiac anomalies was determined. For those cases with right aortic arch the course of the arch and the nature of its branching were identified. The presence or absence of the ductus arteriosus was determined and, when present, its position was identified.

Right Aortic Arch

When only a right aortic arch is present, it shows one of two relationships to the esophagus. In one form, the right aortic arch occupies a retroesophageal position. In the other, the right arch does not pass behind the esophagus but maintains its position to the right of it.

Right Aortic Arch Without Retroesophageal Aortic Segment

In 71 of the 74 specimens with right aortic arch studied, there was no retroesophageal aortic segment. Instead, the arch joined the upper descending aorta as

From the Departments of Radiology and Pathology, University of Minnesota, Minneapolis and the Department of Pathology, United Hospitals-Miller Division, St. Paul, Minnesota.
Supported by USPHS Research Grant 5 R01 HL05694, Research Training Grant 5 T01 HL05570, and Training Grant 1 T12 HE5853 from the National Heart and Lung Institute.
Address for reprints: Jesse E. Edwards, M.D., Department of Pathology, United Hospitals-Miller Division, 125 West College Avenue, St. Paul, Minnesota 55102.
it lay to the right of the esophagus. (In this type of aortic system, the lower portion of the descending aorta shifts toward the left so that the aorta enters the abdomen through a normally positioned aortic hiatus of the diaphragm.)

The 71 cases without a retroesophageal aortic segment were subdivided according to the pattern of branching of the arch as follows: (1) mirror image branching (60 cases), (2) aberrant left subclavian artery (ten cases), and (3) isolation of left subclavian artery (one case) (table 1).

Mirror Image Branching

In this condition, the aortic arch passes over the right main stem bronchus and joins a right-sided proximal descending aorta. The first branch of the arch is the left innominate artery and is followed by the right carotid and the right subclavian arteries, in that order (fig. 1). This pattern was observed in 60 of the 71 cases of right aortic arch without retroesophageal aortic segment.

In each of these 60, congenital cardiac disease was associated, of which the most common type was the tetralogy of Fallot, the latter being observed in 36 of the cases (48%). In 29 of the 36 cases with the tetralogy of Fallot, a stenotic pulmonary tract was present while in seven there was pulmonary atresia along with the other features of the tetralogy, the condition commonly called pseudotruncus arteriosus. The cardiac anomalies among the remaining 24 cases of right aortic arch without retroesophageal aortic segment and with mirror image branching took various forms but each was associated with deficiency of the ventricular septum, including two cases of complete transposition. It was of interest that in three of the cases the cardiac anomaly was tricuspid atresia. Asplenia, along with a variety of intracardiac anomalies, including obstruction to pulmonary arterial flow, was observed in nine cases.

In 57 of the 60 cases with right aortic arch and mirror image branching, the specimens could allow evaluation of the state of the ductus arteriosus or ligamentum arteriosum. Hereinafter, either of these structures will be referred to as ductus arteriosus (DA) (table 1). The DA was absent in 16 of the 57 cases. In 11 cases it was present on the right side, running between that segment of the right arch just beyond the right subclavian branch, above, and the right pulmonary artery, below. In 29 cases, the DA was left-sided, running from the proximal segment of the left subclavian artery (as it arose from a left-sided innominate artery), above, to the left pulmonary artery, below. Bilateral DA were observed in one case. In this case, previously reported as case 2 by Murray and associates, there was no pulmonary trunk and the pulmonary artery of each side arose from a homolateral DA.

In right aortic arch with mirror image branching, there is no retroesophageal vessel. Therefore, compression of either the esophagus or the trachea does not occur.

Aberrant Left Subclavian Artery

In cases with right aortic arch without retroesophageal aortic segment and with aberrant left subclavian artery, the arch passes over the right main stem bronchus and joins a right-sided proximal descending aorta. From before, backward, the branches are the left common carotid, the right common carotid and the right subclavian arteries. The left subclavian artery arises as the fourth branch from the left dorsal aspect of the upper descending aorta. It then runs toward the left arm by crossing the midline behind the esophagus, indenting its posterior wall (fig. 2). As with the right aortic arch and mirror image branching, each case with this aortic pattern was associated with a cardiac anomaly and, in each, the anomaly included deficiency of the ventricular septum. In four of the ten cases, the cardiac anomaly was the tetralogy of Fallot (two classical, two pseudotruncus arteriosus). In contrast to the pattern of mirror image branching, no examples of asplenia were found in the group with aberrant left subclavian artery. The anomalies in the remaining six cases were complete transposition (one case), persistent common atioventricular canal (one case), isolated ventricular septal defect (two cases), ventricular septal defect with pulmonary stenosis, other than the tetralogy (one case), and ventricular septal defect with cor triatriatum (one case).

The DA could be evaluated in each of the ten cases. It was absent in each of the four cases with the tetralogy of Fallot. In only one case was the ductus on the right side, that being the case of cor triatriatum with ventricular septal defect.

Table 1

Nature of Branches in 71 Cases of Right Aortic Arch Without Retroesophageal Aortic Segment and State of Ductus in 57 of the Cases

<table>
<thead>
<tr>
<th>Type of arch</th>
<th>Number of cases (%)</th>
<th>State of ductus arteriosus (57 cases)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mirror image branching</td>
<td>60 (84.5)</td>
<td>Absent: 16, Right: 11, Left: 29</td>
</tr>
<tr>
<td>Aberrant left subclavian</td>
<td>10 (14.1)</td>
<td>Absent: 4, Right: 1, Left: 5</td>
</tr>
<tr>
<td>Isolation of left subclavian</td>
<td>1 (.14)</td>
<td>Absent: 0, Right: 0, Left: 1</td>
</tr>
</tbody>
</table>

*In one case with mirror image branching, the DA was bilateral.
Right aortic arch without retroesophageal aortic segment and with mirror image branching. a. Absent ductus arteriosus (DA). b. Right-sided DA. c. Left-sided DA. Numbers in parentheses indicate state of DA in 56 examples of this type of aortic arch pattern. In the fifty-seventh case, the DA was bilateral.

aberrant left subclavian artery, the ductus was left-sided, running between the left pulmonary artery, below, and the left subclavian artery near its aortic origin, above. In each of the ten cases with aberrant left subclavian artery the posterior wall of the esophagus was indented by the left subclavian artery. When, however, a left DA was present, the esophageal compression was significant and compression of the anterior wall of the trachea by the pulmonary arterial bifuration may also have occurred.

When in a right aortic arch without retroesophageal aortic segment the left subclavian is aberrant, the situation differs from the state of mirror image branching. In the latter, no major vessel lies behind the esophagus. In the former, the left subclavian artery passes behind the esophagus and causes an indentation in the posterior wall of the esophagus. The indentation is less wide than when it is caused by a retroesophageal segment of aorta (to be described). Whether the indentation of the esophagus is significant depends upon the presence or absence of the DA, and its position if present.

When the DA is absent, no significant obstruction of the esophagus results, the situation being comparable to that in the common state of left aortic arch with aberrant right subclavian artery. Similarly, the presence of a right DA has little effect upon the esophagus. If a left ductus is present, it tends to create a vascular ring with resulting esophageal and tracheal compression.

Isolation of the Left Subclavian Artery

In this type of right aortic arch, the configuration of the arch and descending aorta is like that in the two foregoing types. Three branches arise from the arch which from before, backward, are the left common carotid, right common carotid and right subclavian arteries. The left subclavian artery is not attached to the aorta but is connected to the left pulmonary artery through a DA (fig. 3).

Only one case of this type was present in our series and it was associated with the tetralogy of Fallot.

Isolation of the left subclavian artery does not cause a vascular ring, as no vessel lies behind the esophagus. Failure of the left subclavian artery to arise from the aorta creates the potential for a "subclavian steal." 3, 4

Right Aortic Arch With Retroesophageal Aortic Segment

When part of a right aortic arch occupies a retroesophageal position, the pattern of the aortic arch system is as follows.
The solitary aortic arch passes over the right main bronchus to the right of the trachea and esophagus. It then turns abruptly toward the left behind the esophagus. Upon reaching the left side of the esophagus, it joins the proximal end of a left-sided descending aorta. At the junction of the right arch with the descending aorta, a diverticulum protrudes toward the left. From its upper end arises the left subclavian artery while the DA (usually a ligamentum arteriosum) inserts into the lower aspect of the diverticulum. The lower connection of the DA is to the left pulmonary artery (fig. 4). This pattern creates a vascular ring formed by the right arch to the right, its retroesophageal segment posteriorly, the DA to the left and the pulmonary arterial bifurcation anteriorly. The latter is held tightly against the anterior aspect of the trachea. It is commonly the case that in this type of right aortic arch, the arch ascends to a higher level in the thorax than does the right arch that is not associated with a retroesophageal segment. In the pattern under discussion, as well as in the pattern called right aortic arch without retroesophageal aortic segment and with aberrant left subclavian artery, the left subclavian artery arises as the fourth branch of the aorta. By some, these two patterns have been grouped under the term right aortic arch with aberrant left subclavian artery. It should, however, be emphasized that in one it is a segment of the aorta which lies behind the esophagus, while in the other it is the left subclavian artery.

Three cases of right aortic arch with retroesophageal aortic segment were observed in our series. Congenital heart disease was present in only one of this group, the anomaly being a ventricular septal defect of the atrioventricular canal type and associated with a cleft in the septal leaflet of the tricuspid valve.

**Double Aortic Arch**

Among the 78 cases studied in which a right aortic arch was present, the right arch was part of a classical double aortic arch in four instances.

The pattern is as follows. The ascending aorta arises anterior to the trachea and then divides into two arches. Each arch gives rise to the ipsilateral carotid and subclavian arteries; it then passes over its corresponding bronchus to meet the opposite arch behind the esophagus so as to form the descending aorta. The latter tends to lie to the left side of the midline (fig. 5). The DA runs between the left pulmonary artery, inferiorly, and that segment of the left arch between its subclavian branch, anteriorly, and its junction with the descending aorta, superiorly.

A double aortic arch is a classical example of a vascular ring, the ring being formed by the pattern of the aorta itself. In addition, the fact that the DA runs between the aorta and the pulmonary arterial system is an additional factor in causing compression of the anterior wall of the trachea. This occurs in a manner similar to that in right aortic arch with retroesophageal aortic segment. In the latter condition and in double aortic arch, the esophageal compression is caused by a segment of the aorta. The compression is wider than that seen when the retroesophageal vessel is the left subclavian artery in right aortic arch with aberrant left subclavian artery.

In three of the cases, each arch appeared sufficiently wide as to carry a normal complement of blood by itself. In two of these, each arch was of about the same width, while in one of these three the left arch was wider.
RIGHT AORTIC ARCH

In the fourth case, the left arch, although patent, was very narrow in that segment between the left subclavian artery and the descending aorta.

Of our four cases with double aortic arch, a cardiac anomaly was associated in three. The anomalies were as follows: classical tetralogy of Fallot, one case; pseudotruncus arteriosus, one case; and tricuspid atresia, one case, the latter being the one with a stenotic left arch.

Comment

Exclusive of examples of double aortic arch, this review indicates that there are two basic types of right aortic arch. In one, part of the aorta passes behind the esophagus (right arch with retroesophageal aortic segment). In the other, the aorta does not pass behind the esophagus, although in one of its variants the left subclavian artery occupies such a position. While the former creates a vascular ring, the latter does so only if a left-sided DA is present.

In the material reviewed, the type of right aortic arch without a retroesophageal aortic segment was much more common than that with a retroesophageal aortic segment (71:3). This ratio may not reflect the true relative incidence of these two conditions. It may be a byproduct of the fact that right aortic arch without retroesophageal segment is commonly associated with congenital heart disease and that this laboratory attracts cases with congenital heart disease.

In a less selective accumulation of cases with right aortic arch, Felson and Palayew found the two types to be nearly equal in incidence (26 without retroesophageal aortic segment, 33 with retroesophageal aortic segment).

The high incidence of congenital heart disease among cases of right aortic arch without retroesophageal aortic segment found in our studies is consistent with reports of others. 1, 6

References

Right Aortic Arch: Types and Associated Cardiac Anomalies
LAURA KNIGHT and JESSE E. EDWARDS

Circulation. 1974;50:1047-1051
doi: 10.1161/01.CIR.50.5.1047
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1974 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on
the World Wide Web at:
http://circ.ahajournals.org/content/50/5/1047

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/