

# Vascular Anomalies and Tracheoesophageal Compression: A Single Institution's 25-Year Experience

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**Background.** Vascular rings are uncommon anomalies in which preferred strategies for diagnosis and treatment may vary among institutions. In this report, we offer a description of our approach and a review of our 25-year experience.

**Methods.** A retrospective review was conducted of all pediatric patients with symptomatic tracheoesophageal compression secondary to anomalies of the aortic arch and great vessels diagnosed from 1974 to 2000.

**Results.** Thirty-one patients (38%) of eighty-two patients (mean age, 1.7 years), were identified with double aortic arch, 22 patients (27%) with right arch left ligamentum, and 20 patients (24%) with innominate artery compression. Our diagnostic approach emphasized barium esophagram, along with echocardiography. This regimen was found to be reliable for all cases except those with innominate artery compression for which bronchoscopy

was preferred, and except those with pulmonary artery sling for which computed tomography or magnetic resonance imaging, in addition to bronchoscopy, were preferred. Left thoracotomy was the most common operative approach (70 of 82; 85%). Ten patients (12%) had associated heart anomalies, and 6 (7%) patients underwent repair. Complications occurred in 9 (11%) patients and led to death in 3 (4%) patients.

**Conclusions.** In our practice, barium swallow and echocardiography are sufficient in diagnosing and planning the operative strategy in the majority of cases, with notable exceptions. Definitive intraoperative delineation of arch anatomy minimizes the risk of misdiagnosis or inadequate treatment.

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The recent literature on vascular rings demonstrates a certain degree of variability regarding the preferred strategy for the diagnosis of vascular rings [1-5]. Commonly used modalities include magnetic resonance imaging (MRI), computed tomography (CT), barium esophagography, bronchoscopy, and echocardiography. Regardless of the preferred diagnostic approach, the recognized goal is detecting and defining the anomaly so that an appropriate operative approach can be selected and executed while minimizing morbidity and mortality. Our diagnostic approach for the past 25 years has been relatively uniform, relying primarily on plain film radiography and barium esophagography, along with echocardiography to rule out congenital heart anomalies. In this article, we review our 25-year experience with 82 patients and offer a rationale for our preferred strategy for diagnosis and surgical treatment.

## Patients and Methods

We retrospectively reviewed the charts of 82 patients with symptomatic compression of the trachea or esophagus secondary to anomalies of the aortic arch and great vessels, diagnosed from 1974 to 2000. Age at the time of operation ranged from 2 weeks to 12 years (mean, 1.7 years). The male to female ratio was 2 to 1. The number of patients presenting per decade was 17 in the 1970s, 21 in the 1980s, and 44 in the 1990s.

## Results

### *Types of Vascular Anomalies*

Patients were categorized according to six types of vascular anomalies: (1) double aortic arch (31 patients); (2) right arch left ligamentum (22 patients); (3) innominate artery compression (20 patients); (4) aberrant right subclavian artery (4 patients); (5) pulmonary artery sling (3 patients); and (6) aberrant left subclavian artery (2 patients). Out of those patients with double aortic arch, right dominance occurred in 25 patients (81%), left dominance in 5 patients (16%), and codominance in 1 patient (3%). The types and numbers of anomalies are summarized in Table 1.

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Table 1. Distribution of Vascular Anomalies

Type of Anomaly	No. (%)
Double aortic arch	31 (38)
Right arch left ligamentum	22 (27)
Innominate artery compression	20 (24)
Aberrant right subclavian artery	4 (5)
Pulmonary artery sling	3 (4)
Aberrant left subclavian artery	2 (2)

Presentation

Stridor and recurrent upper respiratory tract infection were frequent presentations, occurring in 38 of 82 patients (46%) and 29 of 82 patients (35%), respectively. Stridor was more common among patients with pulmonary artery sling, double aortic arch, and innominate artery compression. Recurrent infection was more common among patients with right arch left ligamentum. Feeding difficulty was the most common presentation of children with a symptomatic aberrant subclavian artery. The presenting features of the various anomalies are summarized in Table 2.

Diagnosis

Although a chest radiograph was obtained in all patients, a barium esophagram was the preferred study for documenting the presence of a vascular anomaly. An esophagram was obtained in all patients referred without a preexisting radiologic or bronchoscopic confirmation of a diagnosis (53 of 82; 65%). The esophagram reliably indicated the presence of a vascular anomaly and an appropriate operative approach. In most cases, the esophagram correctly identified the specific type of anomaly. However, the esophagram was not considered adequate for ruling out innominate artery compression.

Echocardiography (or angiography in the earlier years of the series) was performed in 22 of 82 patients (27%). Bronchoscopy was performed in 17 of 82 patients (21%), predominantly in cases of suspected innominate artery compression. Computed tomography or MRI was obtained in 9 of 82 patients (11%). In most cases, CT or MRI was obtained before referral. However, we did prefer these studies for cases of suspected pulmonary artery sling.

Table 2. Presenting Symptoms of Vascular Anomalies

Symptom <sup>a</sup>	Vascular anomaly				
	DAA (%)	RALL (%)	ASA <sup>b</sup> (%)	IAC (%)	PAS (%)
Stridor	18 (58)	5 (23)	0 (0)	12 (60)	3 (100)
Recurrent infection	7 (23)	11 (50)	2 (33)	8 (40)	0 (0)
Feeding difficulty	5 (16)	3 (14)	4 (66)	0 (0)	0 (0)

<sup>a</sup> Other infrequent presentations are not listed. Therefore percentages do not necessarily total 100. <sup>b</sup> Includes right or left.

ASA = aberrant subclavian artery; DAA = double aortic arch; IAC = innominate artery compression; PAS = pulmonary artery sling; RALL = right arch left ligamentum.

Table 3. Associated Congenital Heart Anomalies<sup>a</sup>

Vascular Anomaly	Congenital Heart Anomaly	
	No. (%)	Anomaly
RALL	7 (32)	VSD (4 patients) PAS <sup>b</sup> (2 patients) CoA (1 patient)
ARSA	1 (25)	TOF
DAA	1 (3)	VSD
IAC	1 (5)	ASD

<sup>a</sup> Percentages refer to the total number of each type of vascular anomaly. Not all congenital heart anomalies required surgery. <sup>b</sup> Includes one patient with left pulmonary artery stenosis and one patient with absent left pulmonary artery.

ARSA = aberrant right subclavian artery; ASD = atrial septal defect; CoA = coarctation of the aorta; DAA = double aortic arch; IAC = innominate artery compression; PAS = pulmonary artery sling; RALL = right arch left ligamentum; TOF = Tetralogy of Fallot; VSD = ventricular septal defect.

Operative Approach

A left thoracotomy was performed in 70 of 82 patients (85%). A right thoracotomy was utilized in 4 of 82 patients (5%). Two of these patients had an aberrant right subclavian artery. One of these patients had innominate artery compression and an atrial septal defect. The other patient had a pulmonary artery sling, which was discovered in the course of exploring the right chest after inadvertent perforation of the trachea during rigid bronchoscopy.

A median sternotomy was performed in 7 of 82 patients (8%). Two of these patients had pulmonary artery sling repaired on cardiopulmonary bypass. One patient had a double aortic arch with concurrent repair of a ventricular septal defect. Three of these patients had a right arch left ligamentum with concurrent cardiac procedures, repair of left pulmonary artery stenosis, reconstruction of absent left pulmonary artery, and repair of coarctation of the aorta and interrupted aortic arch. One patient had an aberrant right subclavian artery and tetralogy of Fallot (see Table 3). One patient with a symptomatic double aortic arch did not undergo surgical treatment secondary to refusal of the operation by the family.

For patients with double aortic arch, the nondominant component of the vascular ring was divided with intraoperative monitoring directing the site of division to preserve the carotid pulse and blood pressure or arterial saturation in the arms and left or right leg. In approximately 45% of patients, a ligamentum or patent ductus arteriosus was also divided. In patients with a right arch left ligamentum, division of the vascular ring was accomplished by dividing the ligamentum or ductus. For both double aortic arch and right arch left ligamentum, the trachea and esophagus were mobilized from surrounding tissues.

A left thoracotomy was also the preferred approach to an aberrant subclavian artery or compression from an innominate artery. A right thoracotomy was used in 2 patients with aberrant right subclavian artery. The aberrant subclavian artery was simply mobilized and divided without reimplantation. For innominate artery compression

Table 4. Complications of Surgical Treatment

Vascular Anomaly	Complication	Outcome
DAA	Aorto-esophageal fistula	Hemorrhage/death
DAA	Stitch abscess	Uneventful recovery
DAA	Esophageal stricture	Dilation/recovery
RALL	Wound infection	Uneventful recovery
RALL	Chylothorax	Ductal ligation/recovery
IAC	Postoperative stridor	Intubation/recovery
PAS	Bronchial leak	Multiple revisions/prolonged stay
PAS	Right heart failure	Exploration artery/death
PAS	Bronchial leak	Mediastinal infection/death

DAA = double aortic arch; IAC = innominate artery compression;  
PAS = pulmonary artery sling; RALL = right arch left ligamentum.

sion, the pericardial reflection at the aorto-innominate junction was secured to the posterior surface of the sternum. In most cases, this was performed with a limited left parasternal incision, although a right parasternal approach may also be used. Intraoperative bronchoscopy was inconsistently used to document relief of compression.

Our experience with pulmonary artery sling was limited to 3 patients. Our preferred approach has been median sternotomy with cardiopulmonary bypass to facilitate resection and reconstruction of the airway. In the case discovered at exploration for tracheal perforation, the trachea was divided at the site of perforation and the airway reconstructed posterior to the artery, utilizing selective intubation of the left bronchus rather than cardiopulmonary bypass. Cardiopulmonary bypass was used in the remaining 2 patients. Both of these patients had resection and reconstruction of the airway; however, 1 patient had repositioning of the pulmonary artery, and the other patient had division and re-anastomosis of the pulmonary artery.

#### Outcome

Complications occurred in 9 of 82 patients (11%) and led to death in 3 patients (4%). The specific complications and interventions are described in Table 4. The mean length of stay for the entire study period was 4.2 days, decreasing from 6.3 days in the 1970s to 3.2 days in the 1990s. The percentage of patients with 2 or less in-hospital days has increased from 7% in the 1970s to more than 30% in the 1990s. Most children demonstrated notable improvement in symptoms shortly after the operation. This was particularly true for children with symptoms related predominantly to compression of the esophagus. Seventy percent of children were free from compressive symptoms within 1 year of the operation. Tracheomalacia was documented by bronchoscopy in 7 patients (9%) in the late postoperative period (4 to 6 months or longer). The remaining patients noted mild

symptoms only with extreme exertion or agitation. Our results are similar to those reported by other authors [6-9].

#### Comment

Gross and Ware [10] defined the operative repair of double aortic arch in 1945 and the surgical management of other vascular anomalies in 1946, indicating the utility of left thoracotomy for division of the ring or symptomatic vessel. In that same year, Neuhauser [11] described the radiographic criteria for diagnosing the various types of anomalies, based on the chest radiograph and the barium swallow. (For a complete description of these criteria, please see Neuhauser's article.) In summary, they incorporate a knowledge of the typical anatomy of the various anomalies and their direct correspondence to certain patterns of effacement of the tracheal air column and esophageal dye column.

Current variations in the diagnostic approach originated with the subsequent availability of newer technologies. The introduction of CT in 1962 was followed 2 years later by a report describing the use of CT to diagnose vascular rings [12]. A similar trend is evident in the use of echocardiography and MRI, described in 1982 and 1986, respectively [4, 13]. The recent literature contains several reports advocating the use of these studies to more completely define the anatomy before operation. In our opinion, there are clear indications for echocardiography, but limited indications exist for MRI or CT. Despite the diagnostic power, CT and MRI can fail to delineate atretic segments. The consideration of cost and benefit and the risk of sedation and intubation frequently required to perform these studies is more relevant, particularly in younger patients with compromised airways. We reemphasize this as our opinion because there are no randomized or retrospective studies, including our own, documenting specific costs or complications associated with MRI or CT in this particular population of patients. (This information was not available from our records as most of these studies were obtained by referring physicians.)

From our perspective, the relevant questions in the evaluation proceed in the following manner: (1) Does this patient have a symptomatic vascular anomaly? (2) Based on the clinical presentation and the known association with certain vascular anomalies, is there a high likelihood of a significant congenital heart anomaly? and (3) Given what is not known from the chest radiograph and barium swallow, would an additional preoperative test provide information that would influence the operative approach? In our experience, a chest radiograph and a barium swallow are sufficient to answer the first question. Negative studies in the context of appropriate symptoms warrant bronchoscopy to rule out innominate artery compression.

The second question requires qualification. Given the frequently cited estimation that approximately 20% of children with symptomatic vascular rings have some type of congenital heart anomaly (CHA) [7, 8, 14], one ap-

Table 5. Composite Series: Relative Occurrence of Congenital Heart Anomalies and Association With Vascular Anomalies

Distribution of CHAs							
Total of 113 CHAs, not all specifically defined (18% of VAs)							
Ninety-four CHAs specifically defined, but not grouped according to VA							
CHA	No. (%)						
Ventricular septal defect (VSD)	34 (36)						
Tetralogy of Fallot (TOF)	30 (32)						
Coarctation of the aorta (CoA)	15 (16)						
Atrial septal defect (ASD)	9 (10)						
Right heart outflow obstruction (RHO)	3 (3)						
Transposition of great arteries (TGA)	2 (2)						
Truncus arteriosus (TA)	1 (1)						
Eighty-eight CHAs specifically defined and grouped according to VA							
Relative Occurrence of CHA per VA							
VA	CHA No.						Total (%) <sup>a</sup>
	VSD	TOF	CoA	ASD	RHO	TGA	
ARSA	5	5	10	2	0	0	22 (61)
RALL	12	9	2	0	2	1	26 (25)
ALSA	3	1	0	3	0	1	8 (27) <sup>b</sup>
PAS	2	1	0	1	0	0	4 (17)
DAA	9	10	2	2	1	0	24 (14)
IAC	3	0	0	1	0	0	4 (4)

<sup>a</sup> Percentage is the proportion of patients with a given VA diagnosed with a CHA. Denominators (total number of specific VA) are based on only those series reporting specific associations between VAs and CHAs. <sup>b</sup> Five patients had an associated Kommerell's diverticulum.

ALSA = aberrant left subclavian artery; ARSA = aberrant right subclavian artery; ASD = atrial septal defect; CHAs = congenital heart anomalies; DAA = double aortic arch; IAC = innominate artery compression; PAS = pulmonary artery sling; RALL = right arch left ligamentum; VA = vascular anomaly.

proach would simply recommend echocardiography for all patients. Although this is not unreasonable, we were curious to evaluate the recent literature regarding information about CHAs and the specific types of vascular rings, because such information might be useful in the context of a more selective application of echocardiography. Recognizing that data from the preechocardiography era, particularly data based on autopsy series, may differ from that of a contemporary population of living patients, we arbitrarily limited our review to clinical series of vascular rings with 10 or more patients reported in the last 20 years (including our series), giving a total of 918 patients [2, 5-9, 14-22]. General information about CHAs was available for 623 patients, whereas specific information about CHAs and specific types of vascular rings was available for 88 patients. The data are summarized in Table 5. Ventricular septal defect and tetralogy of Fallot accounted for over two-thirds of anomalies. Right arch anomalies are typically regarded as the vascular anomaly more likely to be associated with CHAs, consistent with the figure of 25% in this review. However, the percentage for aberrant subclavian artery is surprisingly high and remains more than 25% even if cases of aortic coarctation are excluded. Returning to the original consideration of selective use of echocardiography, and referring to Table 5, patients with innominate artery compression could potentially be excluded. Al-

though we regard the data in Table 5 with skeptical respect, it is our opinion (and more recent practice) that echocardiography should be used liberally.

The third question also merits qualification. There are four types of anomalies for which an approach other than a left thoracotomy may be appropriate: (1) pulmonary artery sling, (2) aberrant right subclavian artery, (3) left arch right ligamentum, and (4) double aortic arch with an atretic segment in the proximal right arch. For pulmonary artery sling, a low anterior compression of the esophageal dye column is the characteristic finding on barium swallow. We agree with others that CT or MRI and bronchoscopy adequately depict the length and degree of tracheobronchial stenosis and are relevant to planning the operative strategy. We favor median sternotomy and cardiopulmonary bypass; however, left thoracotomy is not unreasonable in the uncommon scenario in which airway reconstruction is not indicated. Median sternotomy would also be appropriate for concomitant repair of most types of vascular anomalies and CHAs. For an aberrant right subclavian artery, the need for a right thoracotomy depends on the surgeon's perception of the importance of reimplanting the subclavian artery and the perceived accuracy of the barium swallow. Although we recognize the theoretical concerns supporting reimplantation of the artery, we have not routinely done so in these young patients and have noted no specific compli-

cations. Moreover, we regard the barium swallow as reasonably accurate for depicting the presence of an aberrant subclavian artery; therefore, we do not consider additional preoperative testing necessary or relevant in this context. Another operative consideration is the management of Kommerell's diverticulum. Our limited experience does not provide an adequate basis for offering recommendations. However, given the natural history of most forms of aneurysmal abnormalities of the aorta, resection may be the more appropriate option. Nonetheless, the decision can be made intraoperatively after delineation of the anatomy. Left arch with right ligamentum and double aortic arch with an atretic proximal right arch, anomalies potentially not accurately diagnosed by barium swallow, justify a right thoracotomy (or perhaps median sternotomy). However, of the 918 cases in our review of the literature, there were only 2 patients (0.2%) with a left arch right ligamentum and less than 10 patients (1%) with a double aortic arch and an atretic proximal right arch. In our opinion, the infrequent occurrence of these anomalies does not justify the cost or morbidity of additional testing in all patients. A final consideration is the role of thoracoscopy [23]. Although certain surgeons may have expertise with this approach, general criteria to guide the selection of patients are not completely defined. Although we have no specific experience with this approach, we regard ongoing investigation with interest.

To summarize our experience, barium swallow is a simple, noninvasive test that provides the essential preoperative information. Bronchoscopy is indicated in cases of suspected innominate artery compression. Computed tomography or MRI with bronchoscopy is relevant to the workup of pulmonary artery sling. Echocardiography should be used liberally. In our opinion, all other preoperative testing imparts no significant influence on the operative strategy. The success of this approach mandates definitive intraoperative delineation of anatomy to minimize the risk of inaccurate diagnosis and incomplete or inappropriate treatment.

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## INVITED COMMENTARY

This retrospective analysis is highly provocative. The paper takes us back to the old days before computed tomography and magnetic resonance imaging. Emphasizing the value of a plain barium swallow for diagnosing the anatomy of vascular compression malformations, the authors remind us that sometimes very little can be

gained from these new techniques, if one has competently explored more basic diagnostic procedures beforehand. The point of reconsidering the basics before embarking on a technological overkill with a sick baby definitely has its merits. The diagnosticians, however, have to be sufficiently familiar with these old techniques,