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The presentation and management of vascular rings: An otolaryngology perspective

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VATS;
Thoracoscopic vascular ring repairs;
Vascular ring

Summary

Objective: To review the presentation and natural history of children with vascular rings and present management guidelines.
Methods: Retrospective study of tertiary care pediatric medical center charts from 1991 to 2002.
Results: There were 37 males and 27 females with a diagnosis of vascular rings. At presentation, 91% of patients had airway symptoms and 47% had esophageal symptoms. Airway symptoms included stridor (63%), recurrent respiratory infections (47%), respiratory distress (19%), and cough (17%). The most common esophageal symptom was dysphagia (27%). Pre-operative studies included: echocardiography (96%), chest X-ray (93%), barium swallow (75%), magnetic resonance imaging (MRI) (60%), and computerized tomography (CT) scan of the chest (59%). Surgical management included open ($n = 25$) and thoracoscopic ($n = 39$) approach. Complications included recurrent laryngeal nerve injury in five patients (8%).
Conclusion: Children with vascular rings present with respiratory and/or feeding difficulty. The evaluation should include chest X-ray, echocardiography, and barium

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swallow. Direct laryngoscopy and bronchoscopy are recommended to assess the degree of compression of the airway and/or esophagus, tracheomalacia, and vocal fold motion prior to intervention. Indication for surgical release is given when the diagnosis is made and can be assisted by advanced radiology studies. Surgical options include minimally invasive techniques involving either thoracoscopic or robotic-assisted repairs, as well as open procedures involving thoracotomy.

1. Introduction

Vascular rings are congenital vascular anomalies of the aortic arch complex that result in compression of the trachea and/or esophagus. These lesions often manifest with airway compromise due to extrinsic tracheal compression. Patients with airway symptoms usually present earlier, while those with esophageal symptoms present later in childhood, often associated with the onset of solid feeds. Congenital vascular anomalies can be broadly categorized as anatomically complete rings (as is seen in double aortic arch patients) or anatomically incomplete rings (as is seen in the remainder of vascular rings) [1—4].

As the airway and esophagus may be significantly compressed in patients with vascular rings, the otolaryngologist is often called upon to assist in the evaluation of these patients. Technological advances have led to changes in the evaluation and management of vascular rings. This study was designed to review our institutional experience with the presentation, evaluation, and management of vascular rings over the last decade and to present guidelines for the care of these patients.

2. Methods

A retrospective chart review was performed of all patients undergoing surgical division of vascular rings at the Children's Hospital Boston, covering the period 1991—2002. This study was approved by the Committee for Clinical Investigation at Children's Hospital Boston. Complete patient records were reviewed for this study. Pertinent data extracted from the patient's charts included: demographic information, presenting symptoms, preoperative imaging studies, intra-operative findings, outcomes, and complications. Radiology films and reports were reviewed prior to proceeding with surgical intervention. An unpaired *t*-test was performed to compare patient symptoms (stridor, recurrent infections, respiratory distress, cough, dysphagia, reflux, choking episodes, and failure to thrive) with their specific vascular ring (Statview, North Carolina).

3. Results

From 1991 to 2002, 64 patients (37 males, 27 females) underwent vascular ring division at the Children's Hospital Boston. The mean age at the time of surgery was 3.0 years (standard deviation 5.5 years). The mean weight at the time of surgery was 15.3 kg (standard deviation 20.4 kg). The most common presenting symptoms were related to the airway (92%) and the esophagus (45%). Presenting airway symptoms included stridor (63%), recurrent respiratory infections (47%), respiratory distress (19%), and cough (17%). The presenting esophageal symptoms included dysphagia (27%), reflux (6%), choking (3%), and failure to thrive (3%). One patient was without airway or esophageal symptoms.

The evaluation of patients with a suspected vascular ring included: chest X-ray (89%), echocardiography (95%), and barium swallow (73%). Higher resolution radiographic imaging, MRI (61%) and CT scan (58%) of the chest, were performed in a majority of patients in order to assess the patency of the ring and thereby determine whether a thoracoscopic approach or open thoracotomy approach would be preferable. A thoracotomy would be recommended if the smallest vessel that needed to be divided was at least 2—3 mm in diameter or larger since vessels smaller than that can be safely divided thoracoscopically. The vessel is typically a patent ductus arteriosus, or, in the case of a double aortic arch, may also be the smaller of the two arches. Direct laryngoscopy and bronchoscopy were preformed in 20% patients. Statistical comparison between double aortic arch and right arch left ligamentum arteriosum revealed no significance when comparing the imaging studies obtained with the type of vascular ring ($n = 64$).

The vascular rings identified were double aortic arch ($n = 27$, 42%), right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum ($n = 34$, 53%), and aberrant left subclavian artery ($n = 3$, 5%) with right aortic arch (2 patients) and with left ligamentum arteriosum (1 patient).

Surgical management included an open ($n = 27$) or thoracoscopic ($n = 39$) approach. Eight patients (12%) had complications; five of these were recurrent laryngeal nerve injury. Of all patients undergoing video-assisted thoracoscopic division of the vascular

ring, only one had injury to their recurrent laryngeal nerve; the other four patients with recurrent laryngeal nerve injury initially had a thoracoscopic approach which was converted to an open thoracotomy. Of these four patients, we are unable to comment upon during which phase of the operation (thoracoscopic versus thoracotomy) injury to the recurrent laryngeal nerve occurred.

The three remaining complications included pneumothorax, lung collapse, and chylothorax (all in different patients). There were no deaths in this series.

4. Discussion

The term vascular ring denotes a congenital anomaly of the aortic arch system that results in compression of the trachea and/or esophagus with resulting symptoms.

An understanding of the embryology of the great vessels is imperative to understanding the aberrant development that leads to vascular rings. During embryogenesis, the human aortic arch system develops with six pairs of aortic arches connecting two ventral and dorsal aortae [1,2]. A significant portion of the first, second, and fifth arches regresses [1,2]. The carotid arteries develop from the third arch. In normal development, the right fourth arch regresses resulting in a left aortic arch system [1,2]. In that setting, the dorsal contribution of the sixth arch disappears on the right side, while persisting as the ductus arteriosus on the left side [1—3].

A vascular ring occurs when specific portions of this primitive system persist or regress abnormally. When both fourth arches persist, a double aortic arch occurs [1,2]. When the left fourth arch regresses, a right aortic arch system is created [1,2]. The most common types of vascular ring anomalies that can result in airway/esophageal compression are (in decreasing order of frequency) [3]: (1) double aortic arch, (2) right aortic arch with left ligamentum arteriosum and aberrant left subclavian artery, (3) innominate artery compression, (4) aberrant right subclavian artery (as an isolated finding, this is frequent and often does not require intervention), (5) pulmonary artery sling, and (6) aberrant left subclavian artery [3]. The last four are not true vascular rings.

Vascular rings can be classified as true rings that are anatomically complete or partial rings that are not complete, though both can have similar clinical presentations and treatment algorithms. The complete vascular rings include double aortic arch and right aortic arch with aberrant left subclavian artery and left ligamentum. The incomplete vascular rings include innominate artery compression and pulmonary artery sling.

Double aortic arch is the only complete vascular ring and always necessitates surgical repair. Patients with a double aortic arch present with predominant airway symptoms (stridor, respiratory distress, and cough). They also present earlier than the other vascular rings since the aerodigestive tract is completely encircled by the ring, rather than only being partially compressed [5,6].

The right aortic arch with a left ligamentum arteriosum and aberrant left subclavian artery is almost as frequent as the double aortic arch. As this ring does not constrict the trachea and esophagus as much as seen in a double aortic arch, these patients usually present later, usually within the first few years of life. Airway symptoms in these patients are similar to those with a double aortic arch, though these patients also have dysphagia and can take a long time to finish a meal compared to their peers [1—3].

Innominate artery compression results from an innominate artery that originates more posterior and leftward off the aortic arch. Anterior tracheal compression occurs when this vessel courses rightward towards the thoracic outlet. Airway symptoms are present in these patients. These patients learn adaptive maneuvers (hyperextension) to improve airflow. Surgical management for innominate artery compression is to suspend (or pexy) the innominate artery to the posterior sternum [1—3].

The pulmonary artery sling is a rare vascular ring that occurs when the left pulmonary artery arises off of the right pulmonary artery and encircles the right main stem bronchus and trachea as it passes between the trachea and esophagus prior to entering the left lung, with esophageal compression anteriorly. Due to the significant compression in these patients, they usually present early in life with respiratory symptoms [1—3].

Aberrant right subclavian artery is seen in a patient with a left aortic arch and aberrant origin of the right subclavian artery as the last branch from the aortic arch (from the descending thoracic aorta) and results in posterior indentation of the esophagus. This anomaly is the most common vascular anomaly of the aortic arch system and can be found in 0.5% of humans, although the majority is asymptomatic [1—3].

Of all cardiac anomalies, aortic arch anomalies account for 1—2%. Presenting symptoms correlate with the type of vascular ring. Airway symptoms are found more commonly when the trachea is compressed such as in a double aortic arch, while swallowing symptoms are seen when posterior compression of the esophagus occurs as in the aberrant subclavian artery [5,6].

Table 1 Patients symptoms correlated with vascular ring ($n = 64$)

Patient symptoms	DAA ($n = 27$)	RALL ($n = 34$)	p Value (DAA:RALL)	ASA ($n = 3$)
Stridor	19 (70%)	19 (63%)	.127	2
Recurrent infections	14 (52%)	15 (50%)	.278	1
Respiratory distress	**9 (33%)**	3 (10%)	**.008**	0
Cough	1 (4%)	**10 (33%)**	**.004**	0
Dysphagia	4 (15%)	11 (37%)	.058	2
Reflux	1 (4%)	3 (10%)	.215	0
Choking episodes	1 (4%)	1 (3%)	.435	0
Failure to thrive	2 (7%)	0	.055	0

DAA, double aortic arch; RALL, right arch left ligamentosum; ASA, aberrant left subclavian artery. Totals are over 100% as many patients had multiple symptoms. Statistical significance is determined to be at $p < .05$ (bold typeface).

In our study, patients had airway symptoms according to the type of their vascular ring (Table 1). Patients with a double aortic arch had a significantly higher rate of respiratory distress. Patients with a right arch and left ligamentum arteriosum or aberrant left subclavian artery had significantly higher rates of cough and dysphagia. The evaluating physician may potentially use this information to focus a differential diagnosis of which vascular ring a patient may have (Table 1).

A complete preoperative evaluation is of paramount important to assess the degree of airway symptoms and determine the lack or presence of other conditions such as laryngomalacia, tracheomalacia, and status of vocal cord function. According to several studies, direct laryngoscopy and bronchoscopy is not part of the routine evaluation of patients with vascular rings [3,5—8]. Synchronous airway lesions (i.e. laryngomalacia) are common findings in children with stridor and it is crucial to assess these conditions prior to surgical repair so as to avoid overlooking synchronous airway lesions that may be the true etiology of a patient's symptoms [9]. Vocal fold paresis and/or paralysis is a known complication of surgical repair of vascular ring. A complete preoperative evaluation of vocal cord function and close postoperative follow-up should be part of the treatment protocol. Close follow-up of patients allows timely intervention to ameliorate this injury if the patient's symptoms (aspiration, significantly altered voice quality) want further surgery. Pre-operative bronchoscopy is important in assisting the physician in excluding other diagnoses such as tracheomalacia, which may be confounded by a radiograph demonstrating anterior compression of the trachea with a normal barium swallow, mimicking innominate artery compression [7]. A dynamic view of the airway (via bronchoscopy) would assist in the delineation between these two pathologies [7].

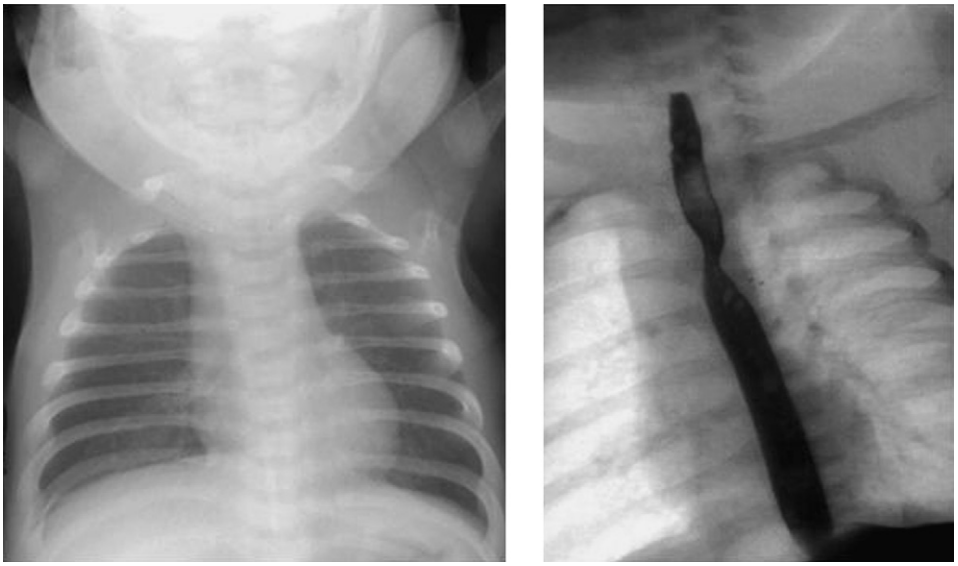

Fig. 1 Anterior—posterior (AP) radiograph of the chest in a child with a double aortic arch. *Note*: Indistinct view of the trachea on the AP view. AP esophagram showing compression of the upper thoracic esophagus by the double aortic arch: the more inferior (caudad) indentation is due to the left aortic arch and the more superior (cephalad) indentation is due to the right aortic arch.

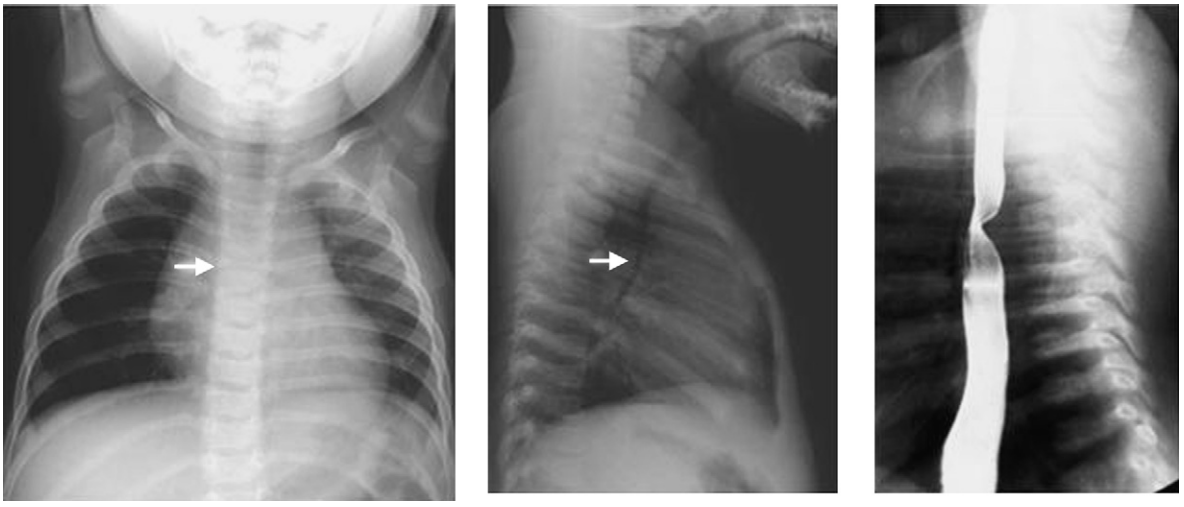

Fig. 2 (Left) Anterior—posterior radiograph of the chest in child with a right aortic arch. *Note*: Displacement of the trachea towards the left indicating a right aortic arch. (Center) Lateral radiograph of the chest demonstrating narrowing and anterior displacement of the trachea by the aberrant left subclavian artery which passes behind it. (Right) Lateral esophagram showing posterior impression of the esophagus by the aberrant left subclavian artery.

Pre-operative evaluation should also include frontal and lateral chest radiographs [9], barium swallow, echocardiography, and MRI or CT of the chest. The importance of frontal and lateral chest radiographs cannot be underestimated as a normal chest radiograph significantly decreases the likelihood of finding a vascular ring in a symptomatic patient [9]. Fig. 1 demonstrates the classic findings in patients with double aortic arch.

Fig. 3 Double aortic arch. Posterior image from a 3-D volume rendered MR angiogram demonstrating a double aortic arch with the left and right arches joining posteriorly. The left and right arches give rise to the left and right common carotid and subclavian arteries, respectively.

Controversy exists regarding the utility of MRI and CT scanning of the chest in patients with vascular rings. Some authors have indicated that MRI and CT scans are of limited utility in further elucidating the vascular pathology [3]. Instead, they emphasize the minimal morbidity and diagnostic ability of a barium swallow [3]. Chen et al., however, advocate the use of MRI and CT to define airway and cardiovascular pathology [8]. The detail obtained from MRI and CT allows the surgeon to thoroughly prepare for surgery and to also assess compression of the airway [11,12]. The detailed imaging gleaned from these advanced radiographic studies is seen in Figs. 2 and 3 and certainly can assist the surgeon in preoperative planning [13]. This is especially important if a thoracoscopic approach is planned, as large patent structures cannot be safely divided thoracoscopically and would require a thoracotomy instead.

Chen et al., note the high incidence of concomitant airway lesions in patients with congenital cardiac anomalies, lending further support to not only advanced imaging, but in performing preoperative direct laryngoscopy and bronchoscopy examinations in these patients [8]. Intraoperative bronchoscopy can be utilized to ensure release of tracheal compression in patients with airway compression. The bronchoscopy can be performed real-time as the ring is divided or the vessels suspended.

5. Conclusion

Children with vascular rings present with respiratory and/or feeding difficulty. The symptoms of the patient can help guide the differential diagnosis regarding which type of vascular ring the patient may have. Evaluation of patients with a suspected vascular ring should include chest X-ray, echocardiography, and barium swallow. Direct laryngoscopy and bronchoscopy are recommended to assess the degree of compression, tracheomalacia, and vocal fold motion prior to intervention. Surgical options include open repairs and thoracoscopic or robotic (surgical telemanipulation) repairs [10,14].

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