Vascular rings in childhood: diagnosis and treatment

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Abstract

Objective: to present the study carried out by the Pediatric Surgery Department of Instituto da Criança at the Medical School of Universidade de São Paulo regarding the diagnosis and treatment of children with aortic arch abnormalities and to define the role of complementary exams for diagnosis.

Methods: retrospective study of 22 patients with diagnosis of tracheoesophageal compression treated at Instituto da Criança from 1985 to 2000, analyzing pre- and postoperative clinical data, diagnostic exams and outcome.

Results: the most frequent diagnosis was right aberrant innominate artery (10 cases), followed by double aortic arch (7 cases) and right aortic arch (5 cases). Respiratory symptoms (86%) and early manifestation (76% since the neonatal period) were predominant. Nevertheless, most cases (60%) had the definitive diagnosis established only after 1 year of life. The most relevant examination for the diagnosis was the esophagogram. The correction of all the anomalies was carried out through left postero-lateral thoracotomy. There were no surgical complications. The outcome was worse in patients with delayed treatment. All children remained symptomatic for up to 6 months, although they had significant improvement in the postoperative period.

Conclusions: the diagnosis of vascular rings should be considered in children with early respiratory symptoms and in the wheezing baby with difficult control. The diagnosis may be established just through the esophagogram. Other image studies add few information and they are unnecessary in most cases. Less severe symptoms may persist for variable periods.

J Pediatr (Rio J) 2002; 78 (3):244-50: airway obstruction, tracheal stenosis, wheezing baby, esophageal stenosis, dysphagia lusoria, abnormalities of the thoracic aorta, surgery of the thoracic aorta.

Introduction

Congenital anomalies of the aortic arch, also known as vascular rings and anomalies of the great vessels, constitute a set of malformations that cause the compression of the esophagus and/or the trachea, being responsible for respiratory and digestive symptoms. These anomalies are rare malformations, classically subdivided into complete rings, such as the double aortic arch (DAA) and the right aortic arch, with persistence of the ductus arteriosus (RA/
Aortic arch malformations were initially described in adults, who mainly presented digestive symptoms. As a consequence, classic reports present the concept of “dysphagia lusoria” for the vascular compression of the esophagus. However, pediatric studies have demonstrated that symptoms often have their onset in the first months of life, with a predominant association with the respiratory system.

Generic diagnosis of tracheal and/or esophageal compression can be easily established; however, in most cases, this diagnosis is often made too late, thus delaying surgical treatment. Such delay can be attributed to the diversity of symptoms, which are often mistaken for pulmonary disorders or gastroesophageal reflux. The lack of information about the diagnosis of vascular rings and, therefore, the reduced level of suspicion are other relevant factors that lead to the delayed definitive treatment.

Several authors have reinforced the importance of differential diagnosis in the investigation of infants with chronic respiratory symptoms (wheezing babies), and have also discussed the adequate sequence of investigation. The use of new imaging techniques has not necessarily brought advantages to the investigation of such anomalies.

The objective of the present study is to present the experience of the Pediatric Surgery Department of the Instituto da Criança at the Hospital de Clínicas of the Medical School of Universidade de São Paulo (ICr-HCFMUSP) in the diagnosis and treatment of aortic arch anomalies, and to define the role of several complementary exams in the diagnosis of such malformations.

**Methods**

From February 1985 to April 2000, 22 infants with vascular rings were admitted to the Pediatric Surgery Department at the Instituto da Criança. Vascular rings were classified according to the nomenclature established by the Congenital Heart Surgery Database Committee and by the European Association for Cardiothoracic Surgery (Table 1).

**Table 1 - Incidence of vascular rings**

<table>
<thead>
<tr>
<th>Type</th>
<th>n</th>
<th>% cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>IAC</td>
<td>10</td>
<td>45%</td>
</tr>
<tr>
<td>DAA</td>
<td>7</td>
<td>32%</td>
</tr>
<tr>
<td>AR/LL</td>
<td>5</td>
<td>23%</td>
</tr>
<tr>
<td>Total</td>
<td>22</td>
<td>100%</td>
</tr>
</tbody>
</table>


Preoperative and postoperative symptoms, diagnostic procedures, and immediate and late postoperative evolution were obtained through a retrospective analysis of medical reports.

**Results**

**Preoperative symptoms**

In 16 infants (73%), the onset of symptoms occurred in the neonatal period; two infants presented acute respiratory insufficiency, requiring orotracheal intubation after birth. For the remaining 14 infants, the onset of symptoms took place between the second and the sixth months of life.

Respiratory symptoms were predominant; bronchospasm crises and permanent wheeze were the most frequent findings, detected in 19 patients (86%). These infants were initially treated as “wheezing babies”, not showing signs of improvement until the correct diagnosis was established. Other respiratory symptoms, such as respiratory stridor and recurrent pneumonia, as well as digestive symptoms, were reported. (Table 2) The latter were the least frequent findings, with low significance, and were never the patient’s major complaint.

**Table 2 - Prevalent symptoms**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>n</th>
<th>% cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic wheezing</td>
<td>19</td>
<td>86%</td>
</tr>
<tr>
<td>Cyanosis at breastfeeding</td>
<td>10</td>
<td>45%</td>
</tr>
<tr>
<td>Estridor</td>
<td>9</td>
<td>41%</td>
</tr>
<tr>
<td>Dysphagia/choking</td>
<td>8</td>
<td>36%</td>
</tr>
<tr>
<td>Total</td>
<td>22</td>
<td>100%</td>
</tr>
</tbody>
</table>

Fourteen infants (63%) had more than one episode of pneumonia prior to definitive diagnosis; five of these infants required mechanical ventilation and intensive therapy care.

**Diagnosis**

Despite the early onset of symptoms, in most cases (60%), diagnosis of compression caused by vascular ring was only established after the age of one.

Except for two patients, all the remaining infants were submitted to barium esophagogram, which allows for the identification of compression in the middle third of the esophagus and for the classification of the probable type of malformation.
Tracheobronchoscopy was performed in 15 infants (68%), generally revealing pulsatile compression in the lower third of the trachea. Two newborns submitted to mechanical ventilation due to acute respiratory insufficiency had their diagnosis established based solely on tracheobronchoscopy, due to the impossibility of performing an esophagogram. Tracheobronchoscopy also allowed for the diagnosis of associated severe tracheomalacia in three patients (14%).

In 10 infants, other imaging exams were carried out. In none of these exams, a computed tomography of the thorax was performed; one was submitted to nuclear magnetic resonance; and three underwent arteriography. These exams only confirmed the diagnosis already established by the esophagogram. None of these exams modified the initial diagnosis, surgical indication or surgical approach.

An echocardiogram was performed in seven infants in order to investigate eventual associated cardiac malformations. Three patients presented heart murmur, with interatrial communication (two cases) and interventricular communication (one case). The echocardiogram was not able to identify vascular rings in any of the seven cases.

Surgery
The age of children at the time of surgery varied from 21 days to four years and four months.

Transpleural left postero-lateral thoracotomy was the preferred approach in all cases.12,13

In cases of double aortic arch (DAA), the ligature of the anterior/left arch, which was non-dominant, was performed, followed by the ligature of the fibrotic ductus arteriosus.

In cases of right aortic arch (RA/LL), a section of the fibrotic ductus arteriosus was performed and the mediastinal masses were released in order to relieve esophageal and tracheal compression.

In cases of anomalous innominate artery (IR), ligature and section of the artery were carried out, without the need for arterial reimplant. The flow in the upper right limb remained normal, and was monitored by pulse oximetry.

An infant with RA/LL was reoperated six days after the first surgery. The decision to perform a new surgery on this patient was based on the persistence of intense symptoms and tracheal compression at bronchoscopy. After extensive dissection, with the release of mediastinal masses, significant improvement was observed.

Postoperative period
Except for two infants, one who needed reoperation and another one who presented with severe tracheomalacia, the remaining patients presented significant improvement in the immediate postoperative period. However, in none of the cases observed, symptoms were completely resolved in the initial period, persisting for up to six months after surgery.

No immediate intra- or postoperative complications were observed. Three infants died in the late postoperative period, two of them due to complications caused by chronic pneumopathy and recurrent infections, at six months and two years after surgery, respectively. These two infants were the first in this series; they had not been diagnosed until the age of four. The anatomopathological exam of the lungs in these cases revealed chronic pneumopathy with signs of bronchiolitis obliterans. The third child died in consequence of complications caused by severe tracheomalacia.

Discussion
Malformations of the aortic arch are rare; however, they are easily diagnosed, with a simple investigation based on nonspecific but significant clinical manifestations. In the present study, all infants presented respiratory symptoms, most of which appeared immediately after birth. Children with symptoms of upper respiratory obstruction (cyanosis at breastfeeding, inspiratory stridor) or presenting difficult-to-control asthma must be early investigated, in order to identify the presence of tracheal compression. Anomalies of the aortic arch must be included among the initial concern of every pediatrician who treats a “wheezing baby”.

As additional differential diagnoses, we should mention other thoracic masses that can cause compression of the trachea or esophagus, such as small bronchogenic cysts, esophageal duplication cysts and benign and malign tumors of the mediastinum. In these cases, non-characteristic radiological and endoscopic findings of vascular rings may indicate the need for complementary investigation, although the procedure is still surgical.

Tracheobronchoscopy is an objective and extremely useful exam for the diagnosis of these malformations.14 In some cases, especially those presenting intense respiratory symptoms, tracheobronchoscopy was the first exam to be requested. In infants intubated due to acute respiratory insufficiency, this exam was also essential for the diagnosis, since these patients were unable to undergo an esophagogram.

Computed tomography (CT) and nuclear magnetic resonance (NMR) are often requested; however, they do not contribute to diagnosis at all. Except when indicated with the specific objective of defining other forms of extrinsic compression of the trachea and esophagus, such as mediastinal tumors and cysts, these exams do not add relevant information to the diagnosis of anomalies of the aortic arch. In addition, in most cases of vascular rings, anomalous vessels are not pervious and, therefore, can not be contrasted, thus complicating the interpretation of the images.10,11,15
Arteriography is an invasive exam and must be indicated specifically in cases of complex cardiopathy, or when there is suspicion of ring in the pulmonary artery (cases with compression of the anterior wall of the esophagus and left bronchus). In the remaining situations, arteriography can be avoided, since the precise definition of the malformation is only established on the surgical act itself and, as already mentioned, in several situations, the structures of the vascular ring are atresic and are not contrasted in the arteriography.

Echocardiogram is useful for the investigation of associated cardiac malformations \(^{16}\); however, it does not allow for a differential diagnosis of vascular rings. In the present study, cardiac malformations presented low recurrence (13.6%), without clinical consequences. For this reason, these malformations did not require concomitant surgical correction.

Esophagogram, along with endoscopic exam, is the main diagnostic method. \(^{17}\) By using only the esophagogram, it was possible to identify the existence of compression in the middle third of the esophagus in 20 cases, and precisely identify the type of malformation in 18 (90%) cases, according to the image obtained. In only two cases, the type of malformation suggested by the esophagogram did not coincide with the intraoperative finding (esophagogram interpreted as DAA with intraoperative diagnosis of RA/LL and preoperative diagnosis of IR in an infant with DAA). However, mistakes observed in the preoperative diagnosis did not interfere with the surgical treatment, since the definition of the malformation and the appropriate correction were carried out through the same surgical access. Characteristic images \(^{17}\) corresponding to each type of vascular ring are described in Figures 1, 2, 3.

With regard to surgical treatment, ligature and section of structures were carried out in all cases, allowing for a complete release of esophageal and tracheal compression, without hindering the vascular flow. Thus, through the ligature of the ductus arteriosus and the atresic or non-dominant aortic arch, it is possible to solve cases of DAA and RA/LL. In cases of IR, although some authors recommend the reimplant of the right subclavian artery to its anatomic position, \(^{18}\) our experience demonstrates that this procedure is dispensable, adding unnecessary surgical morbidity, since distal flow is reestablished immediately after the clamping of the subclavian artery from collaterals, without functional damage. Surgical access through left postero-lateral thoracotomy allows for excellent exposure of vascular and mediastinal structures, and also permits defining the anatomy of the anomaly, as well as the appropriate surgical approach for its correction. In addition, this type of access also allows for the release of fibrotic masses that might persist after vascular ligature, preventing persistent compressive symptoms. Except for pulmonary artery sling, an extremely rare anomaly diagnosed when compression is identified in the anterior wall of the esophagus, the remaining anomalies of the aortic arch do not require extracorporeal circulation for correction.

The incidence of tracheomalacia associated with vascular rings was approximately 14%. Tracheomalacia and chronic

![Figure 1 - Esophagogram of the aberrant right subclavian artery. A- oblique radiography; B-lateral radiograph. Oblique and posterior impression, better viewed with the posterior acute compression in the lateral radiograph](image)
Figure 2 - Esophagogram of the double aortic arch. A - AP radiograph; B - lateral radiograph. Two esophageal compressions: one is larger, at upper position and on the right side, which corresponds to the right aortic arch, and the other one is smaller, at lower position and on the left side, which corresponds to the left aortic arch. In the lateral radiograph, it is possible to observe the large and round compression on the posterior wall of the esophagus, corresponding to the right aortic arch.

Figure 3 - Esophagogram of the aortic arch on the right with arterial ligament on the left. A - PA radiograph; B - lateral radiograph. Esophageal compression on the right, corresponding to the right aortic arch, and another lower compression on the left, corresponding to the left subclavian artery. In the lateral radiograph, evident posterior compression corresponds to the original bulb of the left subclavian artery.
pneumopathies are the most frequent causes of persistent respiratory symptoms in the postoperative period. These residual symptoms may be related to changes in tracheal dynamics, chronic lesions of the pulmonary parenchyma, chronic hyper-reactivity of the upper airways or recurrent infections.\(^7\)\(^1\)\(^9\)\(^2\) In the postoperative period, respiratory physiotherapy and antibiotic prophylaxis are of great importance for the long term treatment of these patients.

The analysis of the present cases allows us to conclude that the diagnosis of vascular rings must be considered in infants with respiratory symptoms in the neonatal period, and in infants with a generic diagnosis of difficult-to-control wheezing episodes ("wheezing babies"). Diagnosis must be established in a simple manner, through esophagogram and tracheal and digestive endoscopy. The remaining imaging exams add little information and are, therefore, dispensable. Finally, respiratory symptoms may remain in the postoperative period for different lengths of time, as a consequence of tracheomalacia or chronic pulmonary affections.

References


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