Surgery for vascular compression of the airway

Jacques Janson

Cardiothoracic Surgery and Paediatric Pulmonology, Tygerberg Children’s Hospital, University of Stellenbosch
Thoracic Surgery

Diagnostic:
- Bronchoscopy
- Lung biopsy
- Lymphnode biopsy
- Thoracoscopy

Therapeutic:
- Tracheal surgery
- Congenital lung lesions
- Bronchiectasis
- Infective lesions
- Diaphragm
- Pleural disease
Tygerberg Children's Hospital Data
Jan 2005- Jun 2017 (Age 0-12yrs)

<table>
<thead>
<tr>
<th>Procedure</th>
<th>N=455</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mediastinal lymph node enucleation</td>
<td>148</td>
<td>33</td>
</tr>
<tr>
<td>Open lung biopsy</td>
<td>65</td>
<td>14</td>
</tr>
<tr>
<td>Lobectomy</td>
<td>50</td>
<td>11</td>
</tr>
<tr>
<td>Pneumonectomy</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>Aortopexy</td>
<td>41</td>
<td>9</td>
</tr>
<tr>
<td>Tracheal resection</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>Double aortic arch</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>Empyema drainage and removal of fibrinous peel</td>
<td>78</td>
<td>17</td>
</tr>
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</table>
## Tygerberg Children's Hospital Data
### Jan 2005-Jun 2017 (Age 0-12yrs)

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<th>Procedure</th>
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<tr>
<td>Excision mediastinal tumor or cyst</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Diaphragm plication</td>
<td>23</td>
<td>5</td>
</tr>
<tr>
<td>Tracheo-esophageal fistula repair</td>
<td>6</td>
<td>1.5</td>
</tr>
<tr>
<td>Diaphragmatic hernia repair</td>
<td>7</td>
<td>1.5</td>
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Tracheal surgery

- 50% of the paediatric thoracic surgery we do is related to tracheal compression or stenosis.
- 33% are TB gland enucleations
- 11% are for vascular tracheal compression
Tracheal procedures

• Tracheal stenosis
  • Congenital (short segment and long segment)
  • Acquired post intubation

• Tracheal compression
  • Vascular Compression
  • Cardiac Causes (Large left atrium or pulmonary arteries)
  • Mediastinal lymph node compression
  • Mediastinal tumors and cysts

• Tracheo-esophageal fistula

• Tracheal tumor
Tracheal compression

• Vascular Compression
  • Cardiac Causes (Large left atrium or pulmonary arteries)
  • Mediastinal lymph node compression
  • Mediastinal tumors and cysts
During the formation of the aortic arch and of the pulmonary arteries, the failure of embryonic structures to fuse and regress normally can cause a wide spectrum of congenital anomalies of the configuration of the great vessels.

The resulting “vascular rings or slings” may represent an uncommon but potentially serious cause of compression of variable degree of the trachea, bronchi, and oesophagus.

Vascular compression

Complete Rings (40%)
- Double aortic Arch
- Right aortic arch

Incomplete Rings (60%)
- Aberrant innominate artery
- Retro-esophageal right subclavian artery
- Left pulmonary artery sling
Complete Rings

Double aortic arch

Right aortic arch with retroesophageal left subclavian artery

Right aortic arch with mirror image branching

57% 25% 18%
Incomplete rings:
Aberrant innominate artery
Incomplete rings: Pulmonary sling

50% associated with tracheal rings and stenosis
Pulmonary sling and tracheal rings
Incomplete rings:
Retro-esophageal right subclavian artery
Vascular compression

Monnier P. Pediatric airway surgery, 2011.
Vascular compression: Symptoms

- Stridor
- Chronic dry cough
- Brassy cough
- Wheezing
- Repeated respiratory infection
- Respiratory distress
- Apnoea
- Feeding problems
- Reflux
The team

- Pulmonologist
- Anaesthesiologist
- Radiologist
- Surgeon
• 209 patients evaluated with chronic cough
• Tracheal compression due to mediastinal vascular anomalies were diagnosed in 68 patients (32.5%).
• 141 children without mediastinal vascular anomalies
  • upper airways cough syndrome 55 patients (39.0%),
  • wheeze/asthma in 16 (11.3%),
  • cough variant asthma in 23 (16.3%),
  • psychogenic cough in 18 (18.8%),
  • gastroesophageal reflux disease (GORD) in 24 (17.0%)
  • pertussis in 5 (3.5%).
Percentage of patients needing surgery

- Aberrant innominate artery 15% surgery
- Right aortic arch 25% surgery
- Double aortic arch 100% surgery
Conservative management rationale:

- This congenital malformation is thought to be related to "mediastinal crowding" and, by three years of age, the aortic arch growth causes the innominate artery to move cephalad, anteriorly, and to the right, away from the trachea. Other factors, including thymic involution, growth of rib cage, and of the supportive tracheal cartilages may also explain the decreased incidence of tracheal compression with advancing age.
Follow-up of surgical correction of aortic arch anomalies causing tracheoesophageal compression: a 38-year single institution experience

Mark Ruzmetov*, Palaniswamy Vijay, Mark D. Rodefeld, Mark W. Turrentine, John W. Brown

Section of Pediatric Cardiothoracic Surgery, James Whitcomb Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, IN 46202, USA

Table 1 Distribution and ages of patients with vascular ring

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<th>Diagnosis</th>
<th>No. of patients</th>
<th>Mean ± SD (median age at surgery)</th>
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<tr>
<td>Double aortic arch</td>
<td>67</td>
<td>1.5 ± 2.8 y (5 mo)</td>
</tr>
<tr>
<td>Right arch dominant</td>
<td>55</td>
<td></td>
</tr>
<tr>
<td>Left arch dominant</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Right aortic arch-left ligamentum</td>
<td>77</td>
<td>1.7 ± 2.9 y (6 mo)</td>
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<td>6</td>
<td></td>
</tr>
<tr>
<td>Aberrant (retroesophageal) right subclavian artery</td>
<td>30</td>
<td>2.6 ± 6.7 y (3 wk)</td>
</tr>
<tr>
<td>Pulmonary artery sling</td>
<td>8</td>
<td>3.9 ± 8.6 y (1.3 y)</td>
</tr>
<tr>
<td>Innominate artery compression</td>
<td>1</td>
<td>2 wk</td>
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<td>183</td>
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<th>Vascular anomaly</th>
<th>Symptoms a</th>
<th></th>
<th></th>
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<tbody>
<tr>
<td></td>
<td>Stridor</td>
<td>Recurrent infection</td>
<td>Feeding difficulty</td>
</tr>
<tr>
<td>Double aortic arch (n = 67)</td>
<td>43 (64%)</td>
<td>28 (42%)</td>
<td>11 (16%)</td>
</tr>
<tr>
<td>Right arch left ligamentum (n = 77)</td>
<td>19 (25%)</td>
<td>41 (53%)</td>
<td>15 (20%)</td>
</tr>
<tr>
<td>Aberrant right subclavian artery (n = 30)</td>
<td>9 (30%)</td>
<td>10 (33%)</td>
<td>19 (63%)</td>
</tr>
<tr>
<td>Pulmonary artery sling (n = 8)</td>
<td>8 (100%)</td>
<td>6 (75%)</td>
<td>1 (13%)</td>
</tr>
<tr>
<td>Innominate artery compression (n = 1)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>TOTAL</td>
<td>80 (44%)</td>
<td>86 (47%)</td>
<td>46 (25%)</td>
</tr>
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a Other infrequent presentations are not listed. Therefore, percentages do not necessarily total to 100.
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Table 2  Associated cardiac diagnosis in 54 patients with vascular rings

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. of patients</th>
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<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>31</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>20</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
<td>13</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>11</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>8</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>5</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>4</td>
</tr>
<tr>
<td>Distal tracheal stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>1</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1</td>
</tr>
<tr>
<td>Mitral valve stenosis</td>
<td>1</td>
</tr>
<tr>
<td><strong>Overall</strong></td>
<td><strong>98</strong></td>
</tr>
</tbody>
</table>

*a* One of the patients had an associated cardiac anomaly.
Vascular compression: Diagnosis

- CXR
- Fibre optic bronchoscopy
  - Dynamic obstruction
- Ba-swallow
- Lung function when possible
- CT-chest
- Echocardiography
  - 10% have other cardiac defects
Ba-swallow patterns

De Leval M, Elliott M. *Vascular Rings*. In Surgery for Congenital Heart Defects. 2006
Vascular rings:
Double aortic arch
Double aortic arch
Dividing left aortic arch
Dividing left aortic arch
Surgeons view

De Leval M, Elliott M. *Vascular Rings*. In Surgery for Congenital Heart Defects, 2006
Outcome of double aortic arch surgery

• Mortality 3 % (30 day)
• Chylothorax 9%
• Left recurrent laryngeal nerve injury 3%
• Persistent stridor 34% (Follow up 6 months)
  – (Stridor was better than before surgery and related to tracheomalacia or tracheal stenosis.
  – Stridor did not require further surgery

Tracheal Compression by the Innominate Artery in Children

W. T. Mustard, M.D., C. E. Bayliss, M.D.,
Blair Fearon, M.D., D. Pelton, M.D., and
G. A. Trusler, M.D.

Vascular anomalies causing tracheobronchial and esophageal compression in children have been well documented by many authors [1–18]. Operative treatment by suspension of the innominate artery from the sternum was first performed by Gross [9] in 1948. Since then we have become increasingly aware that compression by the innominate artery is the commonest of all the vascular anomalies causing tracheal obstruction.

This study spans 16 years, from June, 1952, to June, 1968. During this time the diagnosis of innominate artery compression of the trachea was made in 285 patients ranging from neonates to children 3 years of age. Of these 285 patients with proved innominate artery compression, 39 (13.7%) were operated upon.
FIG. 1. Innominate artery compression occurring in an infant, with normal origin of the artery from the aorta demonstrated by aortography.

Case report

• 6 month old boy presents with recurrent lower respiratory infections
• HIV negative
• Persistent signs of large airway obstructions
• No response to asthma treatments
Aberrant innominate artery
Aberrant innominate artery
Indications for aortopexy

• Moderate to severe tracheal obstruction
• The presence of severe respiratory symptoms
• Presence of a single or of multiple respiratory symptoms that were severely affecting the patient’s quality of life
• No significant improvement after standard treatment of respiratory symptoms or comorbidities detected during the patient evaluation
Aortopexy through left thoracotomy with bronchoscopic guidance
Anterior aortopexy through mini sternotomy
Anterior aortopexy through mini sternotomy
Removing the thymus
Anterior aortopexy through mini sternotomy
Anterior aortopexy through mini sternotomy
Aberrant innominate artery

Pre-op

Post op
Aberrant innominate artery
Intraoperative bronchoscopy
Tracheal Compression as a Cause of Apnea Following Repair of Tracheoesophageal Fistula: Treatment by Aortopexy

By Marshall Z. Schwartz and Robert M. Filler
Galveston, Texas and
Toronto, Ontario, Canada

- Assosiation of TE-fistula repair and intermittent tracheal obstruction
- Up to 50% of patients have tracheomalacia

Journal of Pediatric Surgery, Vol. 15, No. 6 (December), 1980
Fig. 2. Caliber of air filled trachea as it is affected by swallowing (case 2). (A) During a barium meal the trachea is not collapsed when esophagus is relatively empty; (B) Distention of the esophagus causes tracheal compression.
Fig: Chest radiograph illustrating the enlarged hypoperfused left lung with anterior herniation of the left lung across the midline.
Left main bronchus compression

Tracheobronchogram illustrating the short segment narrowing of left main bronchus
Left mainstem bronchial narrowing: a vascular compression syndrome?

Evaluation by magnetic resonance imaging

Fig. 1 Composite drawing of DA positions. (1) Paraspinal, (2) 25–50% of circumference anterior to vertebral body, (3) 50–75% anterior to vertebral body, (4) prespinal

Left main bronchus compression

CT scan illustrating the compression of the left main bronchus between anteriorly displaced aorta and left pulmonary artery.
Left main bronchus compression
Surgery with bronchoscopic guidance

Divide ligamentum arteriosum

Posterior aortapexy
Vascular compression of left main bronchus

Pre-op

Post op
Vascular compression of left main bronchus

Pre-op CXR

Post-op CXR
Vascular compression

Summary

• Vascular compression must be in the differential diagnosis of airway obstruction
• Make the diagnosis
• Degree of obstruction?
• Which vessel is involved?
  – Double aortic arch
    • Most will need surgery
• Can symptoms be tolerated?
• Other contributing factors which can be treated?
• Wait and see or intervene?
Some dragons will go away...some needs to be sorted out