Aortic Uncrossing
For Circumflex Aorta

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Unusual forms of tracheobronchial compression in infants with congenital heart disease.

Three groups of unusual forms of tracheobronchial compression caused by vascular anomalies are presented. Three patients had an encircling right aortic arch with a left-sided descending aorta and ligamentum arteriosum (group 1), two patients had airway compression caused by a pincer effect between a malposed and enlarged ascending aorta and the descending aorta (group 2), and three patients had airway compression after an arterial switch operation for transposition of the great arteries (group 3). Symptoms developed in all patients before the age of 4 months, and six of them had multiple failed attempts at extubation before the surgical intervention directed at relieving the airway compression. Fiberoptic endoscopy was used in all patients as a first-line diagnostic tool and was 100% accurate in establishing the diagnosis. The operations performed were aortic uncrossing in group 1, dissection and aortopexy of the right or left main bronchus in group 2, and dissection of the left main bronchus and

- First reported in 1984 by Planché and Lacour-Gayet, reviewed by Robotin et al. in 1996.
- All 3 pts had previous ligamentum divisions through a left thoracotomy with persistent symptoms.
The “Uncrossing Procedure”

The vascular anatomy present in **circumflex aortas**: the right-sided aortic arch passes over the right main bronchus, then takes a retroesophageal course and joins the left-sided descending aorta.

Schema of the **aortic uncrossing procedure**. The right subclavian artery is divided. The aortic arch is transected proximal to the origin of the right subclavian artery, mobilized, and re-anastomosed to the left side of the ascending aorta and left carotid artery, in front of the trachea.

Lurie Children’s Experience

- 6 Patients – 2002 to 2014
- Age: 1.5, 2, 2.5, 4, 5, 6 years
- Two patients had previous division of left ligamentum without improvement
- Symptoms:
  - Noisy Breathing
  - Frequent Upper Respiratory Illnesses
  - Exercise intolerance
  - Dysphagia
Pre-op Bronchoscopy
Pre-op CT:
Right circumflex cervical aortic arch
Surgical Technique

- Median sternotomy, CPB, Hypothermia (18°C)
- del Nido cardioplegia, Circulatory arrest
- Aorta transected and oversewn distal to head vessels
- Aorta translocated anterior to trachea
- Descending aorta anastomosed to transverse arch
A "circumflex aorta" is a rare vascular ring caused by a right aortic arch with a left ligamentum arteriosum and a descending thoracic aorta that crosses posteriorly from right to left above the level of the tracheal carina (Fig. 1). The trachea and esophagus are compressed from the right aortic arch, the left ligamentum, and the posterior crossing aorta causing the typical symptoms of noisy breathing, dyspnea on exertion, dysphagia, and frequent upper respiratory tract infections. Although ligamentum division would divide the ring, that alone would not relieve the compression produced from this abnormal vascular anatomy. The aortic uncrossing procedure was first reported by Drs. Planché and LaCoeur-Gayet (Fig. 2). They also coined the term "circumflex aorta." They performed the operation in 3 patients, all of whom had previously undergone ligamentum division and had persistent postoperative symptoms. We now have experience with this operation in 4 patients.

All vascular ring operations at our institution begin with a preoperative rigid bronchoscopy to assess for unsuspected additional tracheal pathology such as complete tracheal rings and to evaluate the extent and location of compression from the aorta. A circumflex aorta would have both right-sided compression from the right aortic arch as well as posterior compression produced from the circumflex descending aorta. The "aortic uncrossing" procedure is performed through a median sternotomy with cardiopulmonary bypass, hypothermia, and a short period of circulatory arrest (Figs. 3-13). The heart is arrested with cardioplegia, head vessels are snared, and deep hypothermic circulatory arrest is established. The aorta is transected distal to the takeoff of the right subclavian artery and the proximal stump oversewn. The ligamentum arteriosum is ligated and divided. The right and left recurrent laryngeal nerves must be identified and preserved. The descending aorta is dissected from its posterior attachments and brought up on the left side of the ascending aorta. An arteriotomy is performed on the side of the ascending aorta adjacent to the left carotid artery. An anastomosis between the descending aorta and the arteriotomy is performed. The circulation is resumed and the patient is warmed and weaned from cardiopulmonary bypass. This procedure relieves both the posterior compression caused by the circumflex aorta and the right-sided compression of the trachea from the right aortic arch (Fig. 14).
Aorta pulled from behind trachea

Site of arteriotomy
Aortic Uncrossing

Ascending Aorta
Descending Aorta
Aorta Transected
Proximal Stump Oversewn
Translocated Descending Aorta
Anastomosis
Post-op CTA:
# Aortic Uncrossing Results

<table>
<thead>
<tr>
<th>Pt</th>
<th>Year</th>
<th>Circ Arrest</th>
<th>CPB</th>
<th>Postop LOS-days</th>
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<td>1</td>
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<td>84&quot;</td>
<td>5*</td>
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<tr>
<td>6</td>
<td>2014</td>
<td>26&quot;</td>
<td>126&quot;</td>
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<tr>
<td>Mean</td>
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<td>33&quot;</td>
<td>100&quot;</td>
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*Required reoperation on PO day 14
Aortic Uncrossing
Complications

Patient 1
• Respiratory insufficiency requiring reintubation and mechanical ventilatory support (18 d)

Patient 4
• Temporary tracheostomy
• Right Horner syndrome, bilateral recurrent laryngeal nerve paresis – both resolved
• Arrhythmia – sinus tachycardia

Patient 5
• Readmission with re-operation POD 14 – 14-mm aortic graft extension (esophageal compression)
Patient # 5
Barium Swallow
POD # 10
Pre-Uncrossing CT

Trachea

Esophagus
Pre-Uncrossing CT

Trachea

Esophagus
Pre-Uncrossing CT
Pre-Uncrossing CT
Post-Uncrossing CT
Patient # 5
Barium Swallow
After aortic extension graft
Aortic Uncrossing: Conclusions

- Advanced imaging (CT, MRI) allows identification of rare vascular ring pts with a circumflex aorta.
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- These pts will have marginal benefit from ligamentum division alone and in most cases should undergo aortic uncrossing.
- Note this is a major operation with substantial risk of complications.