Congenital Skills and Decision Making

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Question

What does a Congenital Heart Surgeon have in common with...
A Motorcycle Racer?
The Captain of a Ship?
Answer

Without the Congenital Skills and Decision-making Course
You could…
...Get knocked off track...
…Run Aground…
Or worse, Go Down with the Ship
Reoperation after Vascular Ring Repair

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I have no financial or regulatory disclosures.

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Vascular Rings

- Double Aortic Arch
  - Dominant Right Arch
  - Dominant Left Arch
  - Balanced Arches

- Right Aortic Arch with Left Ligamentum
  - Retroesophageal Left Subclavian Artery
  - Mirror Image Branching
  - “Circumflex” Aorta
Double Aortic Arch

- Dominant right arch 75%
- Dominant left arch 18%
- Balanced arches 7%

J Thorac Cardiovasc Surg 2003;129:1339
Right Aortic Arch, Left Ligamentum, Retroesophageal Left Subclavian Artery
Lurie Children’s Experience 1947-2012

Vascular Ring

<table>
<thead>
<tr>
<th>Condition</th>
<th># of Patients</th>
</tr>
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<tbody>
<tr>
<td>Double aortic arch</td>
<td>145</td>
</tr>
<tr>
<td>Right aortic arch/left ligamentum</td>
<td>153</td>
</tr>
<tr>
<td><strong>Total:</strong></td>
<td><strong>298</strong></td>
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Trends in vascular ring surgery

Carl L. Backer, MD, Constantine Mavroudis, MD, Cynthia K. Rigsby, MD, and Lauren D. Holinger, MD

Objective: We sought to review our experience with infants and children with anatomically complete vascular rings (i.e., double aortic arch and right aortic arch with left ligamentum) and define perioperative trends in diagnostic imaging, operative techniques, and clinical outcomes.

Methods: From 1946 through 2003, 209 patients (113 with double aortic arch and 96 with right aortic arch) underwent surgical repair. Mean and median ages at the time of the operation were as follows: double aortic arch, 1.4 ± 2.4 years and 0.75 years, respectively; right aortic arch, 2.7 ± 3.9 years and 0.9 years, respectively. Fourteen (14.6%) patients with right aortic arch had an associated Kommerell diverticulum. Cardiac diagnoses were present in 26 (12.4%) of 209 patients.

Results: There has been no operative mortality since 1959. In the past 30 years, mean hospital stay decreased from 8 to 3 days. Primary means of diagnosis has shifted from barium swallow and angiography to computed tomographic scanning or magnetic resonance imaging. In the past 10 years, 73% of patients had preoperative or intraoperative bronchoscopy. The technique of operation has shifted to a muscle-sparing left thoracotomy without routine chest drainage. In 7 recent patients with right aortic arch and a Kommerell diverticulum, the diverticulum was resected, and the left subclavian artery was transferred to the left carotid artery as a primary procedure.

Conclusions: At our institution, computed tomographic scanning has replaced barium swallow as the diagnostic procedure of choice for vascular ring evaluation. We recommend both preoperative bronchoscopy and echocardiography. Use of a muscle-sparing thoracotomy without routine chest drainage has decreased mean hospital stay. For patients with a right aortic arch and associated Kommerell diverticulum, we recommend diverticulum resection with left subclavian artery transfer to the left carotid artery.

(J Thorac Cardiovasc Surg 2005;129:1339-1347)
Autopsy Photo: Double Aortic Arch
CTA: 3D Reconstruction
Double Aortic Arch
Primary Indications for Reoperation after Vascular Ring Repair

1. Kommerell diverticulum
2. Circumflex aorta
3. Residual scarring
4. Tracheobronchomalacia requiring aortopexy
Burckhard F. Kommerell
1901-1990

• M.D., University of Tübingen 1925
• Chief of Radiology, Berlin 1934
• Described Aortic Diverticulum 1936
• Practiced Radiology for 30 years
• Pianist and Composer

Kommerell Diverticulum
“… pulsating mass behind the esophagus…consists of an aortic diverticulum from which the right subclavian originates.”

Remnant of the primitive right dorsal aorta (4th aortic arch)
Case Presentation

- 37-year-old woman
  - Right aortic arch
  - Retroesophageal Left Subclavian
  - Kommerell diverticulum

- Age 2: Ligamentum Division, Left Thoracotomy

- Age 35: Developed exercise intolerance with severe progressive DOE

- Dysphagia for solid foods
Reoperation #1, Left Thoracotomy

- Satinsky clamp placed on Kommerell diverticulum – when clamp released
- “Significant amount of bleeding”
- “Pledgetted suture still bleeding”
- “Controlled with index finger”
- “Purse-string suture”
- “Injected Tisseel sealant”
- “Several prolene pledgetted mattress sutures”
- “Quite a bit of chylous leak”
- Left subclavian transferred to left carotid artery (via supraclavicular incision)
Current Symptoms

- Recurrent dyspnea on exertion
- Audible inspiratory wheeze
- Any exercise leads to shortness of breath
- Dysphagia: Meat and Bread
- CT ➔ Residual Kommerell diverticulum, 18 x 20 x 20 mm
- Branch ➔ 50% tracheal narrowing caused by Kommerell diverticulum
Reoperation (#3)

- Median sternotomy
- Cardiopulmonary bypass
- Kommerell diverticulum base controlled with vascular clamp
- Femoral artery blood pressure unchanged
- Kommerell diverticulum resected, base oversewn
- Chylous leak oversewn
One year Later

- Tachycardia x 2 months – resolved
- Paralyzed left vocal cord successful collagen injection and nerve transfer
- Eating greatly improved
- Back to hiking and mountain biking with
- “No problems with breathing” – “fabulous”
Symptoms: Reoperations for Kommerell Diverticulum (n = 18)

- Dysphagia: 8
- Barky Cough: 7
- Wheezing: 5
- Stridor: 2
- DOE: 1
- Recurrent Upper Respiratory Infection: 1
Resection of Kommerell’s diverticulum and left subclavian artery transfer for recurrent symptoms after vascular ring division

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Division of Cardiovascular-Thoracic Surgery, Children’s Memorial Hospital, Chicago, IL, USA
Division of Pediatric Otolaryngology, Children’s Memorial Hospital, Chicago, IL, USA

Objectives: A Kommerell’s diverticulum in patients with a right aortic arch may become aneurysmal and be an independent cause of tracheoesophageal compression, even after ligation and division of a left ligamentum. We review the indications for and results of Kommerell’s diverticulum resection and left subclavian artery transfer in children with a right aortic arch who previously underwent vascular ring (ligamentum) division. Methods: From 1998 through 2001, eight children have been referred with recurrent respiratory symptoms (n = 8) and/or recurrent dysphagia (n = 4) after vascular ring division. Each child had a right aortic arch with a left ligamentum and had undergone division of the ligamentum elsewhere. All had a Kommerell’s diverticulum that was not addressed at the initial operation. All patients had a repeat left thoracotomy with resection of the diverticulum. Five patients had division and reimplantation of the left subclavian artery into the left carotid artery to relieve the sling-like effect of the retroesophageal left subclavian artery on the right aortic arch. One other patient had primary Kommerell’s diverticulum resection and transfer of the left subclavian artery to the left carotid artery. Results: The mean age at the initial operation was 1.7 ± 0.9 years, and the mean age at reoperation was 8.0 ± 3.7 years. In all patients postoperative bronchoscopy confirmed relief of the tracheal compression. There were no complications related to the subclavian artery transfer. Two patients developed postoperative chylothorax, one requiring thoracic duct ligation. The median hospital stay was 5 days. All patients had dramatic resolution of their preoperative symptoms. Conclusions: Kommerell’s diverticulum is an important anatomic structure that can cause recurrent symptoms in patients with a right aortic arch after ligamentum division. In selected patients, reoperation with resection of the Kommerell’s diverticulum and transfer of a retroesophageal left subclavian artery results in relief of symptoms. This technique has become our procedure of choice as a primary operation for children with a right aortic arch and a significant Kommerell’s diverticulum.
Reoperations for Kommerell Diverticulum (n = 18)

- Mean age at initial ligamentum division, 8 years
- Mean age at reoperation, 14 years
- Left Thoracotomy (n = 17)
  - 100 units/kg Heparin prior to clamps
  - Median size of resected Kommerell diverticulum, 1.5 x 2 cm
  - Left subclavian artery transferred to left coronary artery
    (n = 17 in chest, one supraclavicular)
- Median Length of Stay, 5 days
- All subclavian transfers patent
Kommerell Diverticulum Resection

Typical anatomy of right aortic arch, retroesophageal left subclavian artery, and a large Kommerell diverticulum

Resection of a Kommerell diverticulum through a left thoracotomy. A vascular clamp partially occludes the descending thoracic aorta at the origin of the Kommerell diverticulum

Completed repair
Kommerell’s Diverticulum Resection

Preoperative

Postoperative
Right Arch / Left Ligamentum Kommerell Diverticulum

- 8-yr-old
- Dyspnea on exertion
- Chronic cough
- Reflux
- s/p Left ligamentum division 18 months earlier
Reoperation

- Left Thoracotomy Resection of Kommerell Diverticulum
- Transfer Left Subclavian Artery to Left Carotid Artery
Right Arch / Left Ligamentum Kommerell Diverticulum

- 19-yr-old female
- Dysphagia for bread, meat
- Doxycycline tablet lodged in esophagus
- s/p Left ligamentum at age 2 yrs
- s/p Kommerell diverticulum resection at age 6 yrs, retroesophageal LSA left intact
- Right thoracotomy for ligation thoracic duct
Reoperation

- Left Thoracotomy
- Transfer Left subclavian artery to Left carotid artery
Computed Tomographic Imaging

Preop CT axial image of 7-year-old patient with a right aortic arch (R) and a Kommerell diverticulum (KD)

Postop CT axial image of the same 7-year-old patient

(Ann Thorac Surg 2012;94:1612-1618)
Primary Resection of Kommerell Diverticulum and Left Subclavian Artery Transfer.

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Division of Cardiovascular-Thoracic Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois; Department of Surgery, Northwestern University, Chicago, Illinois

BACKGROUND: A Kommerell diverticulum (KD) is an aneurysmal remnant of the dorsal fourth aortic arch. This can be an independent cause of tracheoesophageal compression. We previously reported resection of the KD with left subclavian artery transfer to the left carotid artery for recurrent symptoms in patients with a right aortic arch, left ligamentum, and retroesophageal left subclavian artery after prior ligamentum division. In 2001 we began resecting the KD and transferring the left subclavian artery to the left carotid artery in selected patients as a primary operation.

METHODS: From 2001 to 2011, 20 patients have had primary excision of a Kommerell diverticulum. Diagnosis was with computed tomographic scan (n = 14) or magnetic resonance imaging (n = 6) and bronchoscopy. Sixteen patients had a right aortic arch and 4 had a double aortic arch (right arch dominant). All patients were approached through a left thoracotomy. Fifteen patients had simultaneous division and reimplantation of the left subclavian artery into the left carotid artery.

RESULTS: Mean age at operation was 9.1 ± 6.5 years (range 1.5 to 29.1 years). Symptoms included cough, wheezing, stridor, dysphagia, and dyspnea on exertion. Selection criteria included KD greater than 1.5 times the size of the left subclavian artery and posterior pulsatile compression of the trachea on bronchoscopy. There were no complications related to subclavian artery transfer. No patient required a blood transfusion. No patient had a recurrent laryngeal nerve injury or chylothorax. The mean hospital stay was 4.3 ± 2.5 days. All patients had resolution of their preoperative airway and esophageal symptoms.

CONCLUSIONS: In selected patients with a vascular ring we now recommend resection of the associated Kommerell diverticulum and transfer of the retroesophageal left subclavian artery to the left carotid artery as a primary procedure. This strategy requires comprehensive and precise preoperative imaging with either computed tomography or magnetic resonance imaging.
Aortic Uncrossing
(Anterior Aortic Translocation)

“Circumflex Aorta”: Right aortic arch with left ligamentum and left sided descending thoracic aorta.
### UNUSUAL FORMS OF TRACHEOBRONCHIAL COMPRESSION IN INFANTS WITH CONGENITAL HEART DISEASE

| M. C. Robotin, MBBS, FRACS | 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
|
| J. Bruniaux, MD | |
| A. Serraf, MD | |
| M. Sousa Uva, MD | |
| R. Roussin, MD | |
| F. Lacour-Gayet, MD | |
| C. Planché, MD | |

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

Fig. 1. 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.

Fig. 2. 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 reanastomosed to the left side of the ascending aorta and left carotid artery, in front of the trachea.
Lurie Children’s Experience

- 4 Patients
- Age: 1.5, 2.5, 5, 6 years
- Two patients had previous division of left ligamentum without improvement
- Symptoms:
  - Noisy Breathing
  - Frequent Upper Respiratory Illnesses
  - Exercise intolerance
  - Dysphagia
Surgical Technique

- Median sternotomy
- CPB, hypothermia, circulatory arrest
- Aorta translocated anterior to trachea
- Mean circulatory arrest 34”
- Mean CPB time was 98”
- One patient had associated CoAo
- No mortality or neurological complications
Pre-op CT:
Right circumflex cervical aortic arch
Post-op CT:
Other Creative Strategies for Right Aortic Arch Patients with Recurrent Symptoms

- Grillo and Wright PTFE Sling
- Konstantinov and Puga Arch Division with Interposition Graft
- Martin Elliott Aortopexy
Tracheal Compression With “Hairpin” Right Aortic Arch: Management by Aortic Division and Aortopexy by Right Thoracotomy Guided by Intraoperative Bronchoscopy

Hermes C. Grillo, MD, and Cameron D. Wright, MD

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**Background.** Four patients with severe tracheal obstruction due to right aortic arch, aberrant left subclavian artery, diverticulum of Kommerell, ligamentum or ductus arteriosum, and, additionally, right descending aorta, mild pectus excavatum, and high aortic arch apex, with narrow space between the ascending and descending aortic limbs, underwent division of ligamentum, excision of diverticulum and division (and reimplantation) of aberrant subclavian, either in multiple or single operations, but failed to achieve relief of obstruction.

**Methods.** In addition to the procedures noted, fabric sling aortopexy of ascending and descending aortic limbs around adjacent ribs, with or without aortic division after prosthetic graft between ascending and descending aortic limbs was required, all performed through a right thoracotomy and adjunctive cervical incision, and with flexible bronchoscopic monitoring of each step.

**Results.** Three patients obtained full relief of airway obstruction, which has persisted in follow-up from eight to over 12 years. One who had persistent severe tracheal malacia after prior tracheal resection and resultant chronic pulmonary sepsis died from these complications.

**Conclusions.** In this unusual subset of a rare vascular ring anomaly, radical methods were necessary for correction of airway obstruction after failure of prior conventional procedures.

Surgically Corrected Right-Sided Aortic Arch

deGoma EM, JACC 2011;57:2375
A 32-year-old woman with dysphagia due to an unusual form of right aortic arch and anomalous left subclavian artery had successful repair after two previous failures. The definitive repair was accomplished by resection of the retroesophageal portion of the right aortic arch. The continuity of the aorta was established with a prosthetic graft. The operation was performed through a median sternotomy with cardiopulmonary bypass and circulatory arrest.

(Ann Thorac Surg 2001;72:2121-2123)
Initial Anatomy

Final Repair

Konstantinov IE. *Ann Thorac Surg* 2001;72:2121-2123
Aortopexy as treatment for tracheo-bronchomalacia in children: An 18-year single-center experience*

Emmeline E. Calkoen, MS; Hany O. S. Gabra, MD, FRCSI; Derek J. Roebuck, MRCPCH, FRCR, FRANZCR; Edward Kiely, MD, MB, BCh, FRCSI, FRCS, FRCPC; Martin J. Elliott, MBBS, MD, FRCS

Objectives: To define the factors influencing the outcome of aortopexy as management of tracheo-bronchomalacia.

Design: A retrospective, single-center, observational, cohort study.

Settings: Surgical services in a tertiary care hospital.


Interventions: None.

Measurements and Main Results: Comorbidity (categorized into three groups), surgical approach, and location of malacia were reviewed and analyzed in relation to mortality, need for a second intervention, complications, time to extubation after surgery, intensive care unit stay, and clinical improvement. Median age at surgery was 24 wks (1 wk–541 wks). Two surgical approaches were used: median sternotomy (n = 46) and left anterior parasternal (n = 59). Long-term (>2 yrs) follow-up was available for 73 patients (median = 7.0 yrs [2–18 yrs]); 73% were asymptomatic, 18% had minor symptoms, and 9% needed either ventilation or tracheostomy. The overall mortality rate was 9%, of which one-third was airway-related. Multivariable analysis revealed that major comorbidities were a significant risk factor both for mortality and the need for further procedures (re-do surgery, tracheostomy, internal stents) in contrast to surgical approach and involvement of the bronchus. Intensive care unit stay and days of ventilation after surgery were also significantly higher in patients with major comorbidities.

Conclusions: Aortopexy proved to be an effective treatment for most cases of tracheo-bronchomalacia, but major comorbidity was associated with an adverse outcome. Our data suggest that aortopexy should be considered in most cases of severe tracheobronchomalacia.

(Pediatr Crit Care Med 2011;12:545-551)
Conclusion

- Five percent of vascular ring patients develop recurrent symptoms after ring division
  - Dysphagia
  - DOE
- Evaluation is with CTA and bronchoscopy
- Most common reoperations are:
  - Kommerell diverticulum resection with left subclavian artery transfer
  - Aortic uncrossing for circumflex aorta
  - Aortopexy
- Dedicated multispecialty team is key
  - (Radiology, Cardiovascular Surgery, Anesthesia, ENT)
Q14. The most common indication for reoperation after vascular ring surgery is:

a. Aneurysm formation at the site of the vascular ring repair
b. Inaccurate diagnosis prior to the initial operation
c. A residual Kommerell diverticulum
d. Scar tissue from the previous operation
Q15. The evaluation of a patient for reoperation after prior vascular ring repair is best accomplished by a combination of:

a. Angiography and Bronchoscopy
b. Angiography and CT imaging
c. Bronchoscopy and CT imaging Magnetic
d. Resonance Imaging and Angiography
Q16. The primary feature that makes a right aortic arch vascular ring anatomy consistent with a “circumflex aorta” is:

a. Distal aorta coursing posterior to the trachea superior to the carina
b. Ligamentum arteriosum on the right side rather than the left
c. Mirror-image branching of the right aortic arch
d. Retroesophageal origin of the left subclavian artery