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VASCULAR RINGS AND SLINGS

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DISCLOSURE

The content of this presentation does not relate to any product of a commercial entity; therefore, I have no relationships to report.
Aortic arch and pulmonary artery abnormalities causing airway and esophageal obstruction by encircling them
VASCULAR RINGS AND SLINGS

- History
- Embryology and anatomy
- Fetal echocardiograms
- Clinical presentation
- Diagnosis
- Management
VASCULAR RINGS - HISTORY

- Johan Ludwig Hommell, autopsy, 1737
- Aaron Arkin, First case recognized during life, X-ray, confirmed by autopsy 1926
- Robert Edward Gross, used the term vascular ring for the first time in 1945
Gross

- Surgical relief of vascular ring in 1945 (autopsy in 5-month-old in 1931)
- Used term “Vascular Ring” for the first time
EMBRYOLOGY AND ANATOMY
Basic pattern
Fish
Salamander
Frog
Lizard
Snake
Bird
Mammal

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left

Right
Left
Mammal

Ductus Arteriosus

Right Pulmonary Artery

Left Pulmonary Artery

1

2

3

4

5

6

Mammal

CHILDREN'S HEART CENTER NEVADA FETAL CARDIOLOGY PROGRAM
1830s
Karl Ernst von Baer
(1792-1876)
EMBRYOLOGY

Right

Subclavian artery

Carotid artery

Left

Subclavian artery

Carotid artery

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EMBRYOLOGY

Carotid artery
Subclavian artery

Right

Carotid artery
Subclavian artery

Left

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NORMAL LEFT AO ARCH

NORMAL LEFT AO ARCH
EMBRYOLOGY

LEFT AO ARCH
ABERRANT RSCA
EMBRYOLOGY

Carotid artery
Subclavian artery

Right
Subclavian artery

Left

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VASCULAR RINGS

RIGHT AO ARCH
ABERRANT LSCA
LEFT DUCTUS ART.
vascular rings

Right Ao Arch
Aberrant LSCA
Left Ligamentum Art.
VASCULAR RINGS

RIGHT AO ARCH
ABERRANT LSCA
LEFT DUCTUS ART.
EMBRYOLOGY
VASCULAR RINGS

DOUBLE AORTIC ARCH

Right subclavian
Left subclavian
Right carotid
Left carotid
1877
Journal of Anatomy and Physiology
VASCULAR SLING

Normal

Sling
VASCULAR RINGS - DX

- Prenatal
  - Fetal Echocardiogram

- Postnatal
  - Symptoms
  - Incidental right aortic arch in a CXray
  - Incidental finding in echocardiogram
  - Barium Swallow
  - MRI - CT scan
VASCULAR RINGS - SYMPTOMS

- Occasionally in newborn period
- Airway obstruction
  - Stridor
  - Worse with feeding, crying, resp infections
  - “Asthma” with exercise
- Swallowing problems
  - More frequent in older
  - “Soup sign”
PRENATAL DETECTION OF CCHD

<table>
<thead>
<tr>
<th>Year</th>
<th>Prenatal Detection of CCHD (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2012-2013</td>
<td>70</td>
</tr>
<tr>
<td>2013-2014</td>
<td>75</td>
</tr>
<tr>
<td>2014-2015</td>
<td>80</td>
</tr>
<tr>
<td>2015-2016</td>
<td>90</td>
</tr>
</tbody>
</table>
PRENATAL DETECTION OF VASCULAR RINGS

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>% Prenatal Detection</td>
<td>0%</td>
<td>9%</td>
<td>55%</td>
<td>70%</td>
</tr>
</tbody>
</table>
VASCULAR RINGS - MANAGEMENT

- Surgery to divide ring in symptomatic
- It may be argued that observation may be appropriate if asymptomatic
- Difficult to recommend continued observation given the low surgical risk
Vascular Ring Diagnosis and Management: Notable Trends Over 25 Years

William N. Evans, MD\textsuperscript{1,2}, Ruben J. Acherman, MD\textsuperscript{1,2}, Michael L. Ciccolo, MD\textsuperscript{1,3}, Sergio A. Carrillo, MD\textsuperscript{1,3}, Gary A. Mayman, MD\textsuperscript{1,2}, Carlos F. Luna, MD\textsuperscript{1,2}, Robert C. Rollins, MD\textsuperscript{1,2}, William J. Castillo, MD\textsuperscript{1,2}, and Humberto Restrepo, MD, MPH\textsuperscript{1,2}
**Table 1. Diagnostic Data for All 92 Patients**

<table>
<thead>
<tr>
<th>Description</th>
<th>N = 92</th>
</tr>
</thead>
<tbody>
<tr>
<td>Current age years, mean ± SD</td>
<td>9 ± 7</td>
</tr>
<tr>
<td>RAA-ALS, n (%)</td>
<td>73 (79)</td>
</tr>
<tr>
<td>DAA, n (%)</td>
<td>17 (19)</td>
</tr>
<tr>
<td>Pulmonary artery sling, n (%)</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Isolated VR, n (%)</td>
<td>75 (82)</td>
</tr>
<tr>
<td>RAA-ALS, n</td>
<td>58</td>
</tr>
<tr>
<td>DAA, n</td>
<td>15</td>
</tr>
<tr>
<td>Pulmonary artery sling, n</td>
<td>2</td>
</tr>
<tr>
<td>Dysmorphic syndrome, n (%)</td>
<td>13 (17)</td>
</tr>
<tr>
<td>Prenatal detection 2004-2015, n PNDX/n total DX (%)</td>
<td>23/53 (43)</td>
</tr>
<tr>
<td>VR with intracardiac CHD, n (%)</td>
<td>17 (18)</td>
</tr>
<tr>
<td>RAA-ALS, n</td>
<td>15</td>
</tr>
<tr>
<td>DAA, n</td>
<td>2</td>
</tr>
<tr>
<td>Dysmorphic syndrome, n (%)</td>
<td>9 (53)</td>
</tr>
<tr>
<td>Prenatal detection 2004-2015, n PNDX/n total DX (%)</td>
<td>3/7 (43)</td>
</tr>
<tr>
<td>All dysmorphic syndromes and other malformations, n (%)</td>
<td>22 (24)</td>
</tr>
<tr>
<td>Down syndrome, n</td>
<td>4</td>
</tr>
<tr>
<td>22q11.2 deletion syndrome, n</td>
<td>6</td>
</tr>
<tr>
<td>Miscellaneous noncardiac malformations, n</td>
<td>12</td>
</tr>
</tbody>
</table>

Abbreviations: CHD, congenital heart disease; DAA, double aortic arch; DX, diagnosis; PNDX, prenatal diagnosis; RAA-ALS, right aortic arch, aberrant left subclavian artery and patent ductus arteriosus or ligamentum; SD, standard deviation; VR, vascular ring.
**Table 2. Surgical Data for All 92 Patients.**

<table>
<thead>
<tr>
<th>Description</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated VR, n (%)</td>
<td>75 (82)</td>
</tr>
<tr>
<td>Repaired, n (%)</td>
<td>52 (69)</td>
</tr>
<tr>
<td>RAA-ALS</td>
<td>35</td>
</tr>
<tr>
<td>DAA</td>
<td>15</td>
</tr>
<tr>
<td>Pulmonary artery sling</td>
<td>2</td>
</tr>
<tr>
<td>Asymptomatic to date</td>
<td>23</td>
</tr>
<tr>
<td>RAA-ALS, n</td>
<td>23</td>
</tr>
<tr>
<td>VR with intracardiac CHD, n (%)</td>
<td>17 (19)</td>
</tr>
<tr>
<td>Repaired/palliated, n (%)</td>
<td>17 (100)</td>
</tr>
<tr>
<td>VSD/AVSD, n</td>
<td>7</td>
</tr>
<tr>
<td>TOF/PA VSD, n</td>
<td>7</td>
</tr>
<tr>
<td>UVH, n</td>
<td>2</td>
</tr>
<tr>
<td>D-TGA, n</td>
<td>1</td>
</tr>
<tr>
<td>RAA-ALS, n</td>
<td>15</td>
</tr>
<tr>
<td>DAA, n</td>
<td>2</td>
</tr>
</tbody>
</table>

Abbreviations: CHD, congenital heart disease; DAA, double aortic arch; D-TGA, dextro-transposition of the great arteries; RAA-ALS, right aortic arch, aberrant left subclavian artery, and patent ductus arteriosus or ligamentum; TOF/PA VSD, tetralogy of Fallot/pulmonary atresia ventricular septal defect; UVH, univentricular heart; VR, vascular ring; VSD/AVSD, ventricular septal defect/atrioventricular septal defect.
VASCULAR RINGS - SUMMARY

- Prenatal detection is preferable
- Prenatal detection in Las Vegas is AMAZING!
- Symtoms = surgery
- Low surgical risk
VASCULAR RINGS - IDEALLY

- Prenatal diagnosis
- Observe in NICU for 48 hrs
- MRI/CT at about 4-6mo if asymptomatic
- Elective surgery 18mo - 2 yrs age
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