RINGS
SLINGS
OTHER THINGS
VASCULAR RINGS

Vascular anomalies resulting from abnormal development of the aortic arch complex, causing Compression of the trachea, esophagus or both. May be the cause of some cases of “croup” or “asthma”. May present at any age, including in adulthood, depending on severity

Formerly considered to be < 1% of CHD. Probably THE commonest CHD, due to better diagnosis
2 DAY OLD WITH RESPIRATORY DISTRESS

VASCULAR RING: R ARCH, AB L SUBCL, PDA, LPA
• 5 hour old with R arch.
• Ring?
• `R arch, aberrant L subclavian
• PDA starting to close
• Mild tracheal narrowing and esophageal compression
THE DUCTUS ARTERIOSUS SHRINKS SIGNIFICANTLY FROM A MUSCULAR TO AN ATROPHIC FIBROUS LIGAMENT
“Shriveled Fibrous Remnant of the Ductus Arteriosus”
As the ductus (or atretic segment of a double arch) shrinks, it pulls on one or both ends of the vessels on either side, deforming them.
TUG OF WAR
BETWEEN THE L SUBCLAVIAN ARTERY AND THE LEFT PA

The Ligamentum barely stretches if at all. The arteries enlarge and deform with growth.
THE MAJOR VASCULAR RINGS

COMMON

RIGHT Arch, Aberrant LEFT Subclavian and LEFT Patent Ductus/Ligamentum Arteriosum

DOUBLE Aortic Arch
With or Without Atretic Segments (usually on L)
Severest airway and esophageal compression
Key: 2 symmetric pairs of Carotid and Subclavian arteries on axial imaging

UNCOMMON

Mirror Image RIGHT Arch and POSTERIOR LEFT Patent Ductus/Ligamentum Arteriosum

LEFT Arch, Aberrant RIGHT Subclavian, and RIGHT Patent Ductus/Ligamentum
DOUBLE AORTIC ARCH

- 11 month old with extreme difficulty eating and stridor at rest
7 y.o. asymptomatic boy with suspected double aortic arch

UNUSUAL DOUBLE AORTIC ARCH, WITH SMALLER AND LOWER R ARCH. NO TRACHEAL NARROWING.

OBSERVE.
DOUBLE AORTIC ARCH

First described in 1737. First divided in 1945 (Boston Children’s). Usually present in the first few months of life. Require surgery in the first year of life.

Right/Posterior Arch dominant in 75%
Left/Anterior Arch dominant in 20%
Right = Left Arch in 5%

Formerly considered the commonest vascular ring
DOUBLE AORTIC ARCH
DOUBLE NIPPLE SIGN

3 Day old with mild SOB and tracheal narrowing
Large Diverticulum of Kommerell
Double aortic arch with complete vascular ring.
“Nipples” on both ends of L ligamentum: Kommerell and LPA
At 6 months, resect the Kom, reanastamose LScl, divide L Ligamentum
DOUBLE AORTIC ARCH WITH ATRETIC POSTERIOR L SEGMENT. NIPPLE AT L SUBCLAVIAN>> A LIGAMENT STILL PRESENT PRIOR L LIGAMENT (+/- KOM RESECTION), BUT STILL NB DYSPHAGIA WITH FOOD COMING OUT MOUTH WHEN EATING>> ATRETIC SEGMENT MUST STILL BE INTACT
DOUBLE AORTIC ARCH WITH ATRETIC POSTERIOR L ARCH

14 yo with prior vasc ring surgery, but still drooling saliva. Simulates R arch with aberrant L Scl, but is not, due to symmetric carotids and subclavians. Note air filled esophagus.
Le Gusta Sopita?
RIGHT AORTIC ARCH

Retroesophageal L Subclavian: 65%
Usually no other CHD (5-10%)
70% Tetralogy of Fallot

Mirror Image Branching: 35%
Usually is other CHD (98%)
90% Tetralogy of Fallot
R arch with Mirror image branching, and POSTERIOR Left ligamentum. RING

- R arch with Mirror image branching, and ANTERIOR Left
• 17 mth old who never ate solids. Recent respiratory distress and coin stuck in upper chest
R arch with Mirror image branching, tic of Kommerell and posterior left ligamentum

- 9 mth old with coarse breath sounds.
  Compressed esophagus and lower
R sided CIRCUMFLEX Aortic arch posterior to trachea and esophagus (instead of Kommerell), tic of Kommerell, aberrant left subclavian artery and left ligamentum. Vascular Ring

• 22 week old with R arch and noisy breathing. Echo shows R arch with probable aberrant left subclavian artery.
DIVERTICULUM OF KOMMERELL

Originally described by a Radiologist in a case of L arch and aberrant RSCA: Embryologic Remnant of the posterior part of the left 4th Aortic Arch.

Not a true diverticulum, rather, a focal dilatation/bulge

Decades ago, 50% of patients would present later in life with aneurysmal rupture or dissection, with high mortality.

Resect if Diameter > 1.5 x Diameter of vertical segment of L Subclavian, even if asymptomatic, between 1.5 and 2 years old.

Because of superior resolution of CT over MR, and importance of measuring a Kommerell diverticulum, prefer CT over MR, especially if small, eg <10 yo (DB)
INDICATORS OF A LEFT LIGAMENTUM AND SIGNIFICANT VASCULAR RING (DB)

1. See the ligament (MR only. Low SI in High SI fat)
2. Diverticulum of Kommerell
3. Diverticulum off Diverticulum of Kommerell
4. Stenosis/Occlusion at junction of Kommerell and vertical segment of LSCA
5. Diverticulum at LPA end of the left ligamentum
6. Single or Double “Nipple” on either end of Ligament
7. More Horizontal orientation of Ligamentum.
8. Esophageal lumen with Proximal distension with air.
9. Significant Compression/Indentation/Displacement of Trachea
LIGAMENTUM(L) ON MRI

14 yo with multiple episodes of respiratory distress. R Circumflex Retroesophageal Arch with Kommerell and L Ligamentum
7 week old asymptomatic, but R arch, aberrant L Subcl, Conical Kommerell w/ distal stenosis, Kom/Vertical ratio of 2.0, and proximal esoph dilation. May operate
• 2 day old asymptomatic
• Occluded L subclavian
• No tracheal narrowing
• No appr esoph dilation
• Kom/vertical LScl = 2.0
• Could resect Kommerell
• Could follow to 2 yo
What to do?
Ask Mom.

Surgery now, before symptoms

1. Divide Ligamentum
2. Resect Kommerell
3. Reimplant L subtle into Aorta
R Arch, Aberrant L Subclavian Inferior Diverticulum off Diverticulum of Kommerell
R ARCH, ABERRANT L SCL, HUGE KOM TIC, AND DOUBLE NIPPLES

1 day old with abnormal echo
R ARCH, ABB LSCL, DOUBLE NIPPLE, <0.5 MM DIAMETER PDA

1 day old with abnormal echo
R arch, aberrant L subclavian, Kommerell tic, post L ligamentum division, tic resection and LSCA to LCCA anastamosis

- 21 yr old with 2 surgeries for vascular ring. No longer dysphagia, but new occasional left arm weakness on exertion
LOOSE RINGS (DB)

No, or DELAYED SURGERY

No tracheal narrowing
No proximal esophageal dilatation
No Diverticulum, including Kommerell
Vertical Ligamentum
Pseudo Ring due to “L PDA” being a L Subcl to RPA Collateral. Tetralogy with small stented RVOFT, MPA and branch PAs. Multiple AP collaterals. No trach-esop stenosis.

- 1 month old with R arch descending on the right. Suspected vascular ring with L PDA.
5 month old with suspected double aortic arch and coarctation by echocardiography.

SMOOTH ARCH HYPOPLASIA. DISTAL ARCH TORTUOSITY. MEDIAL L SUBCLAVIAN

NO DOUBLE AORTIC ARCH OR COARCTATION.

SURGERY CANCELLED.
Pseudo Ring
PULMONARY ARTERY SLING
LPA OFF RPA, COMPRESSING R MAINSTREAM BRONCHUS, WITH PS DILATATION

5 week old with supraventricular tachycardia
PA SLING WITH LPA BETWEEN CARINA ANTERIORLY AND ESOPHAGUS POSTERIORLY

5 week old with supraventricular tachycardia
8 wk old with Tetralogy and absent pulmonary valve + PS. Suspected dilated PA’s impinging on airways.

SEVERE PULM STENOSIS AND BRANCH PA DILATATION, NARROWING MAINSTEM, RUL + RML BRONCHI
Tetralogy of Fallot with Absent Pulmonary valve and bilateral bronchial narrowing
Primary Bronchomalacia
CT: Absent RPA, but diverticulum off R PreInnominate Aorta c/w recently occluded AP Collateral to well formed R lung, ballooned to RPA

- 8 day old with non visualized RPA at echo. Aplastic, hypoplastic or discontinuous?
2 day old with respiratory distress. Echo suggesting Pulmonary atresia with MAPCAS
TINY RVOFT BULGE, WITH POSSIBLE TINY COMMUNICATION WITH SEVERELY HYPOPLASTIC MPA. SUGGEST BALLOON VALVULOPLASTY BEFORE UNIFOCALIZATION

2 day old with respiratory distress.
Echo suggested pulmonary atresia with MAPCAS
SEVERE PULMONARY HYPOPLASIA, NOT ATRESIA, PROVEN AFTER INJECTED RVOFT, WIRED, BALLOON VALVULOPLASTY AND PASSED CATHETER.
UNIFOCALIZATION SURGERY AVERTED

2 day old with respiratory distress.
Echo suggesting Pulmonary Atresia with MAPCAS
BERTHOTY BULGES IN CHD

Type 1: (Surgical)

Non Growing Fixed Length Collagen Ligament Between 2 Growing Vascular Structures:

- Diverticulum of Kommerell
- “Nipples” on either end of a Ligamentum.

Type 2: (Interventional Cardiology)

Morphologically Normal but Small Underdeveloped Vessel Beyond It:

- Absent Branch Pulmonary Artery
- Severe Pulmonary Hypoplasia
- Stick a Wire/Catheter/Balloon/Stent in it

Type 3:

Lower Surrounding Resistance Tissue:

- Pseudo IntraPulmonary IVC
- Vascular Malformation
BERTHOTY BULGES IN CHD

Type 4:

Increased Intraluminal Pressure/Flow:
Post-stenotic Dilatation; Aortic Stenosis, Coarctation, Tetralogy of Fallot with Absent Pulmonary valve

Type 5:

Weak Vascular Walls:
Collagen Vascular Disease; Marfan, Ehlers-Danlos, VSD /ASD/Sinus of Valsalva “Aneurysms” Pseudoaneurysm; Infection/Infarction/Surgical
55 yo female with chest pain.
CXR: R paracardiac hernia/fat/cyst/tumor.
CE CT Scan: “SOLID” R paracardiac tumor.
Adult Cardiac surgeon requesting biopsy
Coronary Angiogram: No stenosis
INTRA
PULMONARY
IVC
PSEUDO INTRAPULM IVC

Surrounded by mediastinal fat, that is thin laterally. Lung provides no resistance to the IVC, bulging into it.
ARE VASCULAR RINGS UNDERRATED?

Asymptomatic 16 year old Hiding his emaciation with muscle. Alternative to Lap band, Gastric Sleeve, Gastroenterostomy