Aortic Arch Anomalies: Embryologic Basis and Imaging Correlation

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Goals:

- Understand the embryologic basis of normal and abnormal aortic arch development
- Apply this knowledge to the understanding of normal and abnormal fetal 3 vessel and 3 Vessel tracheal views

Format:

- Simplified overview of cardiac embryology using 3D animations
- Ultrasound images from fetal echocardiograms to highlight the embryologic origins

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Acknowledgement

• Carol Mitchell PhD
Associate Prof in Medicine

• David McDougal/Evan Schultz
Animator and Graphic's Artists

• Sonographers

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Disclaimers

- The Goal of this lecture is to provide a simple easy to understand overview of Cardiac Embryology
- As such it may be an over simplification of some complex processes
- Developmental origins of some structures are hotly debated, and the simplest explanation may be chosen here.

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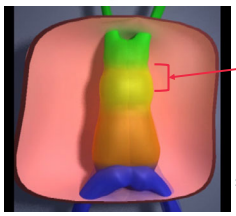
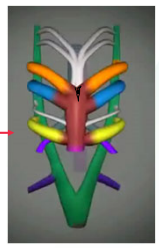
Straight heart tube(23-28 days)



TRUNCUS ARTERIOSUS

AORTIC SAC AND PHARYNGEAL ARCHES

SINUS VENOSUS



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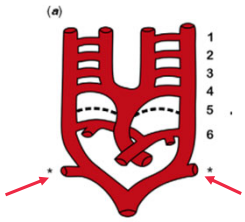
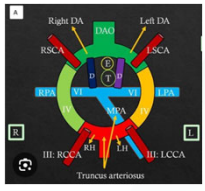
Models of Aortic Arch Development



Rathke's Diagram



Color Coded Edwards Double Arch Model

Sarv Priya et al Cureus 2020 Jul 30

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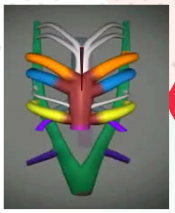
Overview

- Main Components :
  - Aortic Sac and its 2 horns
  - Paired Pharyngeal Arteries (Aortic Arch) 1, 2, **3-4 and 6**
  - Paired Dorsal Descending Aorta
  - 7th intersegmental arteries
- Primitive Paired Aorta
  - Ventral aorta and Dorsal Aorta connected by an arched portion in first pharyngeal arch
  - Aortic sac forms from fusion of paired ventral aorta
  - Subsequent paired pharyngeal arteries develop connecting Aortic sac ventrally to the Dorsal aorta
  - Dorsal aorta fuse just past the 7th intersegmental artery to form the descending aorta



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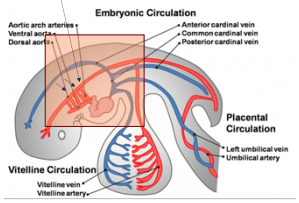
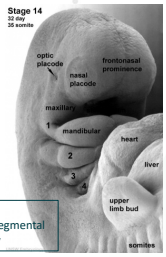
Embryo

ROLE OF NEURAL **CREST CELLS**



Dr Mark Hill UNSW Sydney

32 Day's



7th intersegmental Artery

UNSW Embryology

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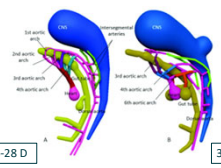
Over View

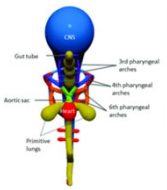
Significant remodeling occurs with lengthening as well as regression and **asymmetric** development of arteries



26-28 D

35-40 D

NOTE the close relationship of the Foregut and Developing Lungs in between the Ventral and Dorsal Descending Aorta



Coral Bravo MD: J Ultrasound Med 2016; 35:237–251

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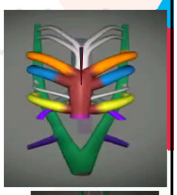
Derivatives of Aortic Arch

Embryonic Arch	RIGHT	LEFT
1st Pharyngeal (Aortic Arch) Artery	Regresses (Small Part of Maxillary Artery)	
2nd Pharyngeal (Aortic Arch) Artery	Regresses (Small contribution to Stapedial Artery)	
3rd Pharyngeal (Aortic Arch) Artery	Rt Common Carotid and part of Internal Carotid A,	Lt Common Carotid and part of internal Carotid A
4th Pharyngeal (Aortic Arch) Artery	Part of Rt Subclavian Artery	Transverse aortic arch between the LCA and LSCA
5th Pharyngeal (Aortic Arch) Artery	? Does not form in Humans	
6th Pharyngeal (Aortic Arch) Artery	Proximal: Proximal RPA Distal Regresses	Proximal: Proximal LPA Distal : Ductus Arteriosus



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NORMAL LEFT AORTIC ARCH

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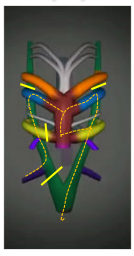
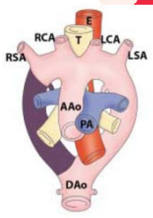
Normal Left Arch



Edwards Model



A Turkvatan KJR 10-176

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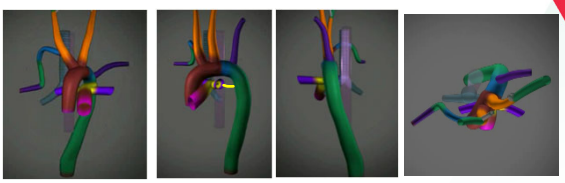
Normal Left Arch



Relationship of the Great Vessels to the Trachea and Esophagus
Head and Neck Vessel Anatomy

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Fetal 3 Vessel View and 3 Vessel Tracheal View

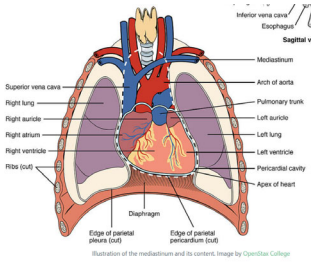

Illustration of the mediastinum and its content. Image by OpenStax College

- 3VV and 3VT outlines the mediastinal anatomy.
- Outlines the course of the great vessels
  - relationship to each other
  - relationship to airway and esophagus.
- Cranial continuation outlines the head and neck vessel anatomy

Excellent for Defining Variations and Abnormalities of the Great Vessels

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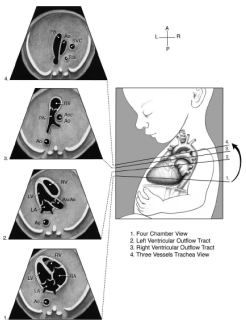
Fetal 3VV/3VT views



3VV and 3VT view are obtained by scanning cranial or cephalad from outflow tract view

4 Chamber
LVOT
RVOT
3VV
3VT
Head and neck vessels.

AIUM 2013 Guidelines

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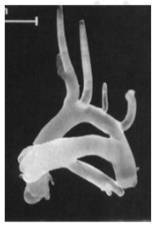
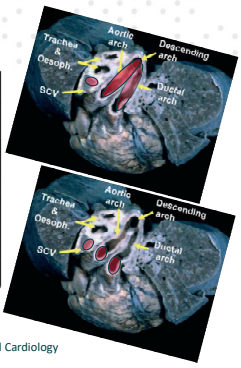
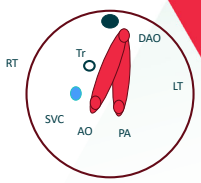
Fetal ductus arteriosus is a large structure





Rt SVC/AO/PV/LT



L Allen: Textbook of Fetal Cardiology

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Areas of Focus

- **SITUS (Right/Left)**
- # of Vessels
- Relative sizes
- **Formation of the "V" at D.Ao**
  - Aortic Arch courses Center to Left
  - D Arch Relatively Straight
  - TIP of the " V" watershed zone
- Direction of flow
- **Relation to the Trachea and each other**
- No arterial structure crossing in front of spine /behind the trachea

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VARIANT ARCH ANATOMY

LEFT Aortic ARCH
- Variations in origin of head and neck vessels
- Aberrant RSCA
- Aberrant innominate artery (IA)
- Isolation of ARSCA/IA
- Cervical aorta

RIGHT Aortic Arch
- Mirror image branching /Variations in head and neck vessels
- Aberrant LSCA
- Aberrant IA
- Isolation LSCA/IA
- Cervical Ao
- **Variations in PDA anatomy**
  - Left from IA
  - Left Dao
  - Rt PDA

Double Aortic Arch
Circumflex Right Arch
Circumflex Left Arch
LPA SLING

Abnormalities in Caliber
- Hypoplasia
- Interrupted Aortic arch
- Coarctation of Aorta

May be seen with any of the other defects

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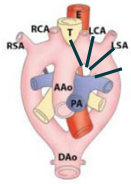
Right Aortic Arch



VARIATION IN head and neck vessels: Interruption at one level along the Left Arch System
- Mirror Image
- Aberrant left Subclavian Artery
- Aberrant Innominate Artery

ISOLATION of a Vessel: Interruption at 2 levels in developing left arch system
- Isolation of LSCA
- Isolation of Innominate Artery

Variations in Origin of Ductus Arteriosus

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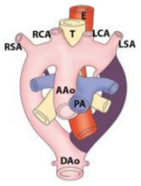
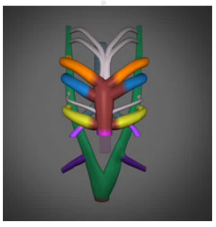
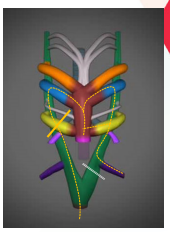
Right Aortic Arch/ Left PDA from Innominate Artery

MIRROR IMAGE BRANCHING







Or, one can have a right PDA (mirror image) with this anatomy

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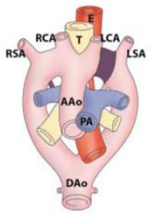
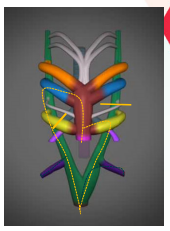
Rt Aortic Arch/ Lt PDA/ ALSCA

VASCULAR RING





Regression Between LSA and 6th Arch : have a left PDA to Desc Aorta with Mirror Image branching a Vascular ring

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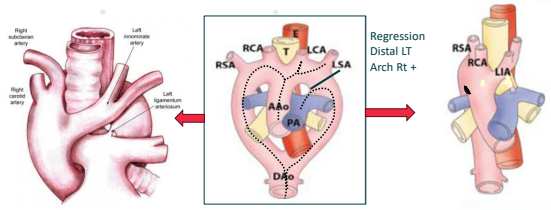
RIGHT ARCH Variations



Regression Distal LT Arch Rt +

Regression Between LSA and 6th Arch : + left PDA to Desc Aorta with Mirror Image branching (Vascular ring)

RAA + Mirror Image Branching and RIGHT PDA to Ds AO. NOT a VASCULAR RING

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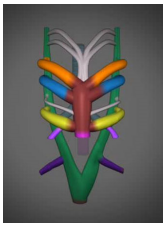
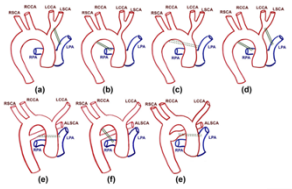
RIGHT AORTIC ARCH: Variations in Ductus Arteriosus





DM Sobh Clinical Radiology 74:2019: 732

Implications in fetal diagnosis

Management of cono-truncal lesions

Some have Vascular ring substrate/ others do not

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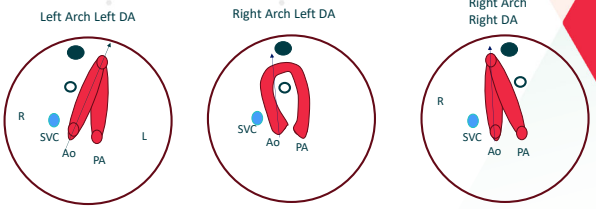
Fetal Diagnosis: Right Aortic Arch 3VV



- Relation of Aortic Arch and Ductal Arch to the Trachea
- Orientation of the aorta

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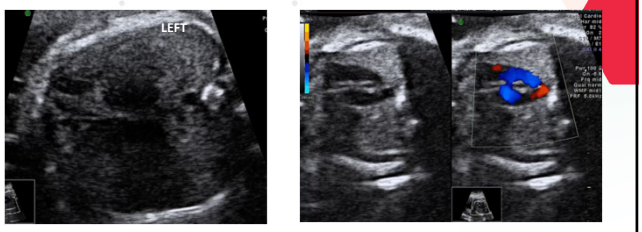
Right Aortic Arch Left Ductus Arteriosus



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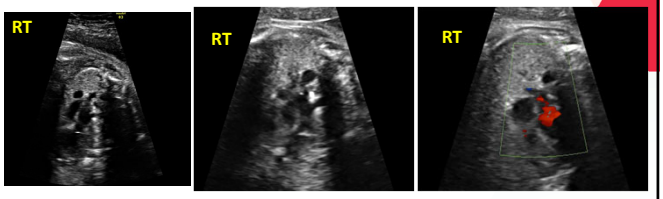
Abnormal 3VV



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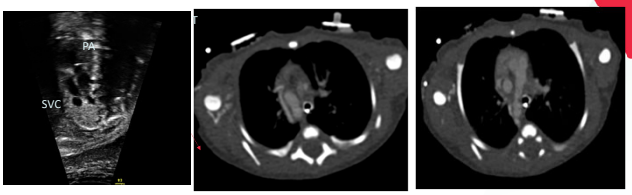
CT Right Arch Right PDA



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Situs Inversus (I,D,D) Right Arch (CH)

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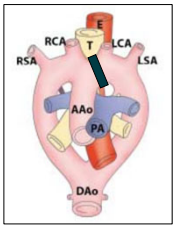
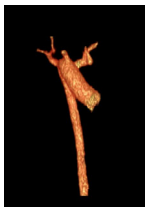
Right Aortic Arch Aberrant Innominate Artery



Regression of Left Horn Aortic Sac



NEWBORN With Tetralogy of Fallot and CHARGE SYNDROME

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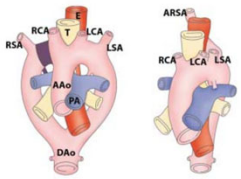
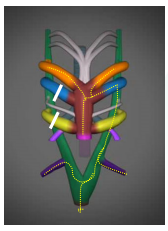
Left Arch Aberrant Right Subclavian Artery



Regression of the right 4th Pharyngeal arch artery



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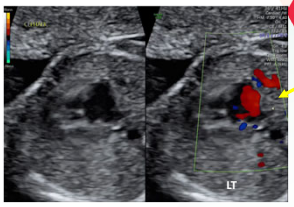
Fetal Echo : ASCA

1. Retro-esophageal course of a vessel
2. Vessel crossing behind the trachea and in front of the spine.
3. May occur in a right or left arch
4. ALSCA: constitute part of a vascular ring in setting of a left DA
5. ARSCA: Usually Benign; changes Aneuploidy risk assessment if other abnormalities are noted.

Left Arch ARSCA



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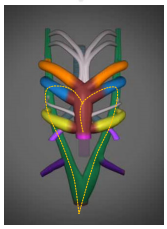
Double arch



1. Right Dominant/ Co Dominant/ Left Dominant
2. Look for Atretic segments
3. Symmetric head and neck vessels
4. More likely to have symptoms especially if fully patent
5. Isolation or with other cardiac defects

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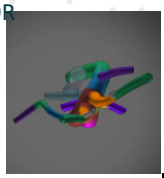
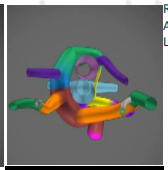
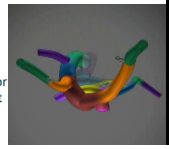
SUPERIOR VIEW

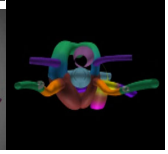
LEFT ARCH/ left PDA



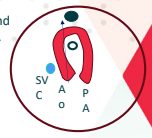


RT ARCH: Mirror Image with Left PDA/ RT PDA

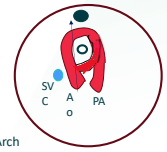




RT ARCH: ARSCA and LEFT PDA



Double Aortic Arch



VASCULAR RINGS

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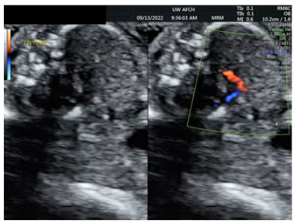
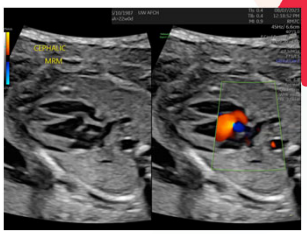
Double Arch



Rt Arch ALSCA



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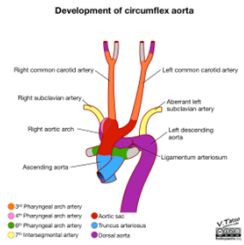
Circumflex Aortic Arch (RT or LT)



RADIOPEDIA

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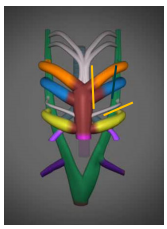
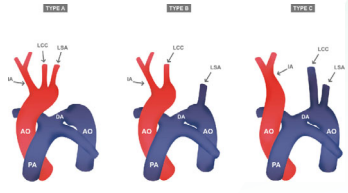
Interrupted Aortic Arch





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Coarctation of the Aorta

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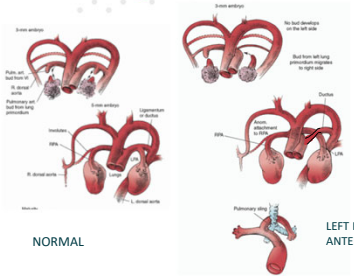
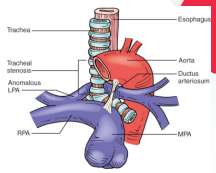
Left PA Sling



NORMAL



LEFT PA BETWEEN TRACHEA and ESOPHAGOUS
ANTERIOR INDENTATION On Barium Swallow

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Summary

- Many Variations in the Aortic Arch/ Head and Neck Vessel Anatomy
- Many are benign, some pathologic and highly symptomatic while others less so
- Understanding the development enhances recognition of variations/ pathology
- Systematic approach to fetal echocardiograms especially the 3VV and 3VT allows for accurate diagnosis in fetal life of most important lesions

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References

- The advantages of naming rather than numbering the arteries of the pharyngeal arches: Robert Anderson et al. Cardiology in the Young , Volume 33 , Issue 11 , November 2023 , pp. 2139 – 2147
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- Aortic Arch Variants and Anomalies: Embryology, Imaging Findings, and Clinical Considerations: Sang Bin Bae et al: J#C$8(/5<$9'#3 $+/4+n#_[__#0':r'[¤a'p_'^i_c_n

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Reading list:

- Larsen's human embryology
- Basics of cardiac development: Ped Research vol 57, no 2 2005
- Development of the human heart Am J Med Genet.2020;184C:7–22 https://onlinelibrary.wiley.com/doi/epdf/10.1002/ajmg.c.31778
- Moorman AFM, Webb S, Brown NA, et al. The development of the heart: (1) Formation of the cardiac chambers and arterial trunks. Heart 2003;89:806–14.
- Robert H Anderson, Sandra Webb, Nigel A Brown, Wouter Lamers, Antoon Moorman. DEVELOPMENT OF THE HEART: (2) SEPTATION OF THE ATRIUMS AND VENTRICLES Heart 2003;89:949–958
- https://www.karger.com/Article/Fulltext/501906; more physiologic transition review not addressed this talk

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