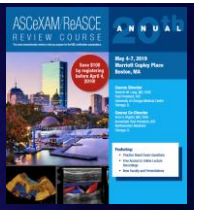
American Society of Echocardiography
20th Annual Review Course
Boston, MA



Congenital Heart Disease
An Approach for Simple & Complex Anomalies

Benjamin W. Eidem, M.D., FACC, FASE
Professor of Pediatrics & Medicine
Divisions of Pediatric Cardiology & Cardiovascular Diseases
Mayo Clinic

MAYO CLINIC

Nothing to Disclose

Congenital Heart Disease & Echocardiography

- **A Congenital Echocardiologist ……**
  - **Presumes that every vein, artery, chamber or valve is abnormal, until it is shown to be normal**
  - **Will use an organized method to examine the cardiovascular system (a step by step approach to avoid missing extra abnormalities)**

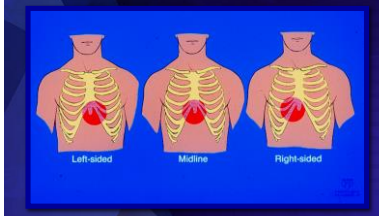
MAYO CLINIC

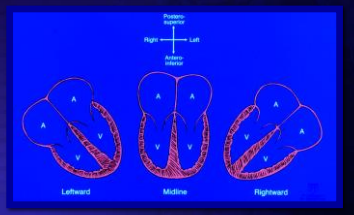
Segmental Approach to CHD
Definition

- A detailed, sequential description of CV anatomy including
  - Cardiac Position and Axis
    - Apex Orientation
  - Determination of Sidedness (“situs”)
    - Abdominal Organ and Atrial Spatial Arrangements
  - Anatomy and Function of each
    - CV Segment and
    - Connections between Segments

MAYO CLINIC

Segmental Approach to CHD
Position vs Axis





Cardiac Position
Where is the Heart?
Left, Right or Midline

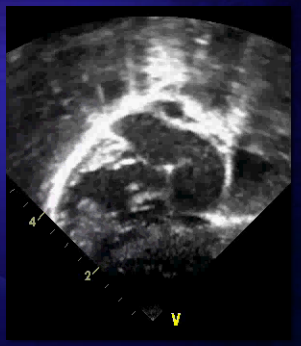
Cardiac Axis
How is the Heart Aligned?
Apex Left – Levocardia
Apex Right – Dextrocardia
Inferior / Midline - Mesocardia

MAYO CLINIC

Segmental Approach to CHD
Position vs Axis

Levocardia

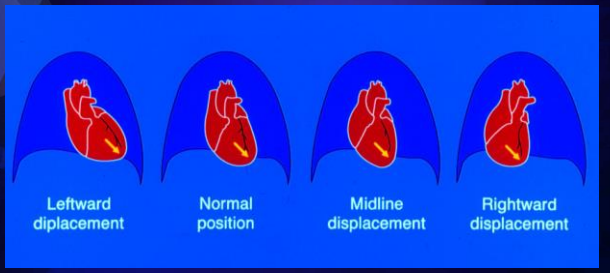
Dextrocardia



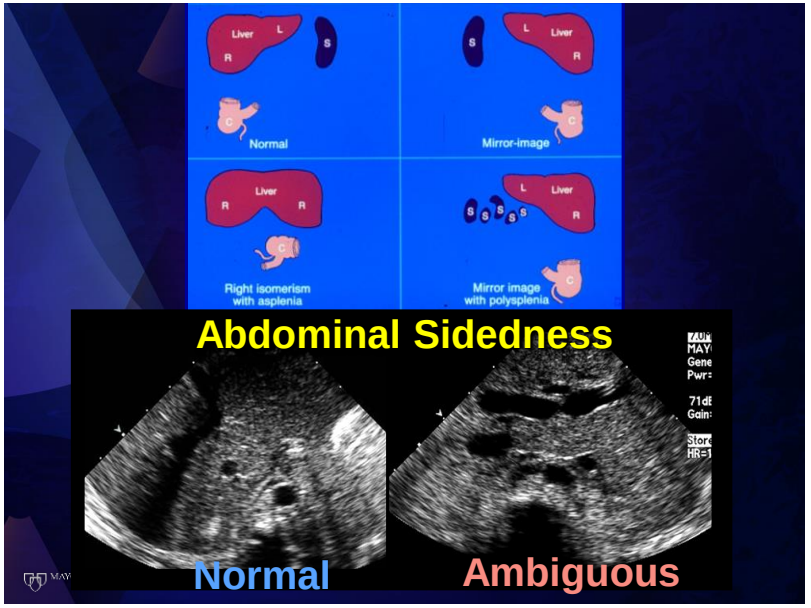
MAYO CLINIC

Why Distinguish Between *Position* and *Axis*?

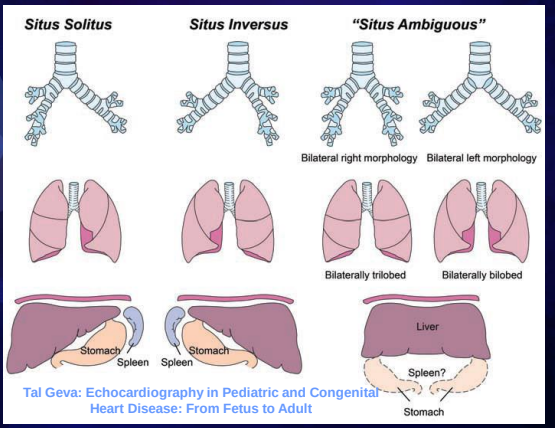
- Cardiac Position can change
- Axis is constant



MAYO CLINIC



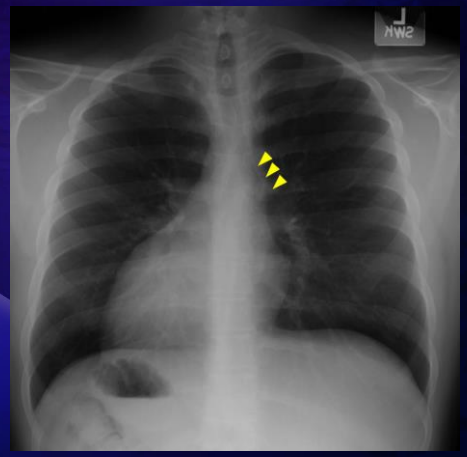
Segmental Approach to CHD
Visceral Anatomy



Tal Geva: Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult

Segmental Approach to CHD
Visceral Anatomy

Situs
Inversus
Totalis



MAYO CLINIC

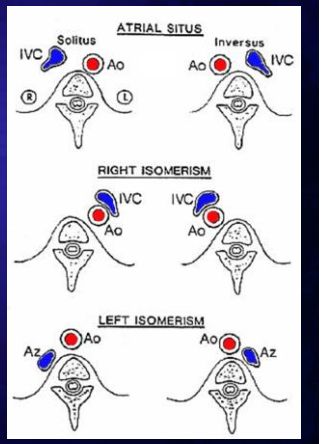
Segmental Approach to CHD
Visceral Anatomy

Situs
Ambiguous

MAYO CLINIC

Segmental Approach to CHD
Systemic Veins

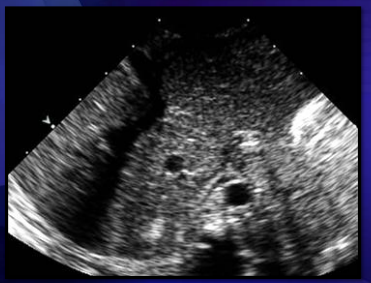
- Inferior vena cava
- Azygos venous system
- Hepatic veins
- Superior vena cava
- Coronary sinus



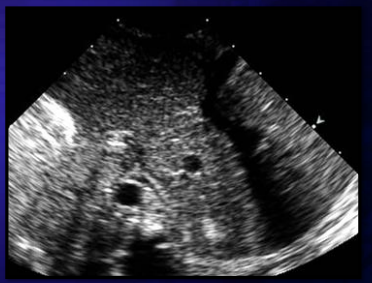
MAYO CLINIC

Segmental Approach to CHD
Systemic Veins

Situs Solitus



Situs Inversus



MAYO CLINIC

Segmental Approach to CHD
Systemic Veins

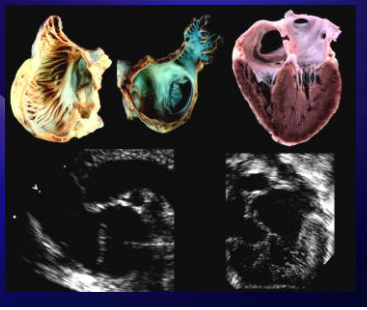
- IVC
  - Always drains into mRA
- Interrupted IVC
  - Azygos continuation to SVC

MAYO CLINIC

Echo Correlates of Atrial Morphology

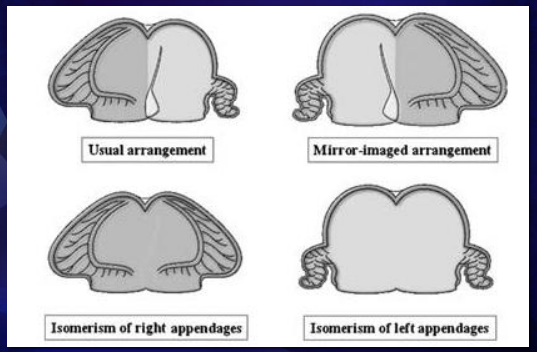
RA Findings
- Pectinate muscles
- Broad appendage
- Thick septal limbus
- Coronary sinus
- Supra-hepatic IVC

LA Findings
- Smooth walls
- Finger-like appendage
- Thin valve of the atrial septum



MAYO CLINIC

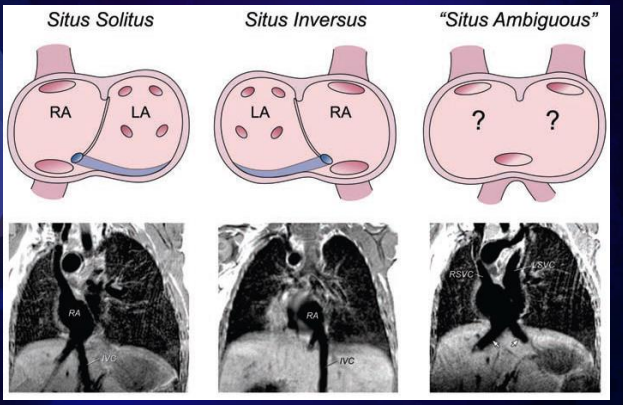
Segmental Approach to CHD
Atrial Anatomy



Jacobs J, Anderson R, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. Cardiology In The Young 2007;17(2):1-28

MAYO CLINIC

Segmental Approach to CHD
Atrial Anatomy



Tal Geva: Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult

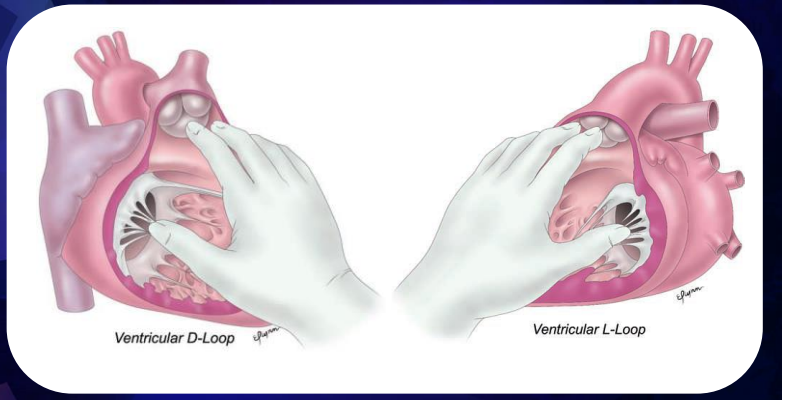
MAYO CLINIC

Segmental Approach to CHD
Ventricular Morphology

How do I tell the RV from the LV ???

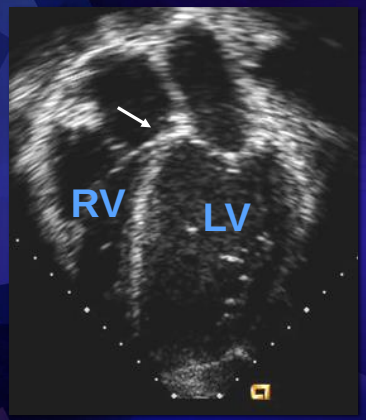
MAYO CLINIC

Segmental Approach to CHD
Ventricular Situs



Tal Geva: Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult

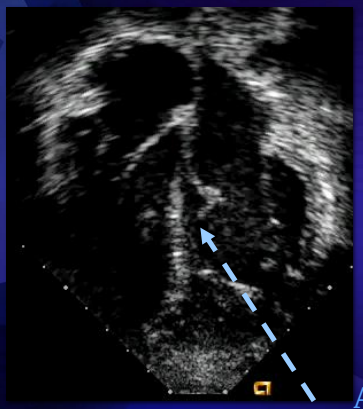
AV Connection, the Internal Crux & Ventricular Morphology



- AV Valve morphology is directly correlated with ventricular type
  - TV ⇒ RV
  - MV ⇒ LV
- Internal Cardiac Crux
  - Septal TV leaflet ***always*** inserts slightly apical to anterior MV leaflet

MAYO CLINIC

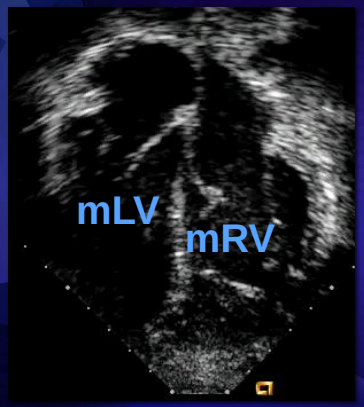
AV Connection, the Internal Crux and Ventricular Morphology



- AV Valve morphology is directly correlated with ventricular type
  - TV ⇒ RV
  - MV ⇒ LV
- Internal Cardiac Crux
  - Septal TV leaflet ***always*** inserts slightly apical to anterior MV leaflet

MAYO CLINIC

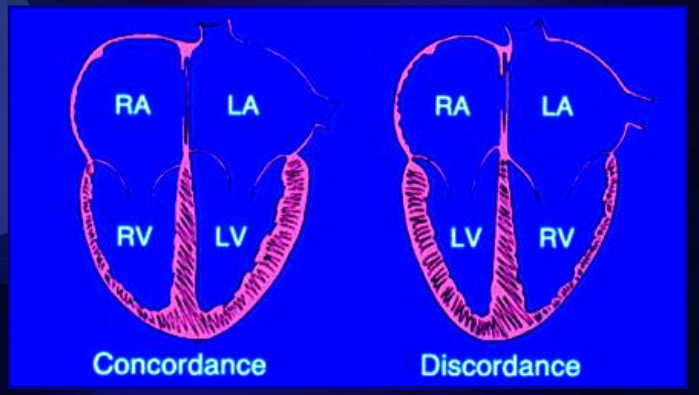
AV Connection, the Internal Crux and Ventricular Morphology



- AV Valve morphology is directly correlated with ventricular type
  - TV ⇒ RV
  - MV ⇒ LV
- Internal Cardiac Crux
  - Septal TV leaflet *always* inserts slightly apical to anterior MV leaflet

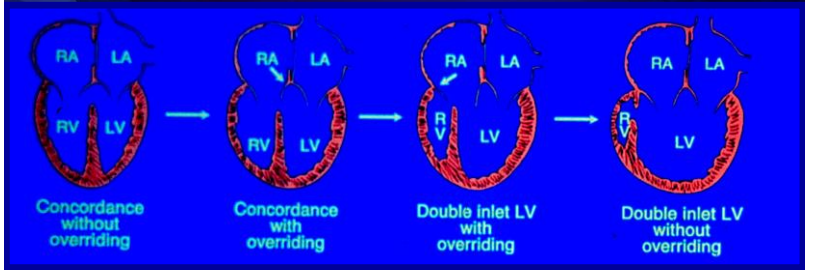
- Additional RV Markers
  - TV attachments to septum
  - Moderator Band
  - Multiple, small PMs
- Additional LV Markers
  - No MV septal attachments*
  - Smooth endocardium
  - Distinct, large PMs (usually just 2)

Types of AV Connection
Biventricular



MAYO CLINIC

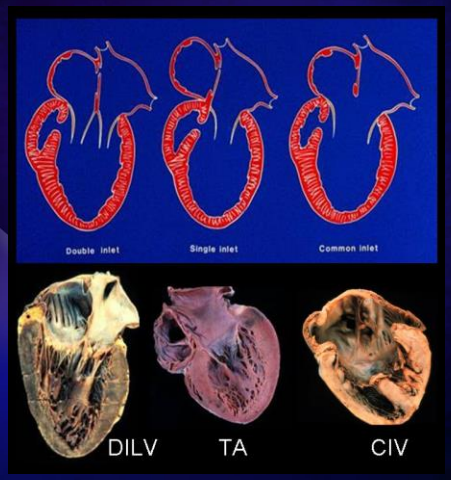
Impact of Annular Override on AV Connection



- Connection is determined by the annulus
  - The 50% rule
- Chordal Straddling does not impact connection

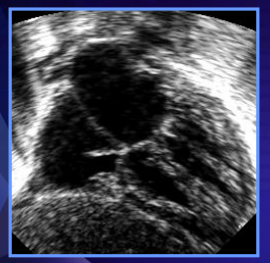
MAYO CLINIC

Types of AV Connection
Univentricular



MAYO CLINIC

Ventricular Morphology in Univentricular AV Connection



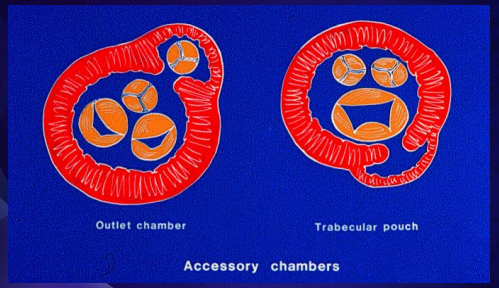
Tricuspid Atresia

HLHS

- Cannot use the internal crux
- Degree of myocardial trabeculation
- Papillary muscle anatomy
  - Septal attachments = TV / RV

MAYO CLINIC

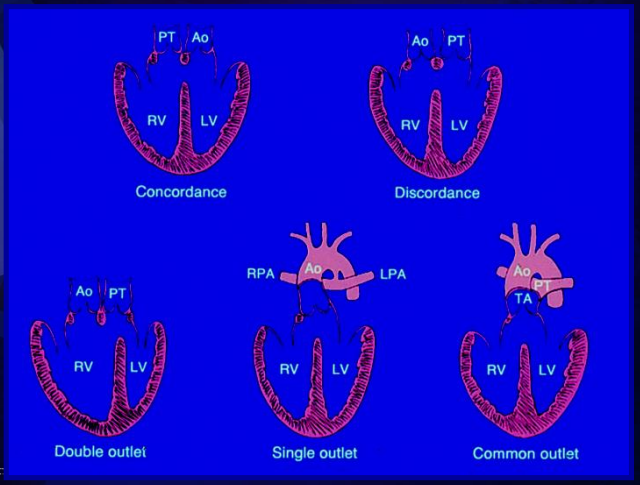
Ventricular Morphology in Univentricular AV Connection



- The ***position*** of the hypoplastic ventricular remnant is most the reliable indicator
  - LV's are Posterior
  - RV's are Anterior

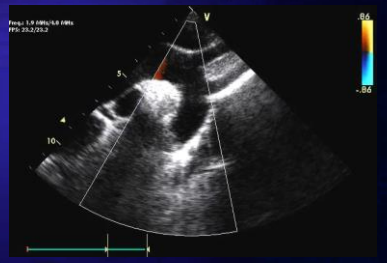
MAYO CLINIC

Ventricular to Arterial Connections



MAYO C

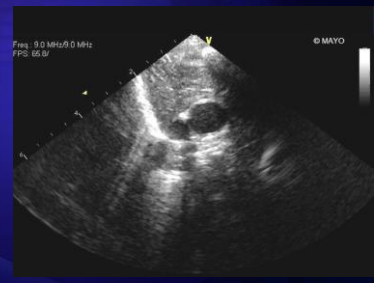
Aortic Arch Sidedness & Branching



Left Aortic Arch with Normal Brachiocephalic Branching

MAYO CLINIC

Aortic Arch Sidedness & Branching



Right Aortic Arch

MAYO CLINIC

Components of the Segmental Approach

CV Segments

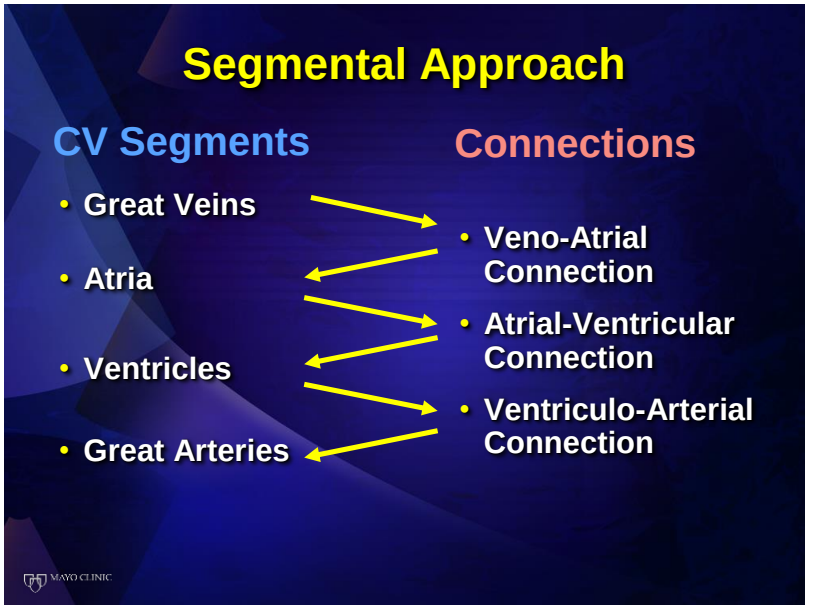
- Position, Anatomy, Size and Function of each component of the segment
- State of Septation between the “right” and “left” components of the segment

CV Connections

- Position, Anatomy, Size and Function of each connection between segments
- Relationship of each connector to the preceding and subsequent segments
  - Malalignment or Straddling?

MAYO CLINIC

Segmental Approach



MAYO CLINIC

Using the Segmental Approach to Evaluate Patients with CHD

CV Segments	Connections
• Great Veins	• Veno-Atrial Connection
• Atria	• Atrial-Ventricular Connection
• Ventricles	• Ventriculo-Arterial Connection
• Great Arteries	

= ⇑ Understanding of CHD

MAYO CLINIC