

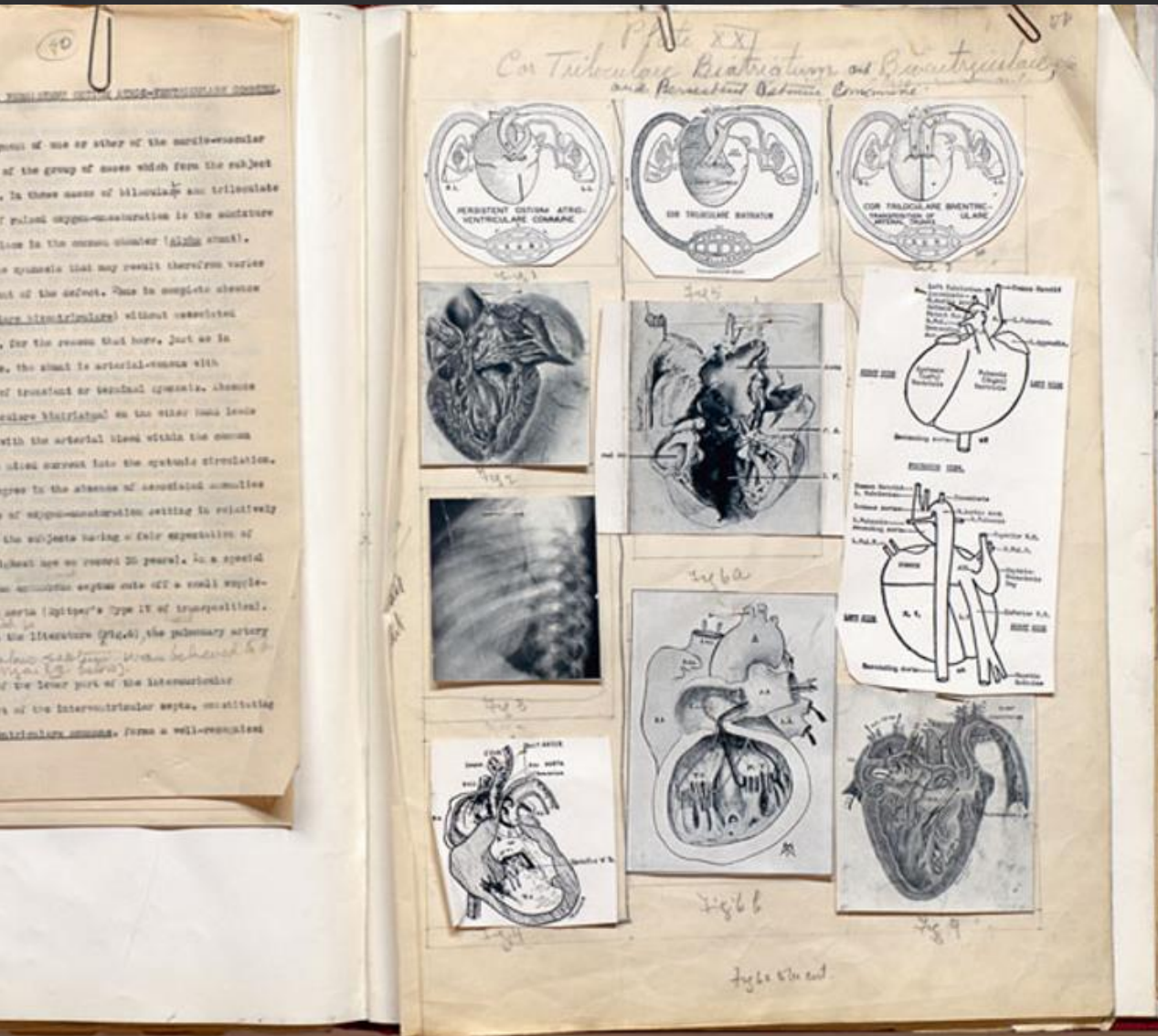
The 2nd Contemporary Morphology Course
Abnormalities of the Atrioventricular Junction
December 2019

Univentricular Atrioventricular Connection

Vitor Guerra

No disclosures

SickKids[®]

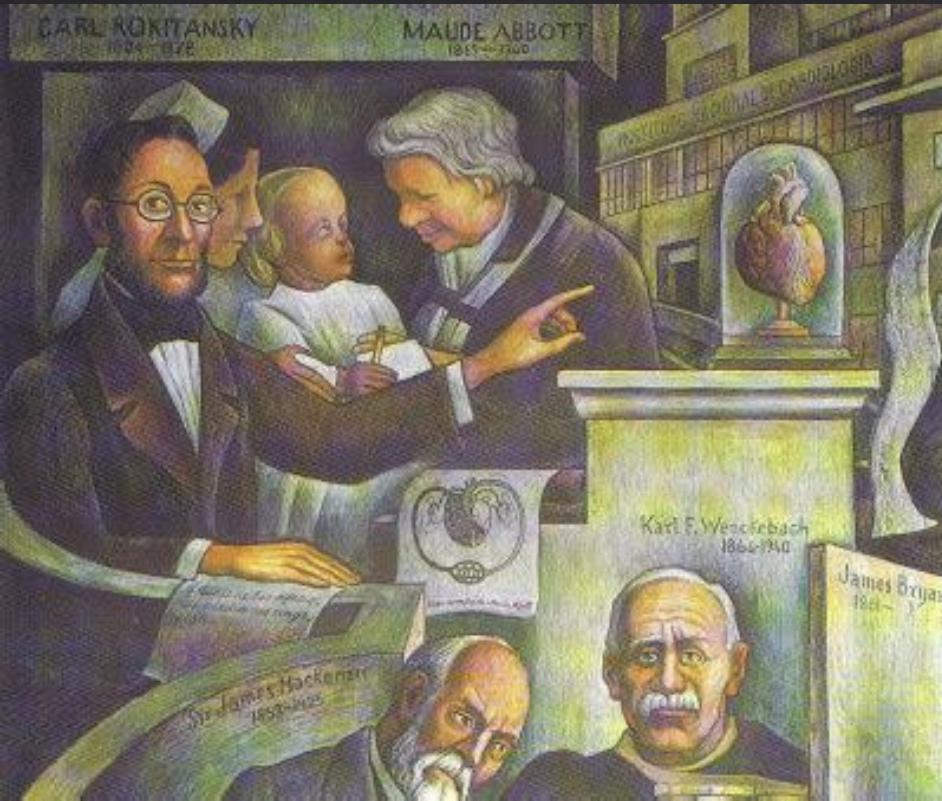


This is a photograph of a draft copy of Maude Abbott's Atlas

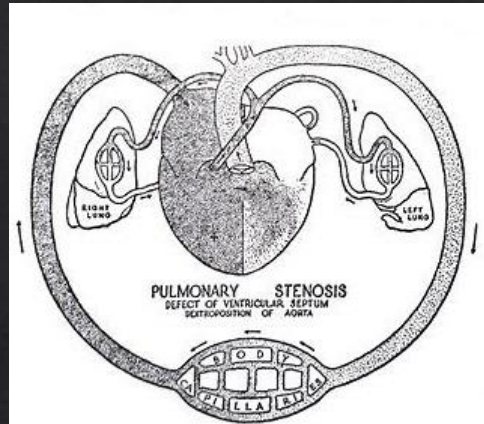
“The simplicity is the last step of wisdom”

(La simplicité est la dernière étape de la sagesse)

Kahlil Gibran – Syria (1883-1931)



Diego Rivera panel
Institute de Cardiologia do Mexico

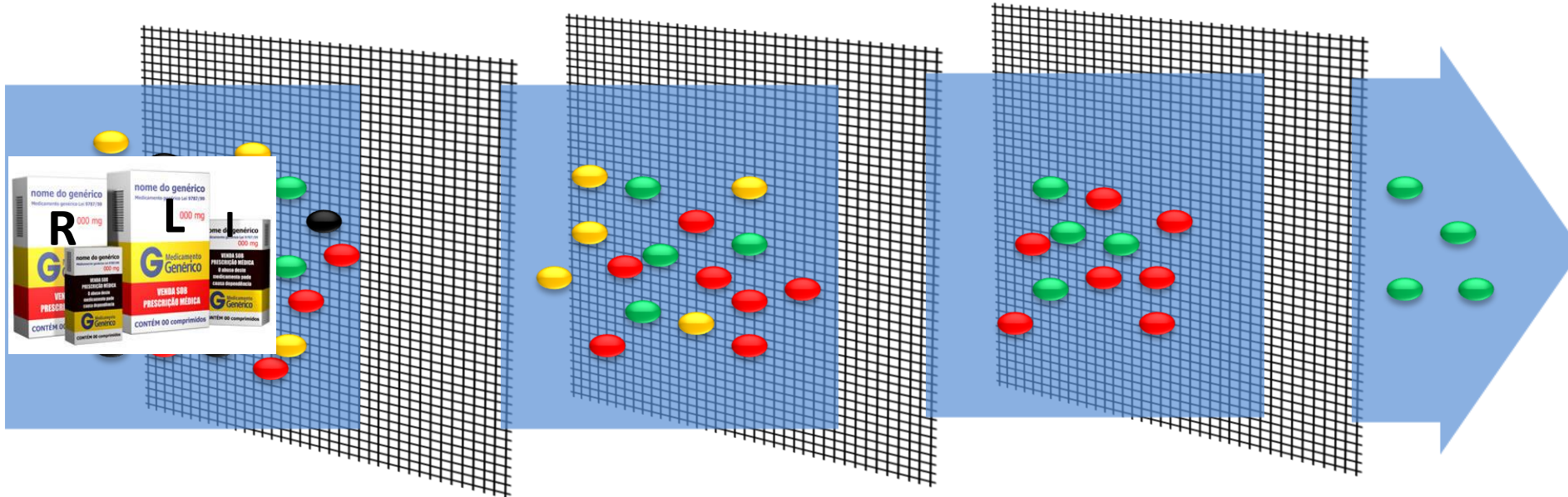


“Holmes Heart”



Maude Abbott

“Univentricular heart: are they generic?”



Sequential
Segmentar
analysis
International
Nomenclature

Anatomical
differences

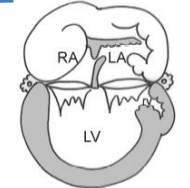
Predict
outcome

Intracardiac
lesions

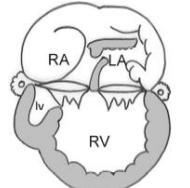
Atrioventricular
valves
Ventricular septal
defect
Sub-aortic outlet

Conduction
System

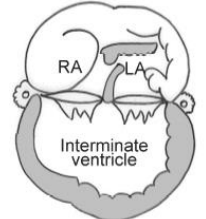
Coronary
Artery System



Double inlet left ventricle

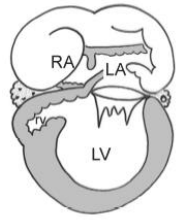


Double inlet right ventricle

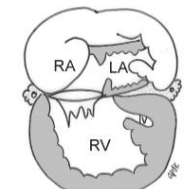


Interminate
ventricle

Double inlet indeterminate
or common ventricle



Absent right
atrioventricular
connection



Absent left
atrioventricular
connection

Who has helped understanding univentricular Atrioventricular connection?

Maria Victoria
de LaCruz



Rules for diagnosis of arterioventricular discordances
and spatial identification of ventricles
*Crossed great arteries and transposition of
the great arteries*

Maria V. de la Cruz, José R. Berrazueta, Manuel Arteaga, Fause Attie, and Jorge Soni
From the Department of Embryology, Instituto Nacional de Cardiología, México 7, D.F. México

Van Praagh



Anatomic Types of Single or Common
Ventricle in Man

Morphologic and Geometric Aspects of 60 Necropsied Cases*

RICHARD VAN PRAAGH, M.D.†, PATRICK A. ONGLEY, M.B., F.A.C.C. and HAROLD J. C. SWAN, M.B.

Rochester, Minnesota

Robert
Anderson



Connexions, relations, discordance, and distortions

Fergus J. Macartney, Elliot A. Shinebourne, and Robert H. Anderson¹

*From the Hospital for Sick Children, Great Ormond Street, London, and Brompton Hospital,
Fulham Road, London*

Who has helped understanding univentricular Atrioventricular connection?

Vera Aiello



Para o Sitor: este haço precisa deslizar. Always, Vera

Vera Aiello Siew Yen Ho Robert H. Anderson

Absence of one atrioventricular connection associated with straddling atrioventricular valve: distinction of a solitary from a common valve and further considerations on the diagnosis of ventricular topology

Unusual presentation of atrioventricular (AV) univentricular connection: Absent left AV connection with a dominant ventricle of left morphology. Anatomical and echocardiographic findings in four cases.

Vitor Coimbra Galvão MD (1), Maria Angélica Bentes MD PhD (2), Vera Diwanji Azeite MD PhD (3), Vera Aiello MD PhD (4), Robert H. Anderson MD PhD (5), Siew Yen Ho MD PhD (6), Vera Aiello MD PhD (7)

(1) Tabor Hospital For Children, Tabor University, School of Medicine - New Orleans, LA, US
 (2) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil
 (3) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil
 (4) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil
 (5) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil
 (6) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil
 (7) Heart Institute of University of São Paulo, School of Medicine - São Paulo, Brazil

Introduction:
 Despite of existing a systematized nomenclature for all types of univentricular AV connection, there are still some cases challenging pediatric cardiologists/echocardiographers. Usually, absence of one AV connection is associated with the presence of a rudimentary ventricular chamber at the same side as the absent connection. Very rarely, the rudimentary chamber is located at the contralateral side of the absent connection, with the dominant ventricle of unexpected morphology for the side of absent connection (see panel I, cartoon C). We sought to clarify this unusual feature by reviewing a series of four cases, including one fetal diagnosis, with this morphological arrangement.

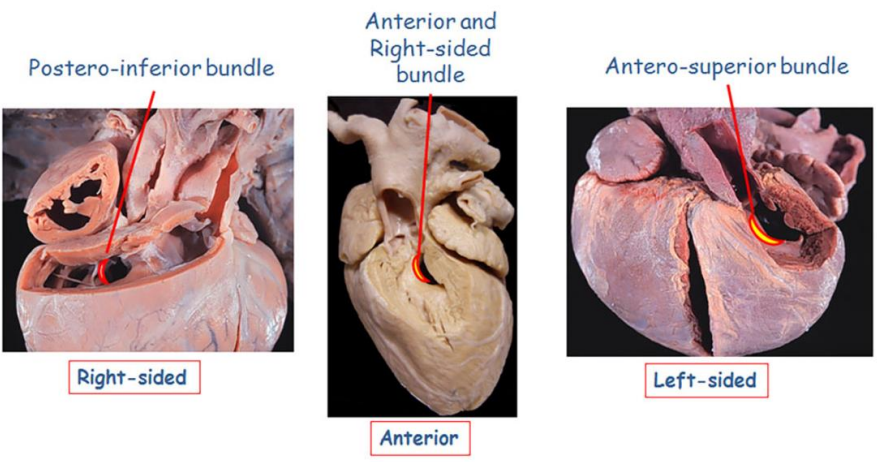
Phenotypes of absence of left AV connection

Material & Methods:
 From the hospital database/chart, data were obtained. Echocardiogram studies were reviewed and correlation with anatomical features from pathological study was done.
 Four cases were studied with absence of left AV connection with a dominant left ventricle and rudimentary right ventricular chamber located at the right side. Three specimens were evaluated from patients who died. The remaining patient has been followed after a palliative surgery.

Results:
 There was agreement between the initial echocardiogram report and pathological features in two cases. In the two remaining, the initial diagnosis was "Mixed atrial", including one fetal diagnosis.
 In all cases there was an early clinical presentation with hypoxia and low cardiac output. Right-catheter direct observation was directed in two patients. Two patients required aortic septostomy for a restrictive PFO. In the remaining patient there was a small PFO, but there was total anomalous pulmonary venous return to the coronary sinus.
 Main echocardiographic features:

| Case | AVD | AVB | Ventricular-septal connection | Location of rudimentary chamber | AVD/AVB | TR/VRD |
|------|--------|--------|-------------------------------|---------------------------------|---------|--------|
| 1 | PRO | Normal | connection | right | Yes | No |
| 2 | Normal | Normal | connection | right | No | No |
| 3 | Normal | Normal | connection | right | No | No |
| 4 | PRO | Normal | connection | right | Yes | No |

Conclusions:
 Echocardiographers must be aware about unusual forms of AV connection. The correct interpretation of absent AV connection by standard views needs a dynamic evaluation and the combination of different views to establish the correct spatial orientation of cardiac chambers and determination of ventricular morphology. Fetal diagnosis requires more views and careful interpretation as well. The sequential segmental analysis is fundamental to achieve the correct diagnosis, both by echocardiography and other morphological methods.



Position of the Rudimentary Right Ventricle

Understanding the Morphology of the Specialized Conduction Tissues in Congenitally Malformed Hearts

Who has helped understanding univentricular Atrioventricular connection?

Shi-Joon Yoo

“The Morphologist of era Modern Era”


Further Morphological Observations on Hearts With Twisted Atrioventricular Connections (Criss-Cross Hearts)

Jeong-Wook Seo, MD, Shi-Joon Yoo, MD,* Siew Yen Ho, PhD, MRCPATH, Heung Jae Lee, MD,[†] and Robert H. Anderson, MD, FRCPath

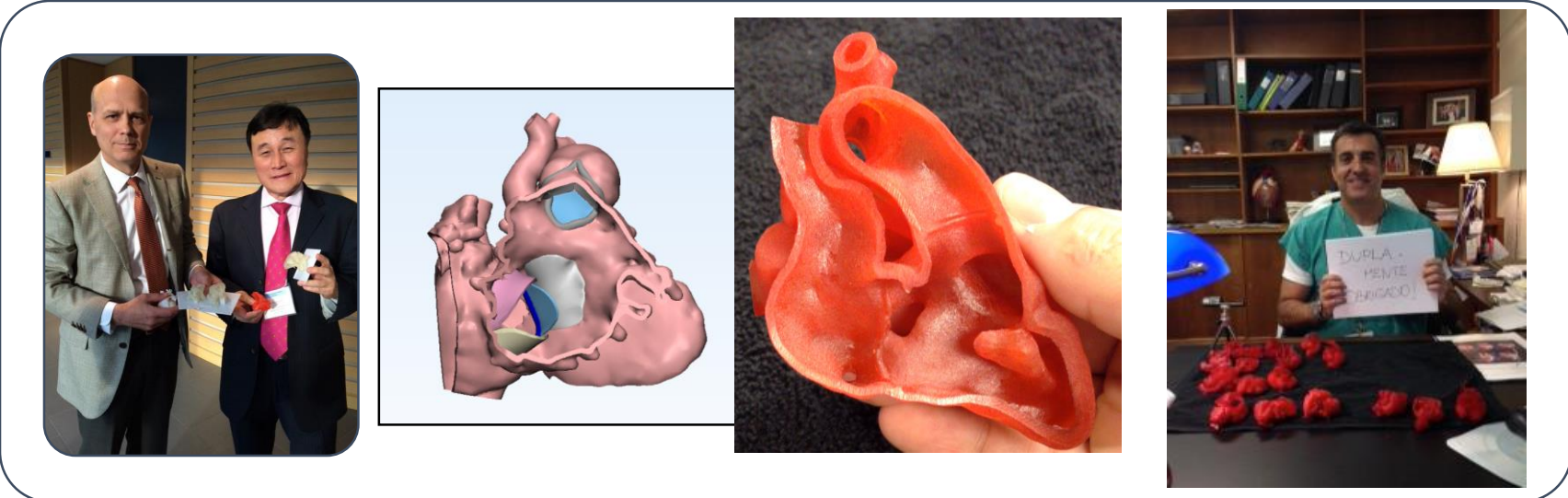

From the Department of Paediatrics, The National Heart and Lung Institute, London, United Kingdom, the [†]Department of Diagnostic Radiology, University of Ulsan-Asan Medical Center, Seoul, and the ^{}Department of Pediatrics, Sejong Heart Institute, Pucheonshi, Korea*

Prenatal diagnosis of topsy-turvy heart

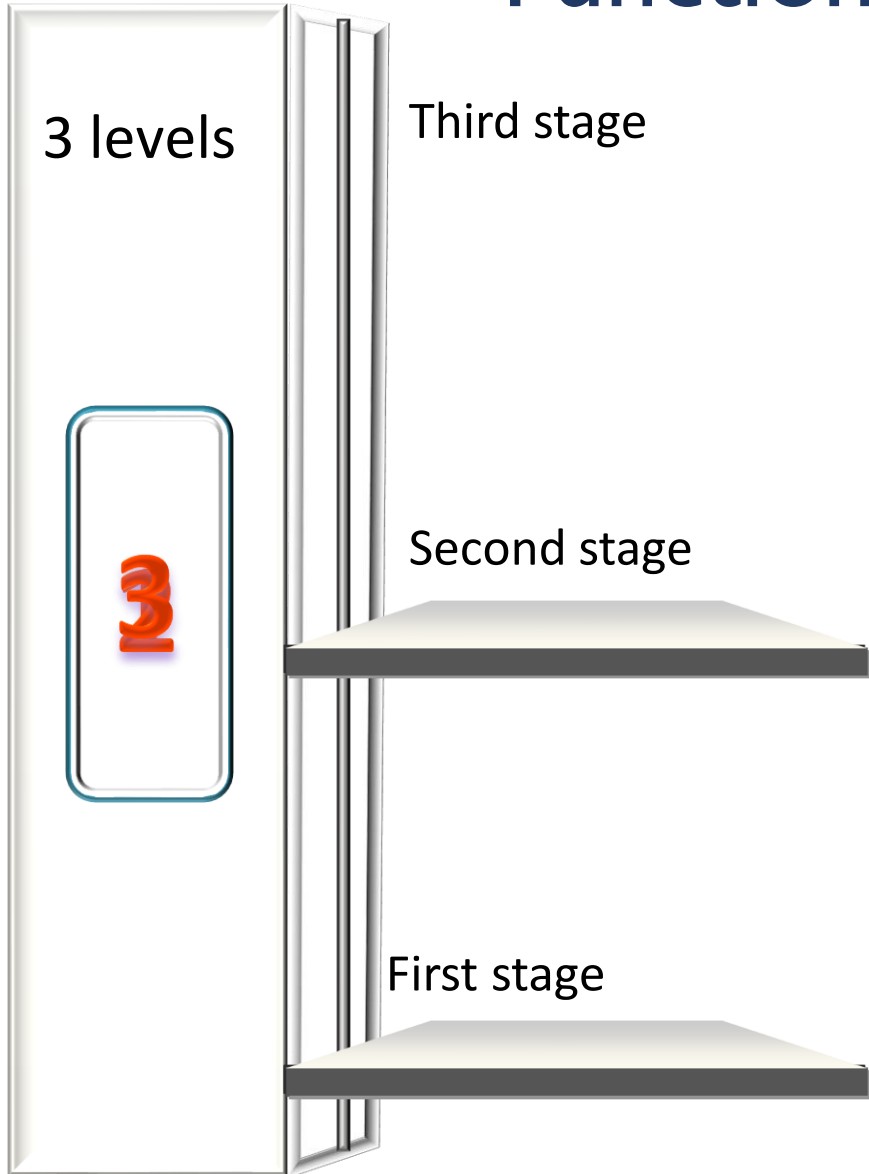
Edgar Jaeggi,¹ David Chitayat,² Fraser Golding,¹ Peter Kim,³ Shi-Joon Yoo^{1,4}

3D printing in medicine of congenital heart diseases 

Shi-Joon Yoo^{1,2*}, Omar Thabit^{1,2}, Eul Kyung Kim⁴, Haruki Ide², Deane Yim², Anreea Dragulescu², Mike Seed^{1,2}, Lars Grosse-Wortmann^{1,2} and Glen van Arsdell³



“Functionally” Univentricular hearts



3

IVC to PA (Fontan)

2

SVC to PA (Glenn)

1

PDA stent
Systemic Pulm shunt
PA Band
DKS and BT shunt
Norwood + BTS /Sano

The entire ventricular mass is assigned to the systemic circulation

Systemic venous return (pulmonary circulation) drains passively to the lungs):

- ✓ Pulmonary pressure
- ✓ Ventricular function
- ✓ Atrioventricular valves
- ✓ “Ventricular septal defects (‘outlet’)
- ✓ Sub-aortic obstruction
- ✓ Electrical stability

Goals of Echocardiogram (any other imaging modality in CHD) for Heart “Functionally” Univentricular:

Sequential Segmentar Analysis & Details of all morphologic aspects

Phase 1



Phase 2



Phase 3



Phase 4



Phase 5



Situs and
Venous
Connection
Atrial Shunt

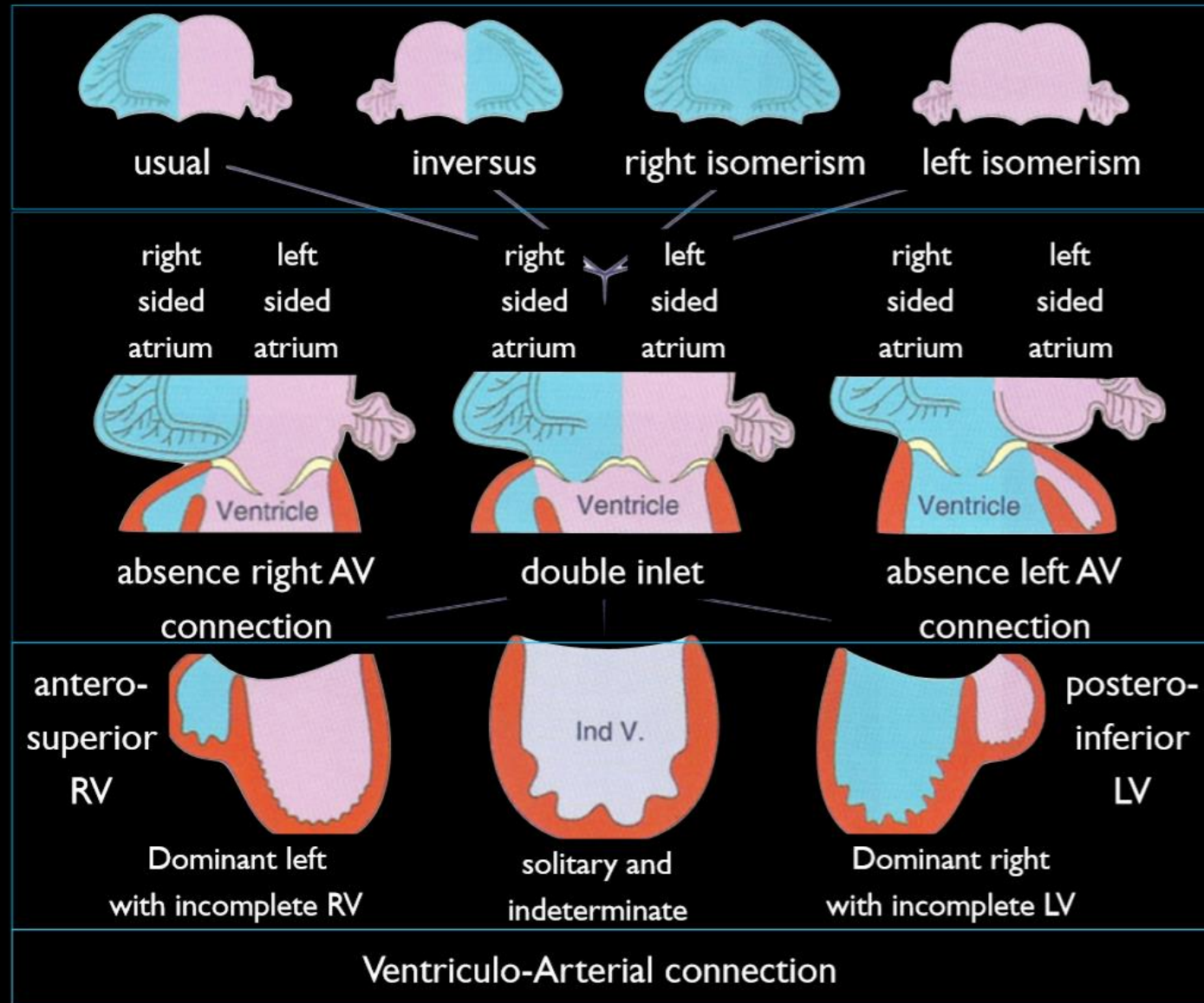
Atrioventricular
Connection

Morphology &
Function of
Atrioventricular
valves

Morphology &
ventricular
function
Ventricular
Shunt

Ventricular –
Arterial
Connection
Associated
lesions

Sequential segmentar analysis: Univentricular atrioventricular connection

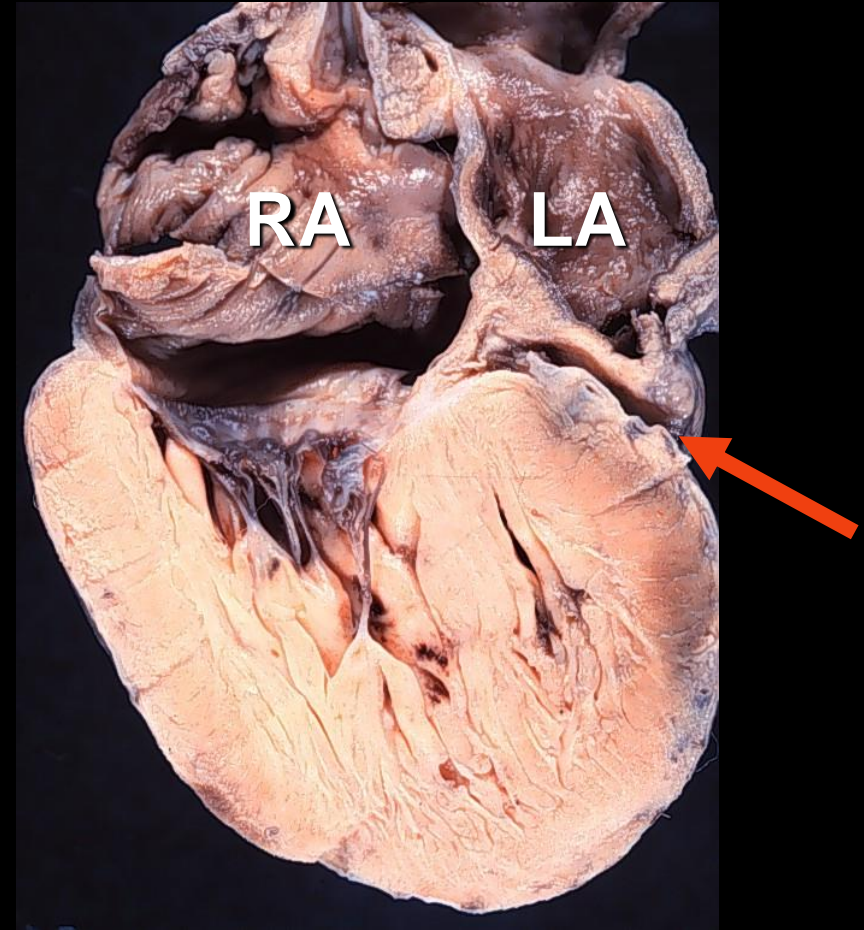
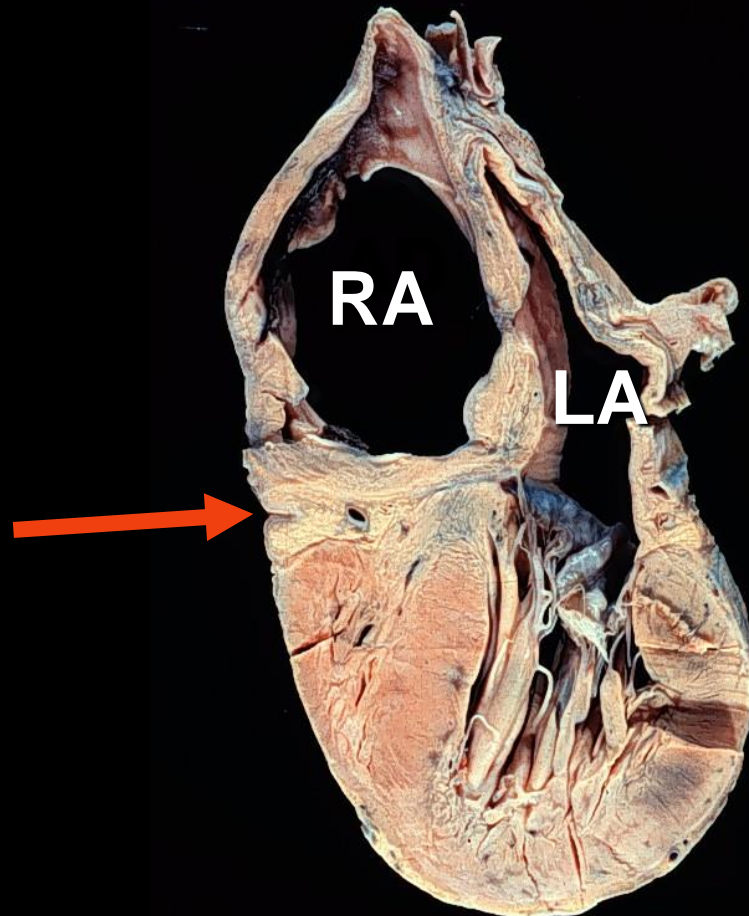


Phenotypes of Univentricular AV connection

Absence of AV connection

Right

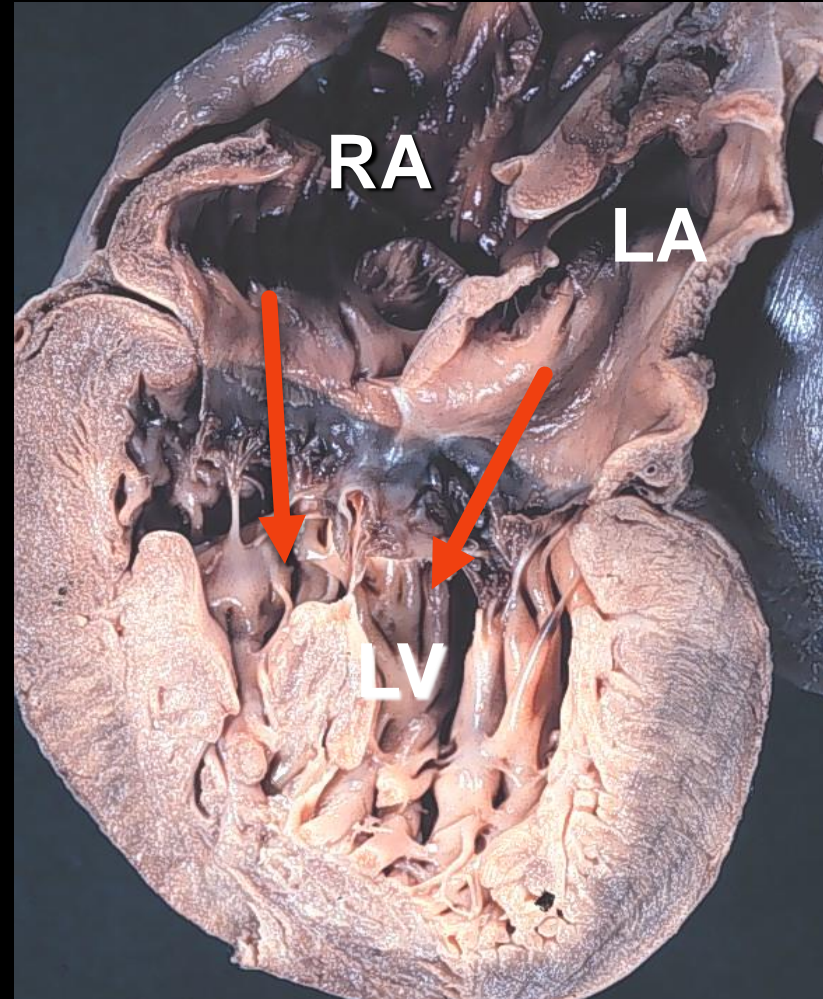
Left



Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

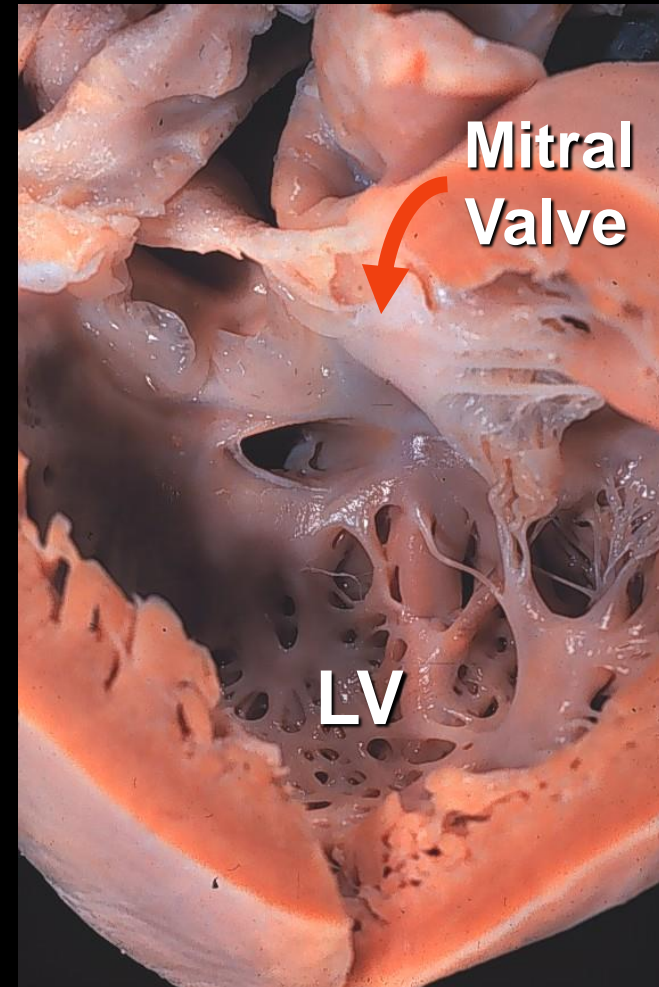
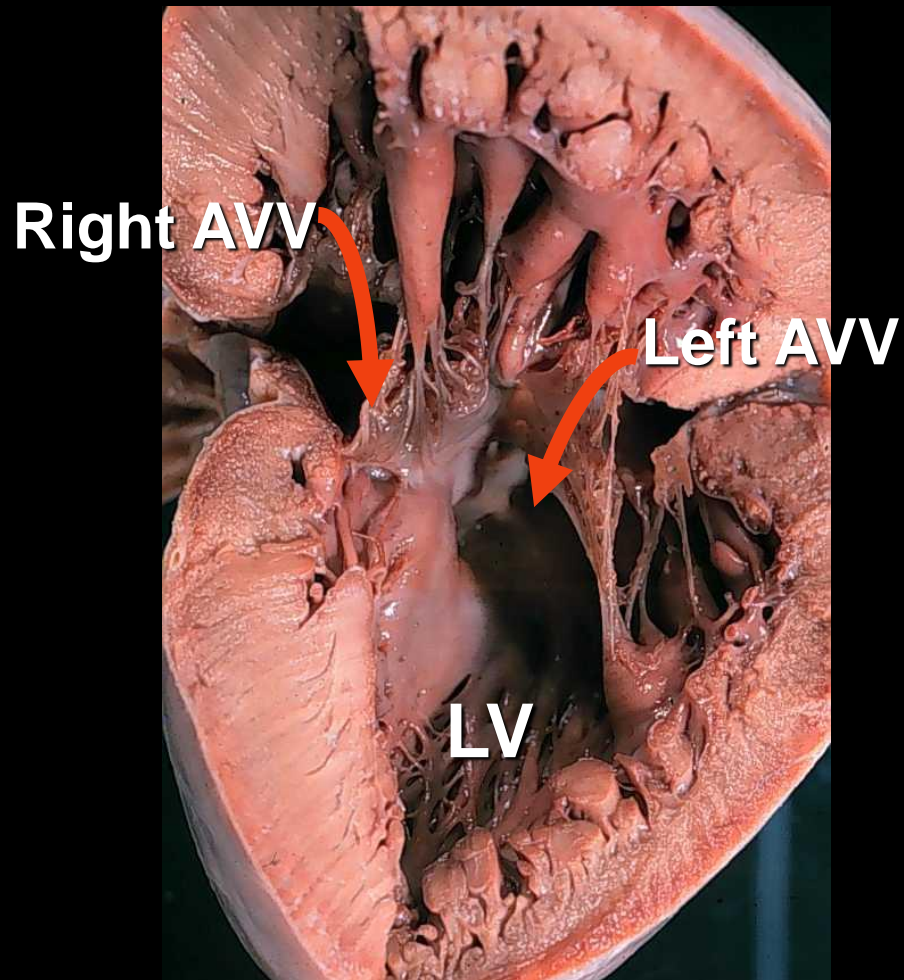
Phenotypes of Univentricular AV connection

Double Inlet



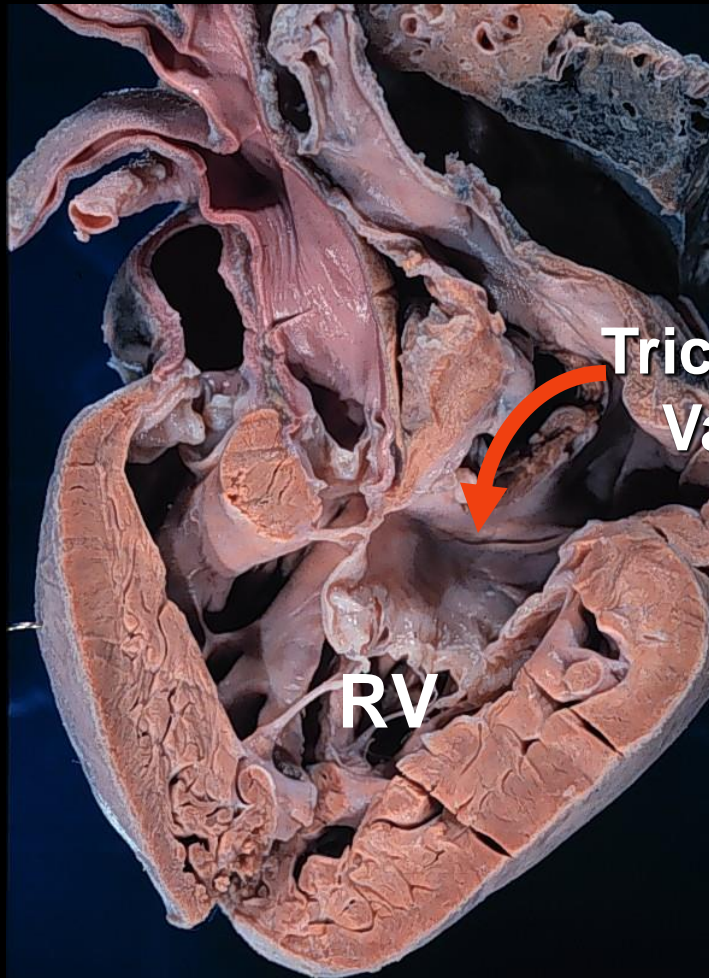
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Morphology of Main Ventricle



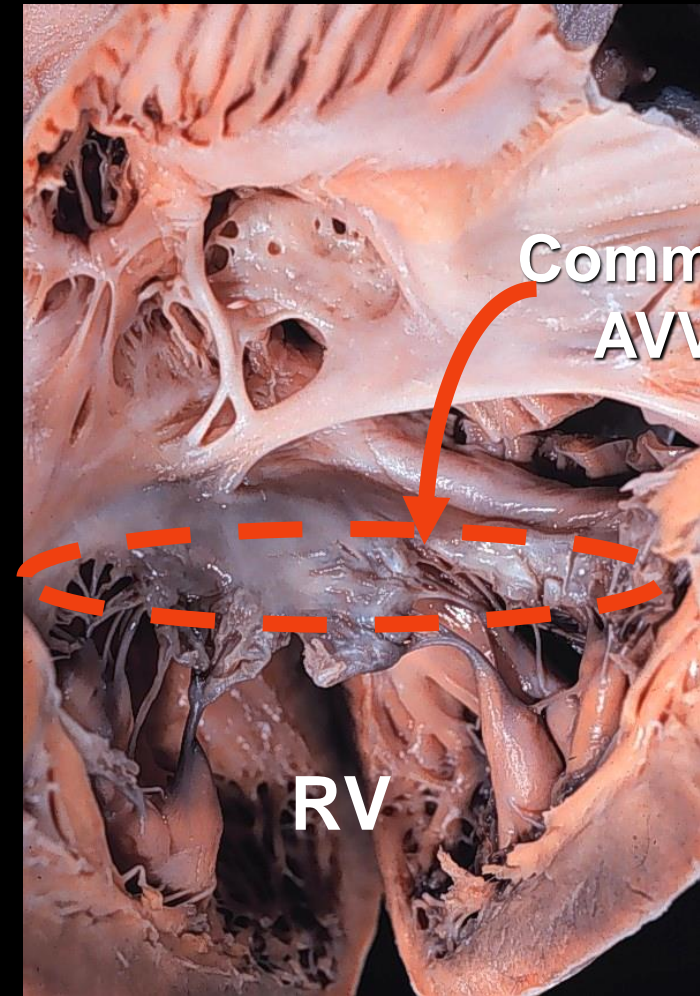
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Morphology of Main Ventricle



Tricuspid
Valve

RV

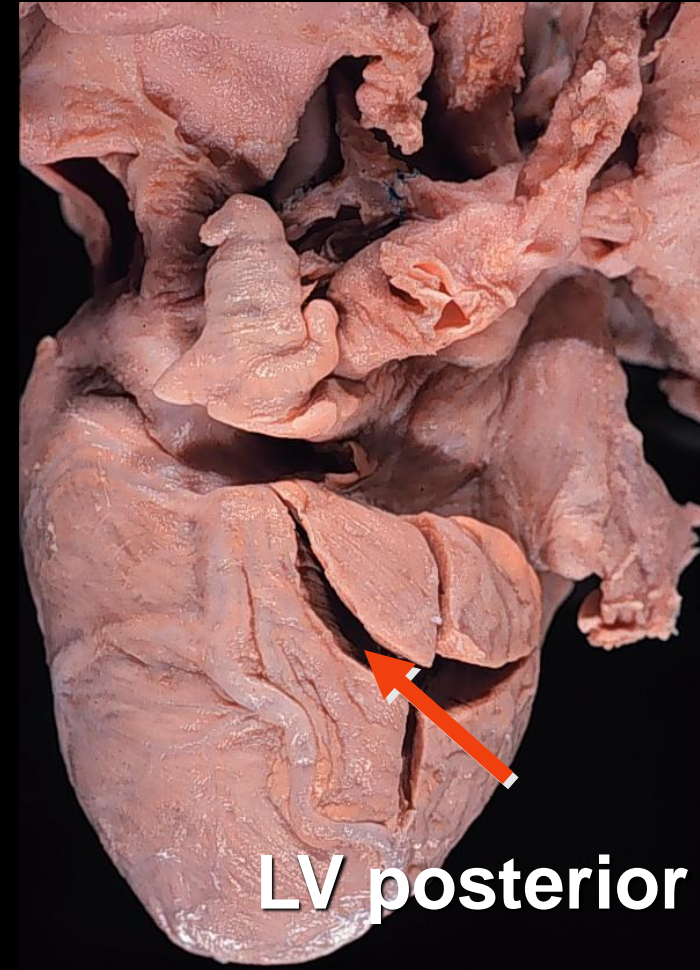
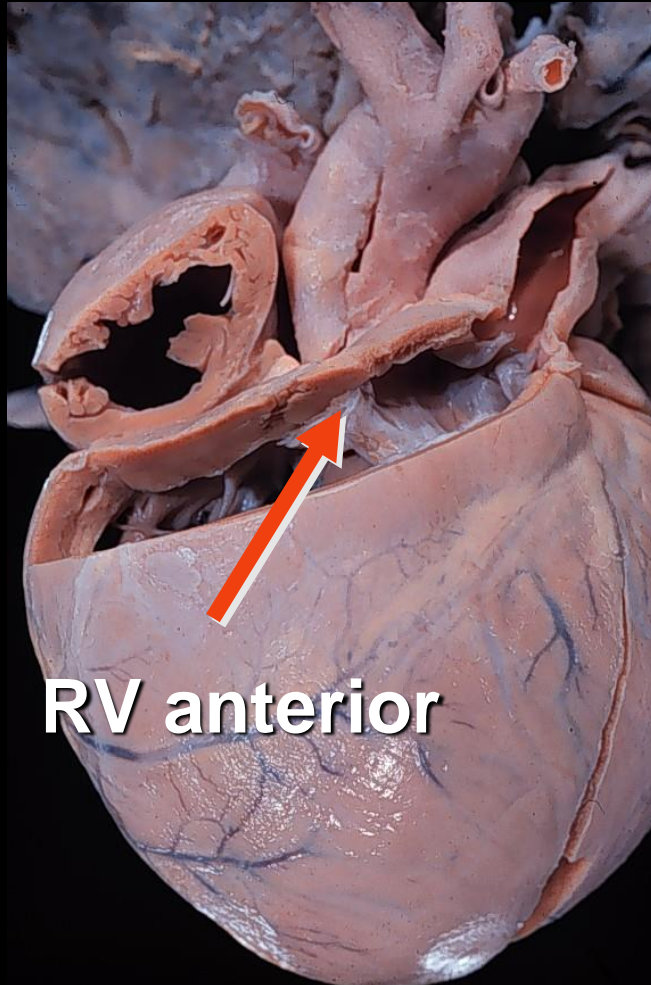


Common
AVV

RV

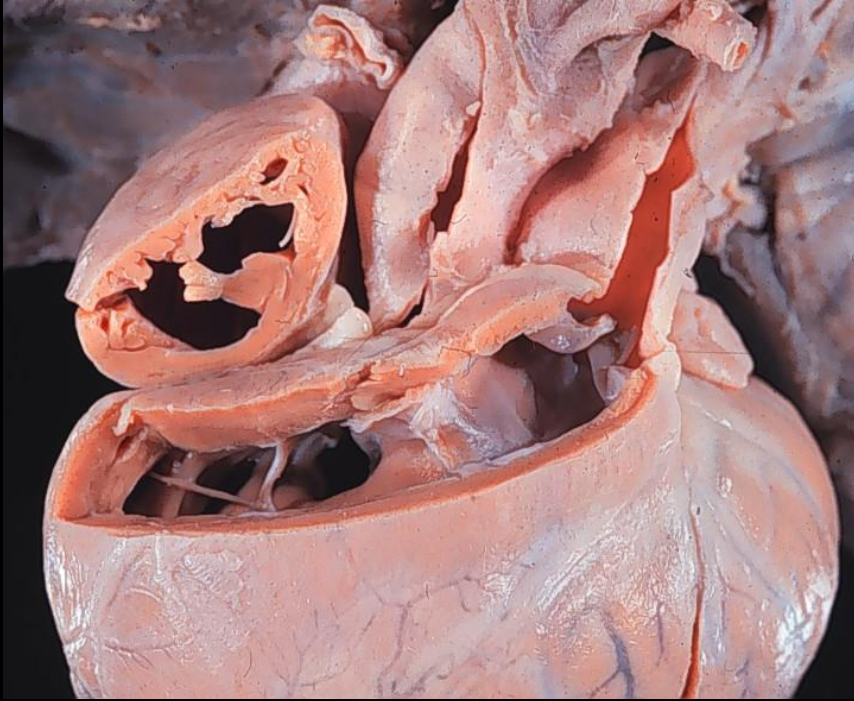
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Morphology & Location of the rudimentary Ventricle

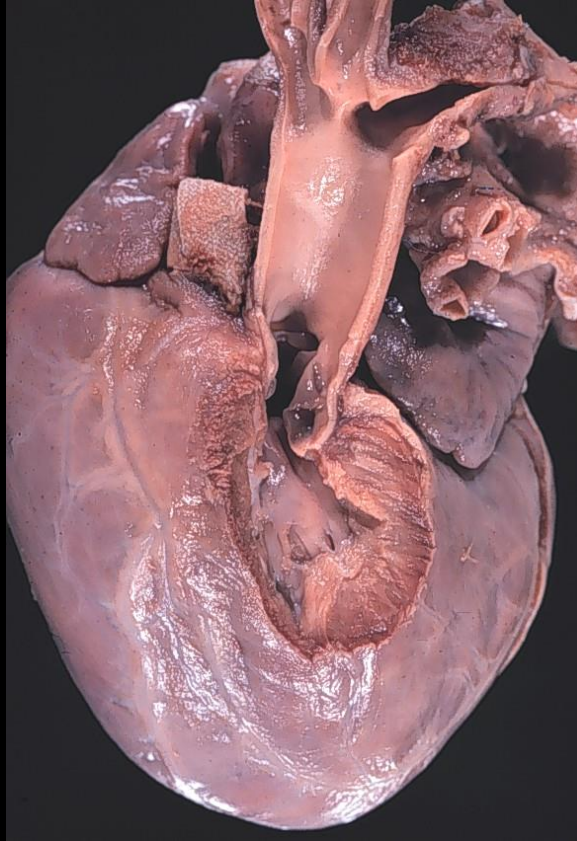


Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Morphology & Location of the rudimentary Right Ventricle



Anterior & Right



Anterior & Central

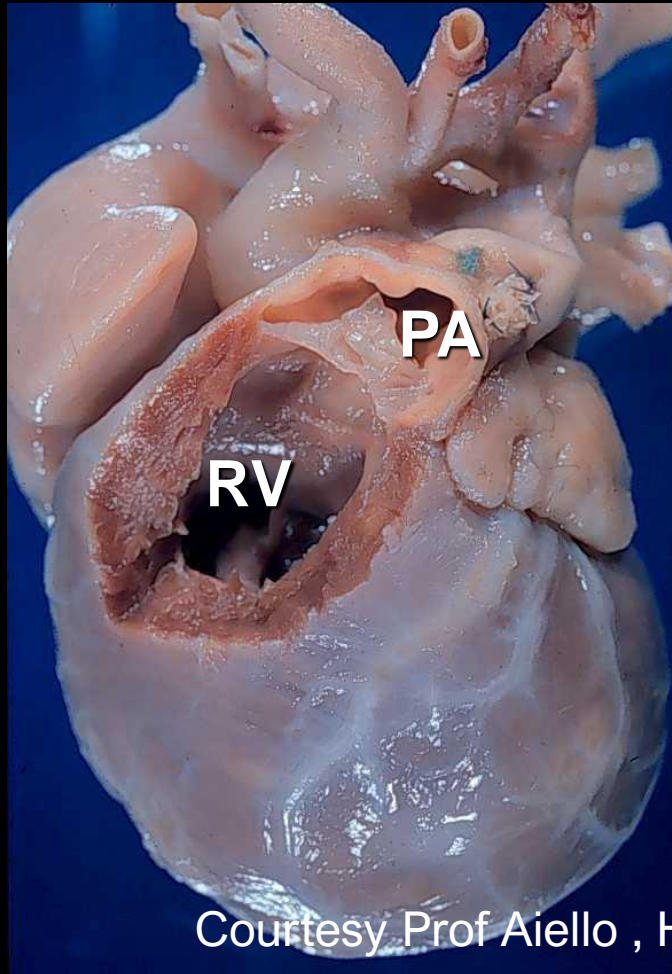


Anterior & Left

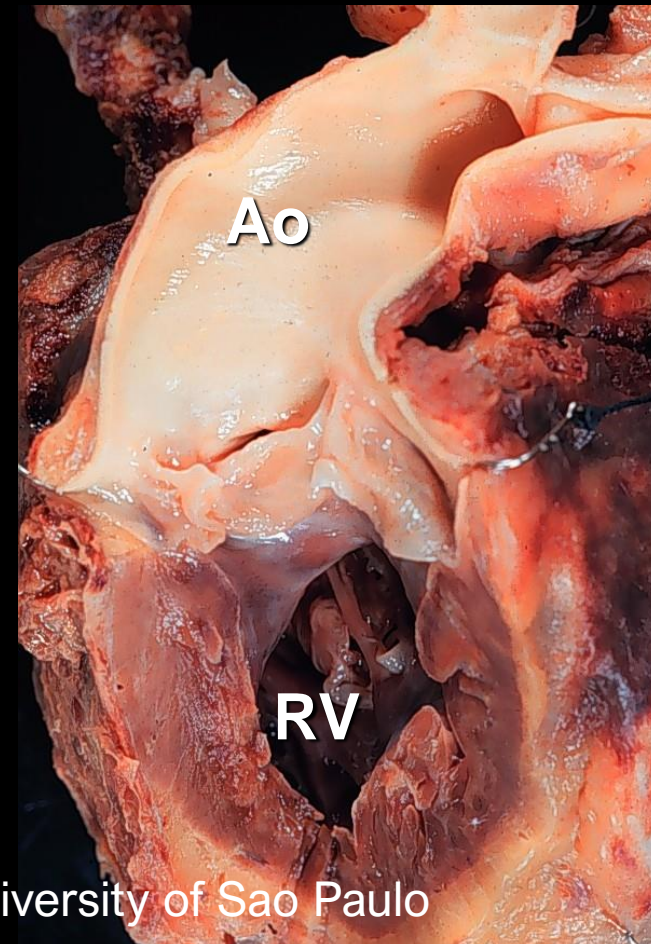
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Ventricular – Arterial Connection

Concordant

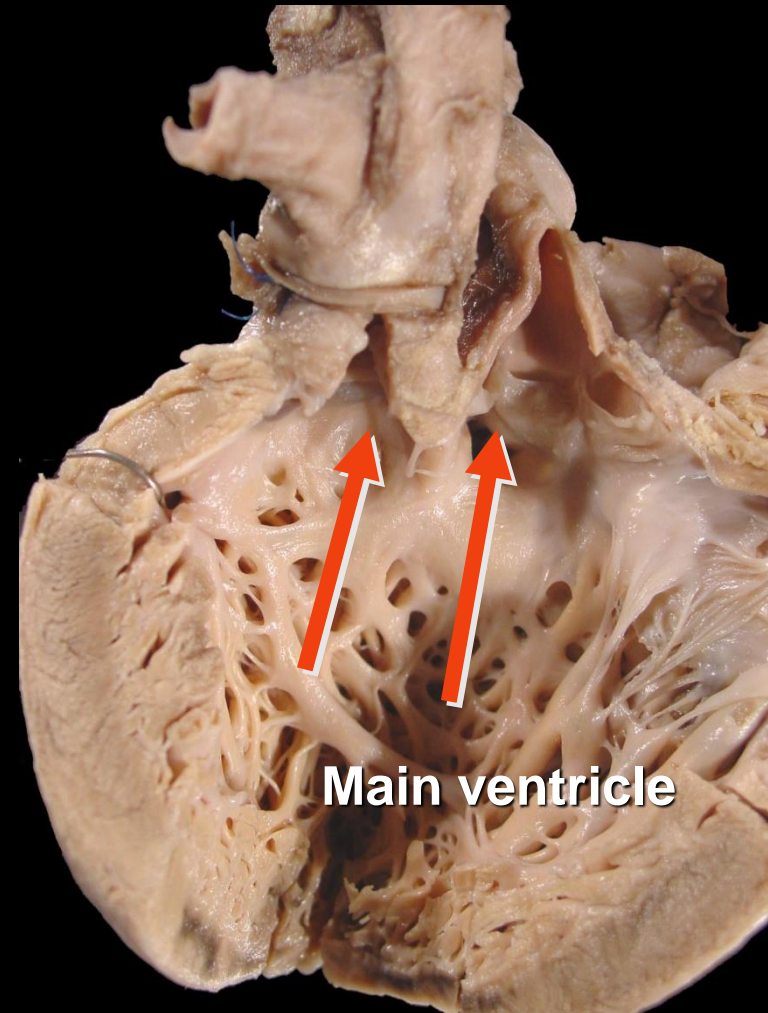
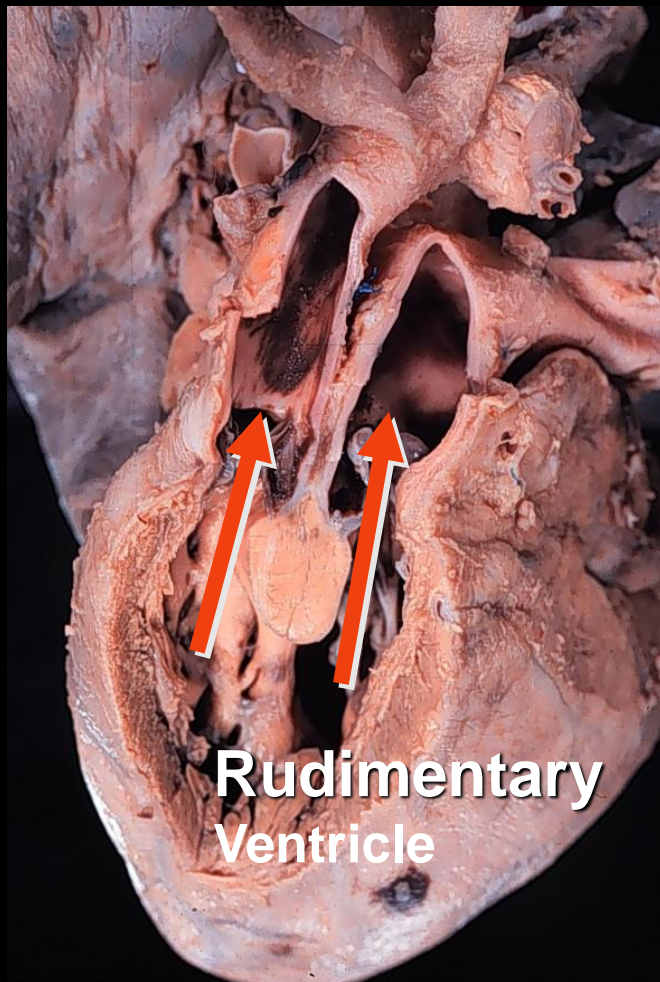


Discordant



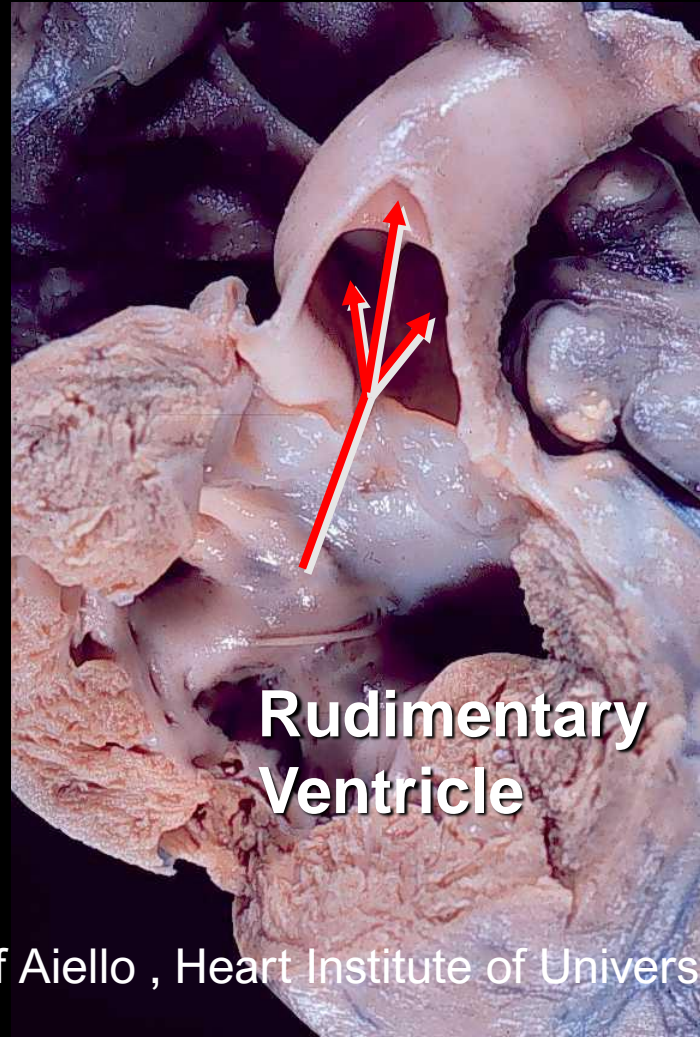
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Ventricular – Arterial Connection: Double Outlet



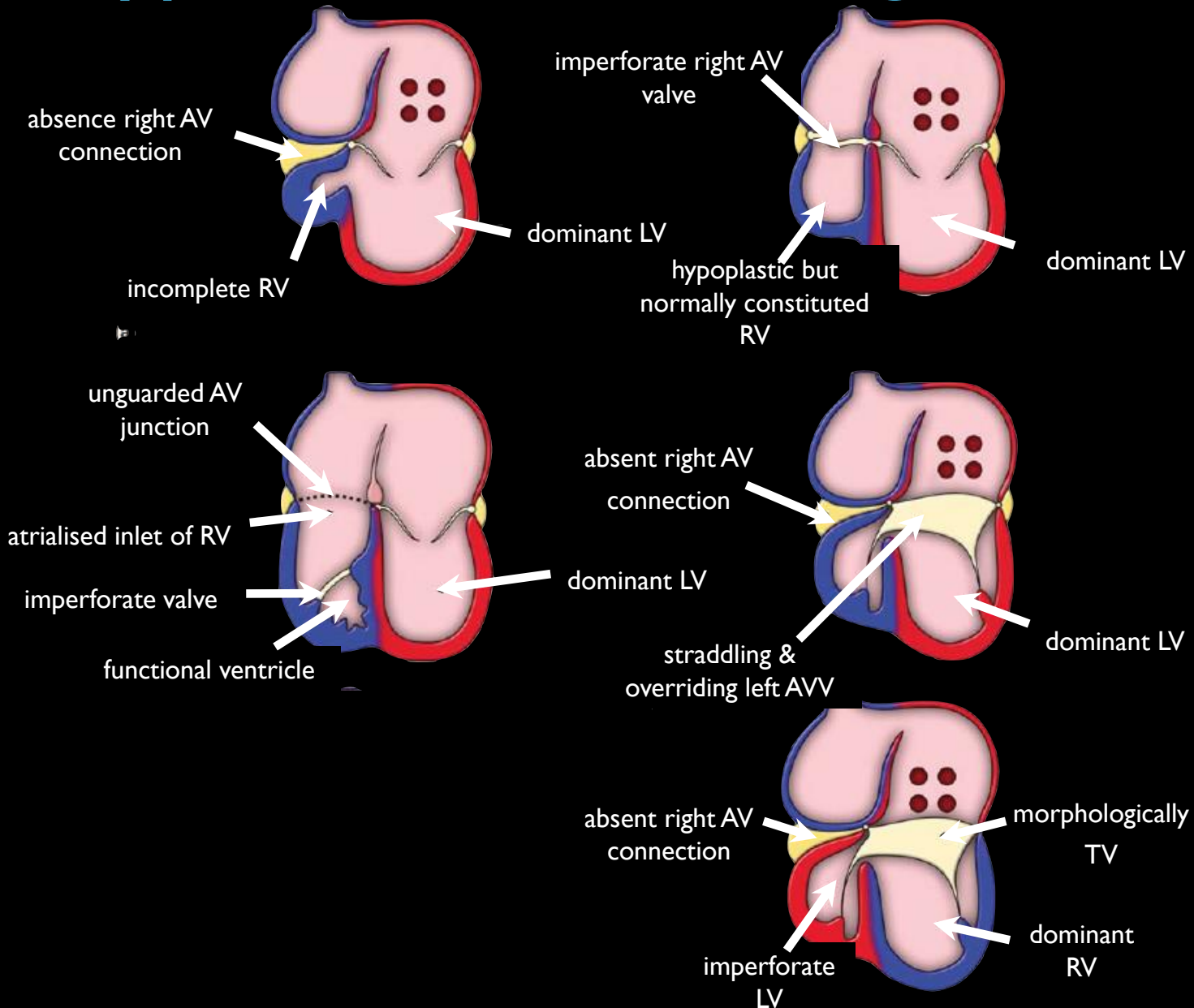
Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Phenotypes of Univentricular AV connection: Ventricular – Arterial Connection: Single Outlet



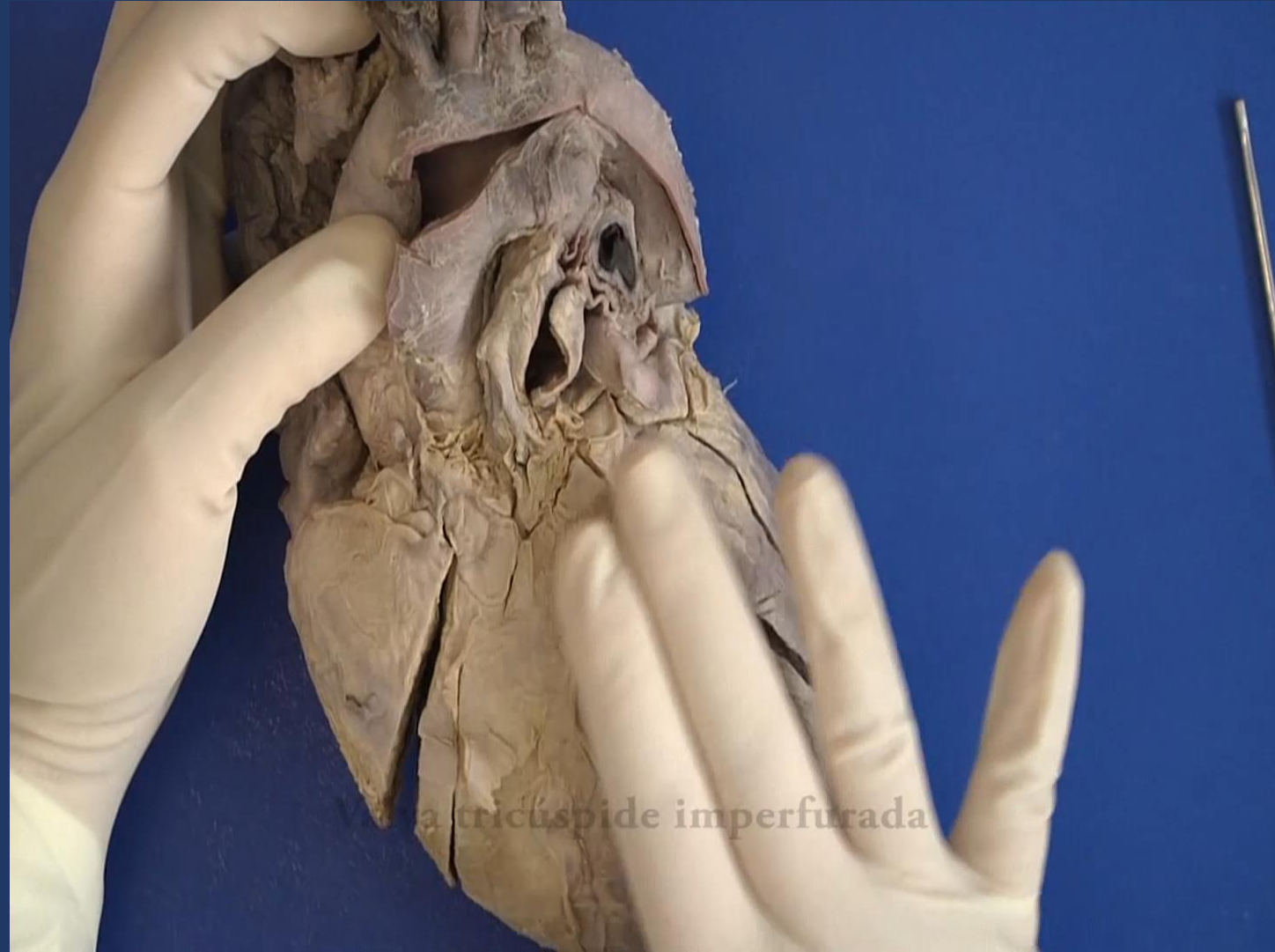
**Truncus
Arteriosus**

Phenotypes of absence of right AV connection



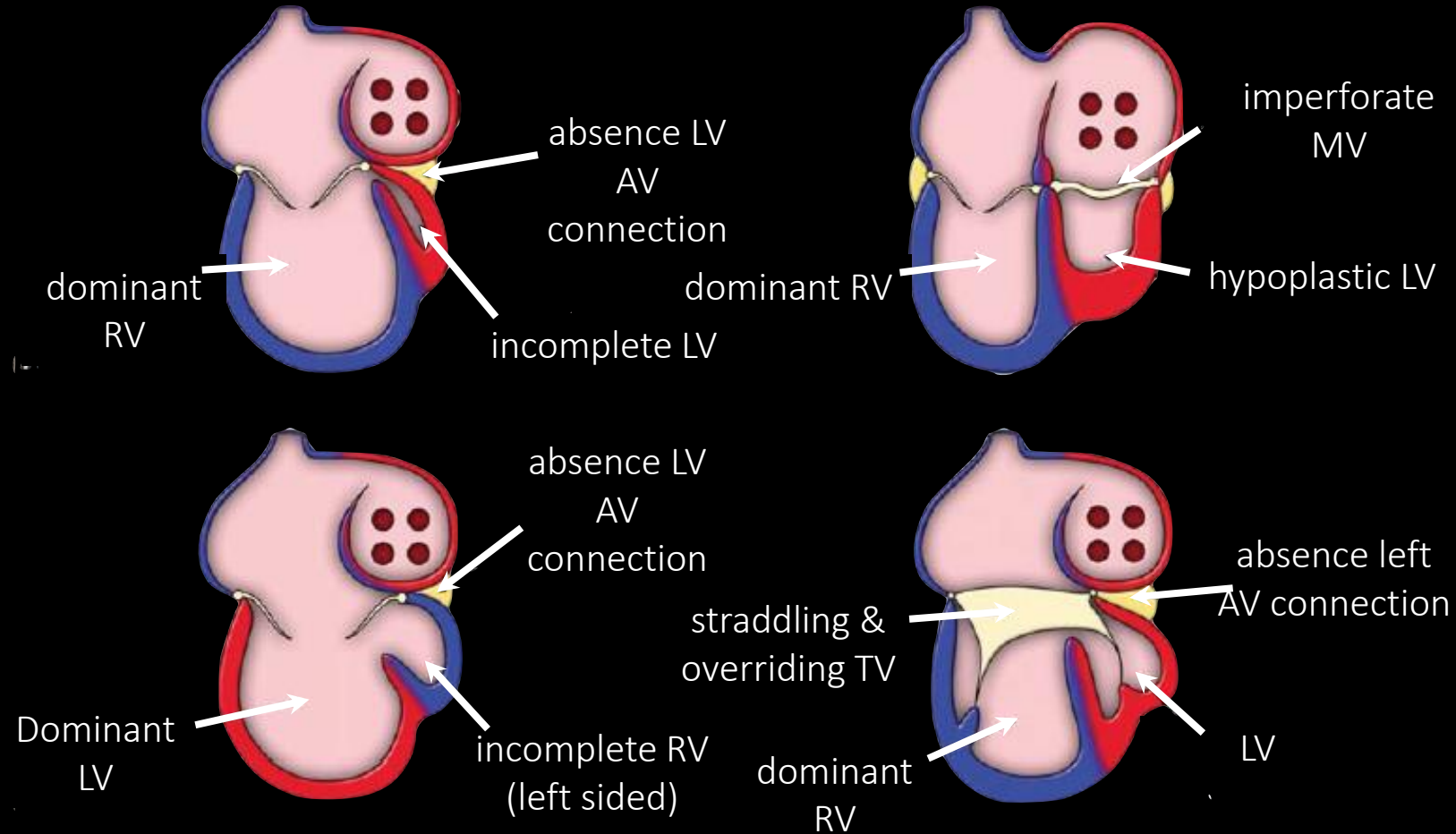


Atresia Tricúspide clássica



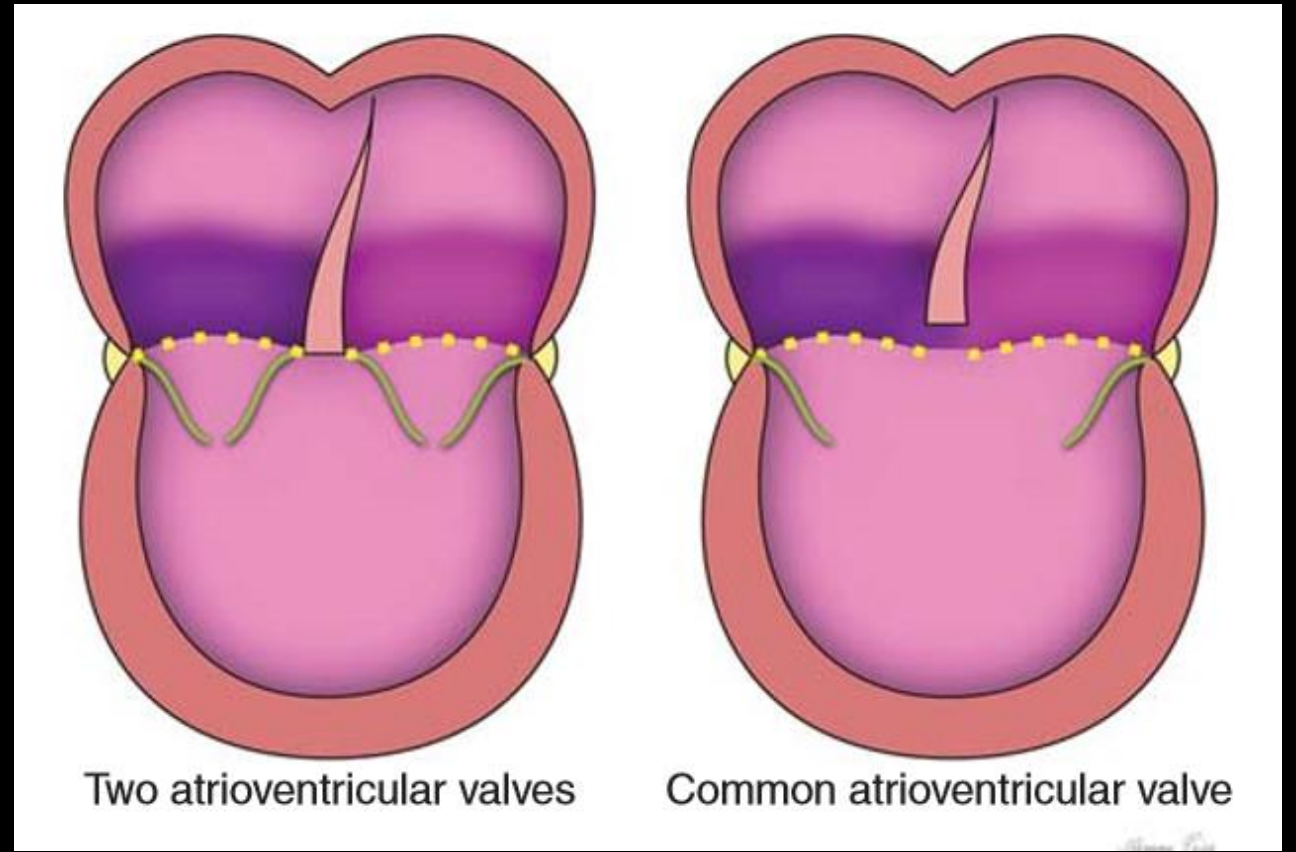
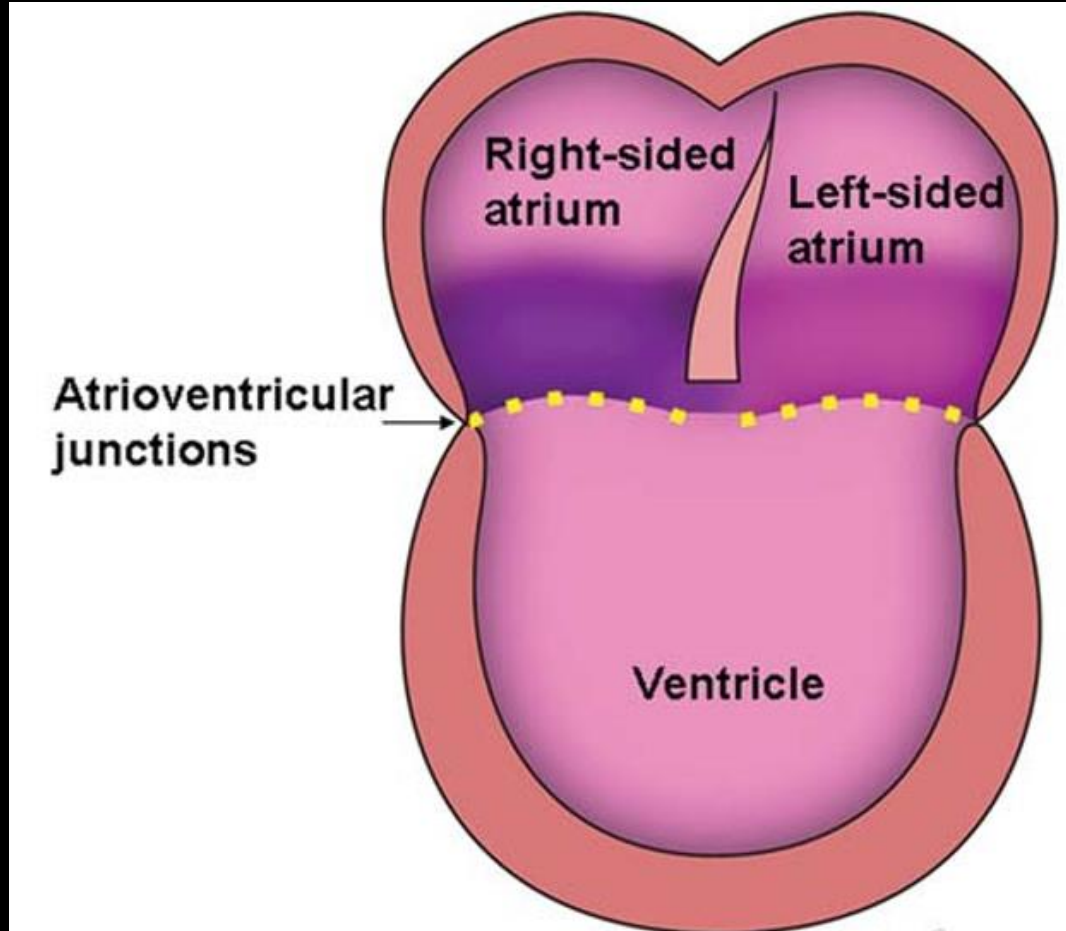
a tricuspide imperfurada

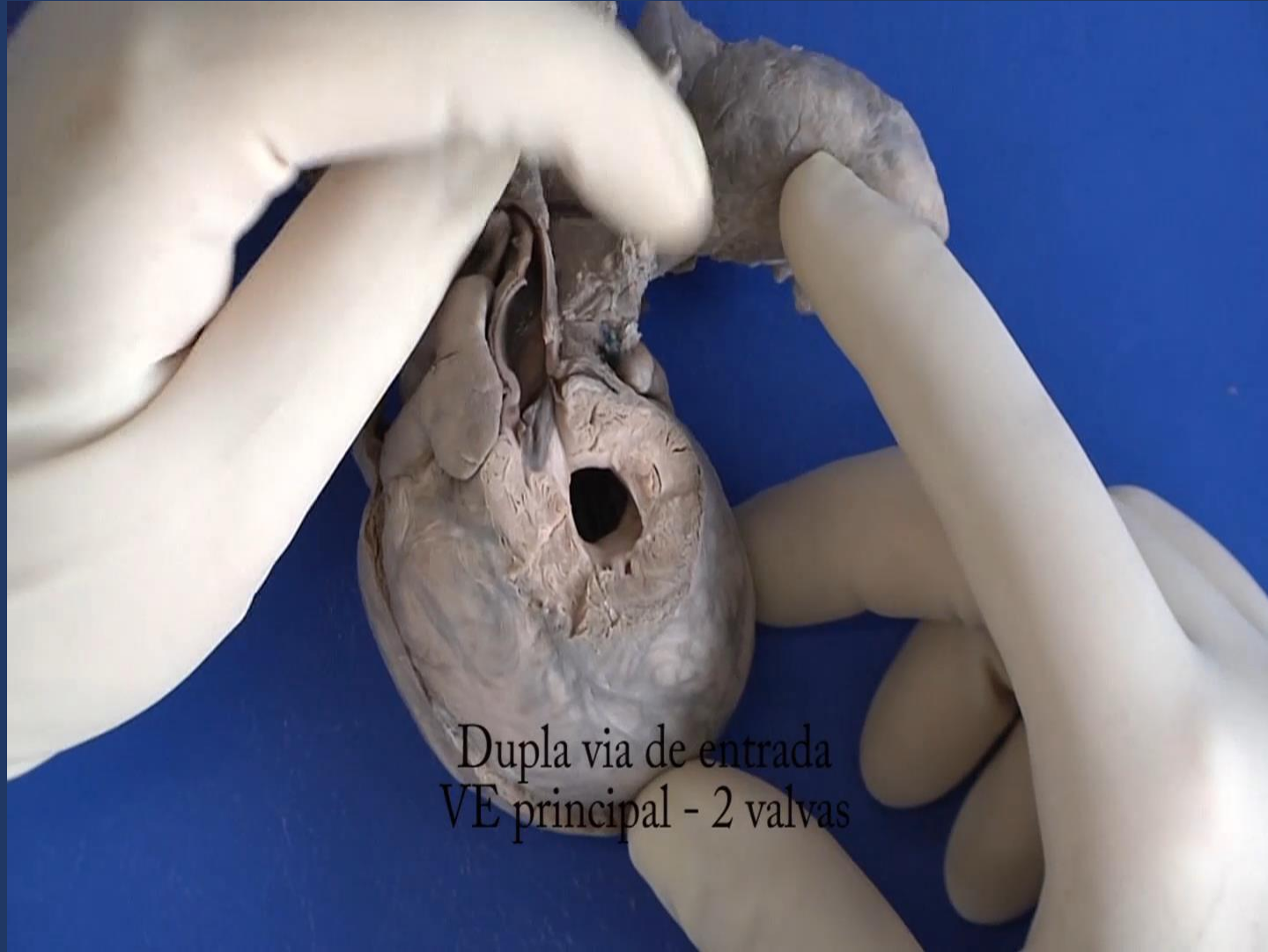
Phenotypes of absence of left AV connection





Double inlet of Ventricle





Dupla via de entrada
VE principal - 2 valvas



Dupla via de entrada
VE principal - valva única

CONGENITAL HEART DISEASES IN YOUR HANDS USING AN ECHO PROBE



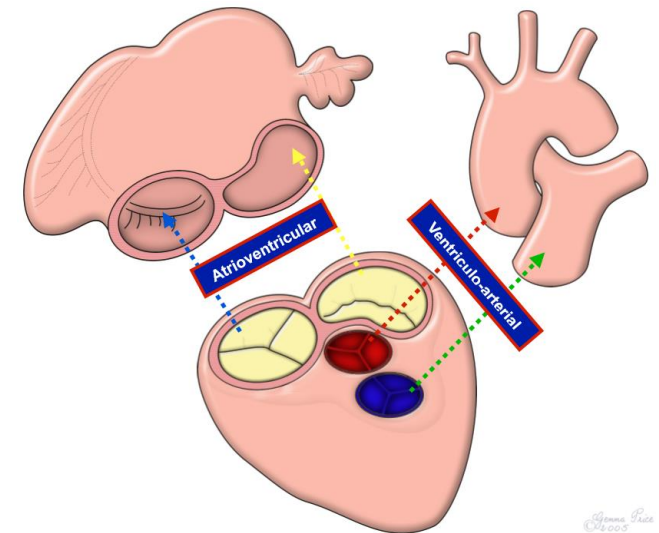
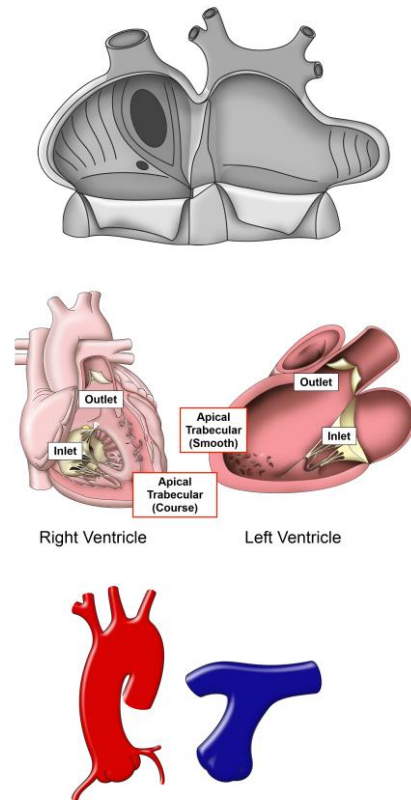
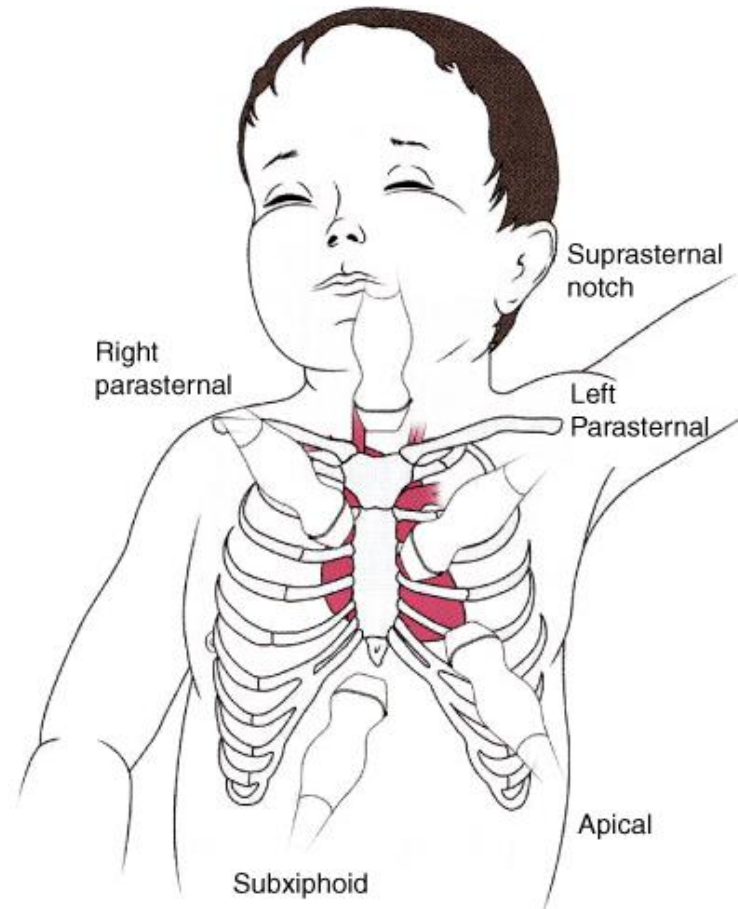
♥ Abnormalities of the Atrioventricular Junction ♥

Acoustic window

Anatomy

Imaging

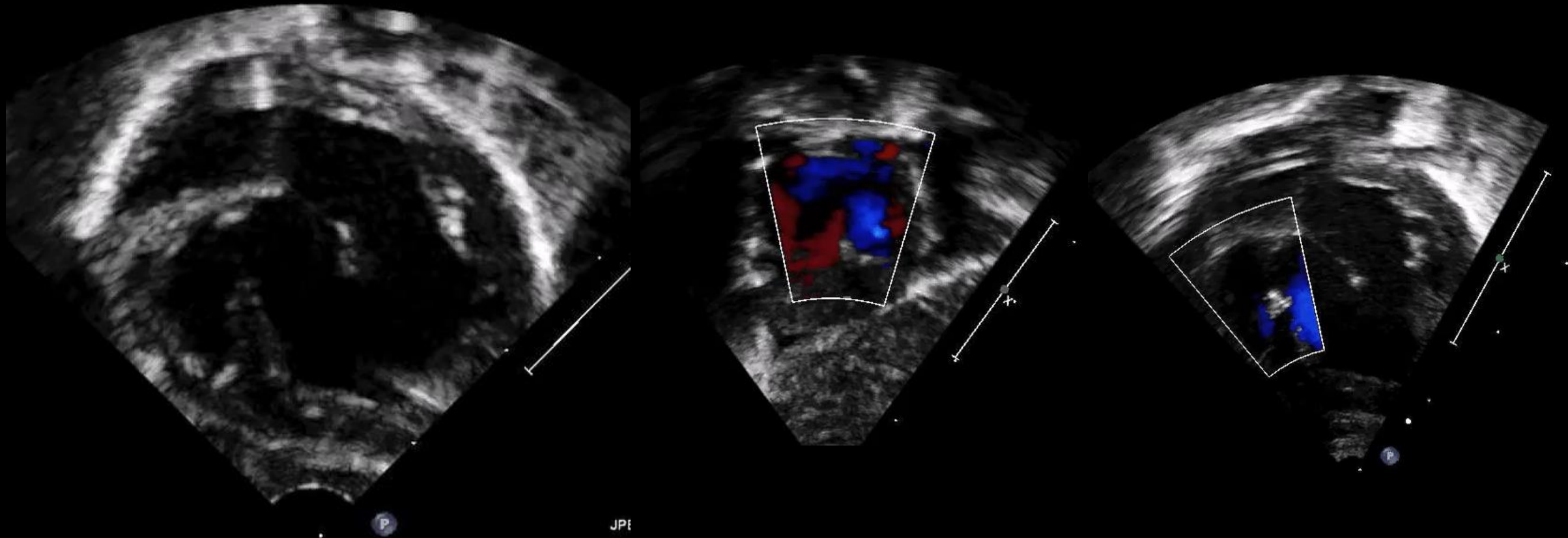
Sequential Segmentar



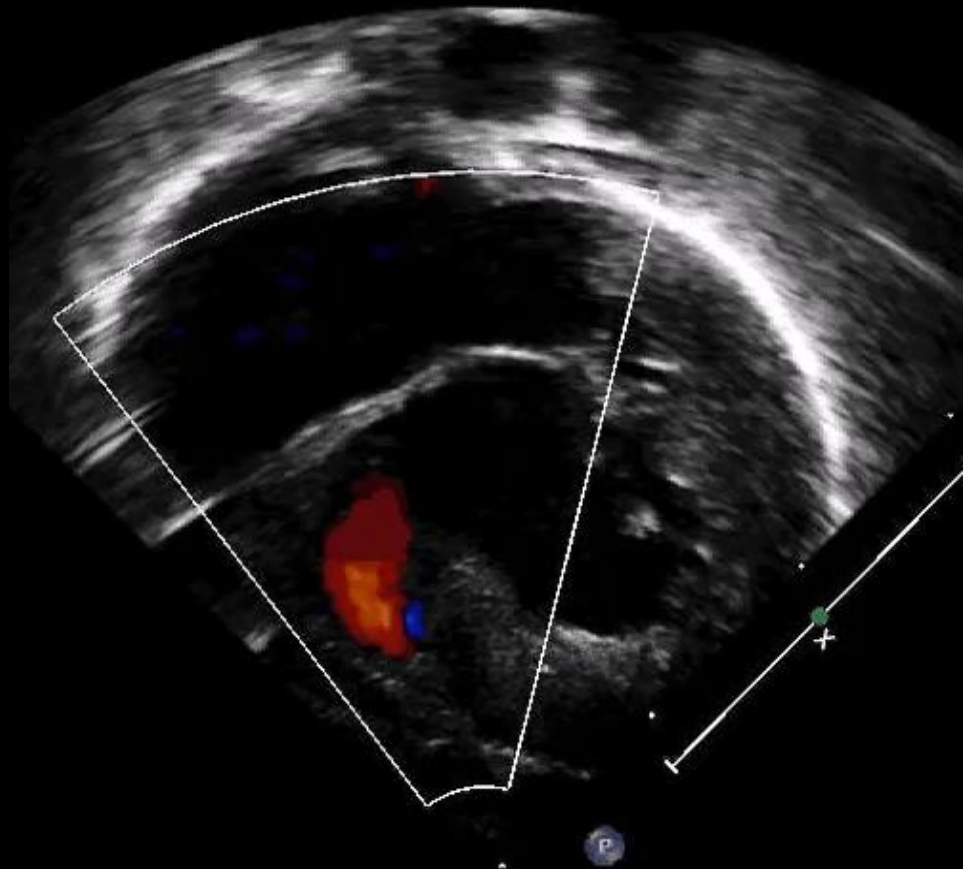
Absence of Right AV connection : Tricuspid Atresia



Absence of Right AV connection

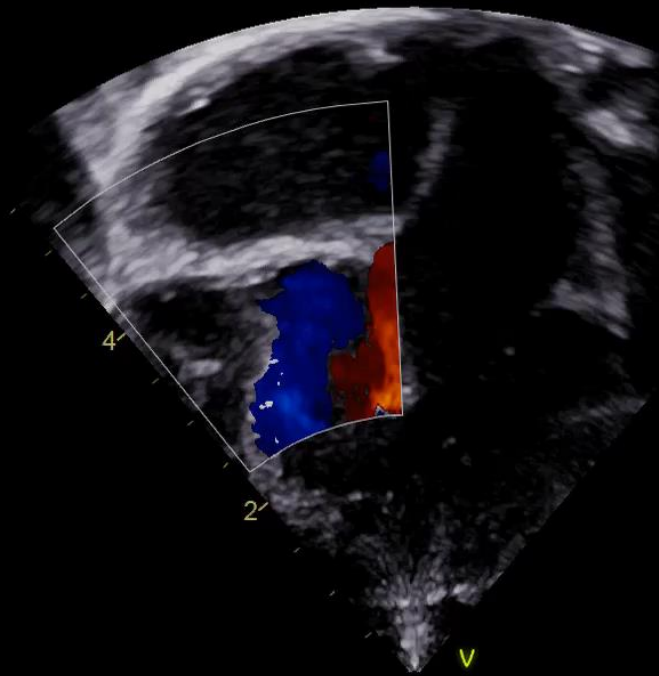


Absence of AV connection: Right



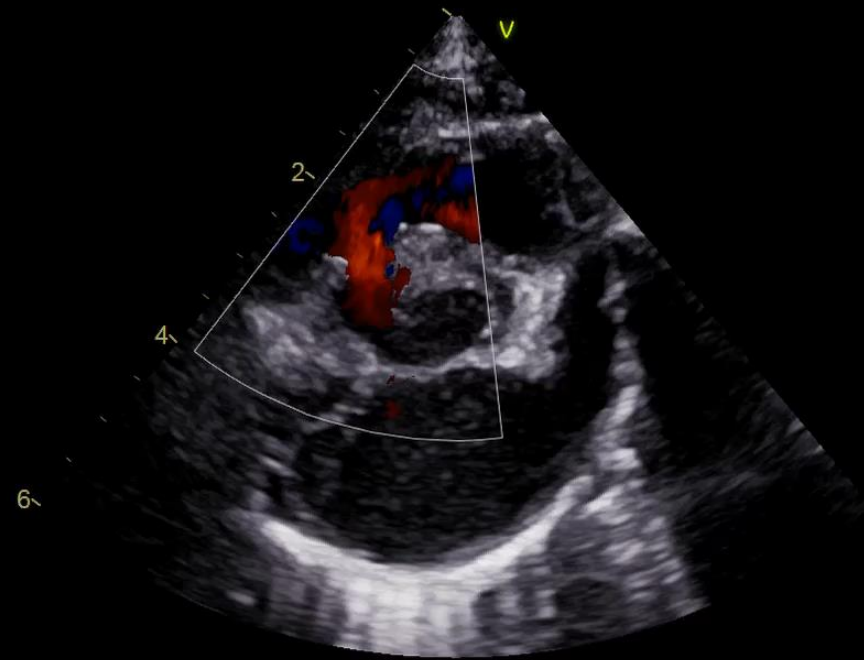
Absence of Right AV connection: Tricuspid Atresia

17:11:31
ACE



Soft
.80
-.80

17:10:13
ACE



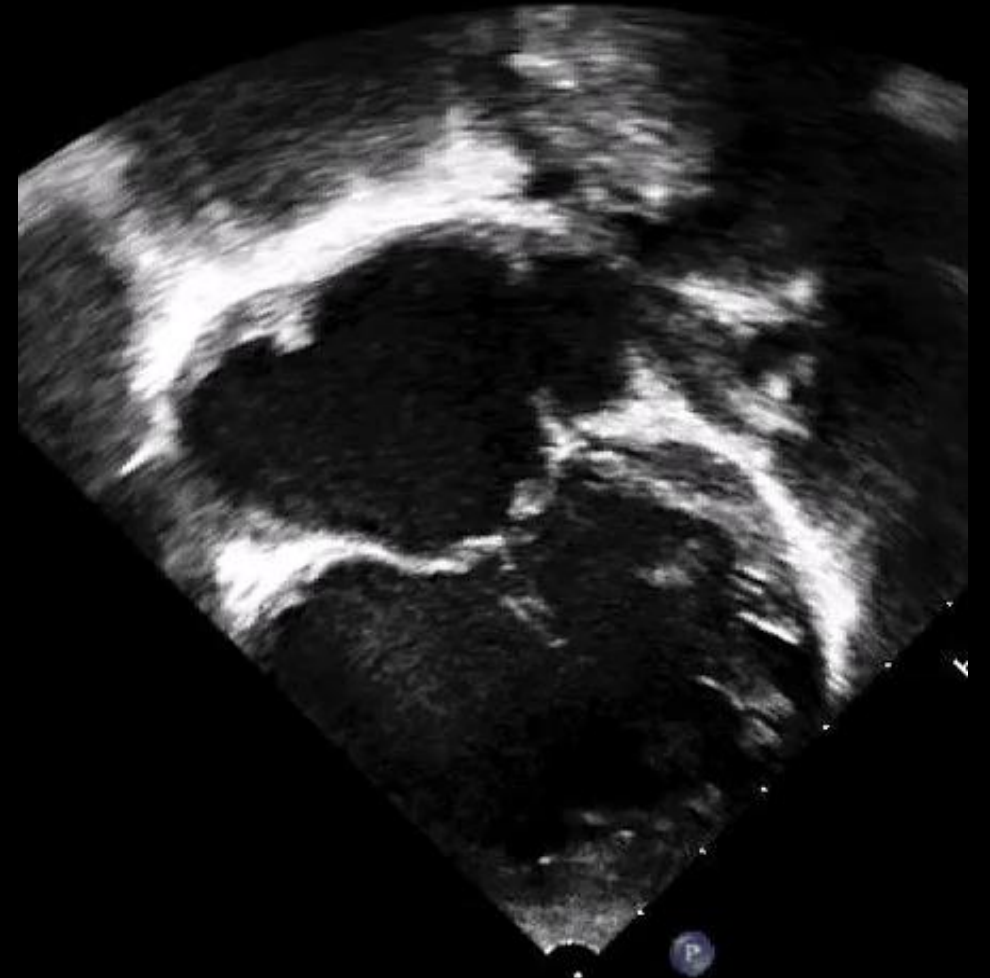
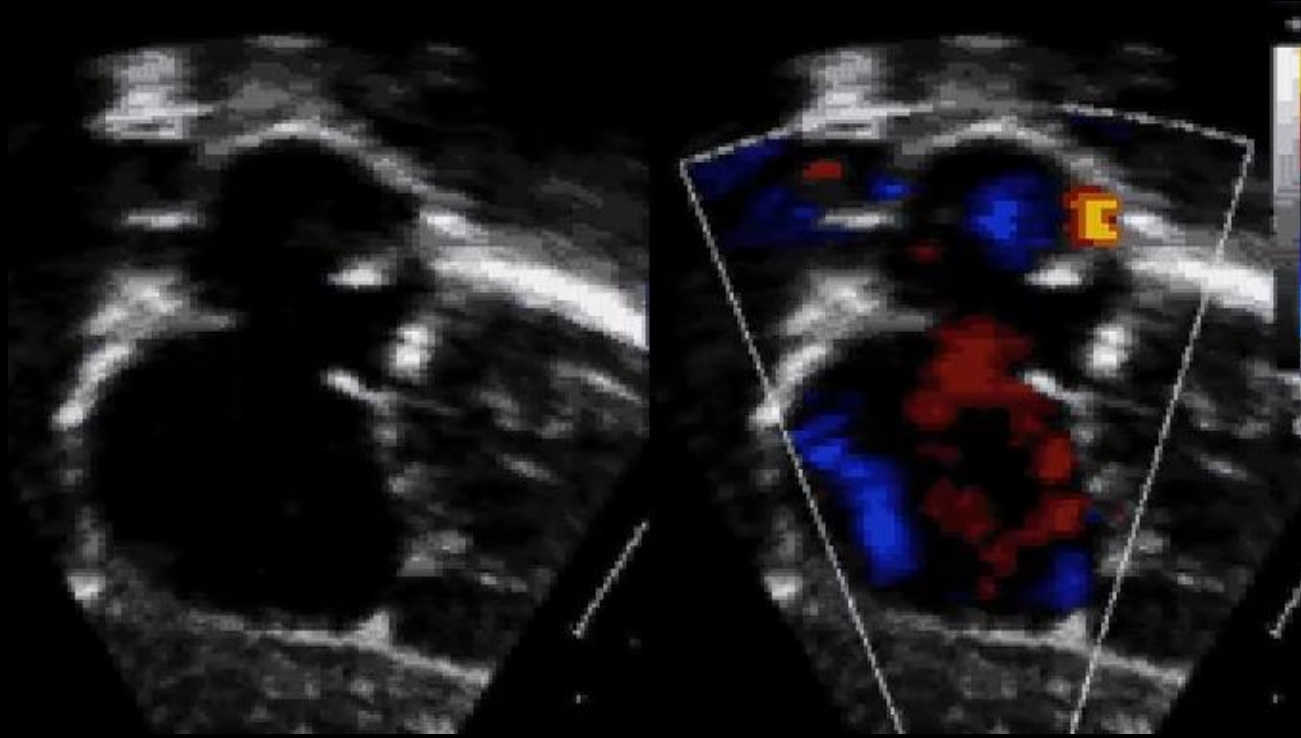
Soft
.80
-.80

Absence of Left AV Connection

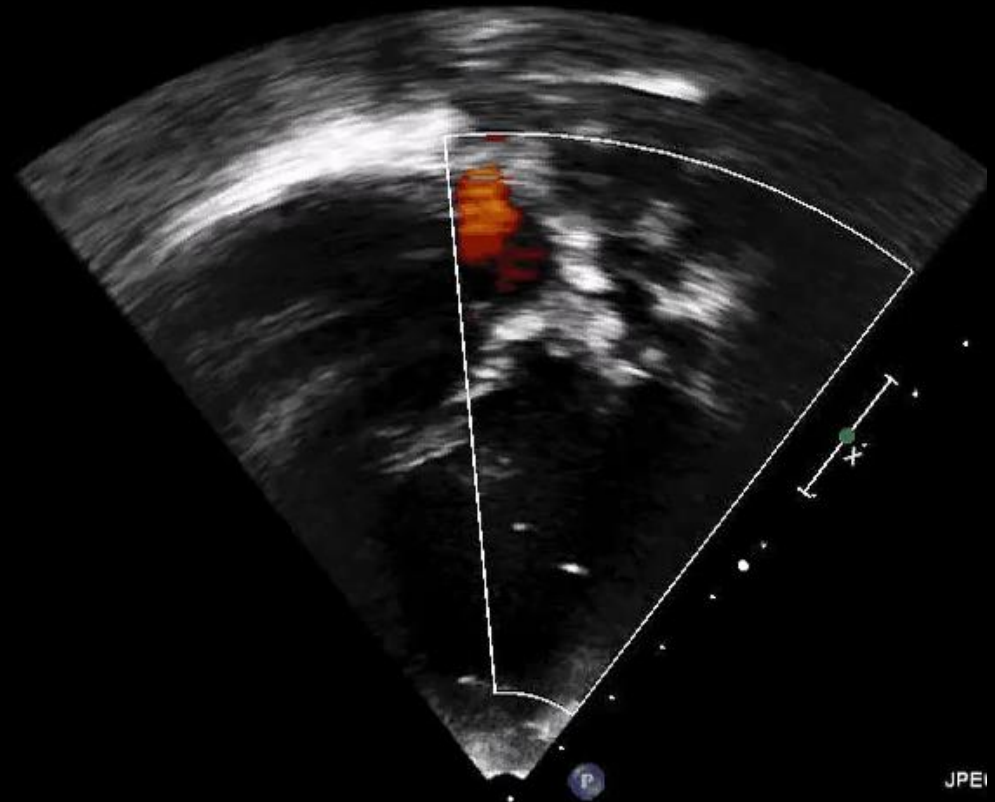
12:02:03



Absence of Left AV connection



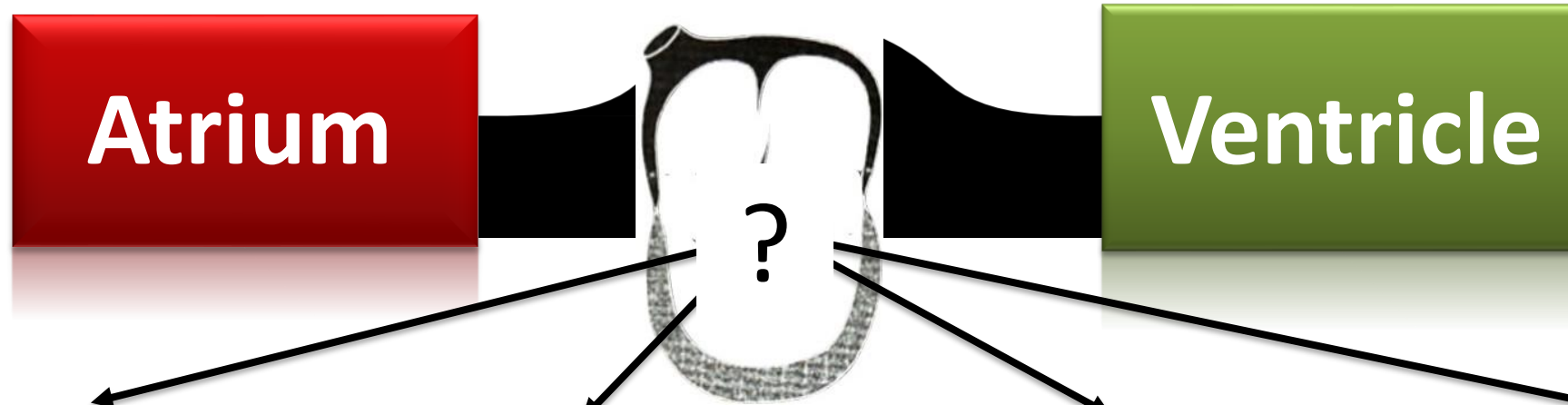
Absence of Left AV connection Double outlet of the Right Ventricle



Double inlet of Ventricle



Atrioventricular Valve (s)



2 atrioventricular valves



Absence of one AVV



Common AVV



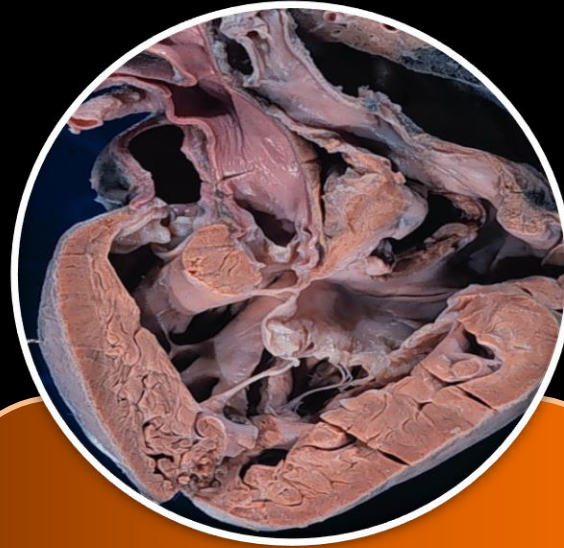
Straddling & Overriding



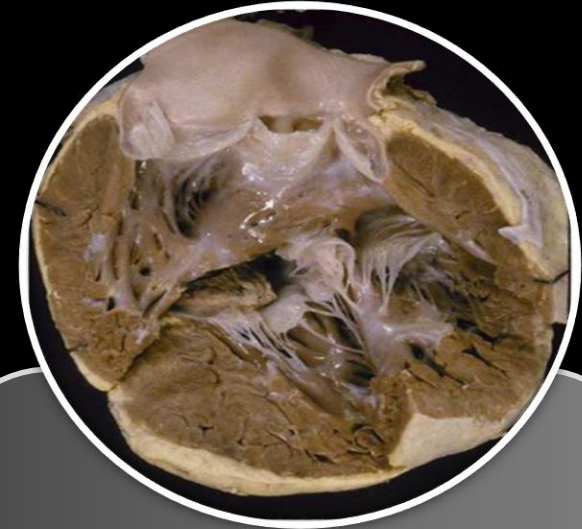
Univentricular AV connection: Atrioventricular Valve(s)



Can not predict the valve morphology (mitral or tricuspid) based on the ventricle morphology.

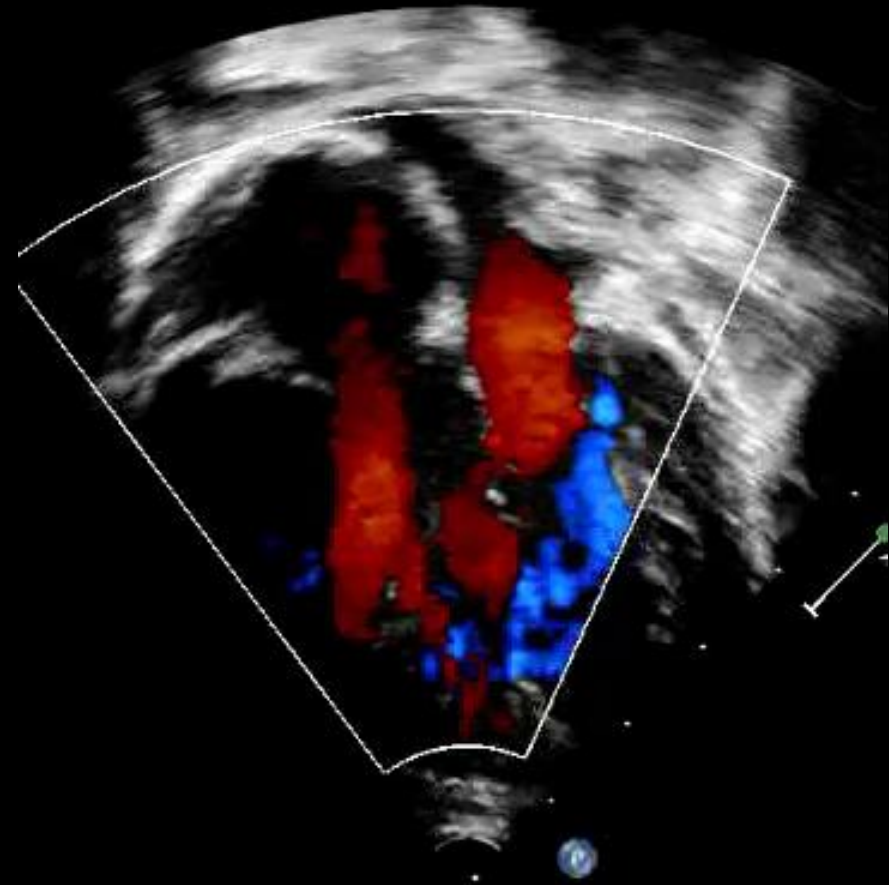
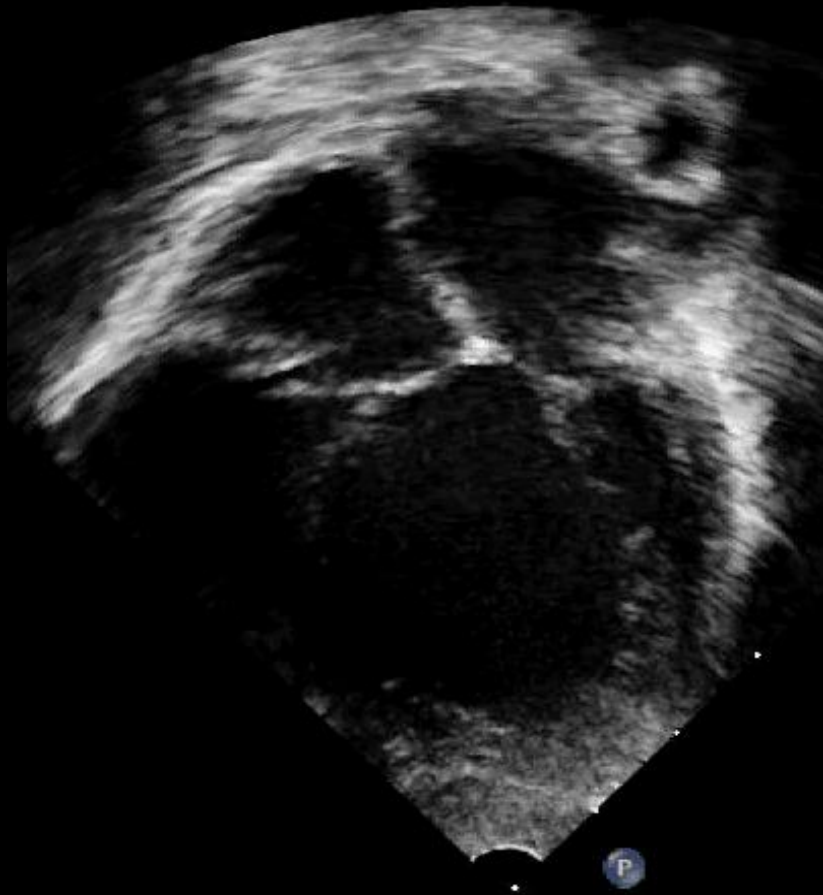


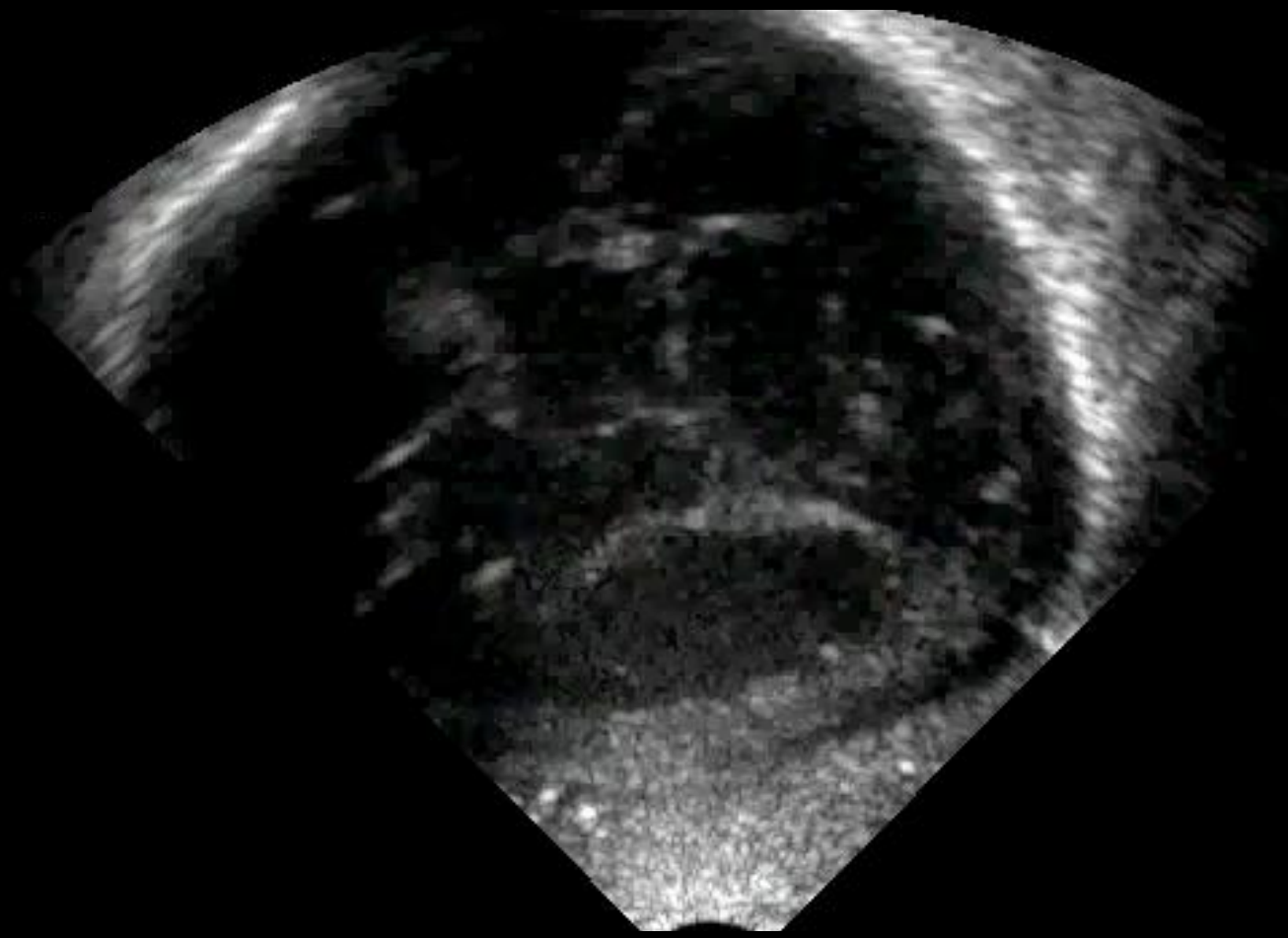
Surgical implications related with type/leaflets relationship/sub valvar apparatus.



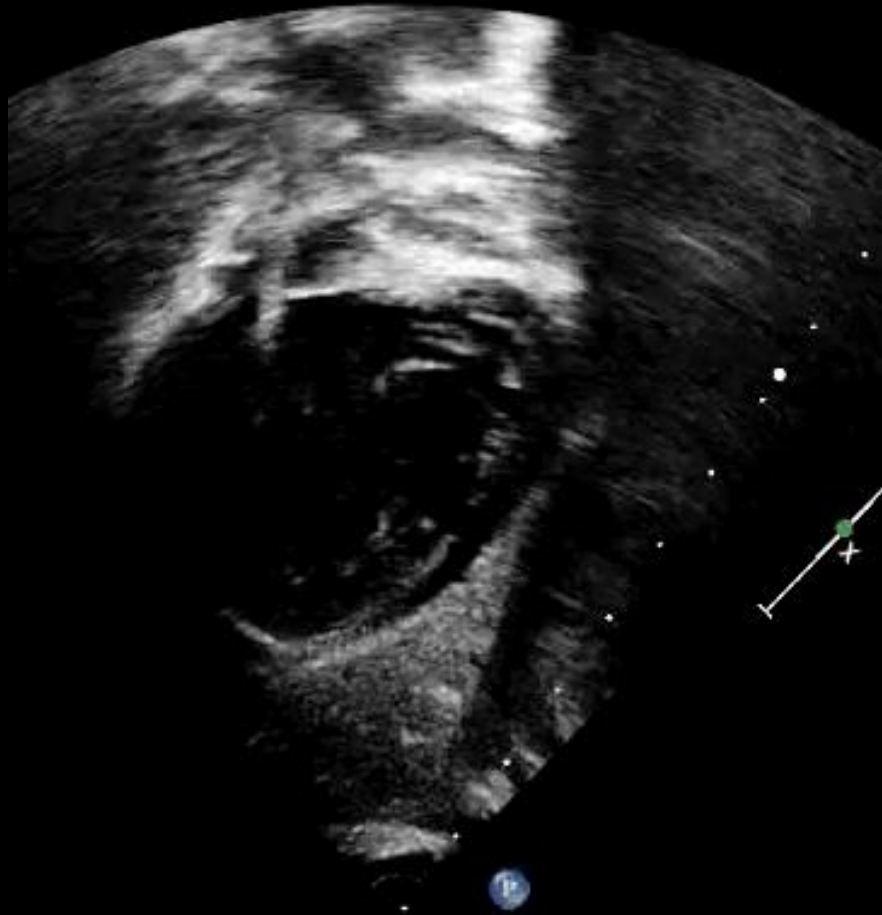
“true” univentricular hearts :
“Thick” trabeculations as sub valvar apparatus

Univentricular AV connection: 2 AVV

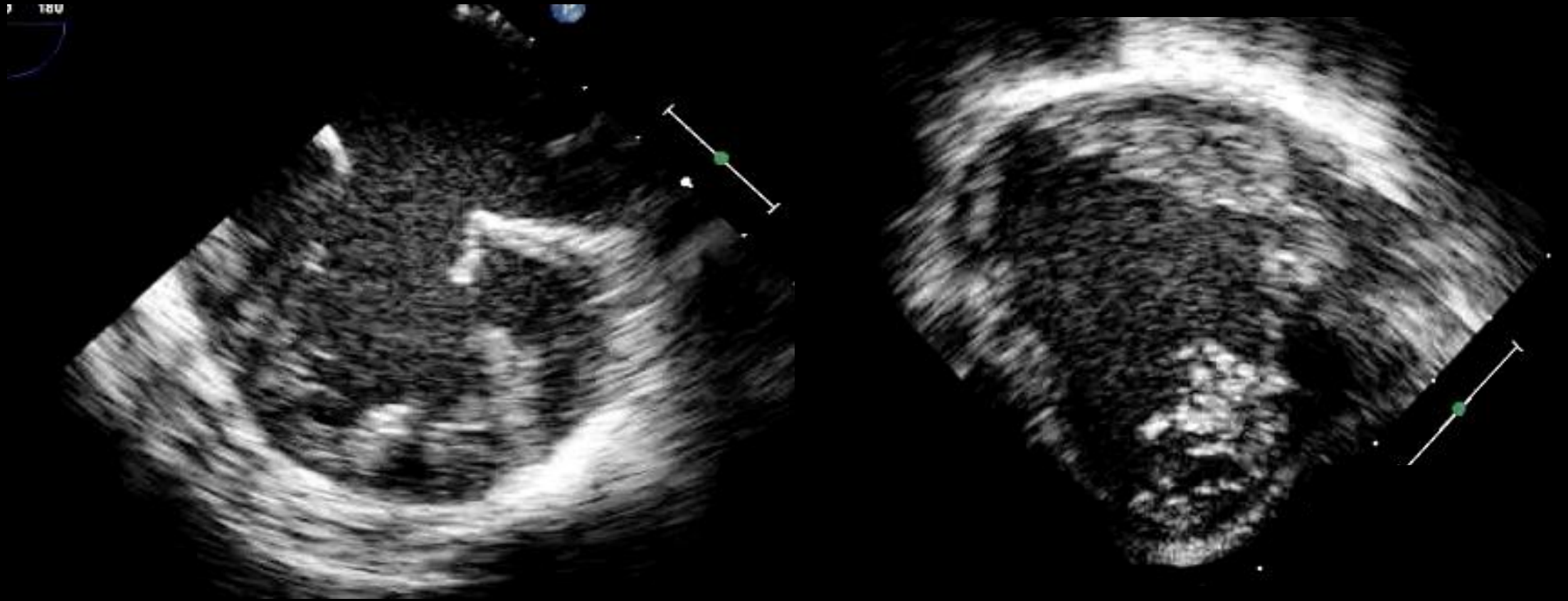




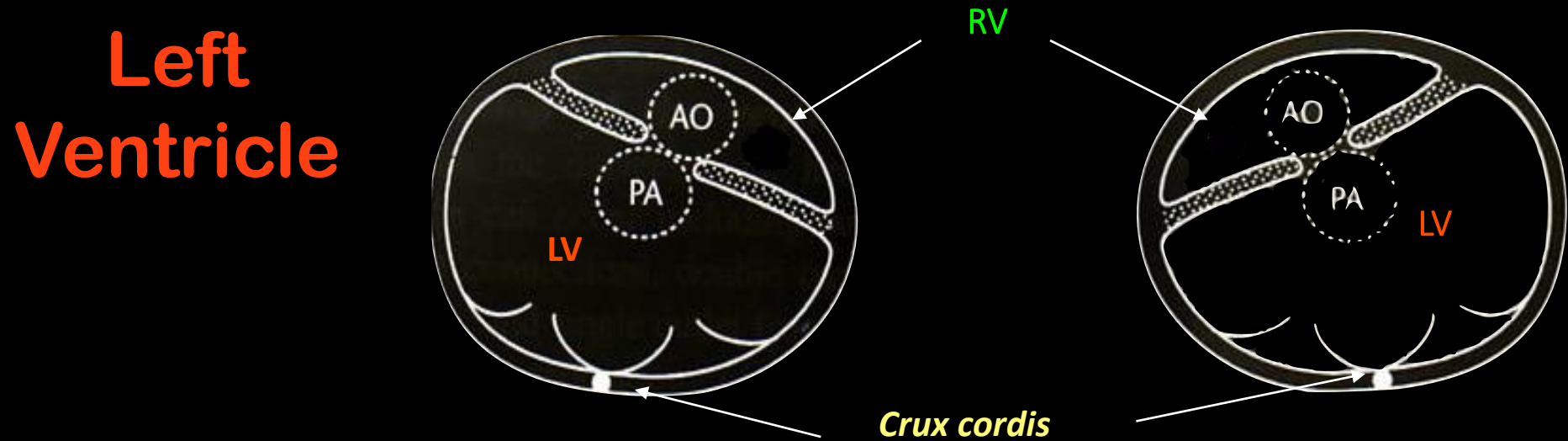
Double inlet: 2 valves:
Which one? Mitral? Tricuspid?



Double inlet : Common AVV



Morphological characteristics of the main Ventricle



- The most common ~70%
- ? Better outcome after Fontan
- Rudimentary chamber is ALWAYS anterior (right or left sided) and main chamber ALWAYS posterior (right or left sided).
- Fibrous continuity between the AVV & artery located more posterior.
- Trabecular septum is always anterior (“far away”) from the crux cordis

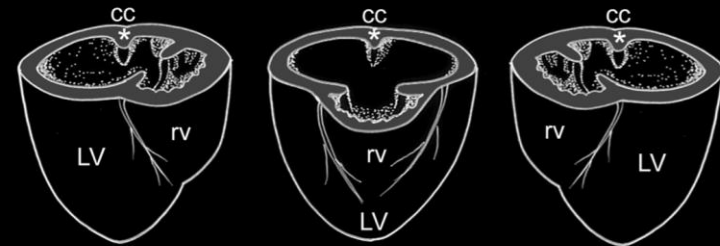
“Simplicity
is the
ultimate
sophistication,”

-Leonardo Da Vinci-

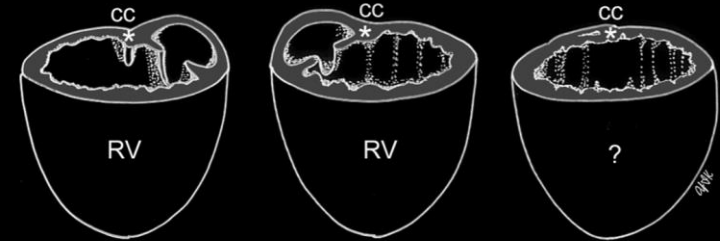


*Dr. Shi-Joon
Yoo*

Dr Yoo is a cardiac radiologist and clinical director of the 3D printing program at the Hospital for Sick Children. He is a professor in the Departments of Medical Imaging and Paediatrics at the University of Toronto. His main clinical and research activities are MR and CT applications in children with cardiovascular disease and fetal echocardiography. He has developed a unique training program for cardiac imaging where both radiology and cardiology fellows collaborate closely together in a harmonious and productive manner. He has introduced 3D printing technology to the program. This technique allows preoperative simulation of the surgical procedures in patients with complex congenital heart disease.

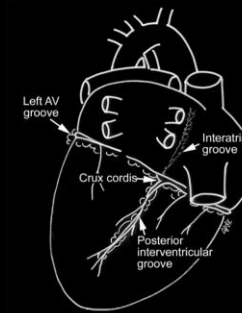


Main chamber with left ventricular morphology
Single LV

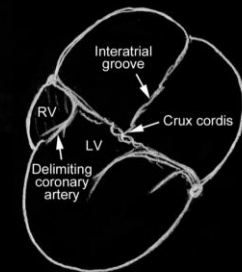
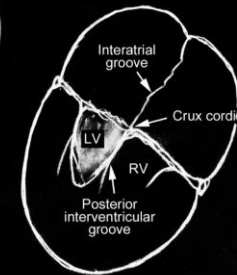


Main chamber with right ventricular morphology
Single RV

Solitary ventricle of indeterminate morphology



Crux cordis

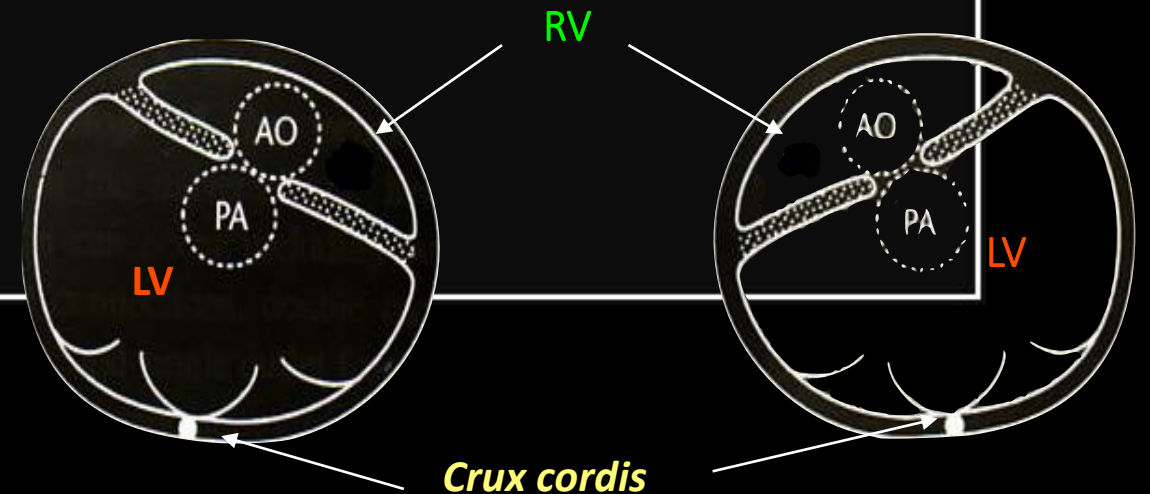




I'm sort of like Costco. I'm big, I'm not fancy and I dare you to understand Crux cordis and how identify the rudimentary chamber

- Vitor Guerra

**Left
Ventricle**



Definition of the crux cordis of the Heart

The **crux cordis** or **crux of the heart** (from Latin "crux" meaning "cross") is the area on the lower back side of the [heart](#) where the [coronary sulcus](#) (the groove separating the [atria](#) from the [ventricles](#)) and the [posterior interventricular sulcus](#) (the groove separating the left from the right ventricle) meet. It is important surgically because the [atrioventricular nodal artery](#), a small but vital vessel, passes in proximity to the crux of the heart. It is the anastomotic point of right and left coronary artery.

Anatomy of the echocardiographic crux cordis in the evaluation of the spectrum of atrioventricular valve atresia

Andrea Magherini, Gaetano Azzolina and Jeanine Careri

Tuscan Heart and Chest Center, Florence, Italy

(Received 11 May 1983; revision accepted 24 July 1983)



1982

“The echocardiographic crux cordis in the normal subject is defined as the spatial interrelation between the atrial and ventricular septal planes and the plane of the AV junction. Its image may be achieved using apical, subcostal, or precordial four-chamber views (planes at right angles to the atrial septum) in which both atrial and ventricular chambers are aligned one next to the other but are separated by their respective septa and AV valves, This plane cuts the atrial and ventricular septum and the AV junction at a level somewhat above the posterior (diaphragmatic) surface of the heart.”

Para sternal Short axis Sweep from the base towards the apex



I'm sort of like Costco. I'm big, I'm not fancy and I dare you to not like me.

- Vitor Guerra

Left Ventricle

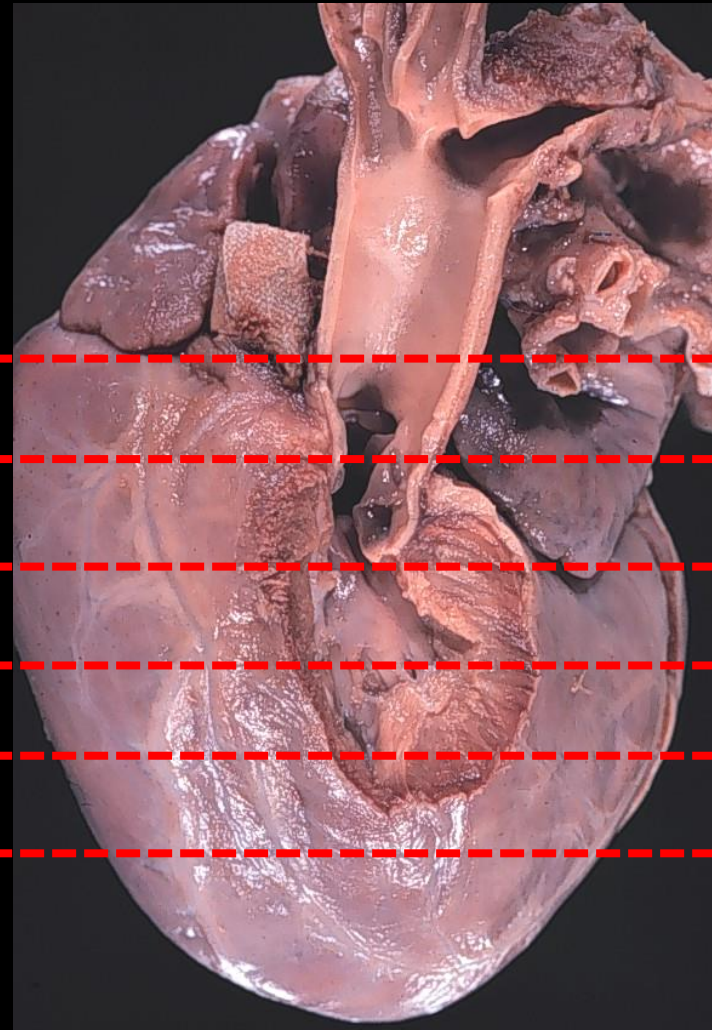
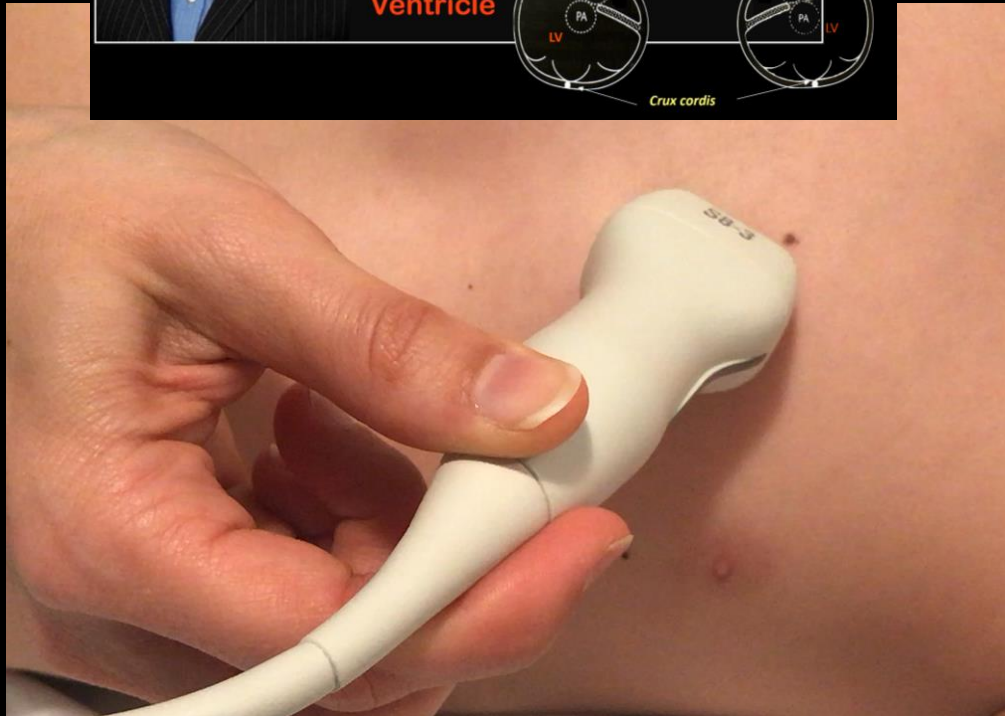
RV

AO

PA

LV

Crux cordis

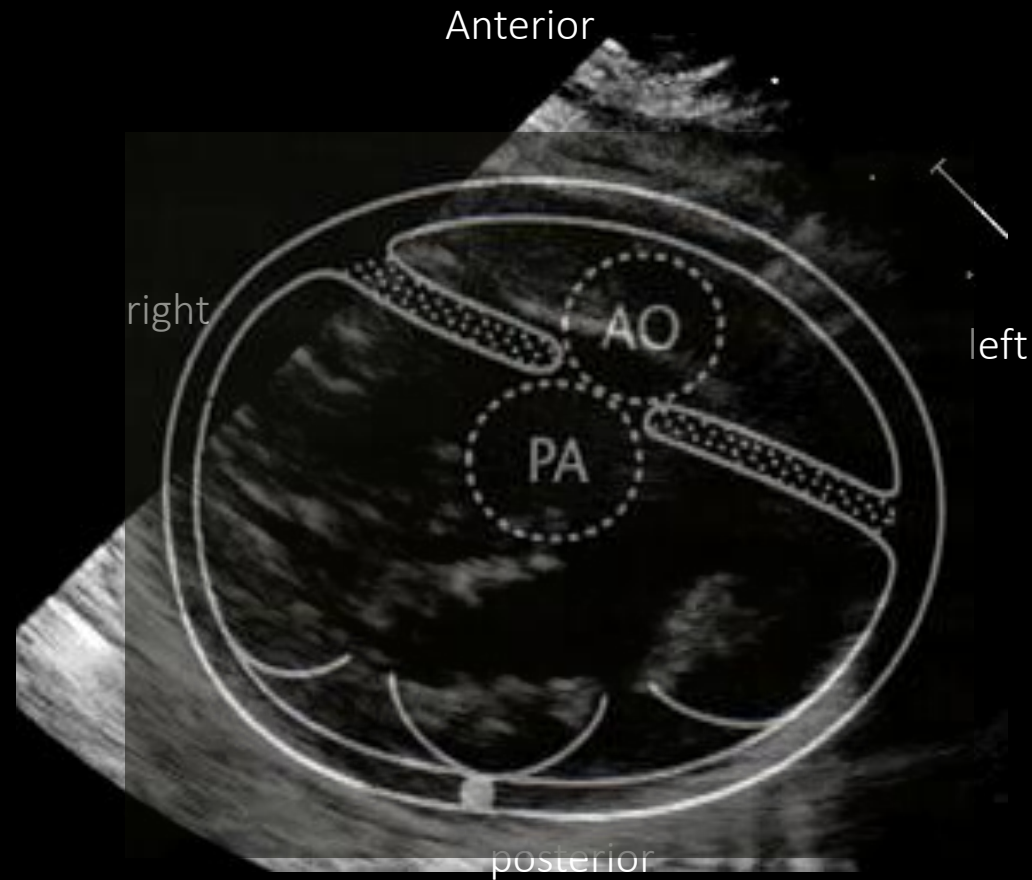


Courtesy Prof Aiello , Heart Institute of University of Sao Paulo

Morphological characteristics of the main Ventricle

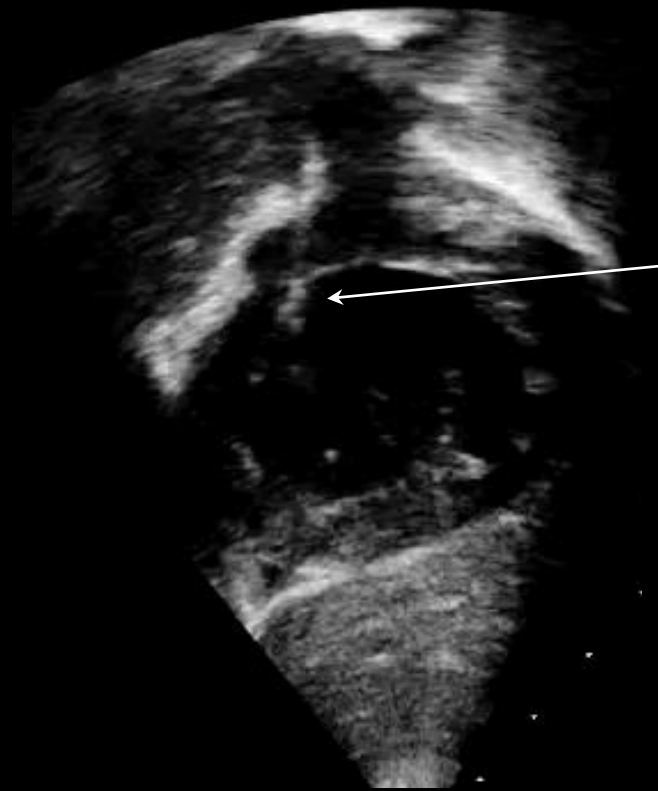
Left Ventricle

- ✓ Parasternal short axis
- ✓ Level of the interventricular septum
(always look at the direction)
- ✓ Level of the AVV valve (s) and crux
cordis



Morphological characteristics of the main Ventricle

Left Ventricle



Fibrous continuity
AVV and great artery

Subcostal view



superior

inferior

Morphological characteristics of the main Ventricle

Left Ventricle

Hypoplastic RV on left side



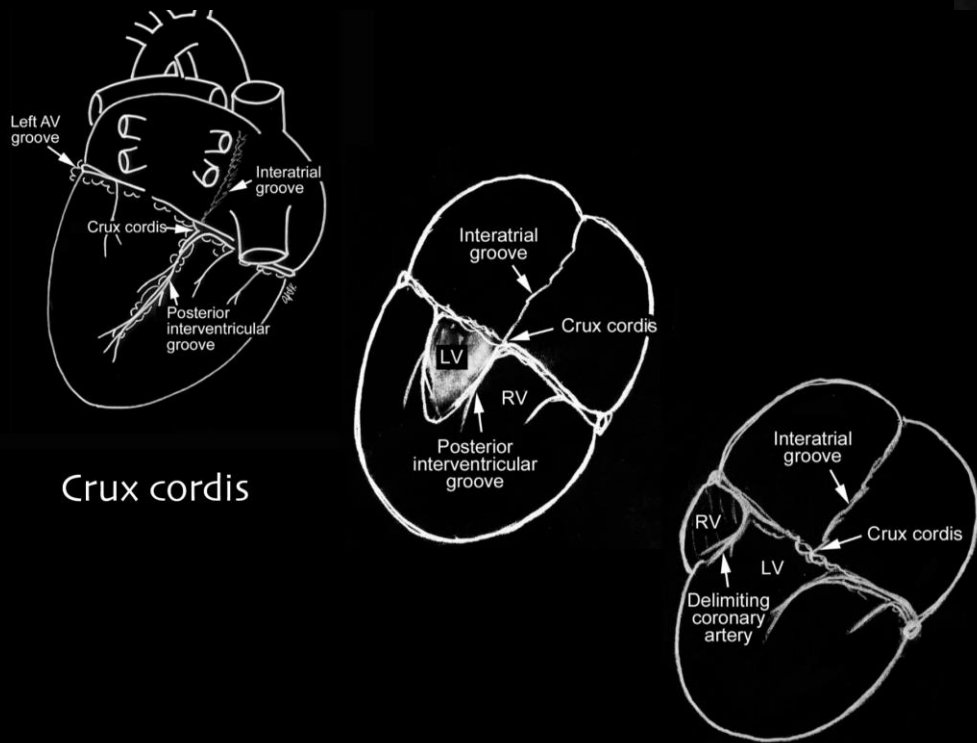
Hypoplastic RV on right side



! From the 4 ch view: sweep anteriorly /posteriorly
to find the rudimentary chamber

Morphological characteristics of the main Ventricle

Left Ventricle



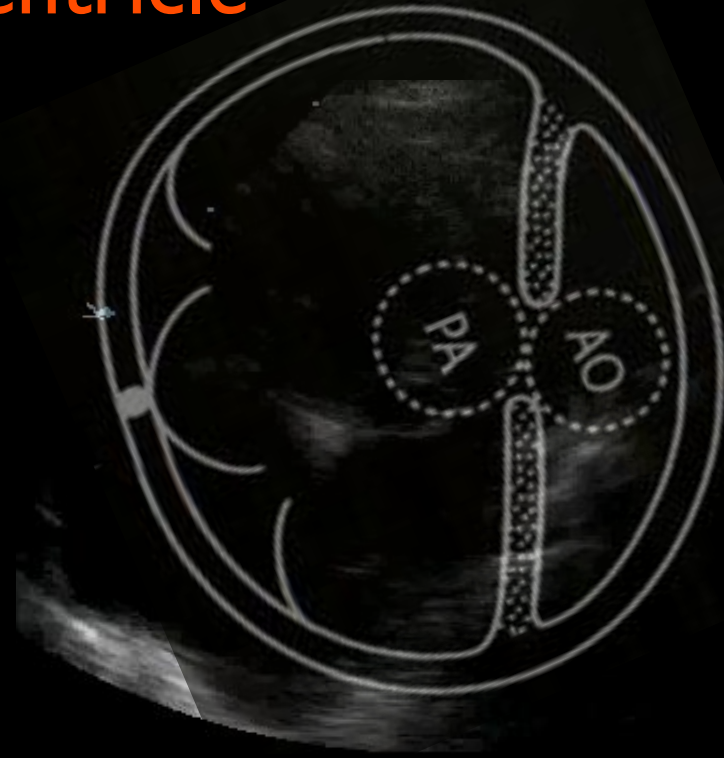
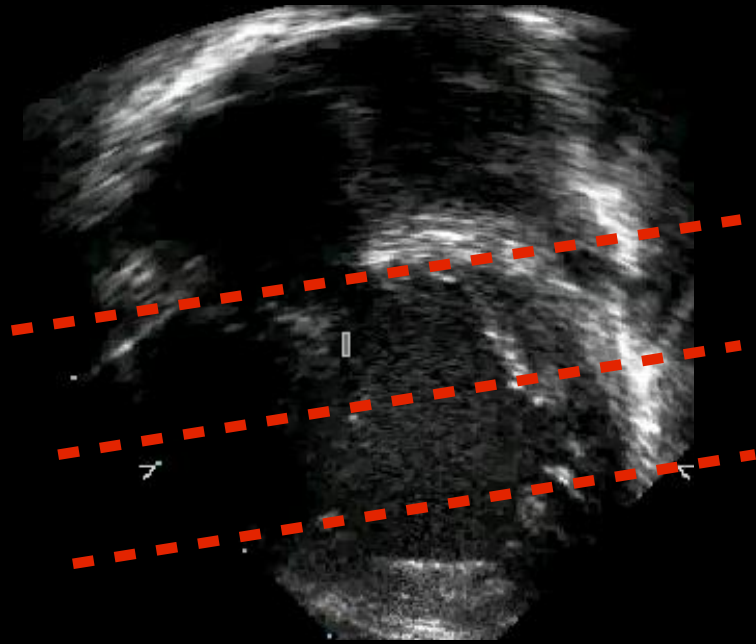
Crux cordis



! From the 4 ch view: sweep anteriorly /posteriorly to find the rudimentary chamber

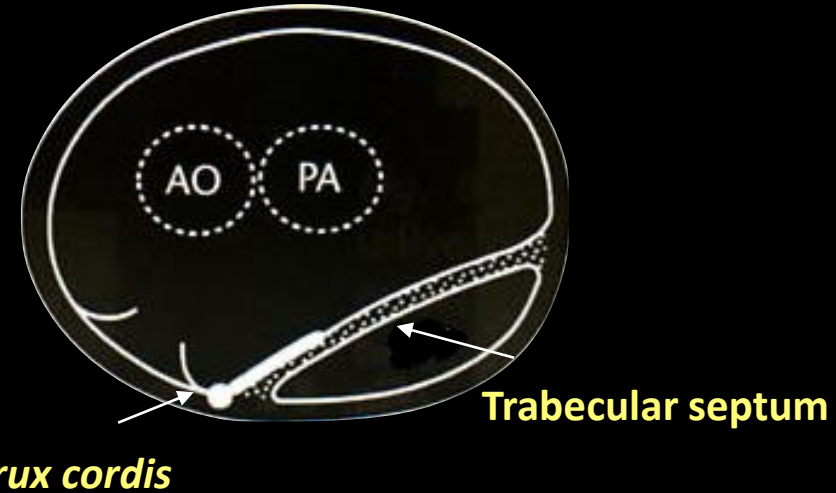
Morphological characteristics of the main Ventricle

Left Ventricle



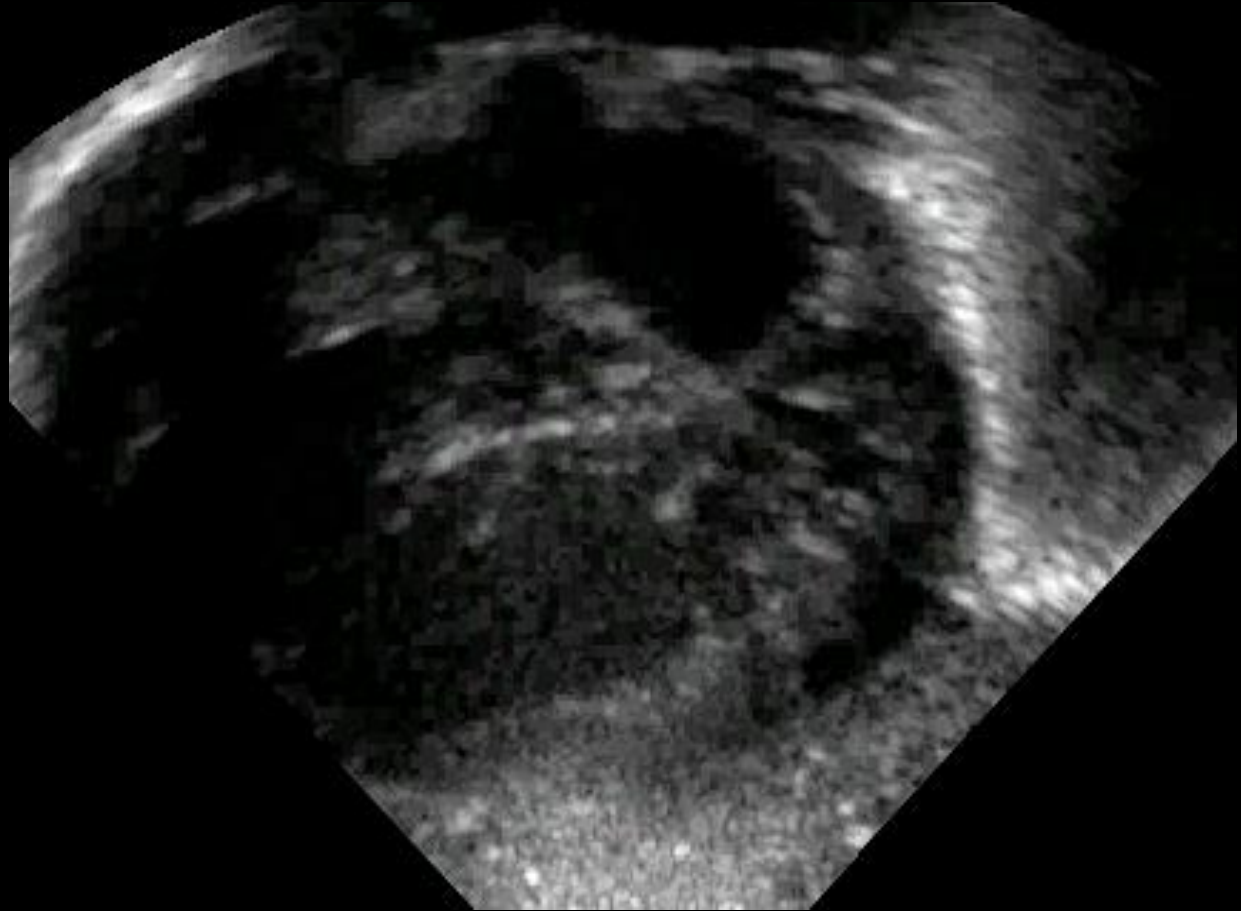
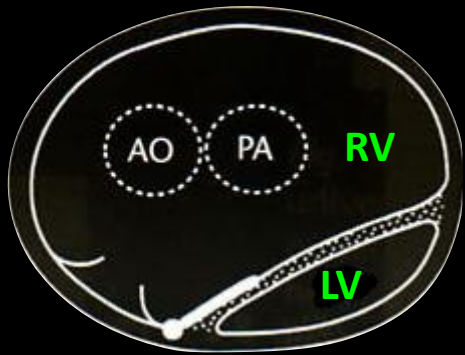
Morphological characteristics of the main Ventricle

Right Ventricle



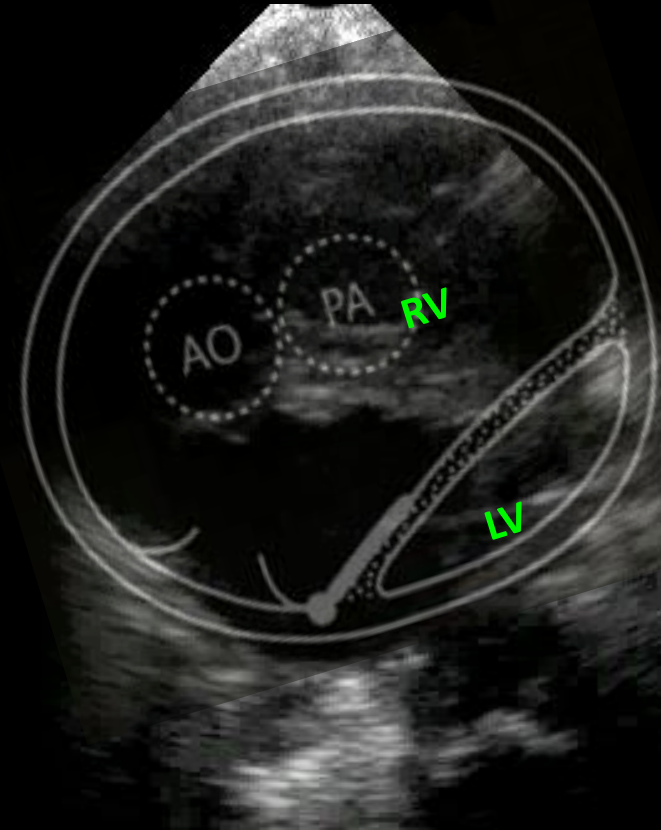
- Rudimentary chamber is located posteriorly (trabecular portion of LV)
- Trabecular septum is posterior to the *crux cordis*
- ?Poor outcome
- No fibrous continuity between AV and great vessels.

Double inlet of RV & Double outlet of RV (DIRV/DORV)

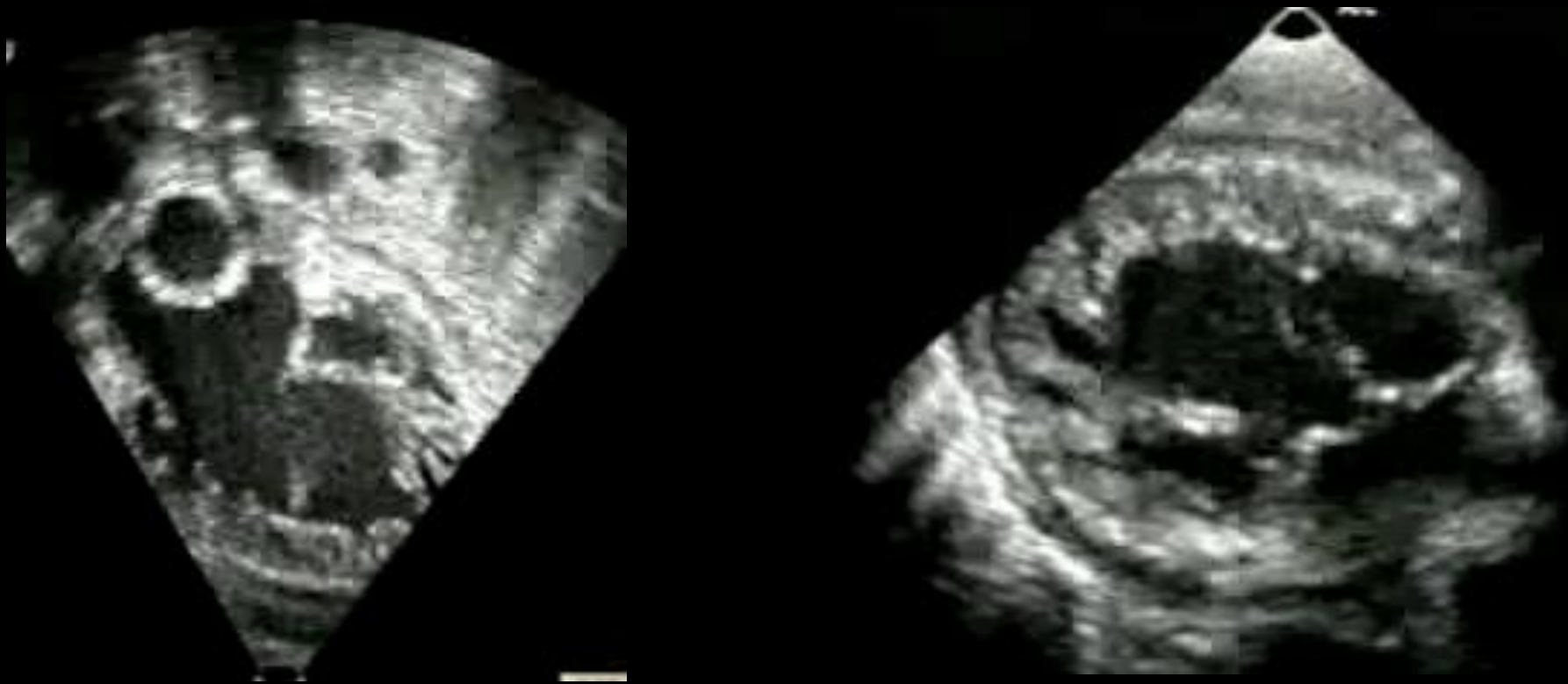


Morphological characteristics of the main Ventricle

Double inlet of Right Ventricle & Double outlet of Right Ventricle

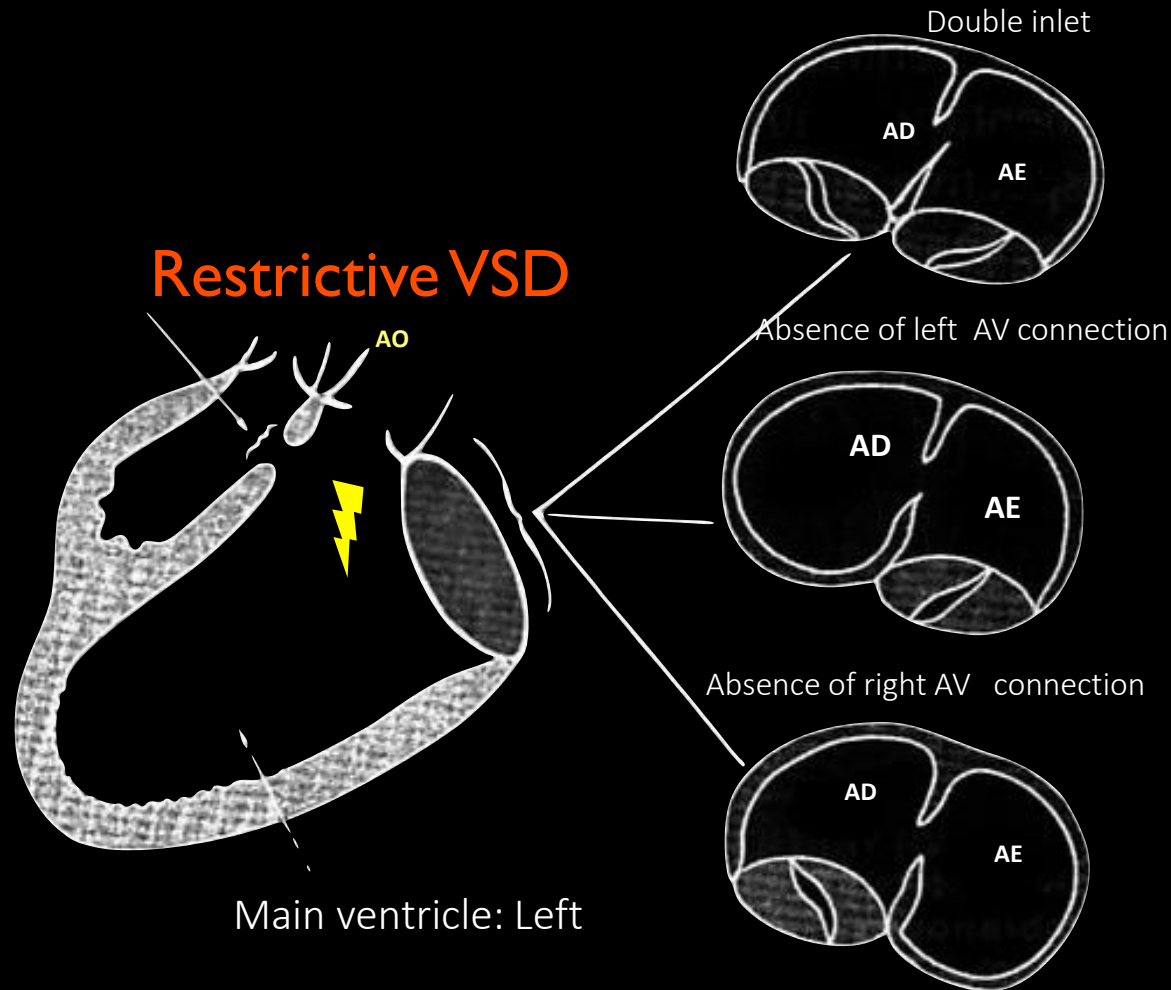


Morphological characteristics of the main Ventricle



Double inlet of RV (Common AVV)

Univentricular AV connection: Ventricular septal defect

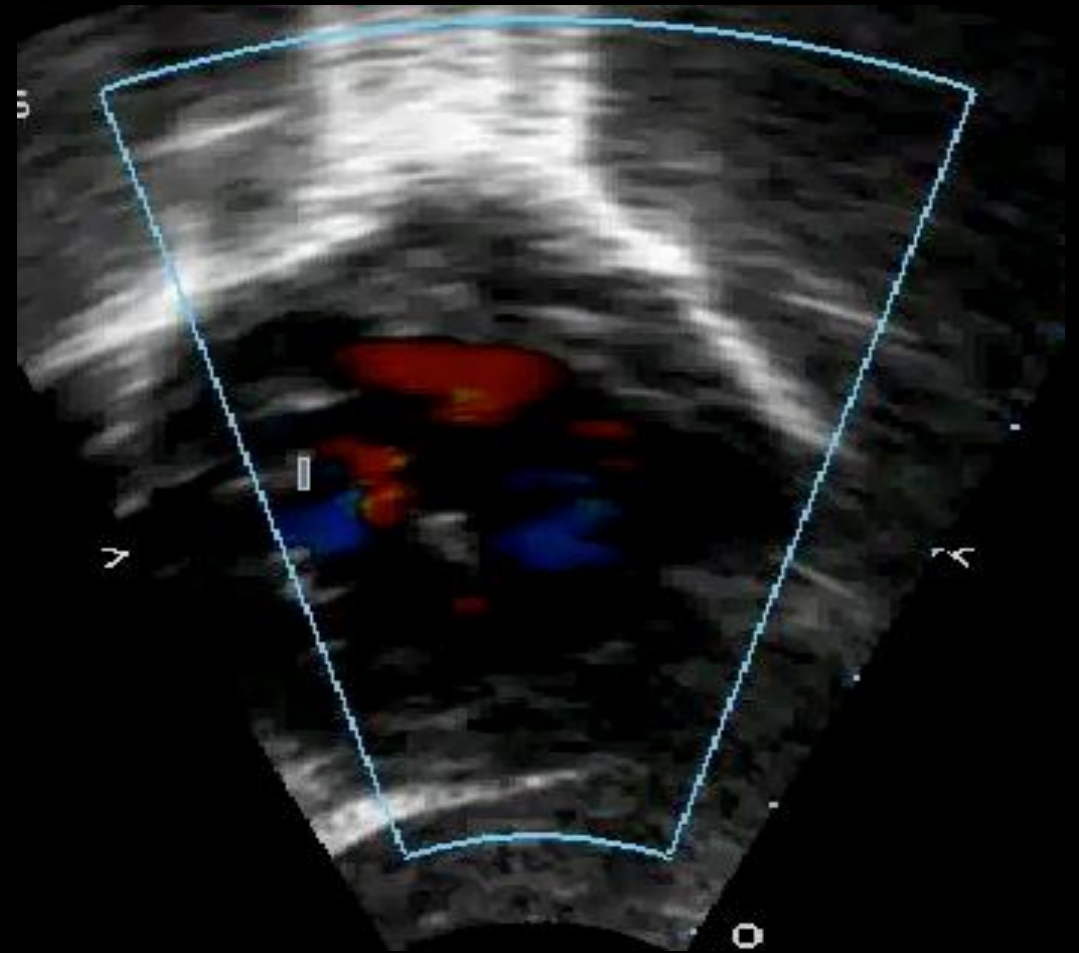
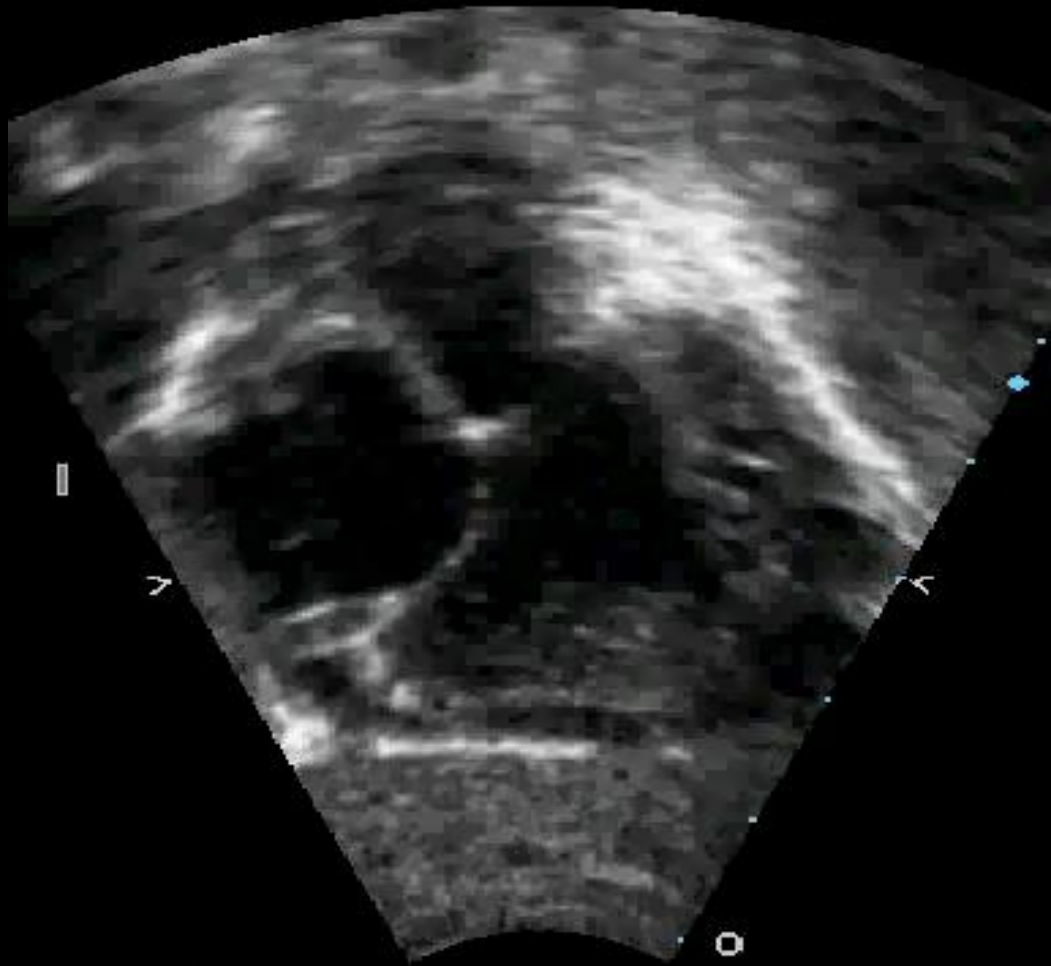


- Predominantly muscular
- ~50%: restrictive
- 21% restrictive (subaortic) : Coarctation and interrupted Ao arch association
- it can become restrictive

Bevilacqua M et al, JACC 1991

VSD remained the location, independently of the AV connection

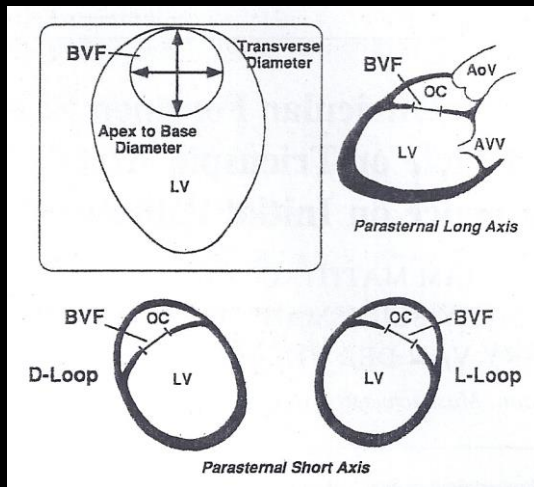
Ventricular septal defect ("Bulboventricular foramen")



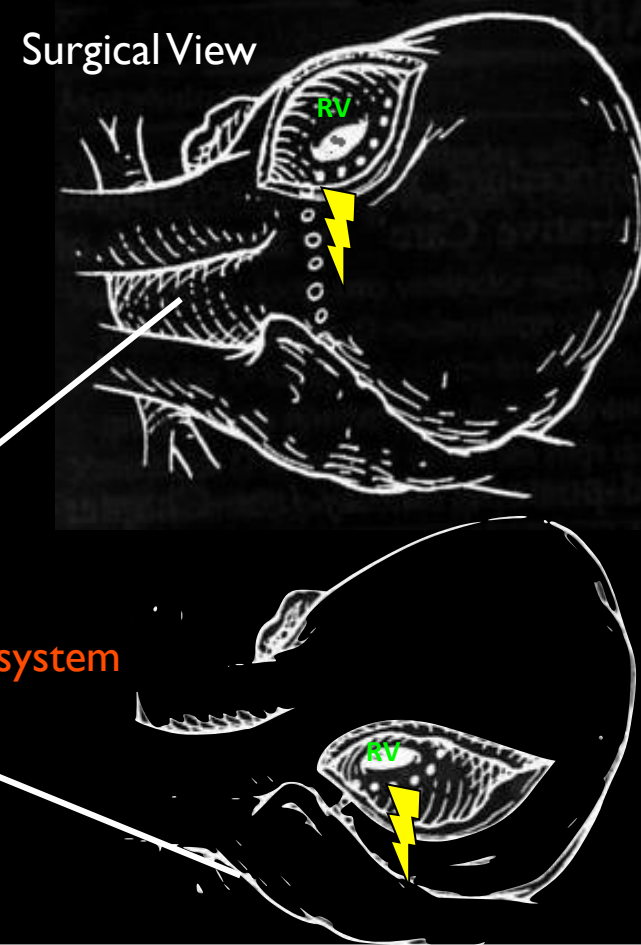
Ventricular septal defect : when it has to be enlarged?

Echocardiogram:

- Doppler gradient can orient but it is not the best predictor (overestimate in presence of PDA)
- Area by $2d < 2 \text{ cm}^2/\text{m}^2$: “high risk”



Matitiau A et al JACC 1992



Conduction system

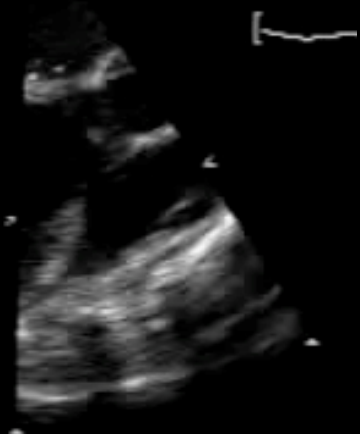
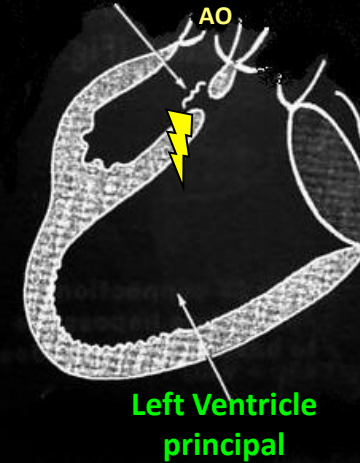
Double inlet of LV : the conduction system it is ALWAYS postero/inferior
no matter where is the rudimentary chamber

Ventricular septal defect

("Bulboventricular foramen")

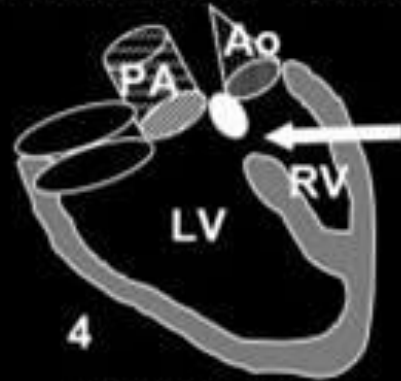


Restrictive VSD



Outlet obstruction in Double inlet of LV

aortic pathway obstruction 5 / 43



4
discordant VA



1
double outlet LV
(with mild overriding)

pulmonary pathway obstruction 19 / 43



2
Ao from RV with
pulmonary atresia

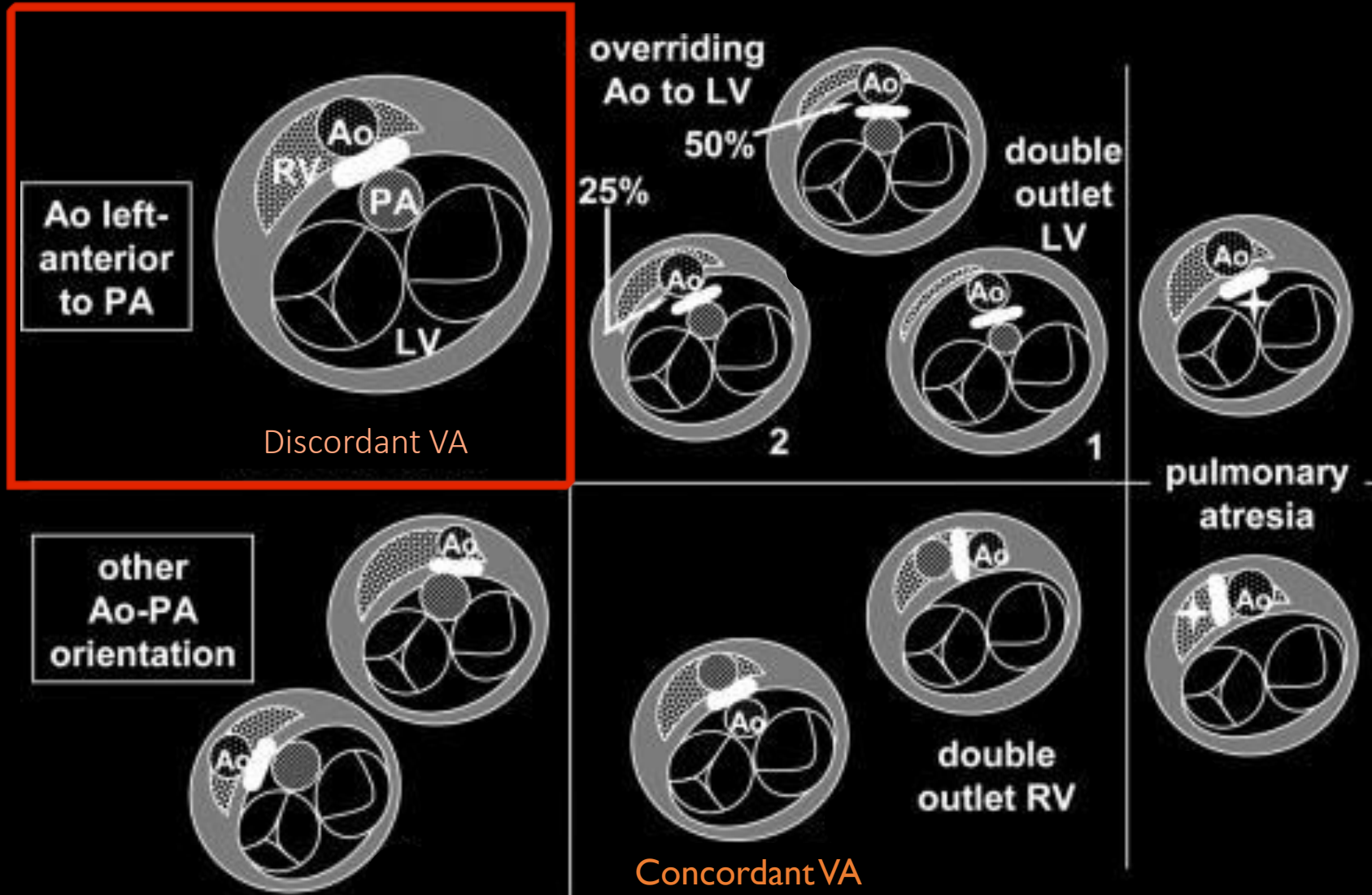


15
discordant VA with
pulmonary stenosis

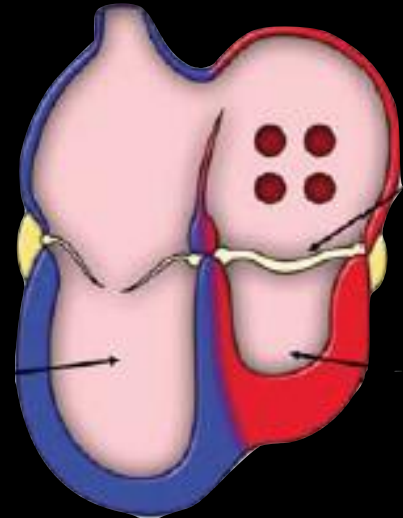
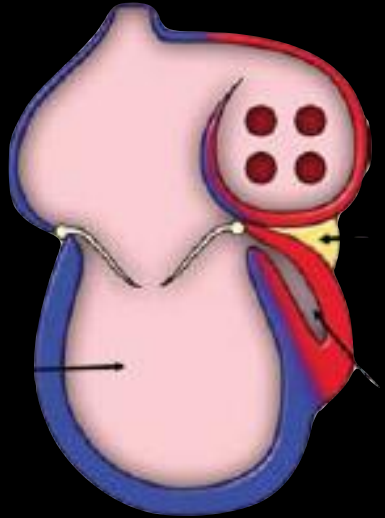


2
concordant VA with
pulmonary stenosis

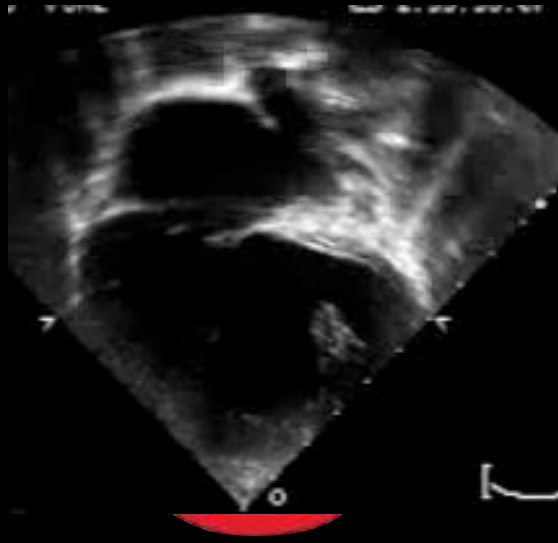
Great Vessels spatial relationship: double inlet LV



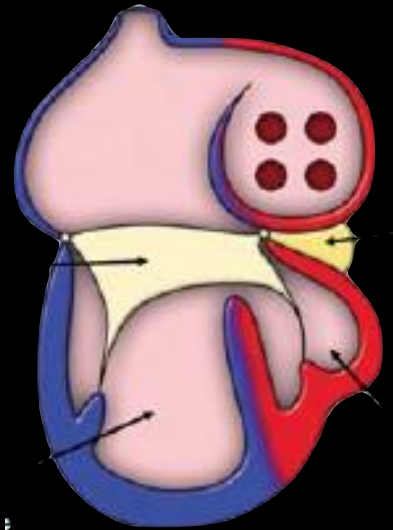
Absence of AV connection: where should be the rudimentary chamber?



(a)



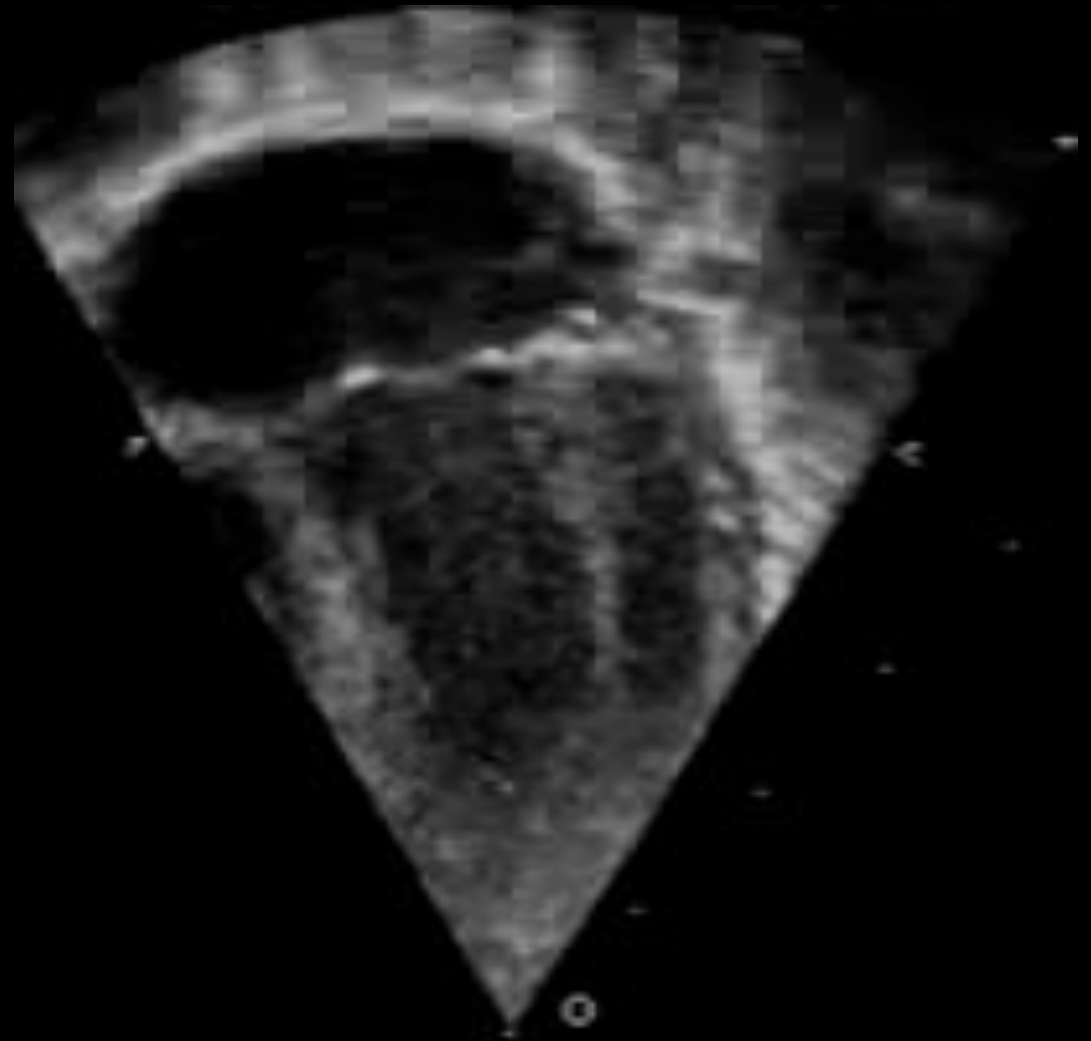
(b)



(c)

Absence of AV connection: where should be the rudimentary chamber?

Absence of left AV connection,
hypoplastic LV on the right side, VA
discordance, sub and Pulmonary
artery stenosis, VSD.....

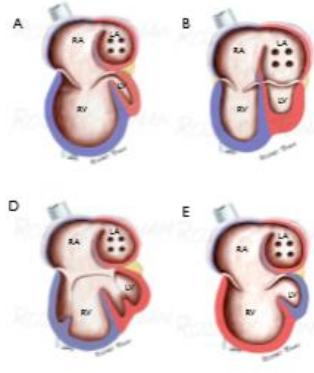


Unusual presentation of atrioventricular (AV) univentricular connection: Absent left AV connection with a dominant ventricle of left morphology. Anatomical and echocardiographic findings in four cases

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 (1) Tulane Hospital For Children, Tulane University, School of Medicine - New Orleans, LA, US
 (2) Heart Institute of University of São Paulo, School of Medicine - São Paulo, SP, Brazil
 vguerra@tulane.edu

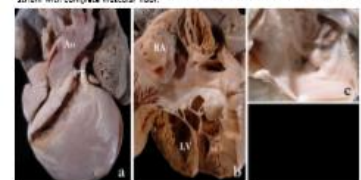
Introduction:
 Despite of existing a systematized nomenclature for all types of univentricular AV connection, there are still some cases challenging pediatric cardiologists/echocardiographers. Usually, absence of one AV connection is associated with the presence of a rudimentary ventricular chamber at the same side as the absent connection. Very rarely, the hypoplastic chamber is located at the contralateral side of the absent connection, with the dominant ventricle of unexpected morphology for the side of absent connection (see panel 1, cartoon C). We sought to clarify this unusual feature by reviewing a series of four cases, including one fetal diagnosis, with this morphological arrangement.

Phenotypes of absence of Left AV connection

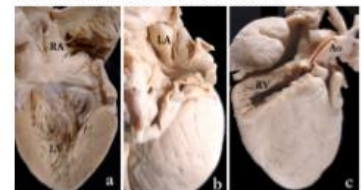


Transesophageal echocardiogram views from patient 1: a) subcostal view showing right atrium, restrictive PFO and hypoplastic left atrium; b) subcostal view, apical, showing left ventricle (LV) connected with Aorta (AO) and the rudimentary right ventricle (RV); c) Apical 4-chamber view showing the right atrium connected (RA) with left ventricle and the hypoplastic RV on the right side.

Grass view of the heart from patient 1 showing in a) Anterior view with the right-sided rudimentary right ventricle connected to the pulmonary trunk; b) morphologically left atrium connected to the left ventricle via a diaphragmatic atrial valve; c) morphologically left atrium with complete muscular floor.



Grass view of the heart from patient 2 showing in a) the morphologically right atrium connected to the left ventricle; b) left atrium with muscular floor; c) anterior view showing right-sided rudimentary right ventricle giving rise to the aorta.



Material & Methods:

From the hospital database/chart, data were obtained. Echocardiogram studies were reviewed and correlation with anatomical features from pathological study was done.

Four cases were studied with absence of left AV connection with a dominant left ventricle and rudimentary right ventricular chamber located at the right side. Three specimens were examined from patients who died. The remaining patient has been followed after a palliative surgery.

Results:

There was agreement between the initial echocardiogram report and pathological features in two cases. In the two remaining, the initial diagnosis was "Tricuspid atresia", including one fetal diagnosis.

In all cases there was an early clinical presentation with hypoxia and low cardiac output. Right outflow tract obstruction was detected in two patients. Two patients required atrial septostomy for a restrictive PFO. In the remaining patient there was a small PFO, but there was total anomalous pulmonary venous return to the coronary sinus.

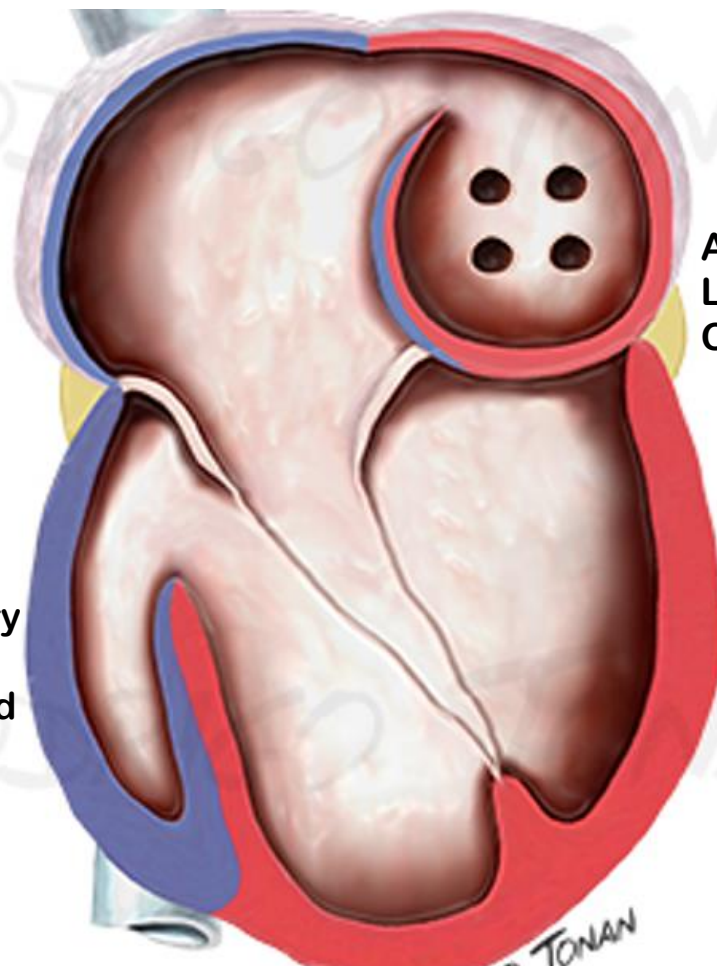
Main echocardiographic features:

| Patient | ASD | VSD | Ventriculo-arterial connections | Location of rudimentary chamber | RVOTO | TAPVD |
|---------|-----------------|--------------------------|---------------------------------|---------------------------------|-------|-------|
| 1 | PFO | Muscular restrictive | concordant | Right | yes | No |
| 2 | Restrictive PFO | Muscular restrictive | discordant | Right | No | No |
| 3 | Secundum ASD | Non restrictive muscular | concordant | Right | No | No |
| 4 | PFO | Non restrictive muscular | concordant | Right | yes | Yes |

Conclusions:

Echocardiographers must be aware about unusual forms of AV connection. The correct interpretation of absent AV connection by standard views needs a dynamic evaluation and the combination of different views to establish the correct spatial orientation of cardiac chambers and determination of ventricular morphology. Fetal diagnosis requires more views and careful interpretation as well. The sequential segmental analysis is fundamental to achieve the correct diagnosis, both by echocardiography and other morphological methods.

Unusual Atrioventricular Univentricular Connection



Absence Left AV Connection

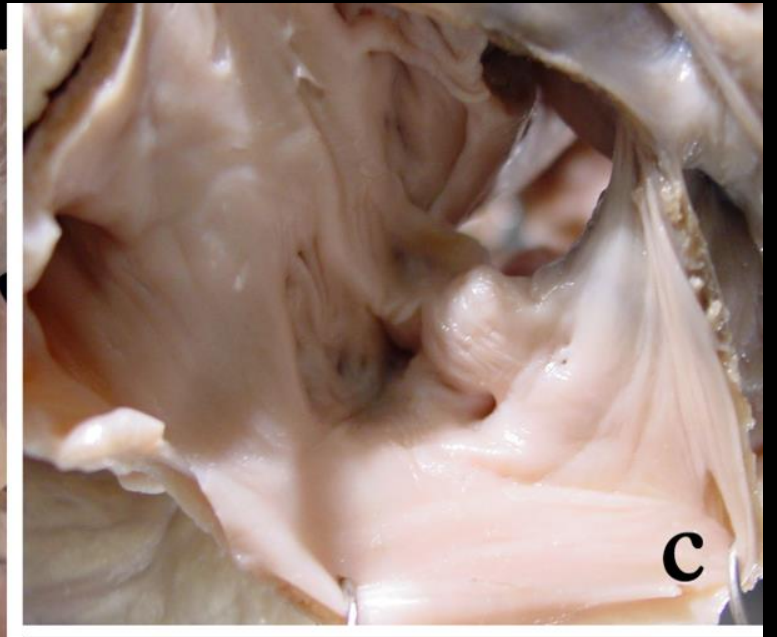
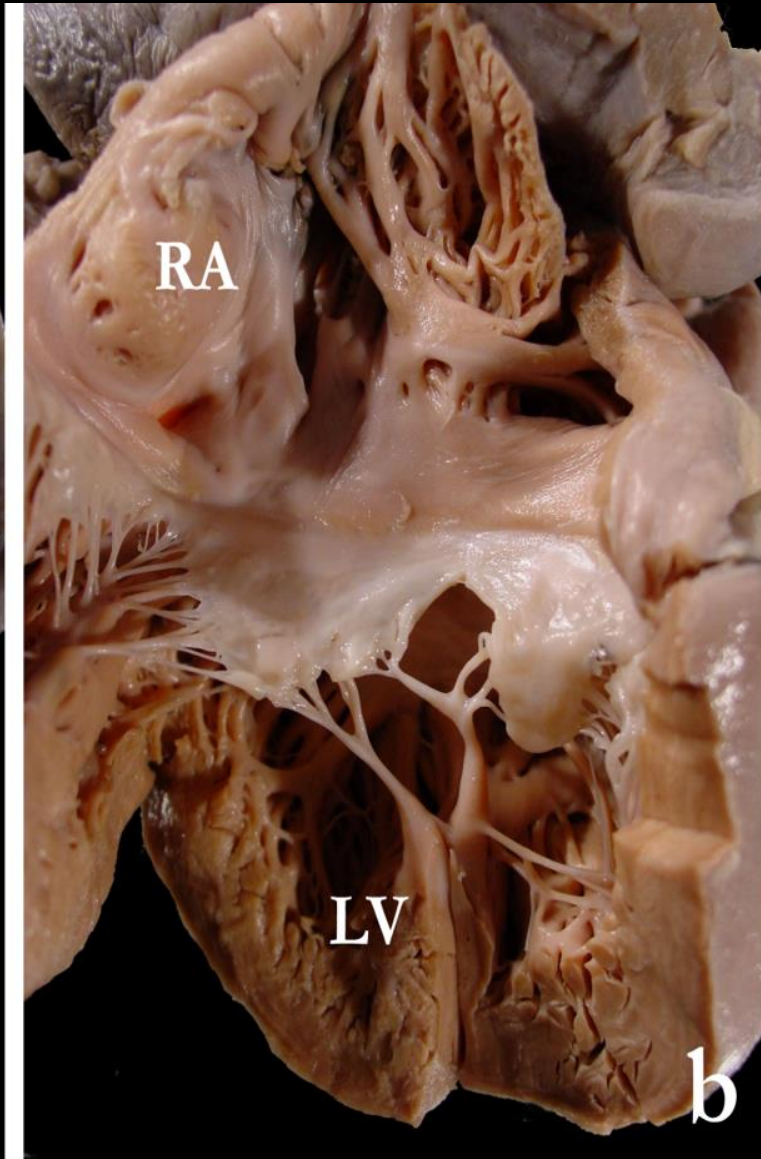
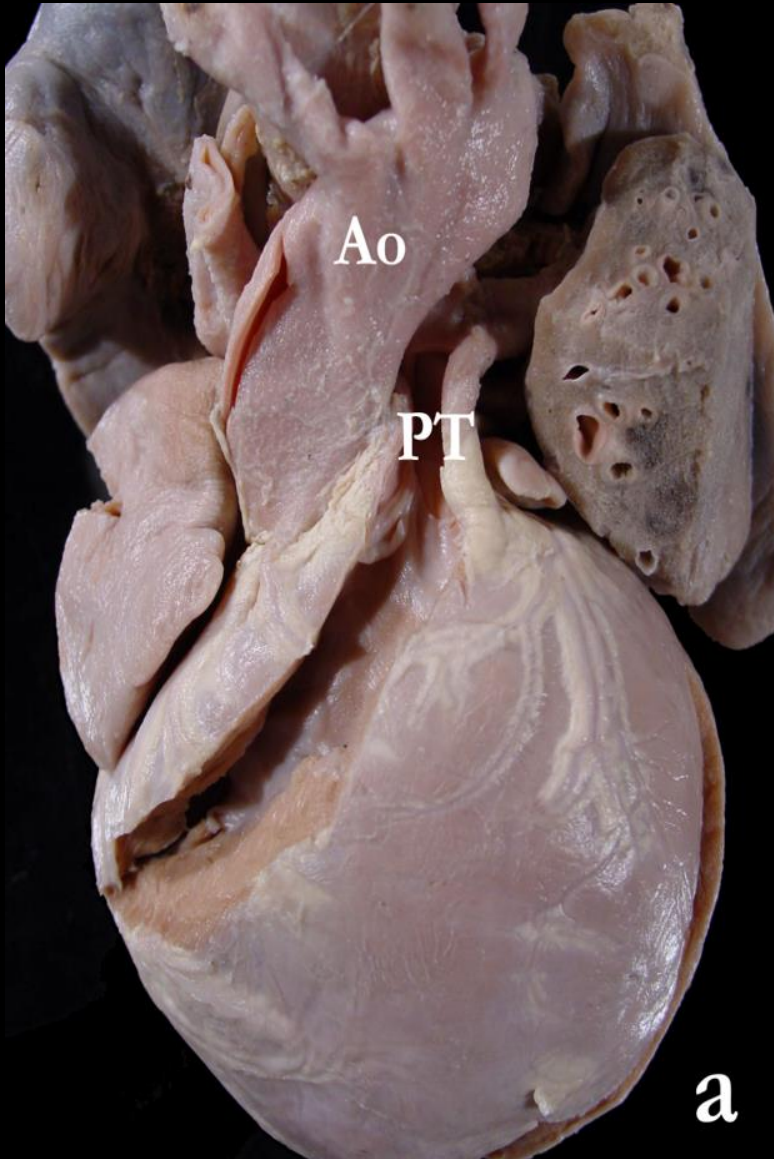
Rudimentary Chamber Anterior and right

RODRIGO TANAN

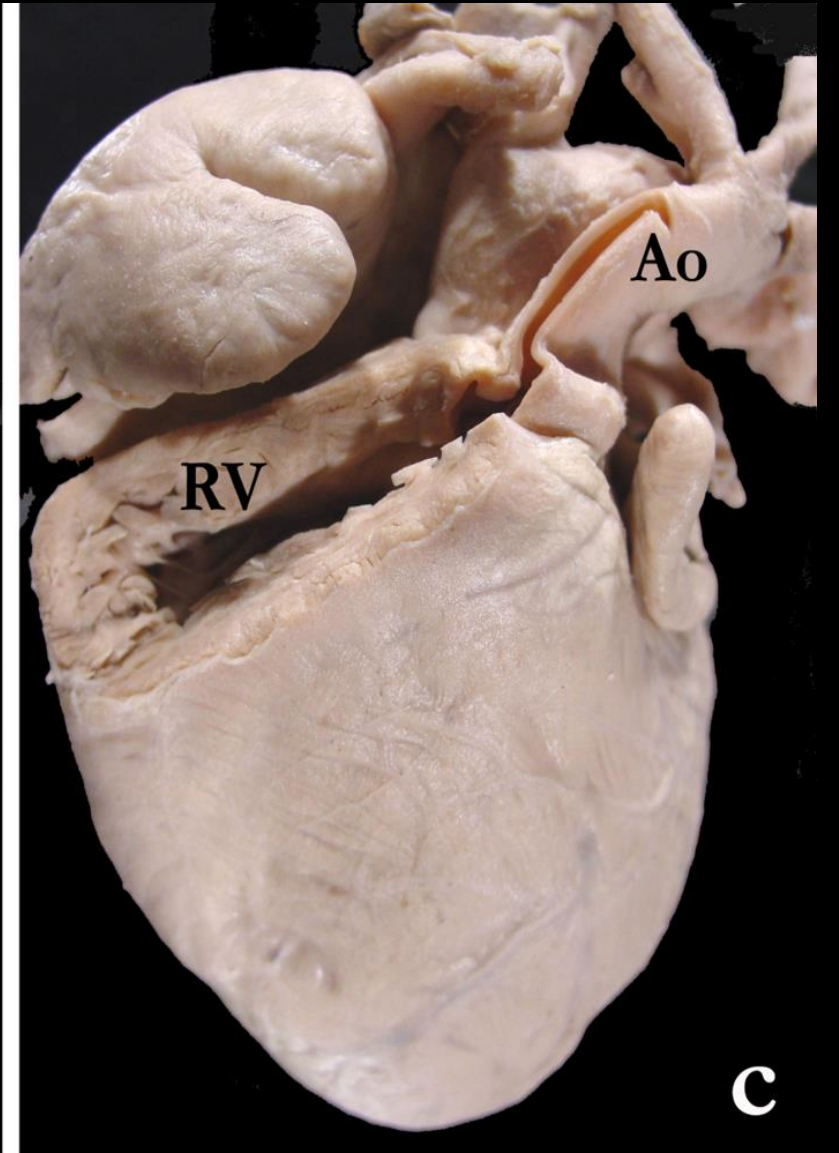
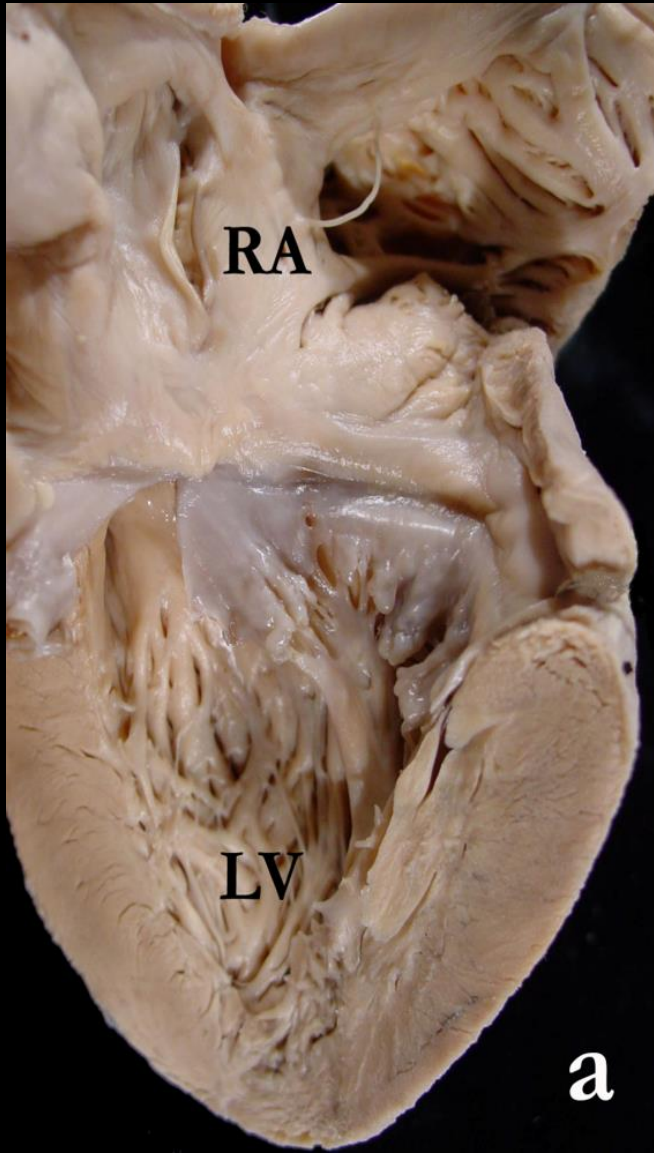


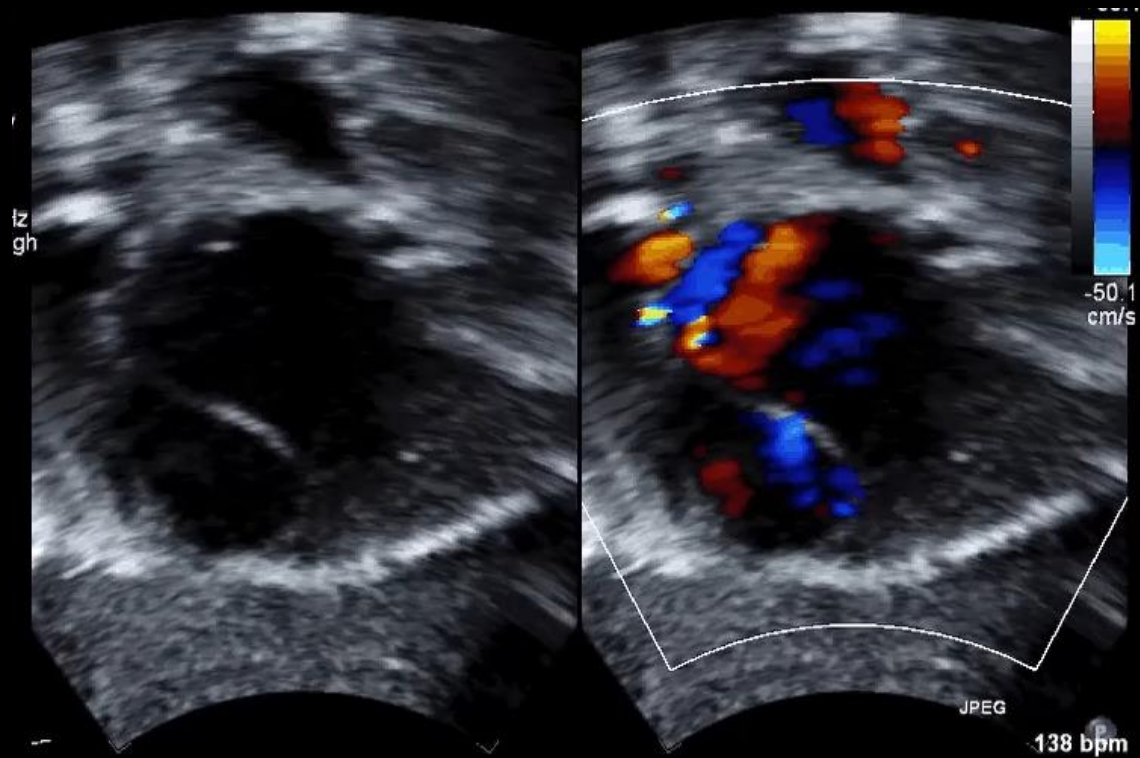
Ausência de conexão AV à esquerda
VE principal

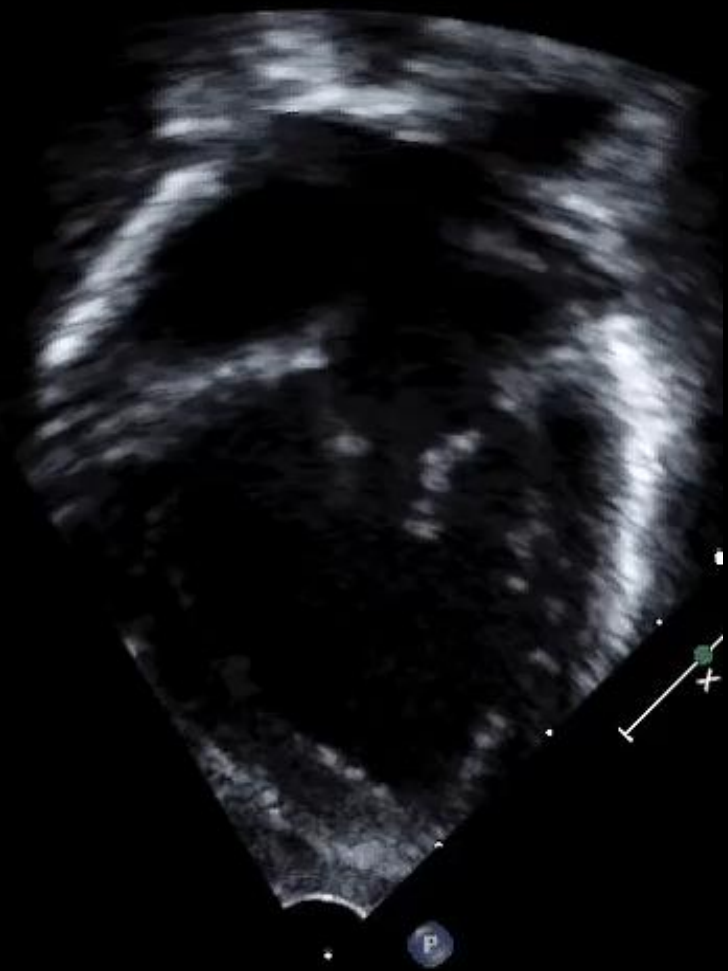
**Absence of Left AV connection & Discordant AV Connection
(rudimentary chamber located anterior and right) & VA Concordance**



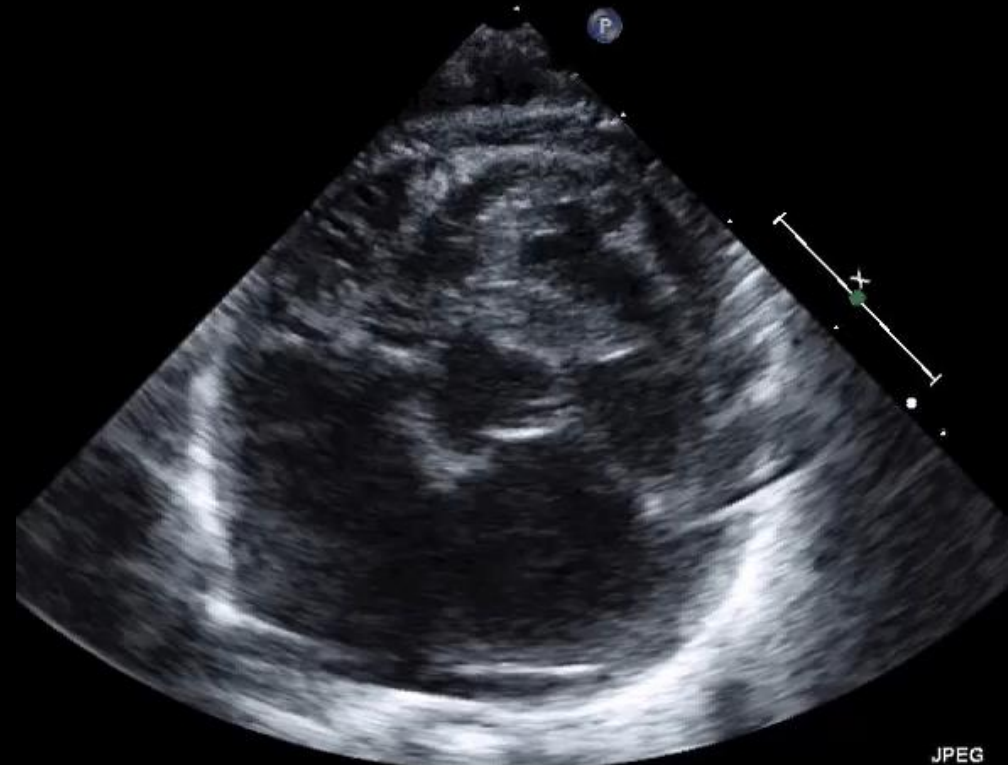
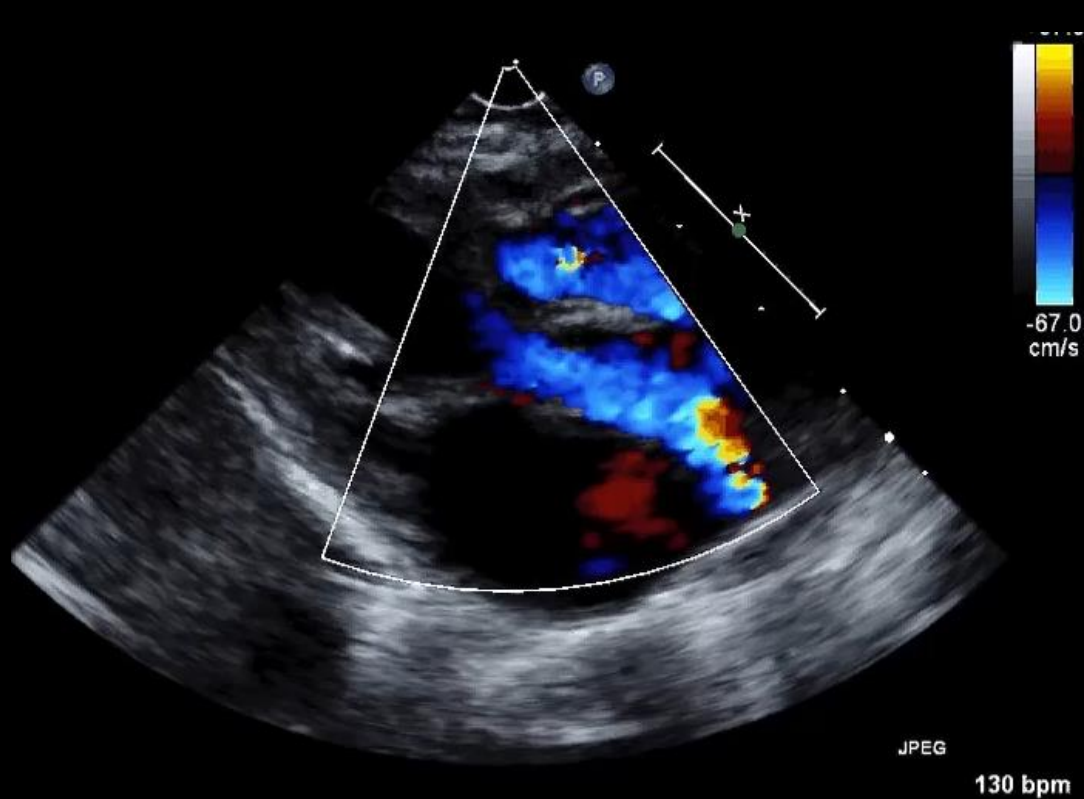
**Absence of Left AV connection & Discordant AV Connection
(rudimentary chamber located anterior and right) & VA Discordance**



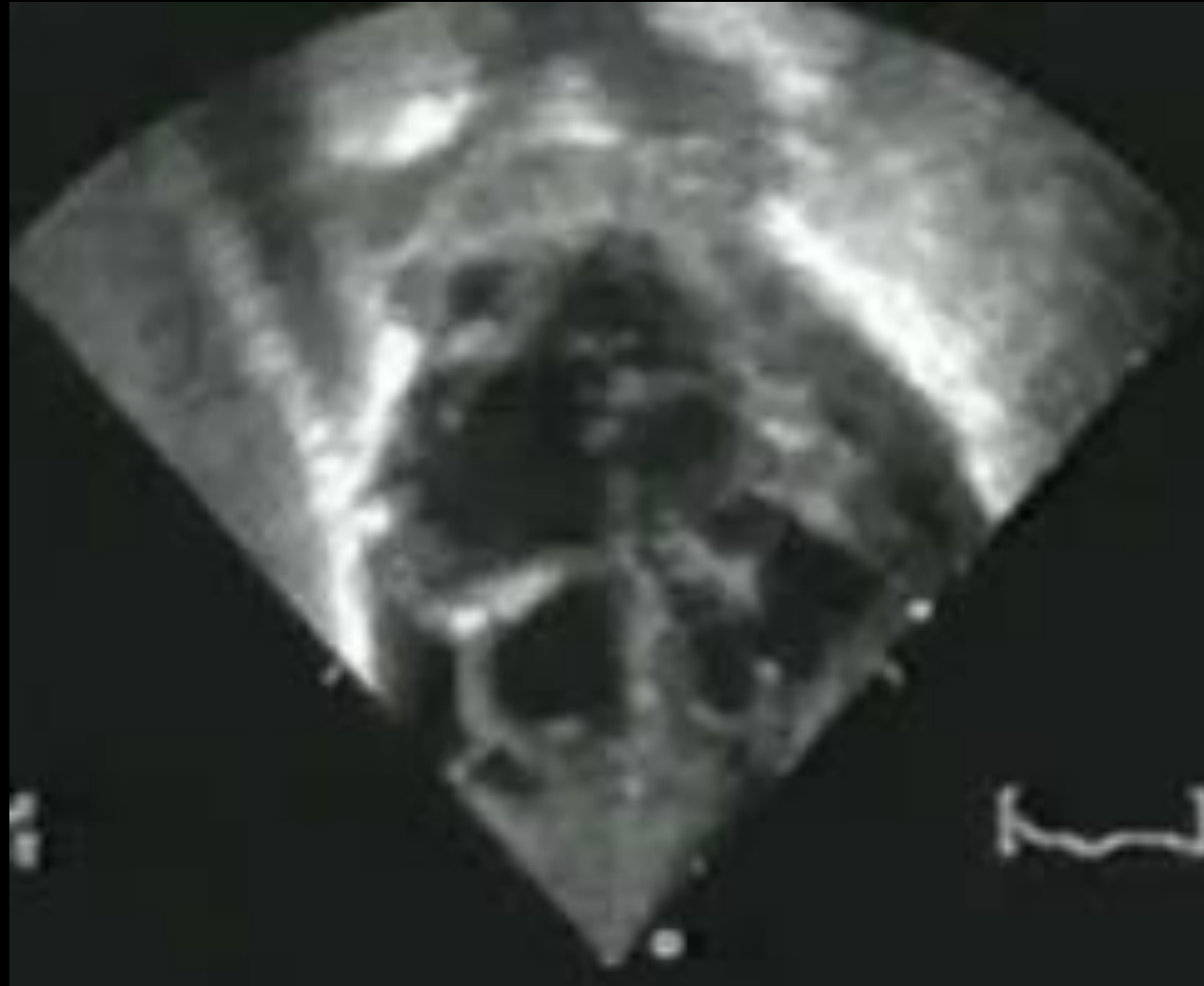




Absence of Right AV connection & Rudimentary chamber located anterior and left) & VA Concordance

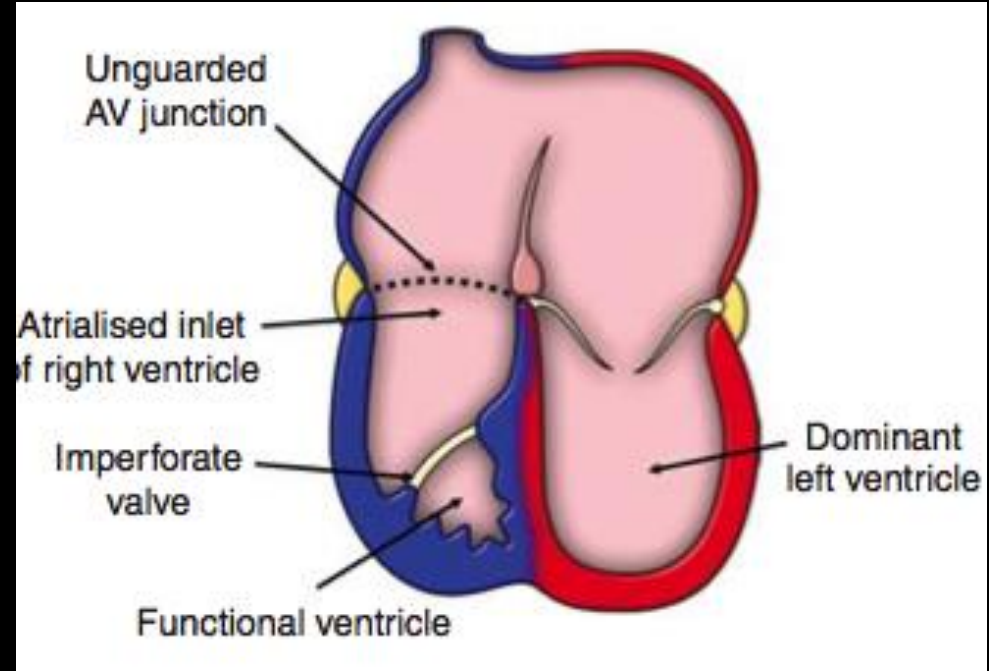
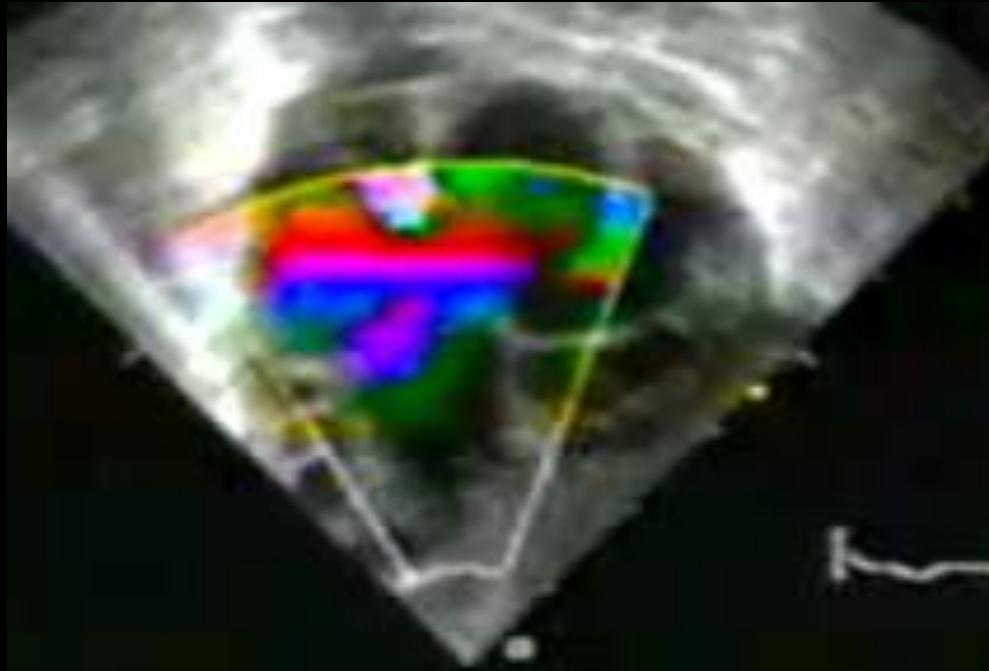


What kind of AV connection?



What kind of AV connection?





ORIFÍCIO MITRAL "DESGUARNECIDO" ASSOCIADO À DISCORDÂNCIA ATRIOVENTRICULAR E VIA DE SAÍDA ÚNICA AÓRTICA DO VENTRÍCULO DIREITO



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 INSTITUTO DO CORAÇÃO (InCor) – HCFMUSP

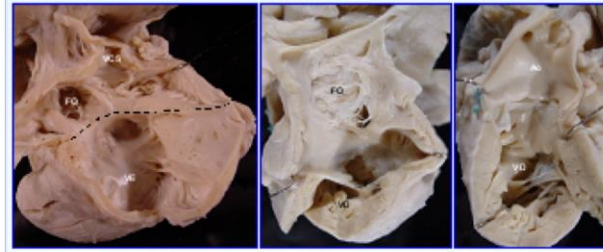
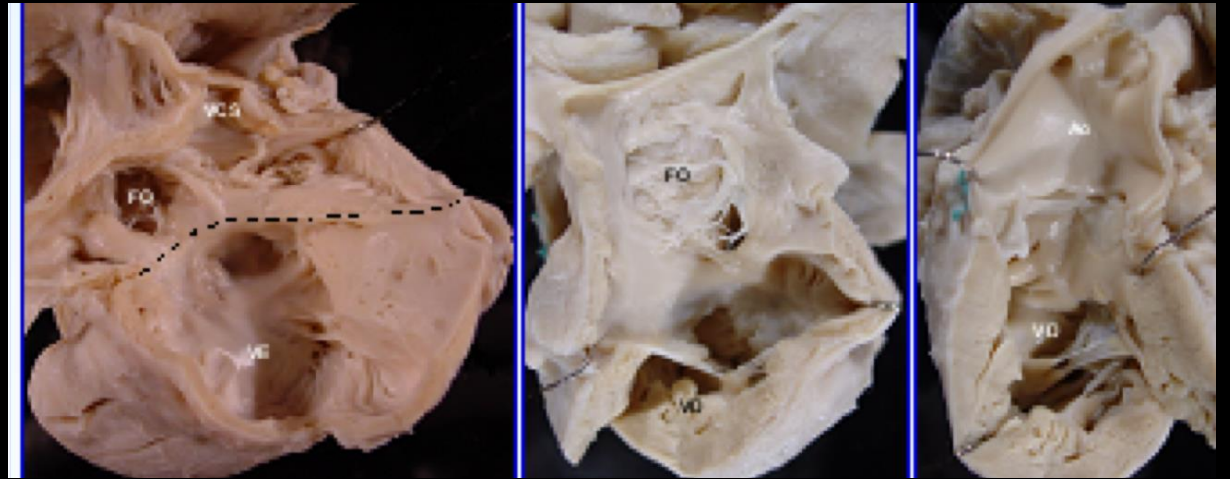


RELATO DE CASO

- ✓ J.N.O, feminino, nascida a termo, com peso de nascimento de 2.165g
- ✓ Evoluiu no 1º dia de vida com ausculta de B2 única, hiperfonética e sopro contínuo.
- ✓ Apresentou desconforto respiratório, com hipoxemia, sendo realizada intubação orotraqueal e administração de Prostin e drogas vasoativas
 - RX Tórax – situs solitus em dextrocardia
 - ECG – onda Q em V1 sugerindo inversão ventricular
- ECO:
 - Situs solitus em dextrocardia
 - Atresia Pulmonar + CIV
 - PCA
- ✓ Admitido no InCor com 11 dias de vida e peso de 2.420 g em regular estado geral, cianose leve (SatO2 86 – 95%), fígado a 2 cm do rebordo costal direito e ausculta cardíaca com B2 única e hiperfonética e sopro contínuo
- ✓ ECO InCor:
 - Situs solitus em mesocardia
 - Aneurisma da fossa oval com comunicação medindo 4,5mm
 - Conexão atrioventricular discordante
 - Via de saída única aórtica do VD, que encontra-se à esquerda
 - Ebstein severo da valva mitral ?
 - CIV's apicais musculares

- ✓ Realizado Blalock modificado (no tronco braqu岸o-cefálico) à direita e ligadura do canal arterial com 17 dias de vida
- ✓ Evoluiu com baixo débito importante, sendo iniciada diálise peritoneal (cateter de Tenckhoff), com piora hemodinâmica e ventilatória progressivas e quadro de abdômen agudo → Enterocolite Necrotizante
- ✓ Indicada Laparotomia exploradora, que observou necrose e perfuração intestinal. Durante procedimento cirúrgico apresentou parada cardiorrespiratória não responsiva às manobras de ressuscitação → óbito
- ✓ Necropsia:
 - Situs solitus. Conexão atrioventricular biventricular e discordante:
 - Orifício mitral desguarnecido
 - Ventrículo direito dilatado, situado à esquerda
 - Valva tricúspide com anel dilatado
 - Átmo morfologicamente direito bastante dilatado. Átmo esquerdo habitual, em conexão com o VD
 - CIA tipo fossa oval, fenestrada e CIA tipo seio venoso (7 mm). Drenagem venosa pulmonar e sistêmica habituais
 - Conexão ventriculoarterial do tipo via de saída única
 - Aorta anterior saindo do VD
 - Atresia da valva pulmonar
 - Tronco pulmonar de calibre normal, atrás da aorta, iniciando em fundo cego
 - Blalock - Taussig modificado à direita, pérvio
 - Ligadura do canal arterial
 - Estenose da artéria pulmonar esquerda

Unguarded "Mitral" valve orifice & Discordant Atrioventricular connection & Discordant ventricular Arterial connection



COMENTÁRIOS

Os primeiros relatos desta rara malformação cardíaca foram descritos por Yasukochi em 1999, através de dois casos de orifício desguarnecido de valva mitral, com situs inversus, discordância atrioventricular, dupla via de saída de VD, CIA ampla, CIV pequena e atresia pulmonar, com artérias pulmonares confluentes e supridas por canal arterial. Caso semelhante foi descrito em 2003 por Earing e colaboradores, diferindo apenas pela ausência de CIV e situs ambiguus abdominal (isomerismo direito). No InCor, em 2005 relatamos caso de orifício desguarnecido de valva mitral com discordância atrioventricular, via de saída única aórtica do VD, com especial atenção para o situs solitus. Em todos os casos o ventrículo esquerdo apresentava paredes finas, ausência de folhetos da valva mitral e do aparato subvalvar. A etiologia desta malformação é ainda desconhecida, embora a apoptose tenha sido sugerida como possível mecanismo, assim como na doença de Uhl; Mas isso não passa de especulação. Novas investigações estão sendo realizadas para esclarecer a sua exata patogênese

REFERÊNCIAS BIBLIOGRÁFICAS

1. Yasukochi S., Satomi G., Park I., Ando M., Momma K. (1999). Unguarded mitral orifice, mirror-imaged atrial arrangement and discordant atrioventricular connections. *Cardiol young* 9 : 478- 483
2. Earing, M.G., Edwards W. D., Page F. J., Cabral A. K. (2003). Unguarded Mitral Orifice Associated with discordant atrioventricular connection, double-outlet right ventricle and pulmonary atresia. *Pediatric Cardiology* : 490 - 492

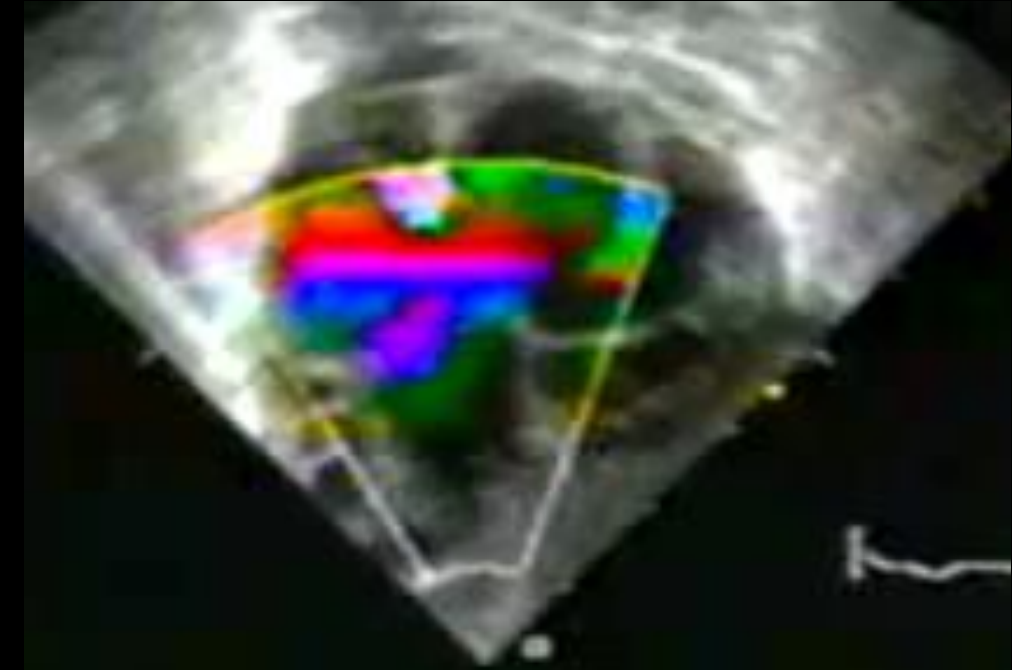


Figura 1: Vista subcostal
Situs solitus



Figura 2: Vista Apical 4 câmaras, mostrando a valva tricúspide e orifício desguarnecido da valva mitral

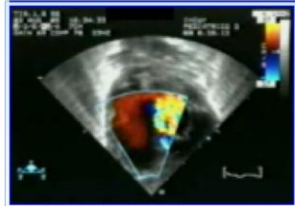


Figura 3: Vista Apical com mapeamento de fluxo a cone. Fluxo retrógrado através do orifício mitral



Figura 4: Vista Apical com mapeamento de fluxo a cone. Fluxo anterógrado através do orifício mitral

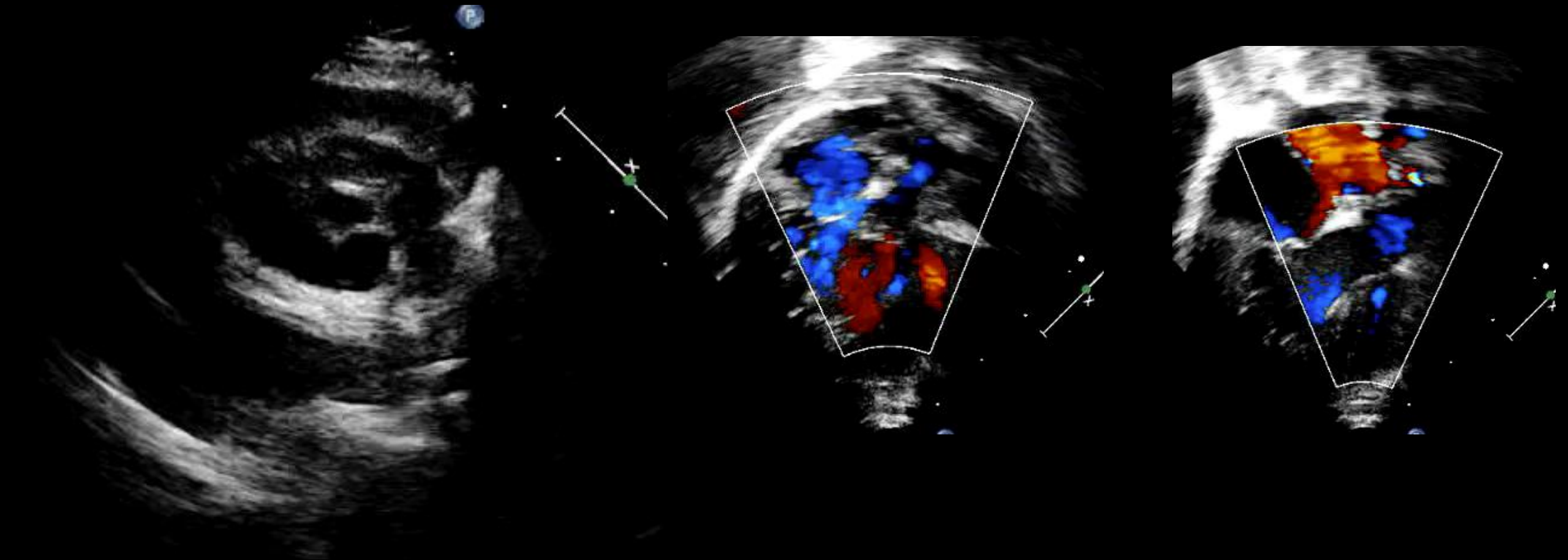


Figura 5: Vista Parasternal Longitudinal : Via de saída única aórtica do VD

Great Vessels spatial relationship:

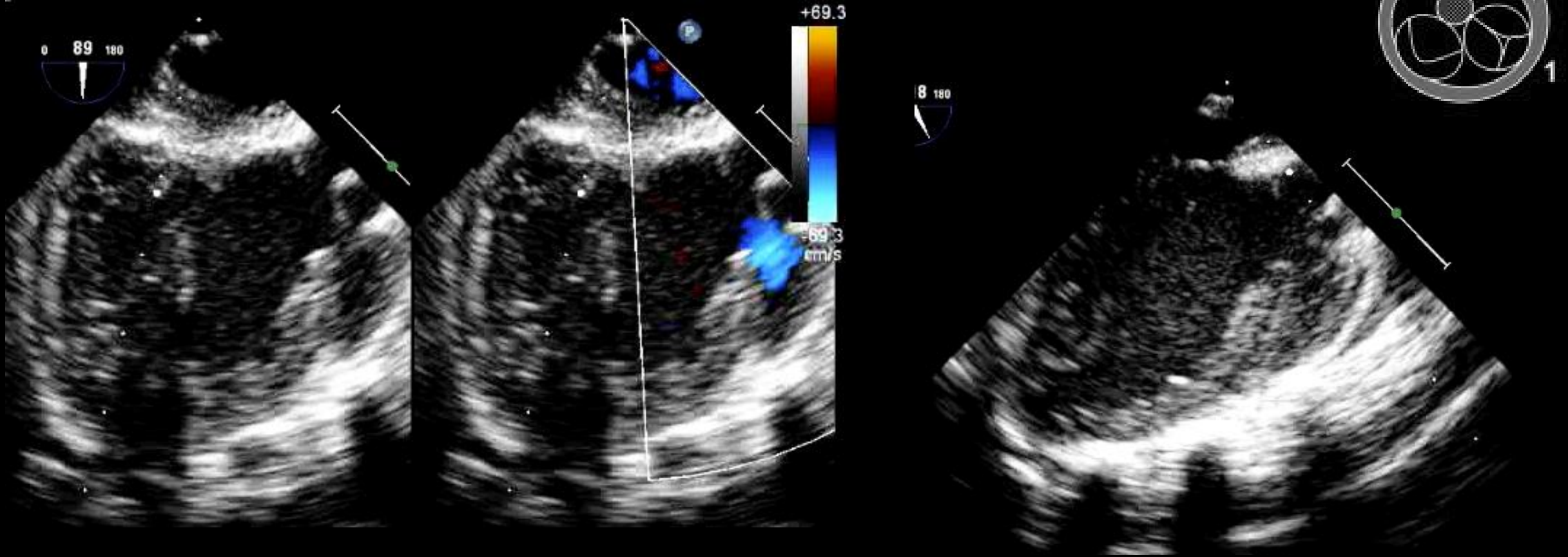
double inlet LV

Double outlet of LV?

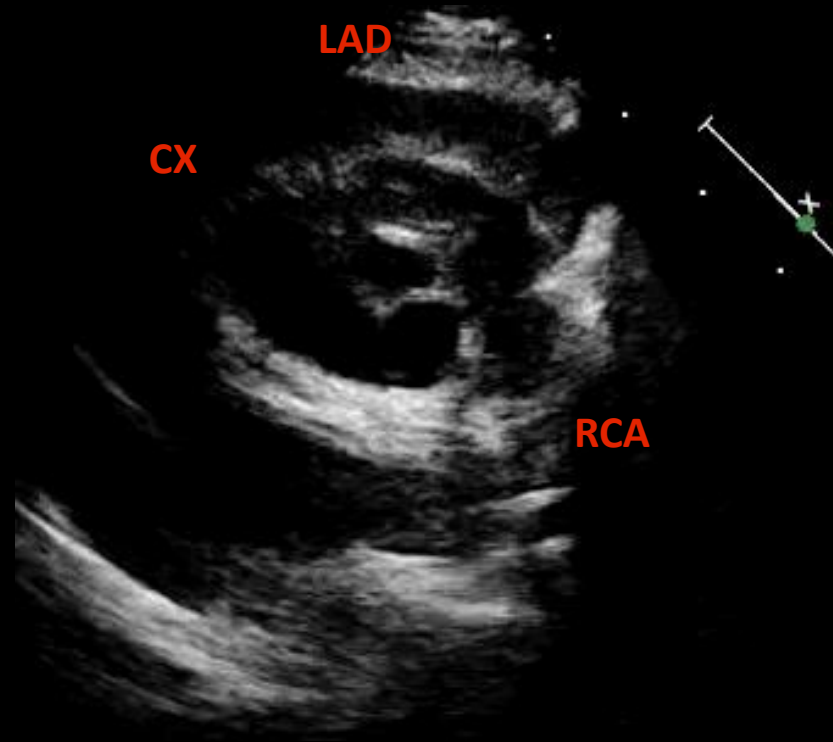
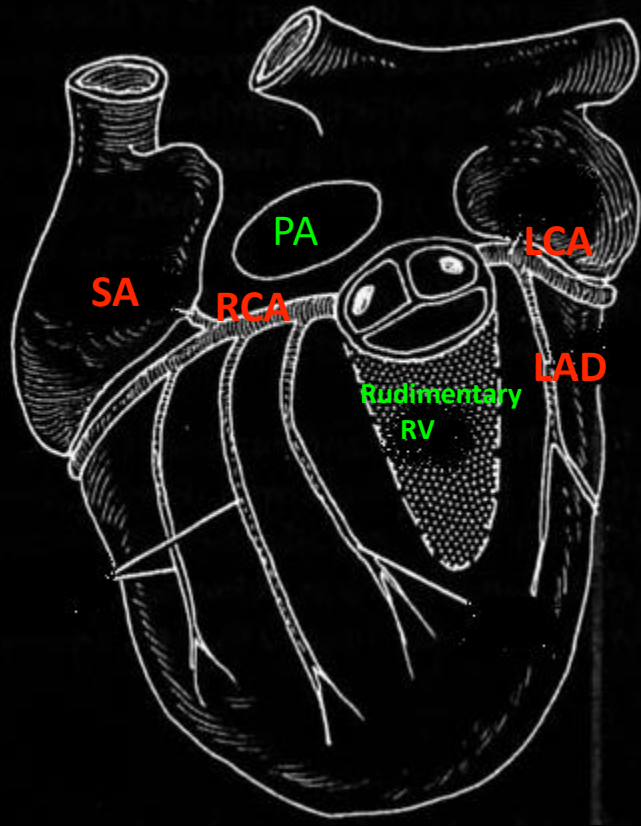


Great Vessels spatial relationship: double inlet LV

Double inlet & outlet of LV (25% overriding aorta)



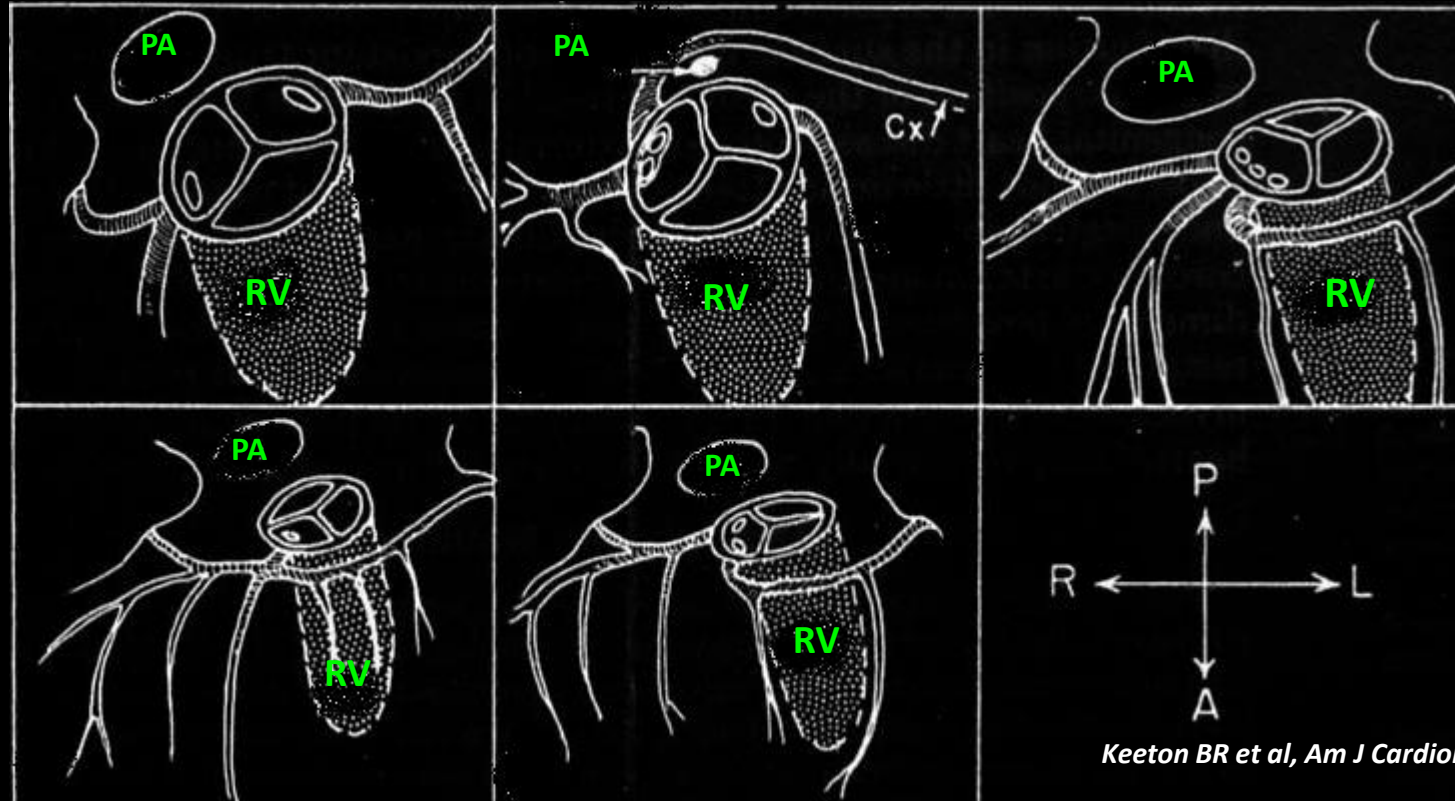
Coronary arteries: Univentricular AV connection



Main Ventricle Left Morphology &
Rudimentary chamber at left sided & ventricular-arterial discordant

Coronary arteries: Univentricular AV connection

Unusual pattern



Keeton BR et al, Am J Cardiol 1979

Main Ventricle Left Morphology &
Rudimentary chamber at left sided & ventricular-arterial discordant

Conduction System: Univentricular Atrioventricular Connections

Main Ventricle: Left

Anomalous anterosuperior or lateral atrioventricular node;

Relationship of the bundle & VSD borders depends on the position of the rudimentary chamber;

Main Ventricle: Right

Ventricular septum does reach the crux cordis if the main ventricle is RIGHT morphology

Regular position of AV node

Undetermined Main Ventricle

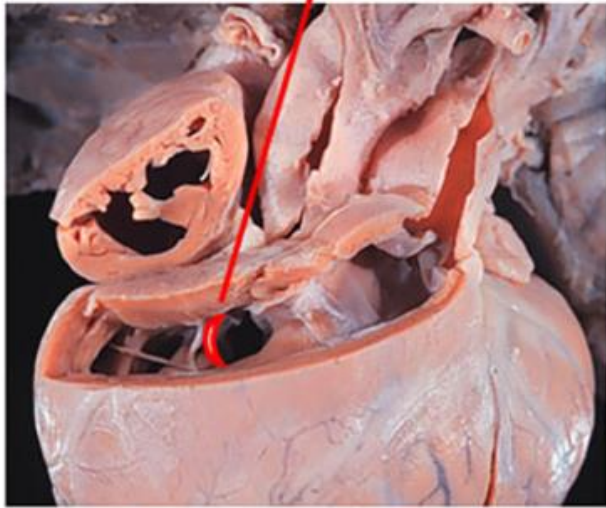
Variable position of AV node and bundle in solitary ventricle

INDETERMINATE morphology

.

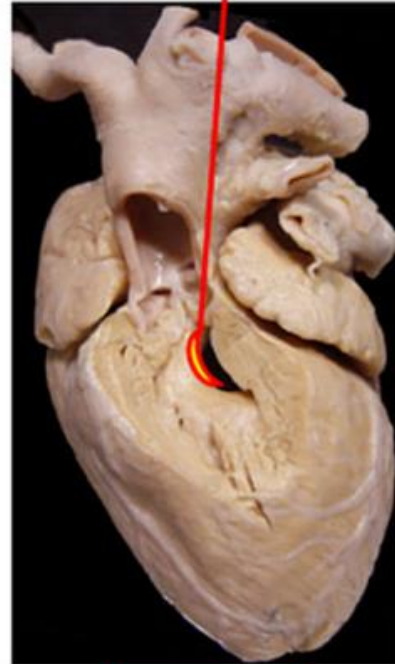
Conduction System: Univentricular Atrioventricular Connections

Postero-inferior bundle



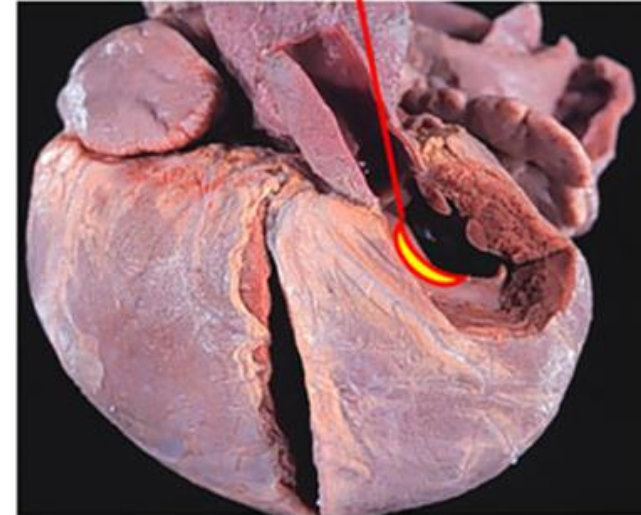
Right-sided

Anterior and Right-sided bundle



Anterior

Antero-superior bundle



Left-sided

Position of the Rudimentary Right Ventricle

From review article : Prof .Vera Aiello

World Journal for Pediatric and Congenital Heart Surgery 2015, Vol. 6(2) 239-249

Take Home message: Univentricular atrioventricular connections

1

Echocardiogram & Pathology share the same approach assessing Univentricular AV connection (= segmentar sequential analysis)

2

Atrioventricular valve(s): morphology and functional assessment is essential for Univentricular repair outcomes (mandatory detailed description by echo).

3

Ventricle Morphology: follow the steps identifying the rudimentary chamber (spatial relationship), trabecular septum, AVV and great vessels fibrous continuity (or not).



4

Ventricular septal defect: can play essential role in the clinical presentation and further surgical approach (be careful evaluating size and gradient by Doppler).

5

Unusual AV Connections : Rudimentary chamber doesn't follow the absence of AV connection side



Special thanks & Disclosure



“There are three methods to obtain wisdom: first, by reflection, the noblest;
Second, by imitation, the easiest;
and the third, by experience, the most bitter.”

Confucius- Chinese
(551-479 BC)

And the fourth way: always be around [Dr.Vera Aiello](#)
... can be virtual, by Skype, Facetime....

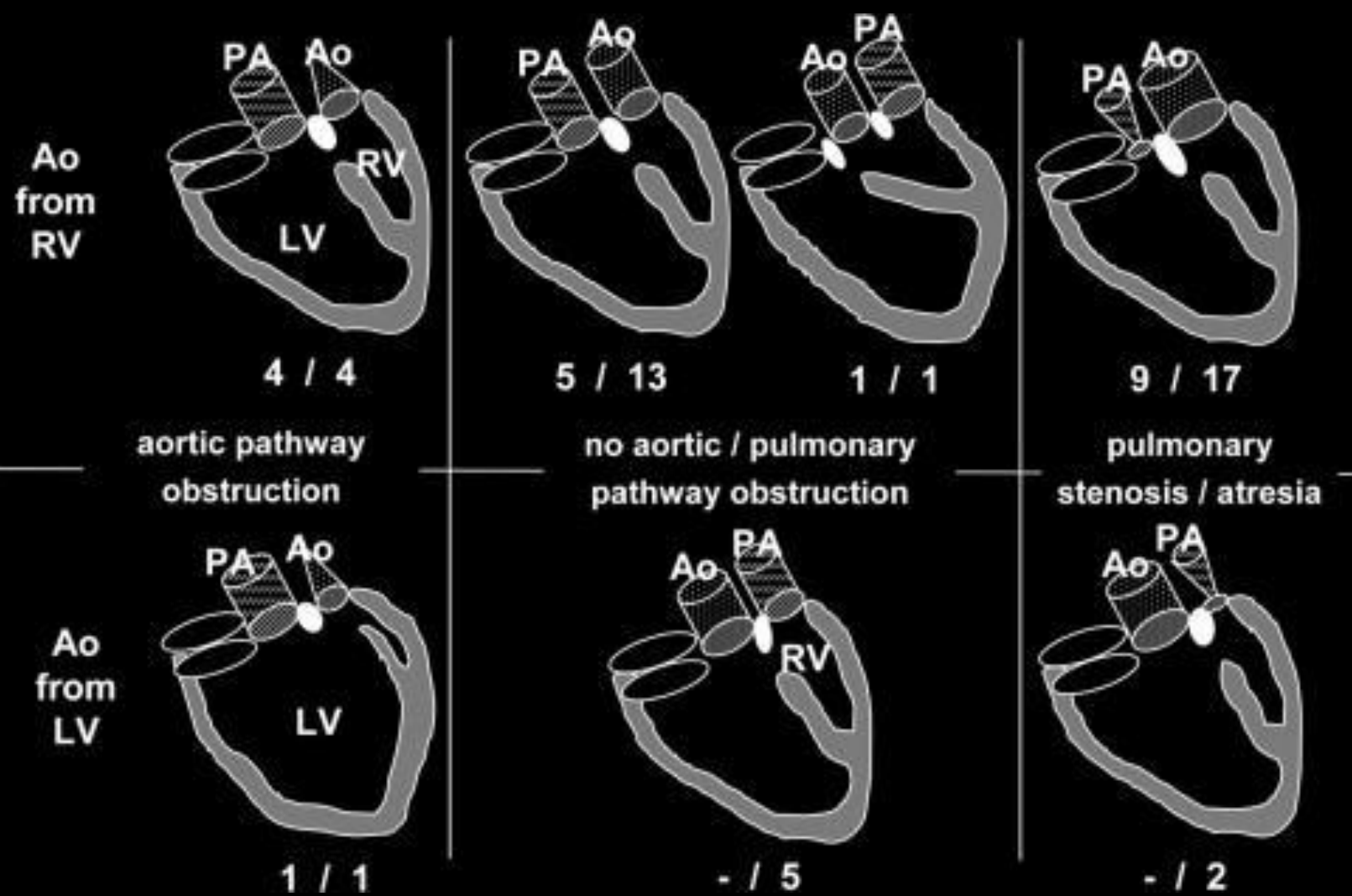
Vitor Guerra.



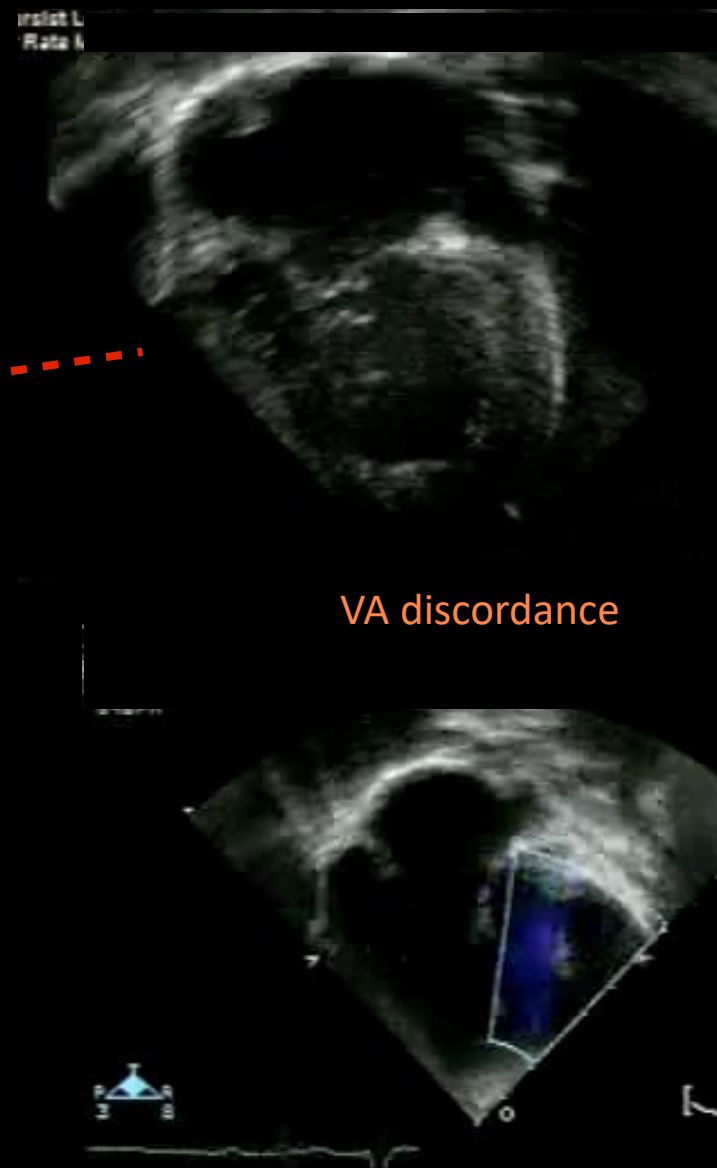
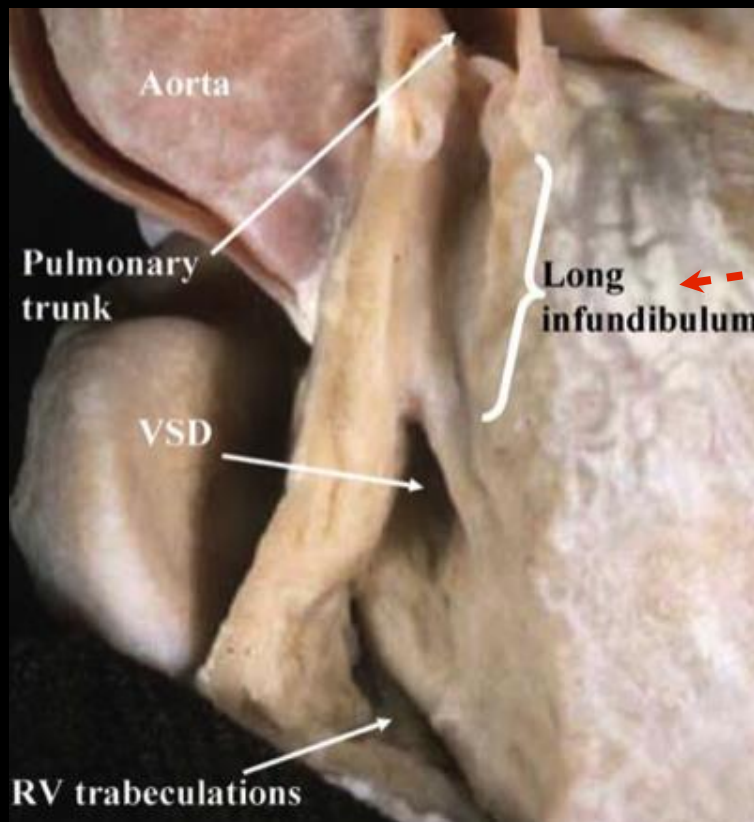


“You see only what you look for .
You recognize only what you Know”

Merril C. Sosman



The Outlets in Univentricular AV connection





overriding Ao to LV

50%

25%

25%

25%



double outlet LV



pulmonary atresia

other Ao-PA orientation



double outlet RV



Great Vessels spatial relationship: double inlet LV

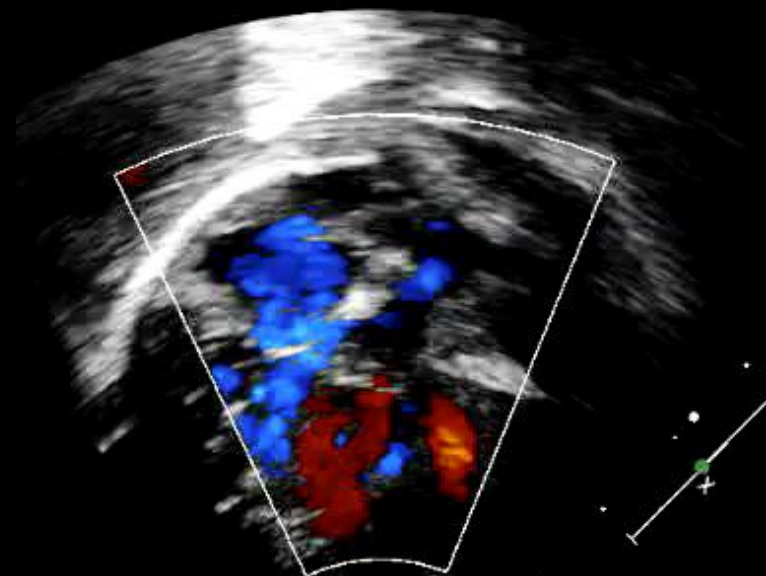
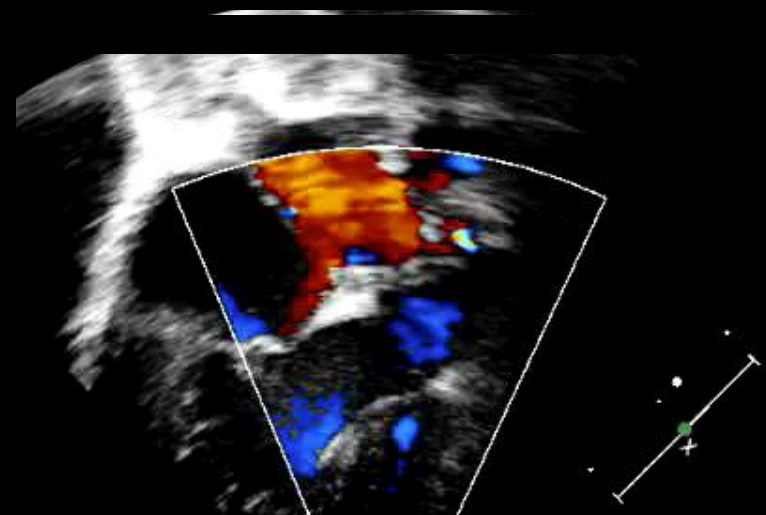
Lossy compression - not

D0011

FR 51Hz
8.1cm

2D
59%
C 59
P Off
HRes

P (R)
2.8 6.6





LUIZ EDUARDO RODRIGUES 30 Aug 06 Tts 1.8 MI 1.1
INCOR PS-3 16mm P_Card/Gen 12:23:40 40 Hz 8.5 c

sp 8
mRg 50dB
instat Low
Rate Med

V0 Pg 0
H0 Pg 0



Main Ventricle Left Morphology &
Rudimentary chamber at left sided & ventricular-arterial discordant

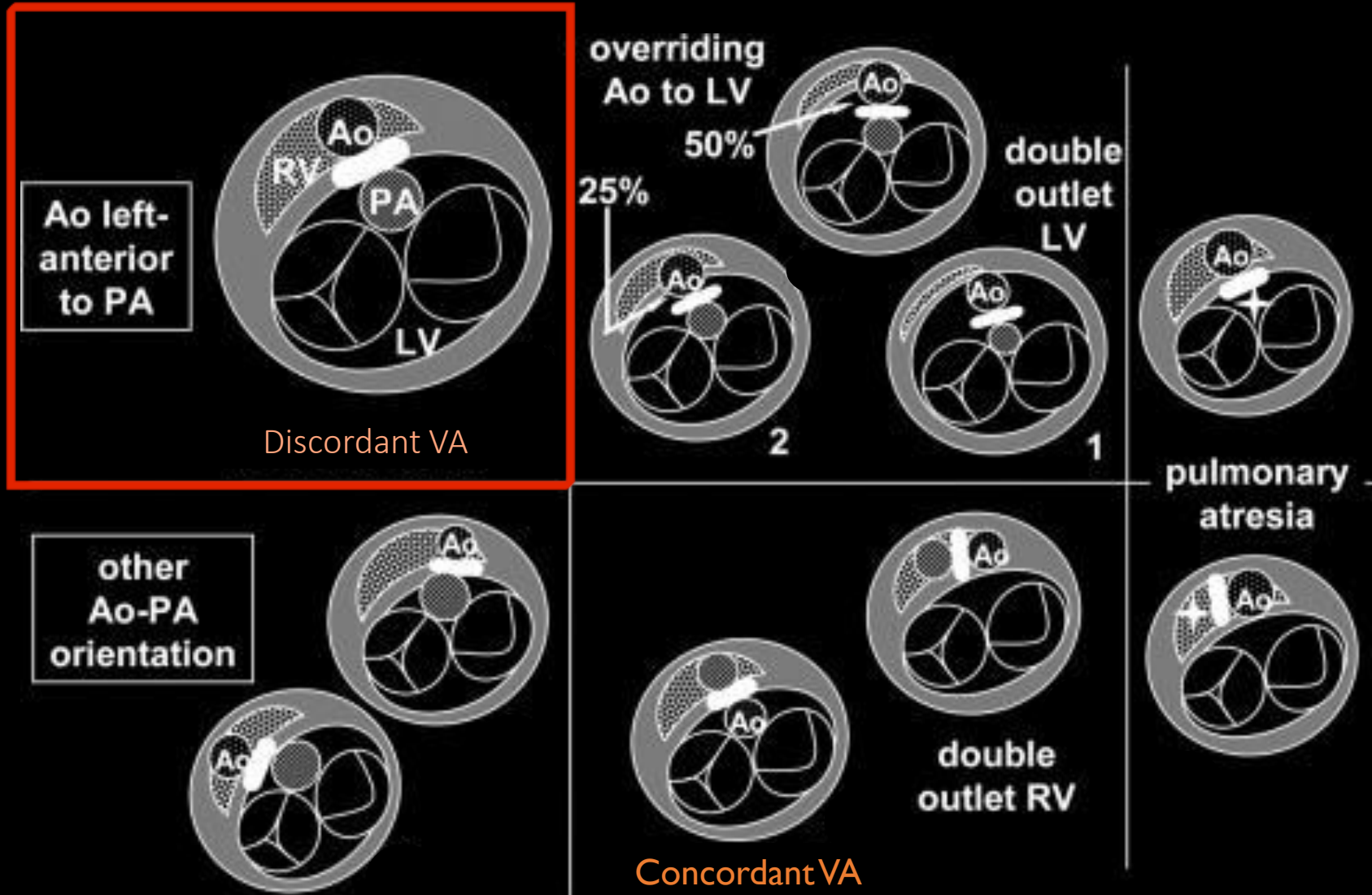


Main Ventricle Left Morphology &
Rudimentary chamber at right sided & ventricular-arterial discordant



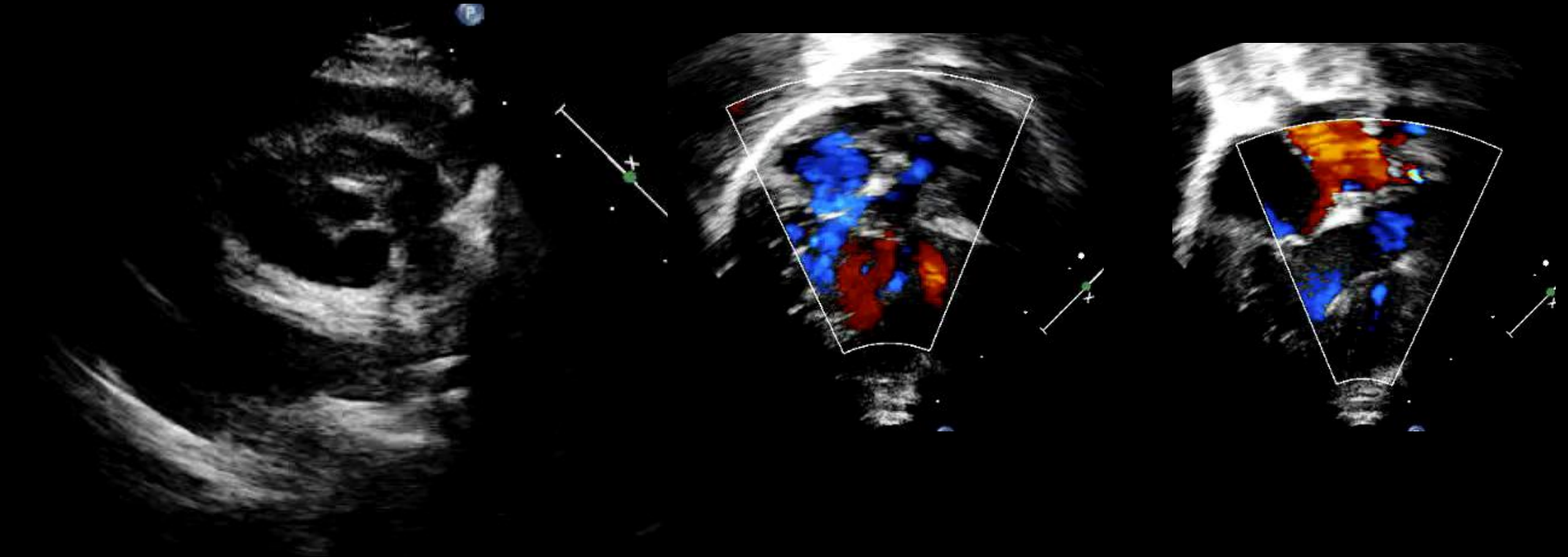
Ventrículo principal tipo esquerdo com câmara rudimentar à **DIREITA**
E **Conexão ventrículo-arterial concordante**

Great Vessels spatial relationship: double inlet LV



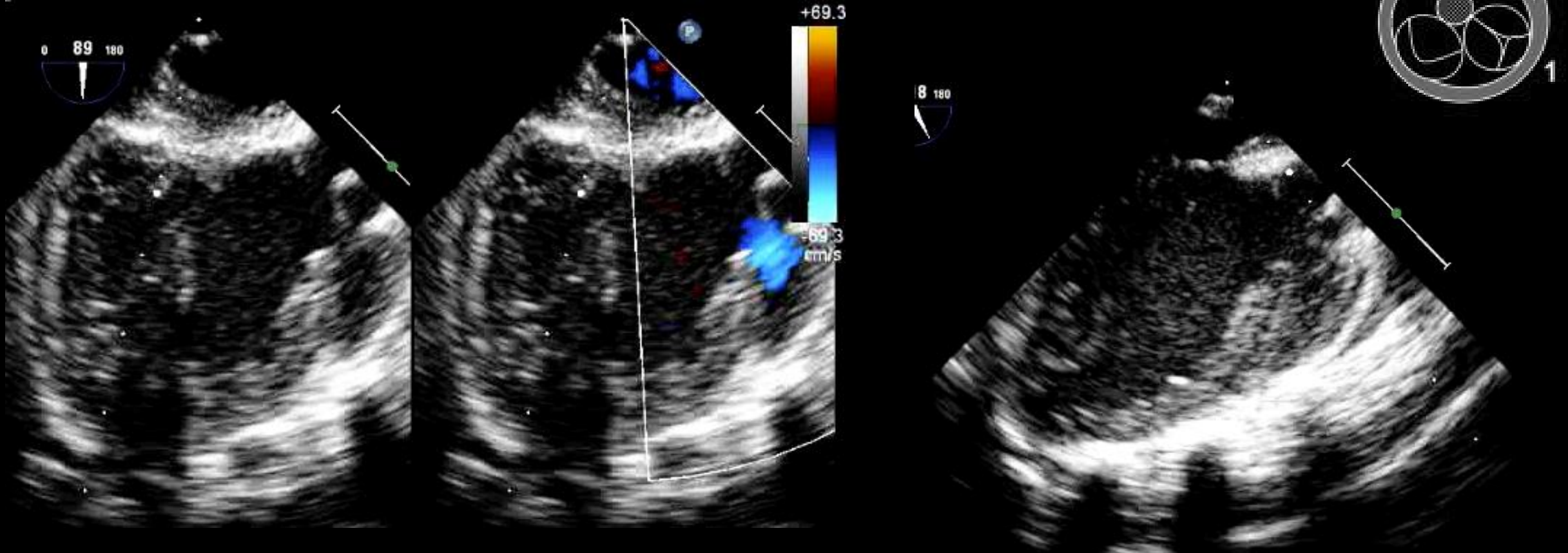
Great Vessels spatial relationship: double inlet LV

Double outlet of LV?

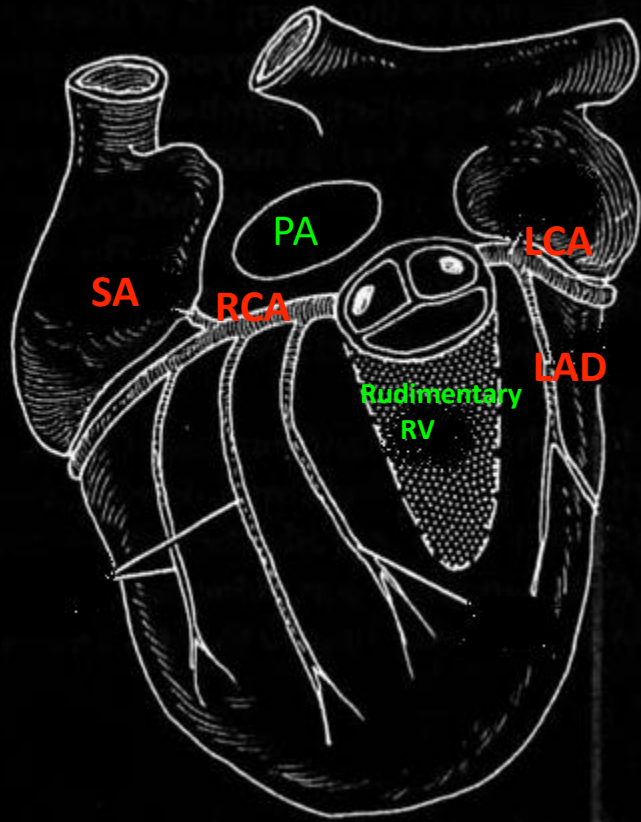


Great Vessels spatial relationship: double inlet LV

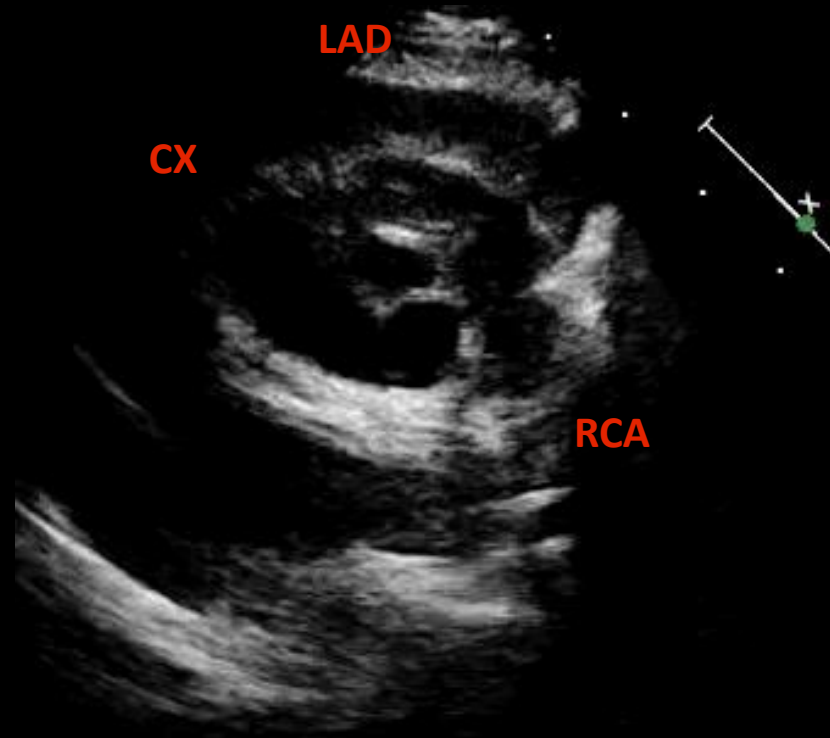
Double inlet & outlet of LV (25% overriding aorta)



Coronary arteries: Univentricular AV connection



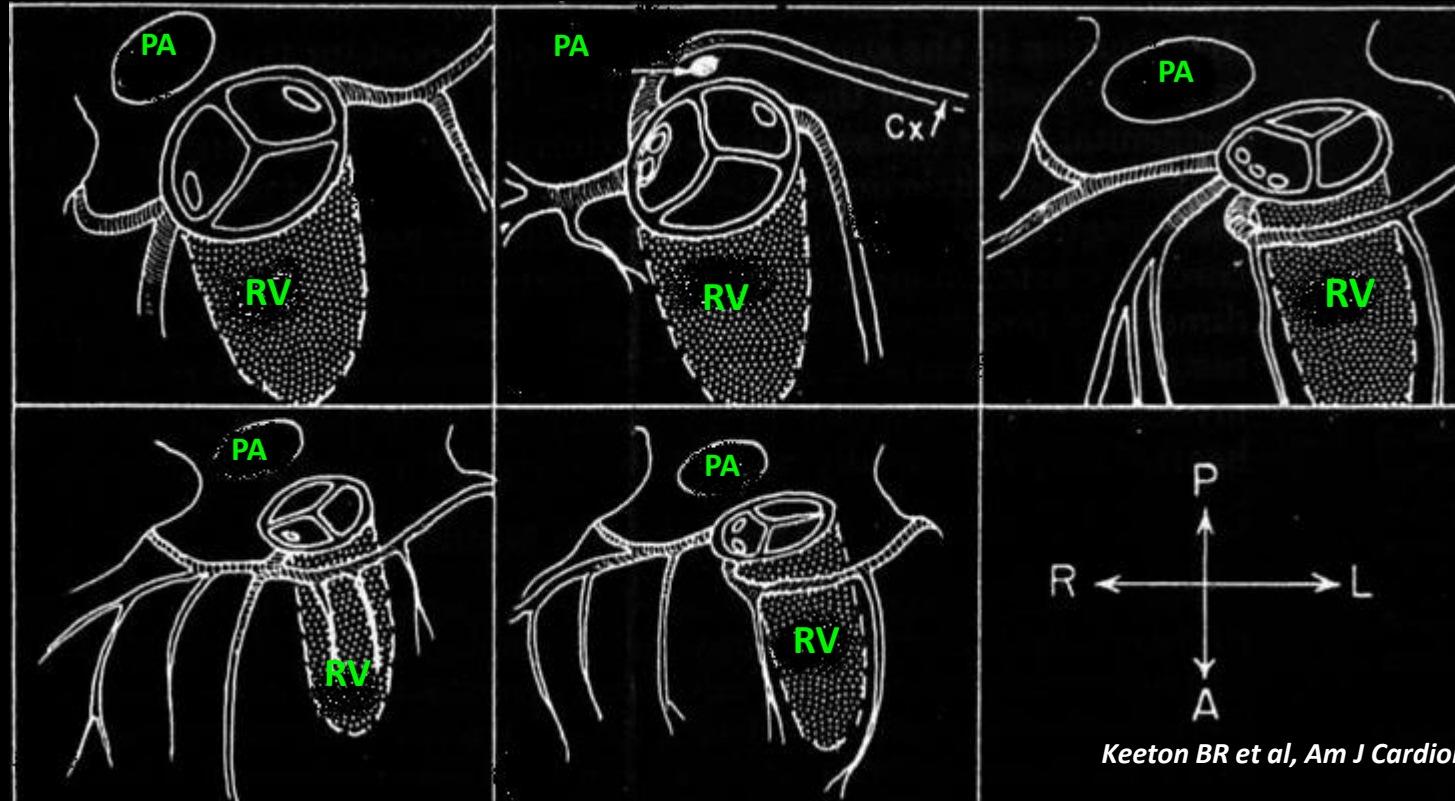
Keeton BR et al, Am J Cardiol 1979



Main Ventricle Left Morphology &
Rudimentary chamber at left sided & ventricular-arterial discordant

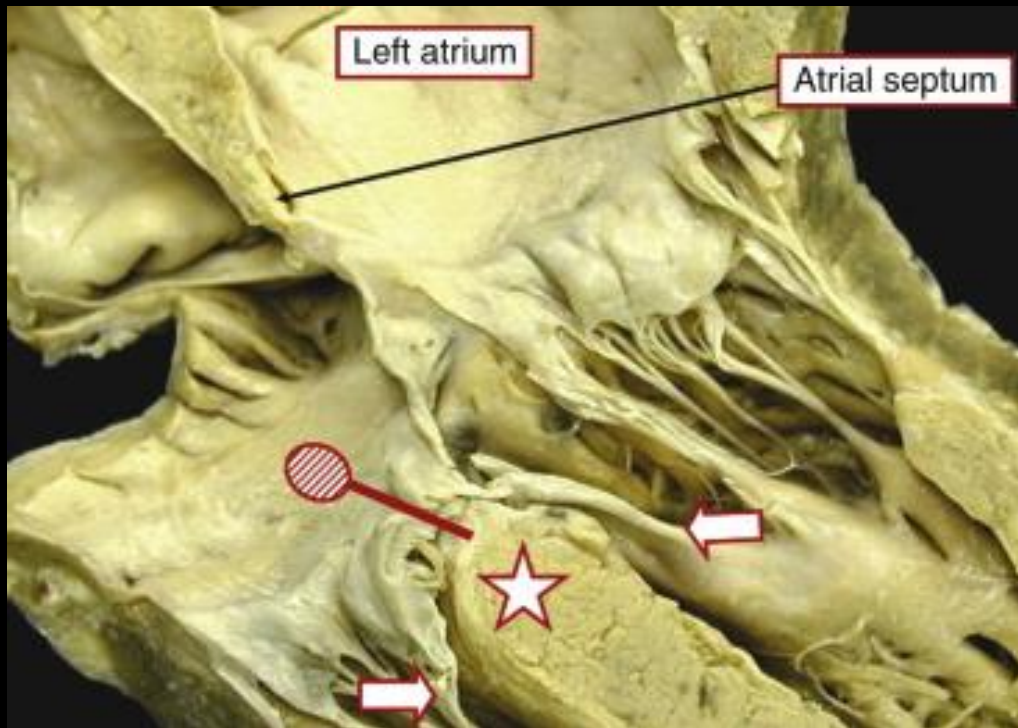
Coronary arteries: Univentricular AV connection

Unusual pattern



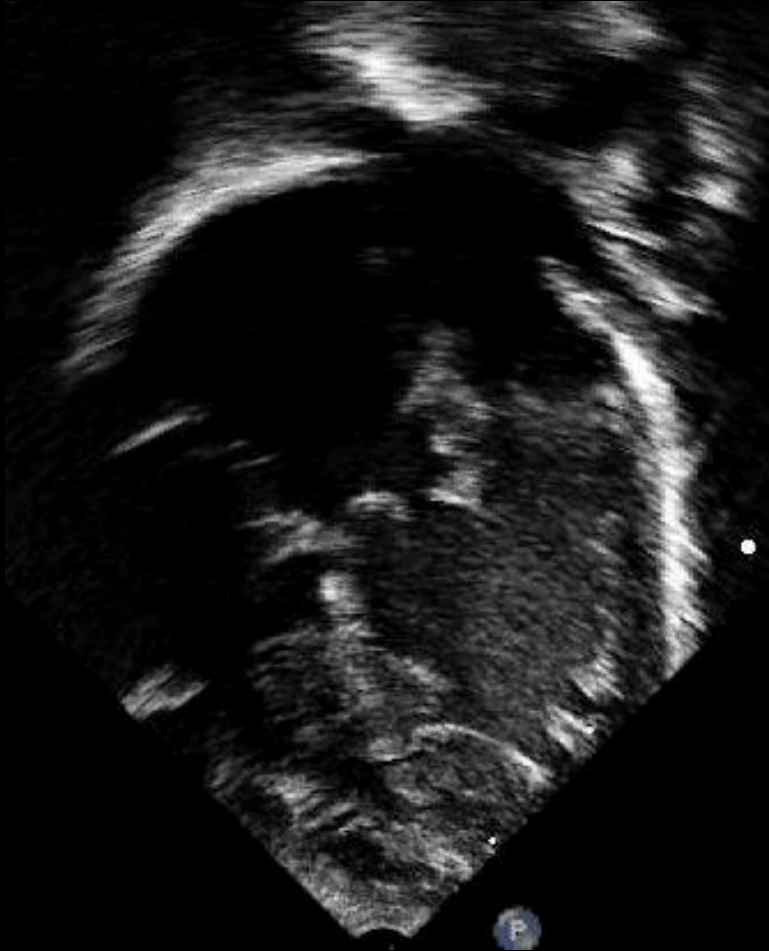
Main Ventricle Left Morphology &
Rudimentary chamber at left sided & ventricular-arterial discordant

Straddling & overriding Tricuspid valve



From Anderson RH, Pediatric Cardiology 3rd edition

Double inlet of Ventricle: Straddling & overriding Right AVV



Double inlet of Ventricle: Straddling & overriding Right AVV



AV Connection : Criss – Cross hearts

