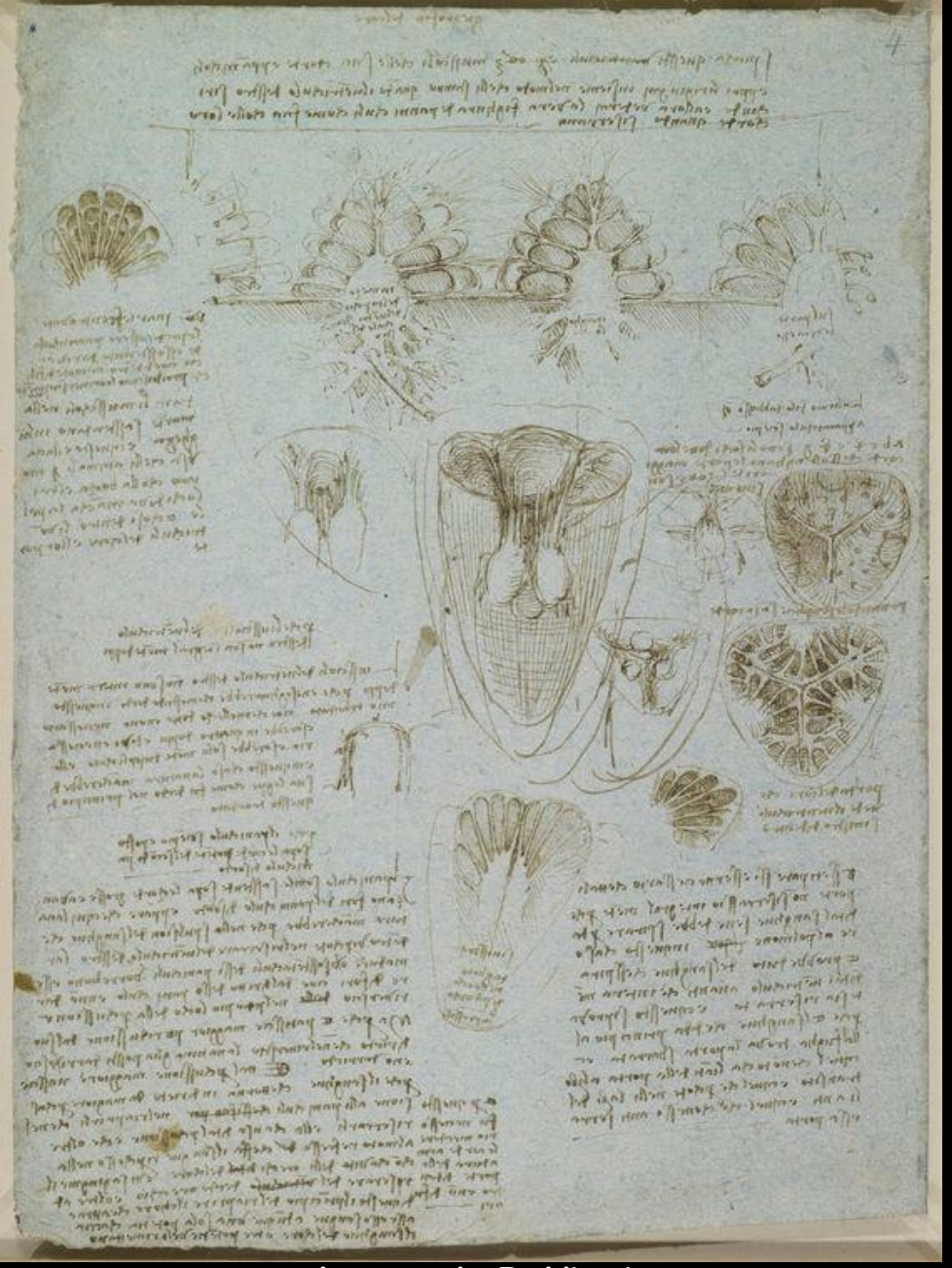


Leonardo DaVinci
“ Sketch of Mitral Valve”
Episcopal Mitre

The 2nd Contemporary Morphology Course with Specimens and 3D Print
Morphology-Imaging - Surgical Correlation

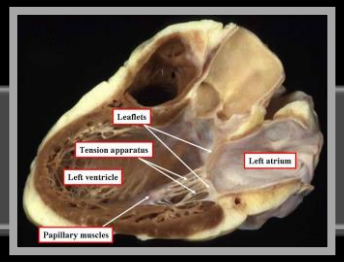
Hypoplastic Left heart Syndrome & related conditions

Vitor Guerra

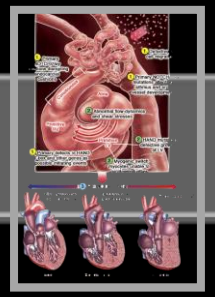
No disclosures

SickKids®

Outlines



Normal Left Heart: What is normal?

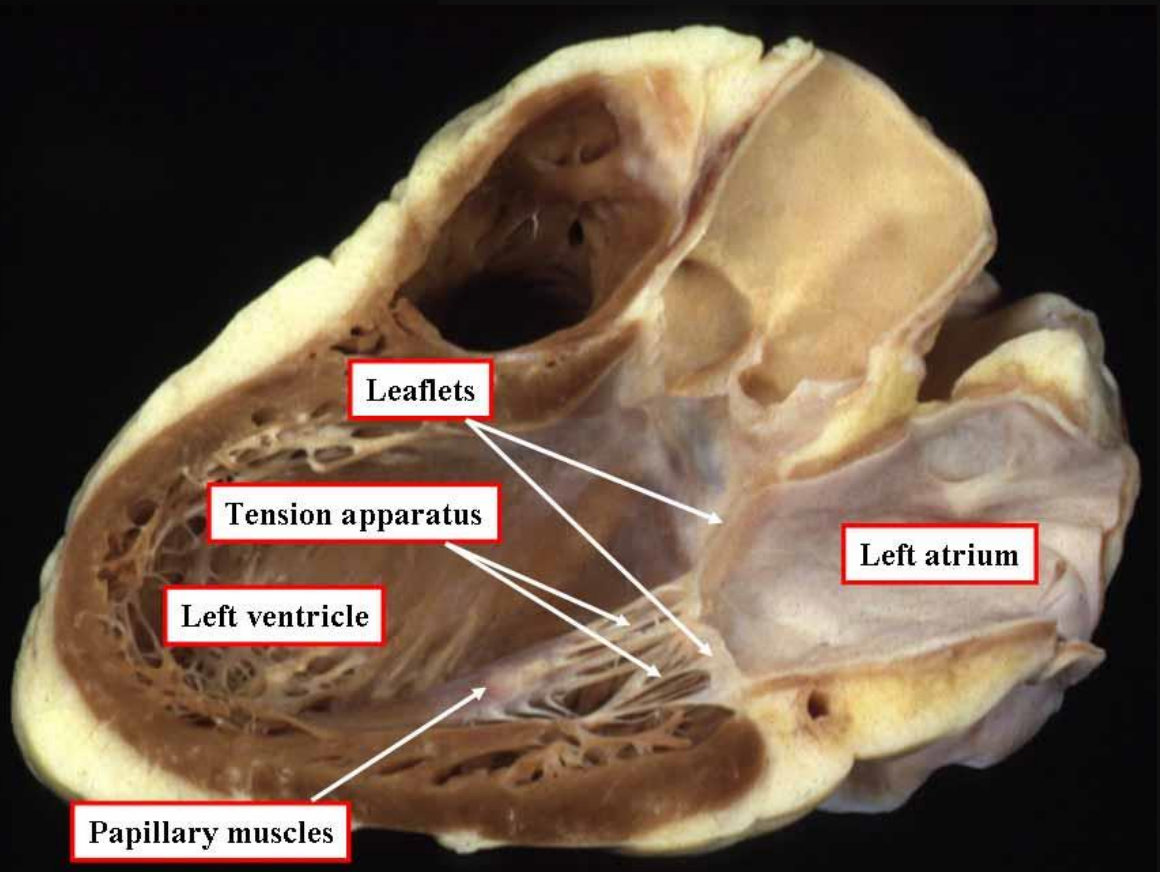


Hypoplasia of the left Ventricle: Embryology & Genetic

Hypoplastic Left heart Syndrome: Echo & Morphology

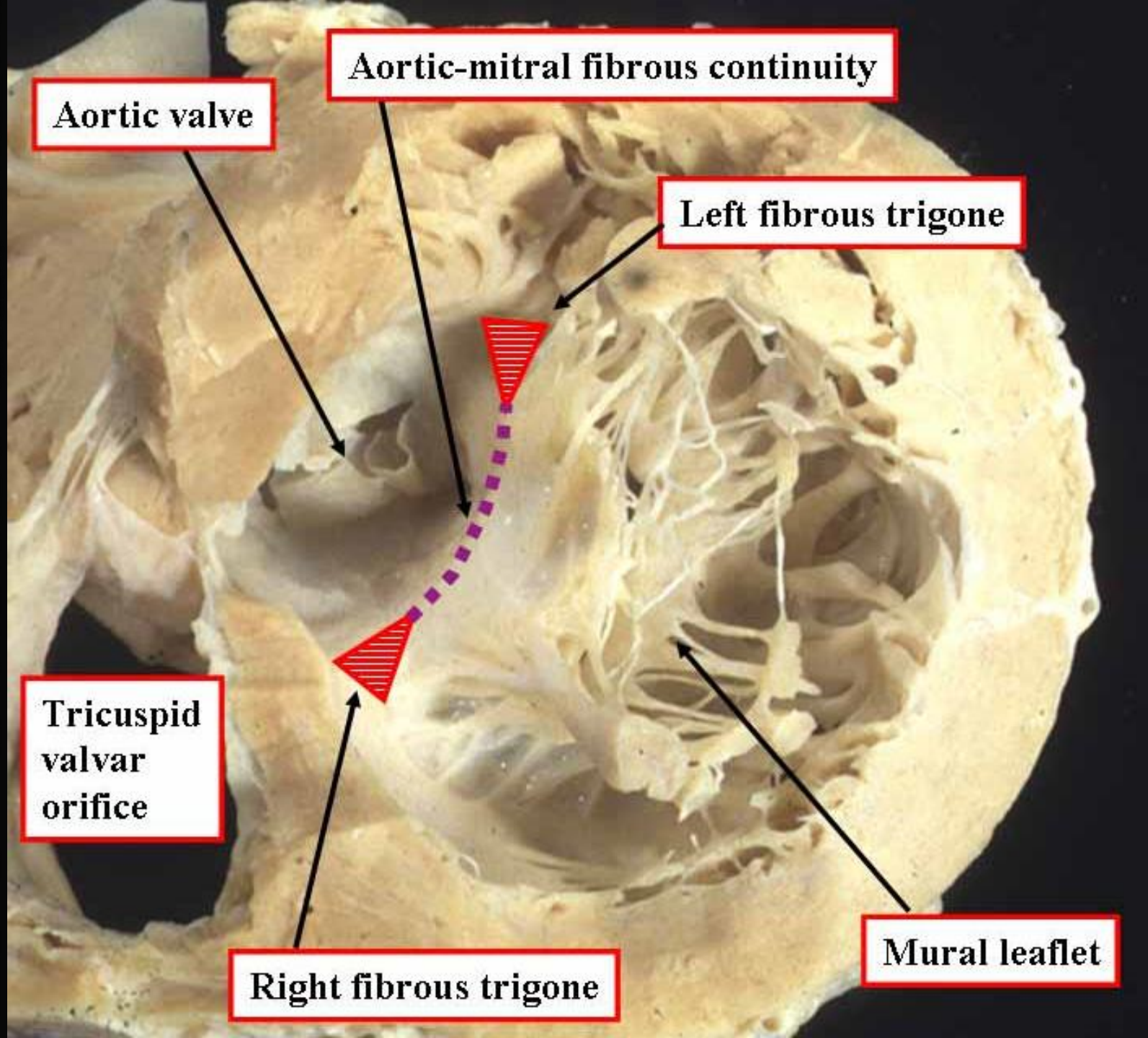
SickKids®

The Normal Left Ventricle

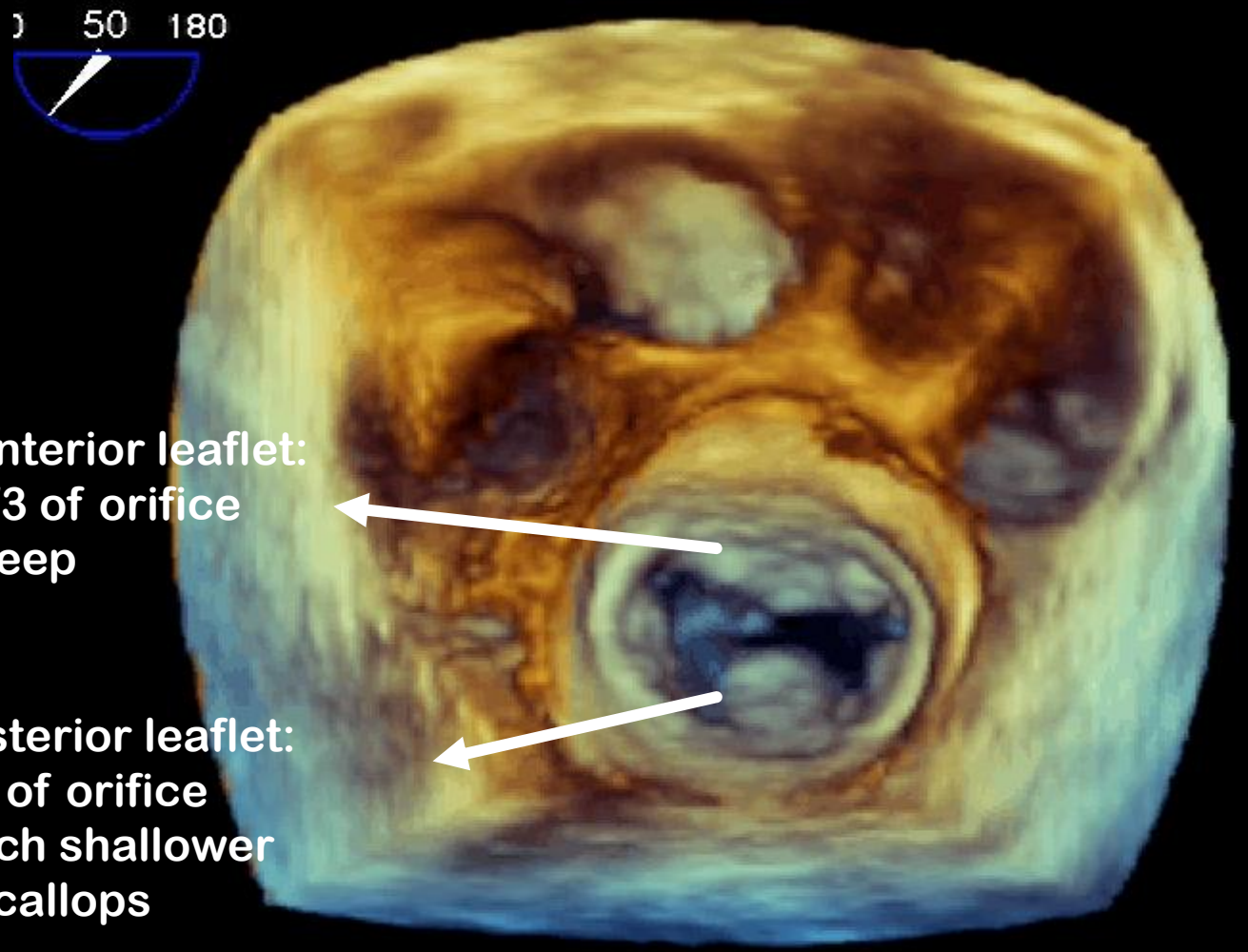


Normal Mitral valve

The anchorage of the fibrous trigones to the basal surface of the ventricular wall secures the aortic-mitral unit in the LV



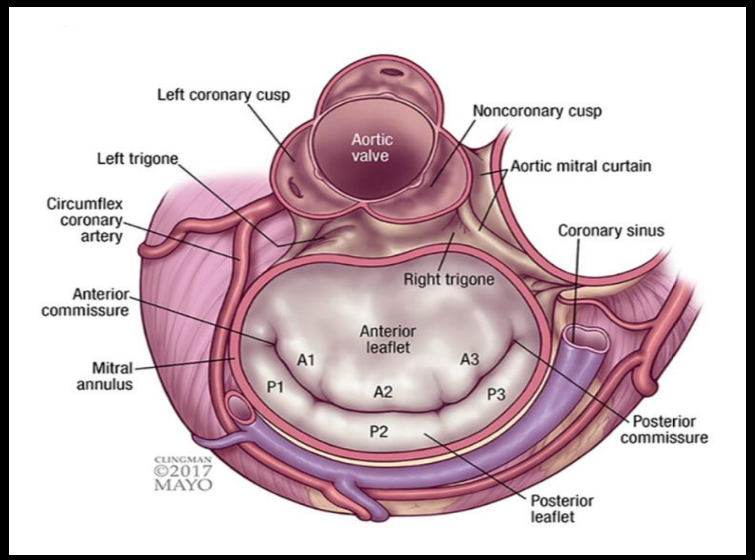
The Normal Mitral Valve



Anterior leaflet:
1/3 of orifice
Deep

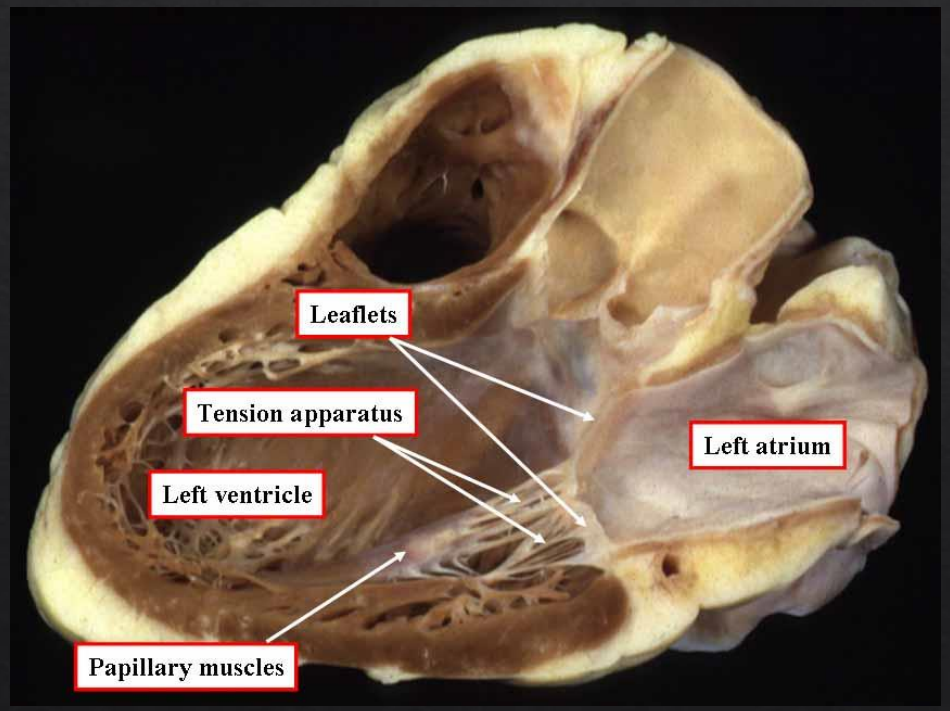
Posterior leaflet:
2/3 of orifice
Much shallower
3 scallops

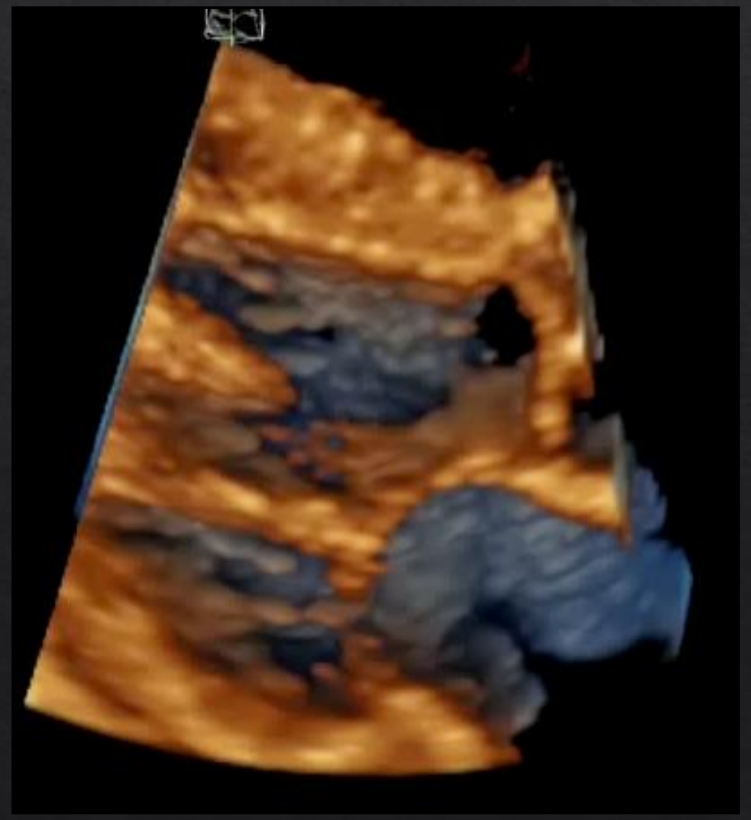
Courtesy Dra Renata Cassar Incor FMUSP



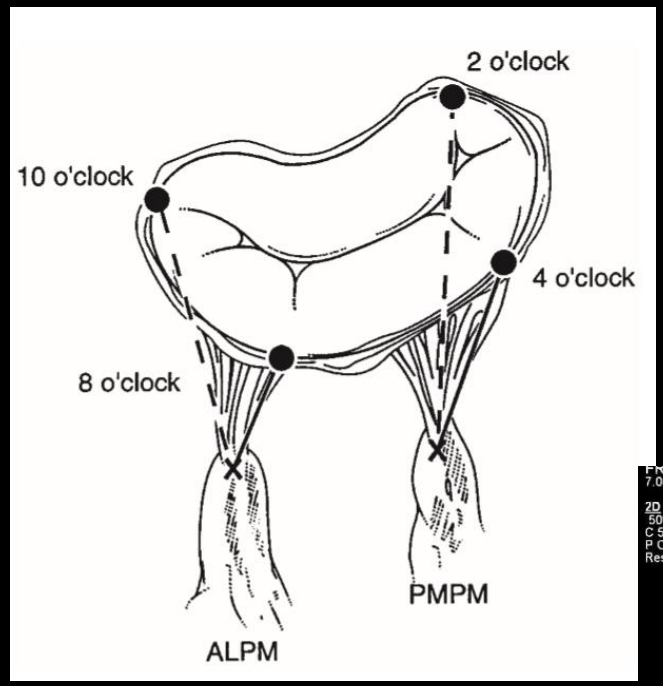
Normal mitral valve & Outflow tract & Aortic Valve

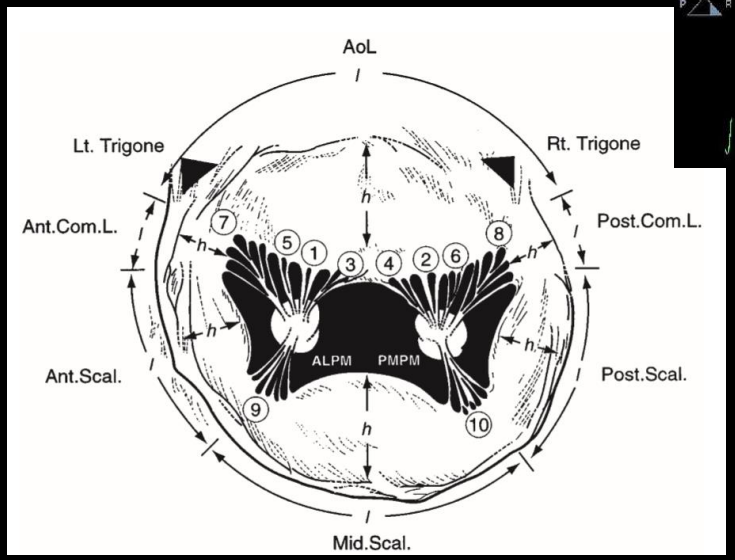
All the components of the mitral valve act in unison and in turn are intricately related to LV function

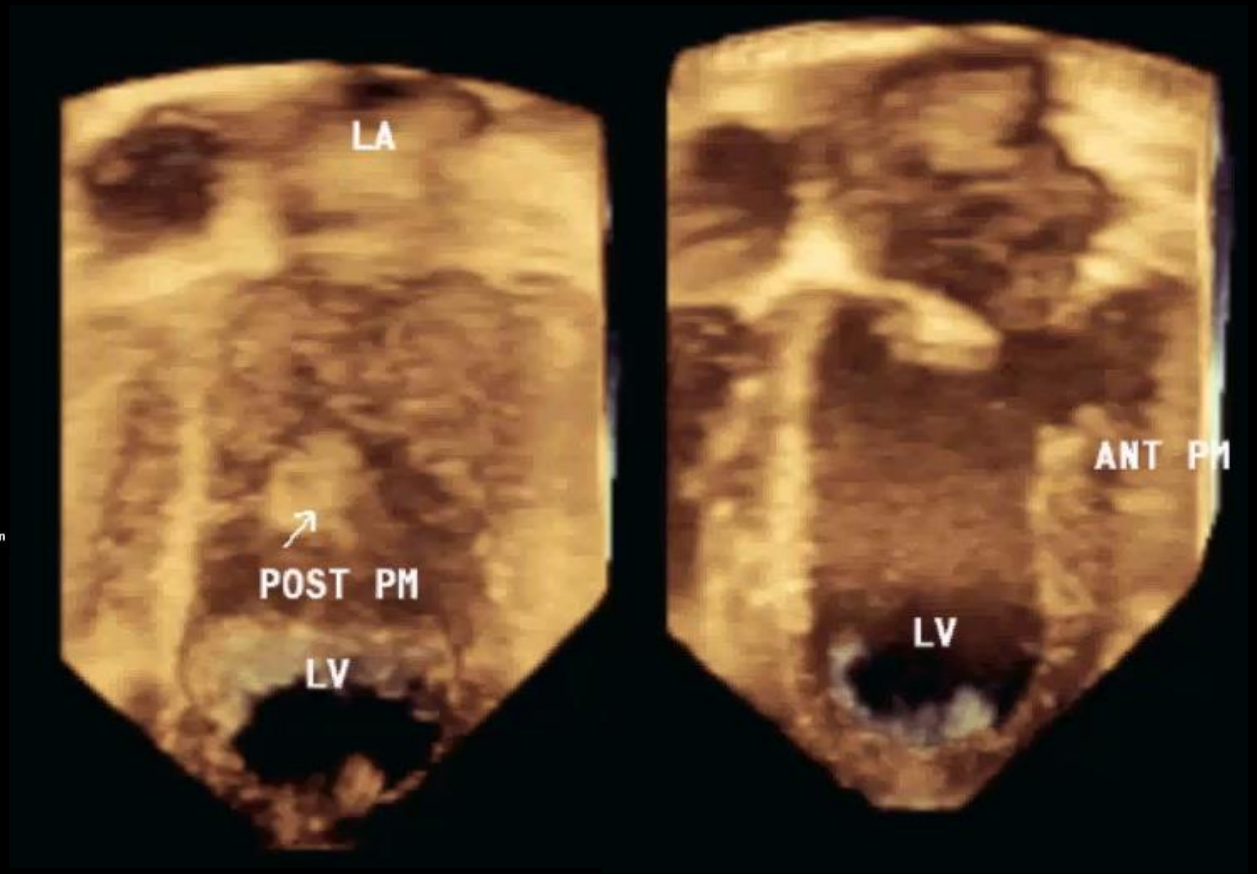




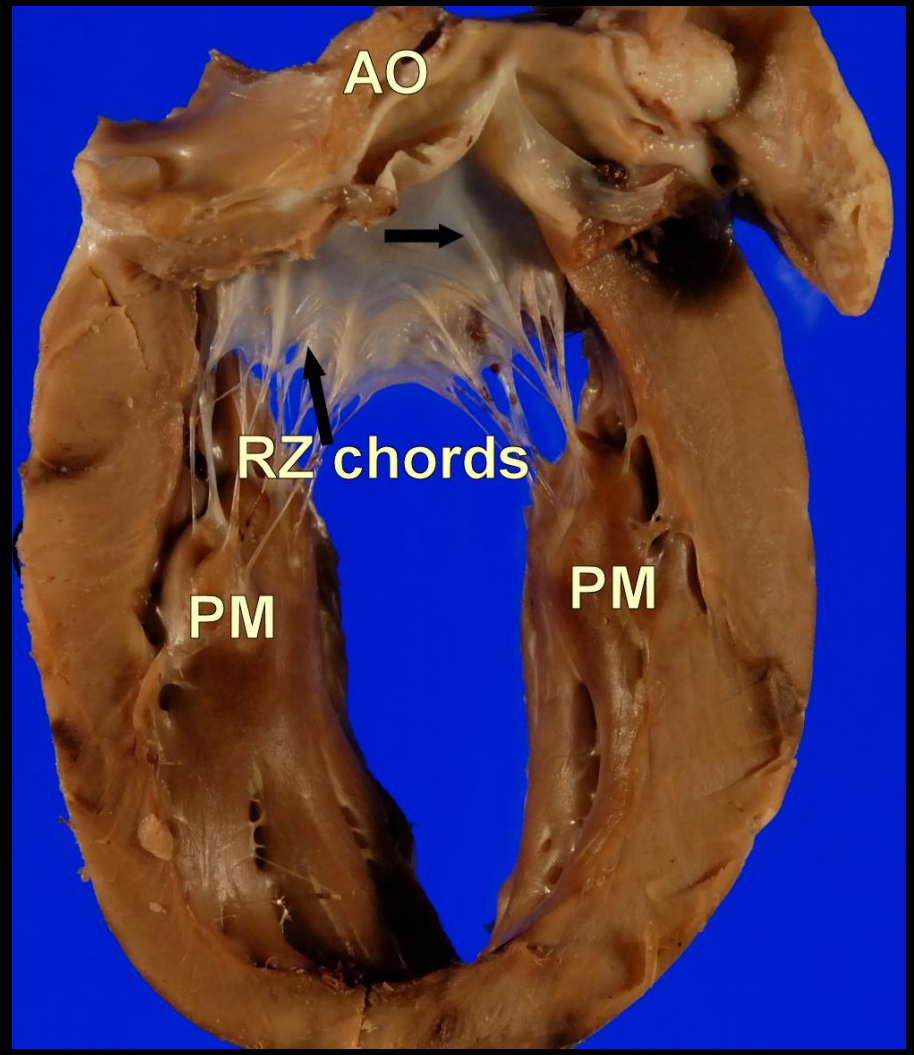
The Normal Mitral Valve : Papillary muscles

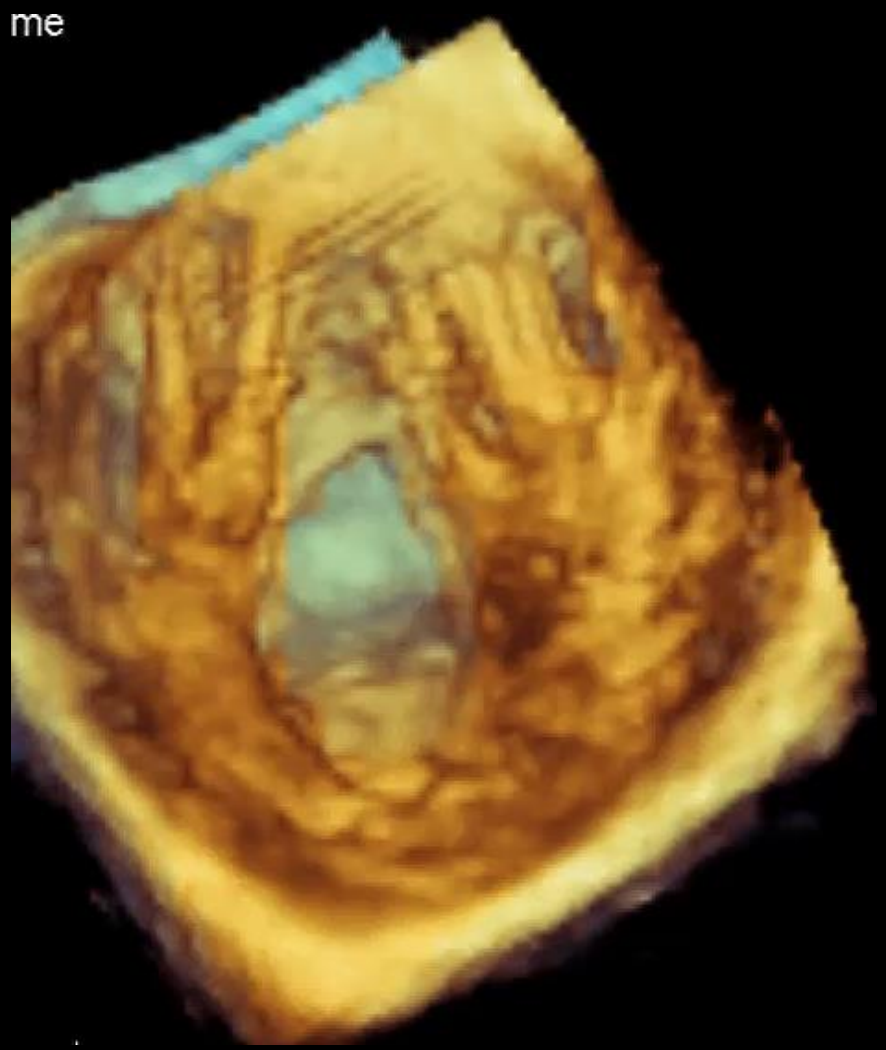






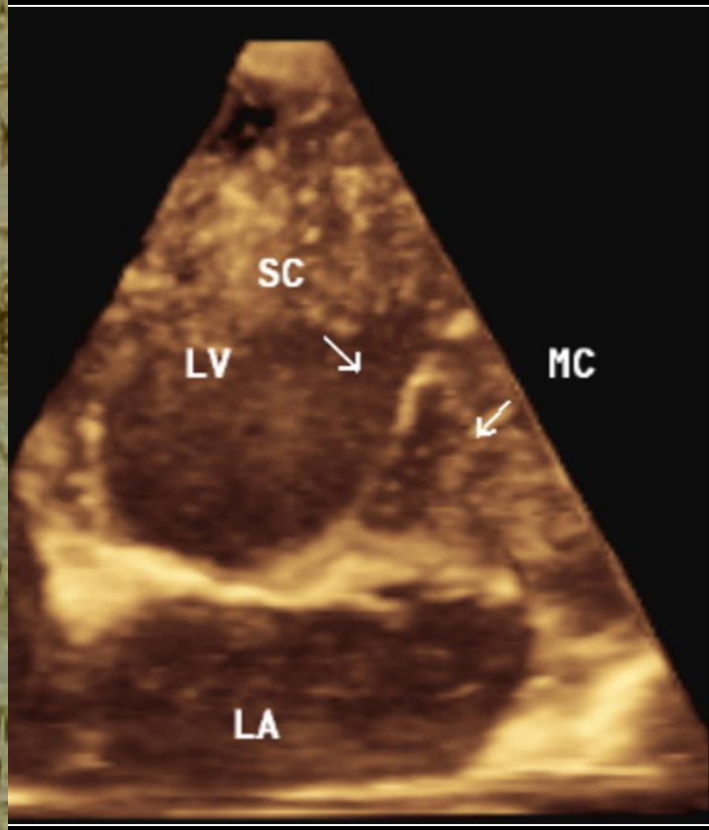
Normal Mitral Valve Chords

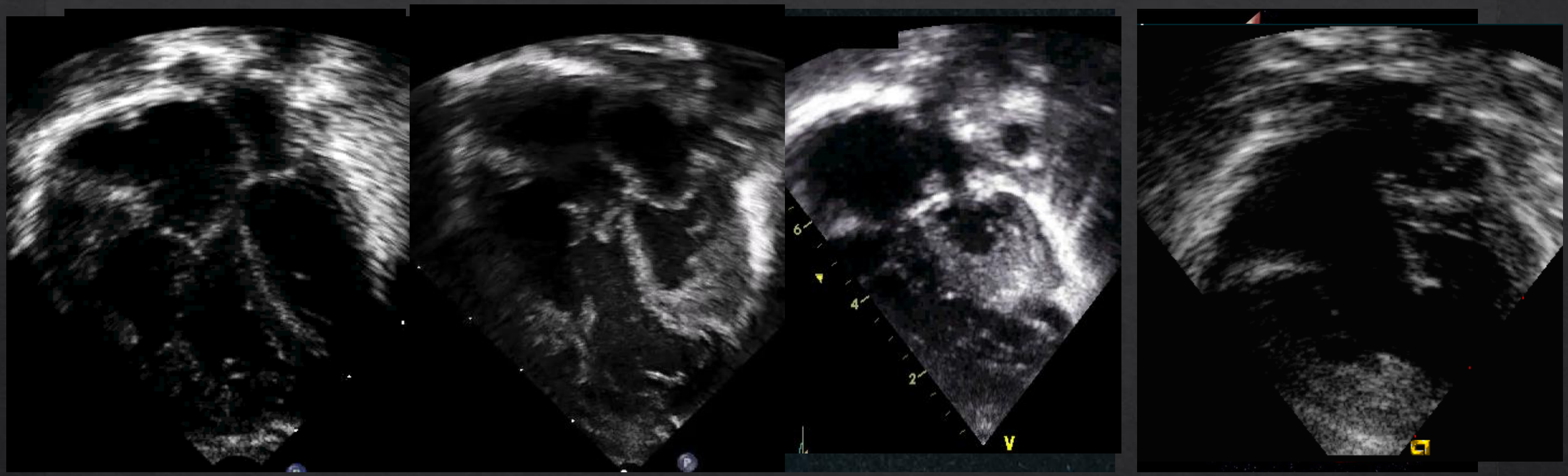




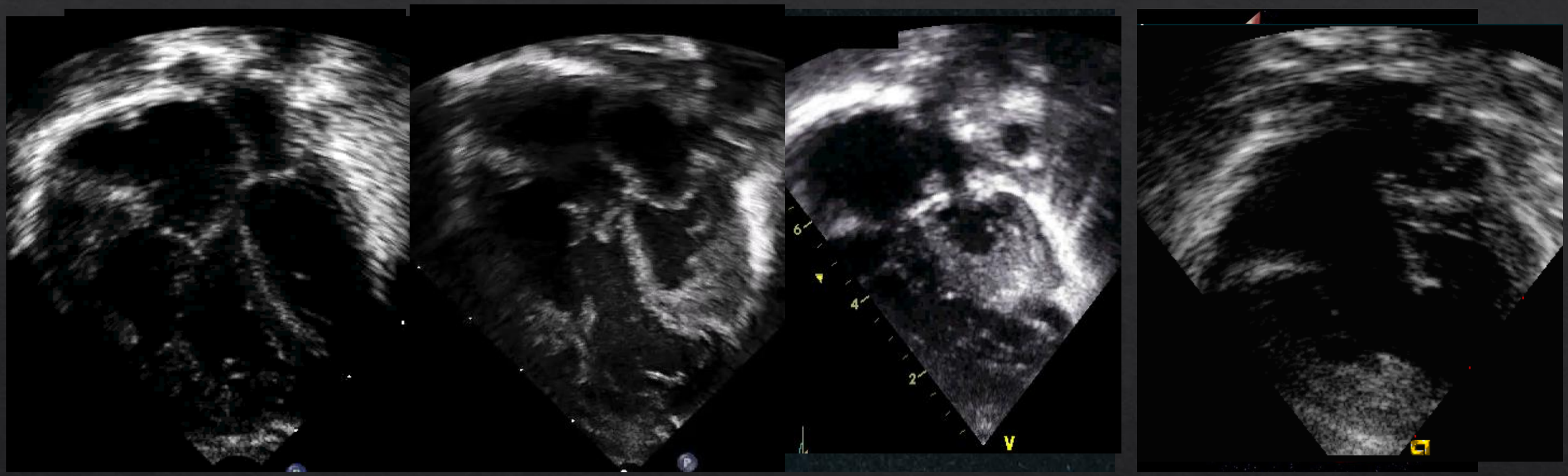
Mitral valve chordal apparatus

- The normal mitral valve: finer marginal first order chordae that support the leaflet tips and prevent prolapse
- Secondary thicker strut chordae that insert symmetrically near the anterior leaflet base



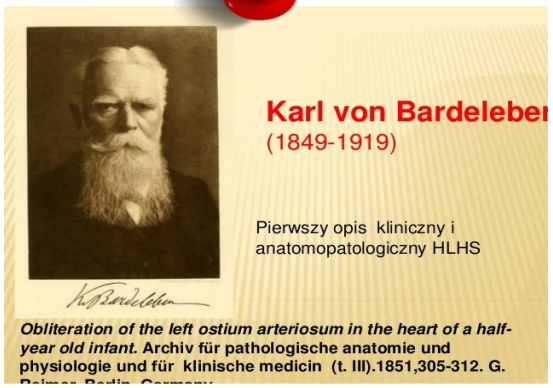


Hypoplasia of the left Heart & Variants



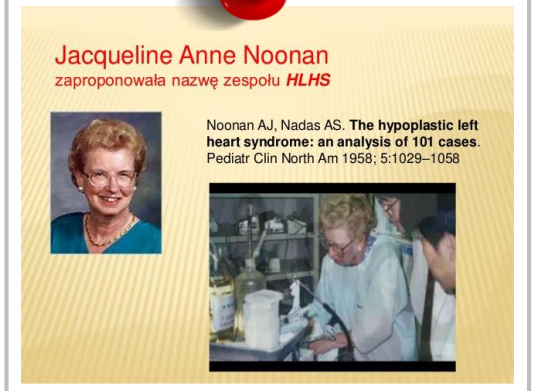
Hypoplasia of the left Heart & Variants

Hypoplastic Left Heart : Morphological Landmarks

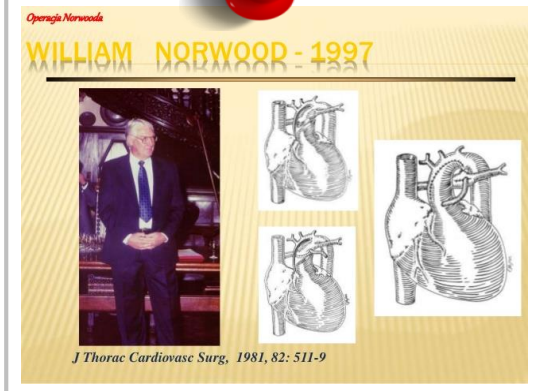


1851

1952



1958



1997

Bardeleben

- Combination of severe mitral and left ventricular hypoplasia along with aortic atresia

Maurice Lev

- Hypoplastic of the aortic tract complex:
- Tendency of hypoplastic and obstructive lesions

Noonan and Nadas

- aortic atresia
- Mitral atresia
- Mitral stenosis
- Atresia of aortic arch
- Hypoplasia aortic arch

William Norwood

- hypoplastic left heart syndrome

Definition of Hypoplastic Left Heart Syndrome

Atresia: “a” means without, “tresis” means perforation – congenital absence or closure of normal orifice

Stenosis: narrowing of duct or a canal

Hypoplasia: incomplete development or underdevelopment of an organ or tissue

Obstruction: act of blocking or clogging

Syndrome: group of signs and symptoms that occur together

Complex: a whole, made up of interrelated parts

The nomenclature, definition and classification of hypoplastic left heart syndrome

Christo I. Tchervenkov,[1] Jeffrey P. Jacobs,[2] Paul M. Weinberg,[3] Vera D. Aiello,[4] Marie J. Béland,[5] Steven D. Colan,[6] Martin J. Elliott,[7] Rodney C.G. Franklin,[8] J. William Gaynor,[9] Otto N. Krogmann,[10] Hiromi Kurosawa,[11] Bohdan Maruszewski,[12] Giovanni Stellin[13]

Cardiol Young 2006; 16: 339–368

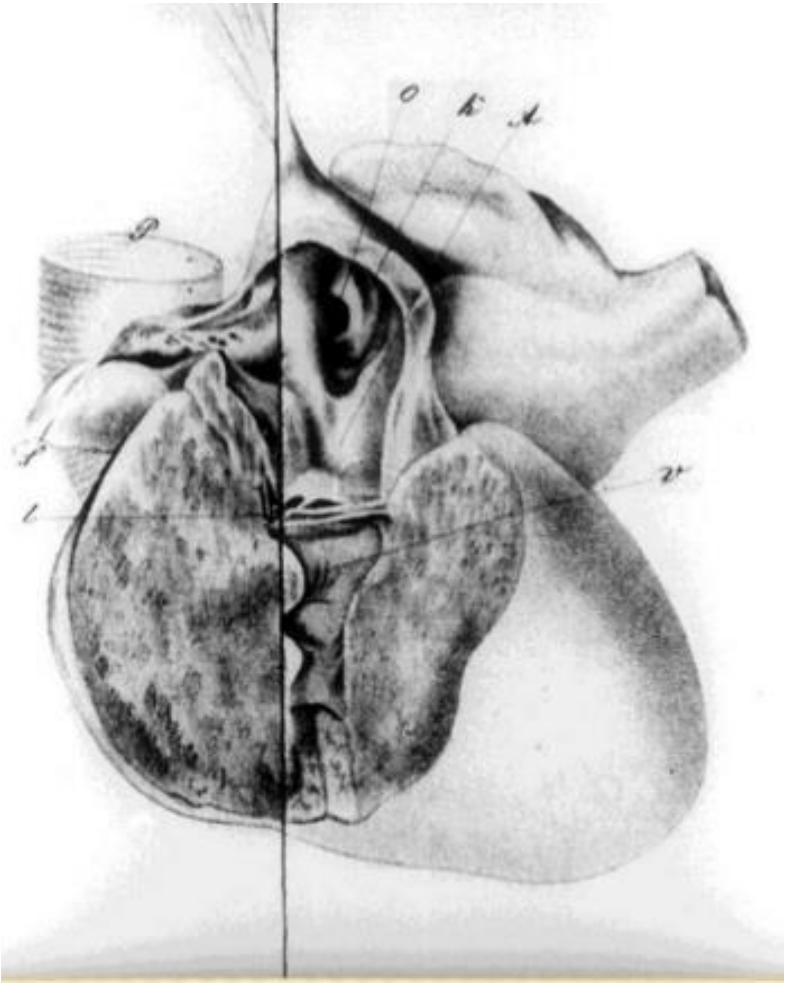
"Hypoplastic left heart syndrome is synonymous with the term hypoplasia of the left heart and is defined as a spectrum of cardiac malformations with normally aligned great arteries without a common atrioventricular junction, characterized by underdevelopment of the left heart with significant hypoplasia of the left ventricle including atresia, stenosis, or hypoplasia of the aortic or mitral valve, or both valves, and hypoplasia of the ascending aorta and aortic arch."

- The concept of SCEH has been expanded for all cases with LV hypodevelopment, which is incapable of maintaining systemic circulation, even if the mitral valve and Ao are present, but hypoplastic

- Recently HLHS : intact interventricular septum

Hypoplastic Left Heart : Morphological Phenotypes

1. Mitral and aortic atresia
2. Mitral atresia, patent aortic root and VSD
3. Aortic atresia with patent mitral valve
4. Aortic valvar Stenosis and patent mitral valve
5. Hypoplastic mitral , aortic valve ,hypoplasia of aortic arch and coarctation of the aorta

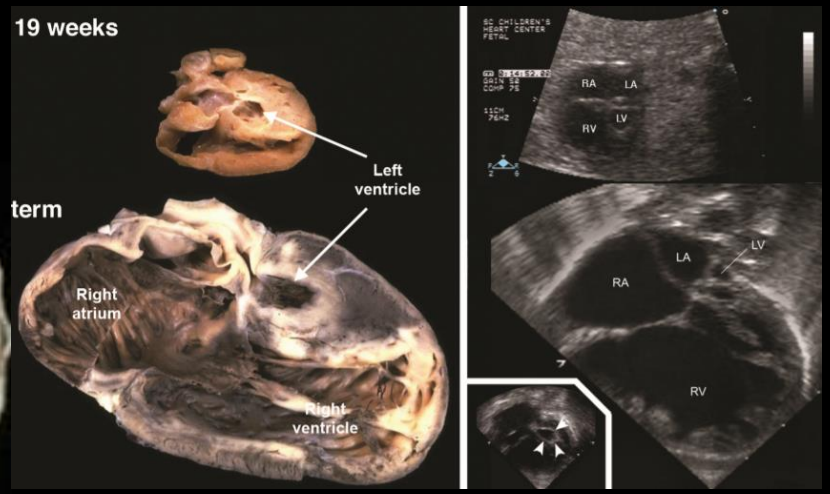


"Obliteration of the left ostium arteriosum in the heart of a half-year old infant: 1851"

Development of Left Heart hypoplasia

Normal Heart

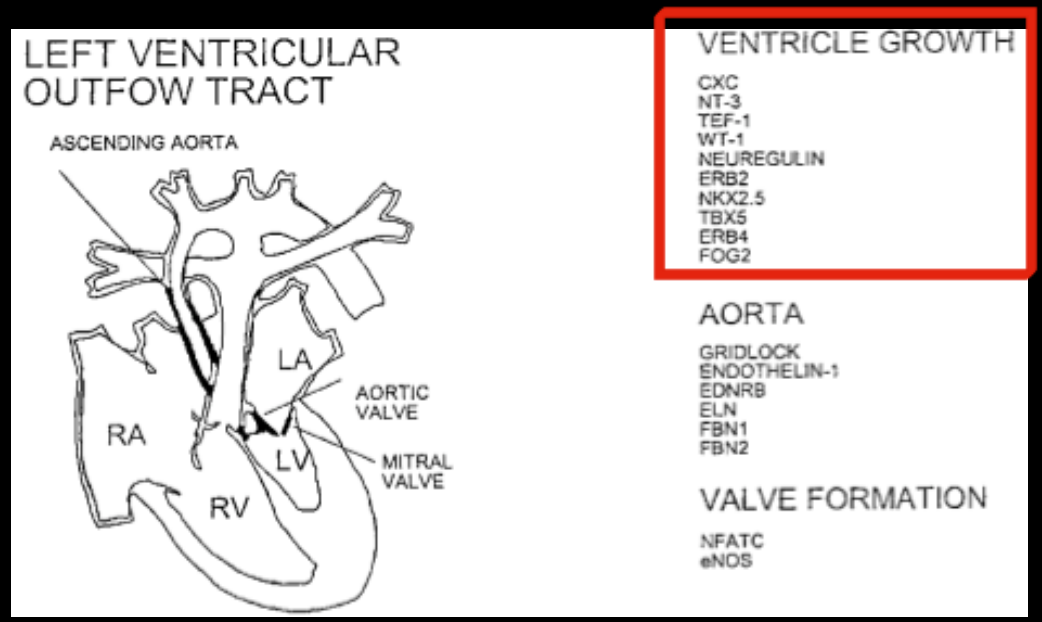
Hypoplastic left heart



Growth of the left heart “structures” through the second half of pregnancy seems to be the main time to detect and/or predict the severity of hypoplasia

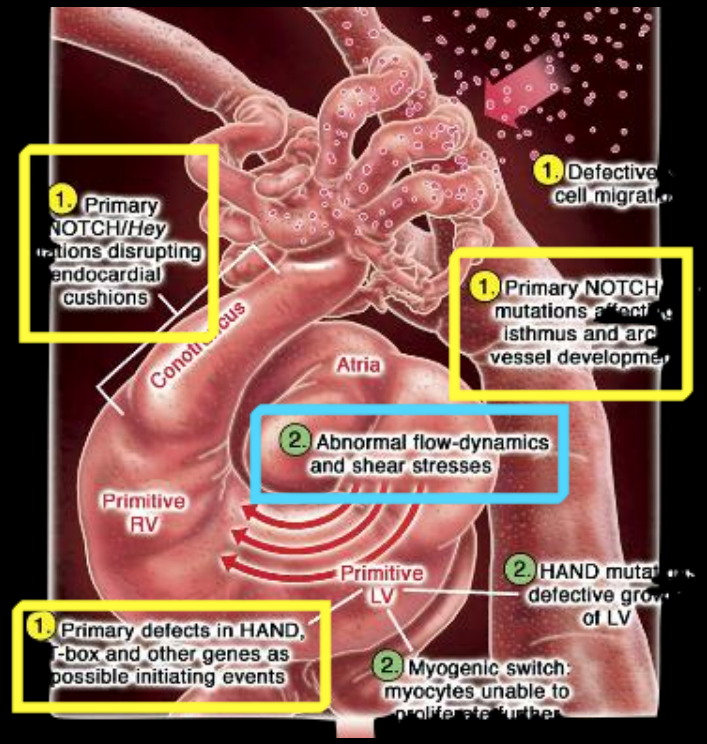
Hypoplastic Left heart : Genetics

- Aortic atresia is the most common manifestation of HLHS;
- Bicuspid AoV: most common finding in first degree relatives of children HLHS (~ 11%).
- Genetic-linkage analysis: shared chromosomal loci (10q22 and 6q23) in the etiology of bicuspid Ao valve and subset of children with HLHS.

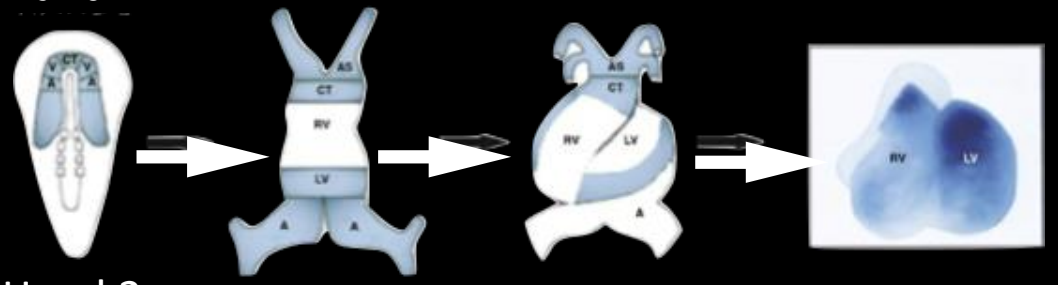


Towbin J t al. Am. J. Med. Genet. (Semin. Med. Genet.) 97:297±303, 2000

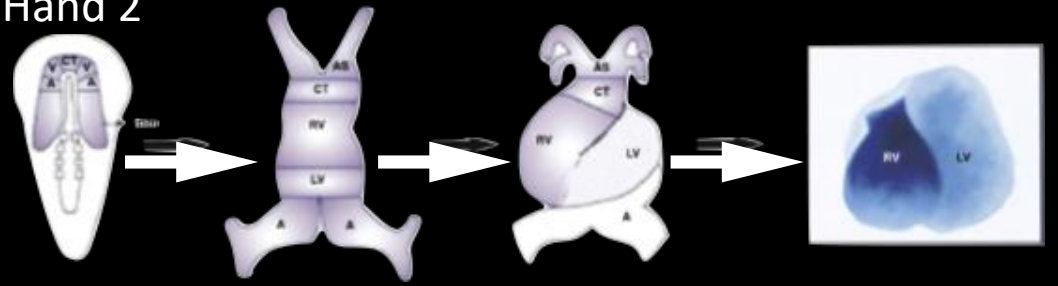
HLHS: Genetics & Embryology background



Hand 1

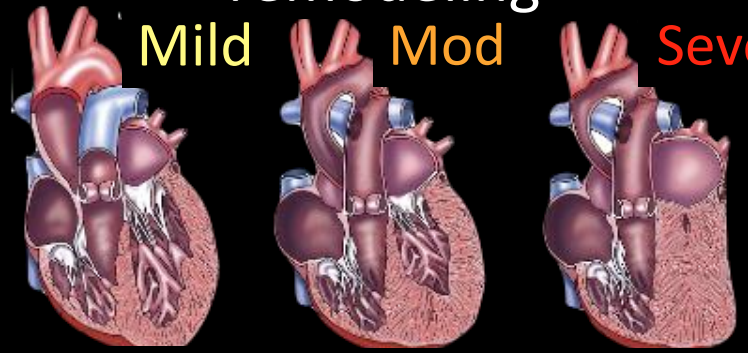


Hand 2



remodeling

Mild Mod Severe



2V repair Borderline LV 1V repair

Hickey et al,, J AM Coll Cardiol 2012, 59

Abnormal flow-dynamics & shear-stresses: trigger the LV remodeling process

HLHS: Physiology & “Hydraulic”& Embryology

Experimental Production of Hypoplastic Left Heart Syndrome in the Chick Embryo

JUNG Y. HARH, MD*†
MILTON H. PAUL, MD, FACC*
WILLIAM J. GALLEN, MD†
DAVID Z. FRIEDBERG, MD†
STANLEY KAPLAN, PhD‡

Chicago, Illinois
Milwaukee, Wisconsin

- First LV hypoplasia model;
- Obliterating the inlet of LV (5 day old chick embryos)
- 20% survival: all degrees of left side hypoplasia;
- “flow-volume hypoplasia was a result of abnormal “flow-volume streaming”

January 1973 The American Journal of CARDIOLOGY Volume 31

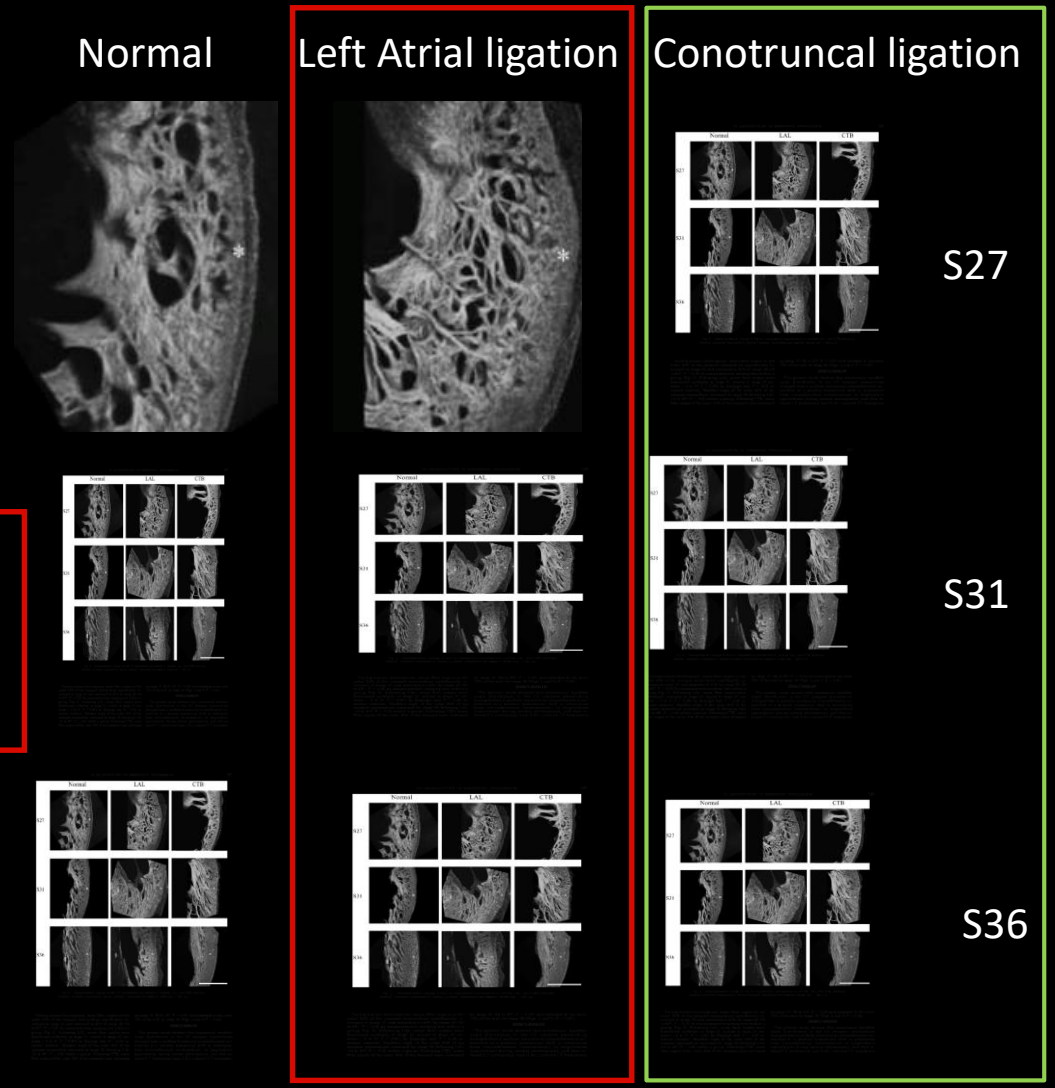
HLHS : Physiology & “Hydraulic”& Embryology

Three-Dimensional Myofiber Architecture of the Embryonic Left Ventricle During Normal Development and Altered Mechanical Loads

KIMIMASA TOBITA,* JASON B. GARRISON, LI J. LIU, JOSEPH P. TINNEY, AND BRADLEY B. KELLER
Division of Pediatric Cardiology, Cardiovascular Development Research Program, Children’s Hospital of Pittsburgh, University of Pittsburgh, Pittsburgh, Pennsylvania

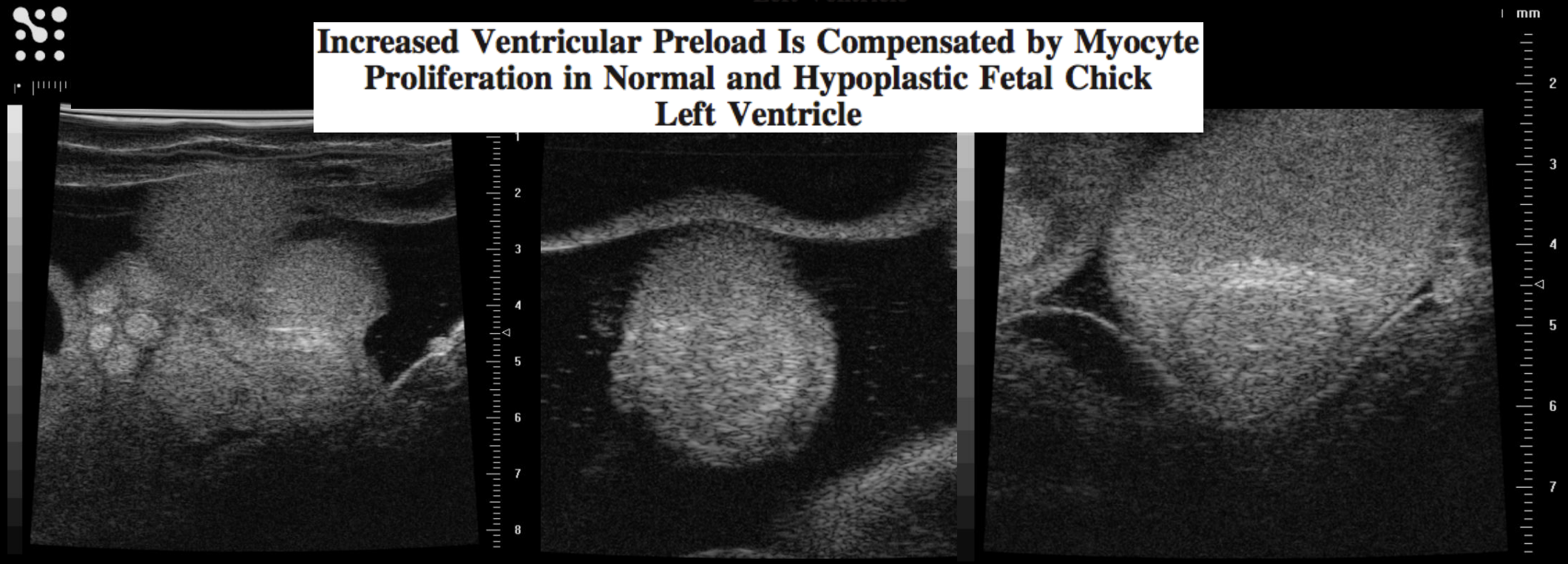
Reducing the preload: <chamber dimensions and myocardial volume (LV hypoplasia); also associated with LV cell proliferation rate.

Increased LV pressure load induces LV chamber dilatation followed by thickening of compact myocardium and acceleration of tertiary trabeculation, myocardial cell proliferation rate increases .



THE ANATOMICAL RECORD PART A 283A:193–201 (2005)

HLHS: Physiology & flow dynamics (Hydraulic)& Embryology



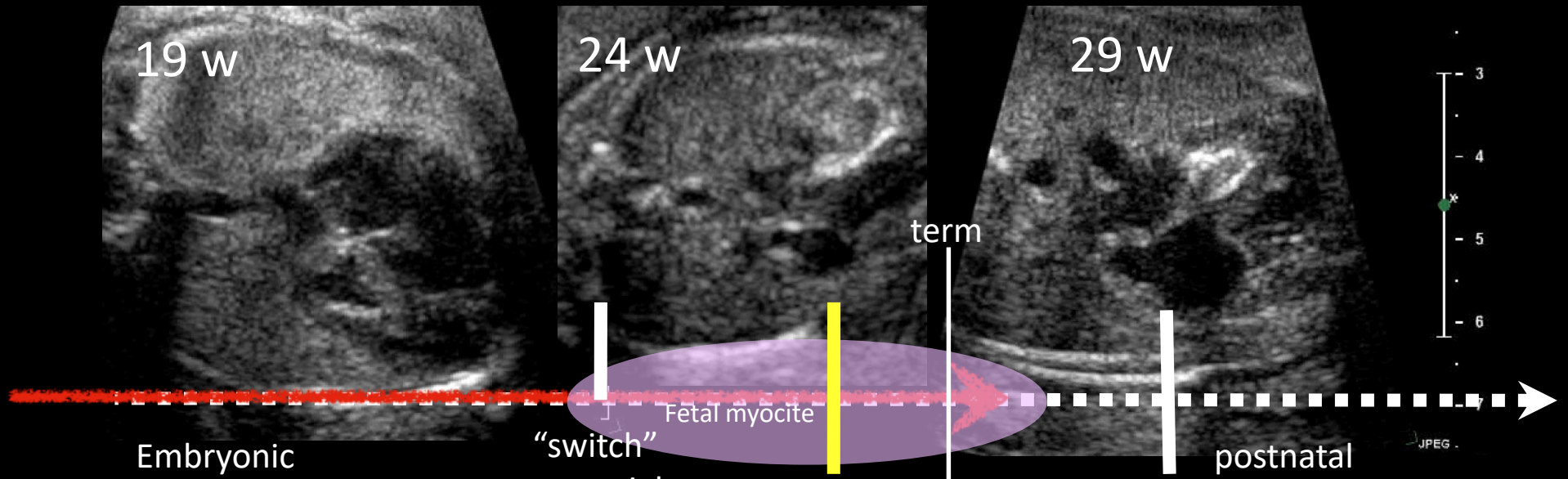
Right atrium “clip” → increased left-sided flow and hyperplastic ventricular response

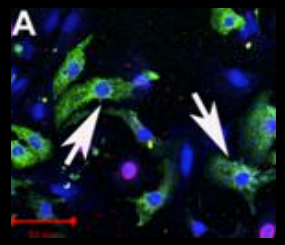
Left atrium “clip” → Decreased myocyte proliferation in the Hypoplastic Left Ventricle model

LV is rescued by increased hemodynamic loading

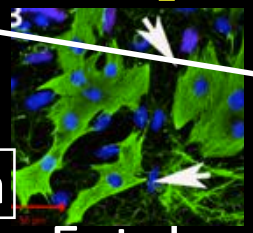
Almeida A et al, Circ Res. 2007;100

Cardiac myocyte proliferation and growth





Primitive myocardium: highly proliferative immature myocytes



Hyperplasia

Trabecular myocites: low proliferation

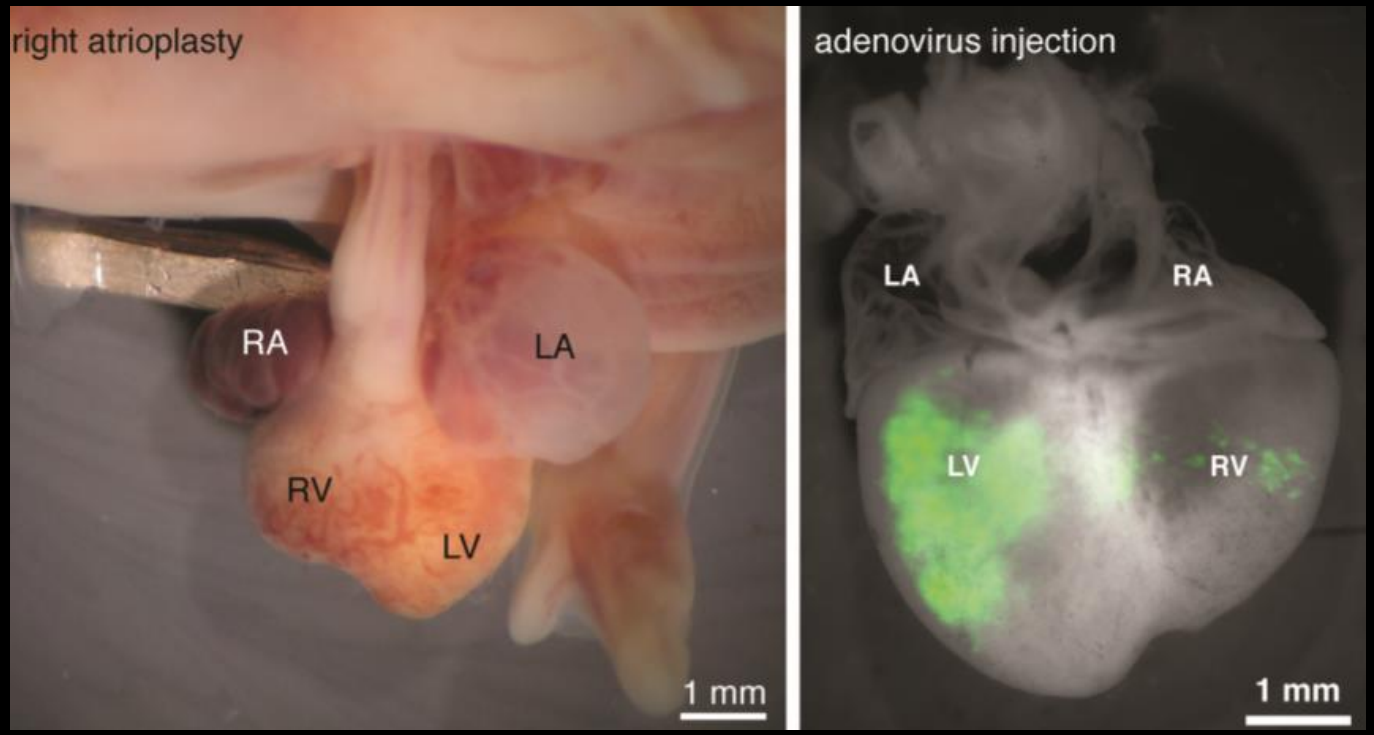
Compact layer: high proliferation

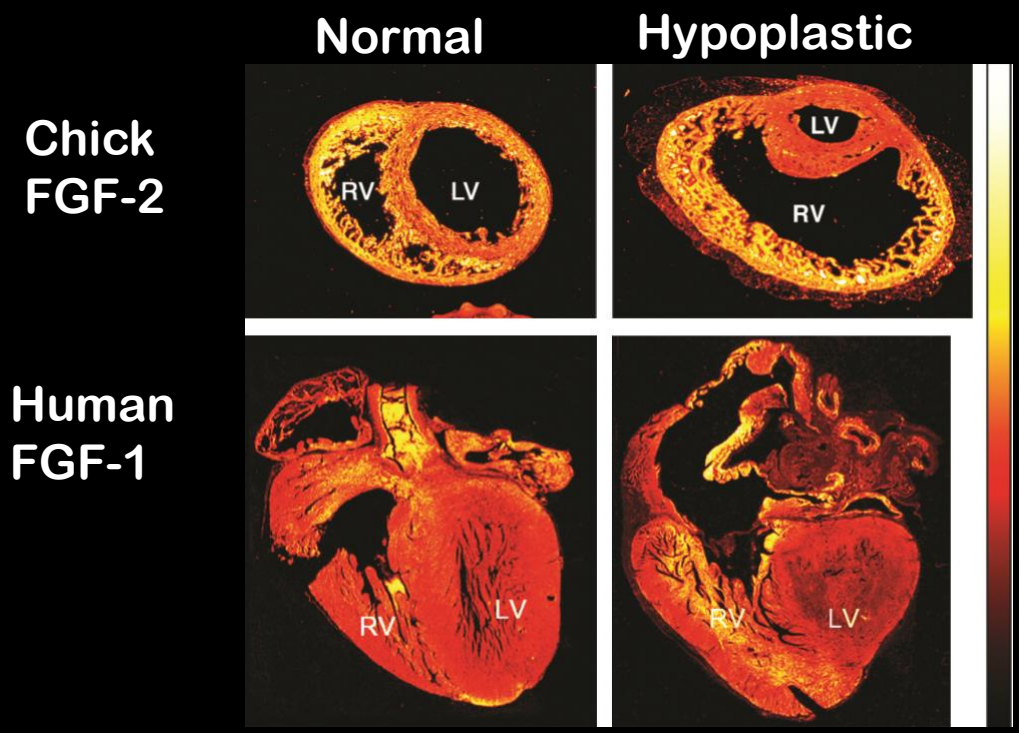
Hypertrophy

Low/no proliferation hypertrophic growth

Cardiac myocyte proliferation and growth
New perspectives

Regulation of cardiomyocytic proliferation by growth factors





HLHS Overall Management

Newborn

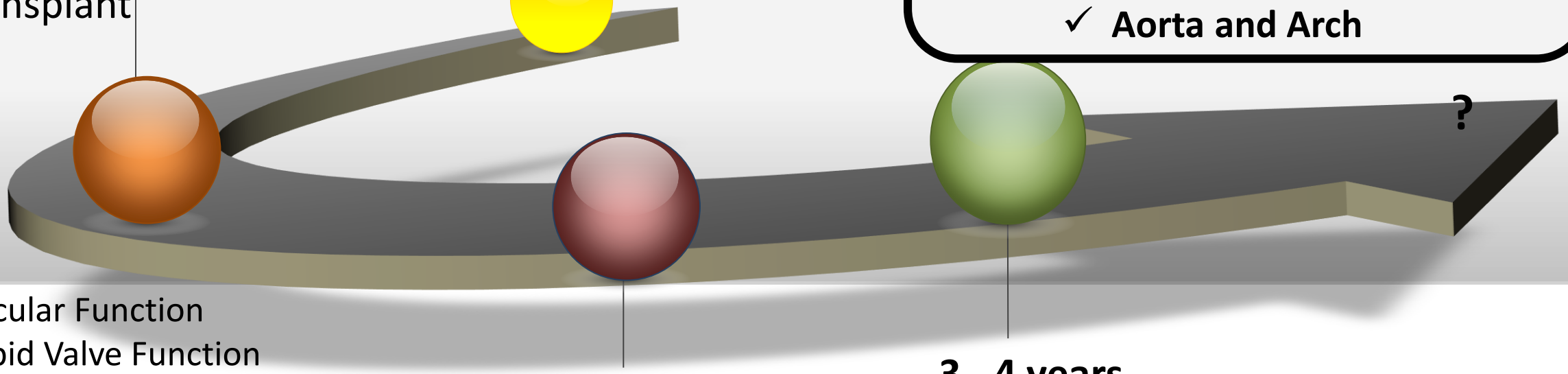
Norwood
Hybrid
Transplant

Fetal

Ductal arch
Atrial septum restriction?
Tricuspid regurgitation?
RV function.

Key Anatomical Points for Outcome

- ✓ **Atrium Septum**
- ✓ **Tricuspid Valve**
- ✓ **Right Ventricle**
- ✓ **Aorta and Arch**



Ventricular Function
Tricuspid Valve Function
Pulmonary Valve Function
Diameter of Asc aorta
Presence of LV to Ca fistula
Aberrant RSCA

4 – 6 months
Glenn Anastomosis

3 - 4 years
Fontan

Echocardiogram Assessment of Hypoplastic Left Heart

Segmental Anatomy & Subtypes

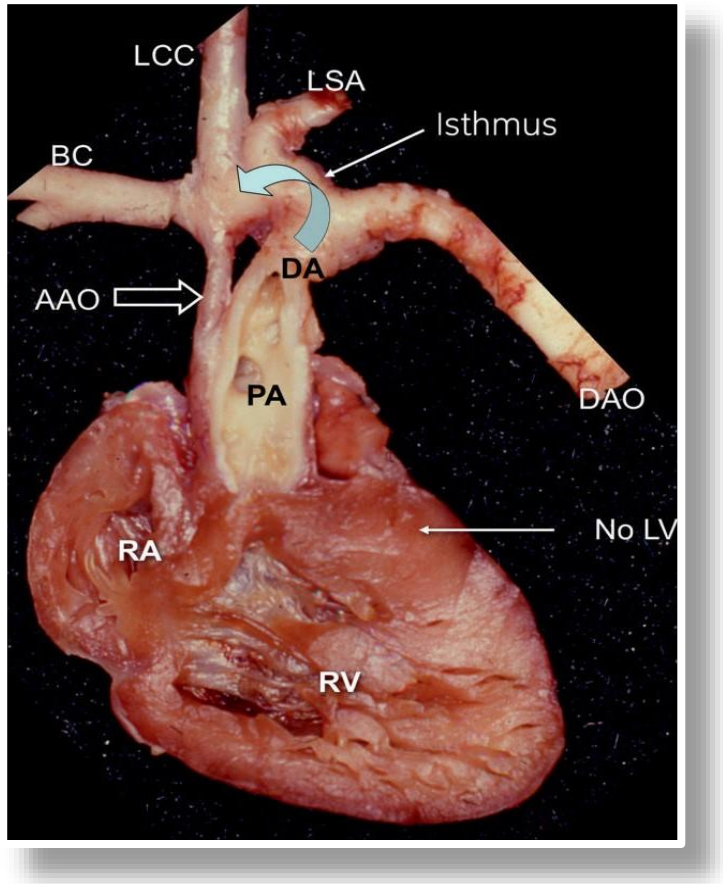
Left atrium, mitral valve, Left ventricular cavity, aortic valve, ascending aorta, aortic arch, and isthmus

Pre-Surgical assessment

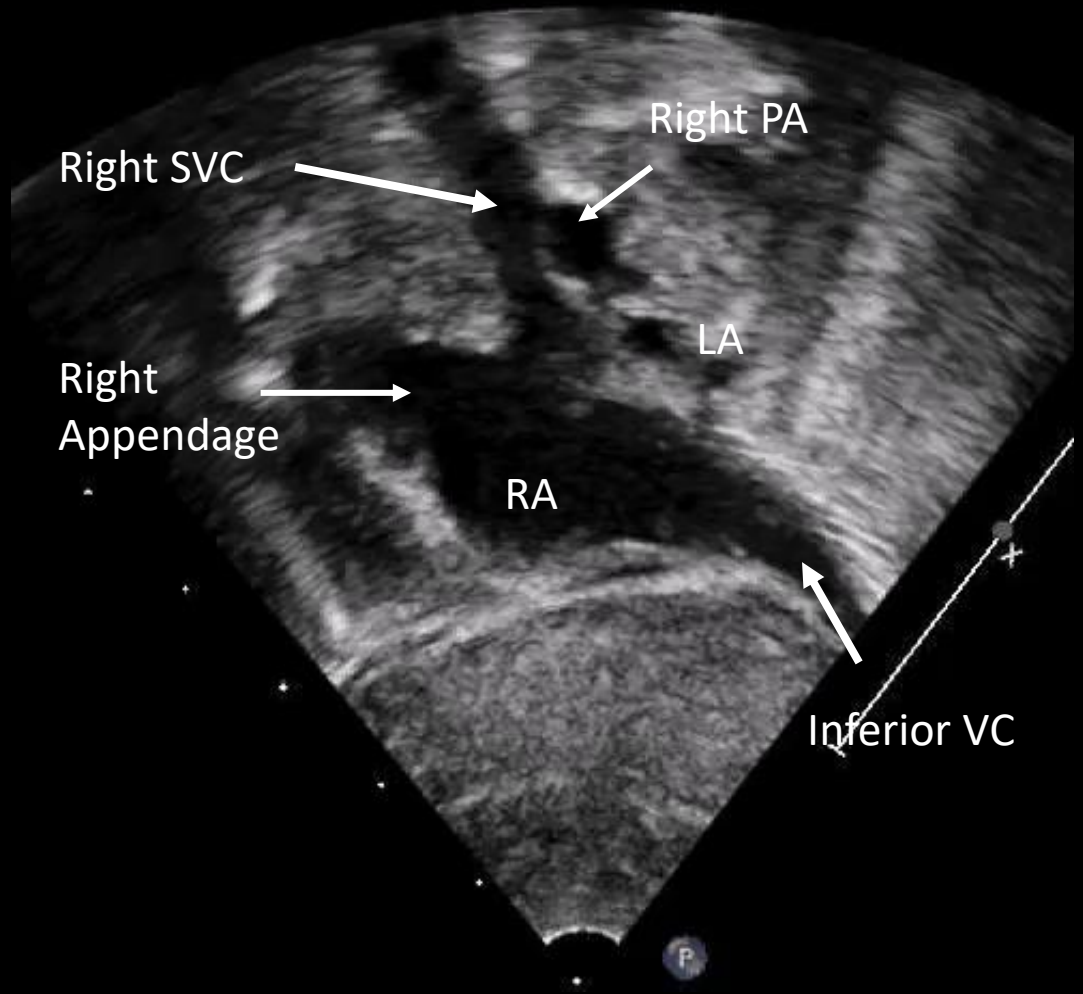
Atrial Communication; Ductal Arch ; TV and RV function

Tips & Traps

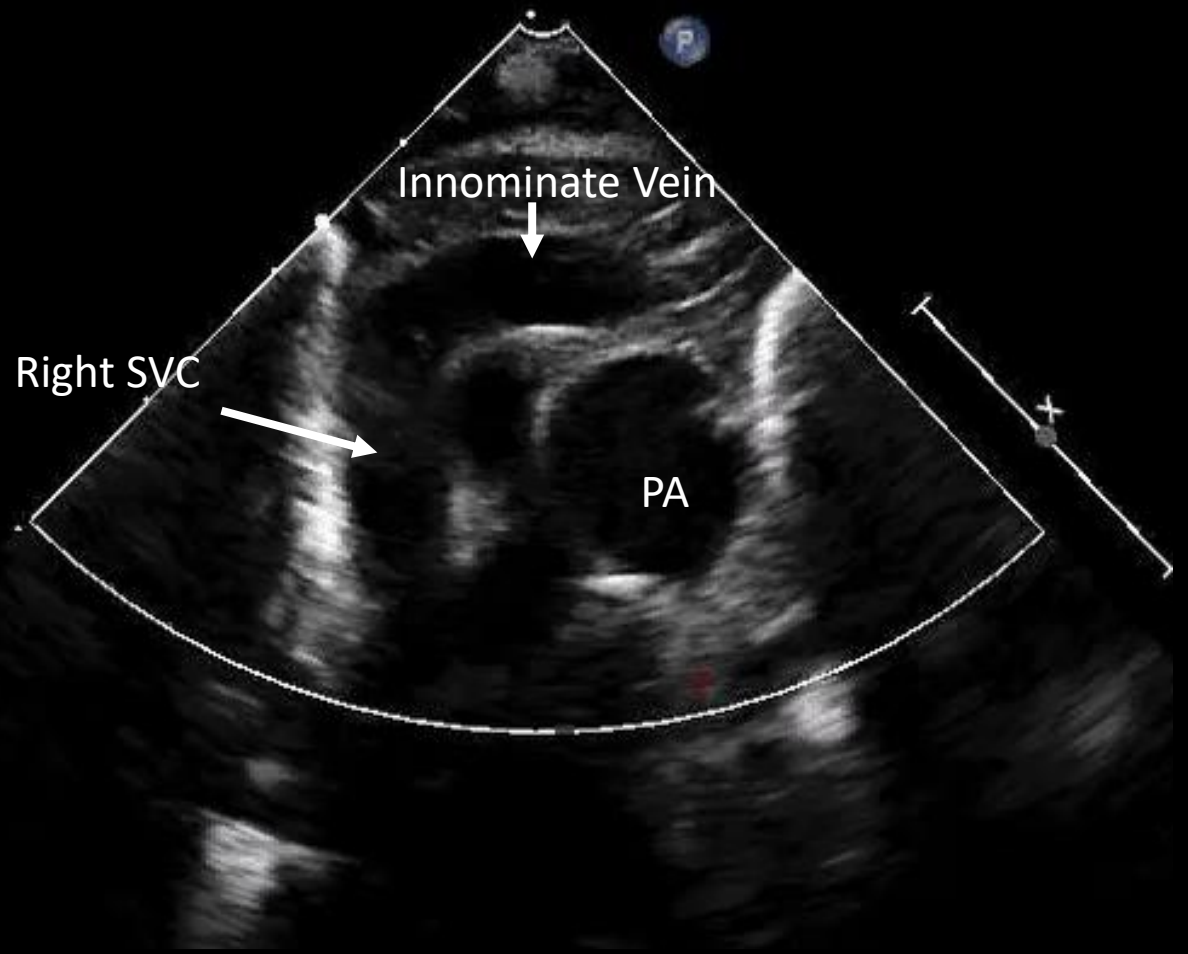
Coronary arteries; Decompressing vein and Arch abnormality



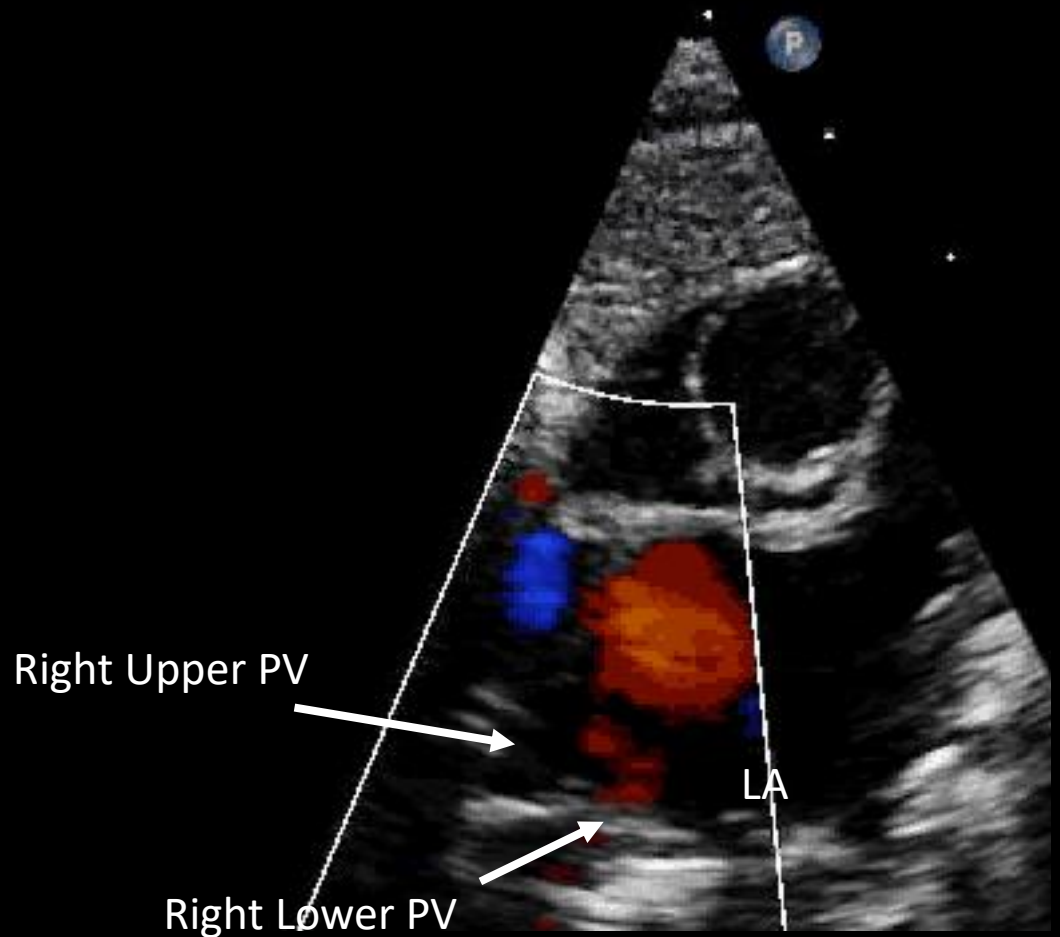
HLHS Venous Anatomy

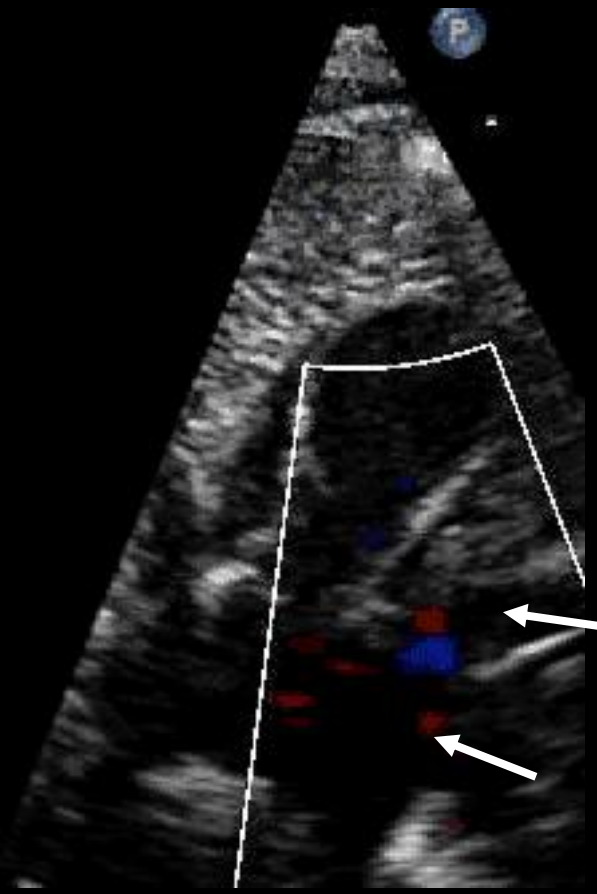


HLHS Venous Anatomy

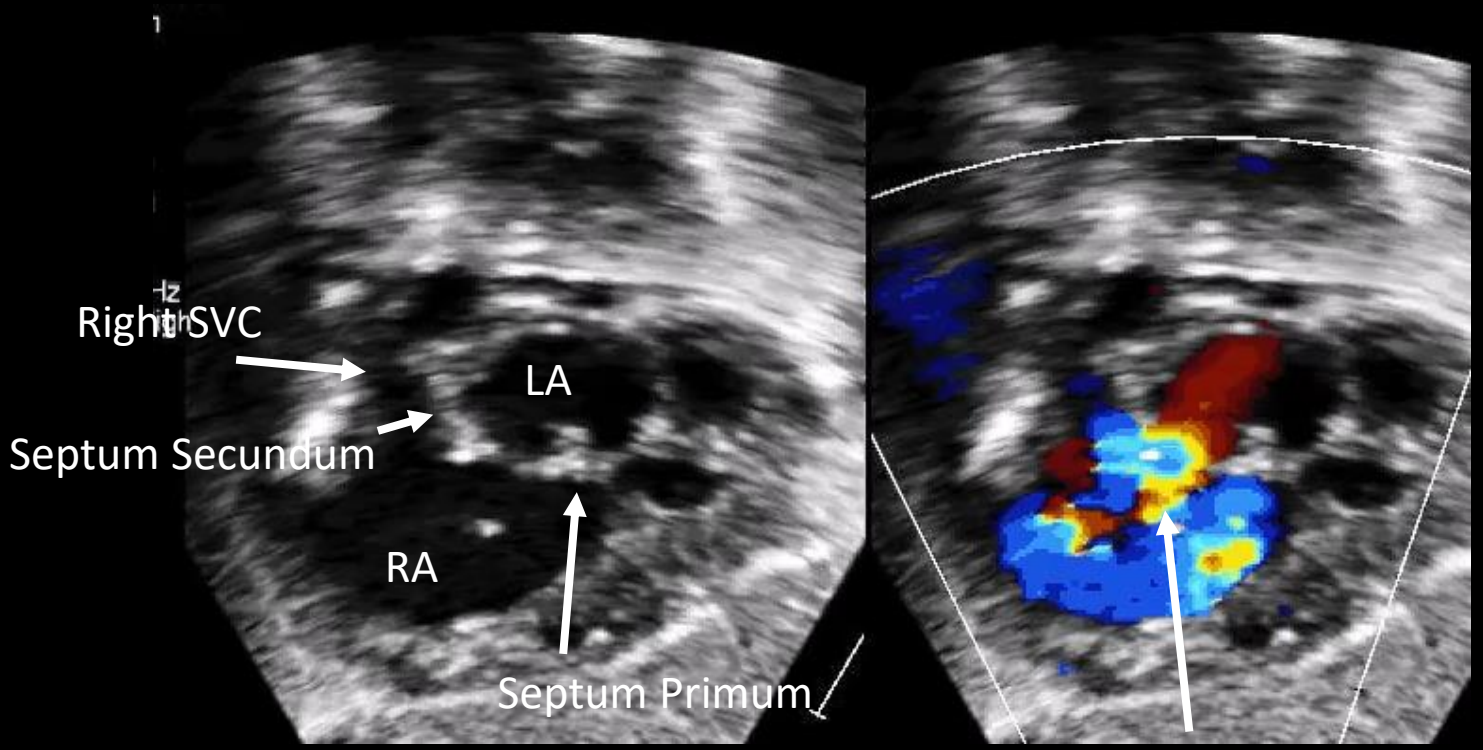


HLHS Venous Anatomy

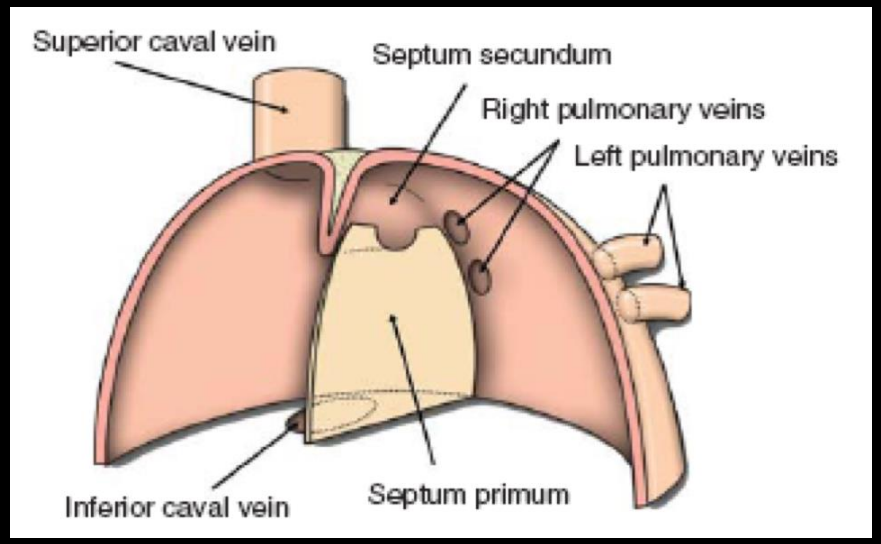


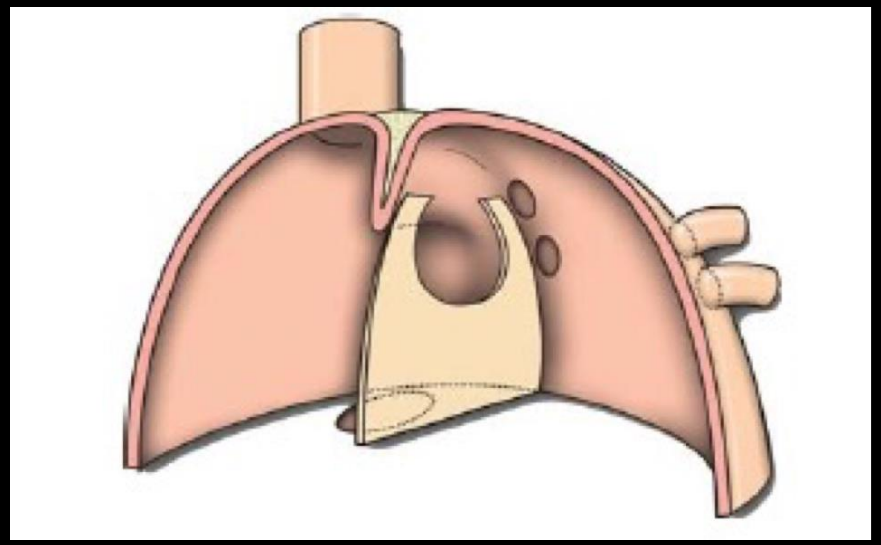


HLHS : Atrium & Inter atrial septum



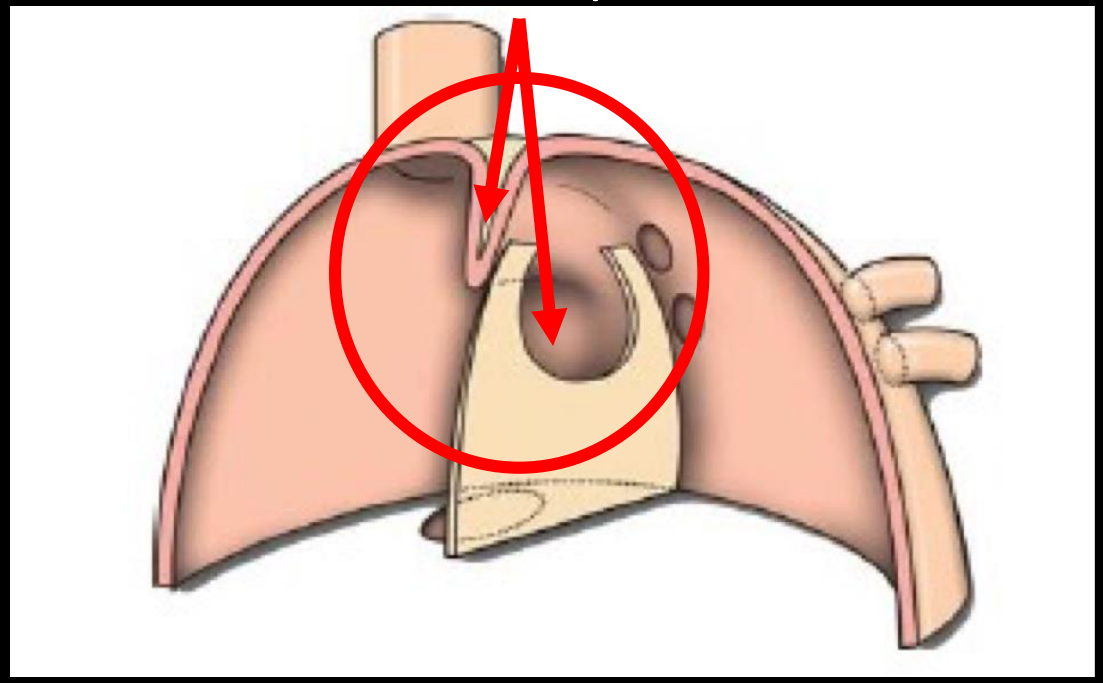
Stretched Foramen Ovale



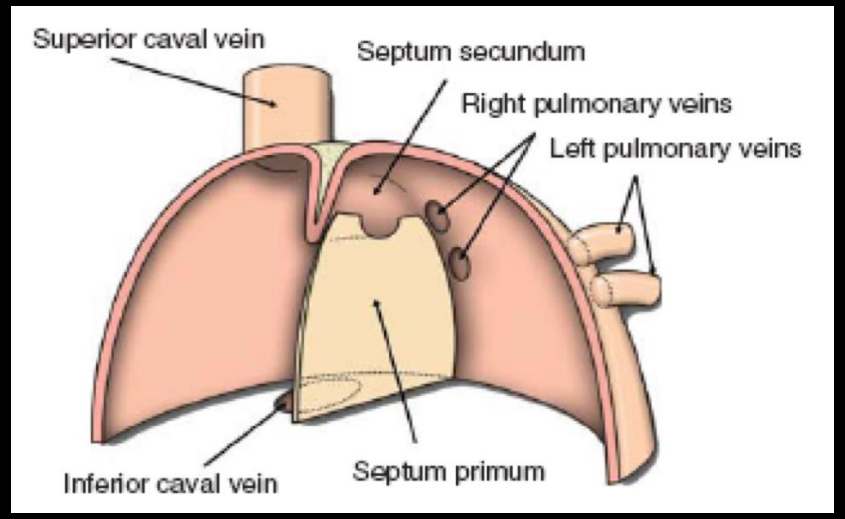


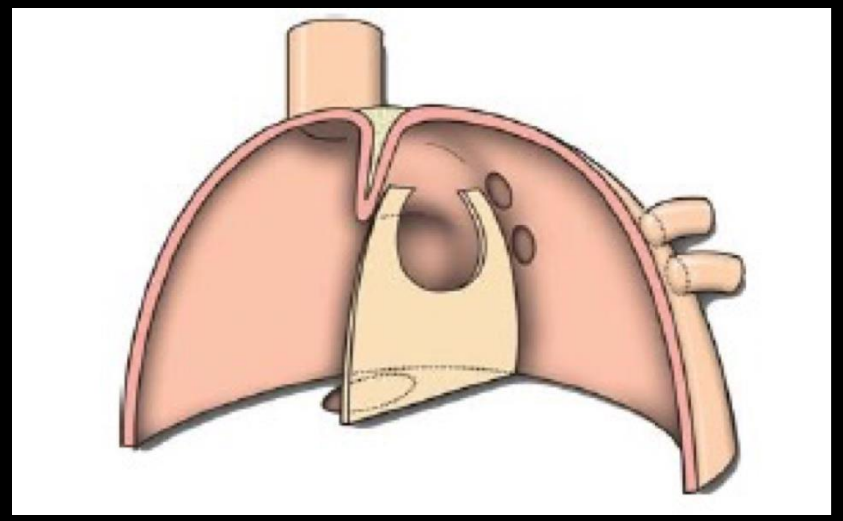
HLHS : Atrium & Inter atrial septum

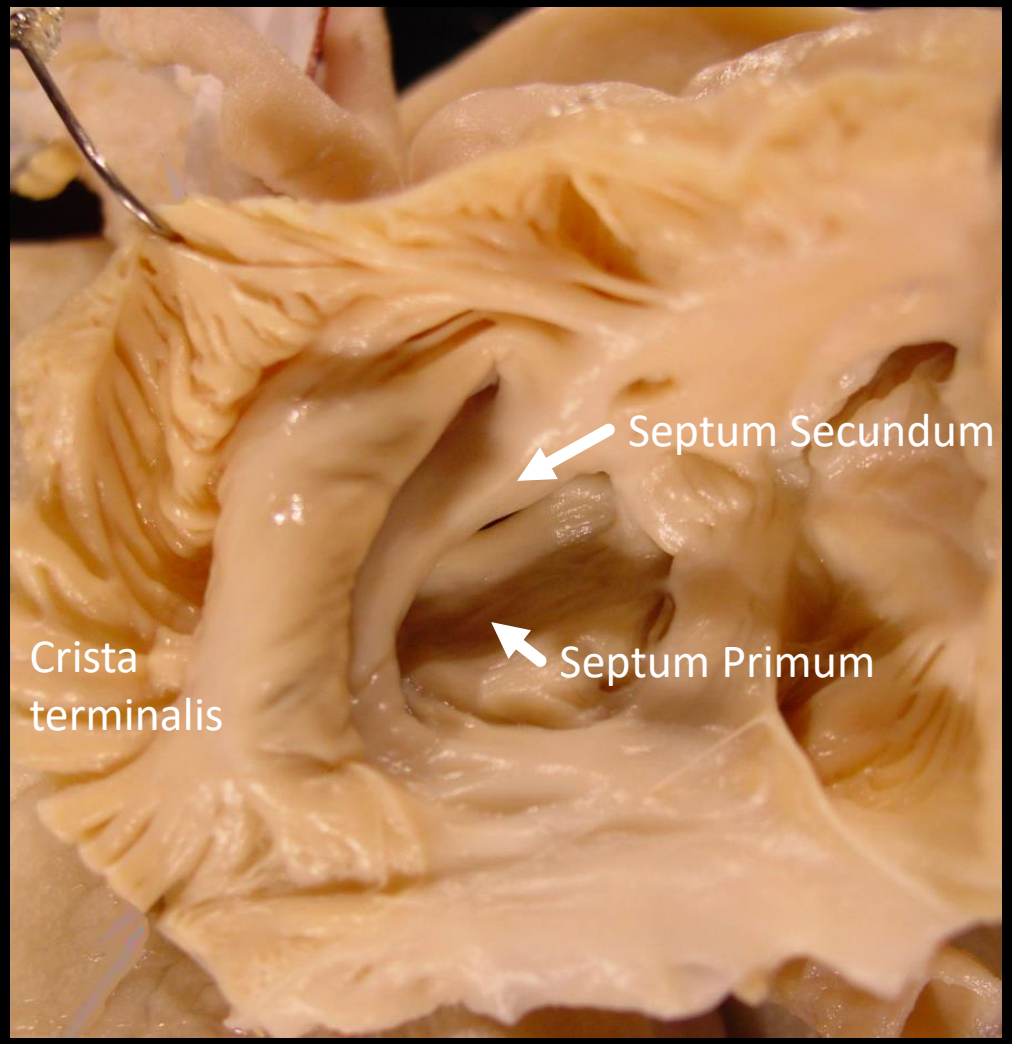
Normal attachment of the septum primum to the infolded superior rim of the foramem ovale, Septum secundum

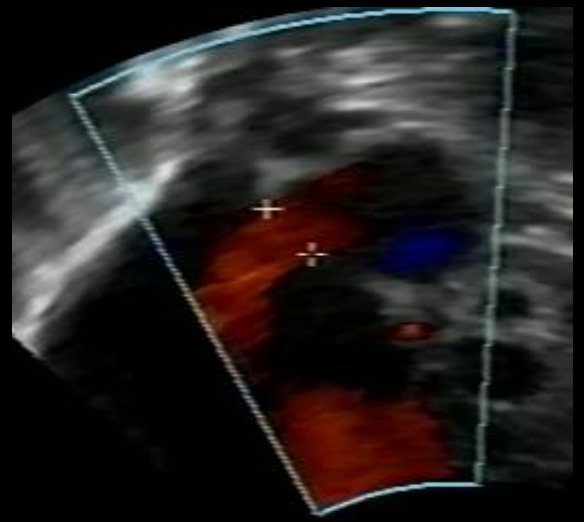


HLHS : Morphology of the Interatrial Septum



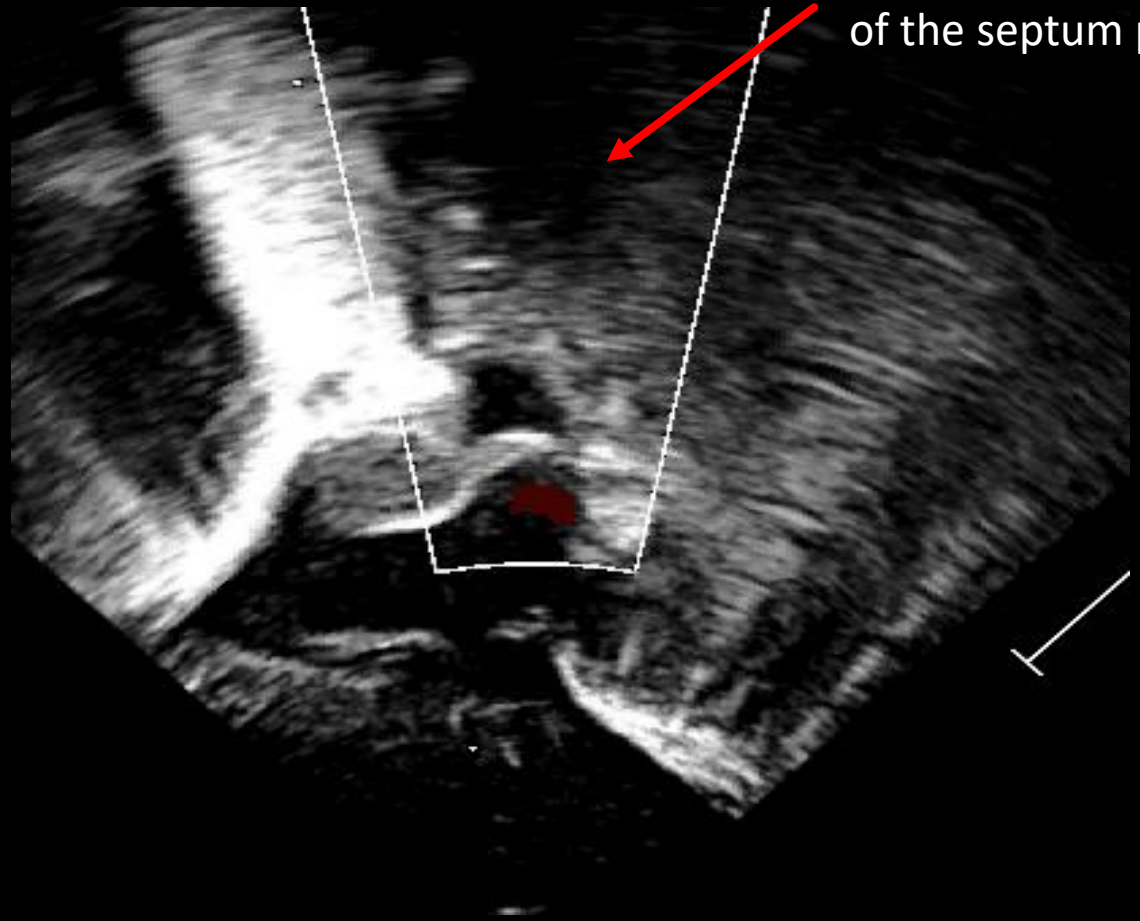




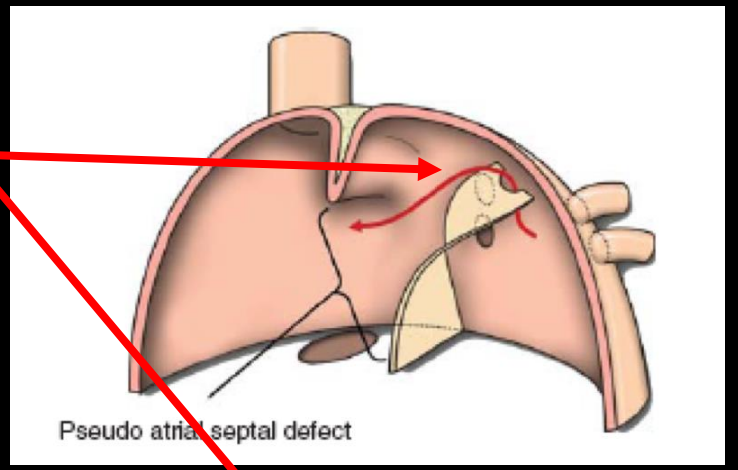


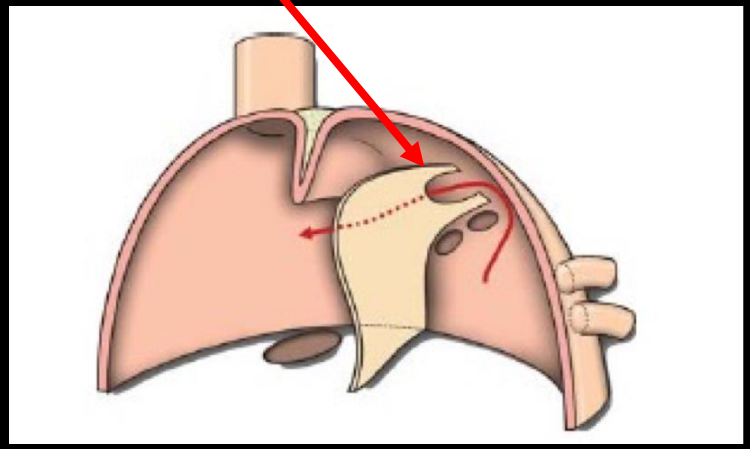
Courtesy Prof. Vera Aiello - Incor FMUSP

HLHS : Morphology of the Interatrial Septum



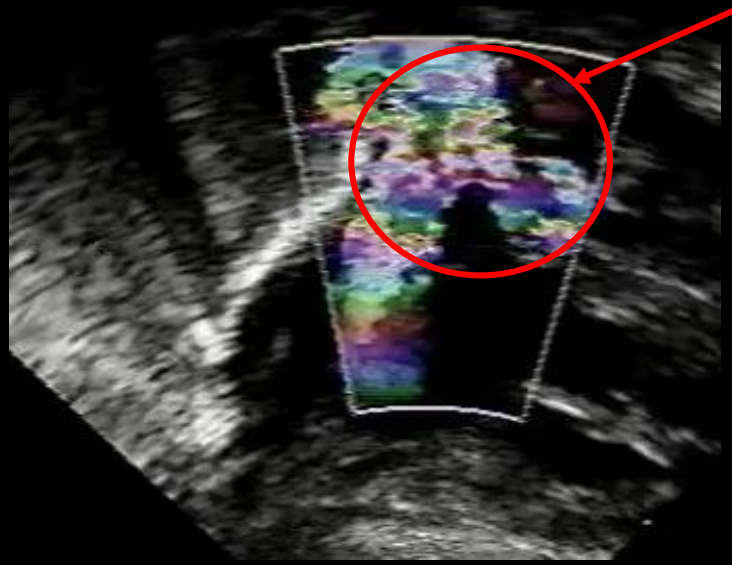
Leftward displacement
of the septum primum

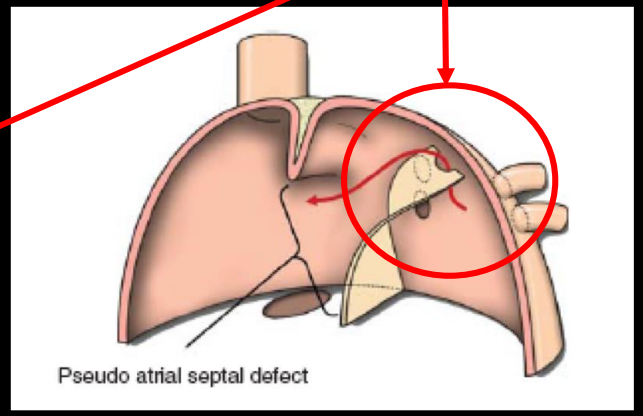


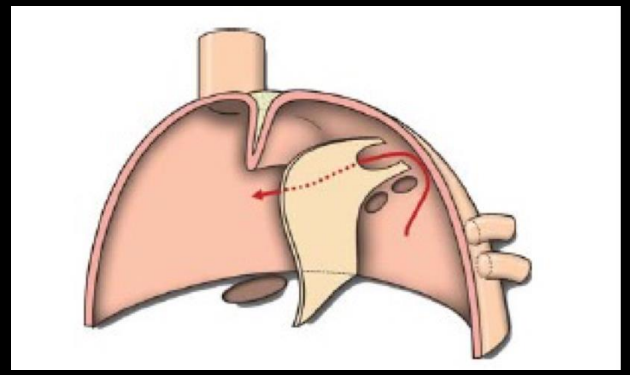


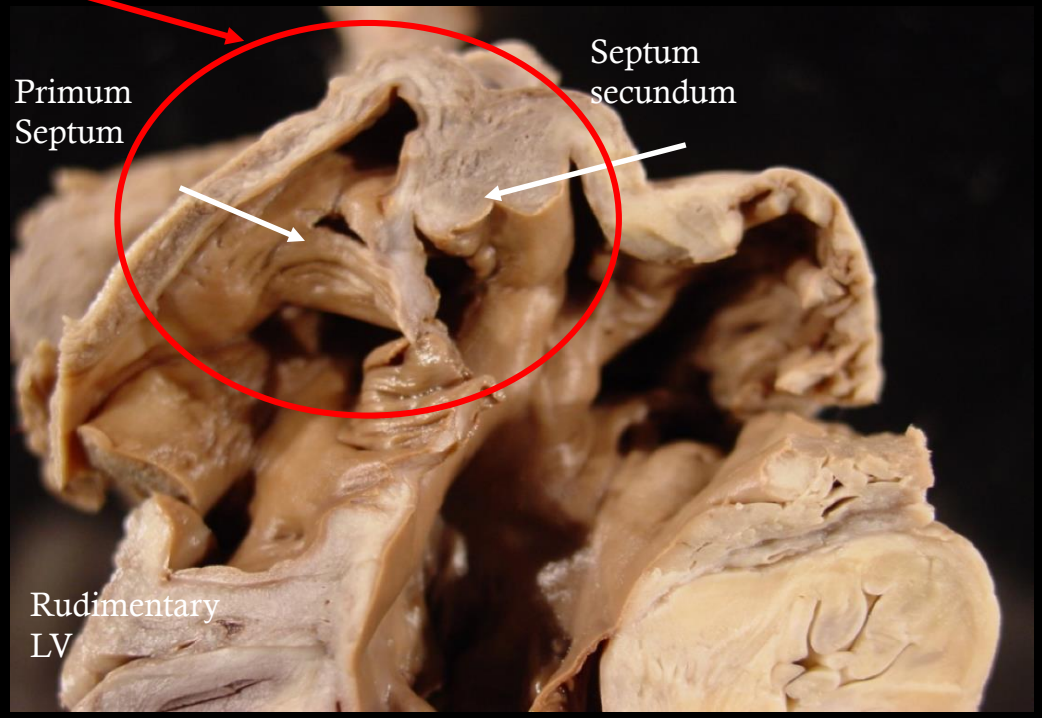
HLHS : Morphology of the Interatrial Septum

Leftward displacement of the flap valve or septum primum: “Pseudo atrial septal defect and bulging of the displaced flap valve formed by the primary atrial septum, or septum primum, due to increased postnatal pulmonary venous return, with the oval foramen partially obstructed as it apposes the roof of left atrium





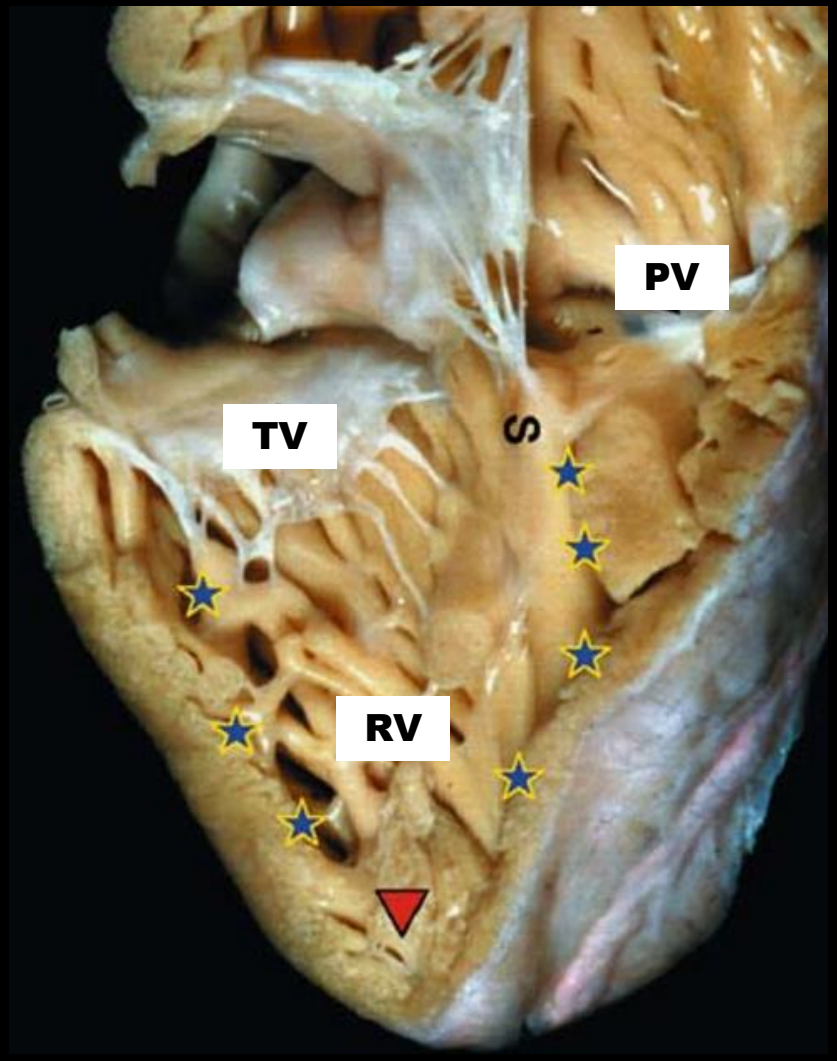




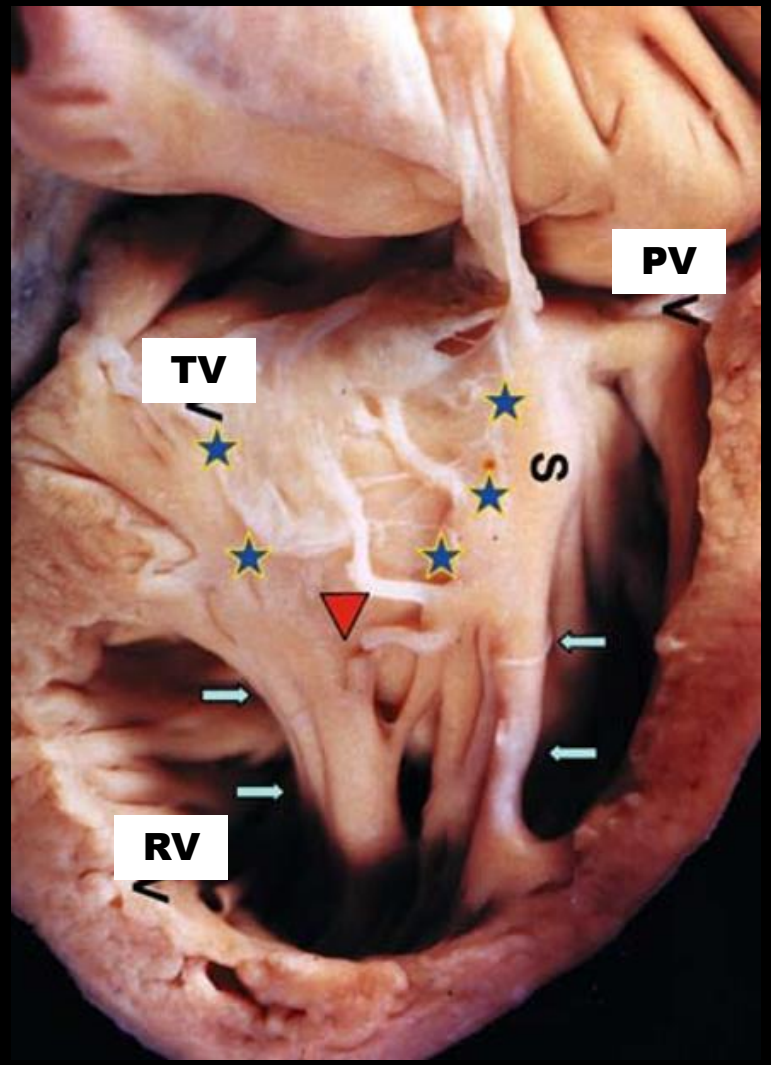
Courtesy Prof. Vera Aiello - Incor FMUSP

HLHS : Morphology of the Right Ventricle & Tricuspid Valve

Normal heart

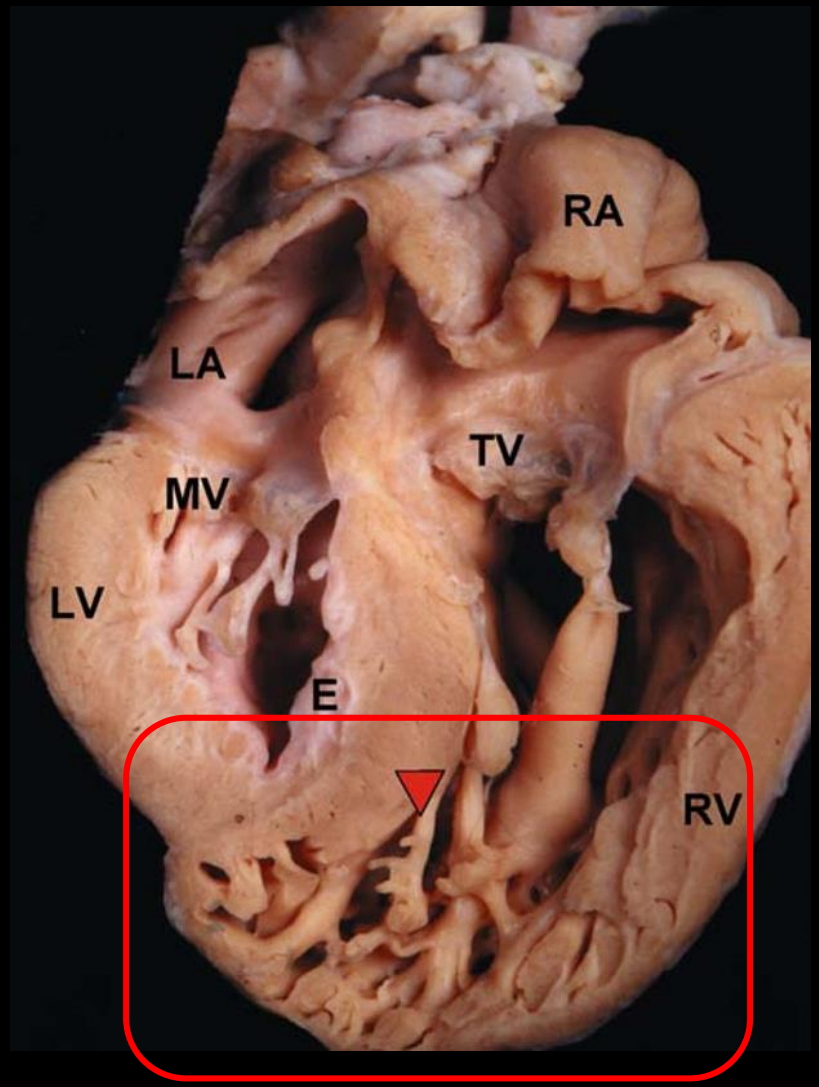


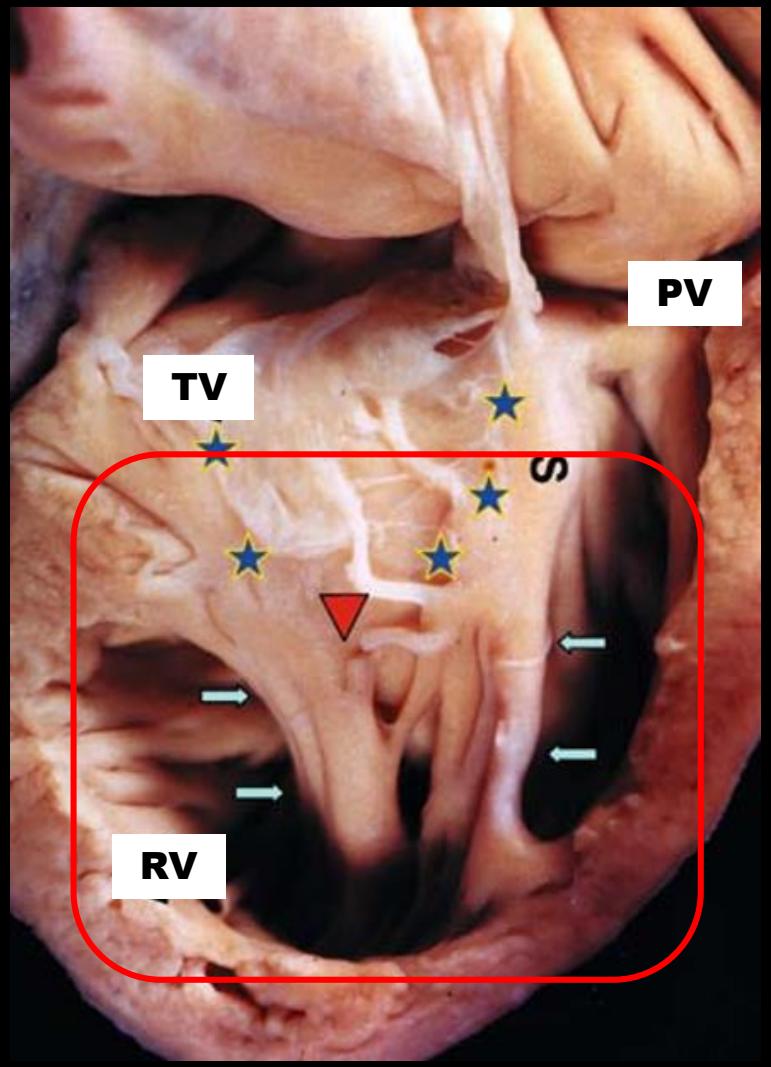
Hypoplastic Left heart

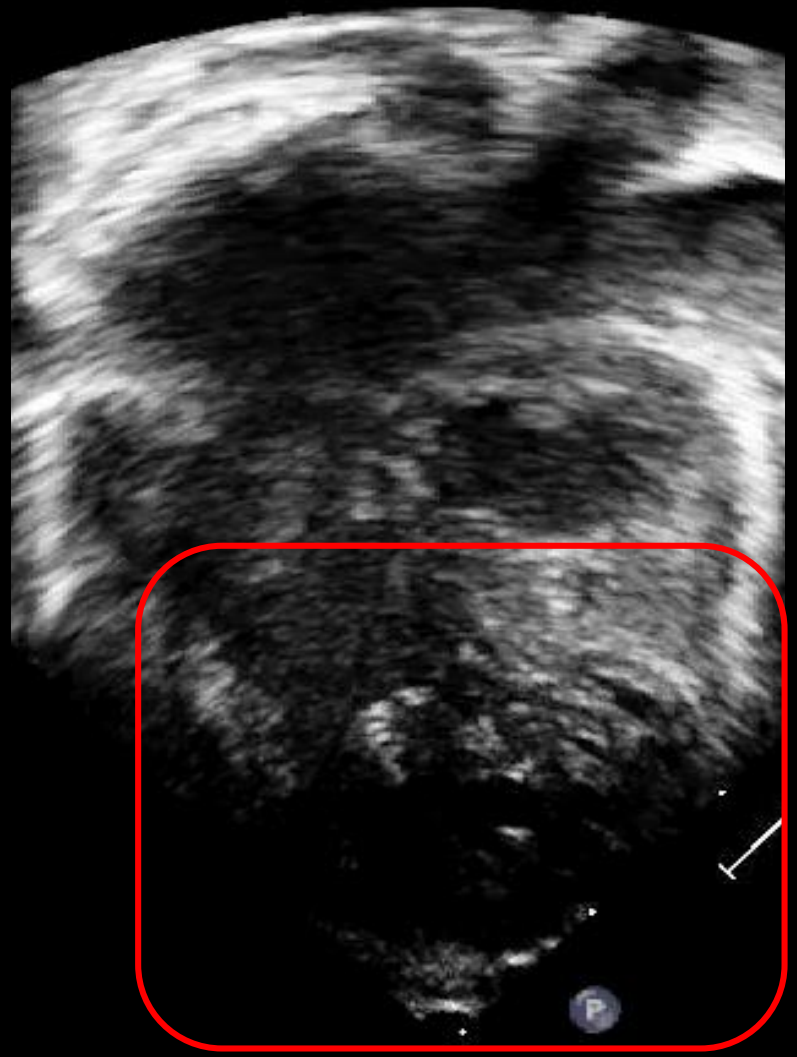


HLHS : Morphology of the Right Ventricle & Tricuspid Valve

Septomarginal Trabeculation is abnormal

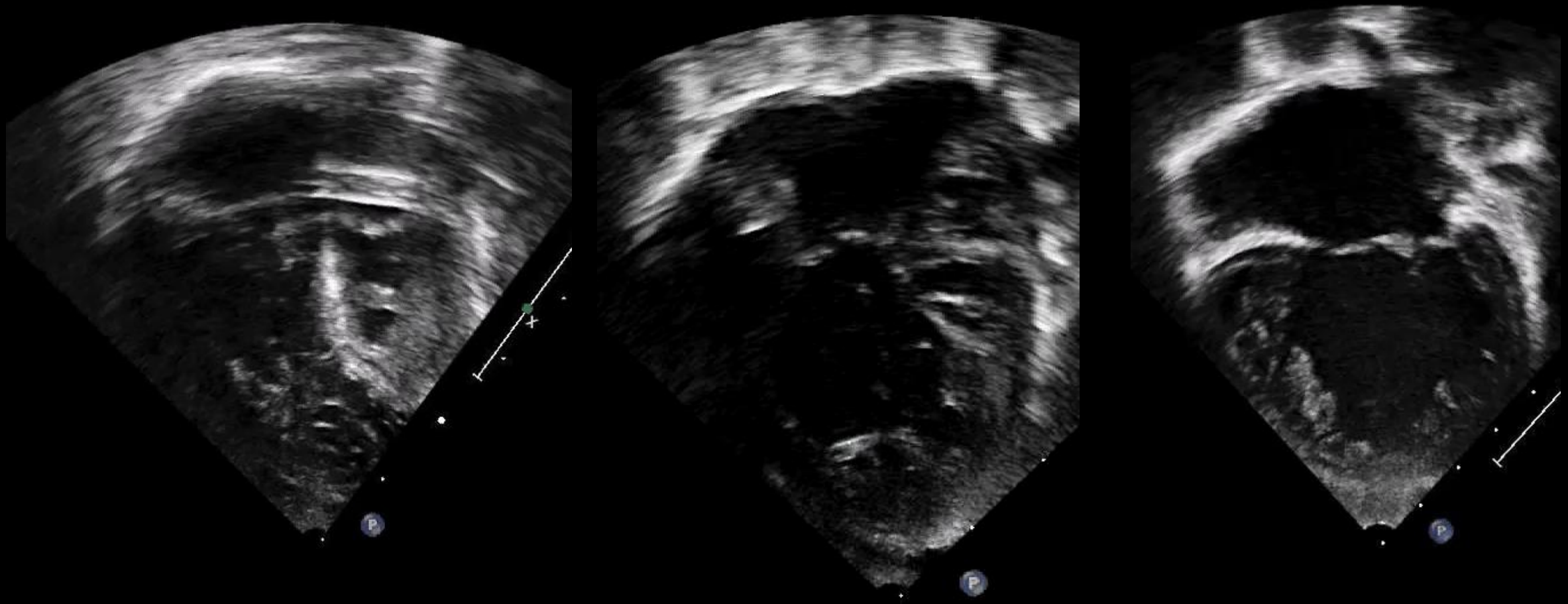




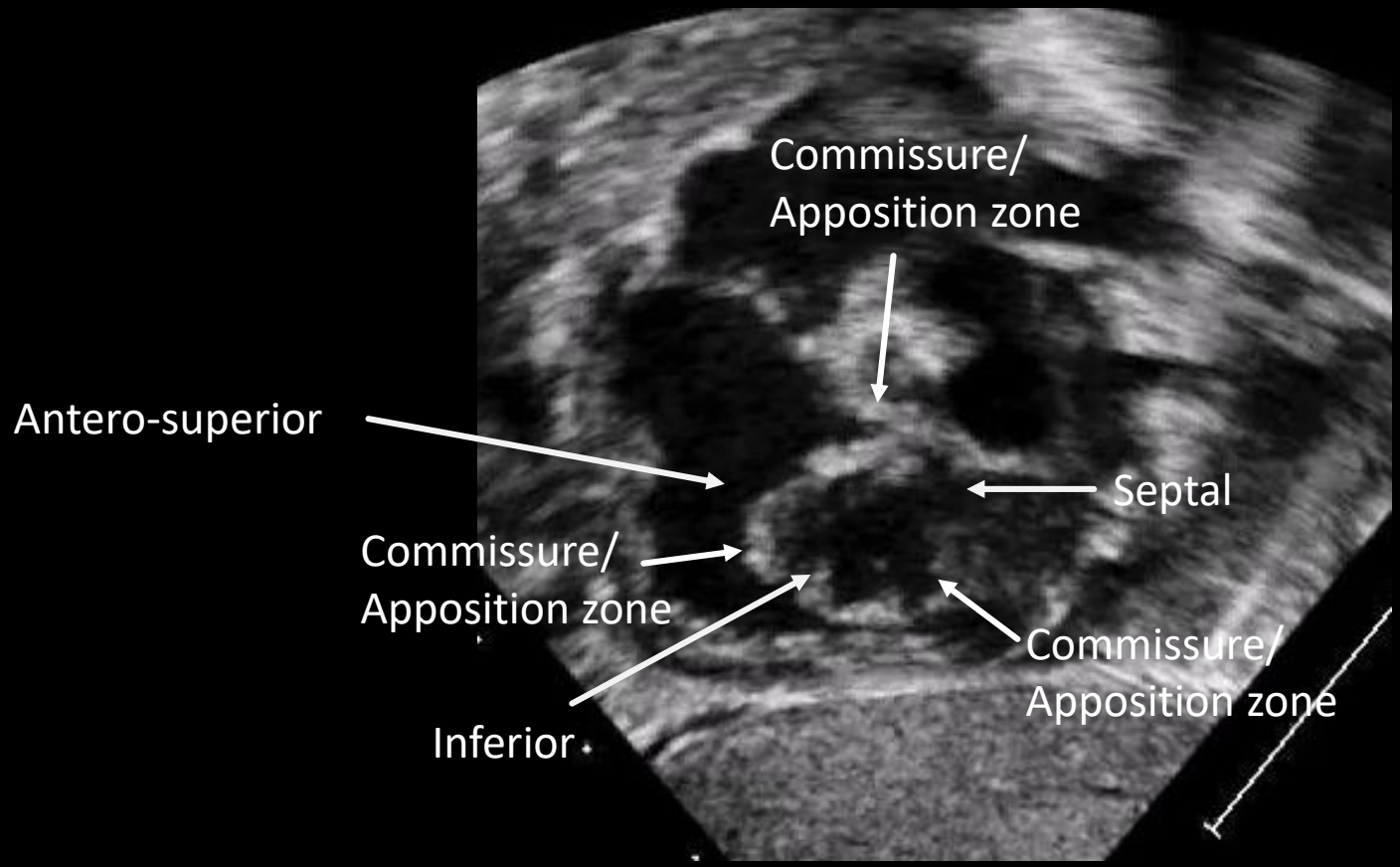


HLHS : Morphology of the Tricuspid Valve

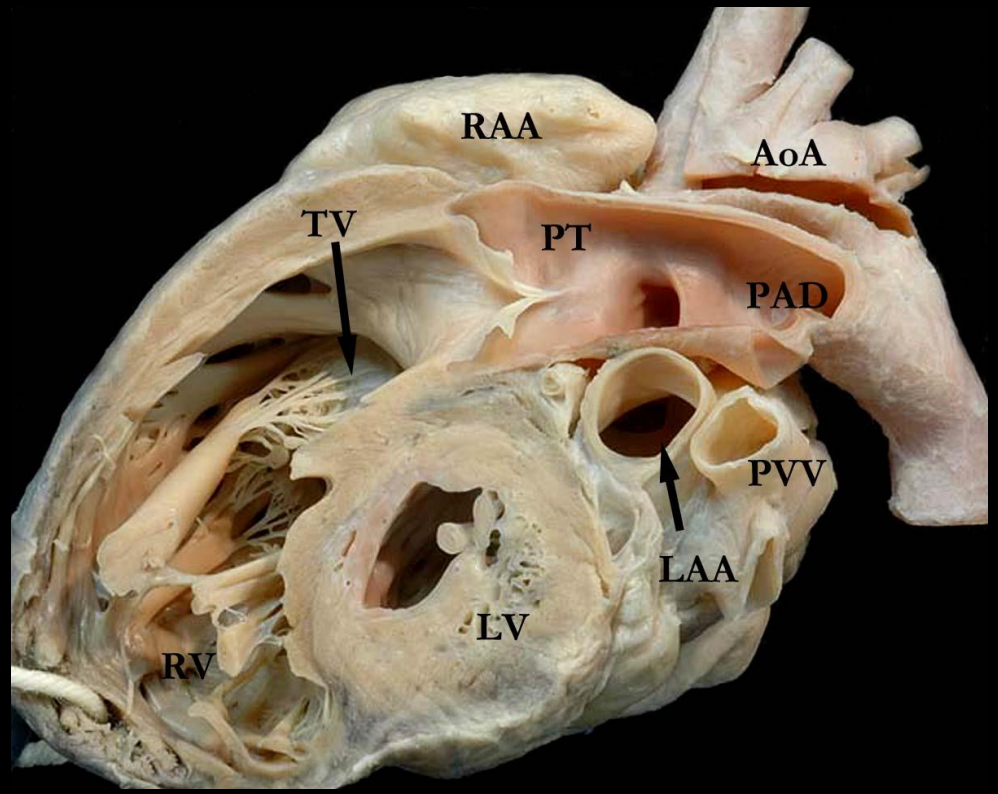
……Normal , dilated annulus, dysplastic (VSD), myxomatous, thickened, redundant, bi – leaflet (“mitralisation of the TV) …..

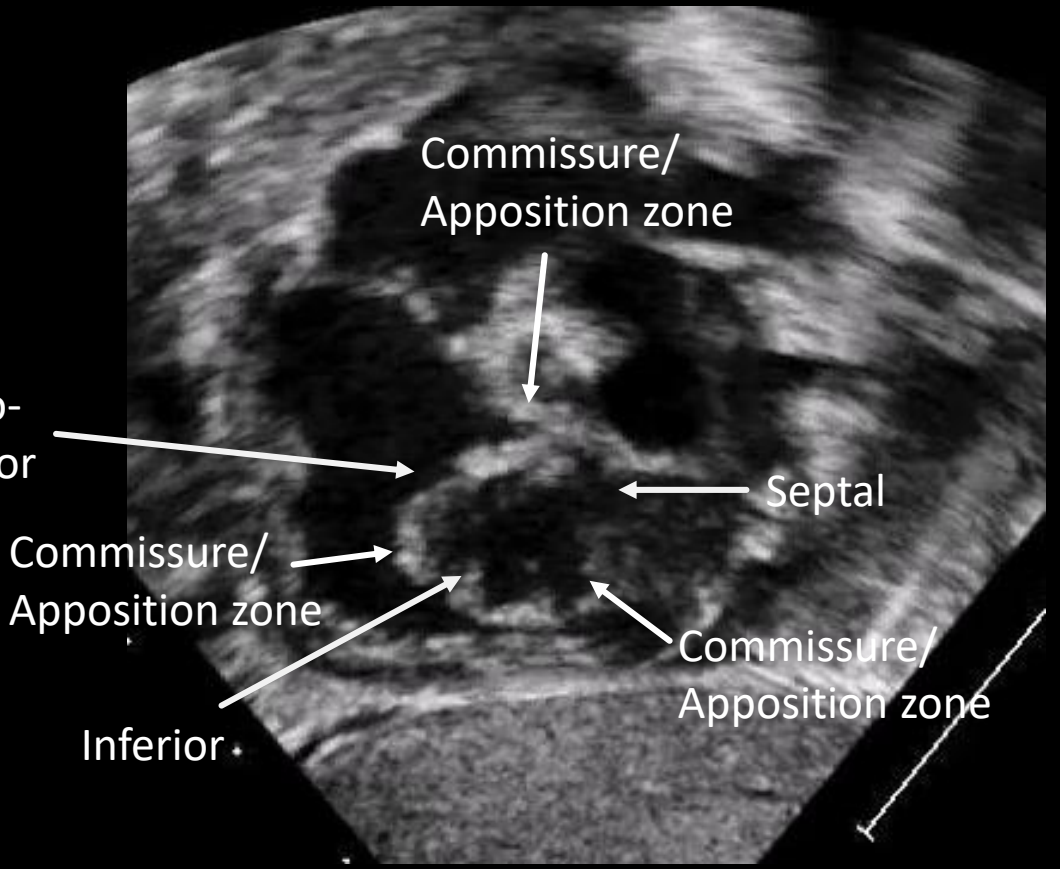


HLHS : Morphology of the Tricuspid Valve



HLHS : Morphology of the Tricuspid Valve





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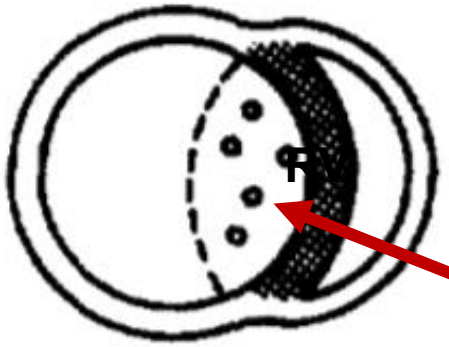
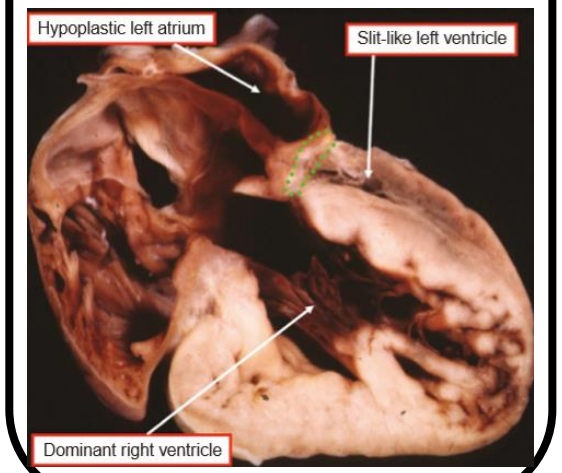
HLHS : Morphology of the Septal Leaflet of the Tricuspid Valve & Relationship with Interventricular Septum

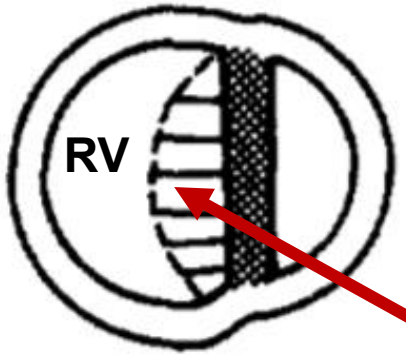
Concave

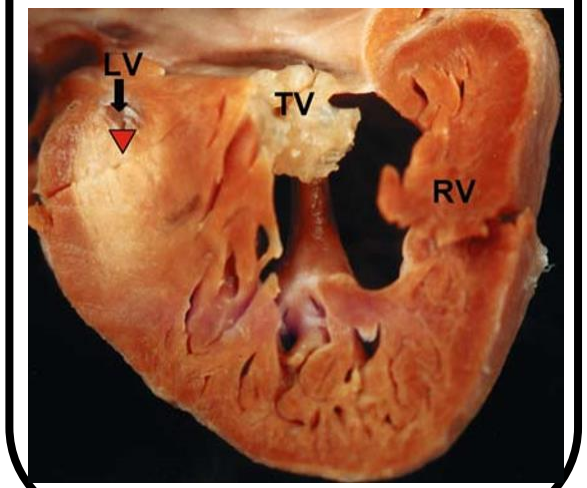


Septal leaflet supporting multiples muscles arising from septal part



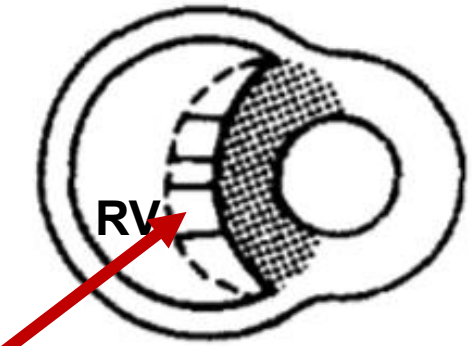
Straight

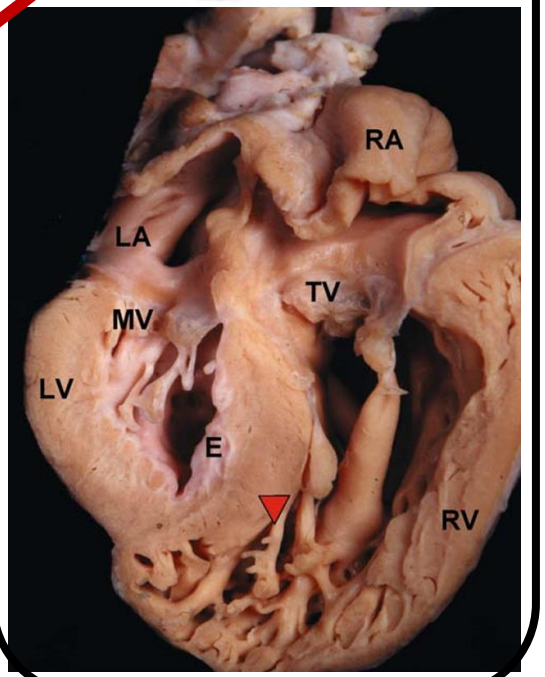




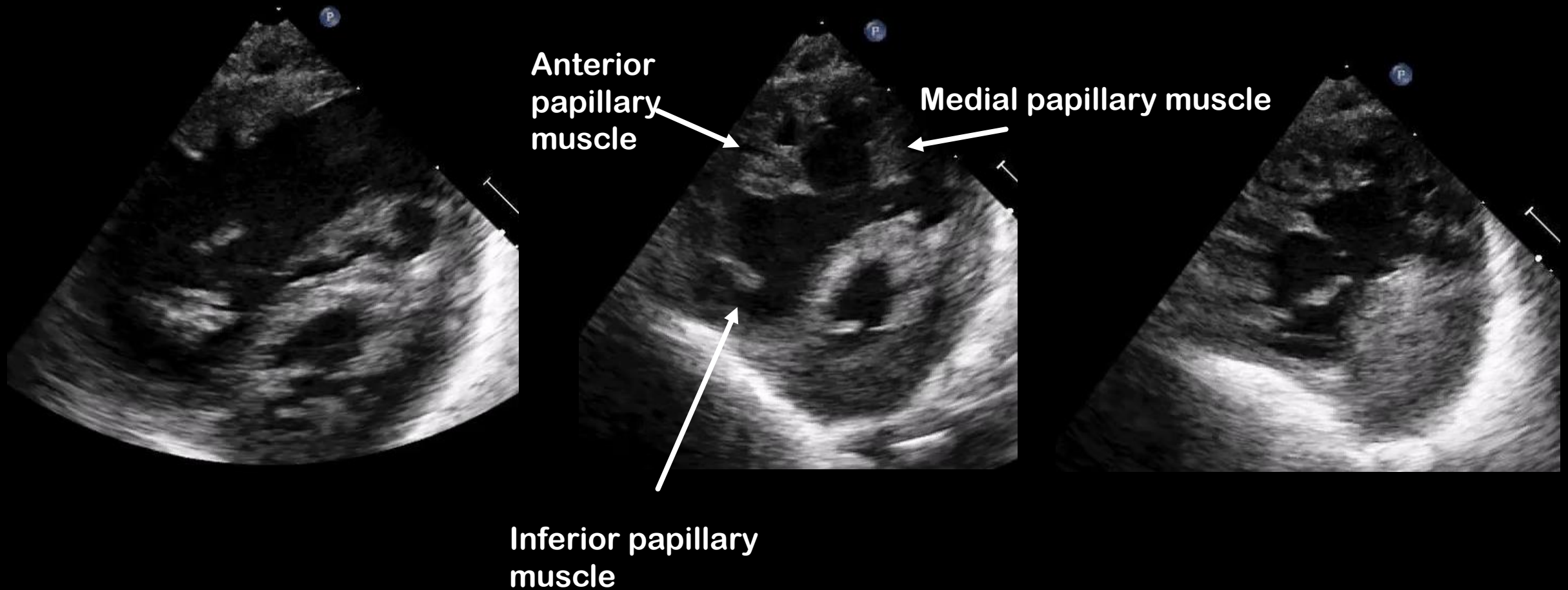
Direct cords arising from the septum supporting septal leaflet

Convex

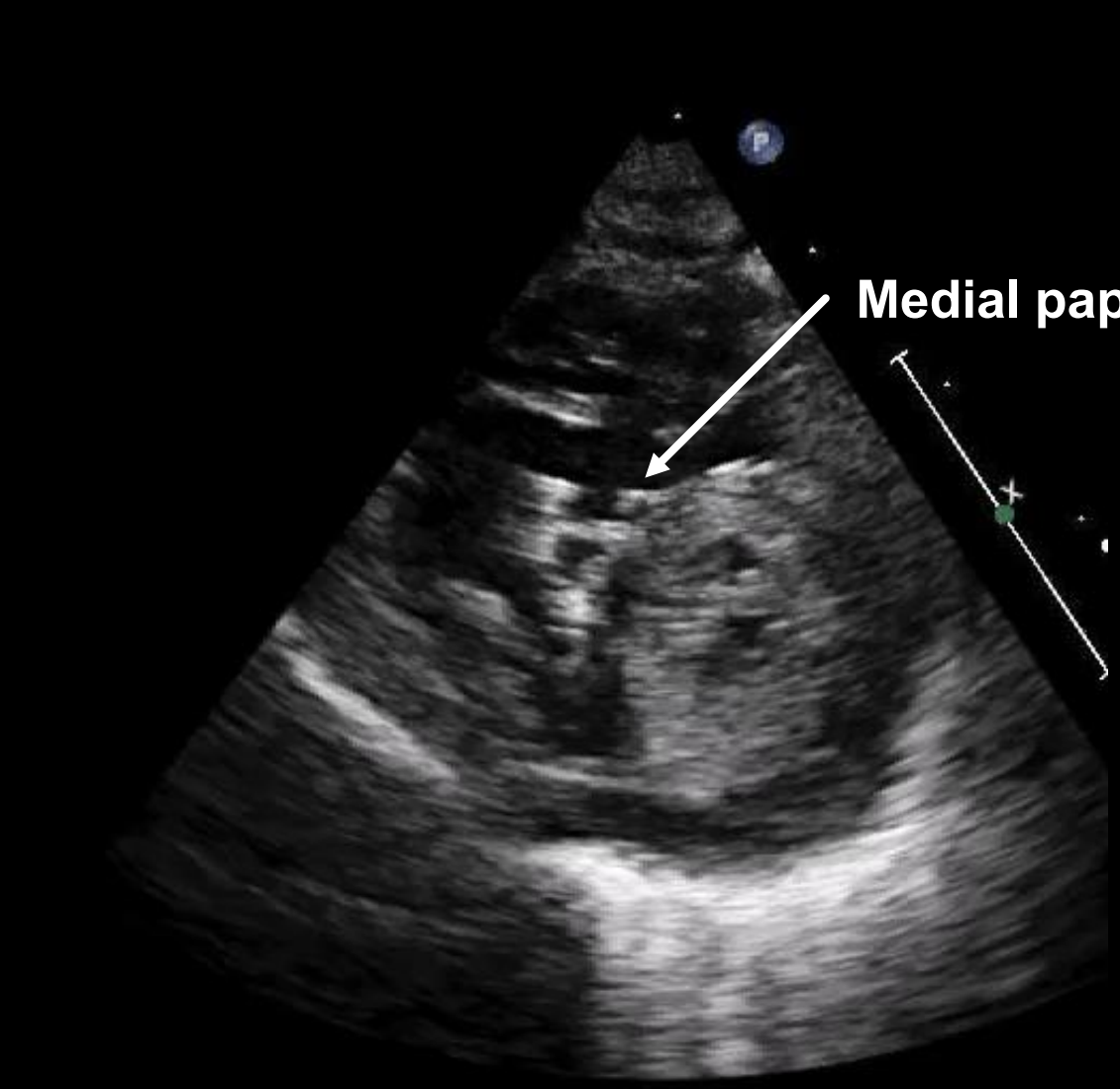


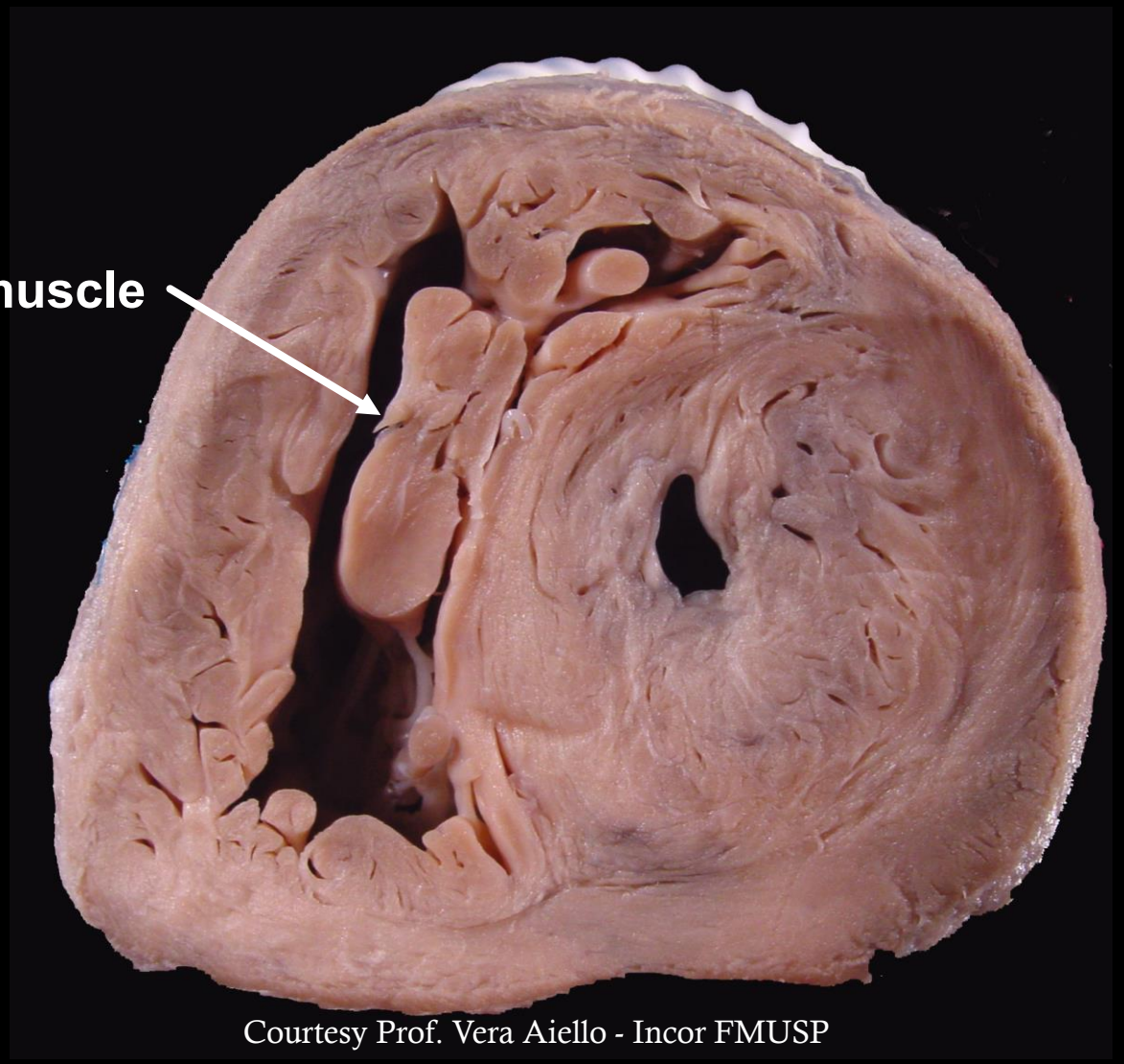


HLHS : Morphology of the Tricuspid Valve



HLHS : Morphology of the Tricuspid Valve





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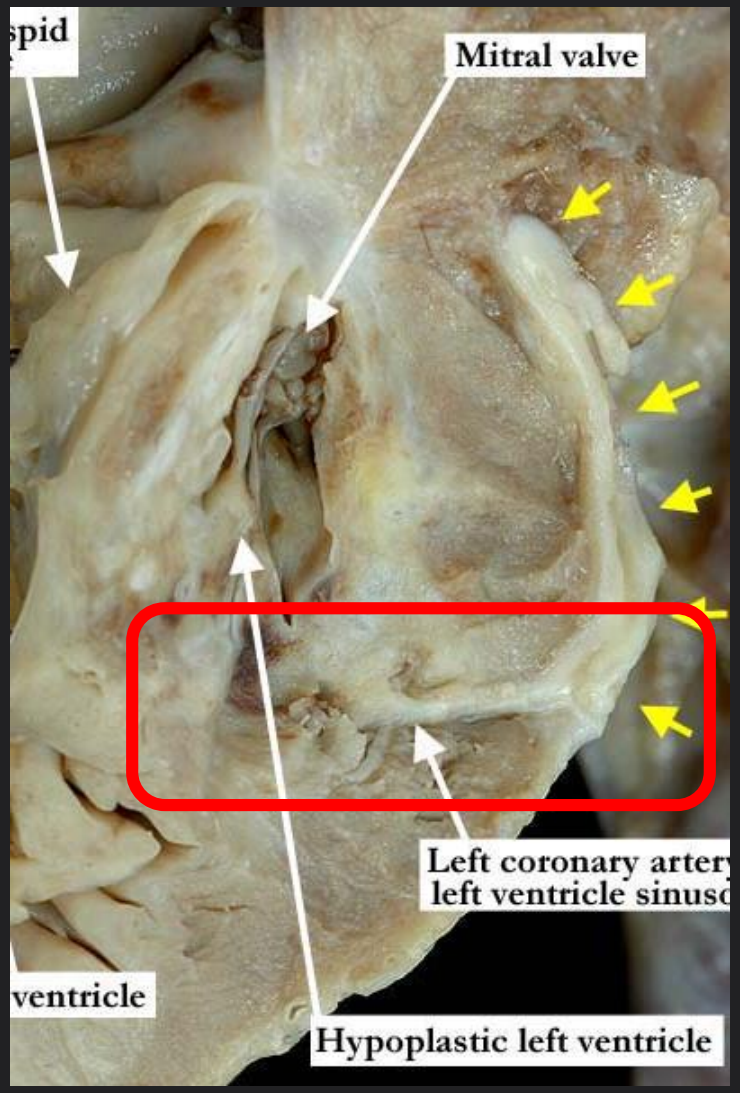
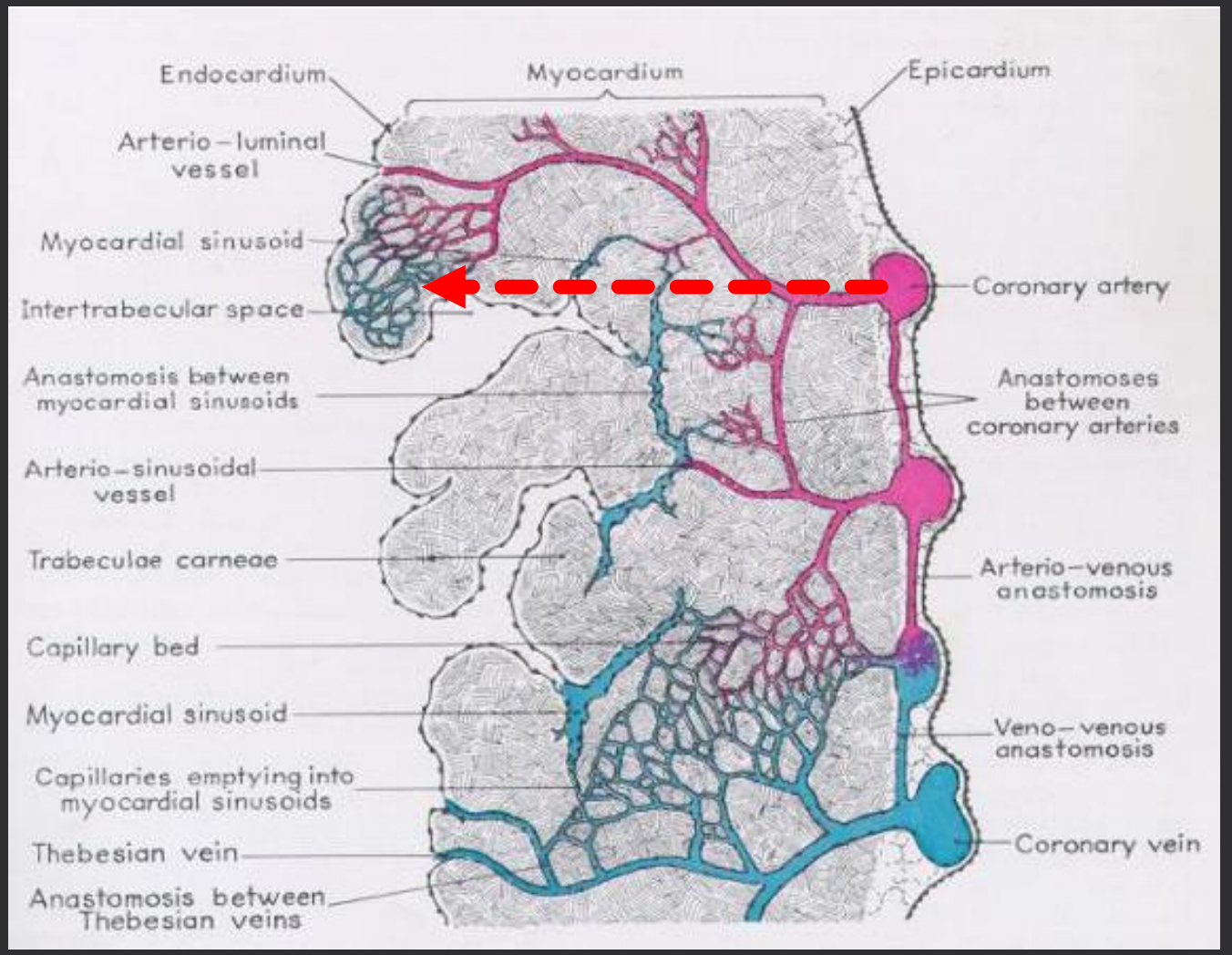
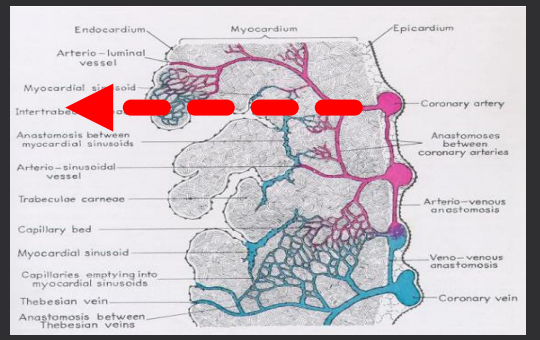
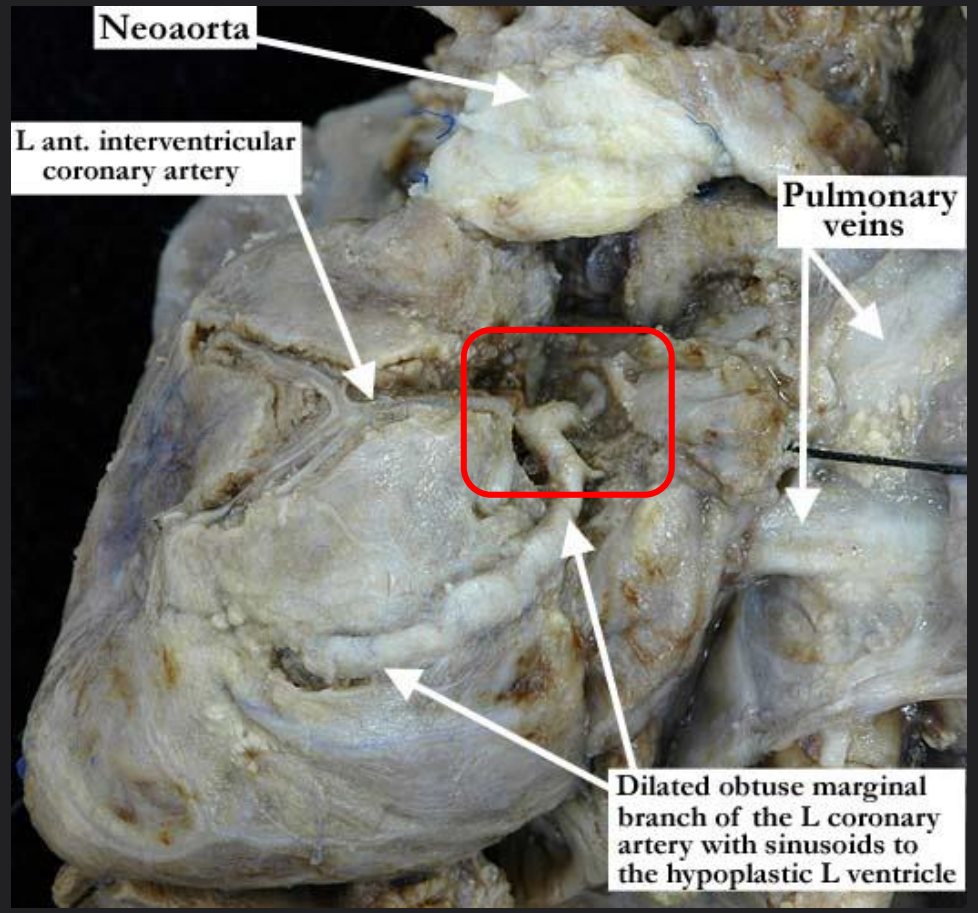
Coronary abnormalities: HLHS
Ventricle-coronary connections

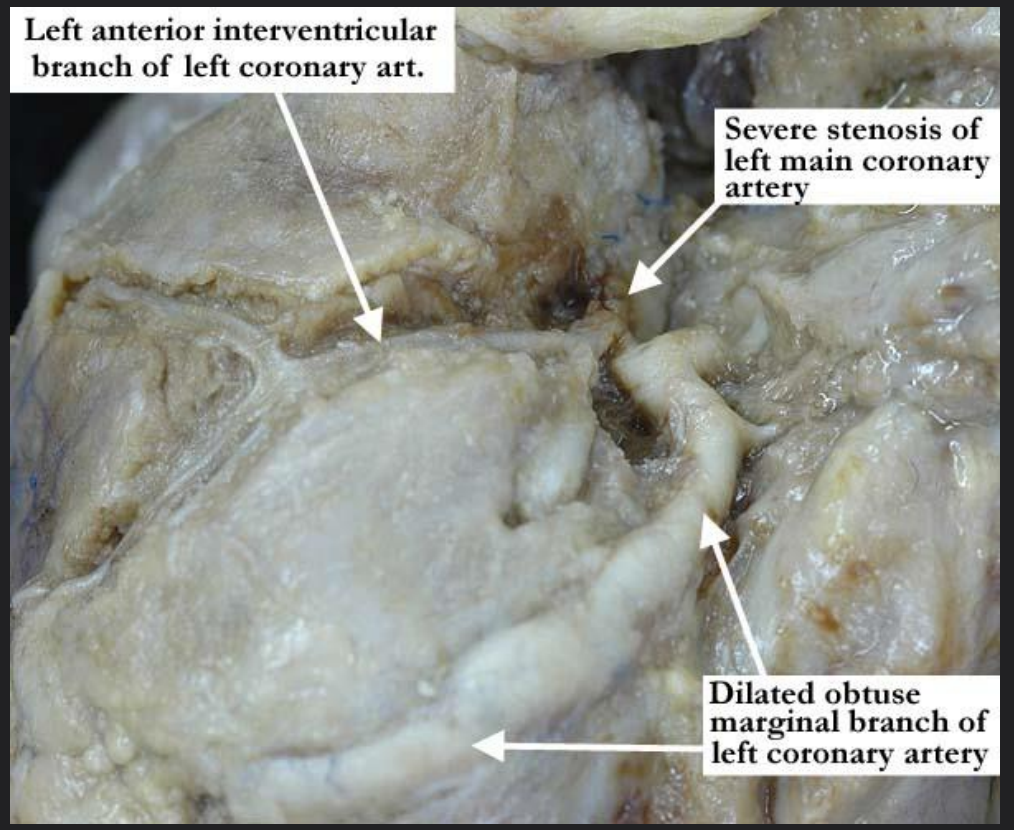




Coronary abnormalities: HLHS
Ventricle-coronary connections







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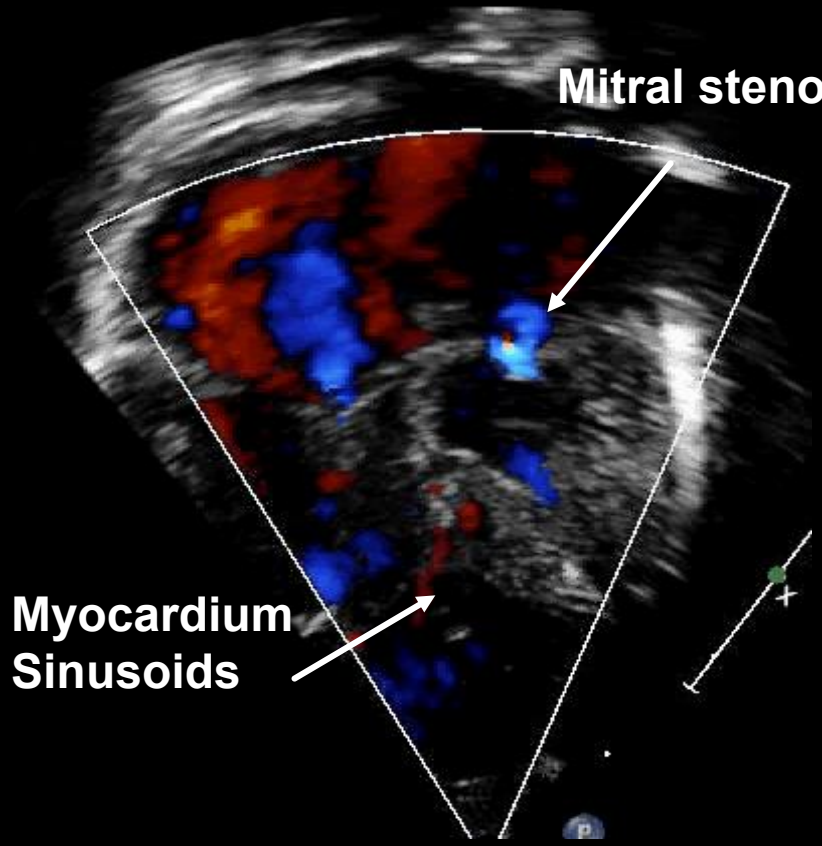
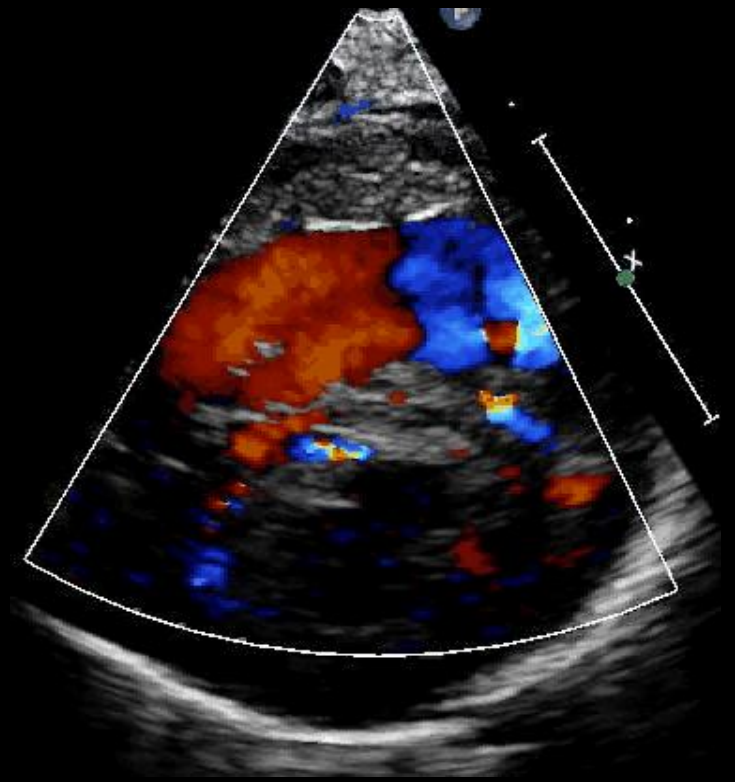
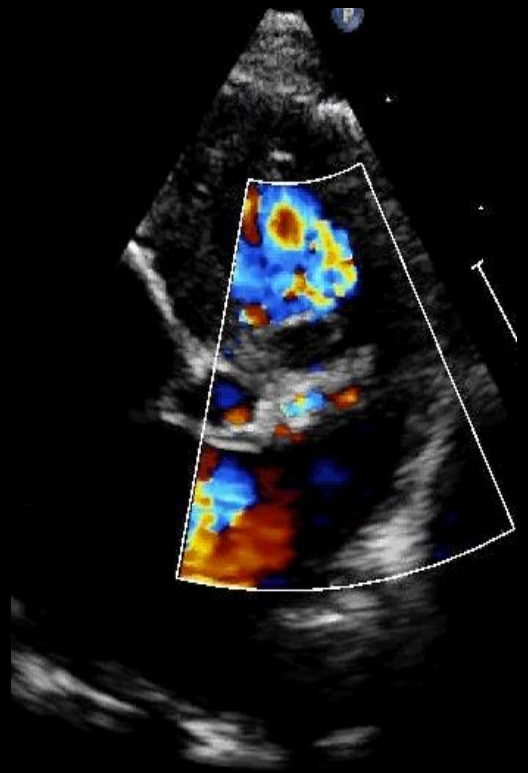
Coronary abnormalities: HLHS
Ventricle-coronary connections

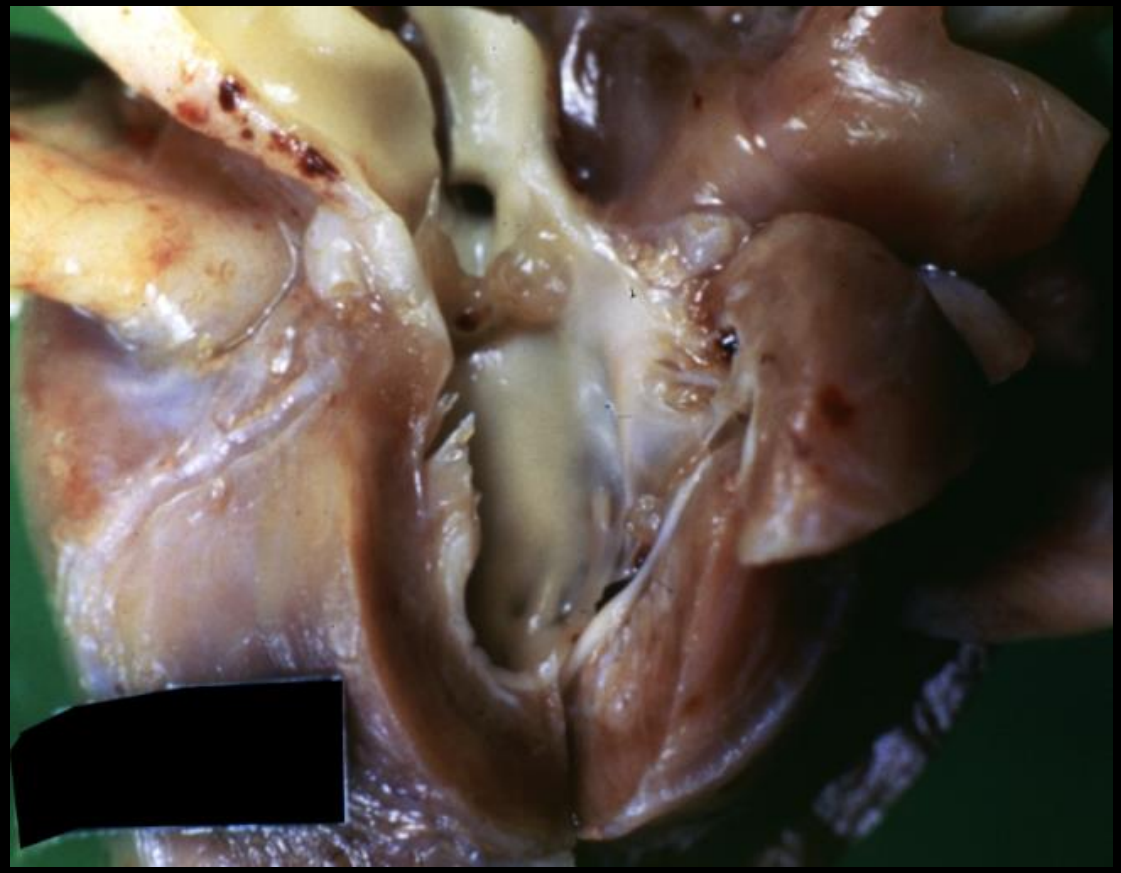
Aortic Atresia and Patent Mitral Valve



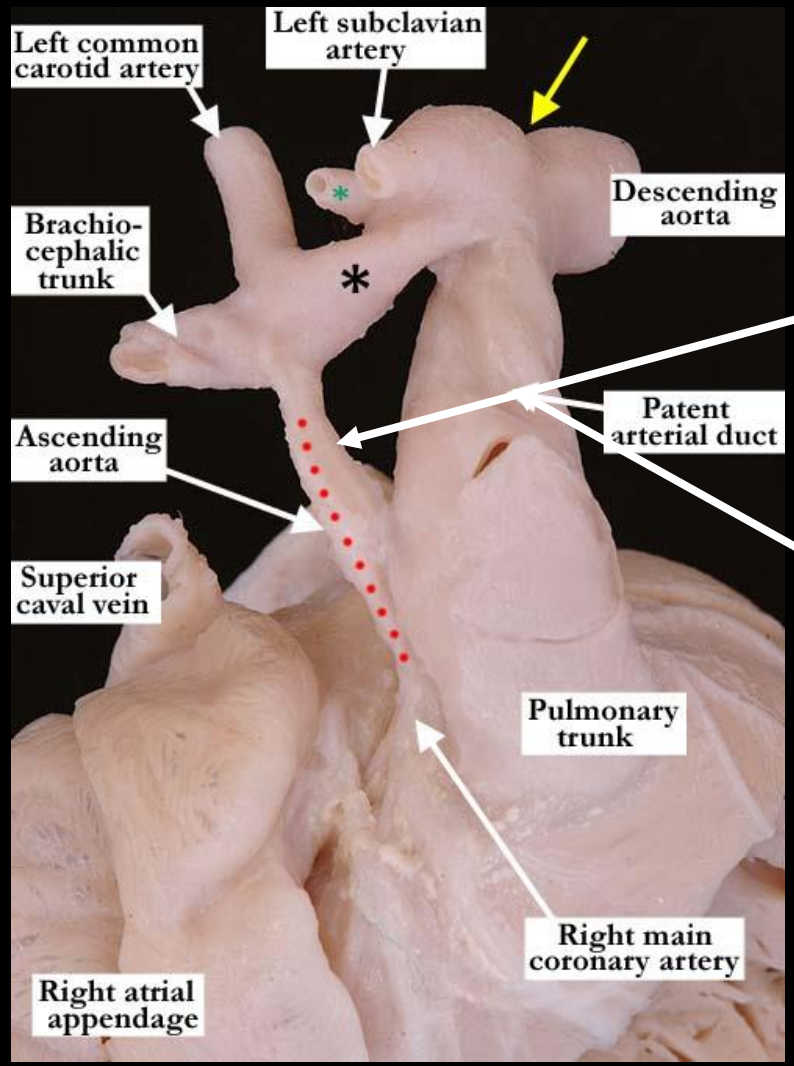


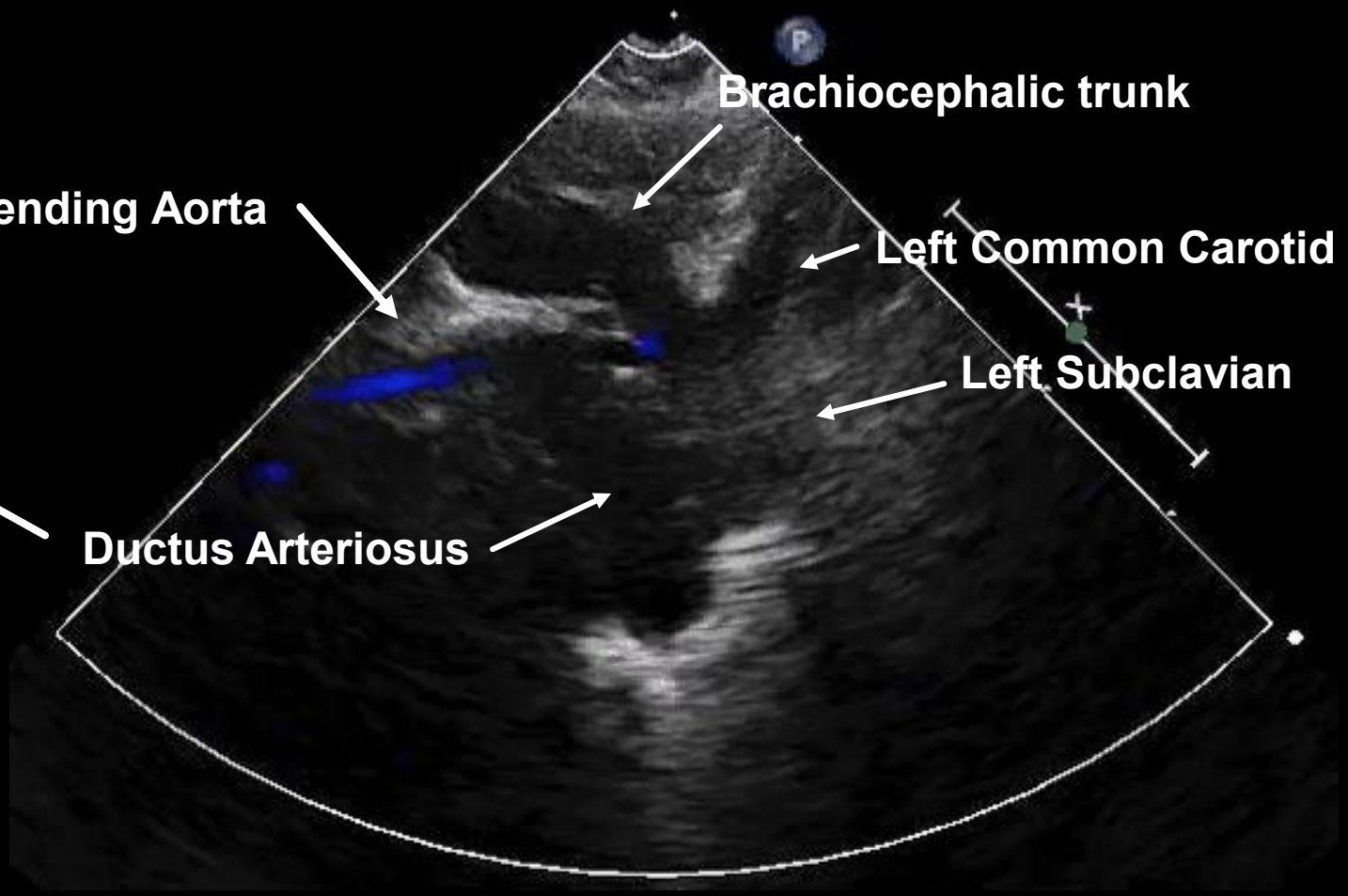


HLHS : Endocardial fibroelastosis & Coronary Artery Abnormalities



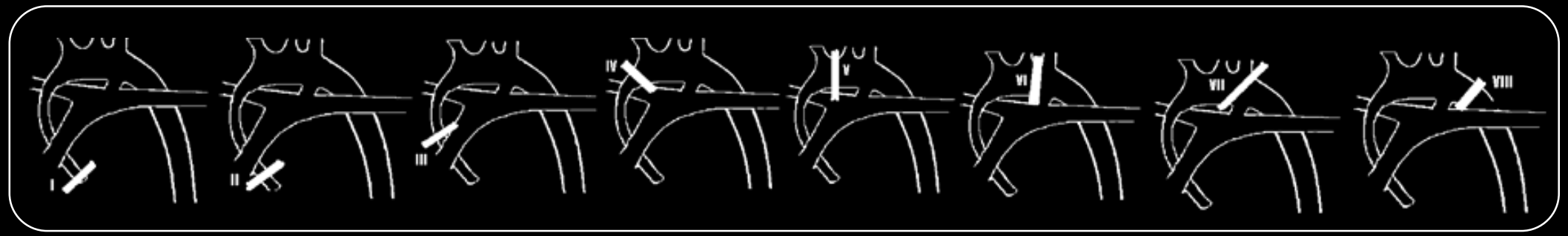
HLHS: Ascending aorta, Aortic Arch and Ductus Arteriosus

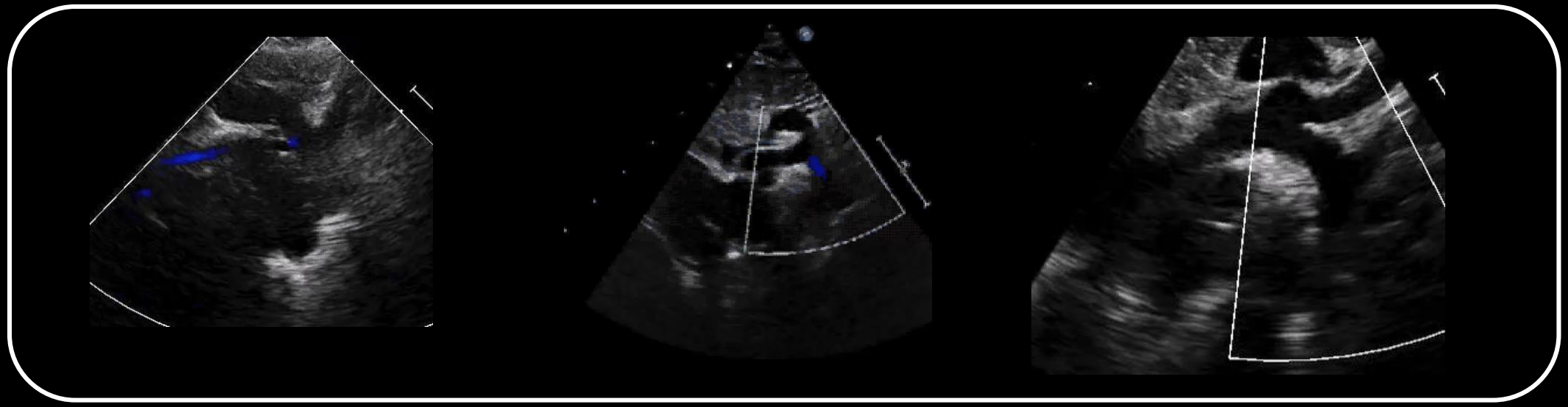




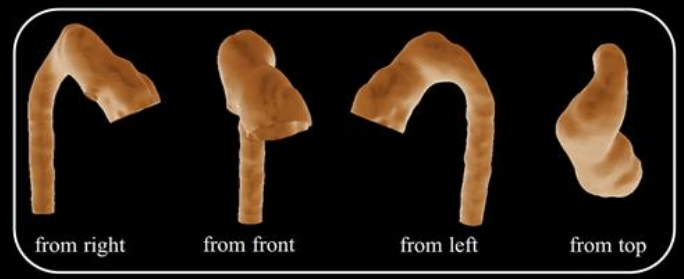
HLHS: Ascending aorta & Aortic Arch geometry &dimensions

Pre – op Aorta and Arch Dimensions & Shape

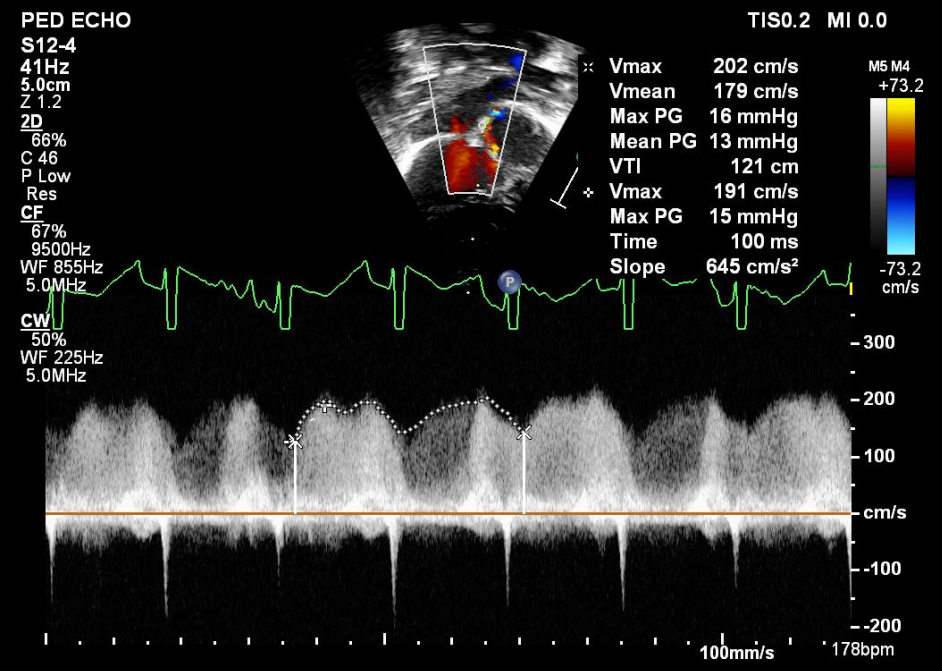




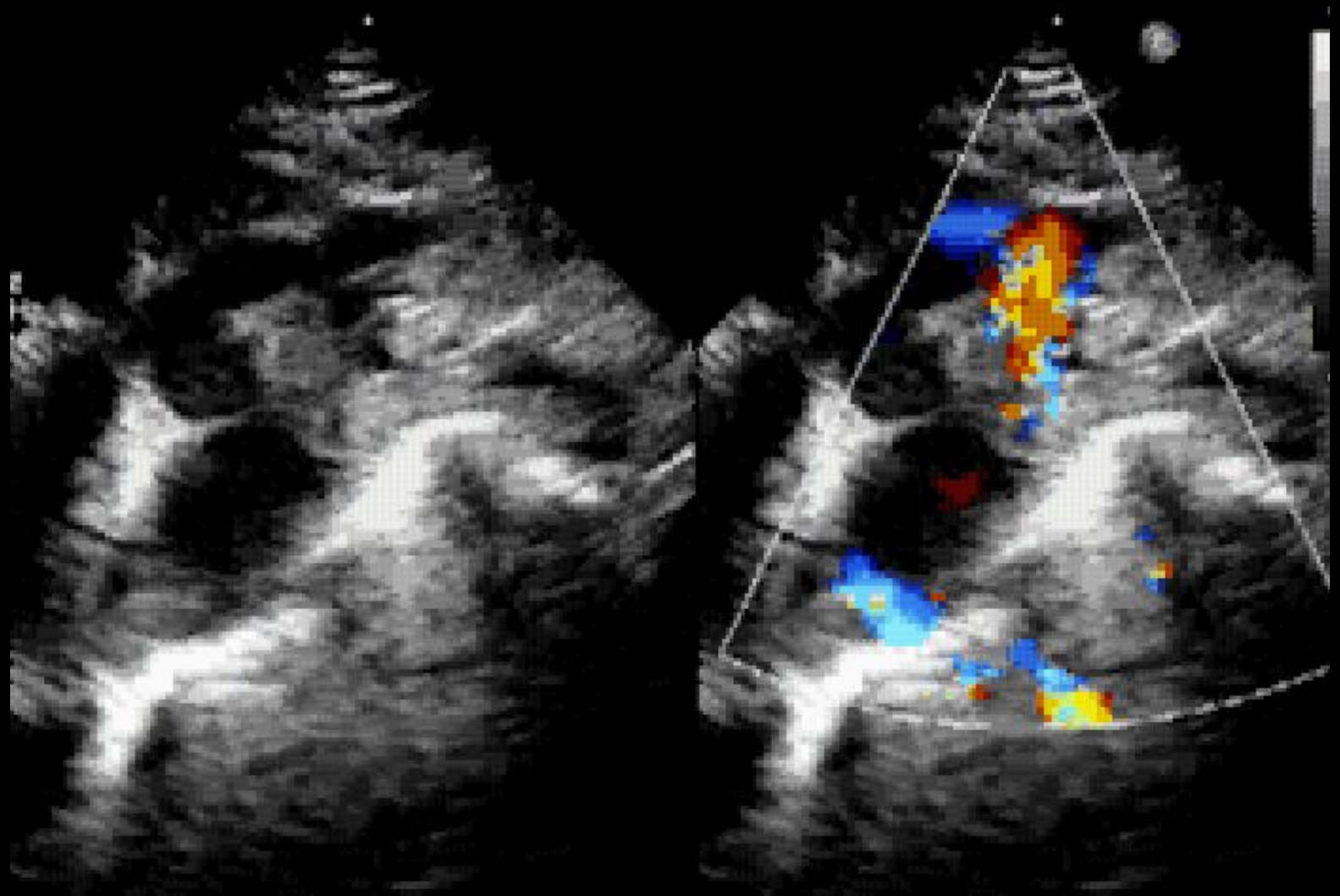
Pos – Norwood operation

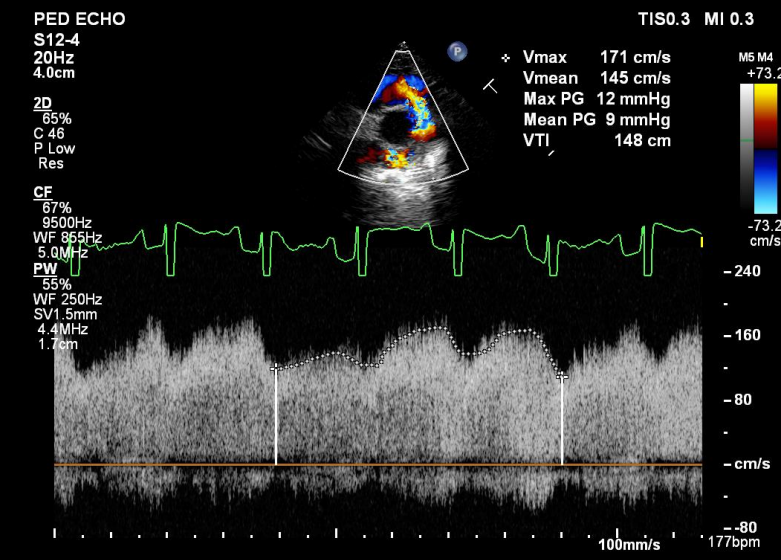


HLHS: restrictive atrial septum communication

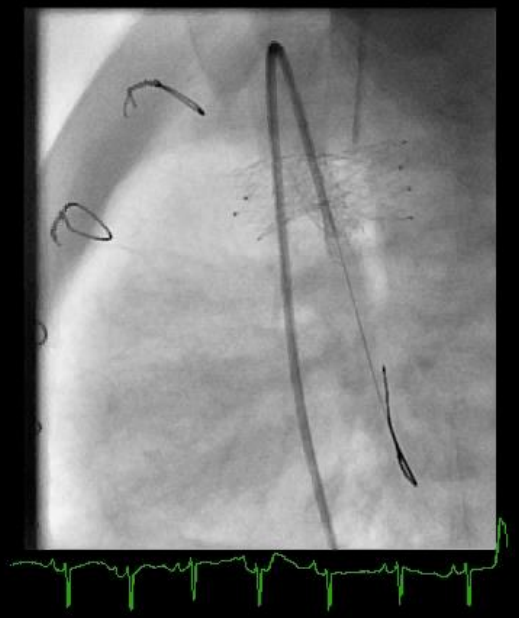


HLHS: restrictive atrial septum communication & Levoatriocardinal vein

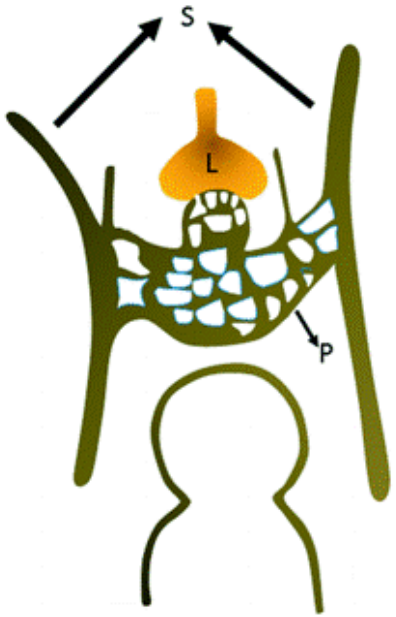




HLHS: restrictive atrial septum communication & Levoatriocardinal vein

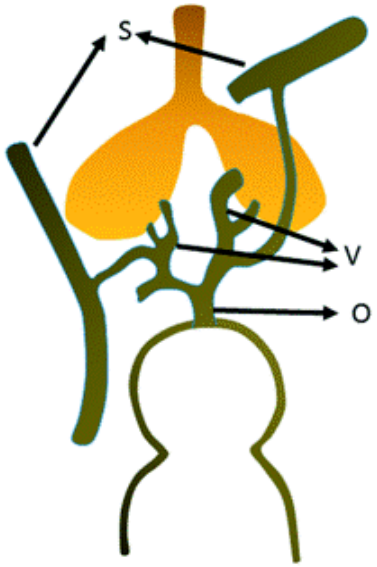


Embryology of the Levoatriocardinal Vein



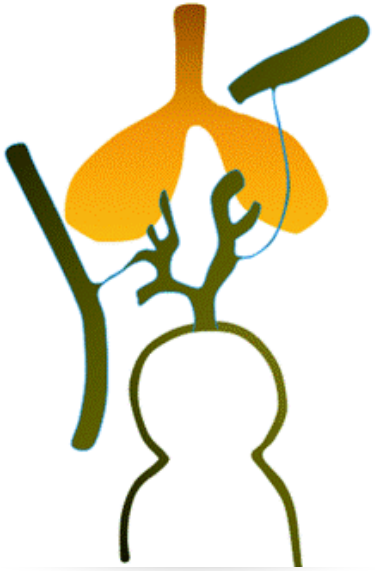
Phase 1

Capillary plexus surrounds the embryonic foregut And communicate with cardinal system



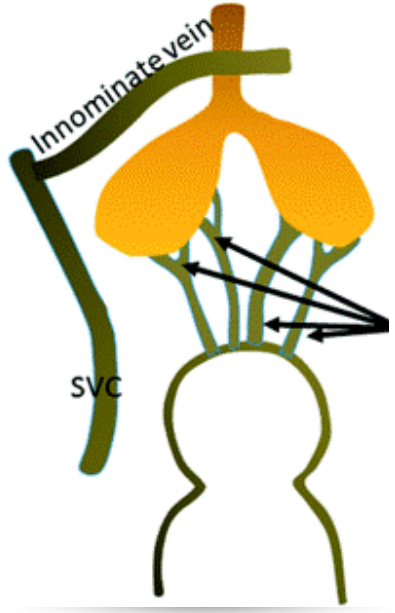
Phase 2

Plexus evolves into pulmonary veins and communicates with sinoatrial region



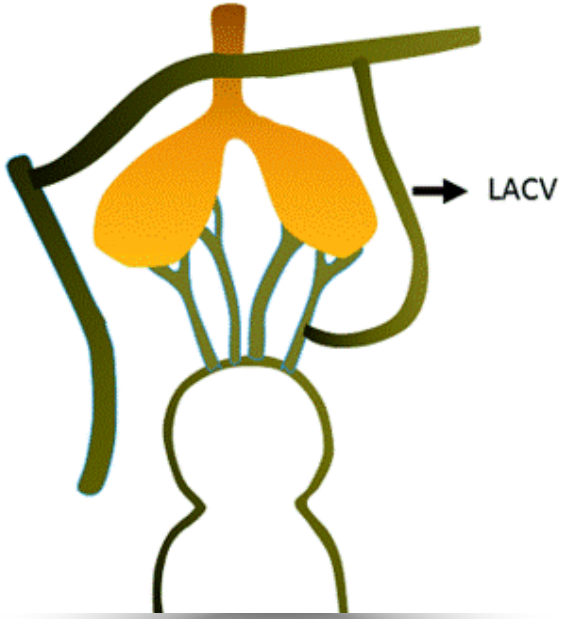
Phase 3

gradually loosing connection with cardinal system



Phase 4

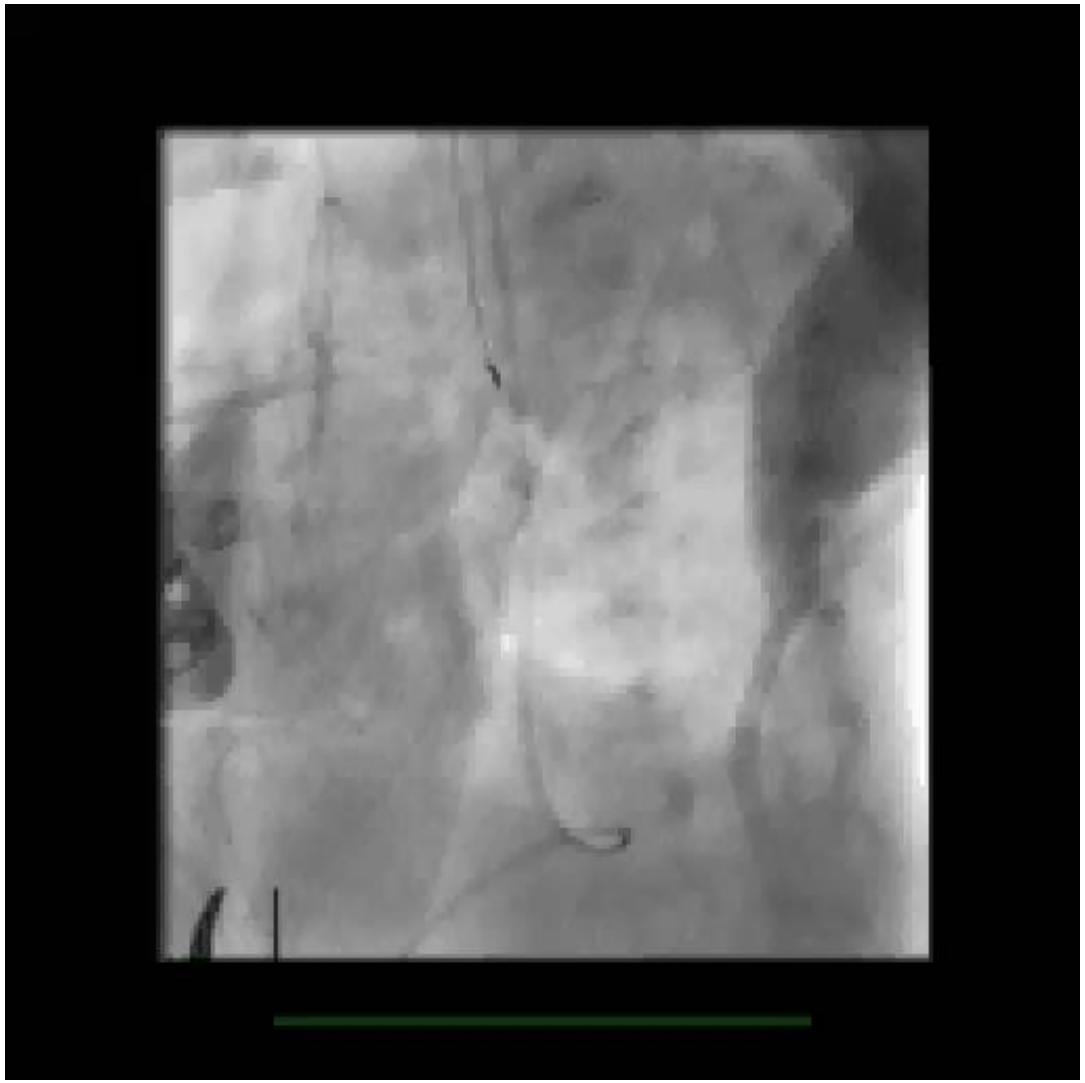
Full developed pulmonary veins connected with left atrium

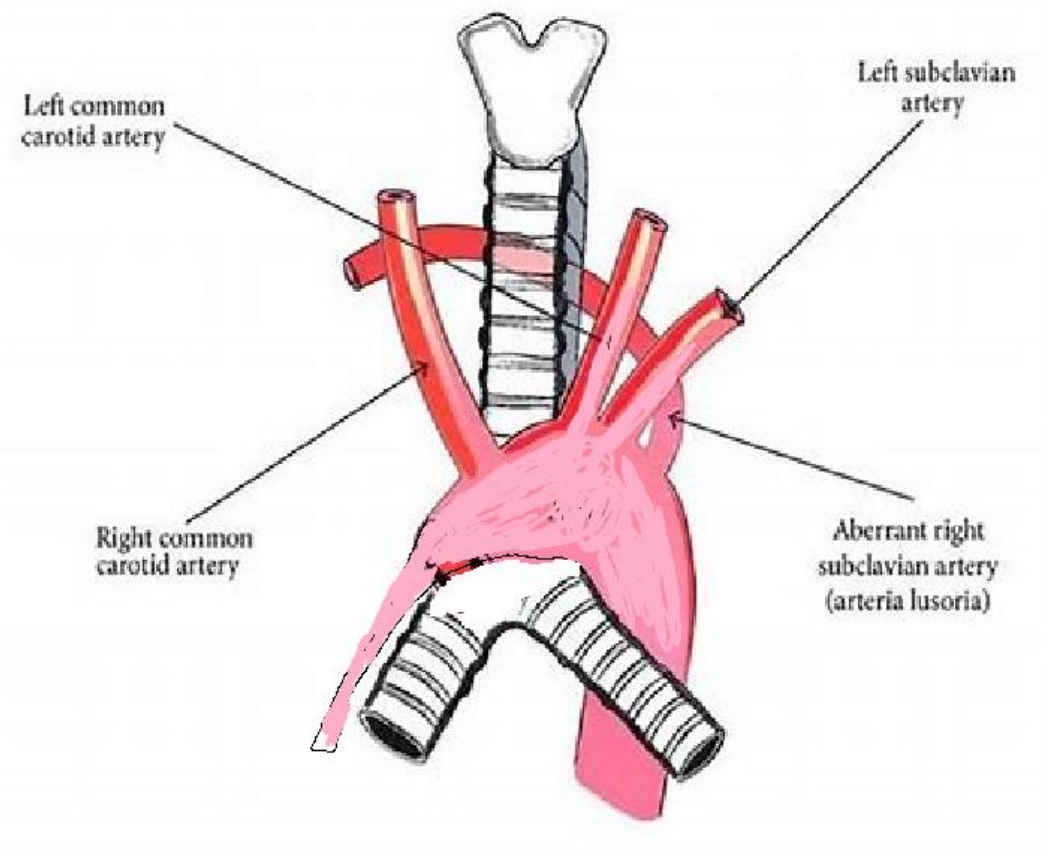


Phase 5

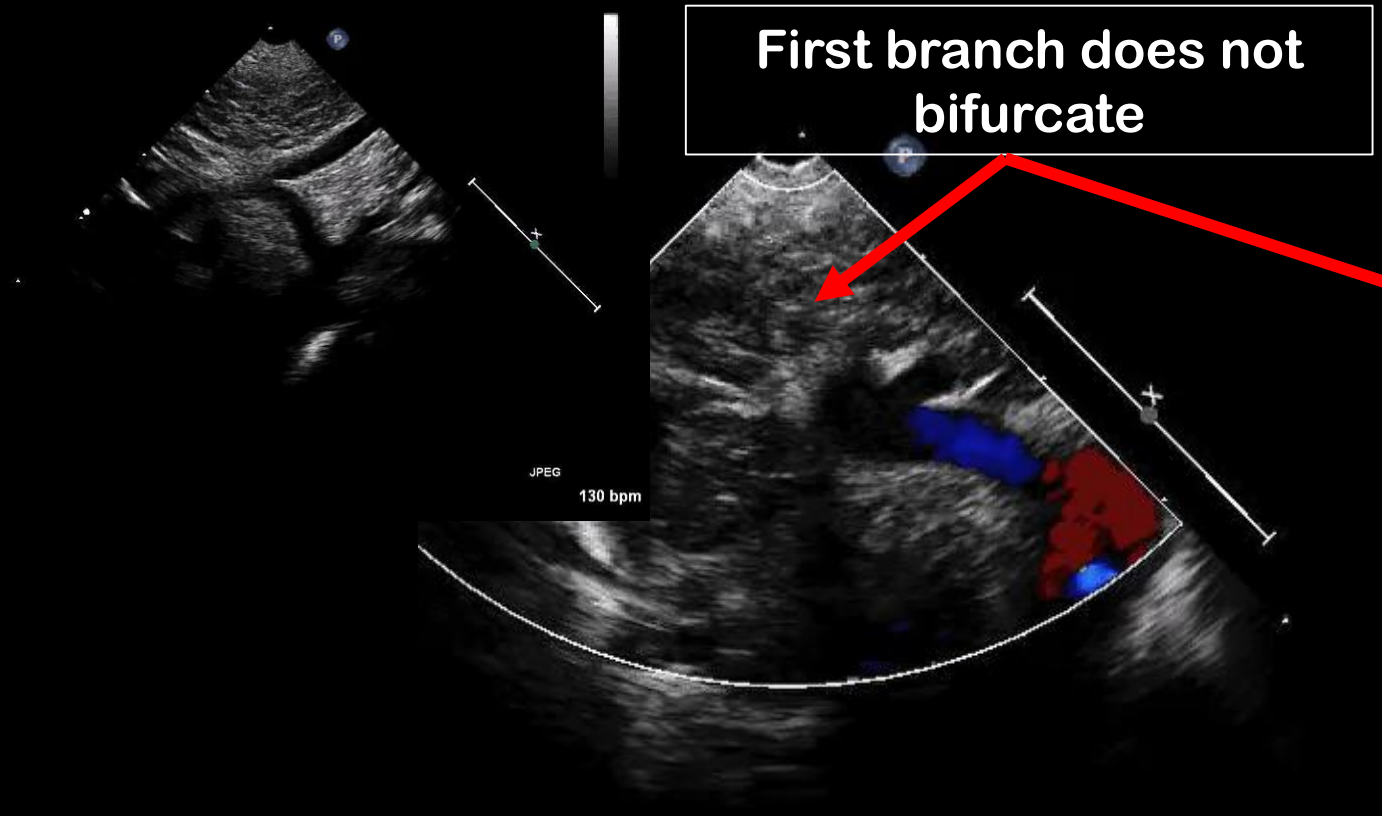
Persistent of primitive connection with the cardinal system: presence of obstructive left lesions

HLHS : Aortic Arch & Aberrant Right Subclavian Artery

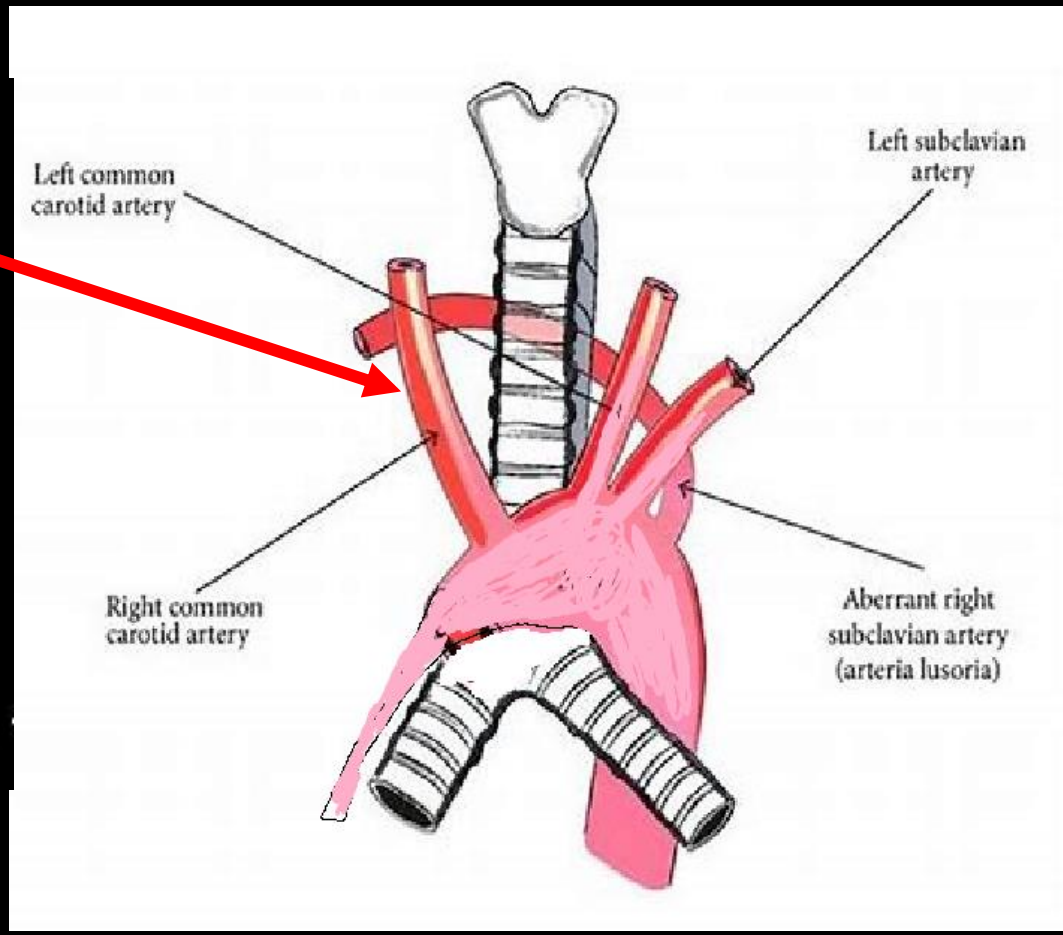




HLHS : Aortic Arch & Aberrant Right Subclavian Artery



First branch does not bifurcate



Take Home Message : Hypoplastic Left Heart Syndrome

Point 1

Spectrum of cardiac malformation : significant underdevelopment of the left side structures. Severe form : Mitral and aortic atresia

Point 2

Genetic & Embryology % Flow: Myocardial maturation and Left ventricle development

Point 3

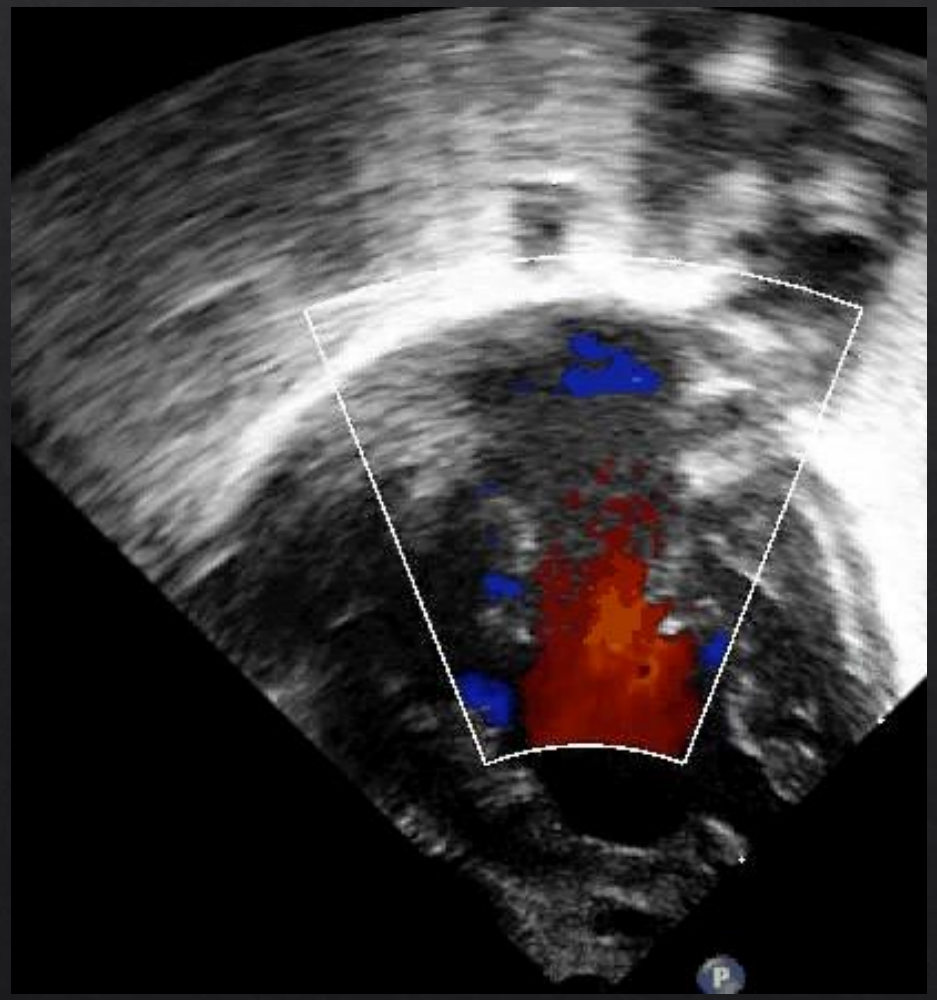
Atrial Septum: leftward deviation ;
Tricuspid valve : range of anomalies ; mainly subvalvar apparatus
Right Ventricle: abnormal septomarginal trabecula

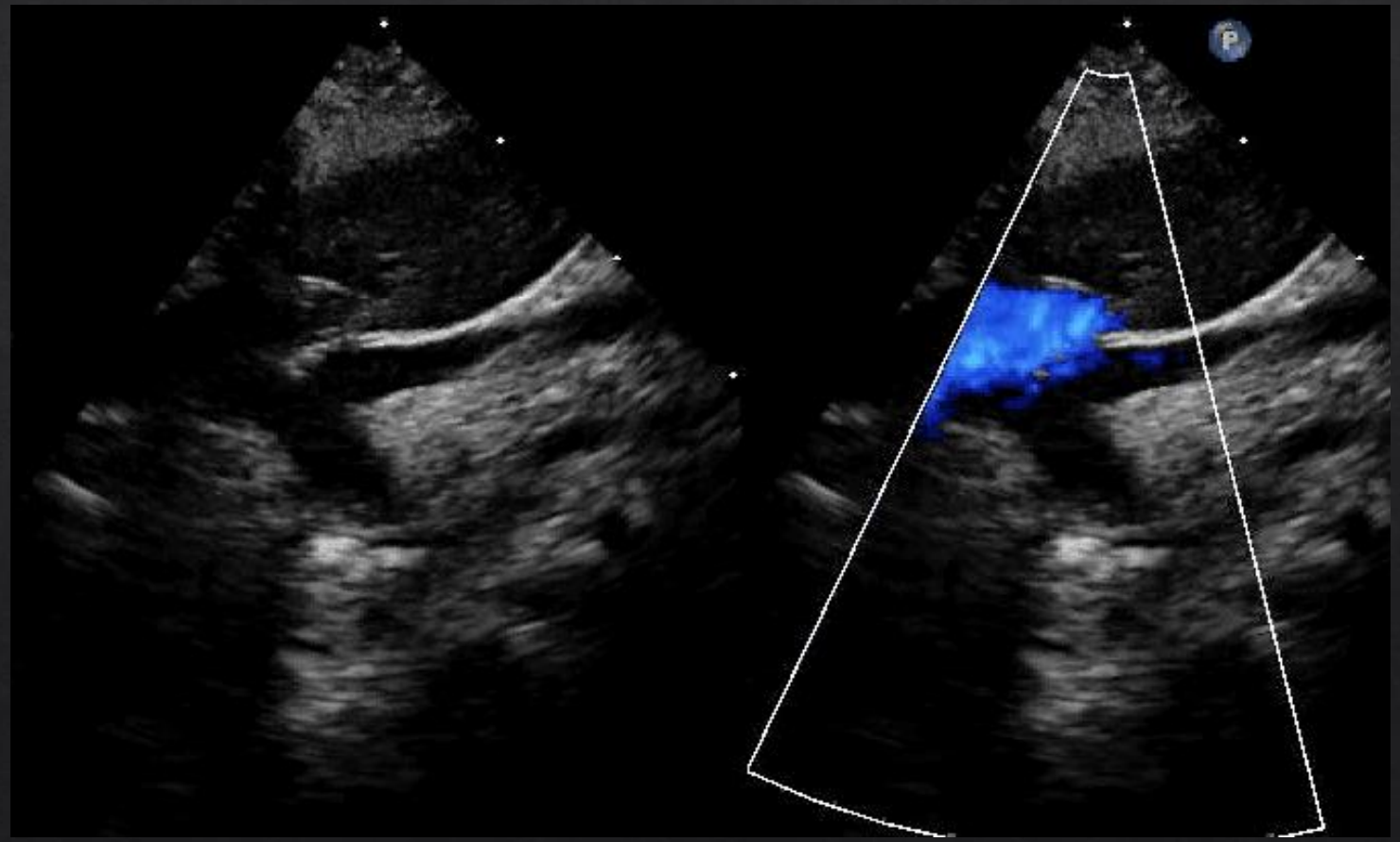
Point 4

Coronary Artery Sinusoids: > Patent MV
Hypoplastic ascending aorta & Arch geometry: log term outcome
Recognize Levoatriocardinal vein
Aortic Arch: Aberrant Right Subclavian Artery

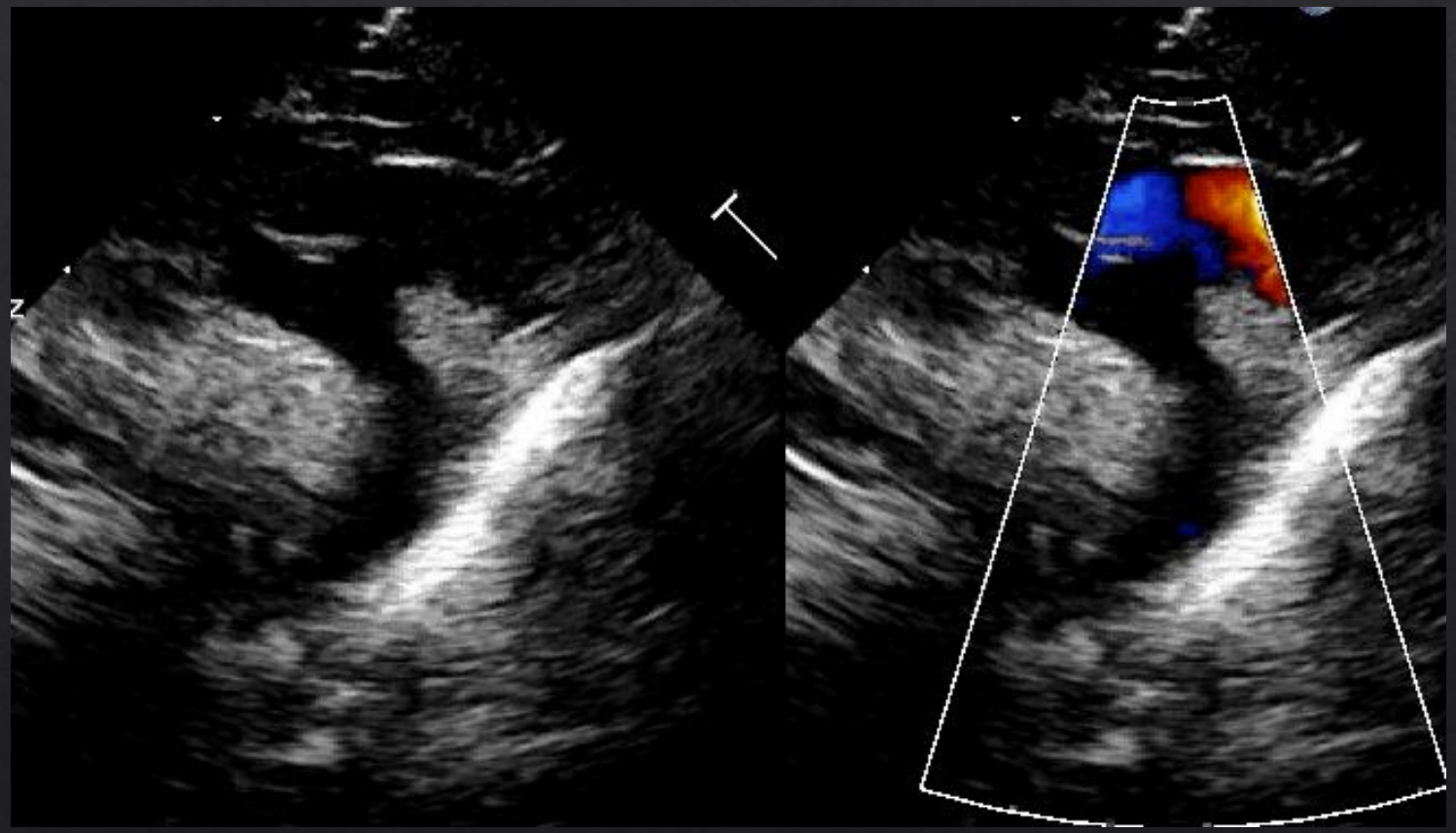
vitor.Guerra@Sickkids.ca

Hypoplasia of the left Heart: restrictive atrial septum communication & levocardinal vein



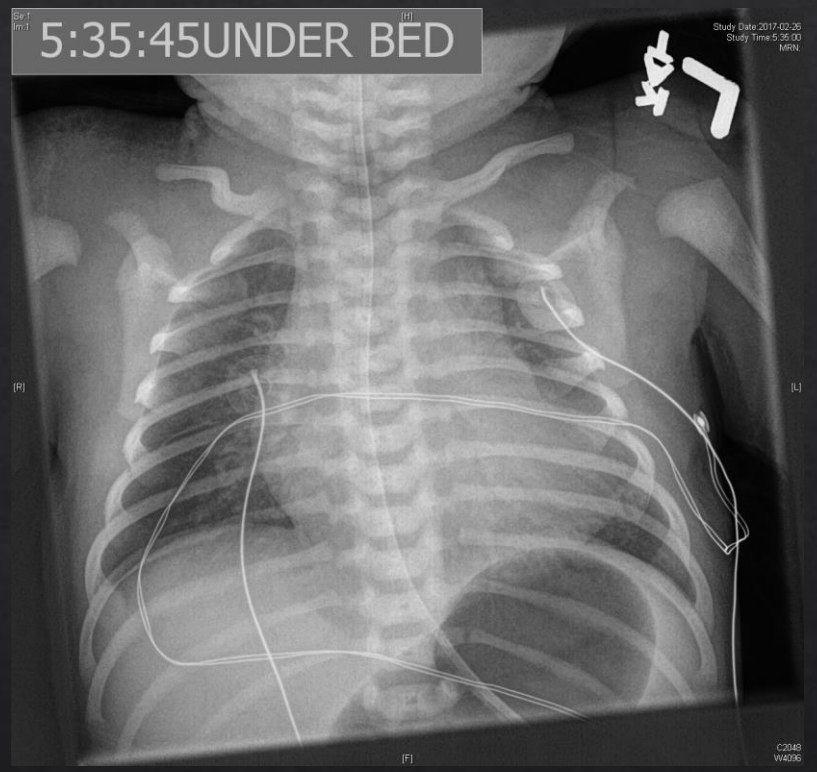


Hypoplasia of the left Heart: restrictive atrial septum communication & levocardinal vein



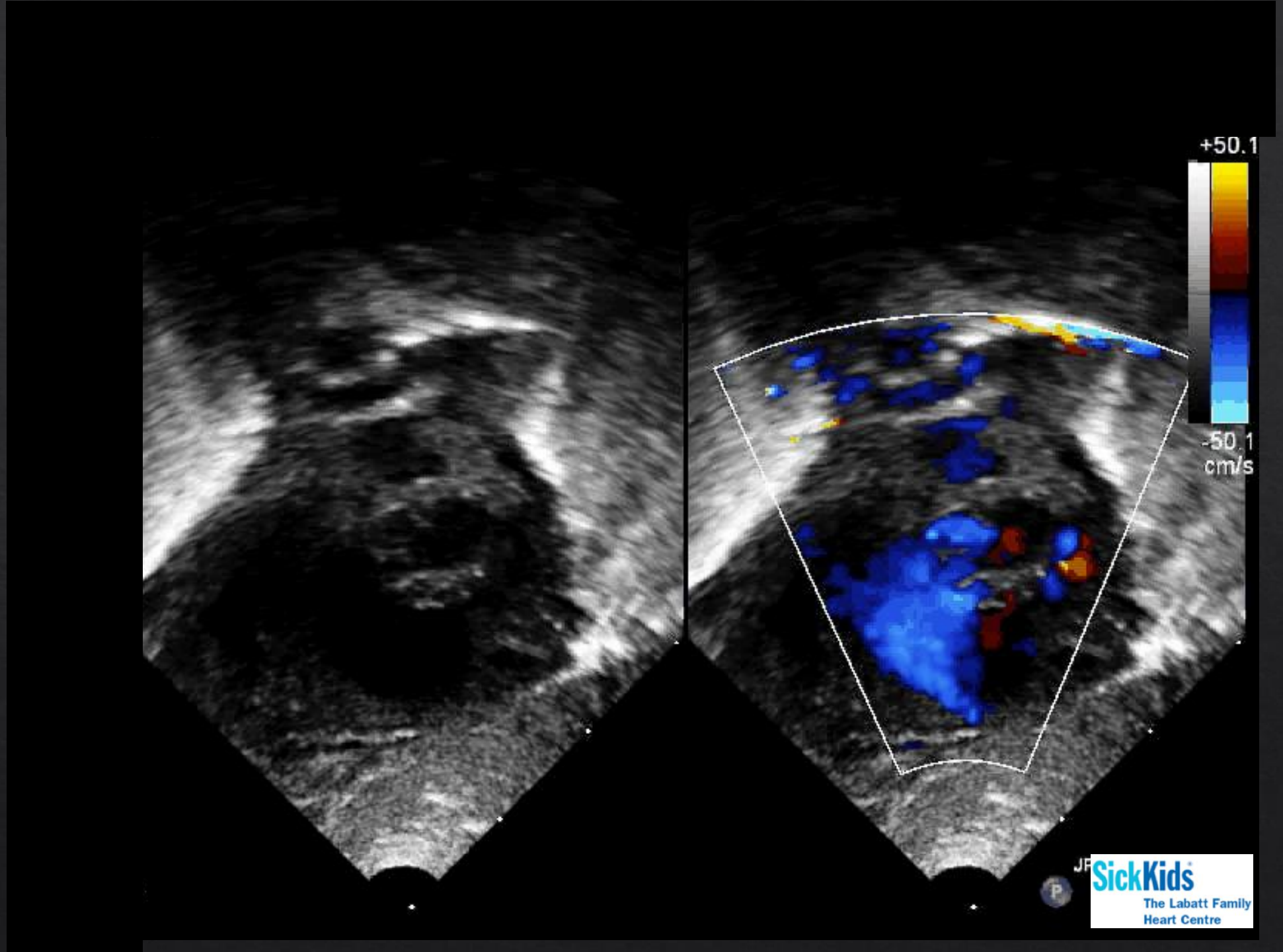
Case 1

- Newborn, prenatal diagnosis of "HLHS", very restrictive interarterial septum (intact septum?), severe pulmonary veins obstruction, ? levo atrial vein.
- Fetal Counseling : active management ; family aware about high risk/ mortality
- Parents denied : amniocentesis / Fetal MRI
- Delivery by C section, 39 weeks, ECMO stand by, 2.7 kg, Sats 30-60 %, chest x ray



SickKids
The Labatt Family Heart Centre


JPEG
SickKids
The Labatt Family
Heart Centre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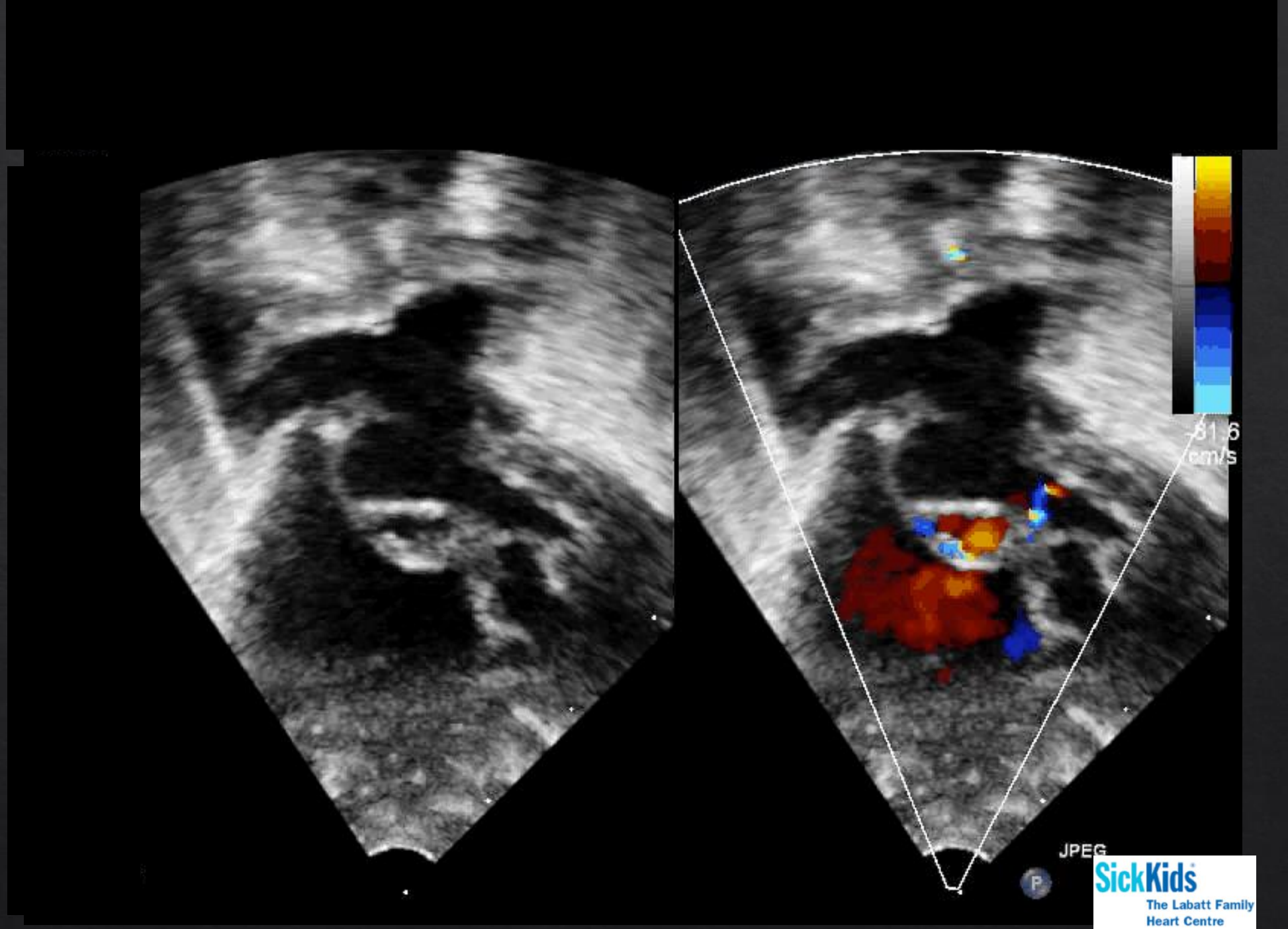
-50.1
cm/s
SickKids
The Labatt Family
Heart Centre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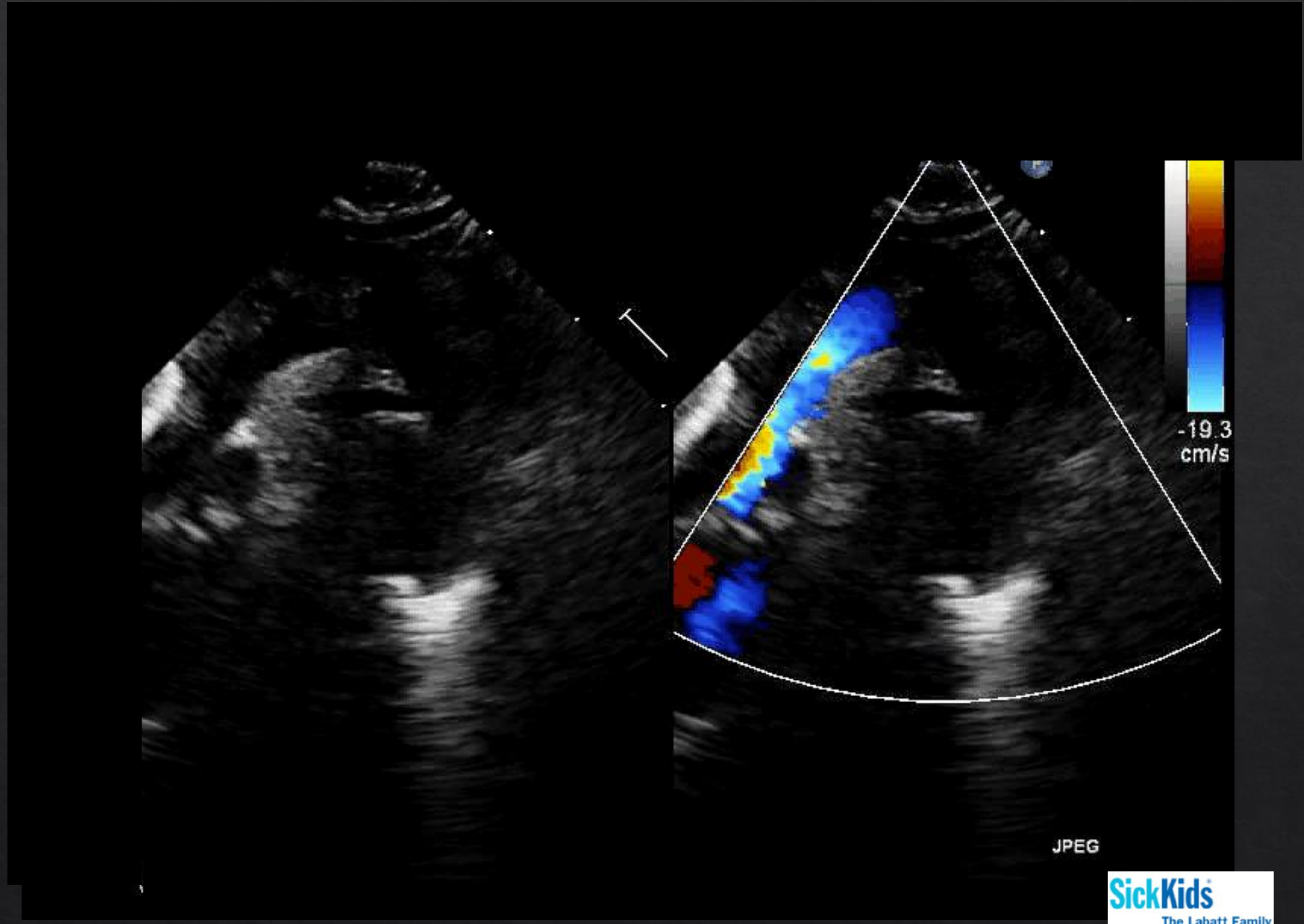
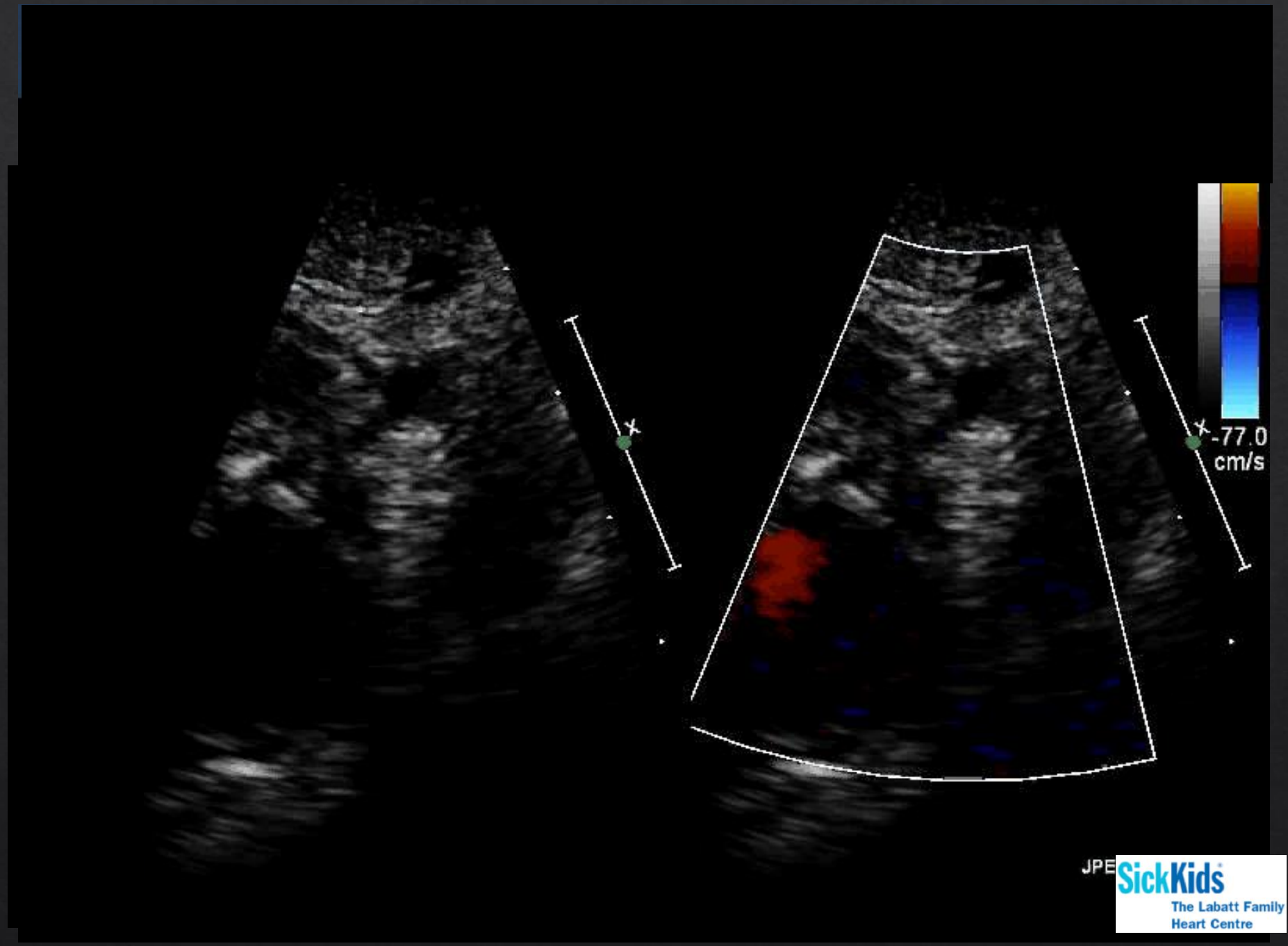
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre

SickKids
The Labatt Family Heart Centre


-67.0
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre



SickKids
The Labatt Family
Heart Centre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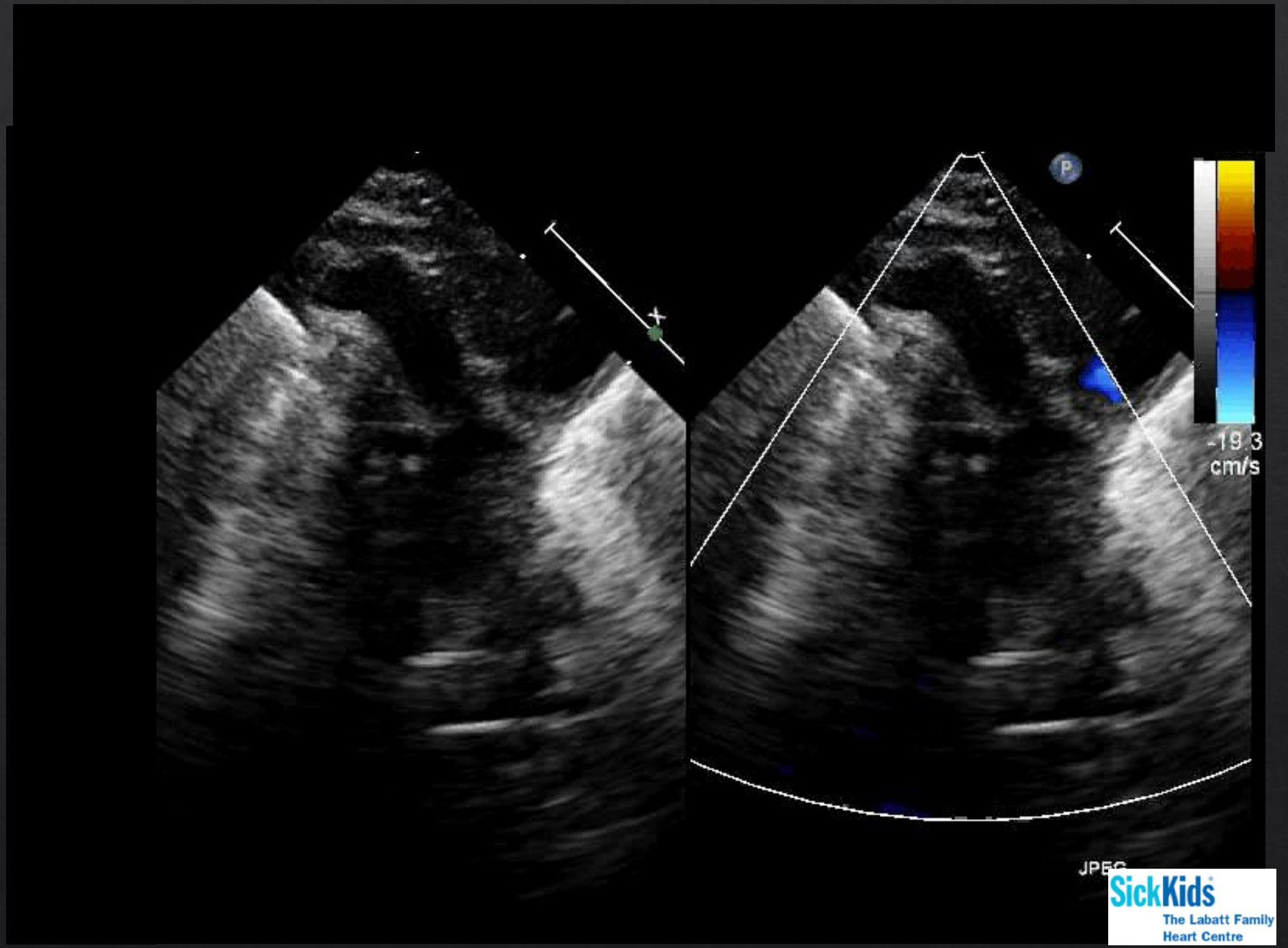
SickKids
The Labatt Family
Heart Centre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cm/s
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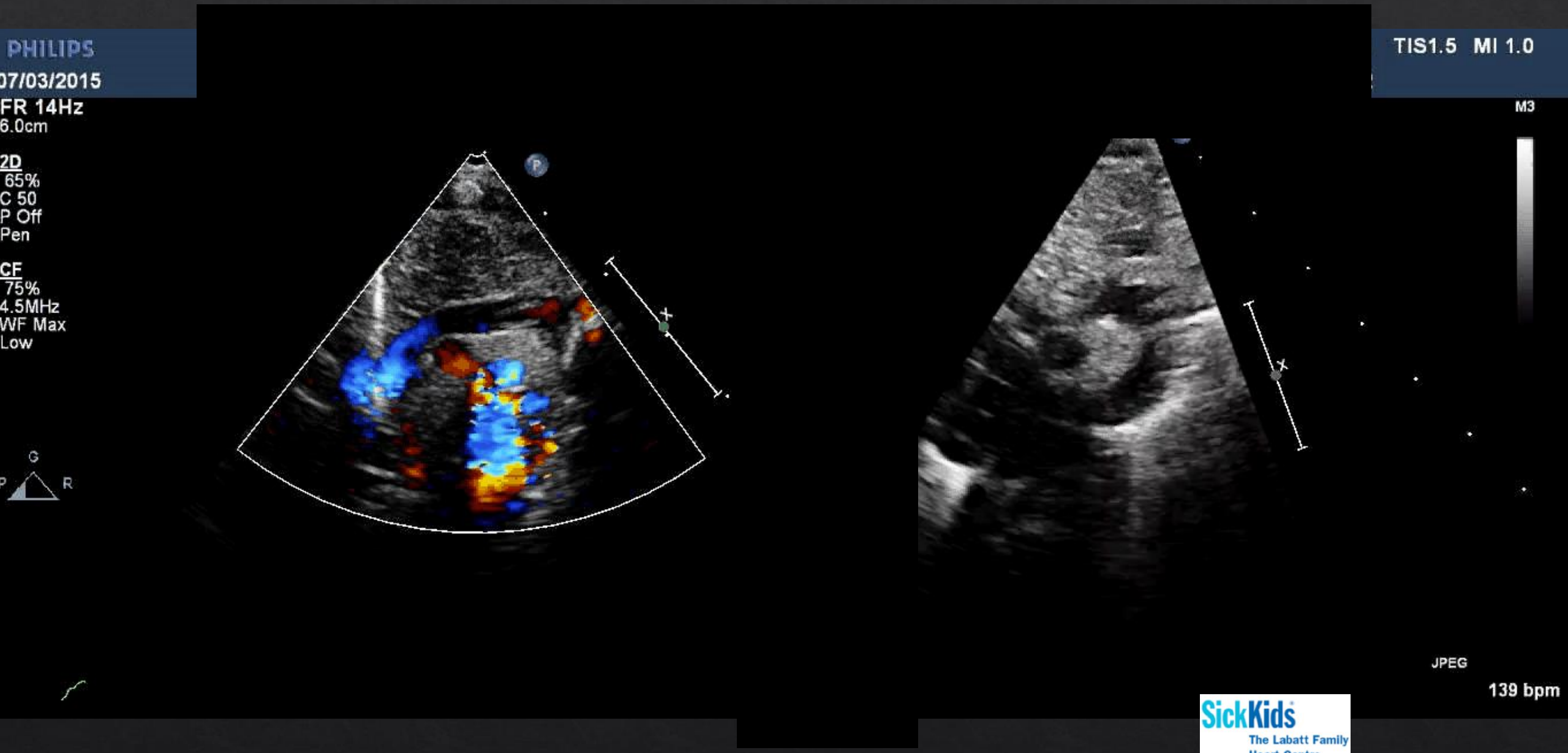
SickKids
The Labatt Family
Heart Centre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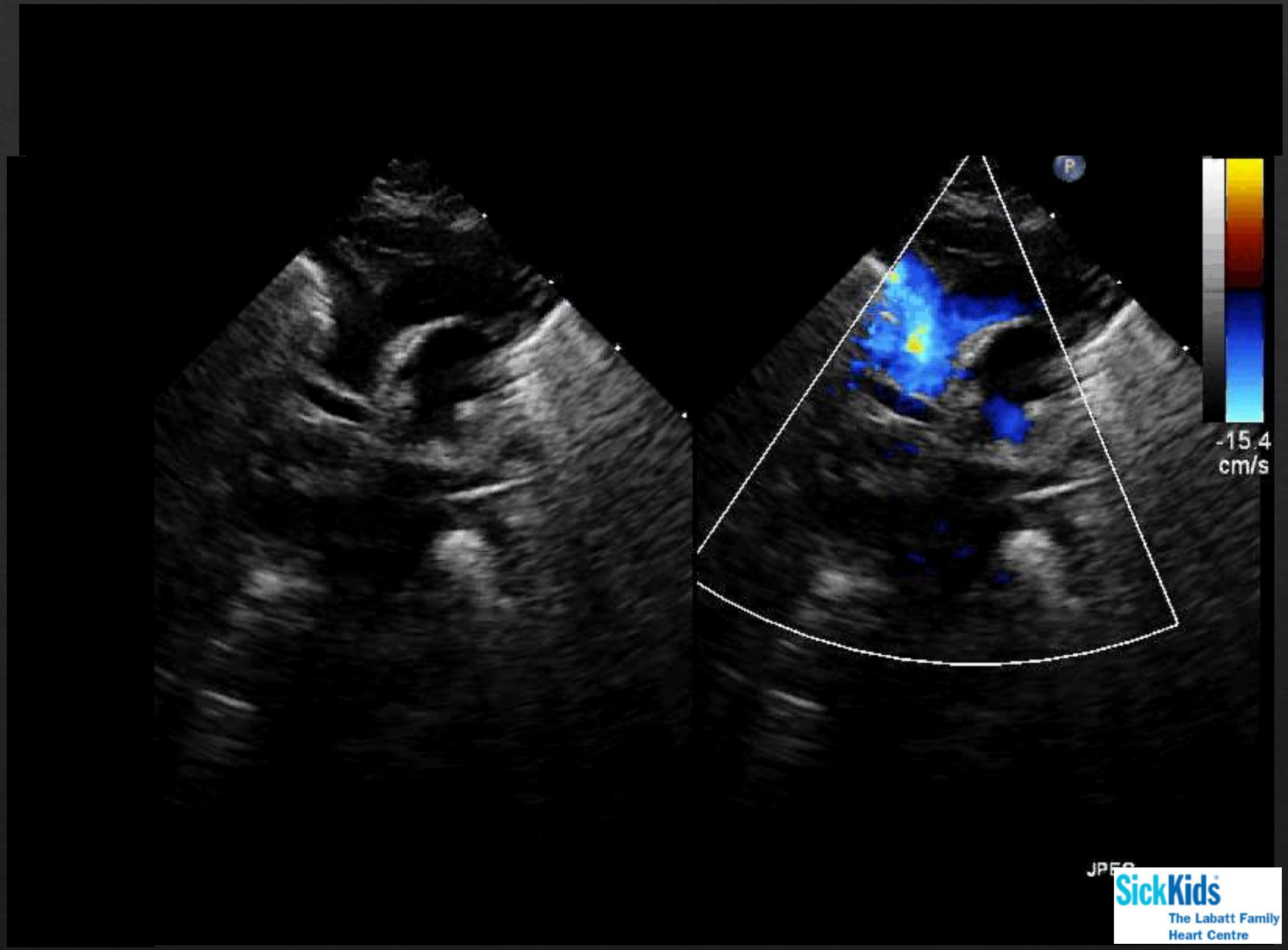
07/03/2015
FR 14Hz
6.0cm
2D
65%
C 50
P Off
Pen
CF
75%
4.5MHz
WF Max
Low
G
P
R
TIS1.5 MI 1.0
M3
JPEG
139 bpm
SickKids
The Labatt Family
Heart Centre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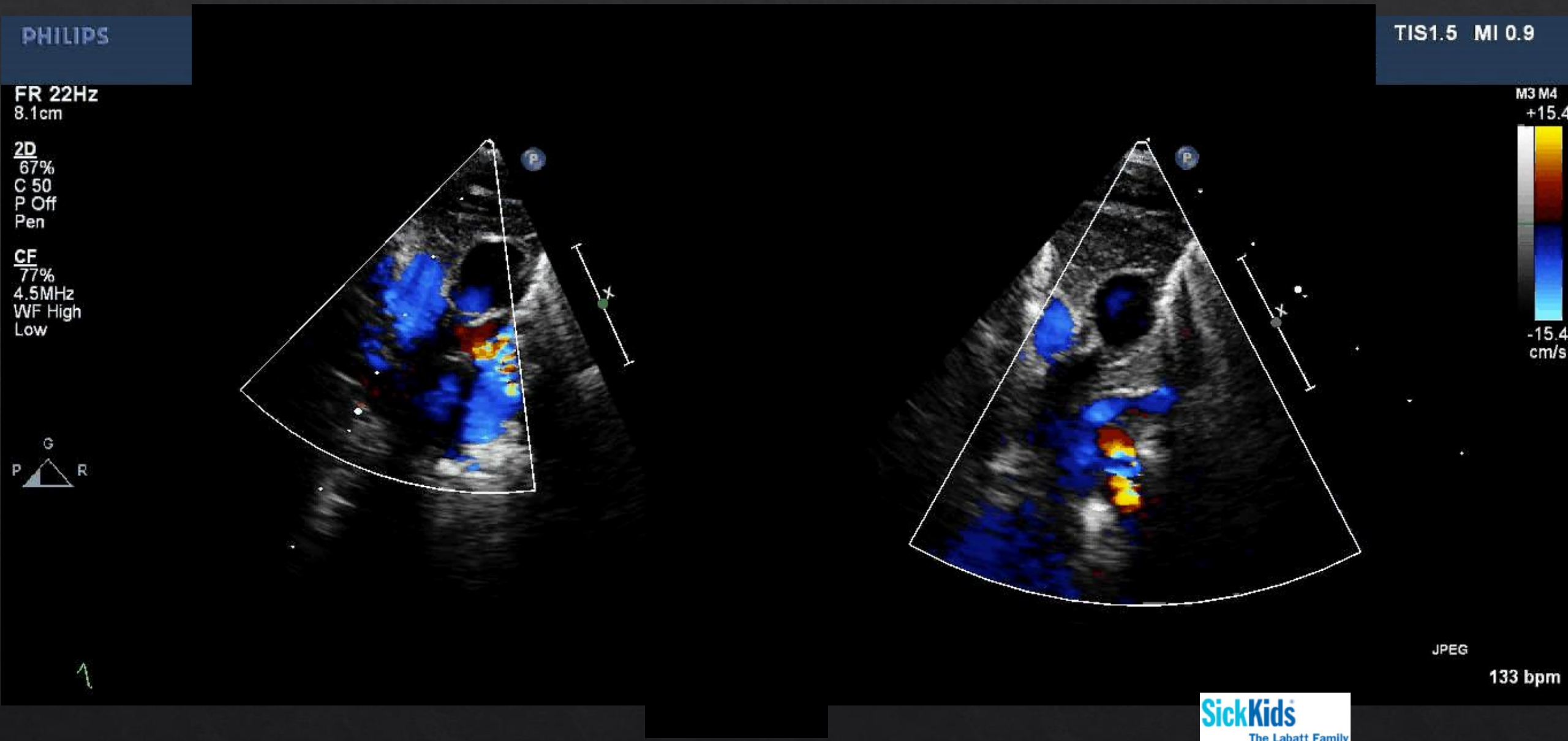
cm/s
SickKids
The Labatt Family
Heart Centre


PHILIPS
TIS1.5 MI 0.9
FR 22Hz
8.1cm
2D
67%
C 50
P Off
Pen
CF
77%
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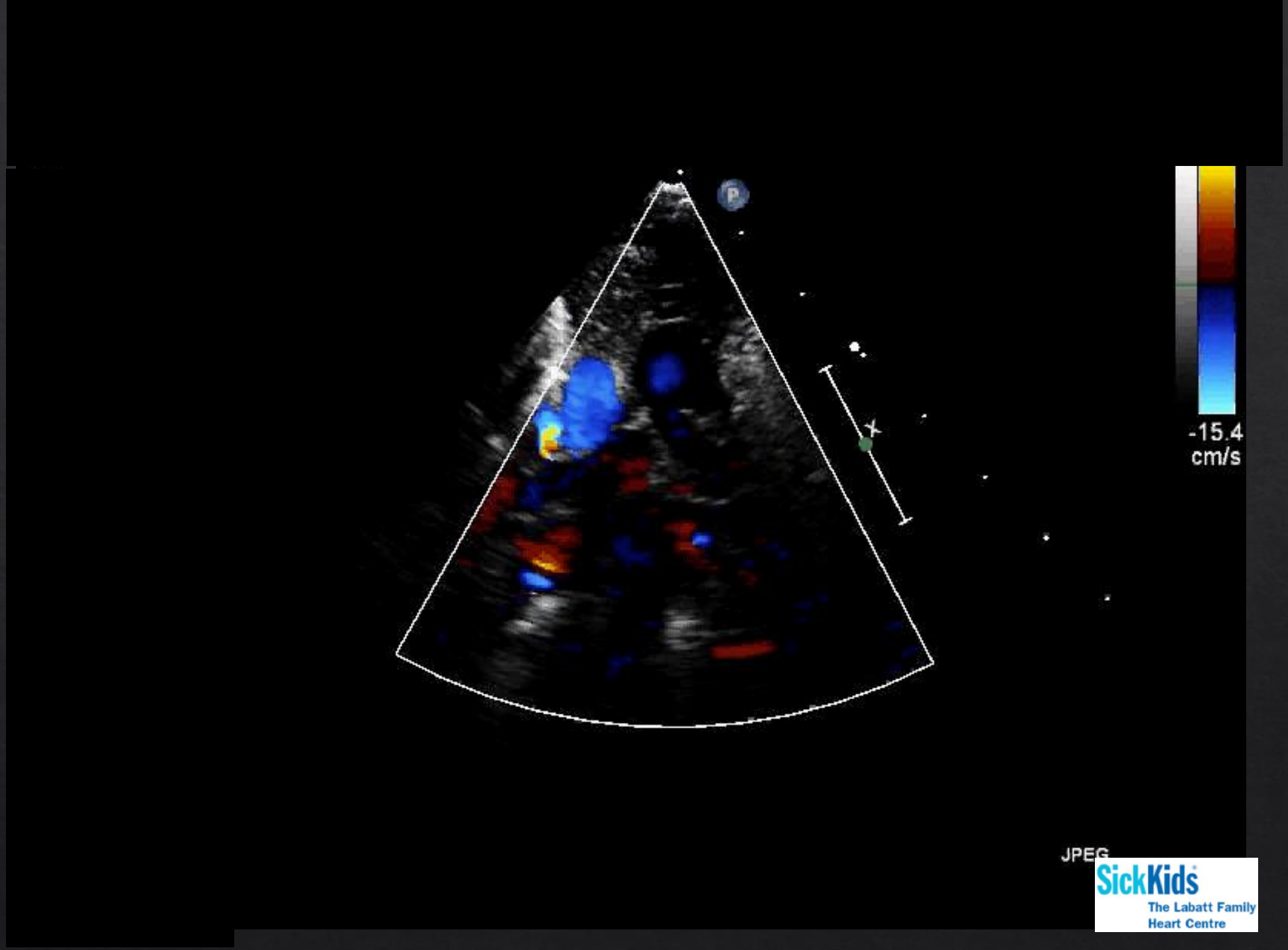
-15.4
cm/s
G
P
R
JPEG
133 bpm


-15.4
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre

Outcome

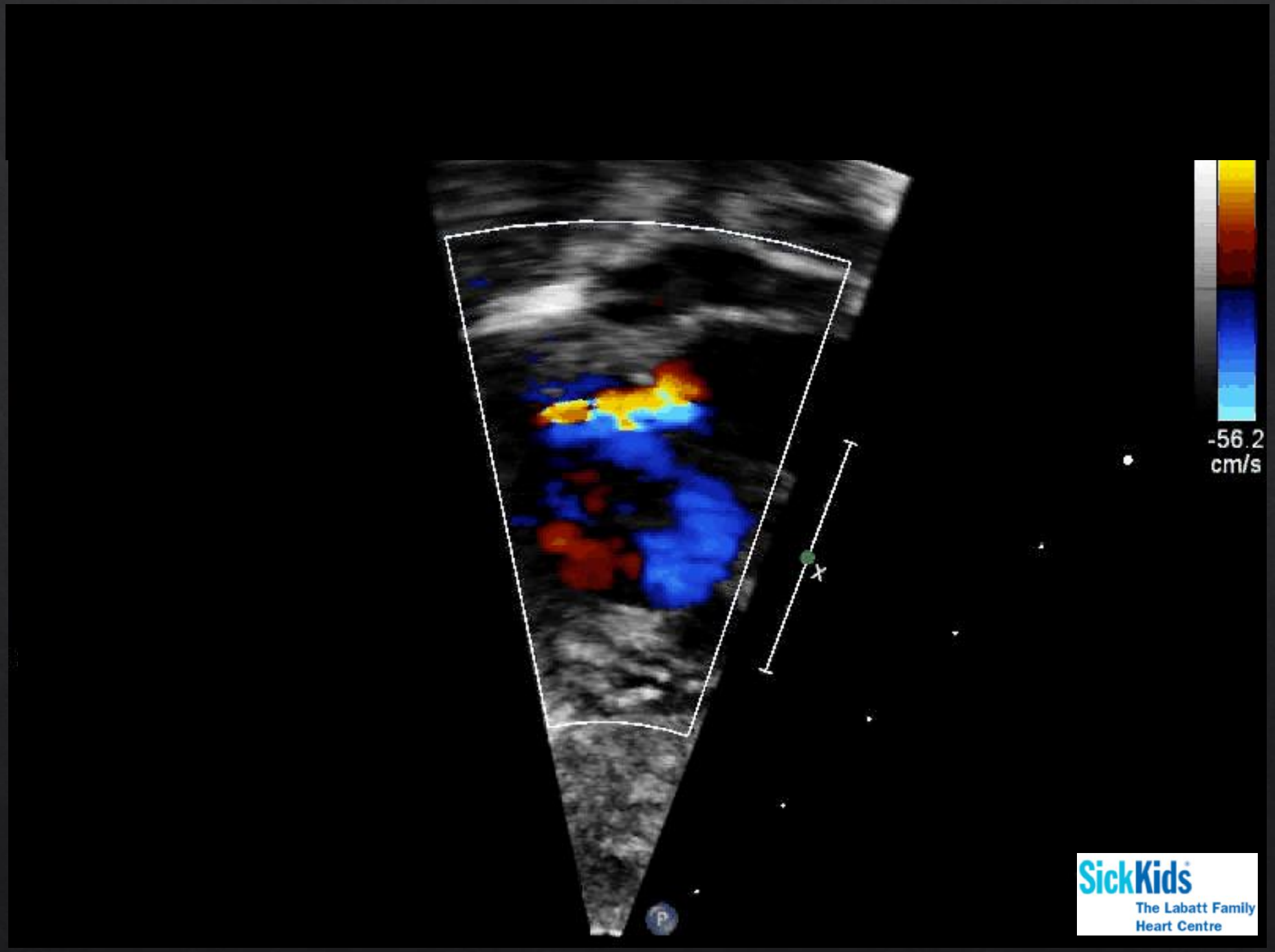
- Norwood / Shunt + Atrioseptectomy (Thick Cortriatriatum and partial unroofed coronary sinus)
- Post operative course: Low Sats / Pulmonary veins hypoplasia
- ECMO….

Case 2

- Newborn, pre natal diagnosis of HLHS, possible multiple small muscular VSDs.
- Family counseling : active management.
- Delivery spontaneously : 39 weeks, 3.1 kg

SickKids
The Labatt Family Heart Centre


-56.2
cm/s
SickKids
The Labatt Family
Heart Centre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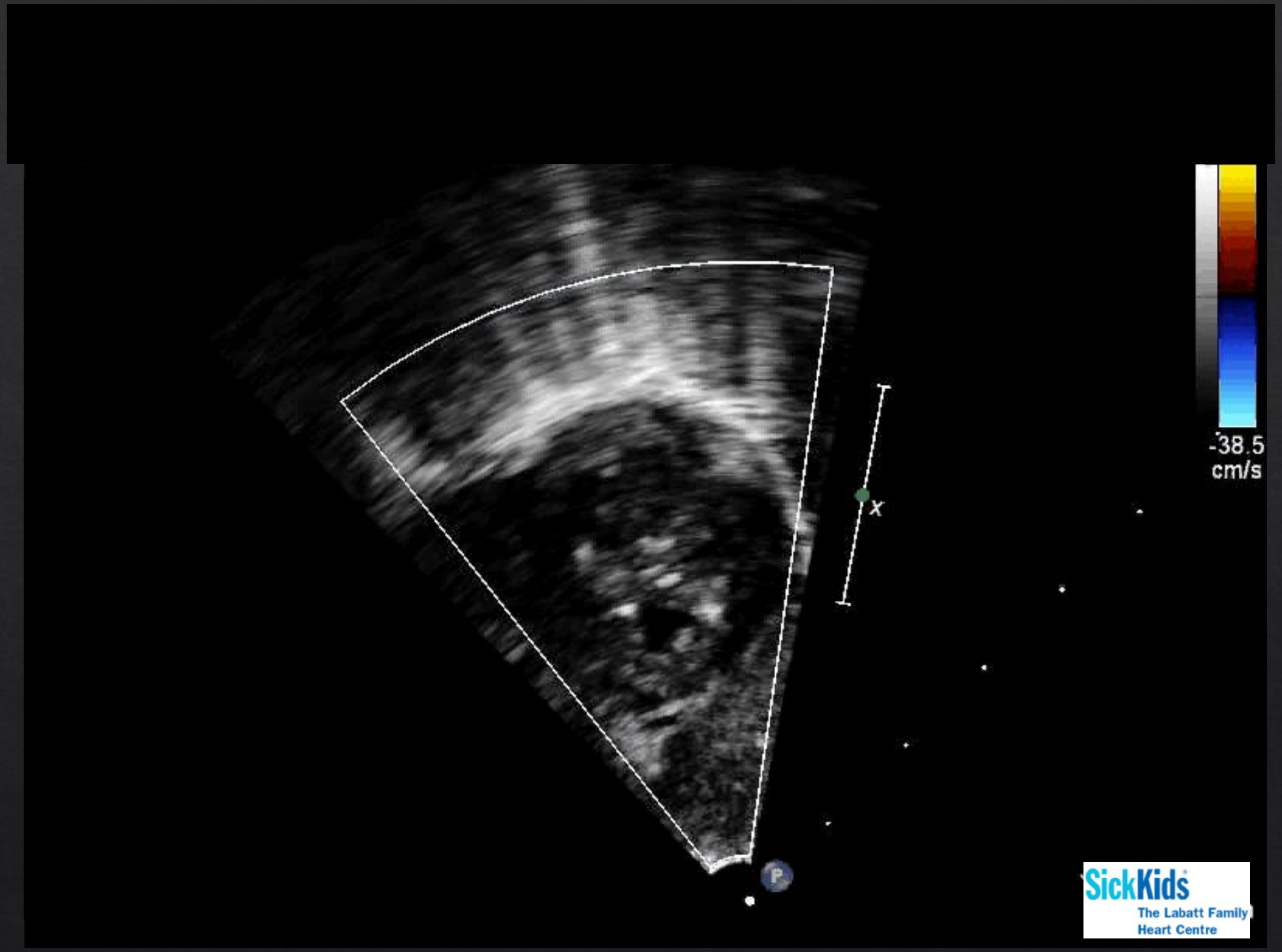
cm/s
SickKids
The Labatt Family
Heart Centre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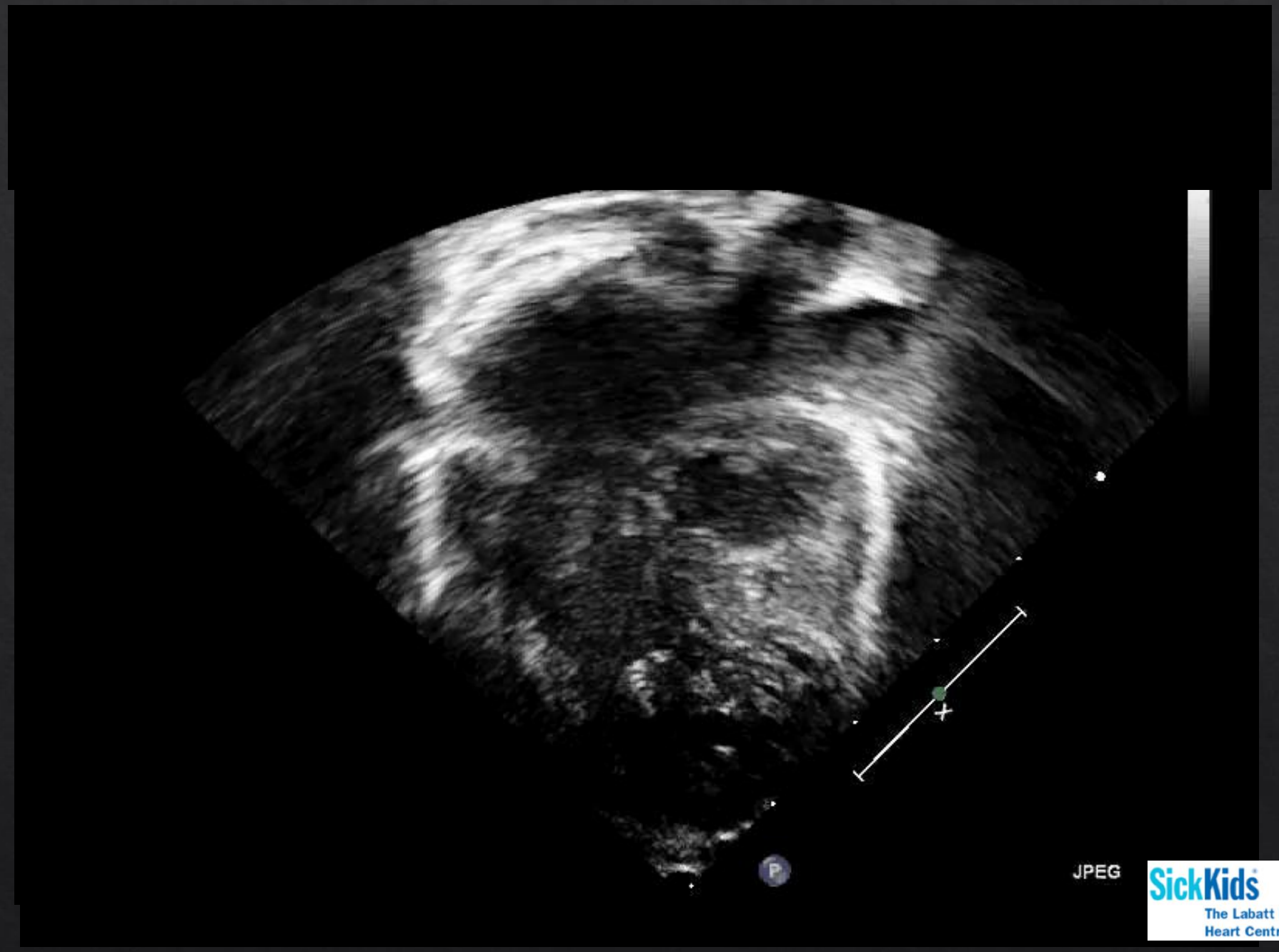
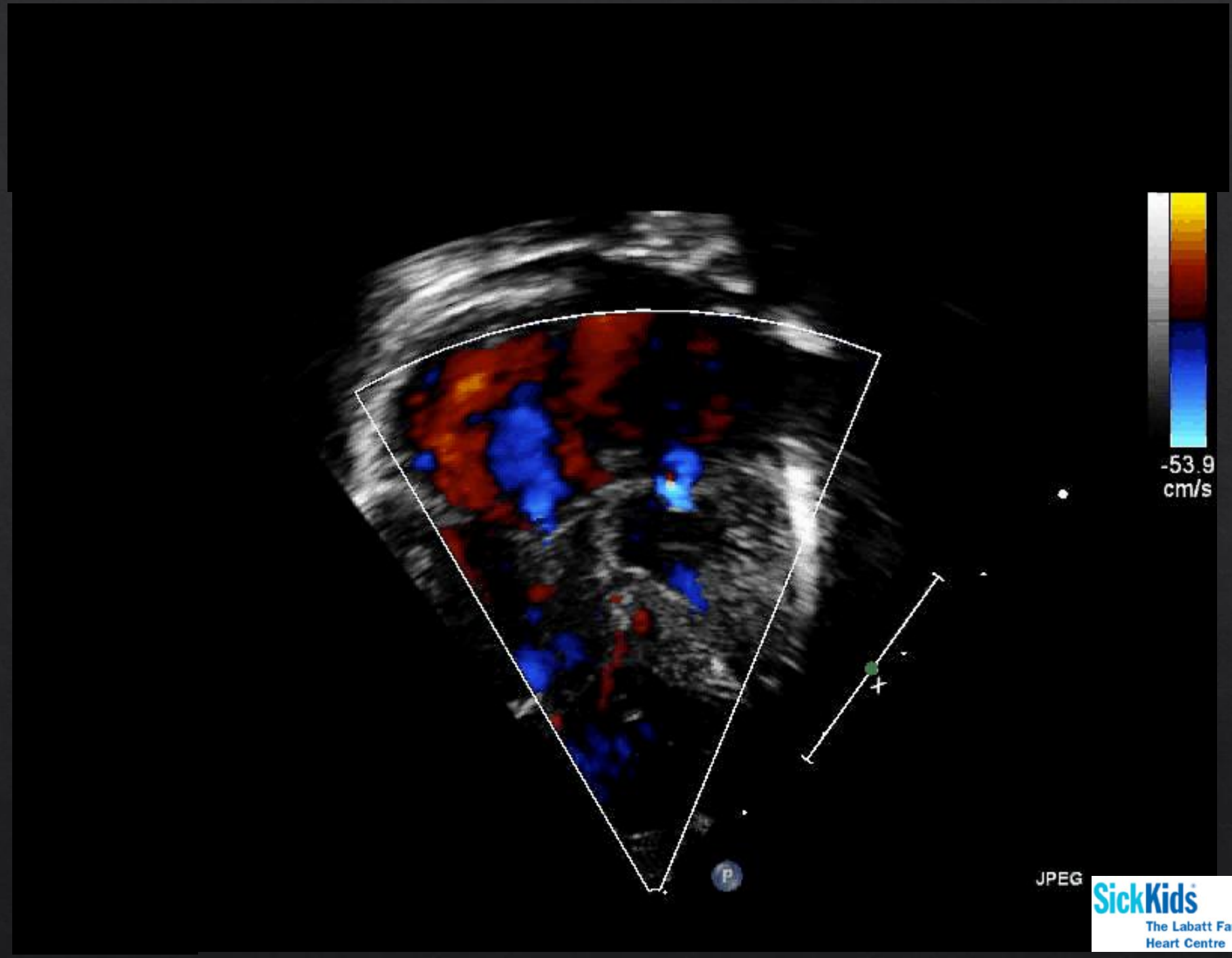
The Labatt Family
Heart Centre


P
JPEG
SickKids
The Labatt Family
Heart Centre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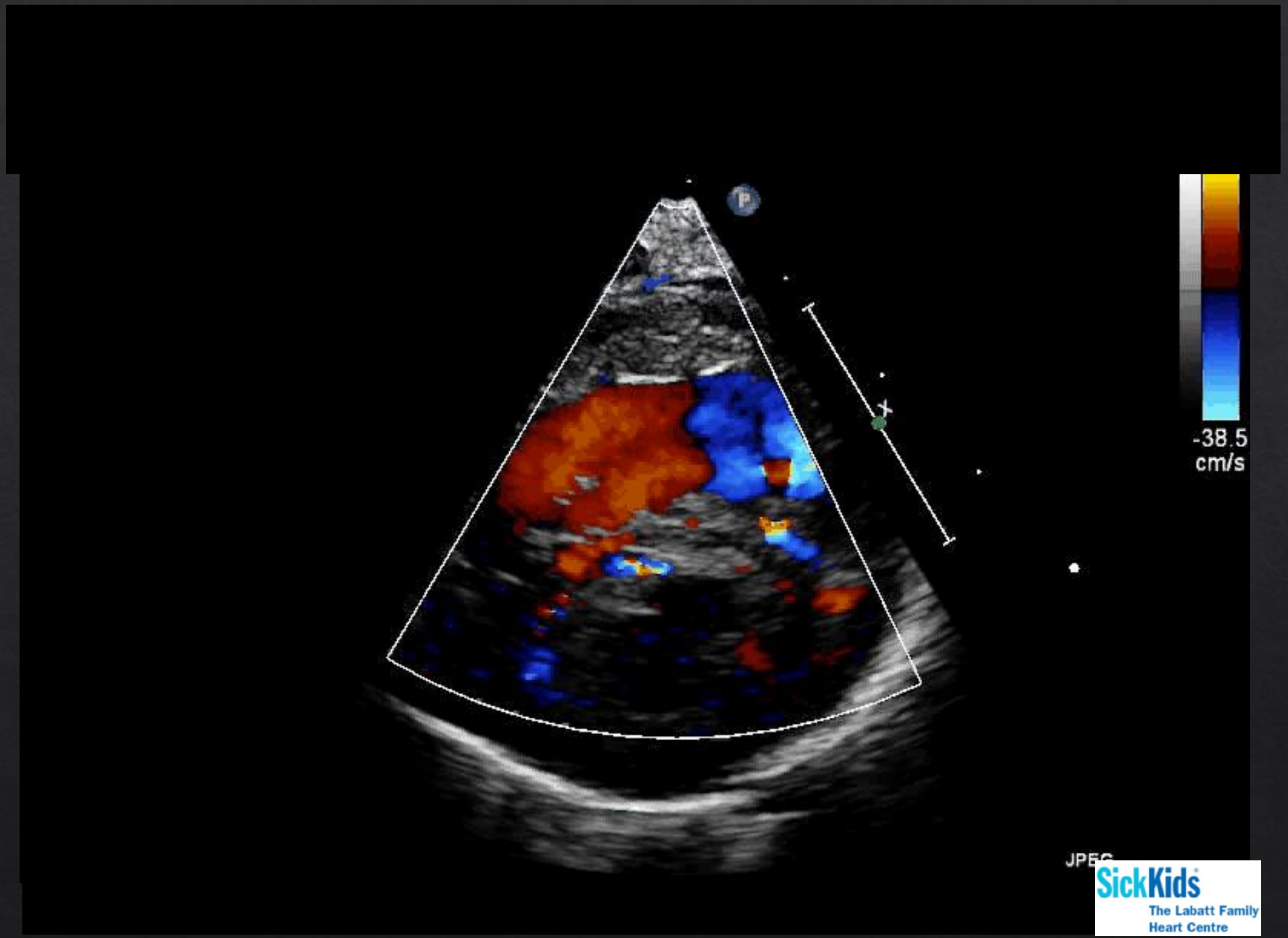
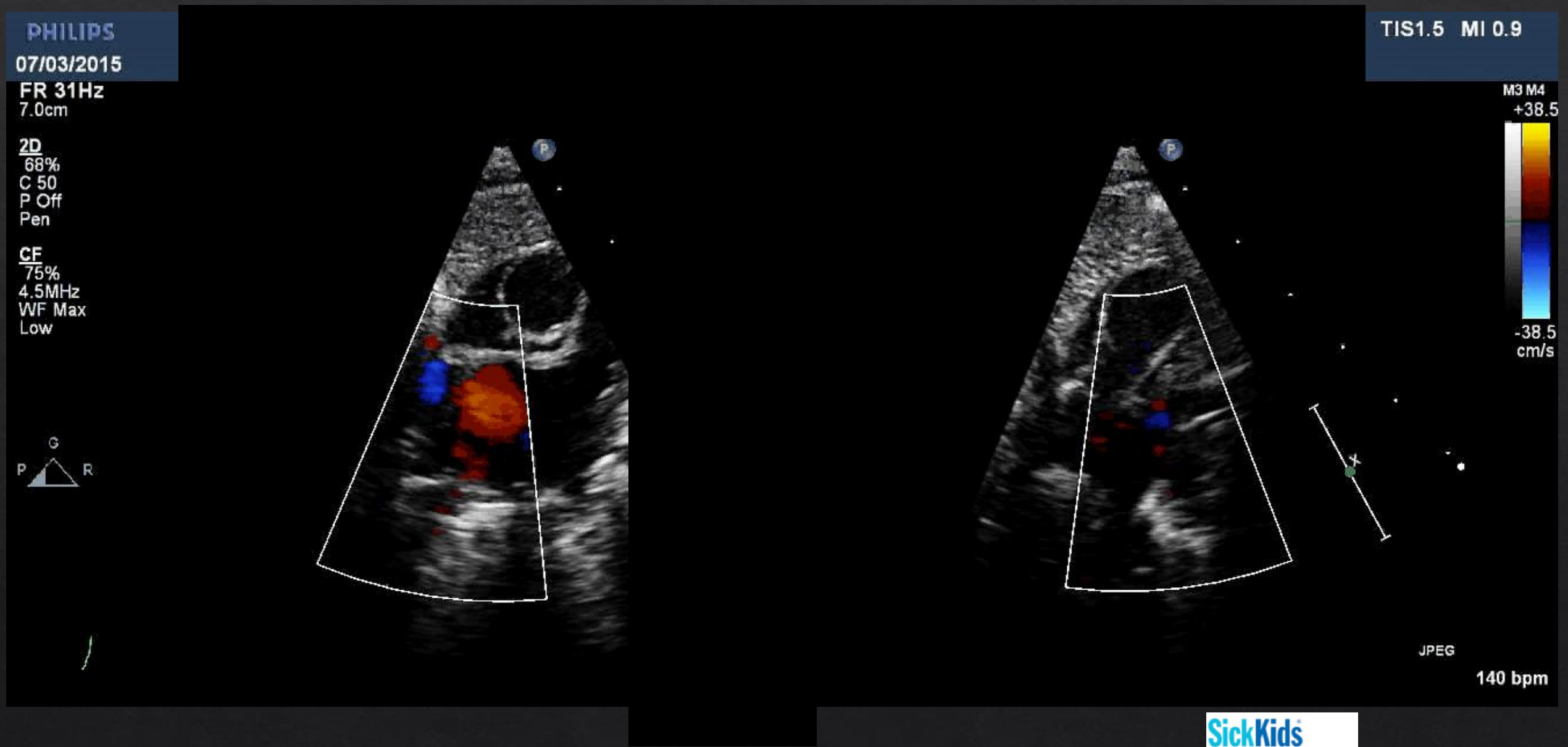
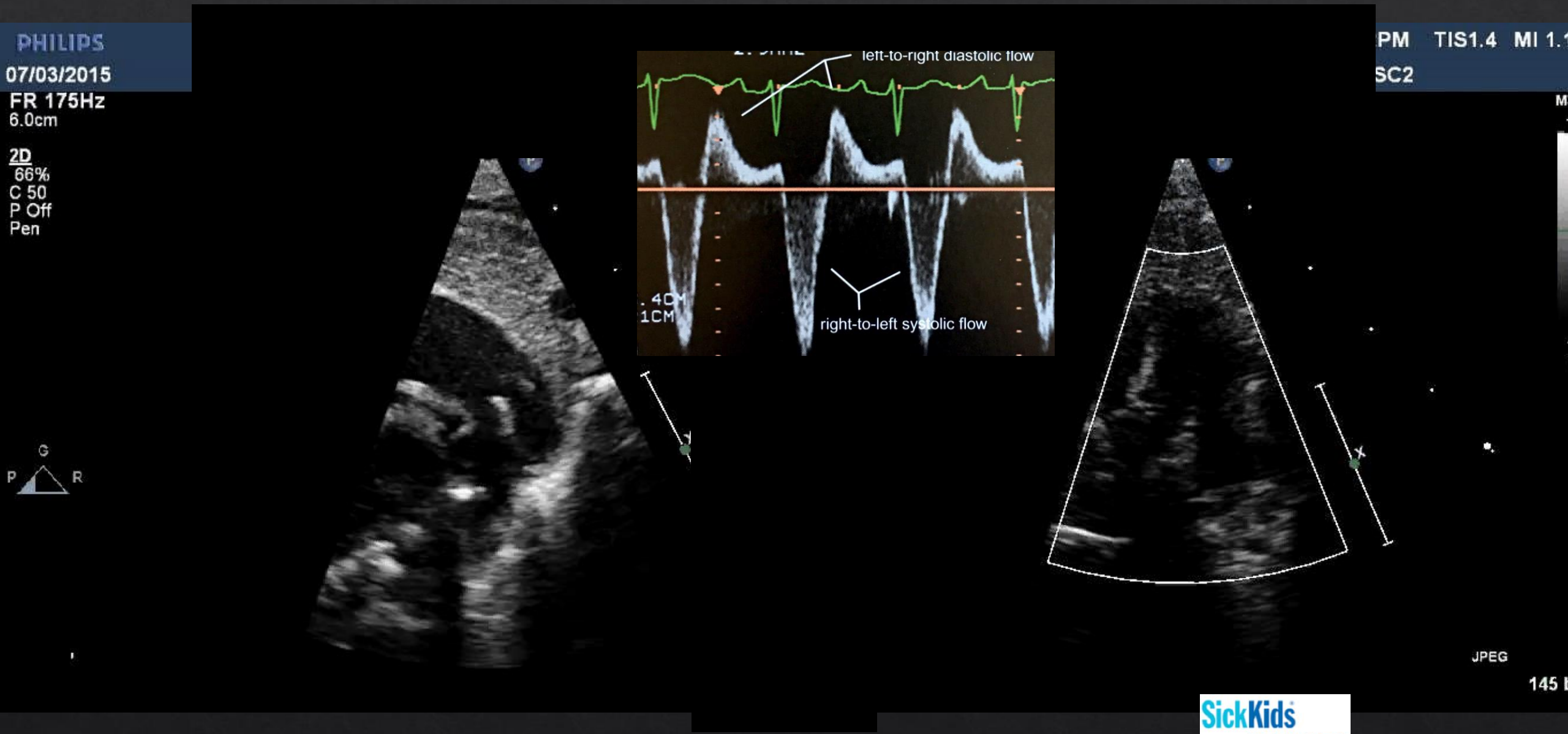
cm/s
P
JPEG
SickKids
The Labatt Family
Heart Centre


-38.5
cm/s
SickKids
The Labatt Family
Heart Centre

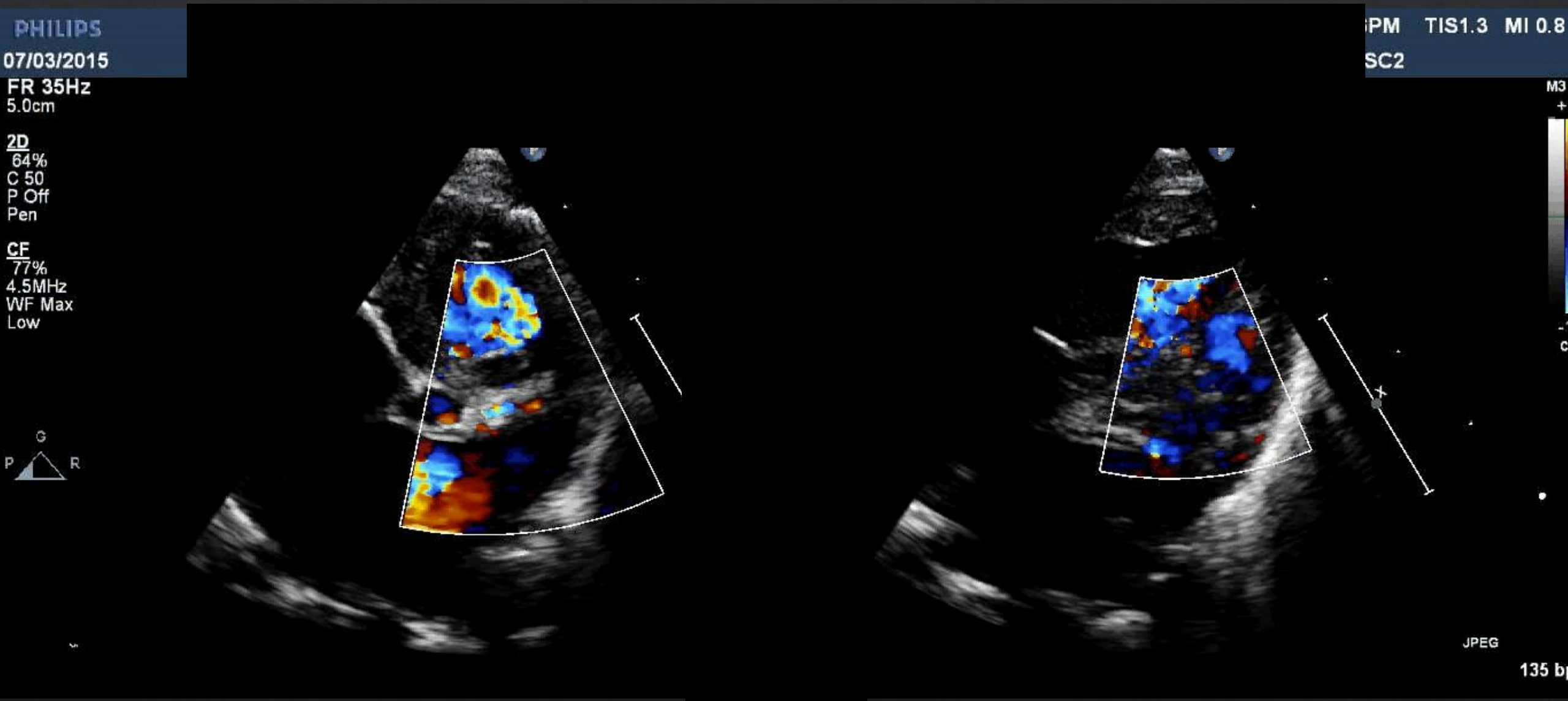

+63.9
-63.9
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre



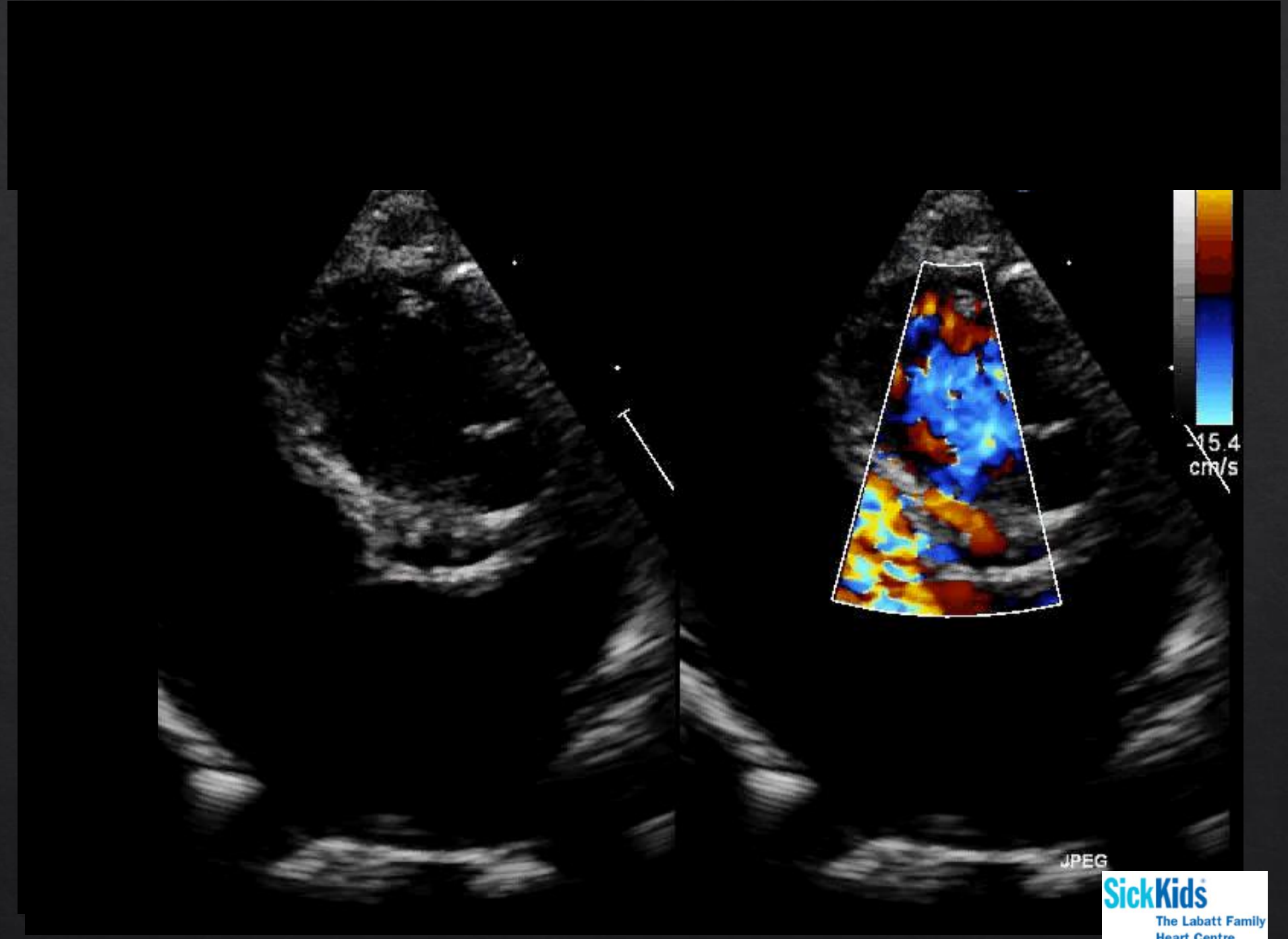
SickKids
The Labatt Family Heart Centre



SickKids
The Labatt Family Heart Centre



SickKids
The Labatt Family Heart Centre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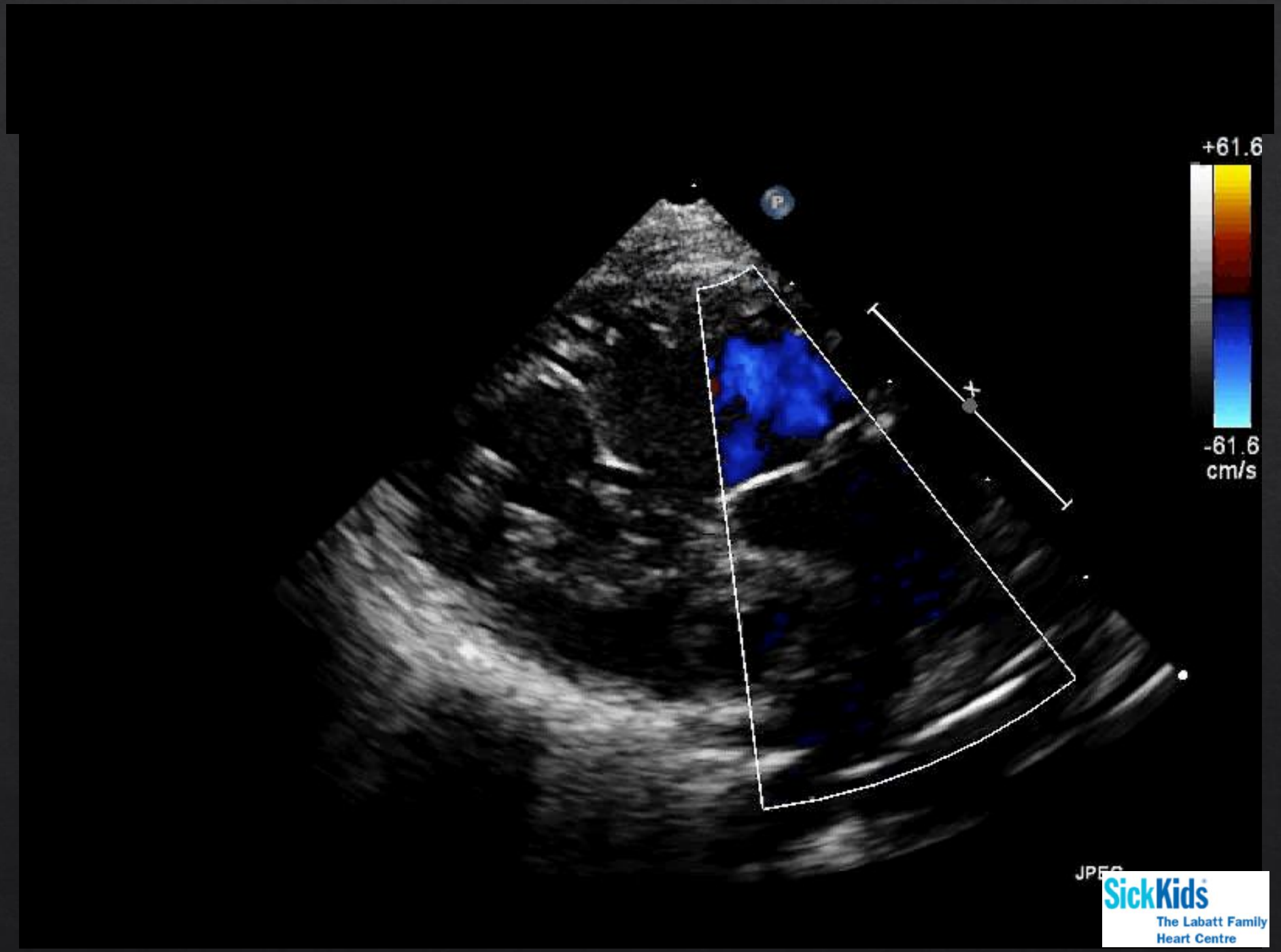
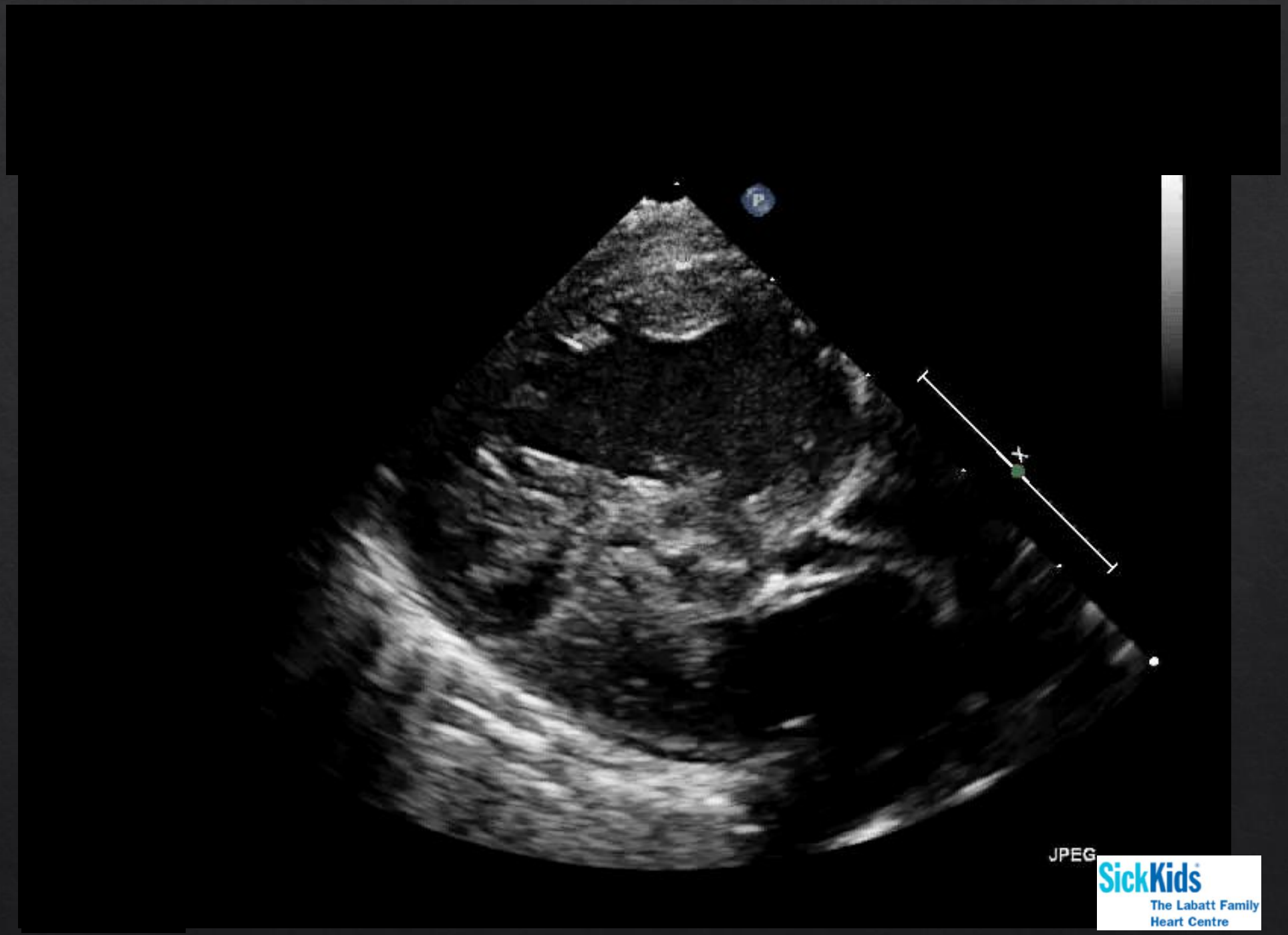
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre


+61.6
-61.6
cm/s
SickKids
The Labatt Family
Heart Centre


JPEG
SickKids
The Labatt Family
Heart Centre

Outcome

- Norwood Sano
- Pre Glenn cath: no coronary stenosis, multiple sinusoides.
- Glenn anastomosis : 6 month old
- Waiting Fontan : pre Cath - adequate pressures.

Case 3

- Newborn, prenatal diagnosis: HLHS, restrictive PFO ….

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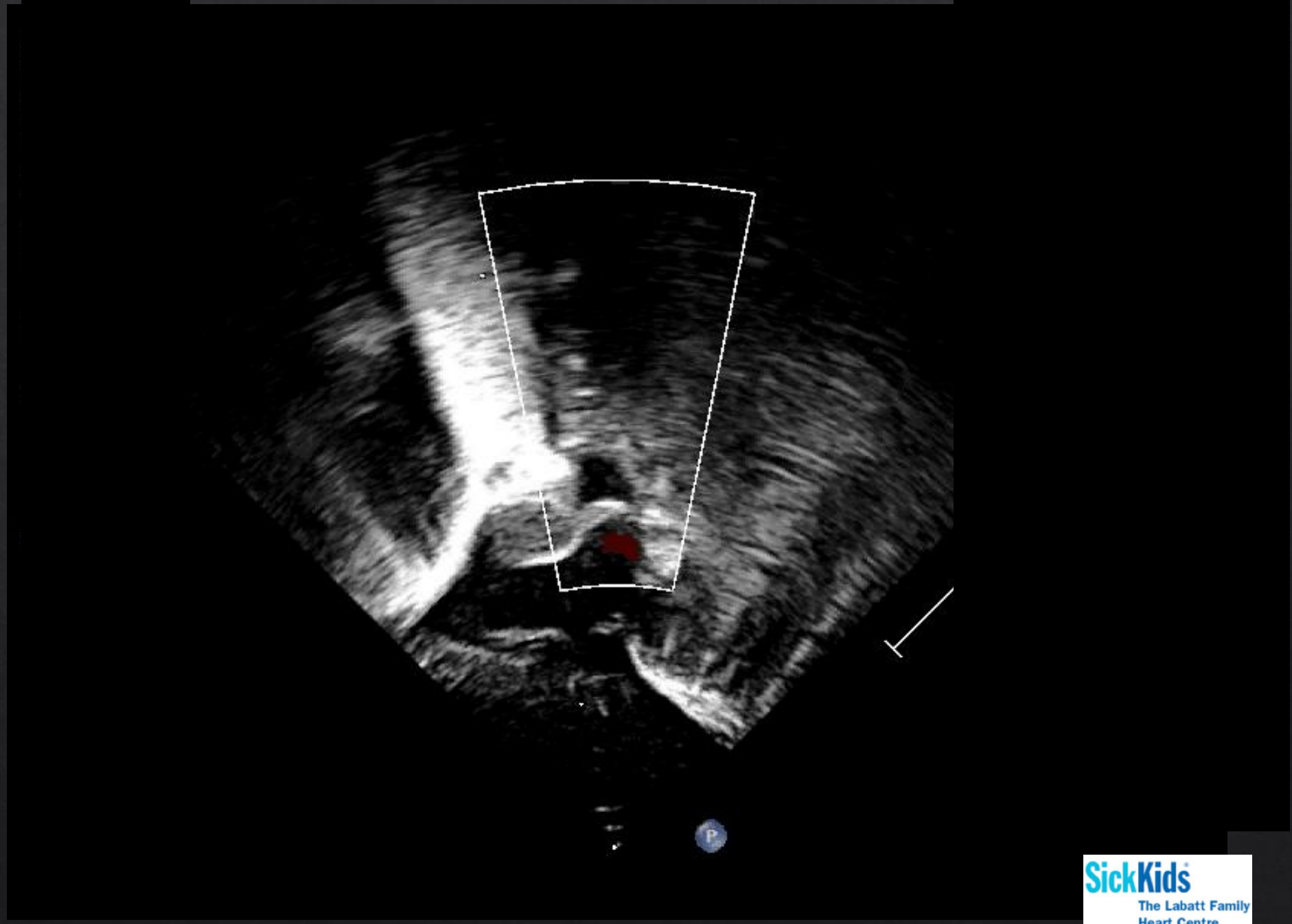
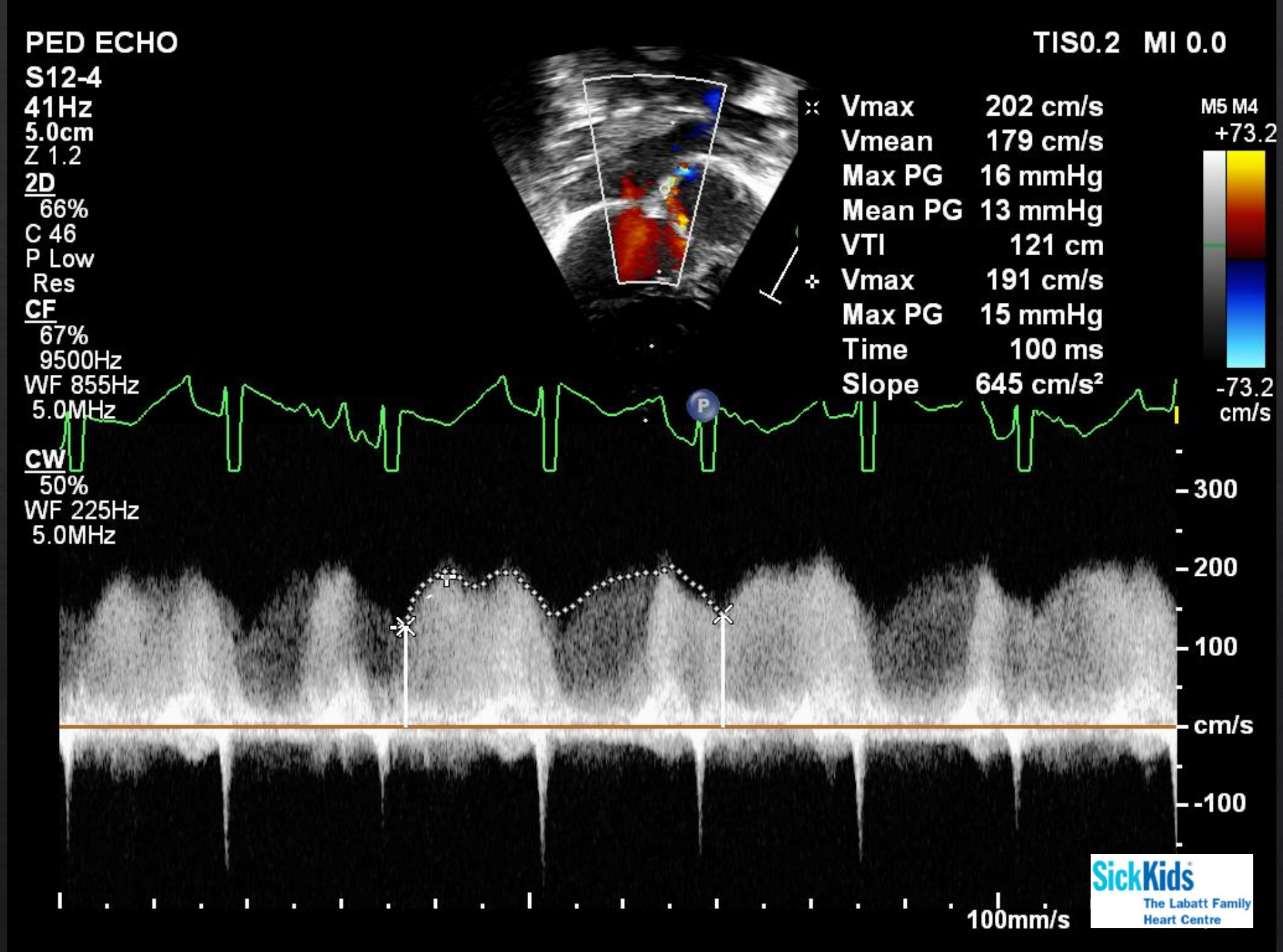
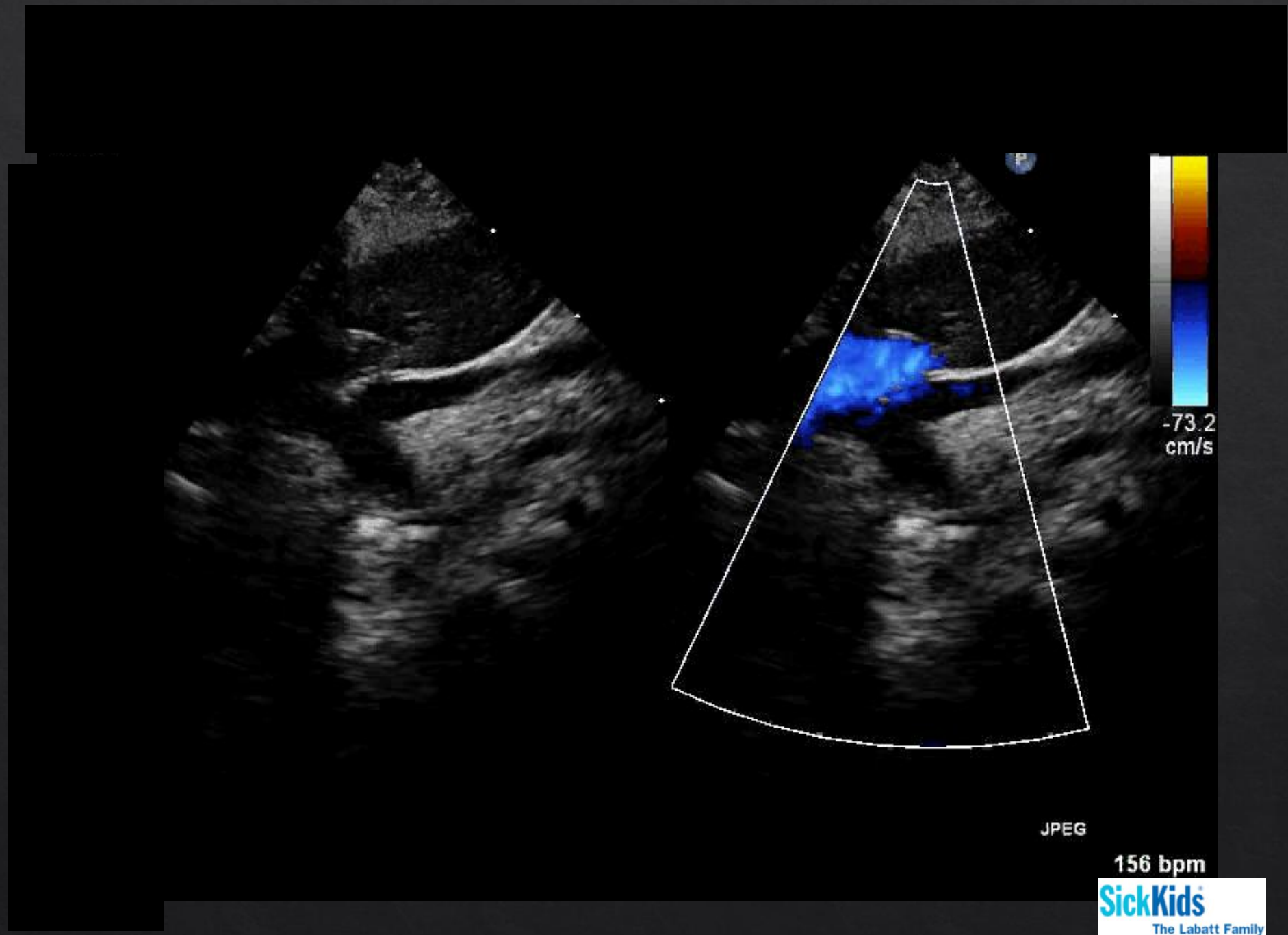
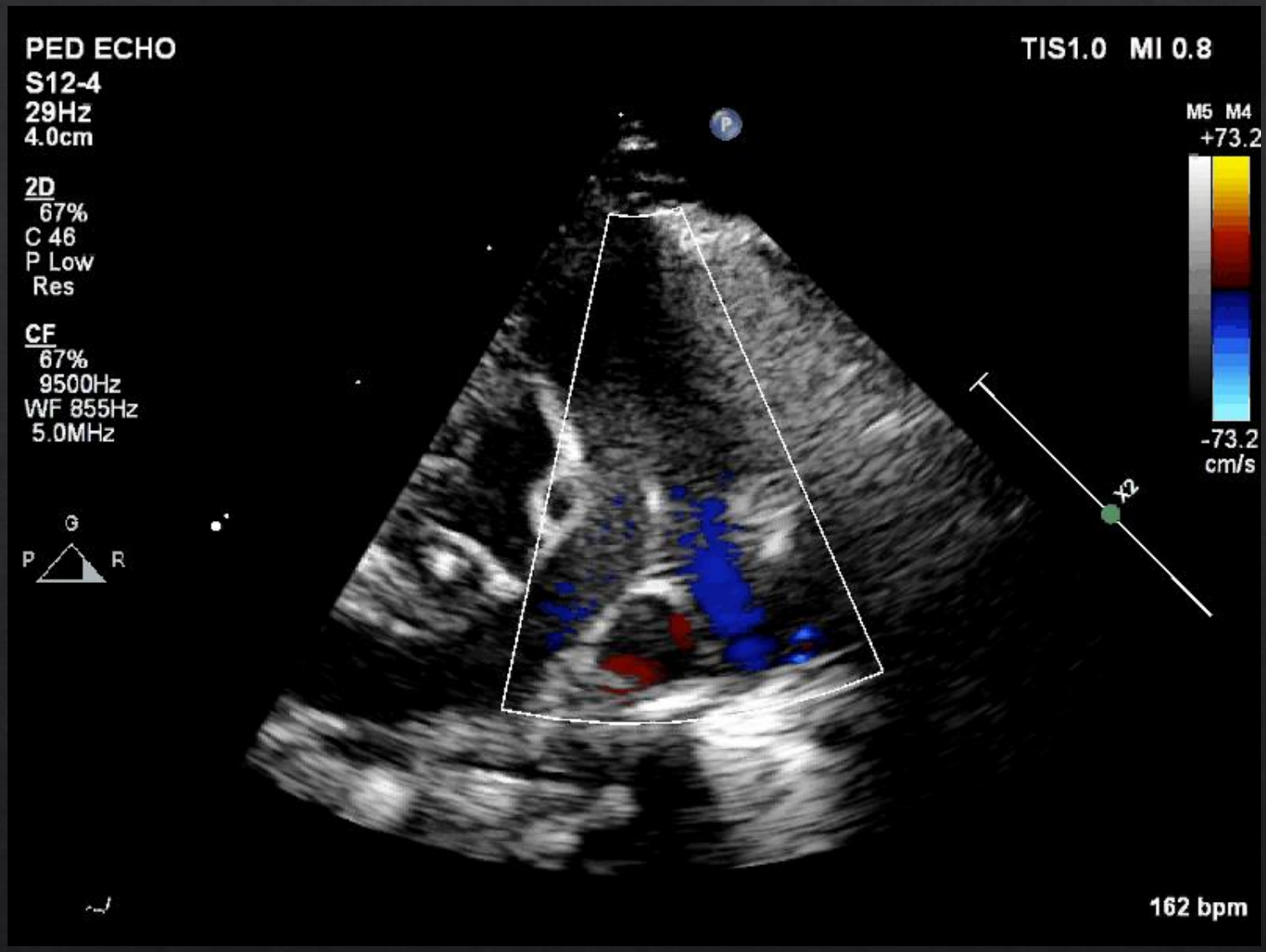


SickKids
The Labatt Family Heart Centre

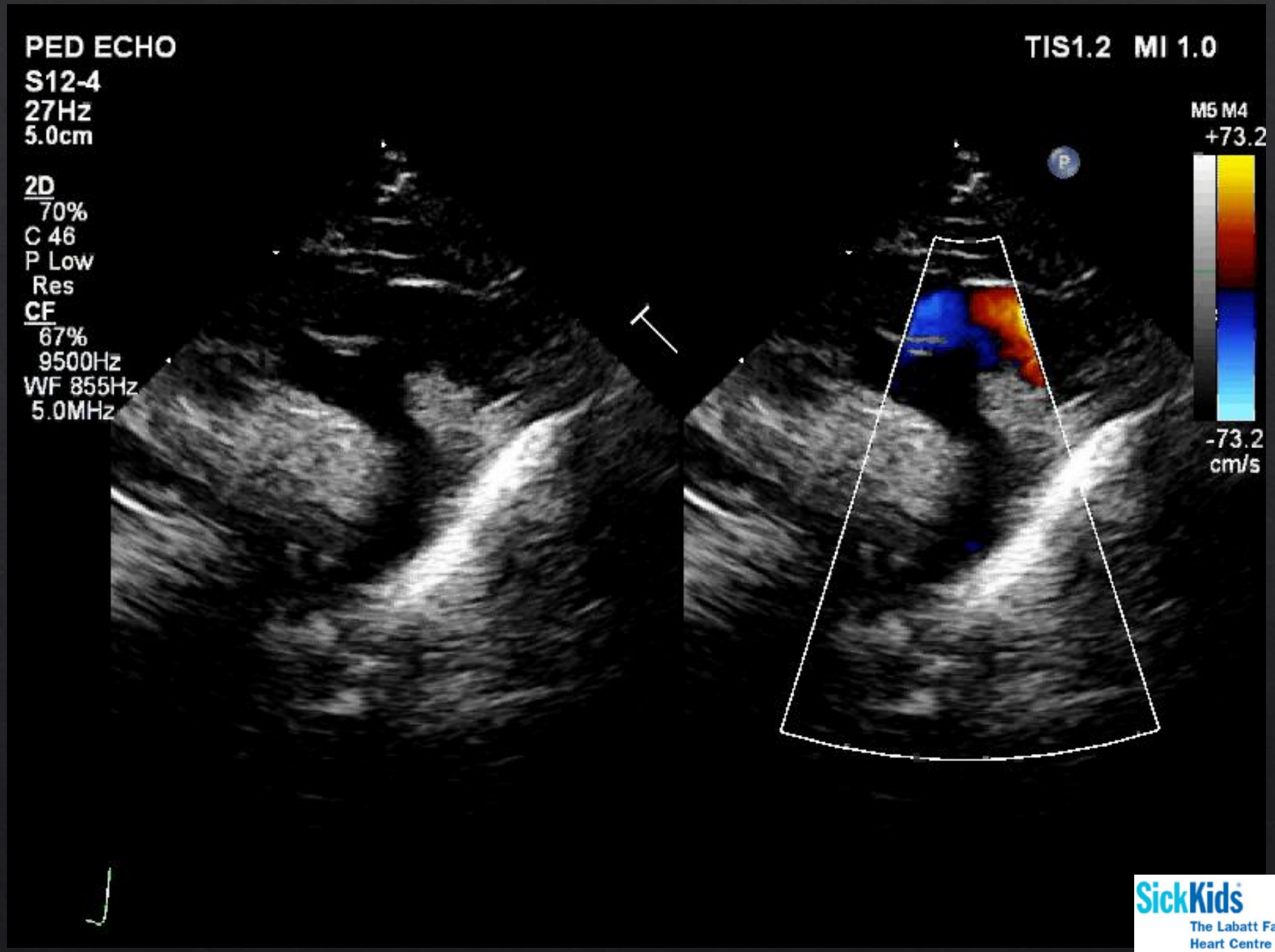

PED ECHO
S12-4
41Hz
5.0cm
Z 1.2
2D
66%
C 46
P Low
Res
CF
67%
9500Hz
WF 855Hz
5.0MHz
CW
50%
WF 225Hz
5.0MHz
TIS0.2 MI 0.0
Vmax 202 cm/s
Vmean 179 cm/s
Max PG 16 mmHg
Mean PG 13 mmHg
VTI 121 cm
Vmax 191 cm/s
Max PG 15 mmHg
Time 100 ms
Slope 645 cm/s²
M5 M4
+73.2
-73.2
cm/s
300
200
100
cm/s
-100
100mm/s
SickKids
The Labatt Family
Heart Centre

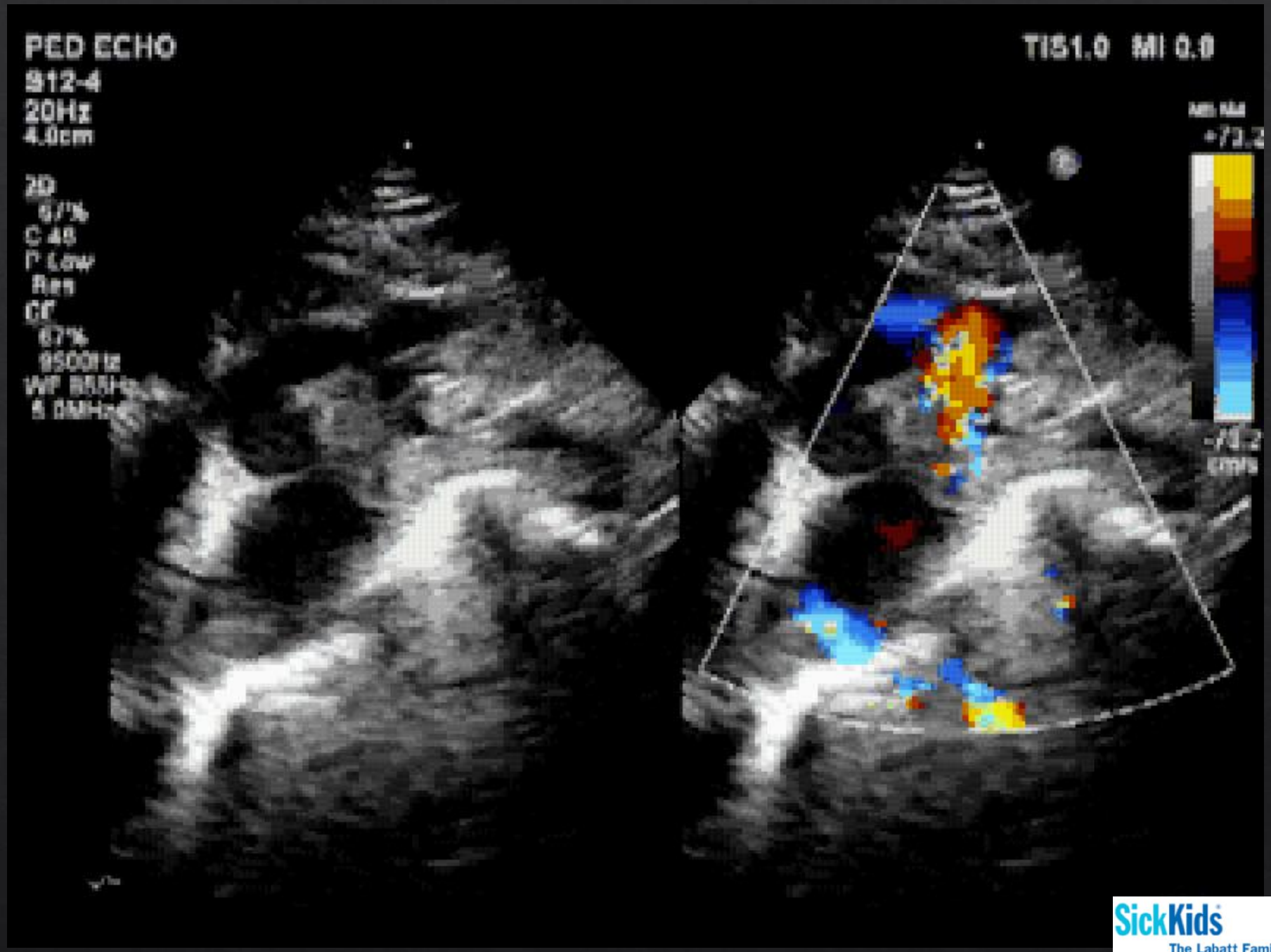

-73.2
cm/s
JPEG
156 bpm
SickKids
The Labatt Family
Heart Centre

SickKids
The Labatt Family Heart Centre

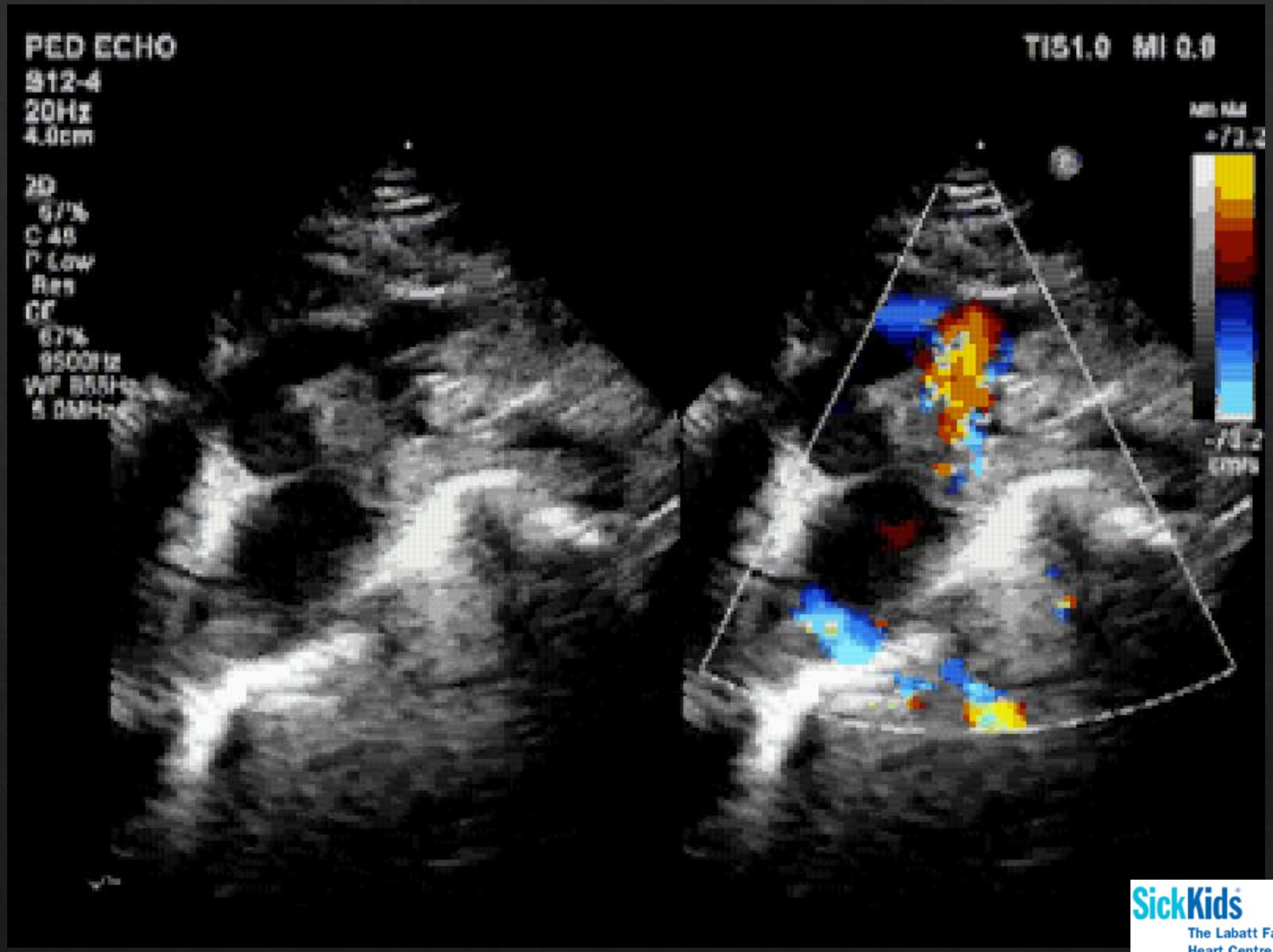


SickKids
The Labatt Family Heart Centre


PED ECHO
S12-4
27Hz
5.0cm
2D
70%
C 46
P Low
Res
CF
67%
9500Hz
WF 855Hz
5.0MHz
TIS1.2 MI 1.0
M5 M4
+73.2
-73.2
cm/s
SickKids
The Labatt Family
Heart Centre



SickKids
The Labatt Family Heart Centre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912-4
20Hz
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2D
C 49
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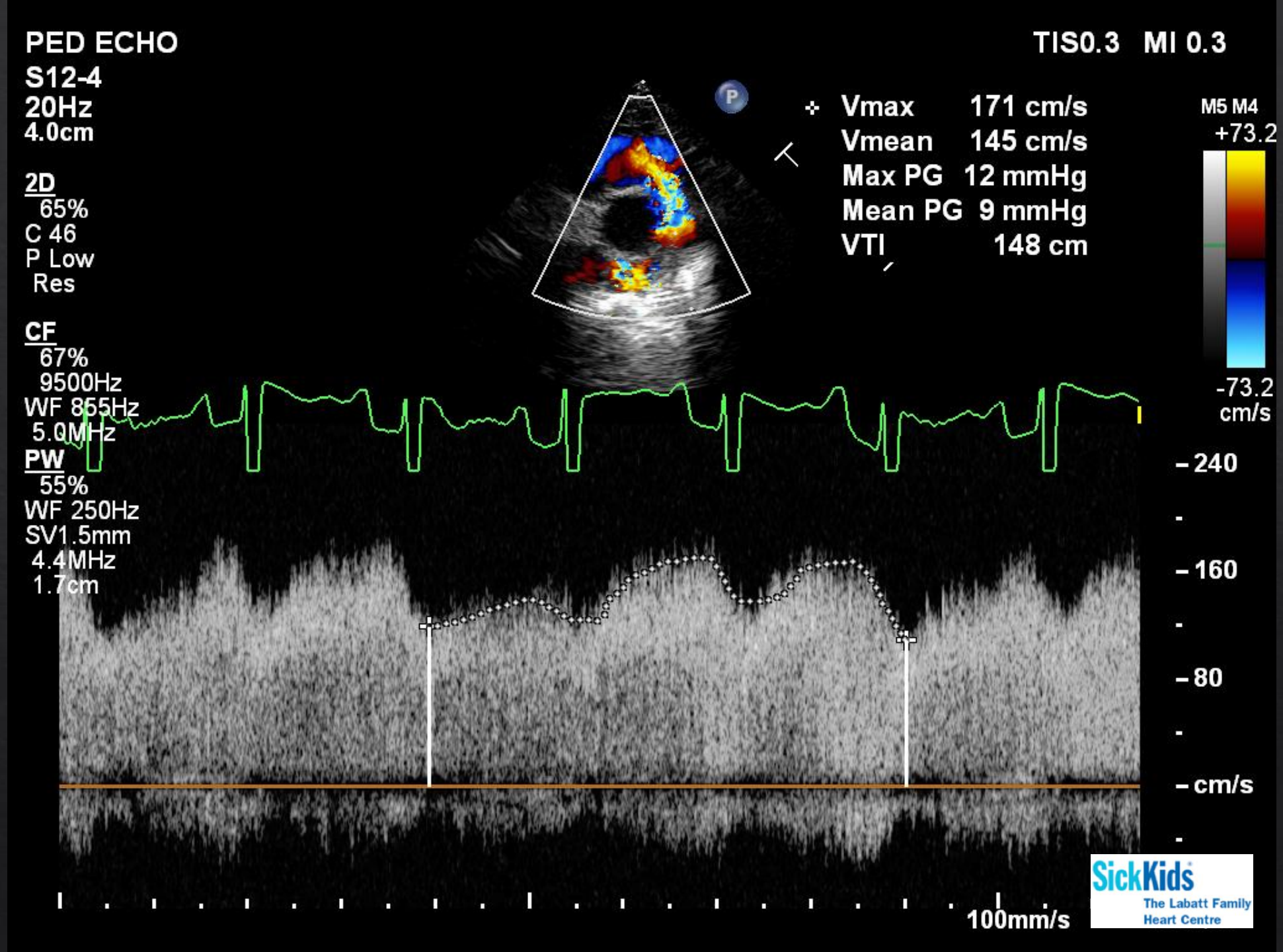
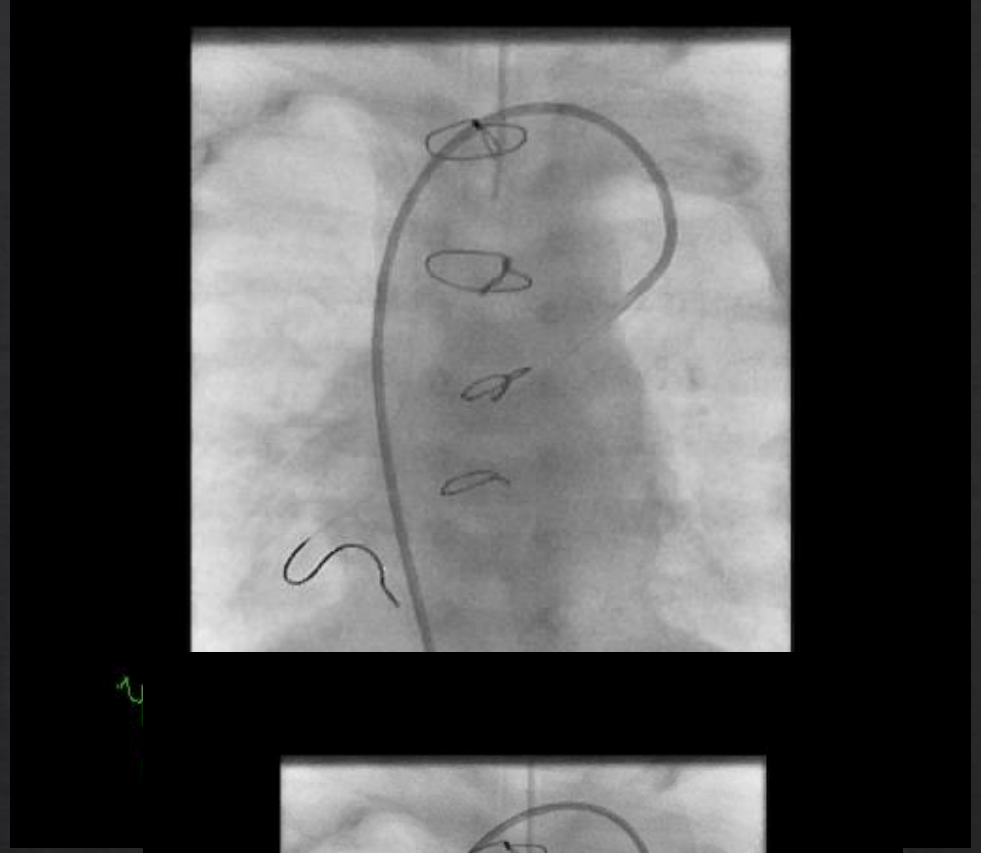
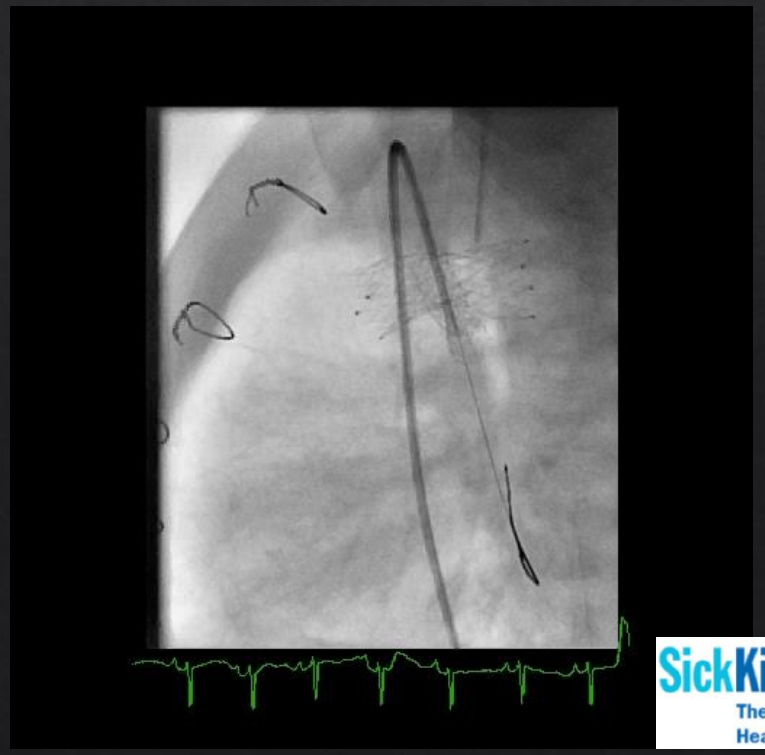
Res
CF
67%
TIS1.0 MI 0.8
SickKids
The Labatt Family
Heart Centre


PED ECHO
S12-4
20Hz
4.0cm
2D
65%
C 46
P Low
Res
CF
67%
9500Hz
WF 855Hz
5.0MHz
PW
55%
WF 250Hz
SV1.5mm
4.4MHz
1.7cm
TIS0.3 MI 0.3
Vmax 171 cm/s
Vmean 145 cm/s
Max PG 12 mmHg
Mean PG 9 mmHg
VTI 148 cm
M5 M4
+73.2
-73.2
cm/s
240
160
80
cm/s
100mm/s
SickKids
The Labatt Family
Heart Centre





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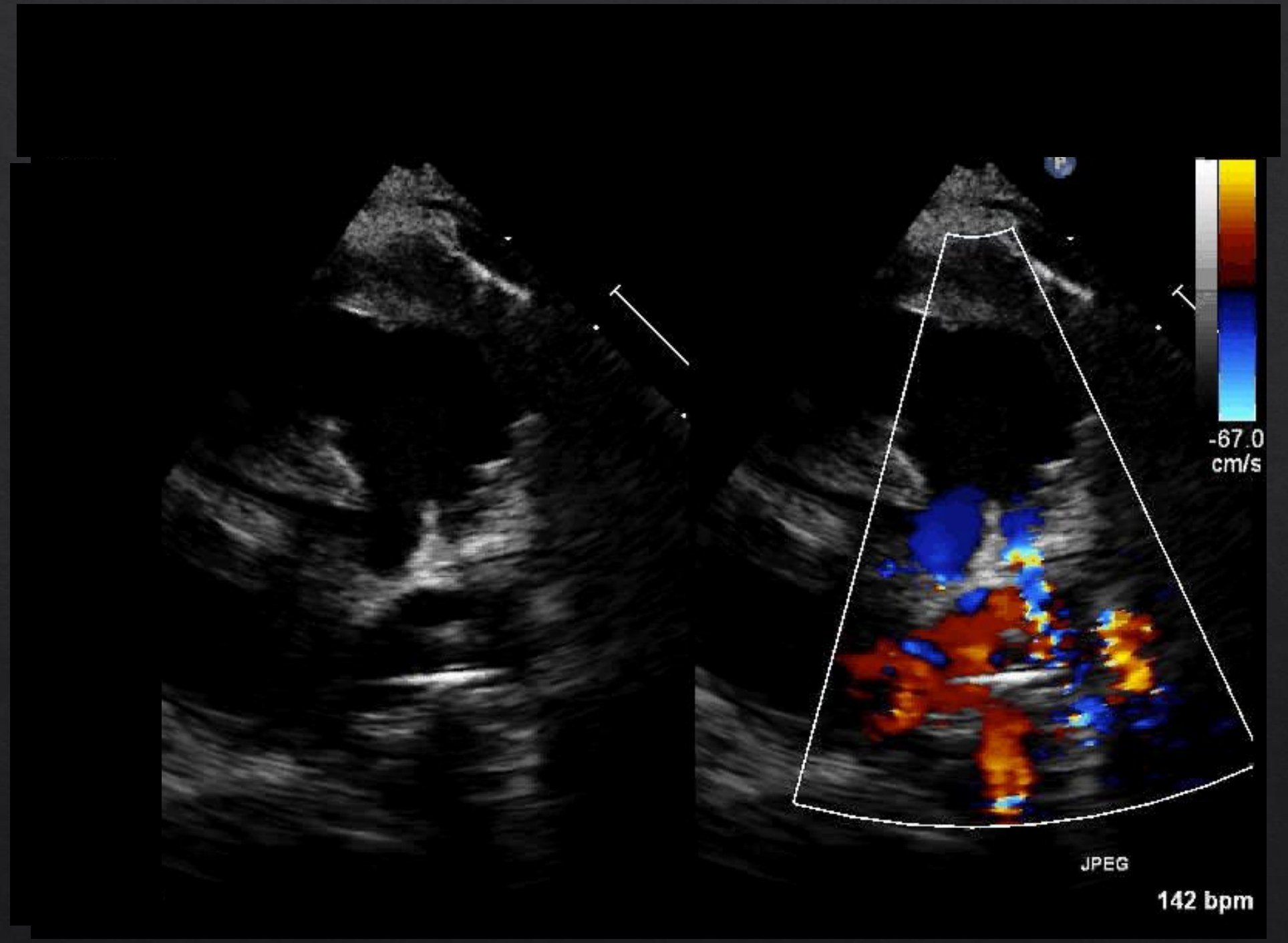
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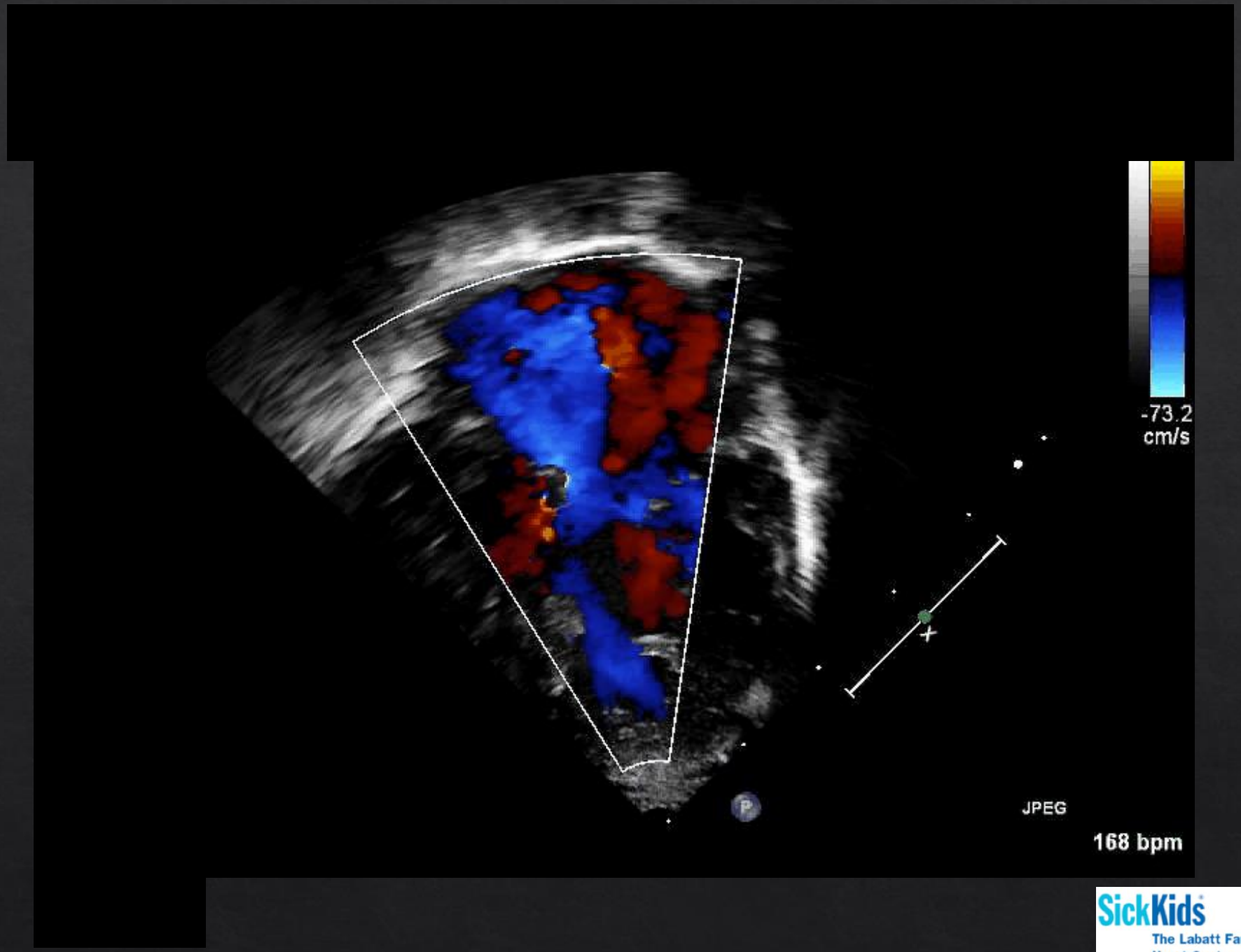

SickKids
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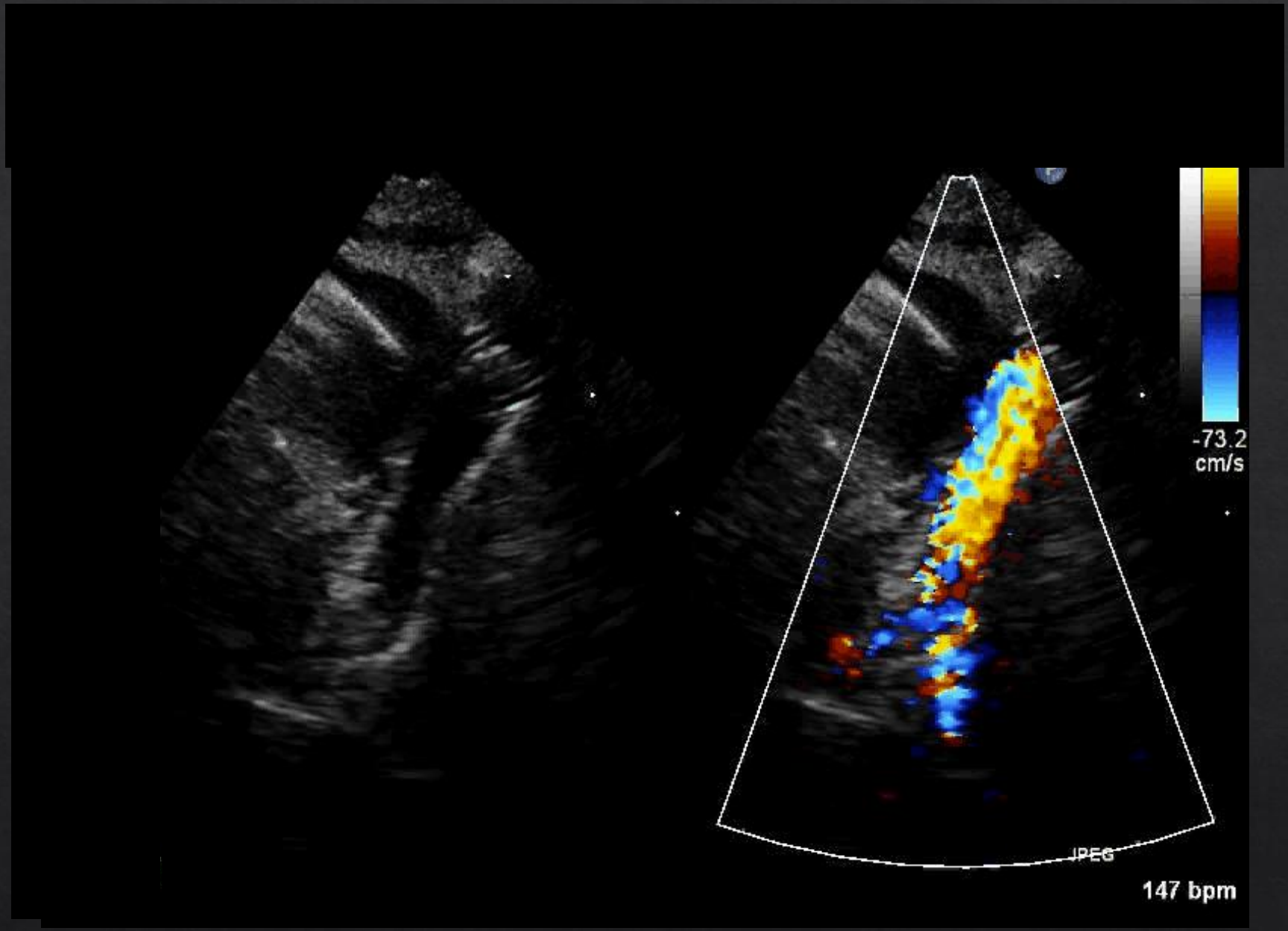
Heart Centre


P
-77.0
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre



SickKids
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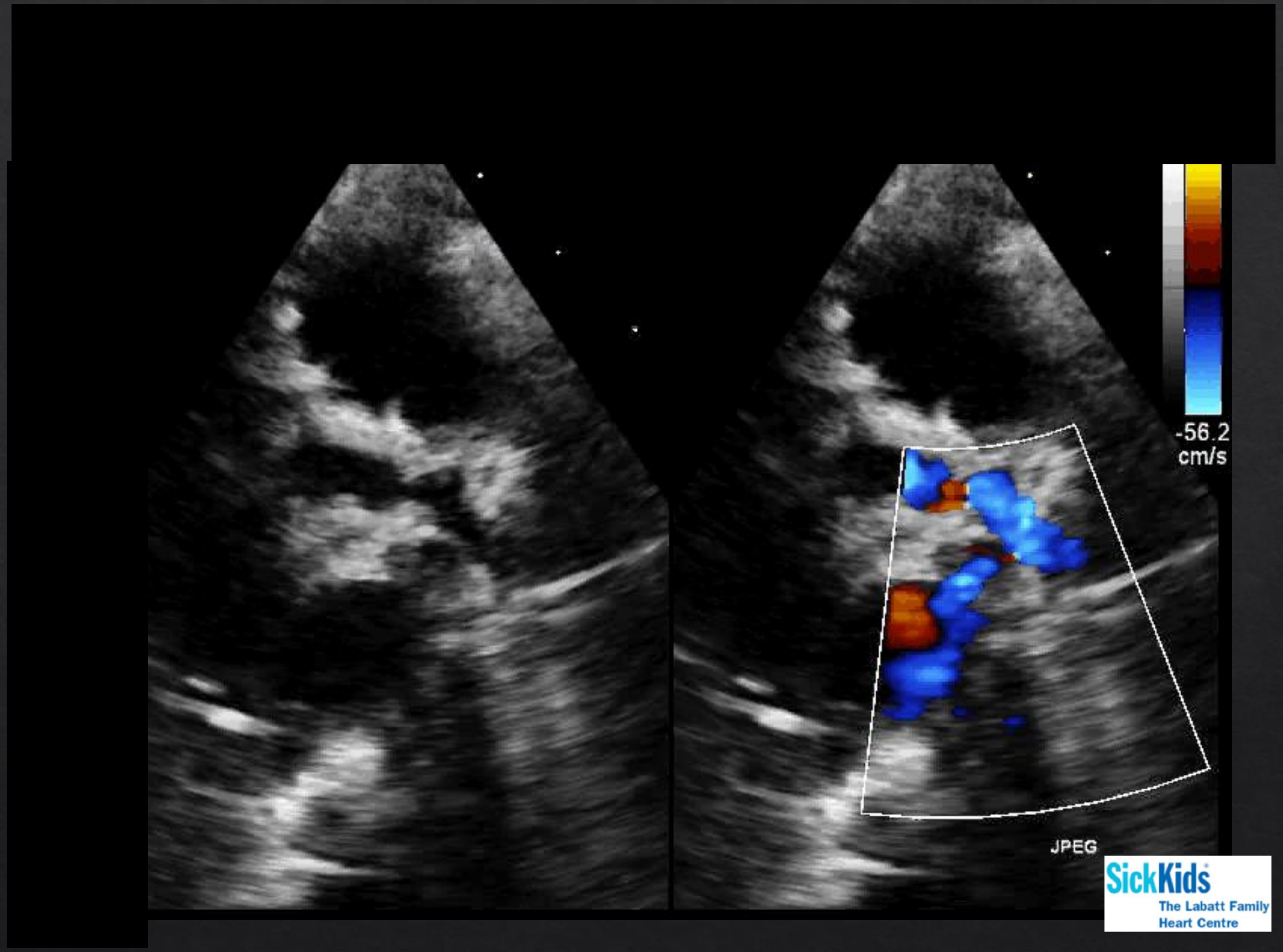
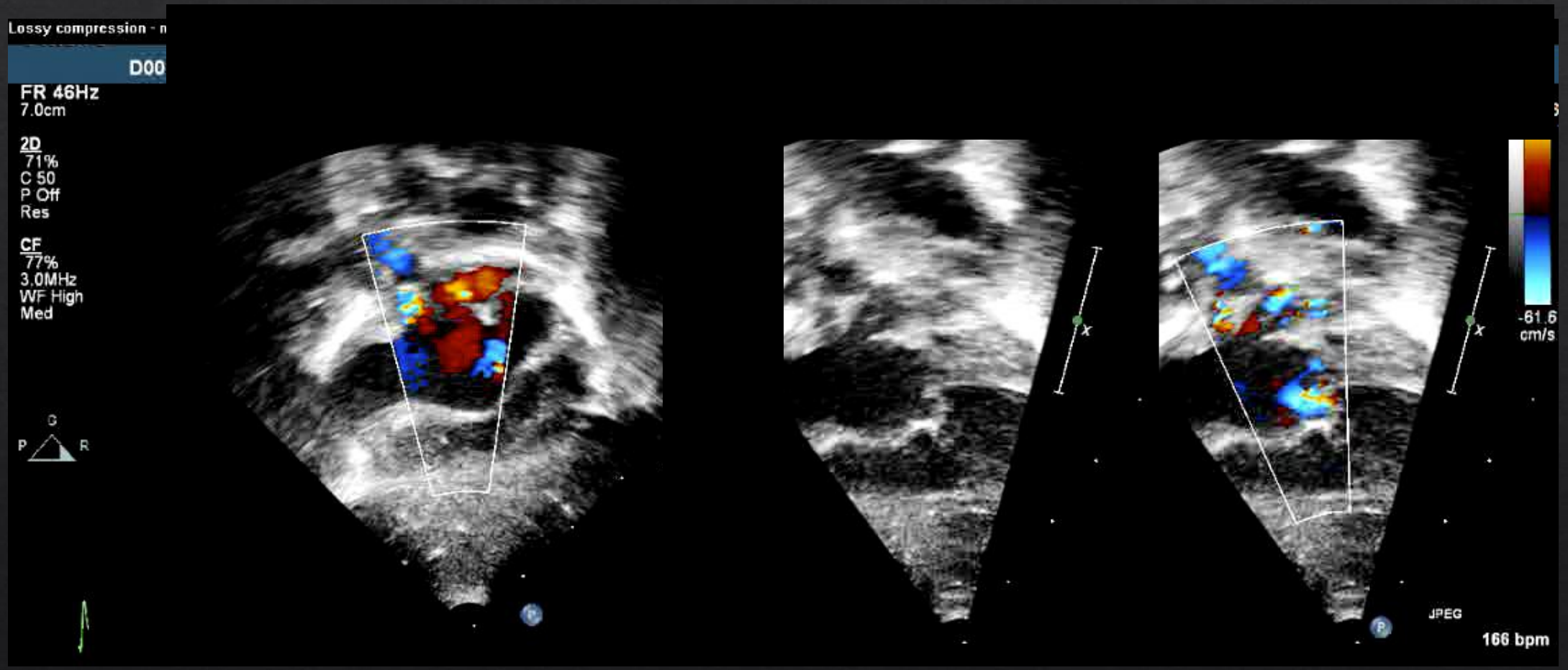
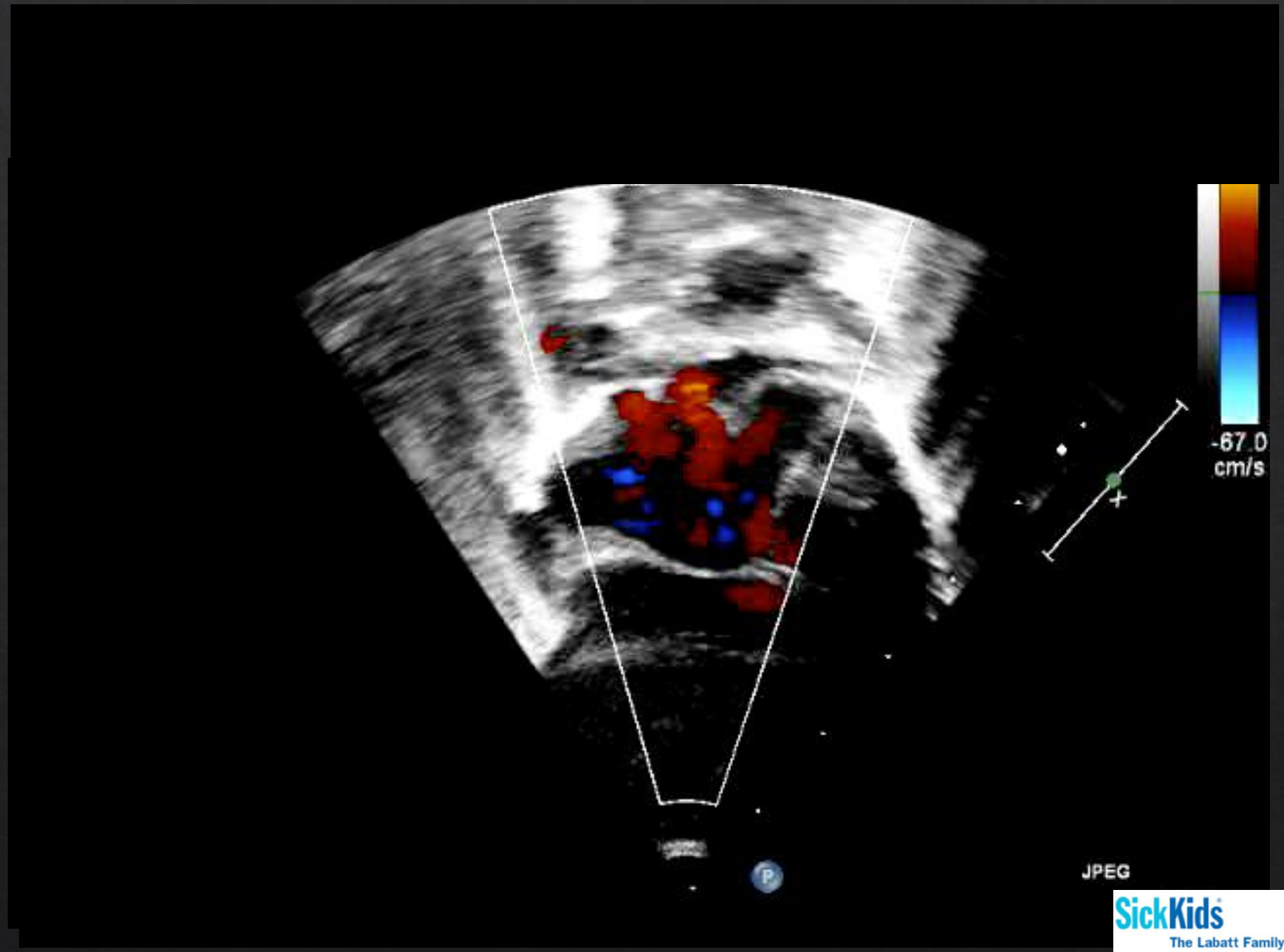

-56.2
cm/s
JPEG
SickKids
The Labatt Family
Heart Centre

case 4



SickKids
The Labatt Family Heart Centre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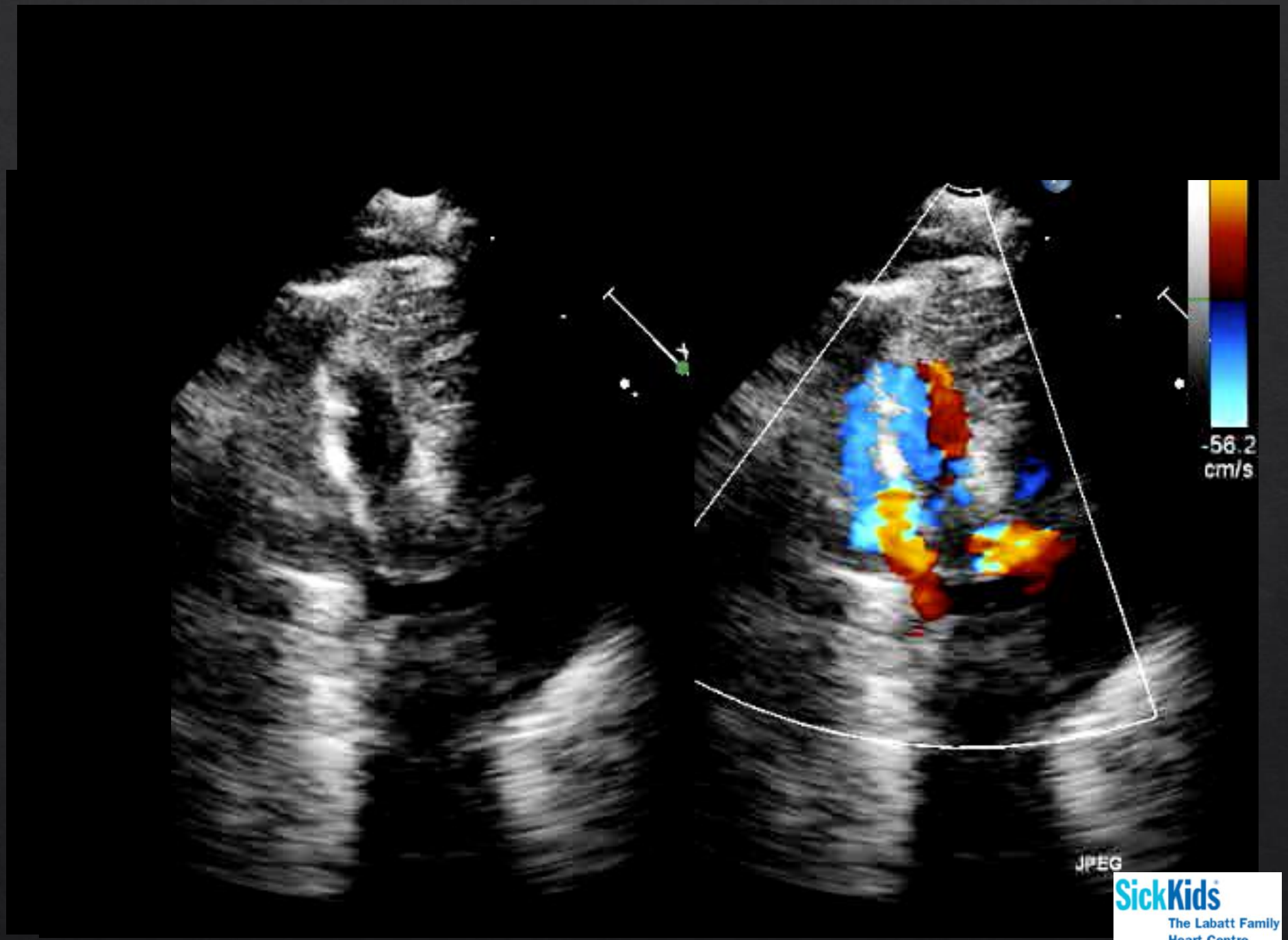
The Labatt Family
Heart Centre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cm/s
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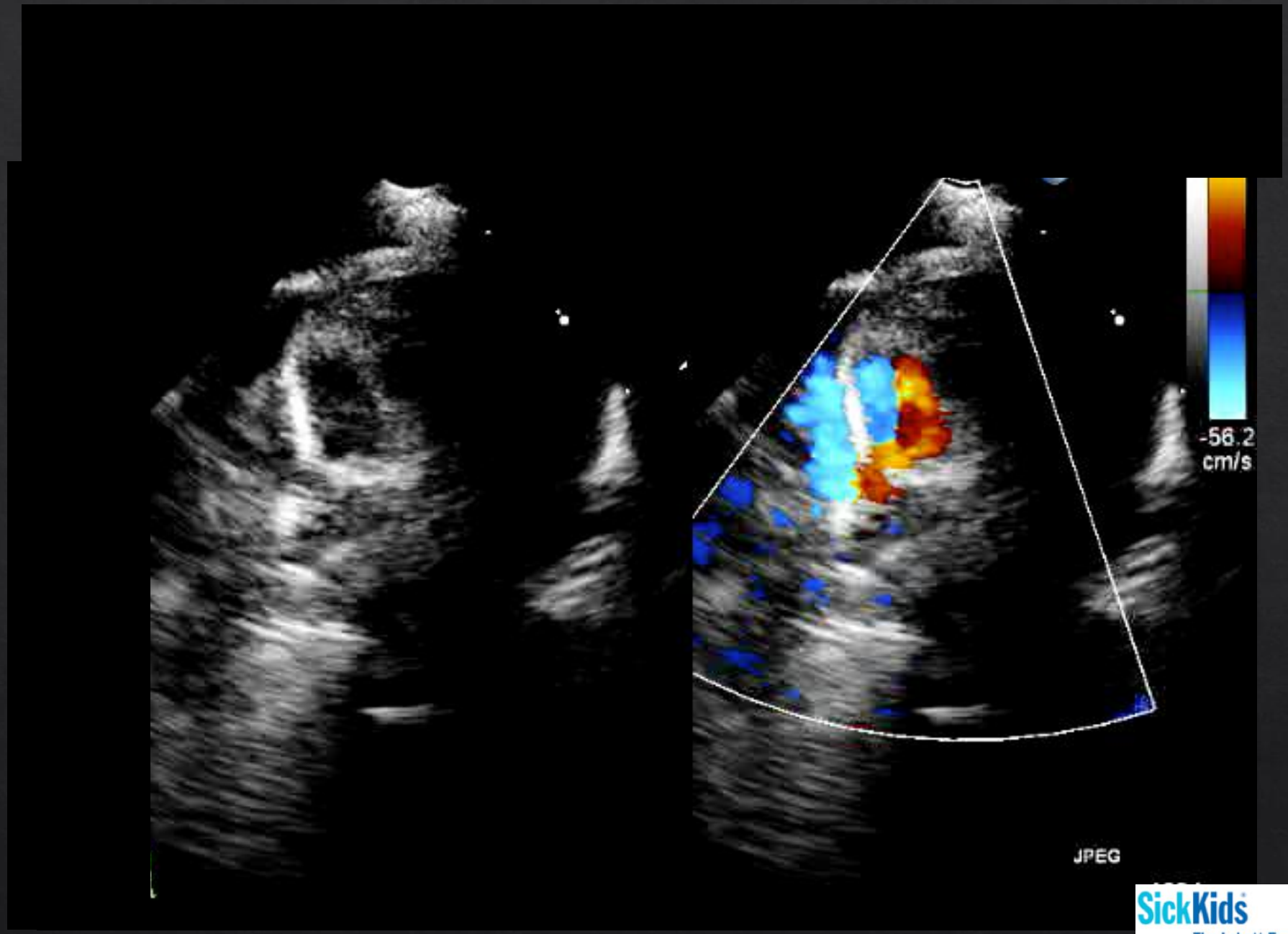
SickKids
The Labatt Family
Heart Centre



SickKids
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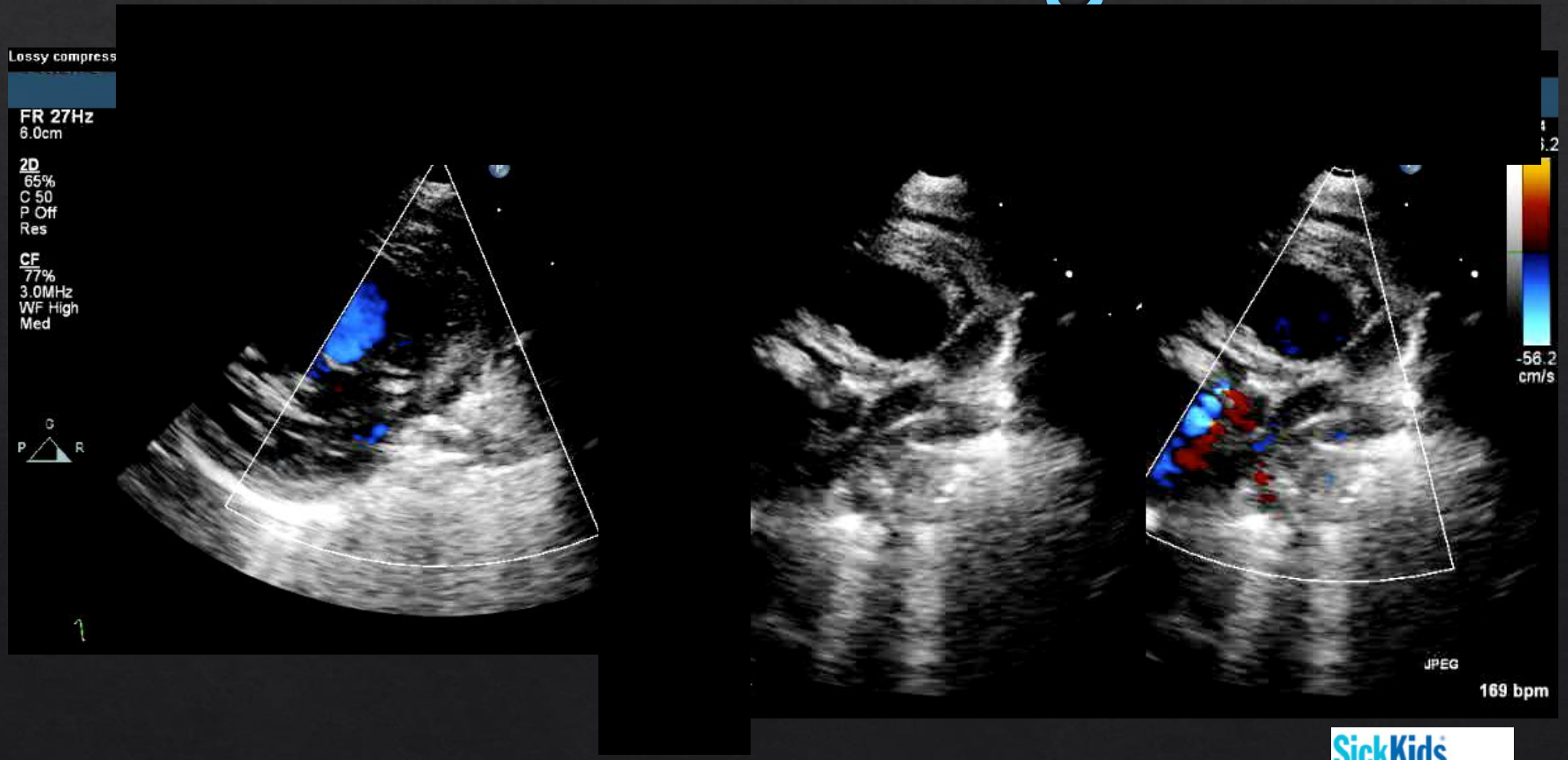
Left SVC draining into CS

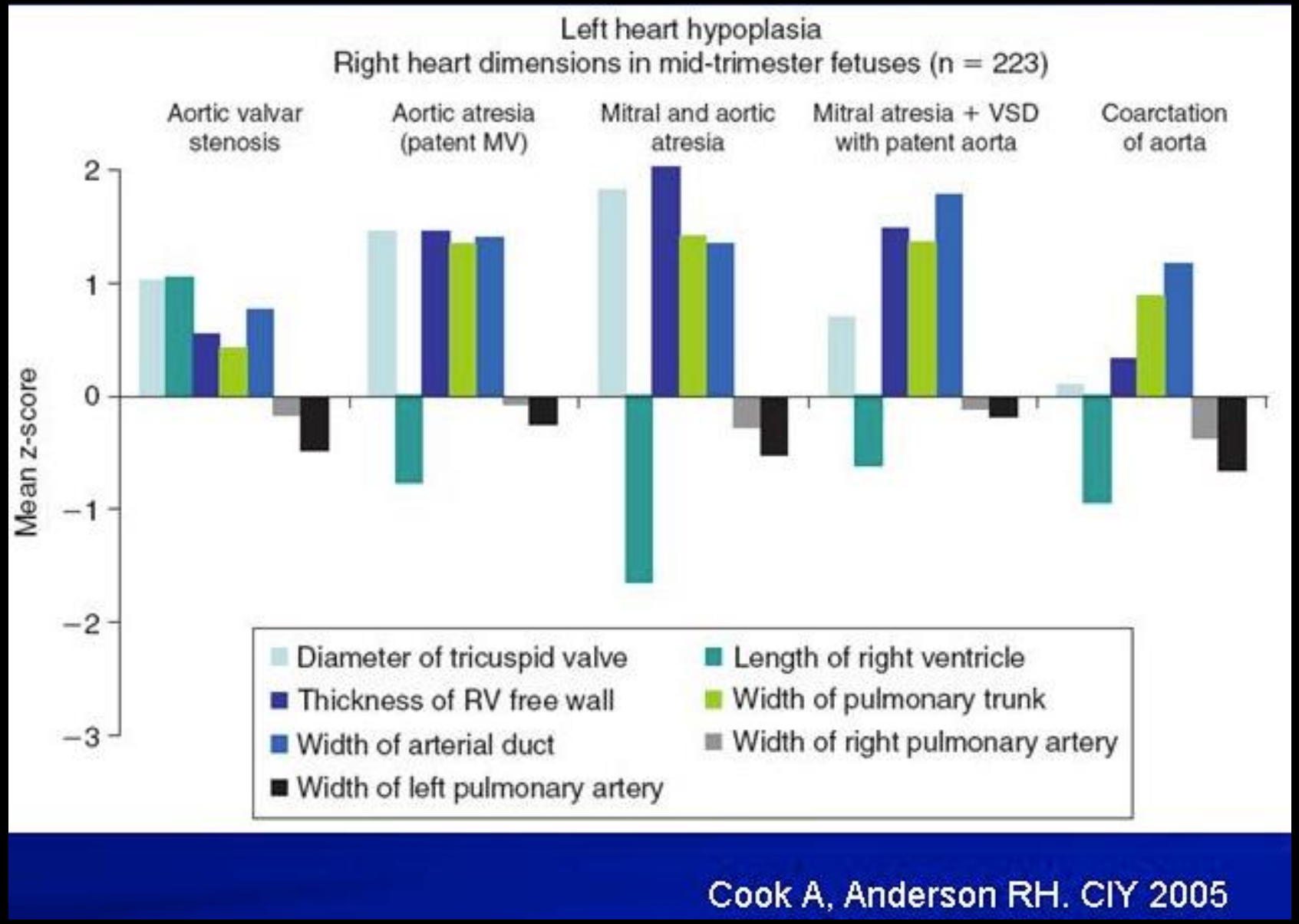


SickKids
The Labatt Family Heart Centre

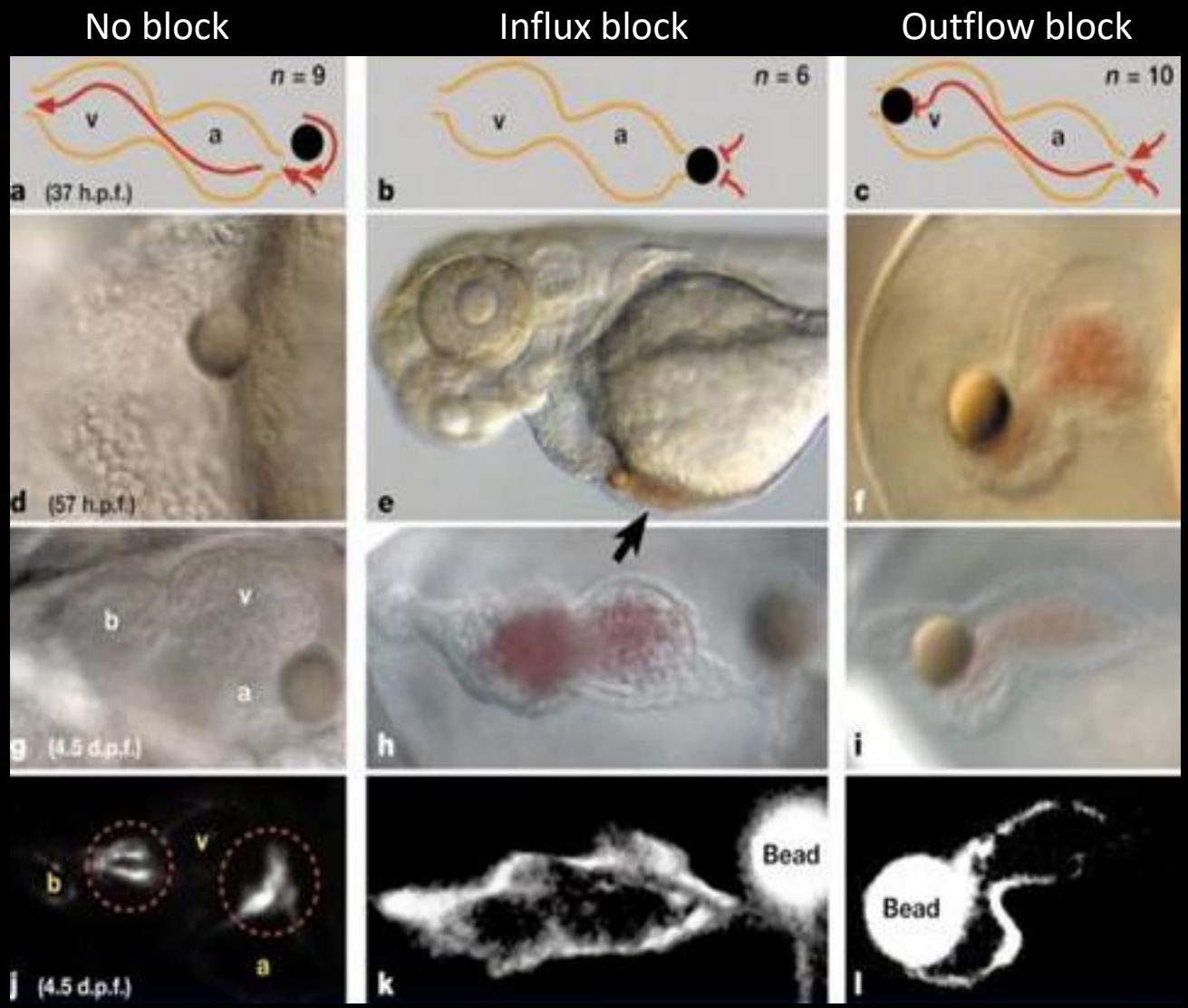
Outcome

- Norwood Sano
- Developed LPA stenosis : LPA dilatation / stent.
- Bilateral Glenn

What about the right side?



HLHS: Physiology & flow dynamics (Hydraulic)& Embryology



Hove JR et al. Nature 2003