Arch obstruction & LVOTO

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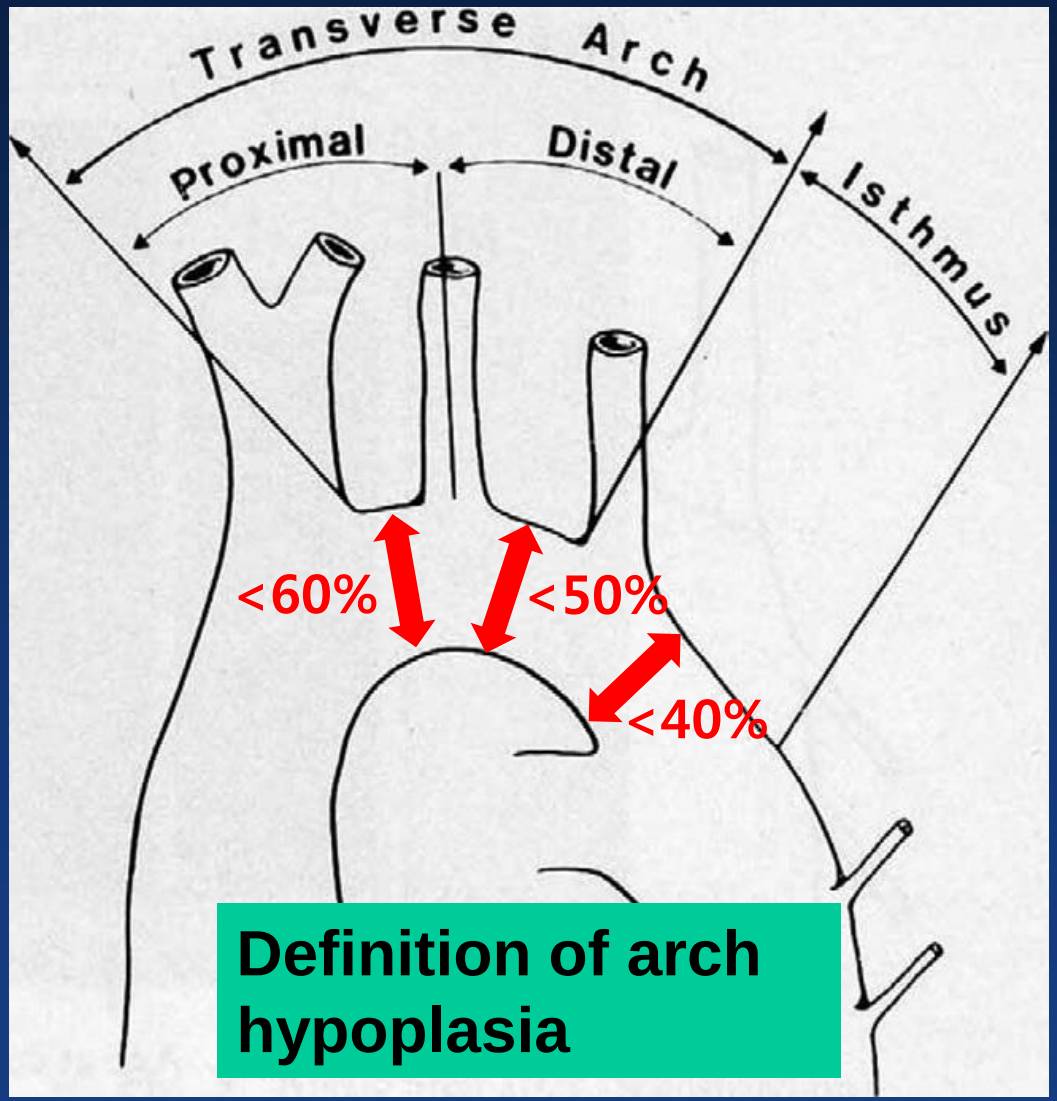
소아심장외과
박 천 수

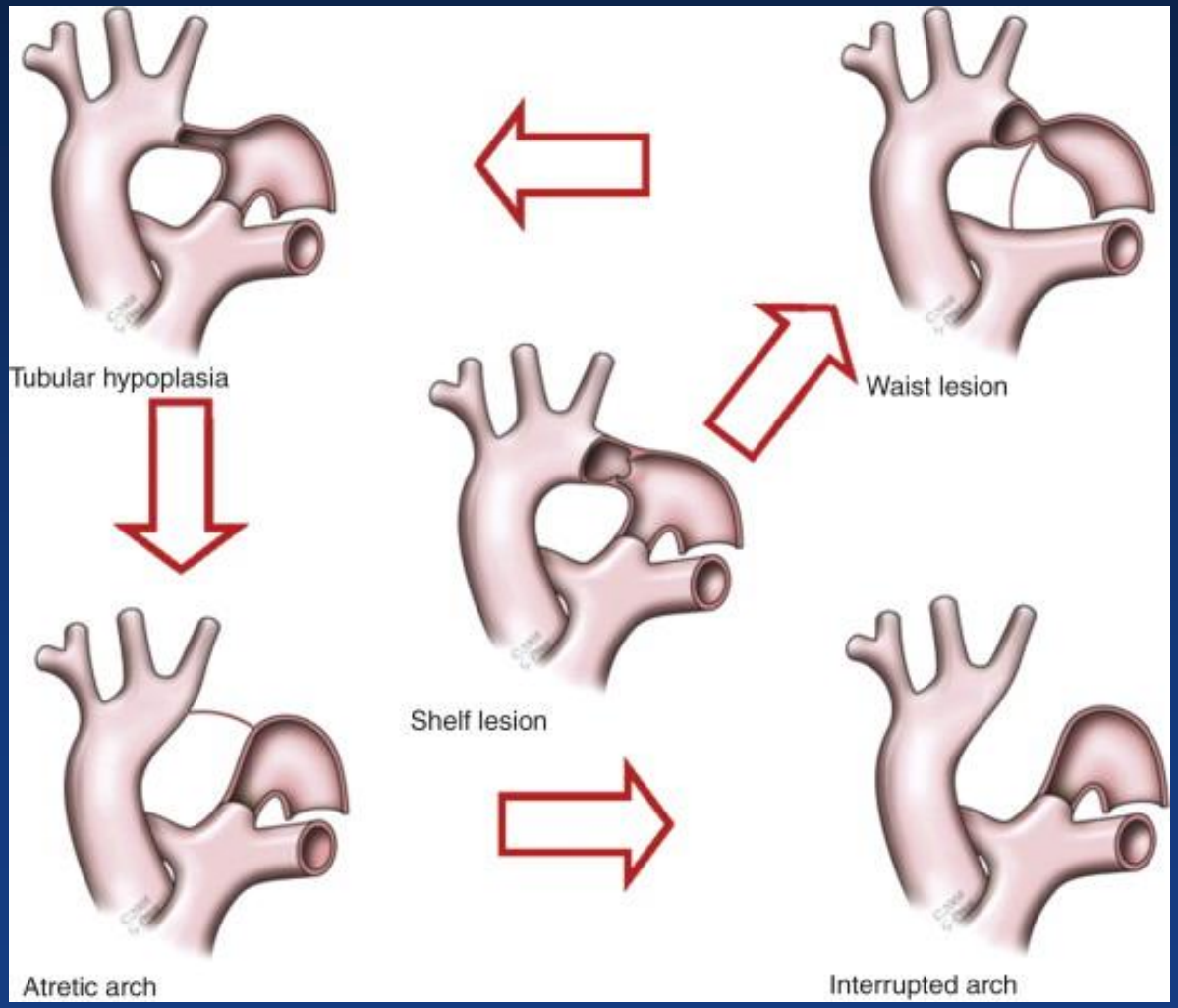
Arch Obstruction

Aortic Arch


Transverse Arch
Proximal
Distal
Isthmus
<60%
<50%
<40%
Definition of arch hypoplasia

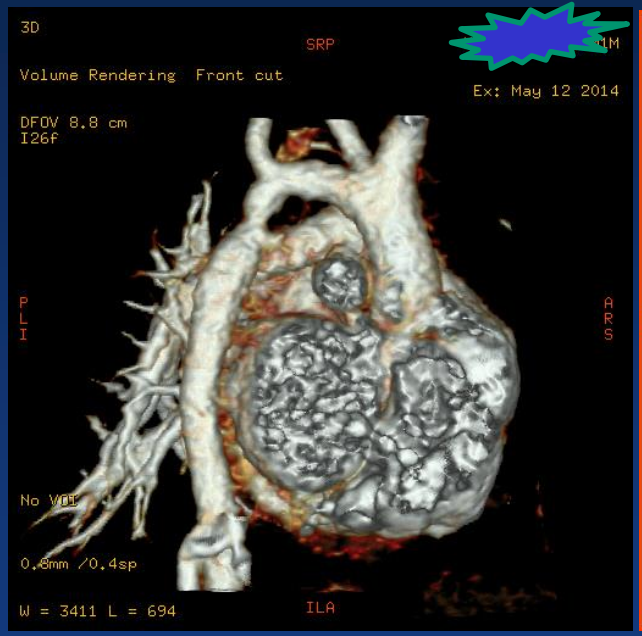
Aortic Arch Obstruction

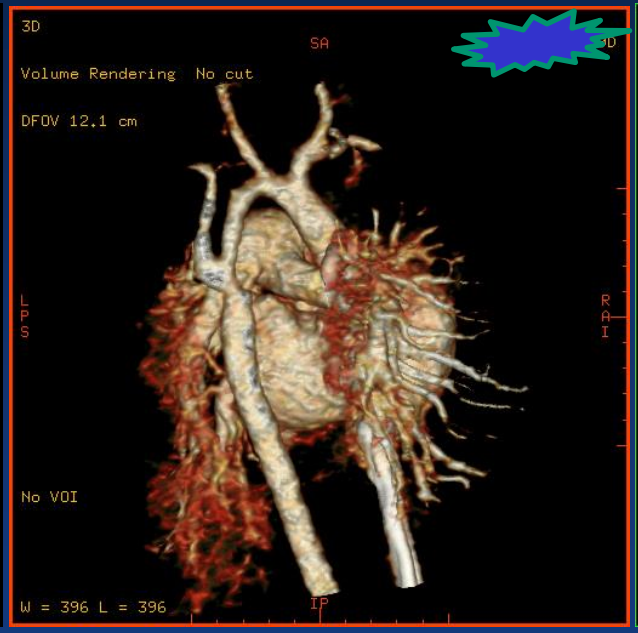
- Morphological Spectrum -

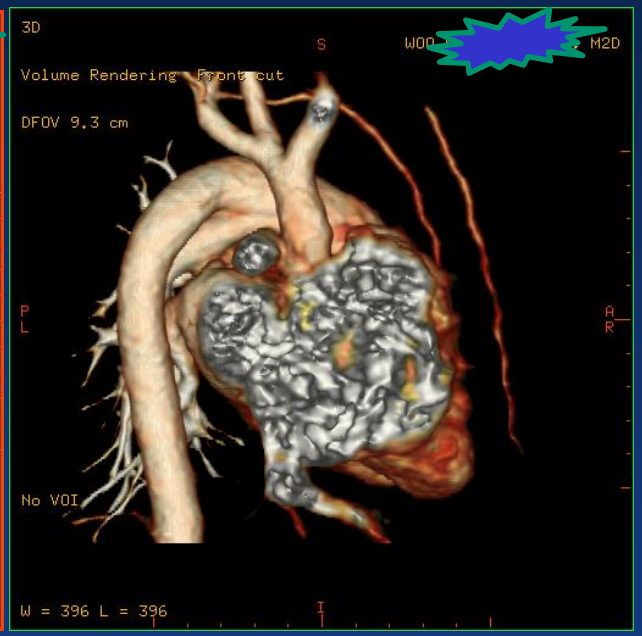


Aortic Arch Obstruction

- Morphological Spectrum -



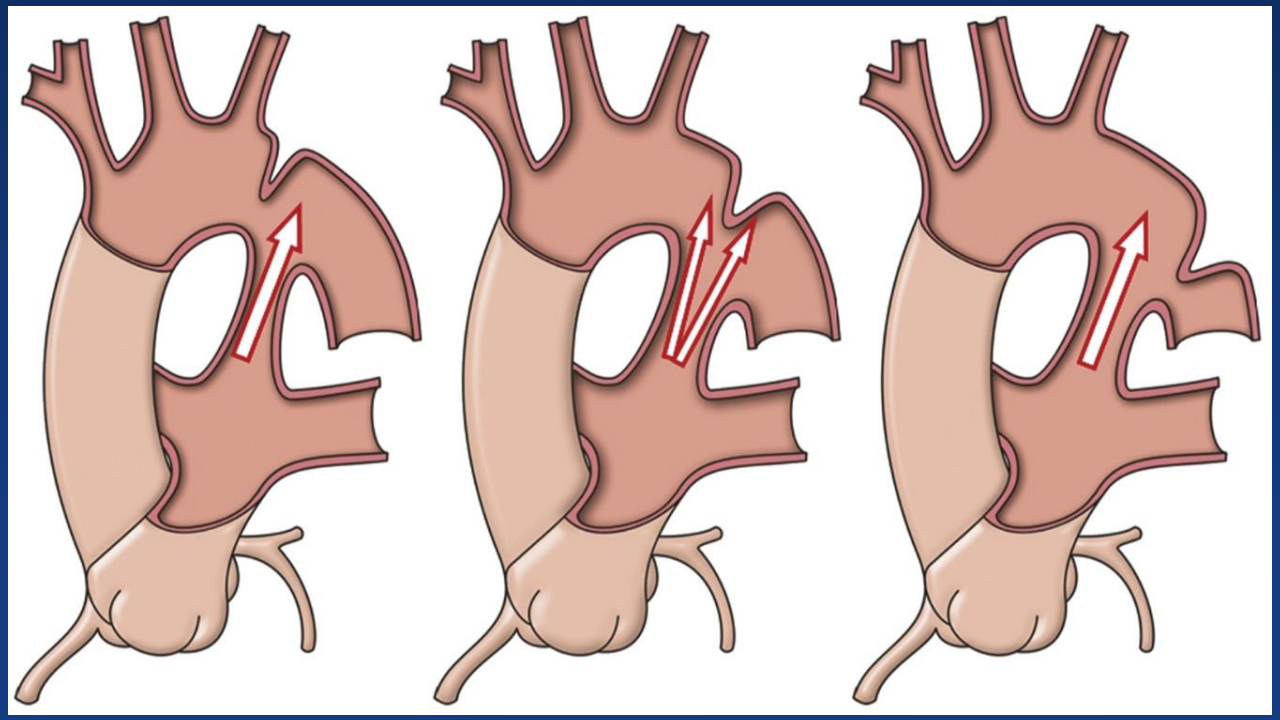




Discrete CoA | **CoA with arch hypoplasia** | **Arch interruption**

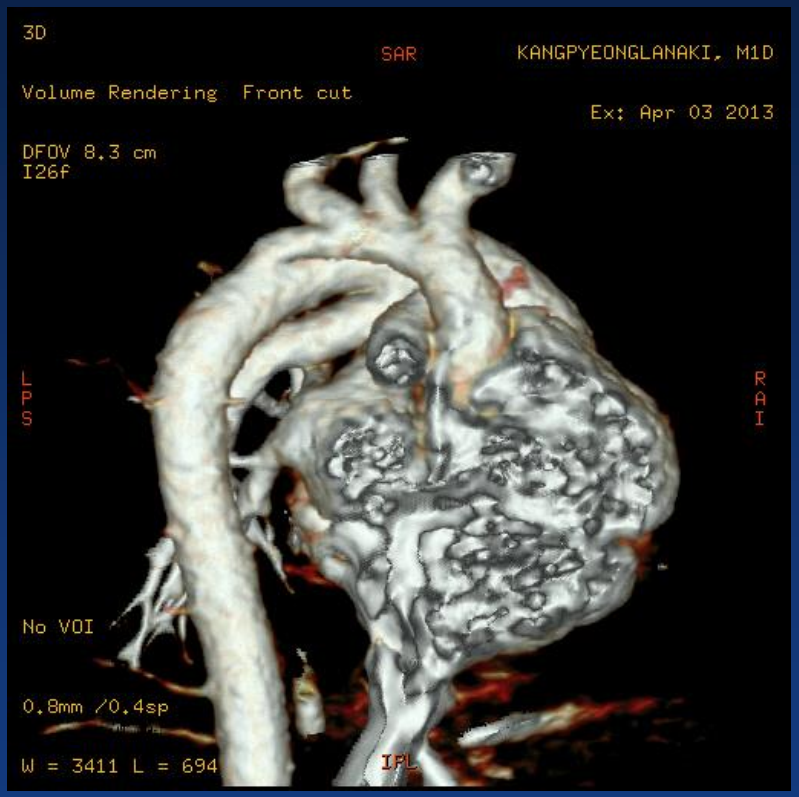
Aortic Arch Obstruction

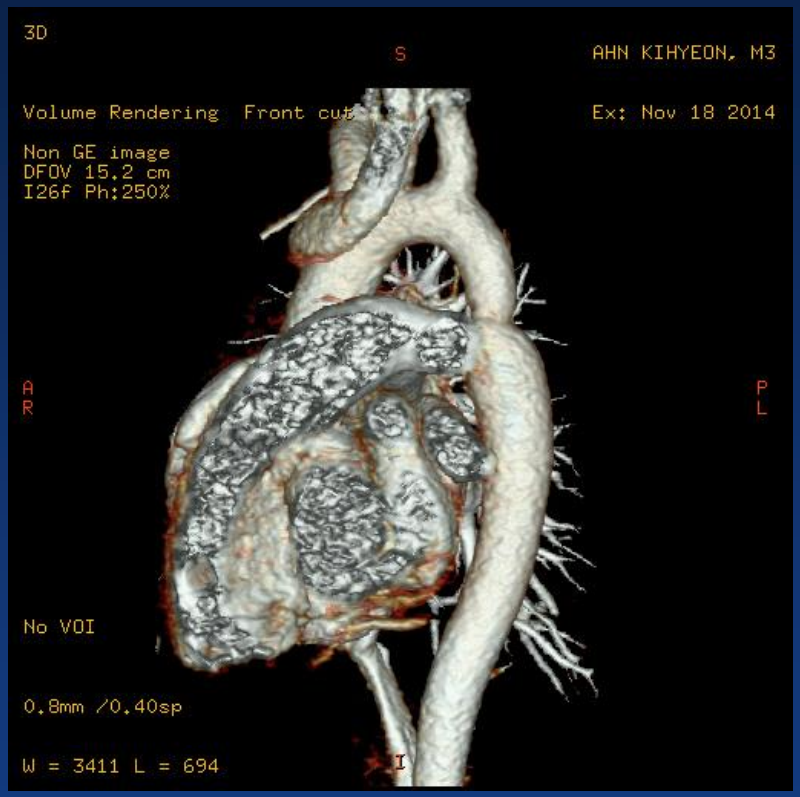
- **Coarctation of Aorta**
  - ✓ **Site of coarctation**
  - ✓ **Concomitant arch hypoplasia**



Aortic Arch Obstruction

- **Coarctation of Aorta**



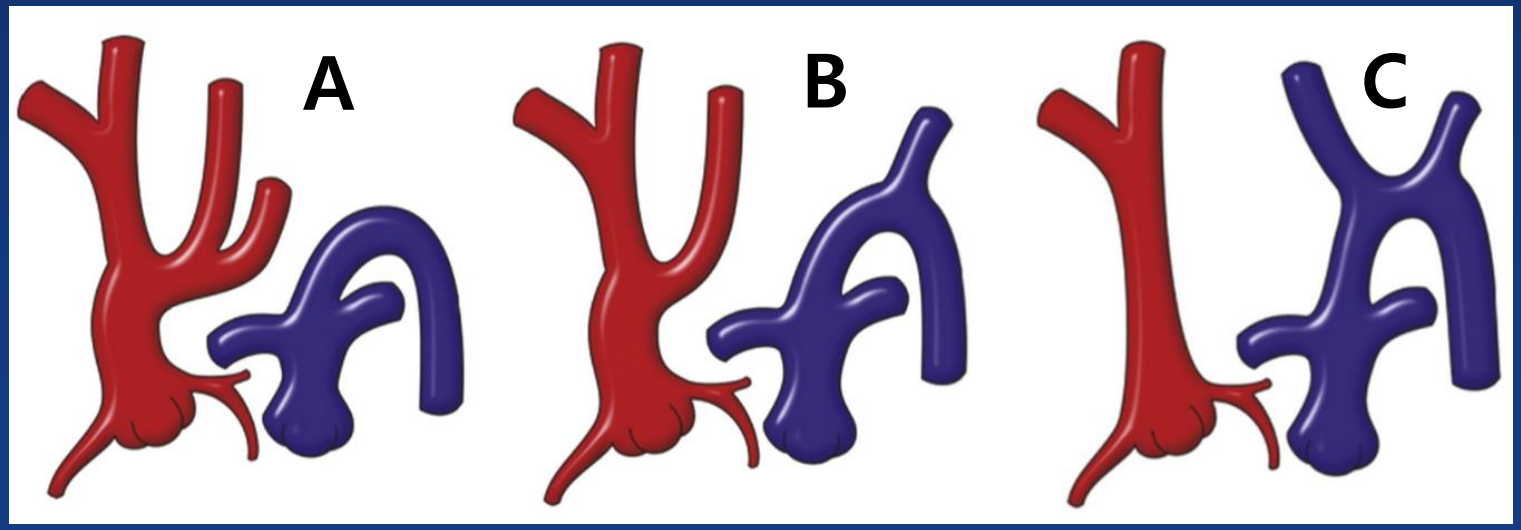


- Preductal
- Arch hypoplasia
- Infantile CoA

- Postductal
- Discrete
- Adult CoA

Aortic Arch Obstruction

- **Interrupted aortic arch**
  - ✓ **Type A : isthmus**
  - ✓ **Type B : distal arch (↑aberrant SCA)**
  - ✓ **Type C : proximal arch**



Presentation

- **Heart failure**
  - **Mostly < 3months of life**
  - **A significant number < 1st week of life**
- **Circulatory collapse**
  - **Ductal closure: ↓ in lower body perfusion**
  - **Falling PVR: preferential flow of blood to the pulmonary circulation**

  → development of acidosis and shock

Initial Management

- **Stabilization**
  - **Maintain ductal patency : Prostaglandin E1**
    - **Maximal response: 15 minutes ~ 4 hours**
    - **Less effective: older or closed duct**

    → Strongly suspected → Start PGE1!!

    → Antenatal transfer to tertiary center
  - **Mechanical ventilator**
    - **Reduce the systemic O2 demand**
    - **Improve heart failure**
    - **Maneuvers to ↑ PVR: ↑ lower body perfusion**

Surgical Correction

- Extent of arch obstruction
- Combined anomaly
  - Simple septal defects: VSD, ASD
  - Complex septal defect: CAVSD
  - Left heart anomaly: LVOTO, AS, MS, LV hypoplasia…
- Arch obstruction + simple septal defect

→ One-stage correction

Surgical Consideration

- Extent of Arch Surgery -

- End-to-End anastomosis
- End-to-Side anastomosis

  *Extended repair: proximal to 1st branch
- Subclavian flap; (reverse flap)
- Prosthetic patch
- Prosthetic interposition graft

Surgical Consideration

- DHCA vs. regional perfusion -

	DHCA	SCP
Advantage	• Clear operative field • More accurate correction especially in small babies • ↓ exposure to CPB	• Potentially neuroprotective
Disadvantage	• Poor neurodevelopmental outcome → Safe duration of circulatory arrest :???	• Crowded operative field • Technically demanding • Lack of randomized trial

Surgical Consideration

- DHCA vs. regional perfusion -

	DHCA	SCP
Advantage	• Clear operative field • More accurate correction especially in small babies • ↓ exposure to CPB	• Optimal perfusion flow: ? • Optimal perfusion pressure: ? • Is it really neuroprotective?
Disadvantage	• Poor neurodevelopmental outcome → Safe duration of circulatory arrest :???	• Crowded operative field • Technically demanding • Lack of randomized trial

Surgical Consideration

- DHCA vs. regional perfusion -

Perfusion

No perfusion

Surgical Consideration

- DHCA vs. regional perfusion -

Perfusion is better !

Surgical Consideration

- Cerebral Protection -

- Selective cerebral perfusion
  - Our standard
  - Monitoring
    - Blood pressure
    - Flow rate : 50~70ml/kg/min (30~ 50%)
    - Near-infrared spectroscopy
    - Lactic acid

Surgical Consideration

- Cerebral Protection -

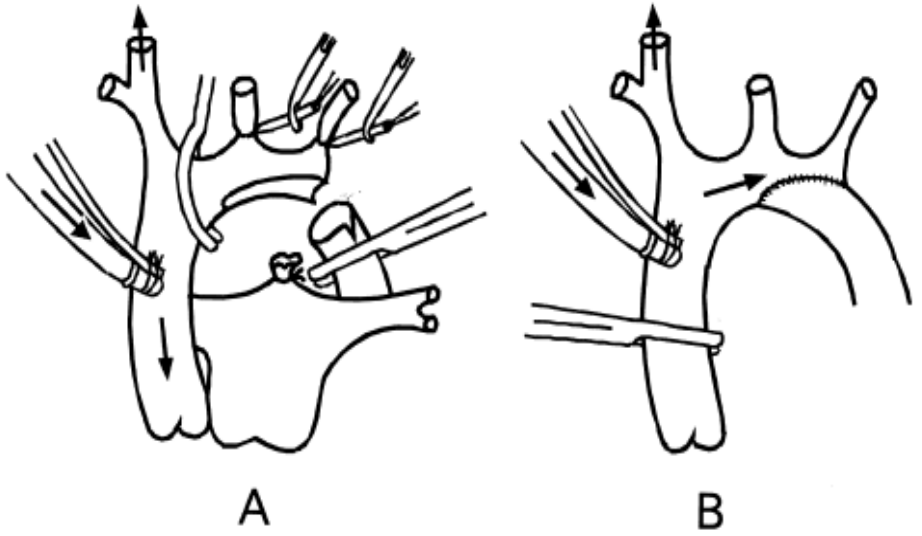


Fig. 2. Isolated cerebral and myocardial perfusion are established by clamping the aortic arch between the innominate artery and left carotid artery. (A) Coarctation repair is carried out with brain perfusion and the heart beating. (B) The clamp is repositioned onto the ascending aorta, and the VSD is closed with cardioplegic arrest.

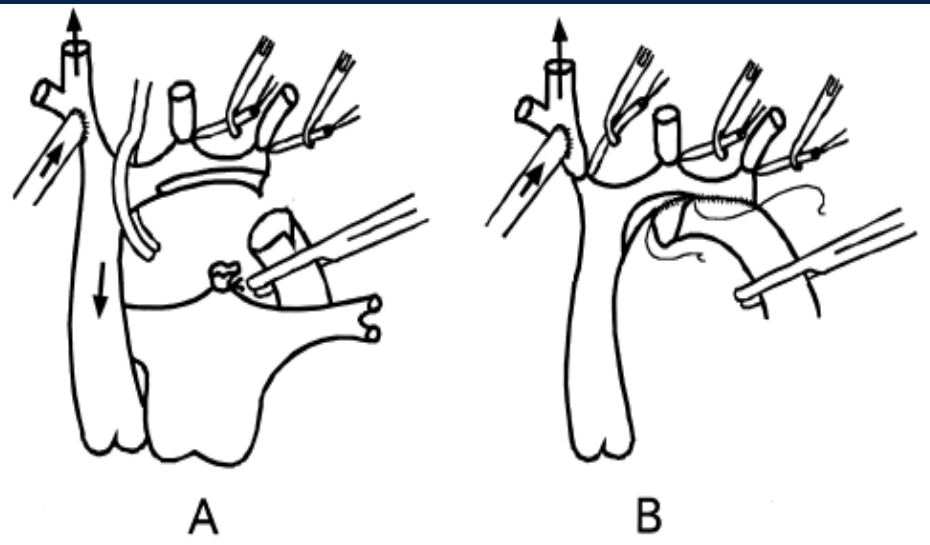


Fig. 3. (A) In a case of coarctation plus hypoplastic arch, about two-thirds of the arch anastomosis is accomplished with isolated cerebral and myocardial perfusion. (B) The innominate artery just proximal to the PTFE tube is snared, the arch is unclamped, and the arch anastomosis is extended with isolated cerebral perfusion.

Ishino K et al. Eur J Cardiothorac Surg 2000;17:538-42.

Surgical Consideration

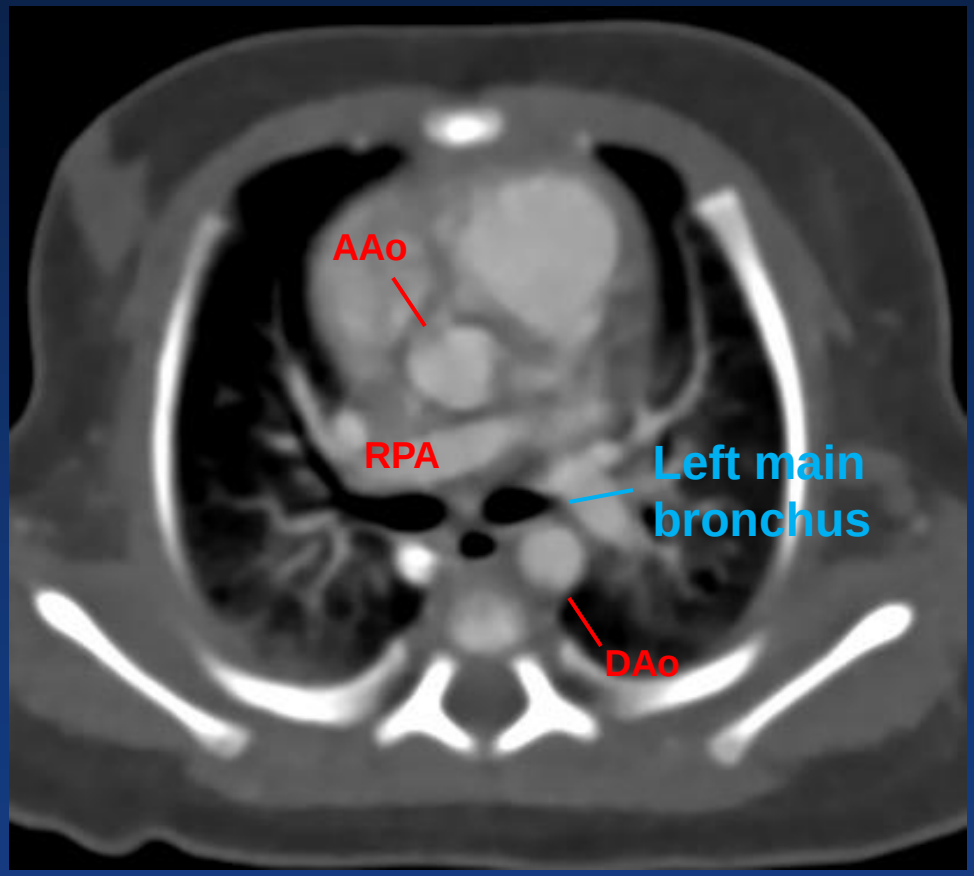
- Selective cerebral perfusion
- Perfusion is better !

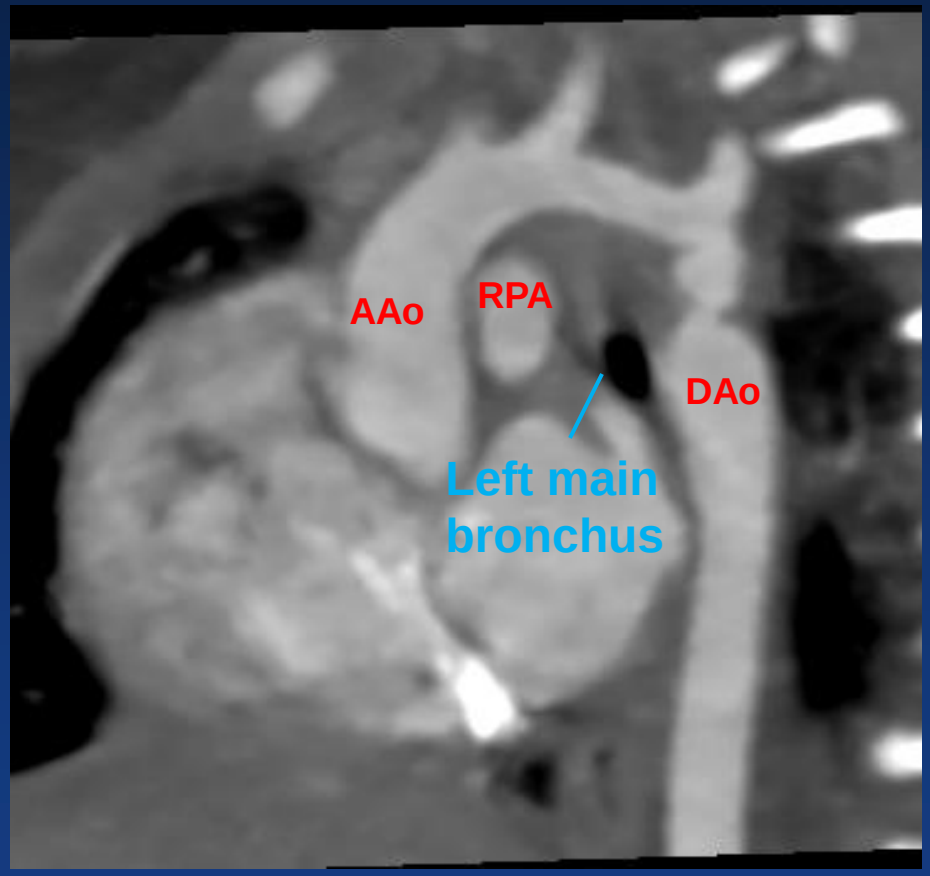
+ Selective myocardial perfusion

→ Selective C & M perfusion

Surgical Consideration

- Airway -





Left main bronchus compression!!

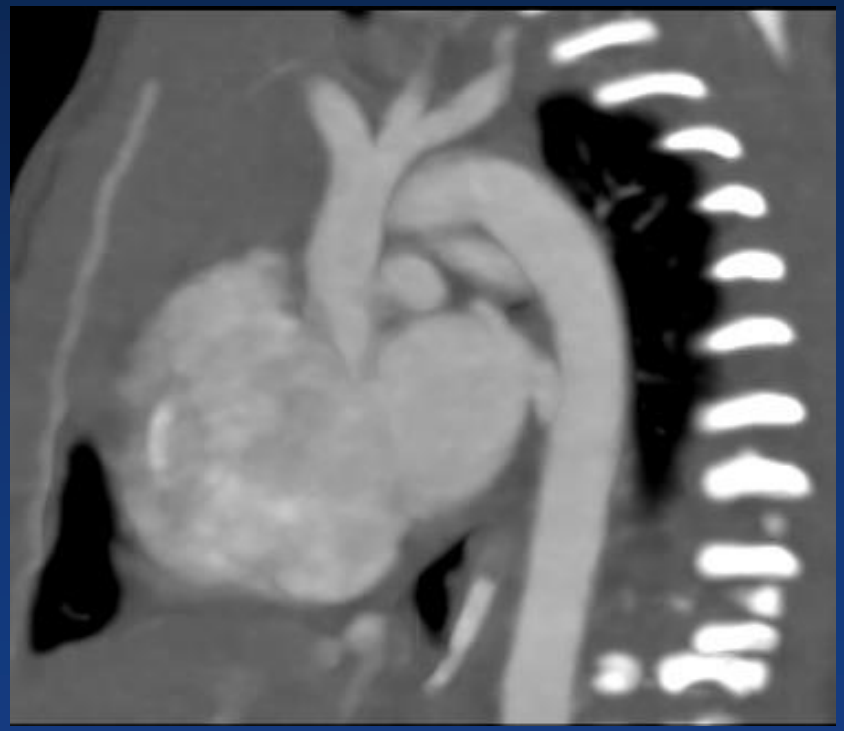
Surgical Consideration

- Airway -

- **To avoid airway problem**
  - ✓ **extensive dissection of arch vessels and descending aorta**
  - ✓ **arch repair using autologous MPA patch**
  - ✓ **RPA anterior translocation**

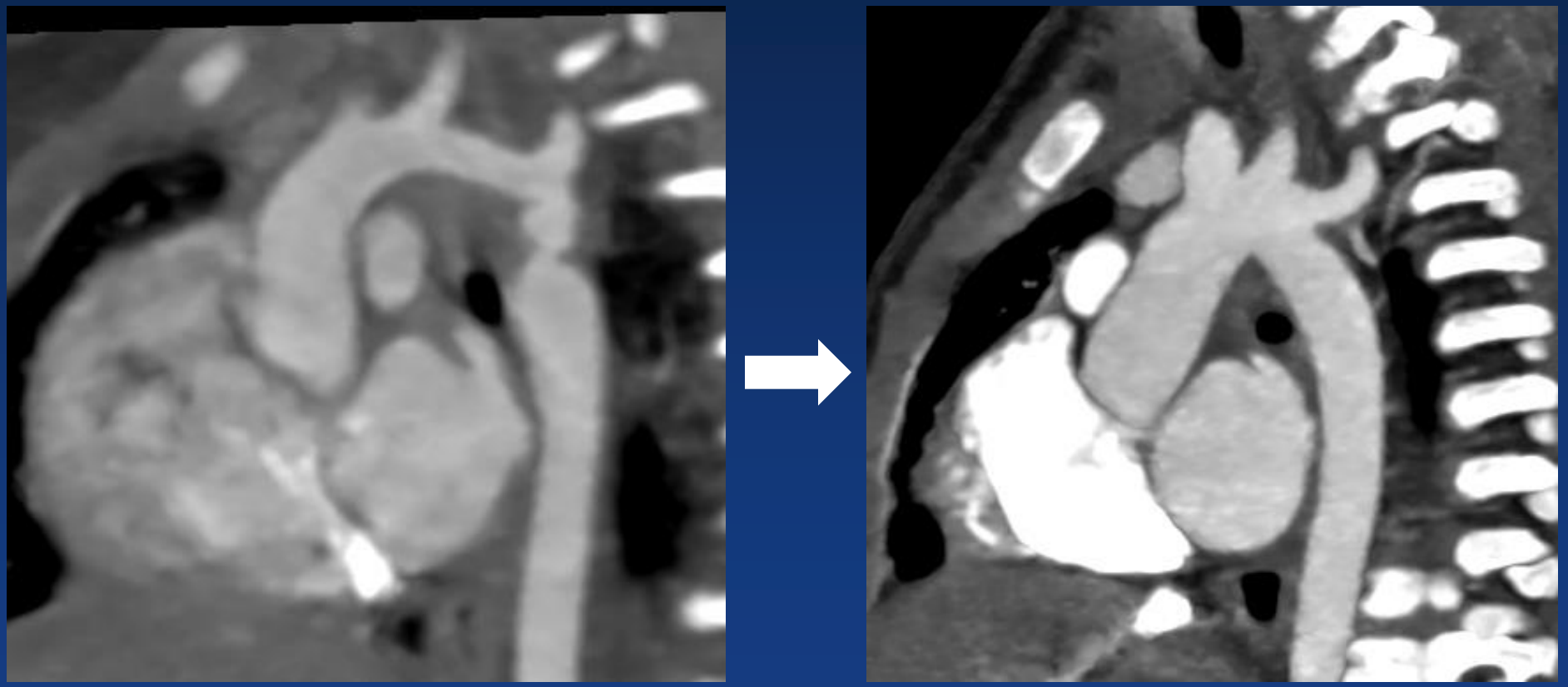
Surgical Consideration

- Arch repair using autologous MPA patch -



Surgical Consideration

- RPA anterior translocation -



Case 1

- Discrete CoA -

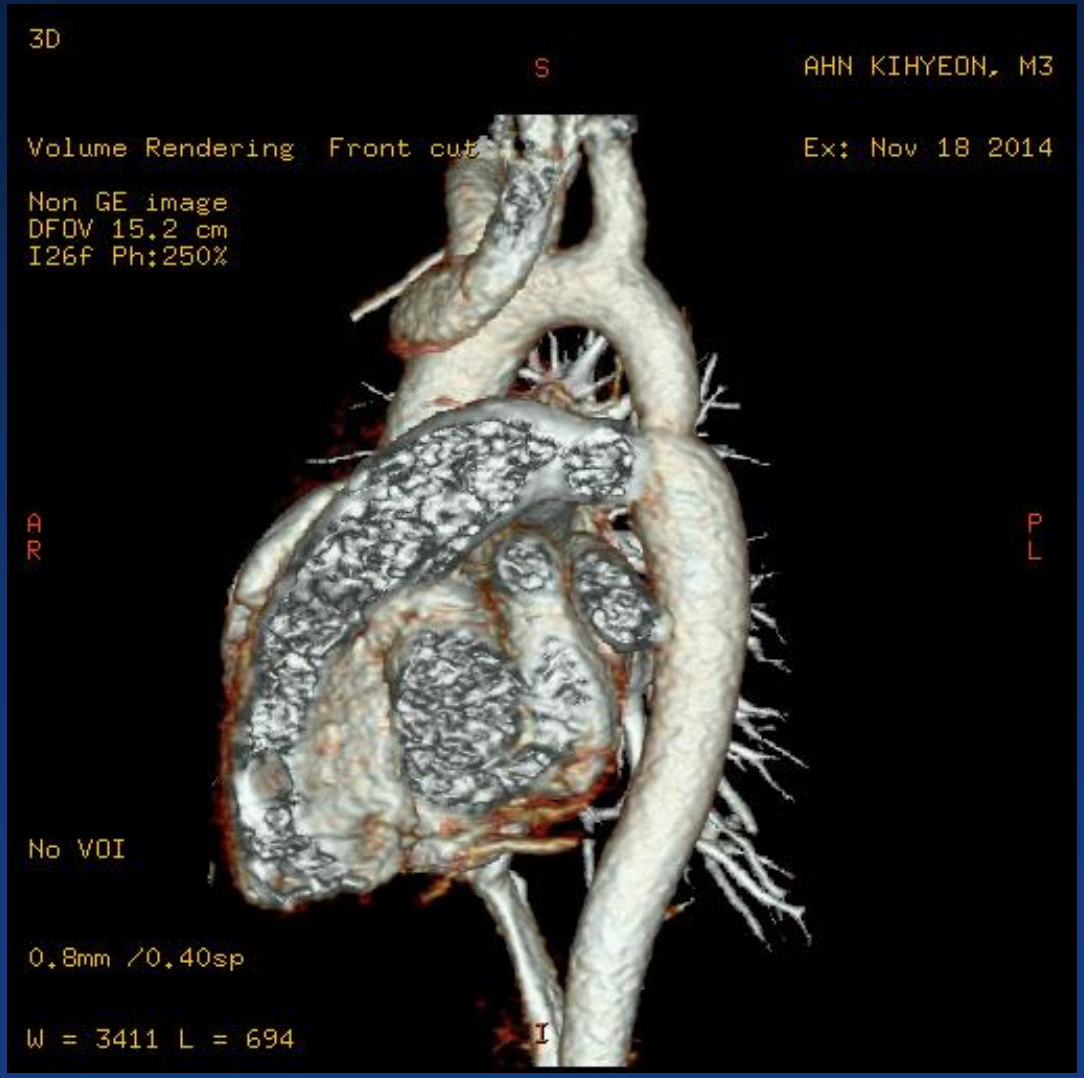
- M/ 4years
- Asymptomatic
- Murmur
- Preop. cuff blood pressure
  - ✓ UE: 105/65, LE: 97/66, PG 8
- EchoCG/ Heart CT

Case 1

- Discrete CoA -

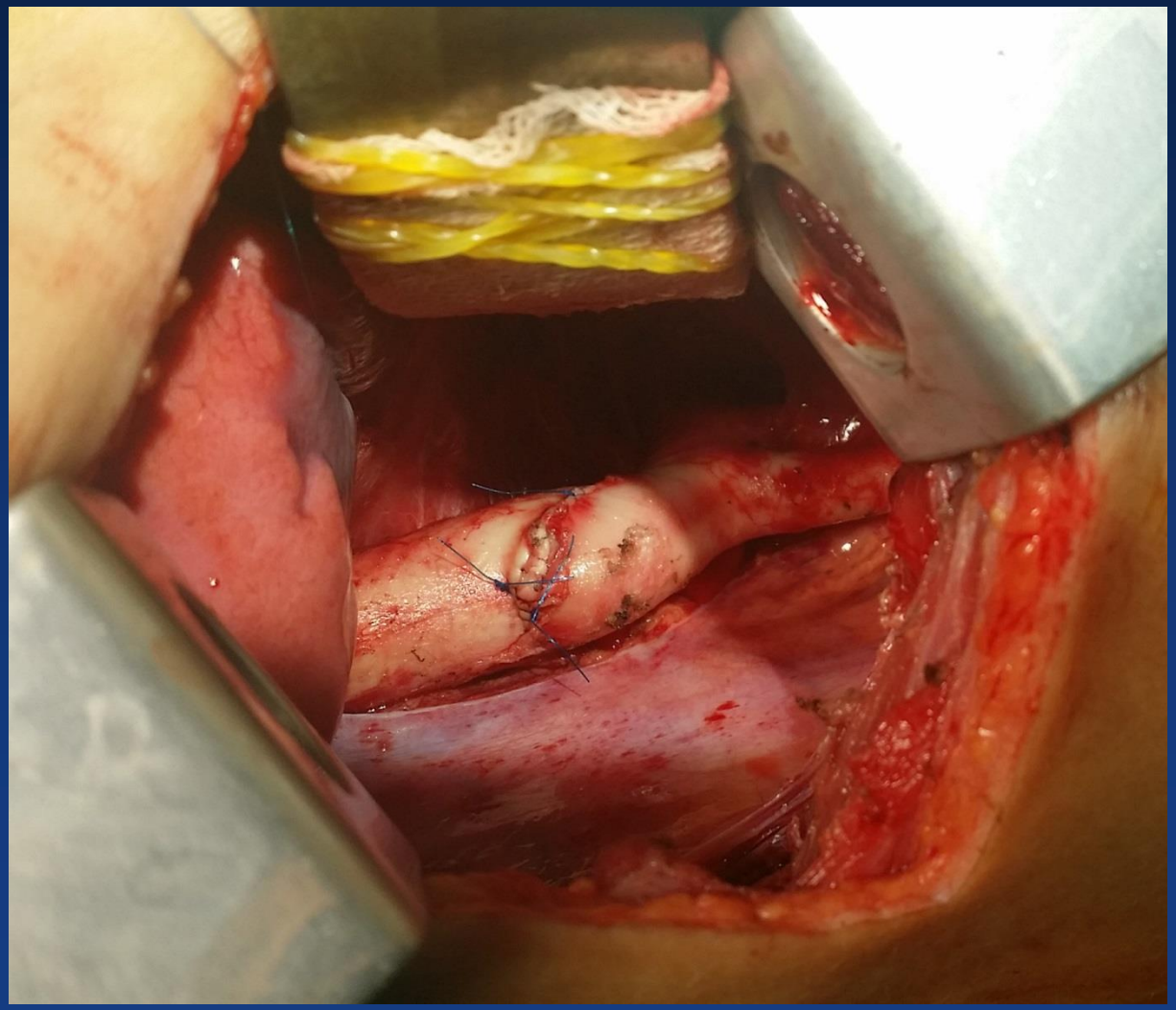
Case 1

- Discrete CoA -



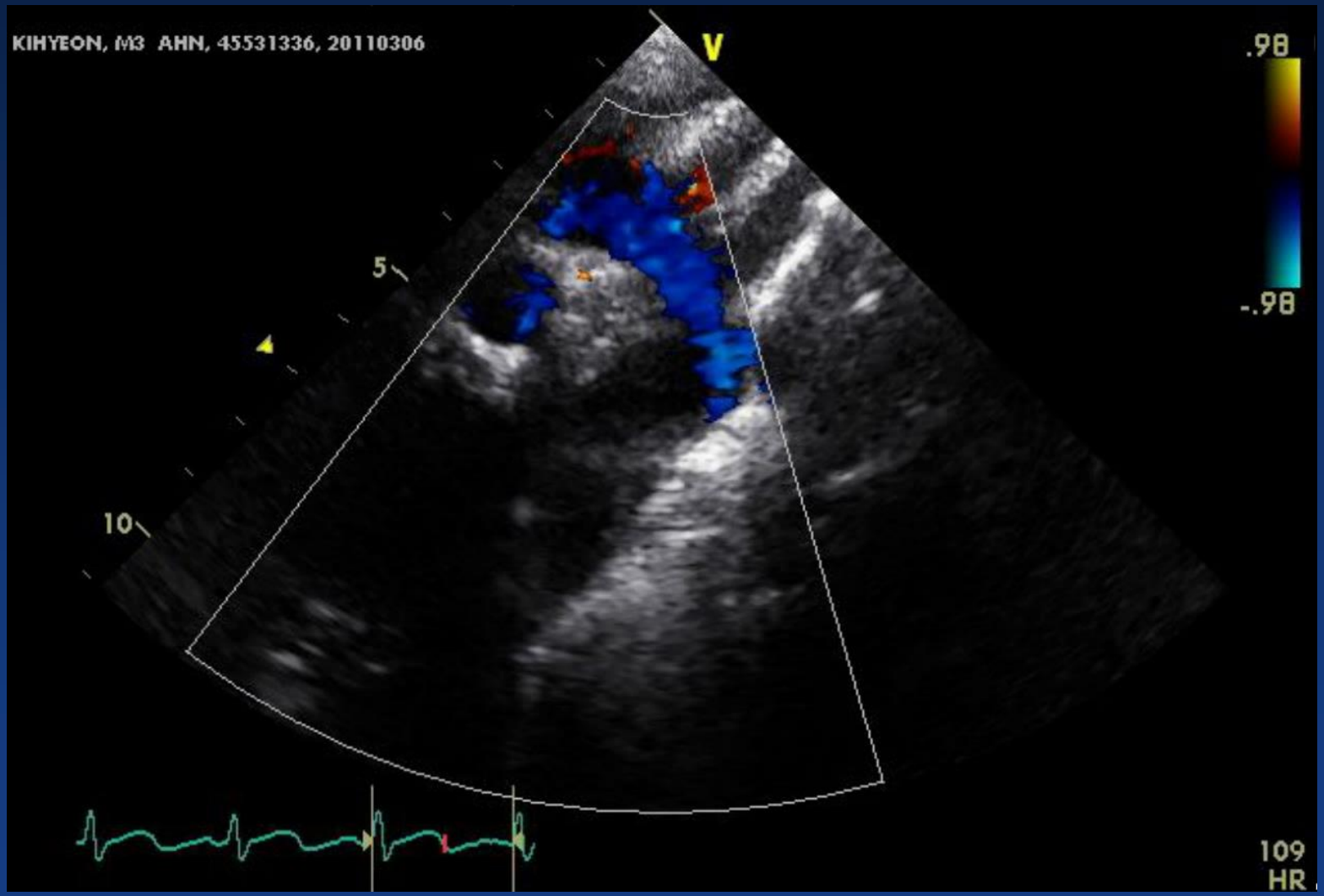
Case 1

- Discrete CoA -



Case 1

- Discrete CoA -



Case 2

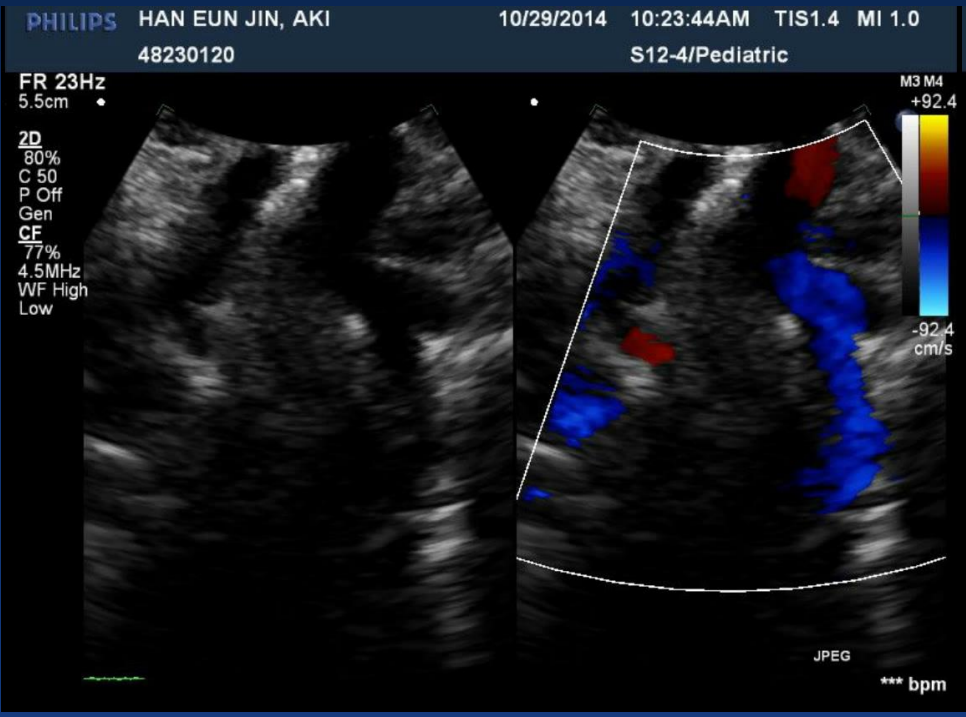
- CoA with Arch Hypoplasia -

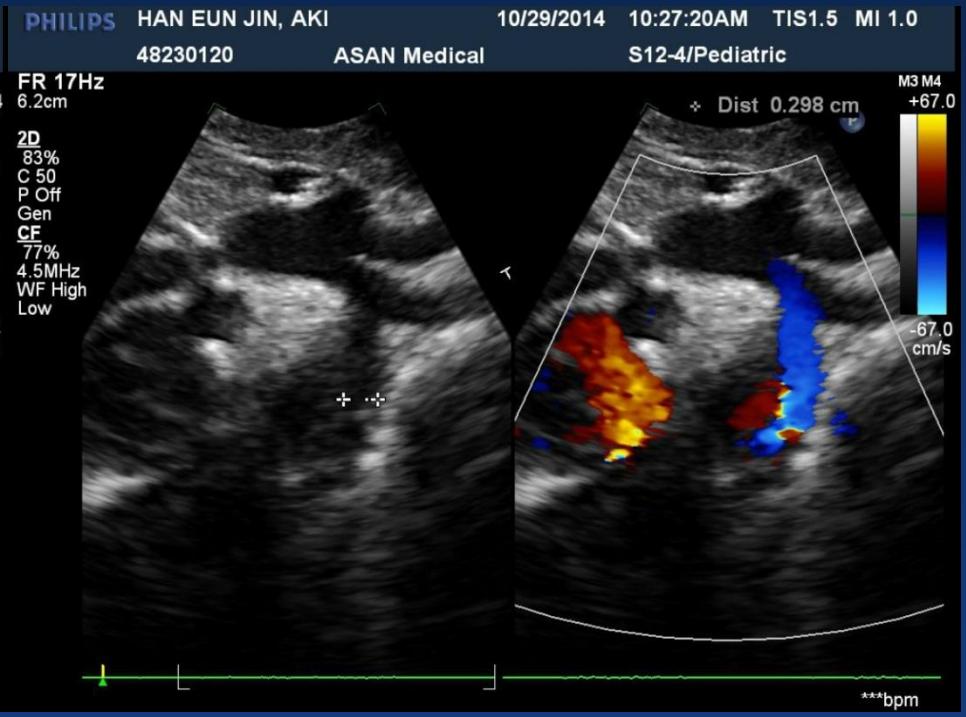
- Prenatal Dx. (+)
- GA 39+2wks, birth weight 3590 gm

  → Admission to NICU
- E-glandin 0.004mcg/kg/min
- Mechanical ventilator (-)
- BNP 1724

Case 2

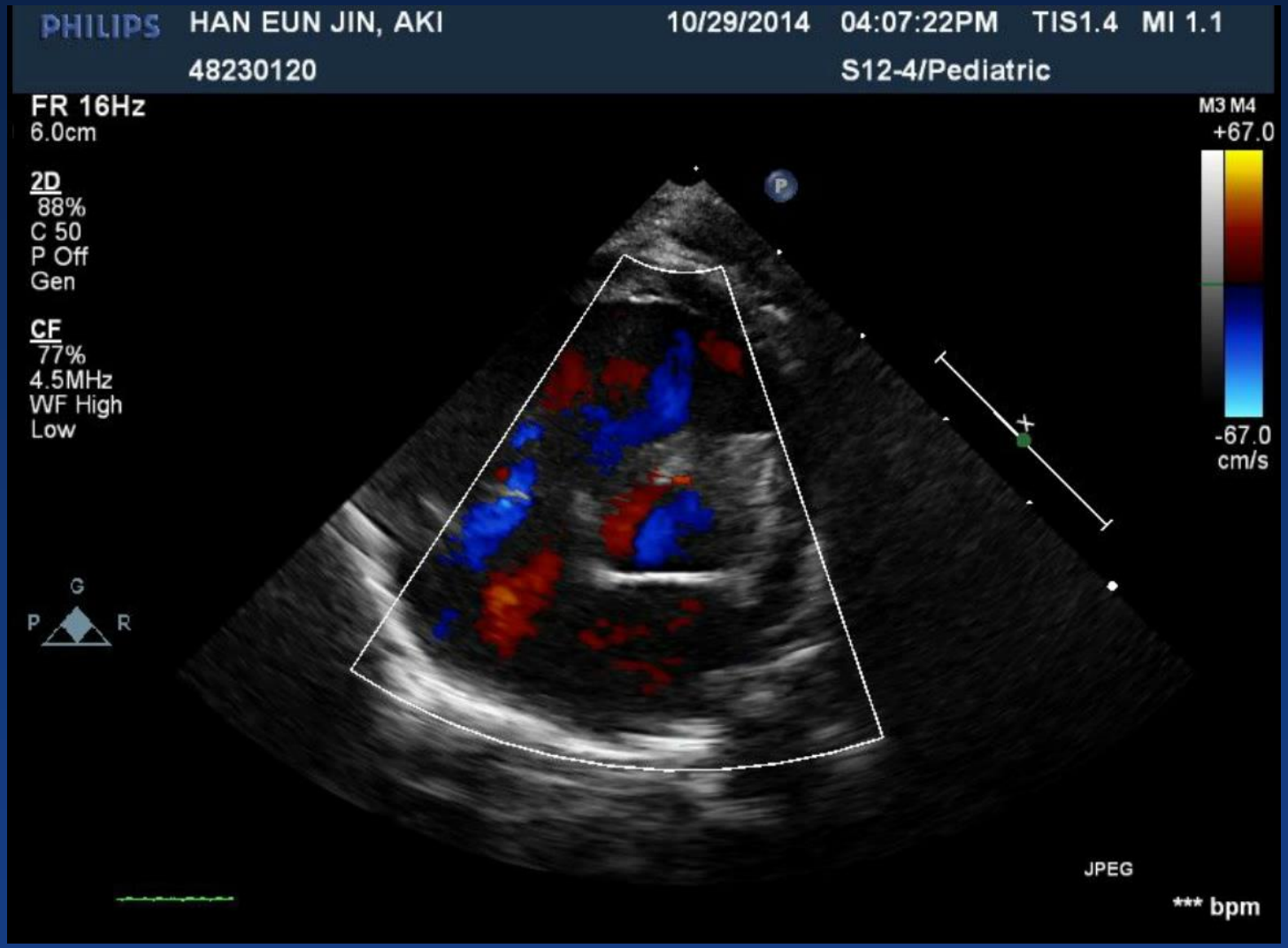
- CoA with Arch Hypoplasia -





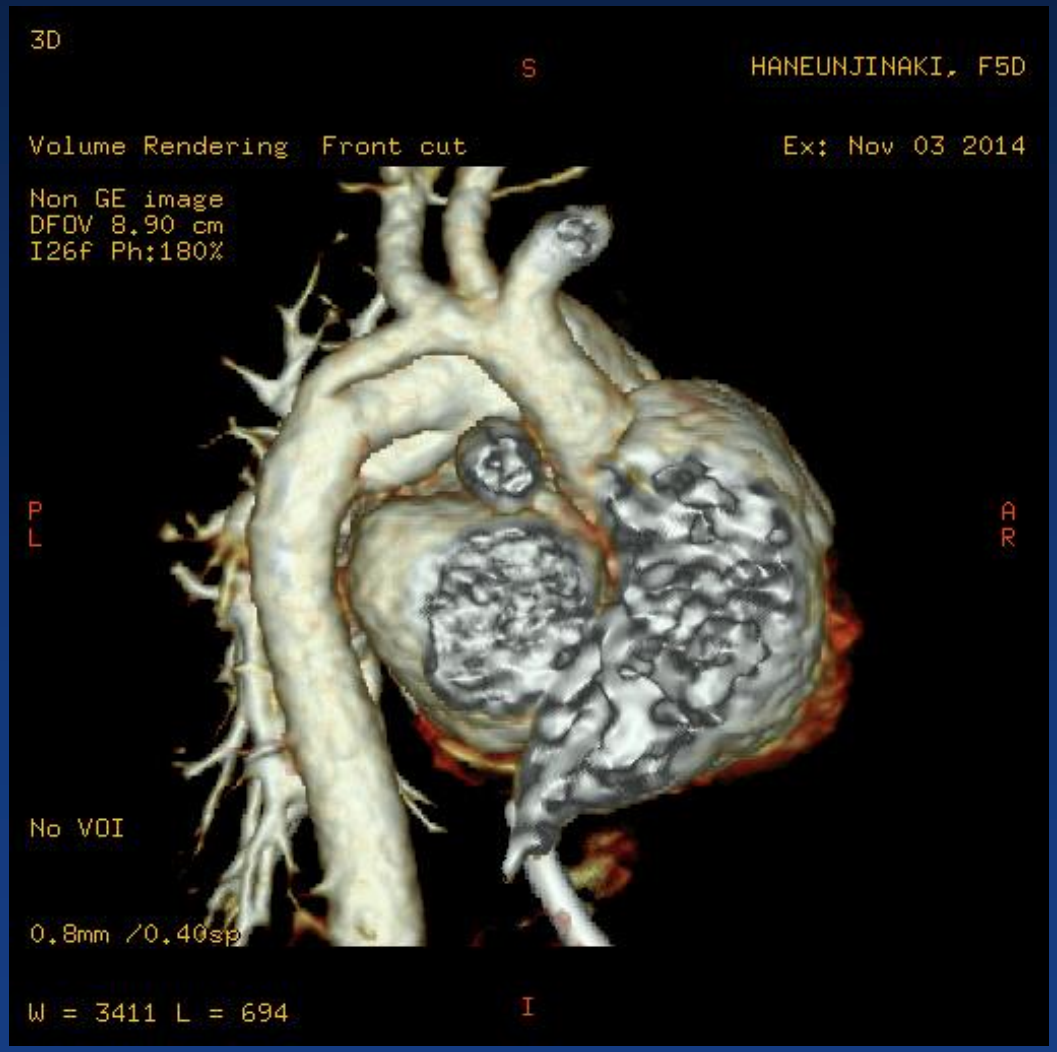
Case 2

- CoA with Arch Hypoplasia -



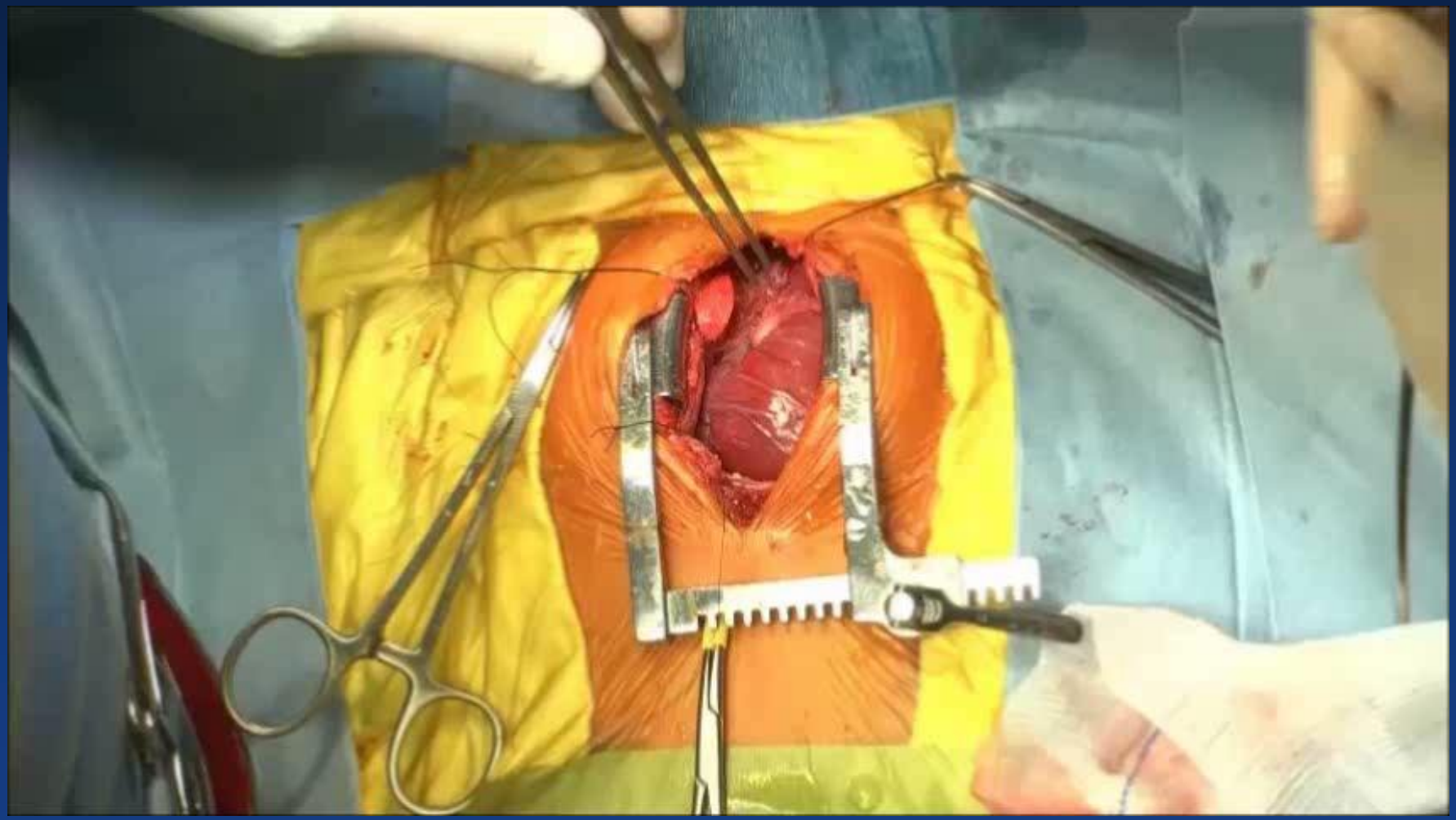
Case 2

- CoA with Arch Hypoplasia -



Case 2

- 14 days, 3.58 kg -

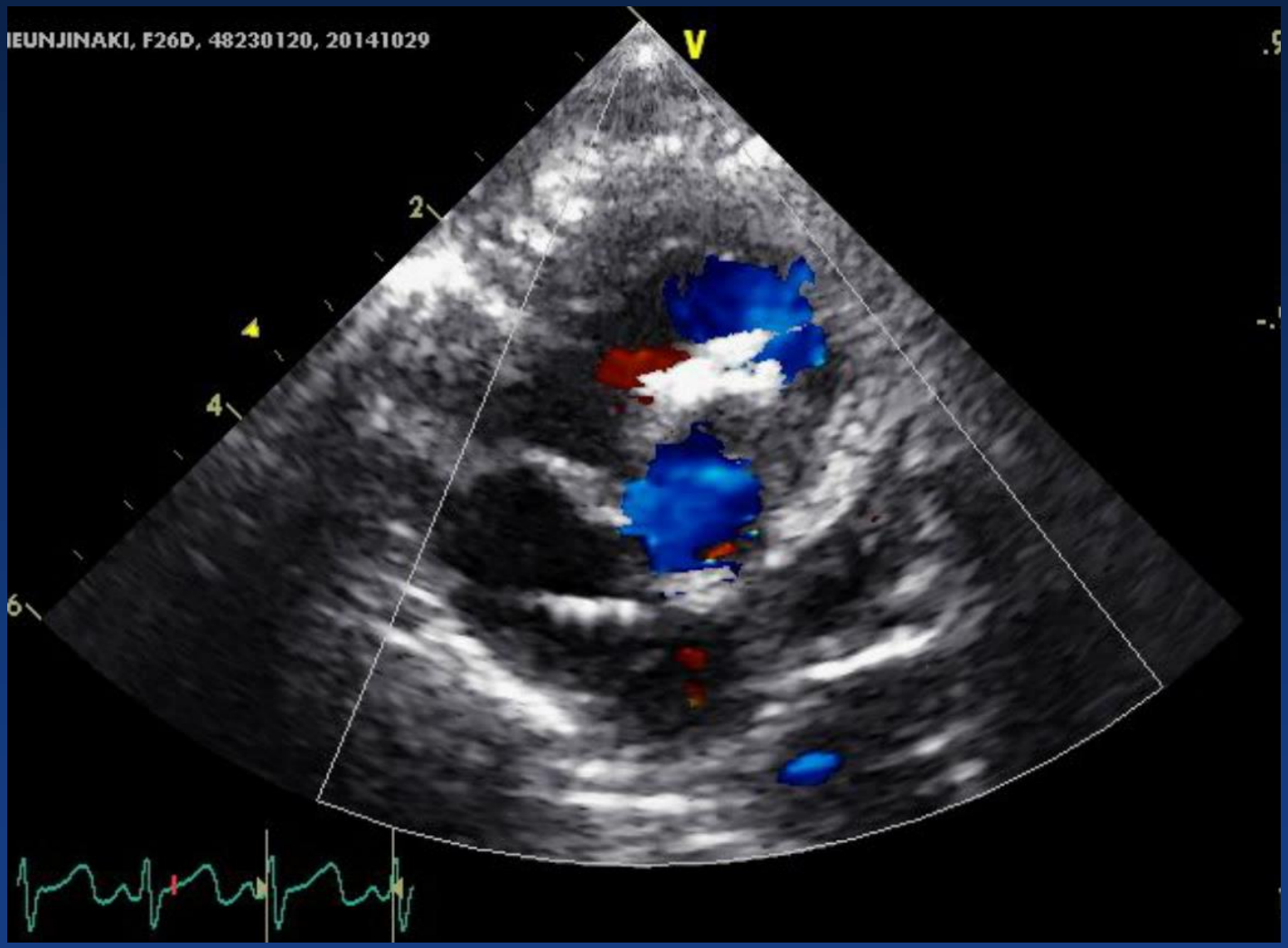


Case 2

- CoA with Arch Hypoplasia -

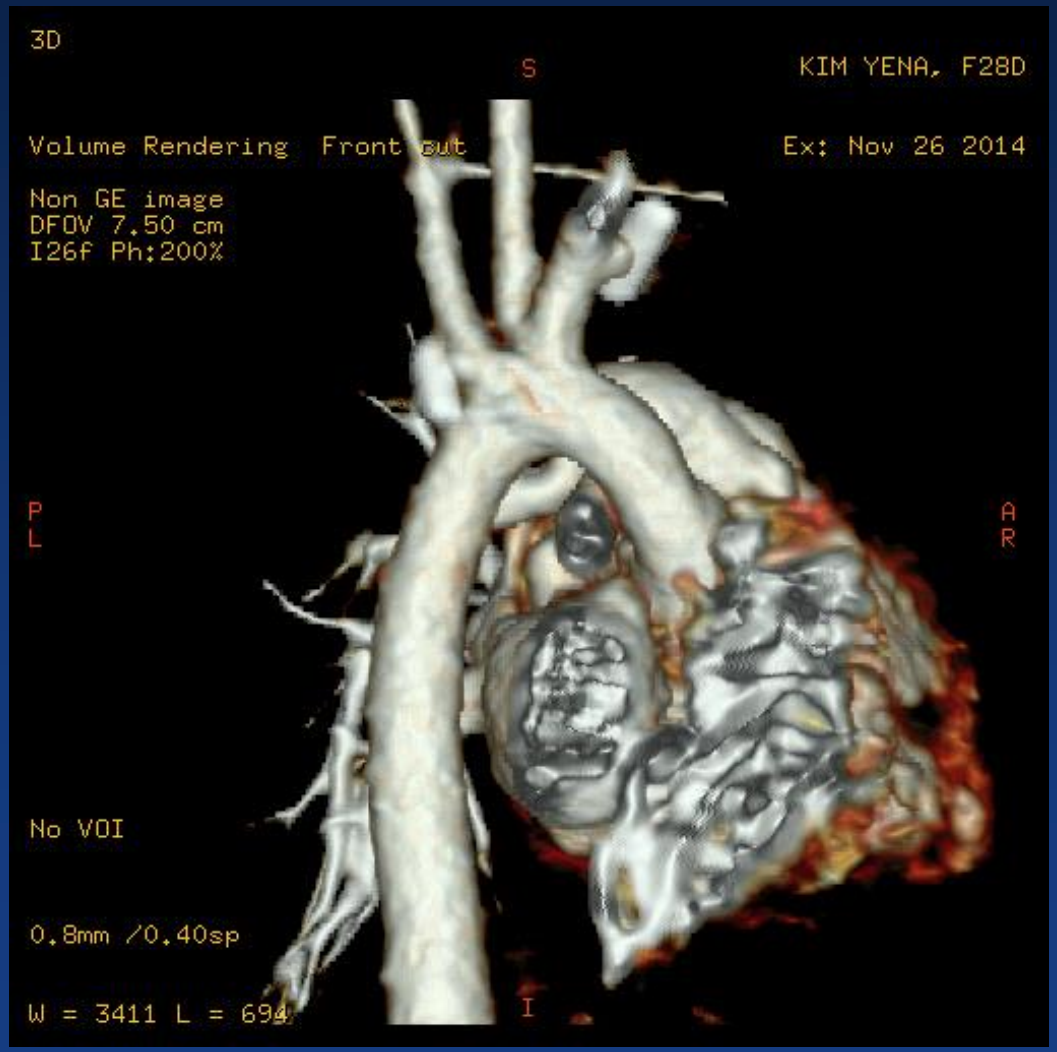
Case 2

- CoA with Arch Hypoplasia -



Case 2

- CoA with Arch Hypoplasia -



Complications

- **Paradoxical hypertension**
  - **Correction of CoA (cause of HTN) → Hypertension ??**
  - **Postcoarctectomy syndrome**

    : sudden ↑ of blood pressure → mesenteric arteritis → ischemia

    → Strict BP control, NPO for 1~2days

Complications

- **Paraplegia**
  - **Related to**
    - ✓ **Prolonged distal clamp time**
    - ✓ **Intraoperative hyperthermia**
    - ✓ **Hypotension/ Acidosis**
  - **To avoid**
    - ✓ **Reducing clamp time (fast and perfect)**
    - ✓ **Local hypothermia**
    - ✓ **Hypertension**

Complications

- **Recoarctation**
  - **Can be avoided by**
    - ✓ **Using native tissue**
    - ✓ **Low tension anastomosis**
    - ✓ **Wide anastomosis**
    - ✓ **Completely removing ductal tissue**

LVOTO

LVOTO

- Definition -

- **Obstruction at various levels between LV and ascending aorta**

Congenital Heart Surgery Database and Nomenclature Project: Aortic Valve Disease

(Ann Thorac Surg 2000;69:S118–31)

AS, NOS
AS, Valvar
AS, Supravalvar
AS, Subvalvar

LVOTO

- Valvar
- Subvalvar
  - ➢ Fixed subvalvar
    - ✓ Discrete
    - ✓ Diffuse (tunnel)
  - ➢ Hypertrophic CMP
- Supravalvar

Related to genetic disorder

LVOTO

- Valvar -

- **Valvar AS**
  - **Incidence: 3~6% (~70% of LVOTO)**
  - **Male : female = 3~5 : 1**
  - **Associated anomalies**
    - ✓ **VSD**
    - ✓ **PDA**
    - ✓ **CoA**
    - ✓ **LV hypoplasia : 2V vs. 1V**

LVOTO

- Valvar -

- **Valvar AS**
  - ➢ **a wide spectrum**

Mild AS

± bicuspid AoV

2V

1V

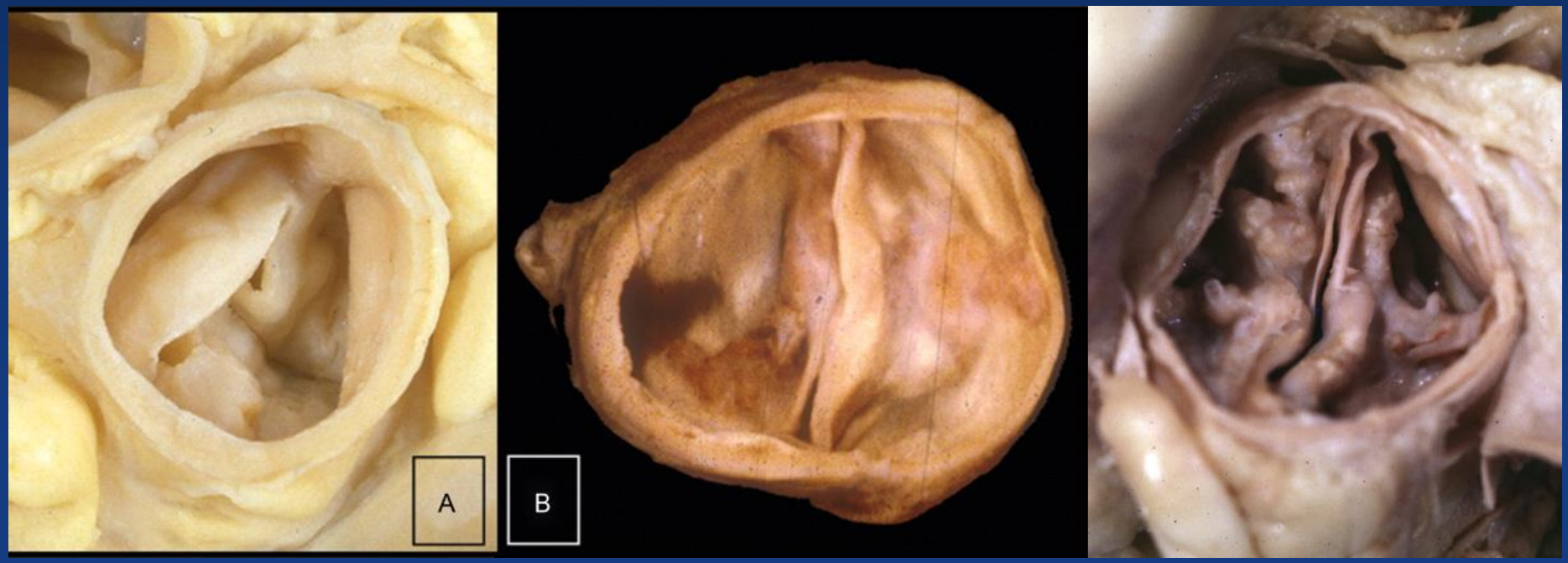
Critical AS

Ductus dependent systemic circulation

LVOTO

- Valvar -

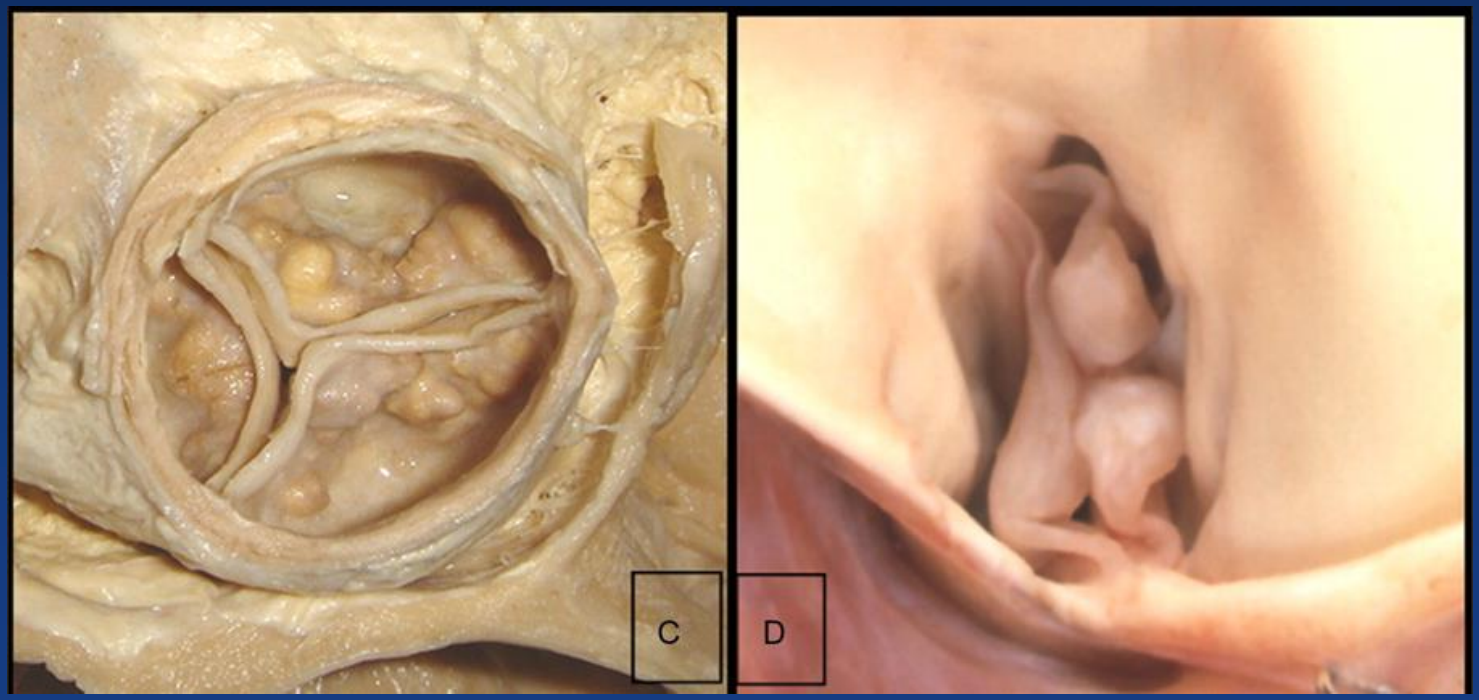
- **Morphology of the valve**
  - **Bicuspid, dysplastic leaflet, fused commissures**



LVOTO

- Valvar -

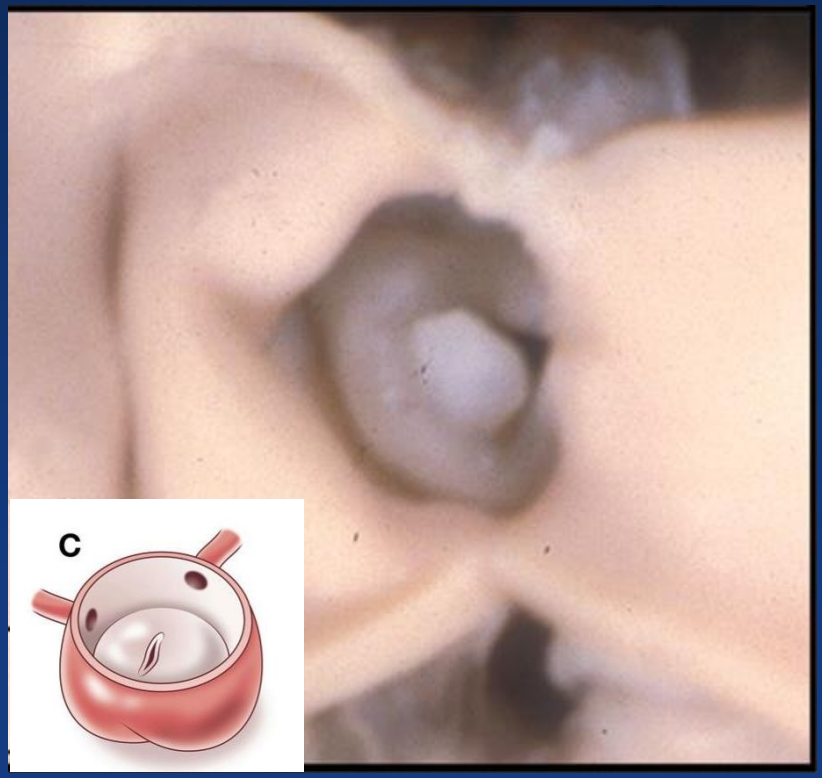
- **Morphology of the valve**
  - **Tricuspid, dysplastic leaflet, fused commissures**



LVOTO

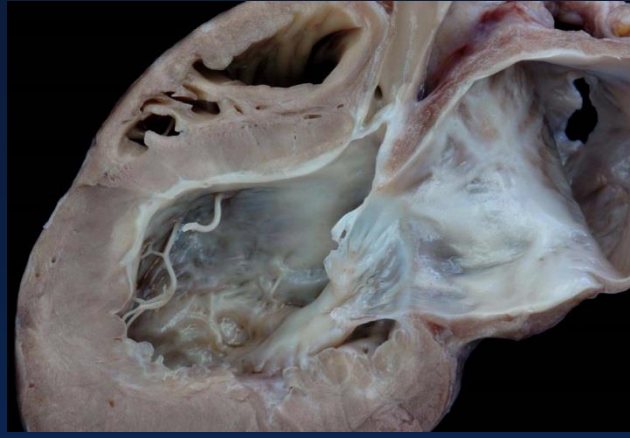
- Valvar -

- **Morphology of the valve**
  - **Unicuspid**



LVOTO

- Valvar -



- **AS in neonate and infant**
  - **Pathophysiology (severe AS)**

→ ↑ afterload → ↑ wall tension & workload → hypertrophy → dysfunction

→ PG at valve level → coronary/myocardial perfusion mismatch → myocardial ischemia (esp. subEndoC) → EFE → dysfunction

LVOTO

- Valvar -

- **AS in neonate and infant**
  - **Postnatal course depends on**
    - ✓ **Severity of valvar obstruction**
    - ✓ **Degree of LV dysfu**
    - ✓ **Degree of LV hypoplasia**
    - ✓ **Shunt flow at atrial & ductal level**
  - **Symptoms: exertional chest pain, easy fatigability, syncope**

R→ L (+adequate forward flow through AoV): differential cyanosis

LVOTO

- Valvar -

- ❖ Underestimated
  - ▪ ventricular dysfunction
  - ▪ R to L shunt through PDA
- ❖ Flow direction (PDA, arch, AAo.)

  → 2V vs. 1V

✓ Severity of stenosis (doppler)

LVOTO

- Valvar -

- **AS in neonate and infant**
  - ➢ **Cardiac catheterization**
    - ✓ **Therapeutic >> Diagnostic**

LVOTO

- Valvar -

- **AS in neonate and infant**
  - ➢ **Indication for intervention**
    - ✓ **CHF**
    - ✓ **Ductal dependent circulation**
    - ✓ **Symptoms**
    - ✓ **Pressure gradient (P-P) > 50mmHg**

→ Left side structures: adequate for sustaining systemic circulation??

LVOTO

- Valvar -

- AS in neonate and infant

Borderline LV

- ✓ Pressure gradient (P-P) > 50mmHg

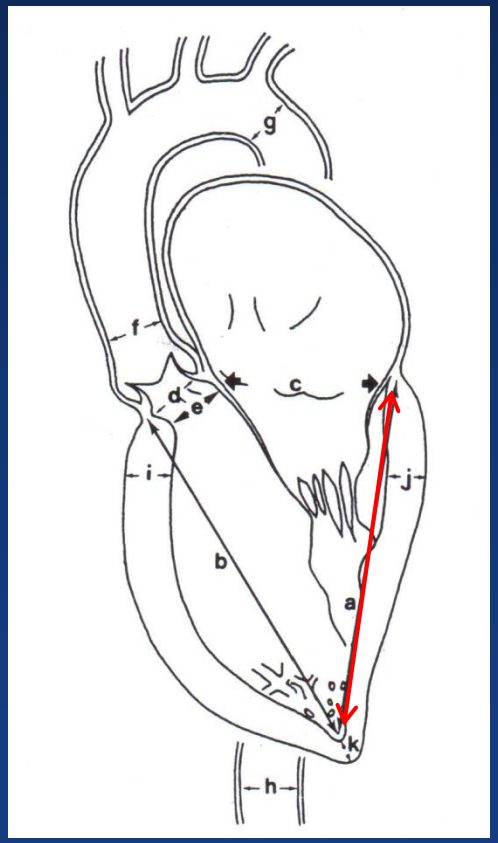
→ Left side structures: adequate for sustaining systemic circulation??

Critical aortic stenosis in early infancy

Anatomic and echocardiographic substrates of successful open valvotomy

Maurice P. Leung, MBBS, MRCP,* Roxane McKay, BA, MD, FRCS, Audrey Smith, FIMLS, MPhil, PhD, Robert H. Anderson, BSc, MD, FRCPath,** and Robert Arnold, MB, ChB, FRCP, *Liverpool, England*

J Thorac Cardiovasc Surg 1991;101:526-535



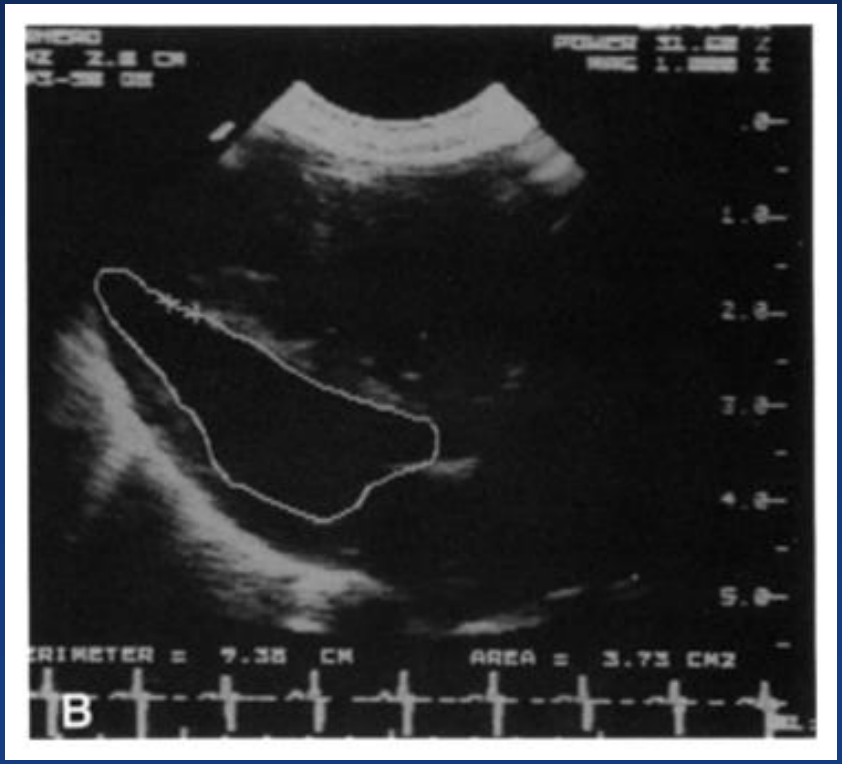
- **LV inflow dimension <25mm**
- **VA junction <5mm**
- **MV orifice <9mm**

Echocardiographic Estimation of Critical Left Ventricular Size in Infants With Isolated Aortic Valve Stenosis

MARK K. PARSONS, MD, GORDON A. MOREAU, MD, THOMAS P. GRAHAM, JR., MD, FACC, JAMES A. JOHNS, MD, ROBERT J. BOUCEK, JR., MD

Nashville, Tennessee

J Am Coll Cardiol 1991;18:1049-1055



- **LV cross-sectional area $<2cm^2$**
  - **LVEDD <13mm**
  - **LVEDV < 20ml/m^2 (Cath)**

Predictors of Survival in Neonates With Critical Aortic Stenosis

Larry A. Rhodes, MD; Steven D. Colan, MD; Stanton B. Perry, MD;
Richard A. Jonas, MD; and Stephen P. Sanders, MD

Circulation 1991;84:2325-2335

- **Threshold Score (Sum): <2 → 2V favor**
  - ✓ LV to heart (long axis) ratio ≤ 0.8
  - ✓ Indexed aortic root diameter ≤ 3.5cm/m^2
  - ✓ Indexed mitral valve area ≤ 4.75cm^2/m^2
  - ✓ LM mass index ≤ 35 g/m^2

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$$\text{Score} = 14.0\ (\text{BSA}) + 0.943(\text{ROOT}_i) + 4.78\ (\text{LAR}) + 0.157\ (\text{MVA}_i) - 12.03$$

Rhodes score

SV **-0.35** **BV**

CRITICAL AORTIC STENOSIS IN THE NEONATE: A MULTI-INSTITUTIONAL STUDY OF MANAGEMENT, OUTCOMES, AND RISK FACTORS

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Christo I. Tchervenkov, MD[e]
Rekwan Sittiwangkul, MD[b]
Richard A. Jonas, MD[f]
Congenital Heart Surgeons Society

J Thorac Cardiovasc Surg 2001;121:10-27

Table VI. *Independent factors predictive of percent survival benefit at 5 years after entry for Norwood procedure versus biventricular repair**

Variable	*Parameter estimate (SE)*	P *value*
Intercept	–86.47 (6.36)	
Higher grade of endocardial fibroelastosis	12.14 (0.96)	<.001
Lower z-score of aortic valve at the level of the sinuses of Valsalva	–6.20 (0.25)	<.001
Younger age at entry (d)†	30.55 (1.79)	<.001
Larger ascending aorta diameter (mm)‡	23.33 (2.24)	<.001
Absence of moderate or severe tricuspid regurgitation	–28.30 (2.60)	<.001
Lower z-score of the LV length	–0.70 (0.22)	.02

SE, Standard error.
*Adjusted for missing values for all variables; model R^2 = 0.888; root mean square error = 9.858.
†Inverse of (age at entry + 1) transformation.
‡Logarithmic transformation.

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J Thorac Cardiovasc Surg 2001;121:10-27

CHSS score

Survival benefit = 30.55 (inverse of age at study entry [d] + 1)
− 6.20 (aortic root *z*-score)
+ 12.14 (echocardiographic grade of EFE)
+ 23.33 (logarithm of ascending aortic [mm])
− 28.30 (presence of moderate or severe tricuspid regurgitation)
− 0.70 (LV long-axis length *z*-score) − 86.47

- **Positive value: SV favor**
- **Negative value: BV favor**

Validation and Re-Evaluation of a Discriminant Model Predicting Anatomic Suitability for Biventricular Repair in Neonates With Aortic Stenosis

Steven D. Colan, MD, Doff B. McElhinney, MD, Elizabeth C. Crawford, RDCS, John F. Keane, MD, James E. Lock, MD

Boston, Massachusetts

J Am Coll Cardiol 2006;47:1858-1865

- **Colan Score** = 10.98 (BSA) + 0.56 (aortic valve annulus z-score) + 5.89 (LAR) – 0.79 (presence of grade 2 or 3 EFE) – 6.78.

  → Cutoff : -0.65 (~90% predictive)

- Score (EFE omitted) = 12.16 (BSA) + 0.59 (aortic valve annulus z-score) + 5.73 (LAR) – 7.02 (→ Cutoff : -0.46)

Critical left ventricular outflow tract obstruction: The disproportionate impact of biventricular repair in borderline cases

Edward J. Hickey, MD,[a] Christopher A. Caldarone, MD,[b,*] Eugene H. Blackstone, MD,[c] Gary K. Lofland, MD,[d] Thomas Yeh, Jr, MD,[e] Christian Pizarro, MD,[f] Christo I. Tchervenkov, MD,[g] Frank Pigula, MD,[h] David M. Overman, MD,[i] Marshall L. Jacobs, MD,[j] Brian W. McCrindle, MD,[b,*] and the Congenital Heart Surgeons' Society

J Thorac Cardiovasc Surg 2007;134:1429-1437

TABLE 1. Incremental risk factors for time-related death for patients who had an initial procedure indicating an intended biventricular repair pathway

Covariate	Estimate	*P* value
Intercept	−.484	<.001
Presence of moderate or severe tricuspid regurgitation	−.279	<.001
Z-score of mitral valve annulus	+.030	<.001
Presence of large VSD	−.312	<.001
Length of apex-forming ventricle (cm)*	+.715	<.001
Minimum diameter of the LVOT (cm)†	+.892	<.001
Presence of left ventricular dysfunction‡	+.230	<.001
Grade of endocardial fibroelastosis§	+.165	<.001
Diameter of the mid-aortic arch (cm)‖	−.187	<.001

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chss score for neonatal critical aortic stenosis

calculator

for neonatal critical aortic stenosis

data collection

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Weight (Kg):	
Height (cm):	
BSA:	
Presence of moderate or severe tricuspid regurgitation :	none-mild
Mitral valve annulus: (cm)	
Presence of large ventriculo-septal defect:	none
Length of apex-forming ventricle (mm):	
Minimum diameter of the left ventricular outflow tract(mm):	
Presence of left ventricular dysfunction:	0 = none
Grade of endocardial fibroelastosis:	0 = none
Diameter of the mid-aortic arch (cm):	
	Click here to get score :
CHSS Score:	

Revised CHSS score

www.chssdc.org

Calculations for the Borderline Left Ventricle

(Rhodes Score, Discriminant Score, & CHSS Scores)

Inputs

Height (cm):

Weight (kg):

Age (days):

Mitral Valve Annulus, LAX (mm):

Mitral Valve Annulus, A4C (mm):

Left Ventricular Long Axis (mm):

Heart Long Axis (mm):

Minimum LVOT Diam (mm):

Aortic Valve (mm):

Ascending Aorta (mm):

Results

Rhodes

CHSS-1

Discriminant

CHSS-2

http://dev.parameterz.com/borderline

LVOTO

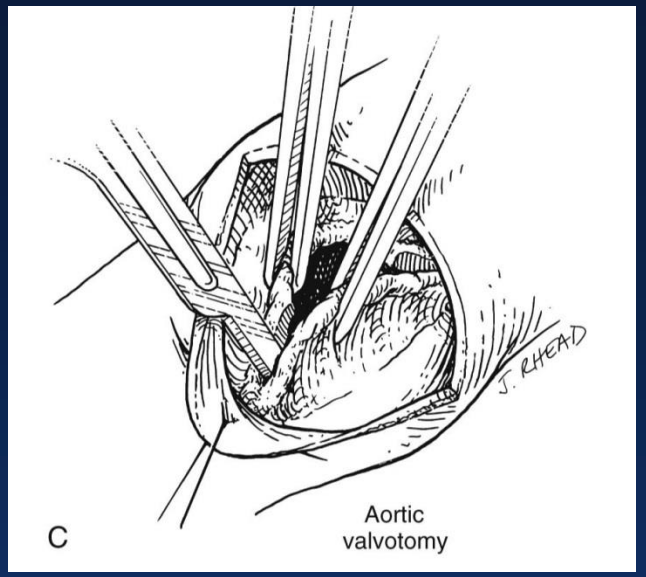
- Valvar -

- **AS in neonate and infant**
  - **Treatment options**
    - ✓ **Percutaneous balloon valvuloplasty**
    - ✓ **Surgical valvotomy/ valvuloplasty**
    - ✓ **Aortic valve replacement with pulmonary autograft (Ross)**

      : with or without annular enlargement

LVOTO

- Valvar -



- **AS in neonate and infant**
  - ➢ **Surgical valvotomy**
    - ✓ **Do not reach aortic wall: even one millimeter enough to sufficiently enlarge aortic opening**
    - ✓ **Do not touch false raphe**
    - ✓ **As conservative as possible**

**** AR → ↑ probability of reoperation**

LVOTO

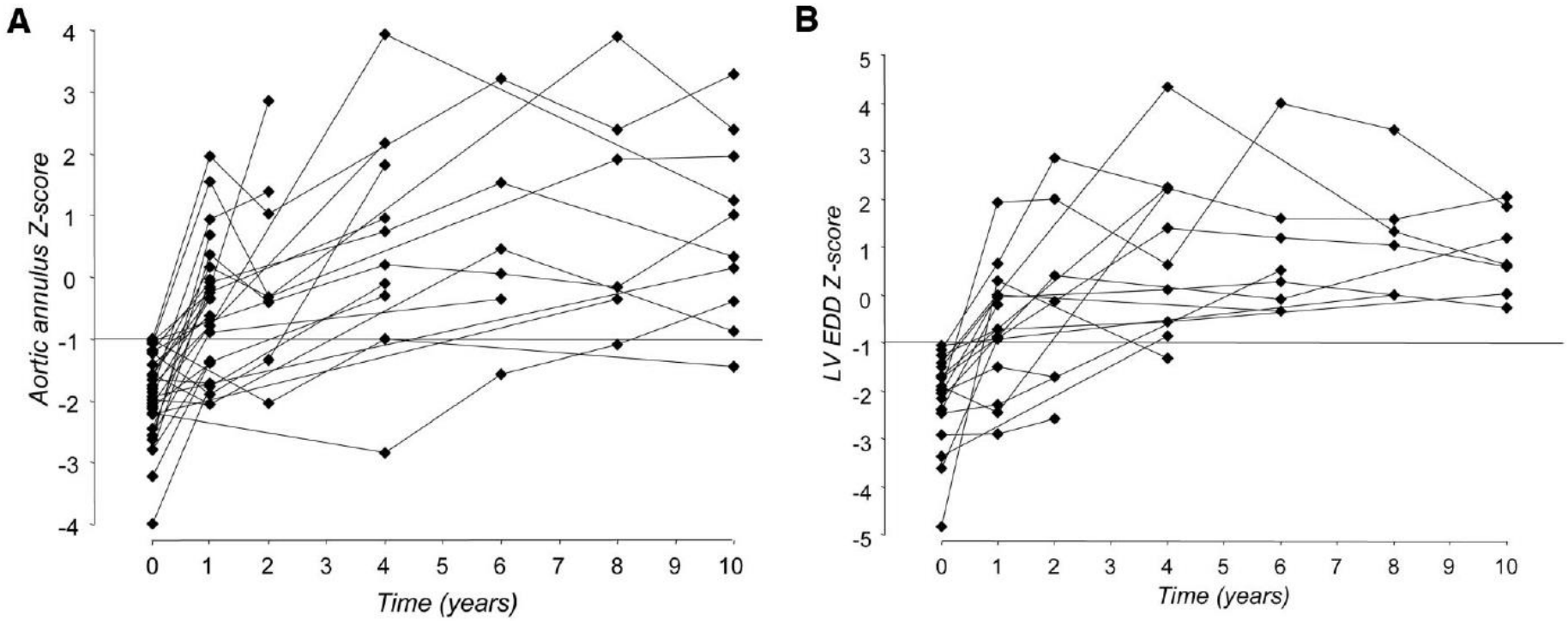
- Valvar -

- **AS in neonate and infant**
  - **Outcomes**
    - ✓ **Balloon vs. surgical: similar in mortality and reintervention for AoV**
    - ✓ **↑ AS in surgery vs. ↑ AR in balloon**
    - ✓ **Outcome improvement: importance of patient selection (borderline LV)**
    - ✓ **Catch-up growth of left heart structures**

Left Heart Growth, Function, and Reintervention After Balloon Aortic Valvuloplasty for Neonatal Aortic Stenosis

Doff B. McElhinney, MD; James E. Lock, MD; John F. Keane, MD;
Adrian M. Moran, MD; Steven D. Colan, MD

Circulation 2005;111:451-458

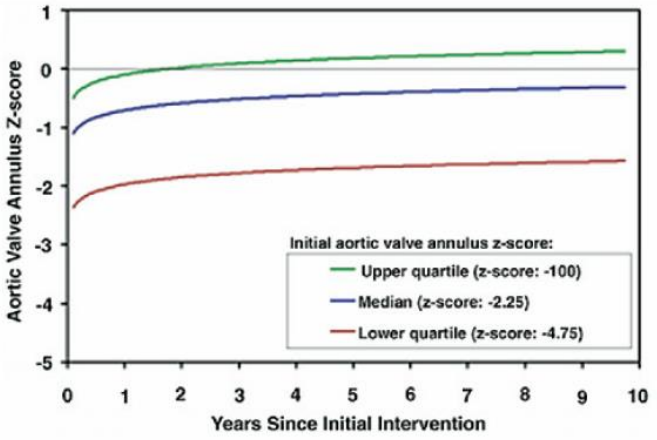


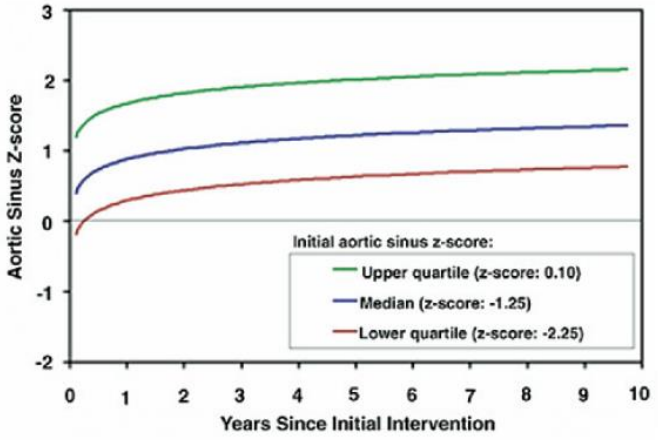
Outcome and Growth Potential of Left Heart Structures After Neonatal Intervention for Aortic Valve Stenosis

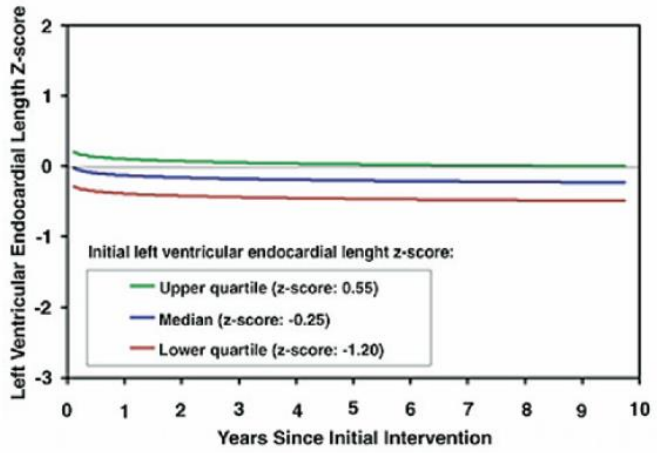
Ra K. Han, MD, FRCPC,* Rebecca C. Gurofsky, BSc,* Kyong-Jin Lee, MD, FRCPC,*
Anne I. Dipchand, MD, FRCPC,* William G. Williams, MD, FRCSC,†
Jeffrey F. Smallhorn, MD, FRCPC,* Brian W. McCrindle, MD, MPH, FRCPC*

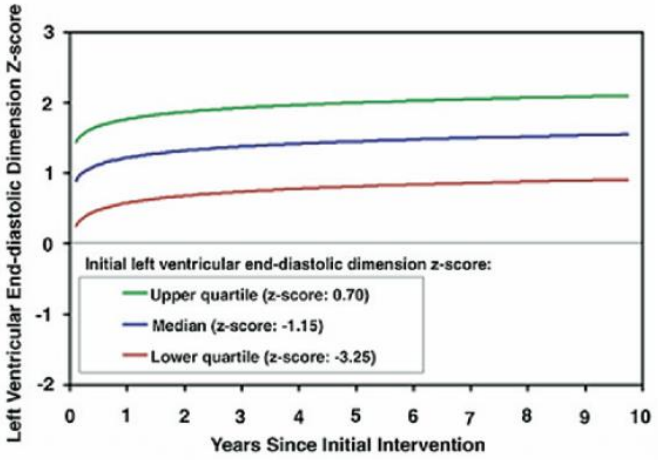
Toronto, Ontario, Canada

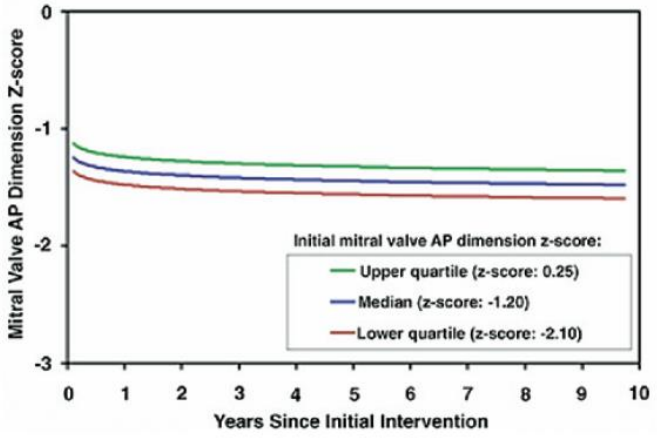
J Am Coll Cardiol 2007;50:2406-2414

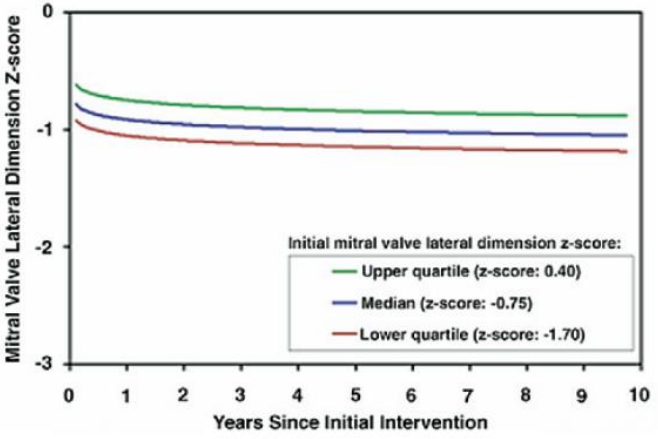

Aortic Valve Annulus Z-score
Initial aortic valve annulus z-score:
Upper quartile (z-score: -100)
Median (z-score: -2.25)
Lower quartile (z-score: -4.75)
Years Since Initial Intervention


Aortic Sinus Z-score
Initial aortic sinus z-score:
Upper quartile (z-score: 0.10)
Median (z-score: -1.25)
Lower quartile (z-score: -2.25)
Years Since Initial Intervention


Left Ventricular Endocardial Length Z-score
Initial left ventricular endocardial lenght z-score:
Upper quartile (z-score: 0.55)
Median (z-score: -0.25)
Lower quartile (z-score: -1.20)
Years Since Initial Intervention


Left Ventricular End-diastolic Dimension Z-score
Initial left ventricular end-diastolic dimension z-score:
Upper quartile (z-score: 0.70)
Median (z-score: -1.15)
Lower quartile (z-score: -3.25)
Years Since Initial Intervention


Mitral Valve AP Dimension Z-score
Initial mitral valve AP dimension z-score:
Upper quartile (z-score: 0.25)
Median (z-score: -1.20)
Lower quartile (z-score: -2.10)
Years Since Initial Intervention


Mitral Valve Lateral Dimension Z-score
Initial mitral valve lateral dimension z-score:
Upper quartile (z-score: 0.40)
Median (z-score: -0.75)
Lower quartile (z-score: -1.70)
Years Since Initial Intervention

LVOTO

- Valvar -

- **AS in older children**
  - ➢ **Indication for intervention**
    - ✓ **Symptoms with PG >50mmHg**
    - ✓ **Asymptomatic but PG >70mmHg**
    - ✓ **Asymptomatic, PG 50~70mmHg**

      → controversial

      → early intervention is beneficial

LVOTO

- Valvar -

- **AS in older children**
  - **Treatment options**
    - ✓ **Percutaneous balloon valvuloplasty**
    - ✓ **Surgical valvotomy/ valvuloplasty**
    - ✓ **Aortic valve replacement with pulmonary autograft (Ross)**

      : with or without annular enlargement

LVOTO

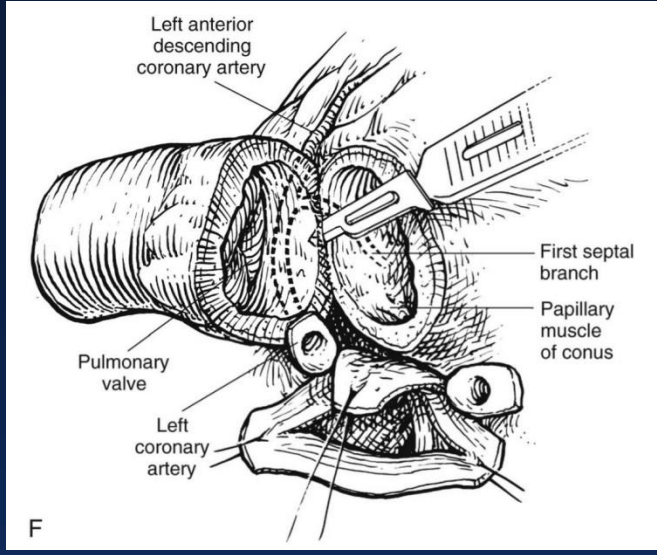
- Valvar -

- **AS in older children**
  - ➢ **AVR**
    - ✓ **Significant No. need AVR, eventually.**
    - ✓ **Prosthetic AVR in the young**
      - ❖ **Limited size and lack of growth**
      - ❖ **Early degeneration in bio.**
      - ❖ **Life-long anticoagulation in mech.**

      → **Pulmonary autograft (Ross)**

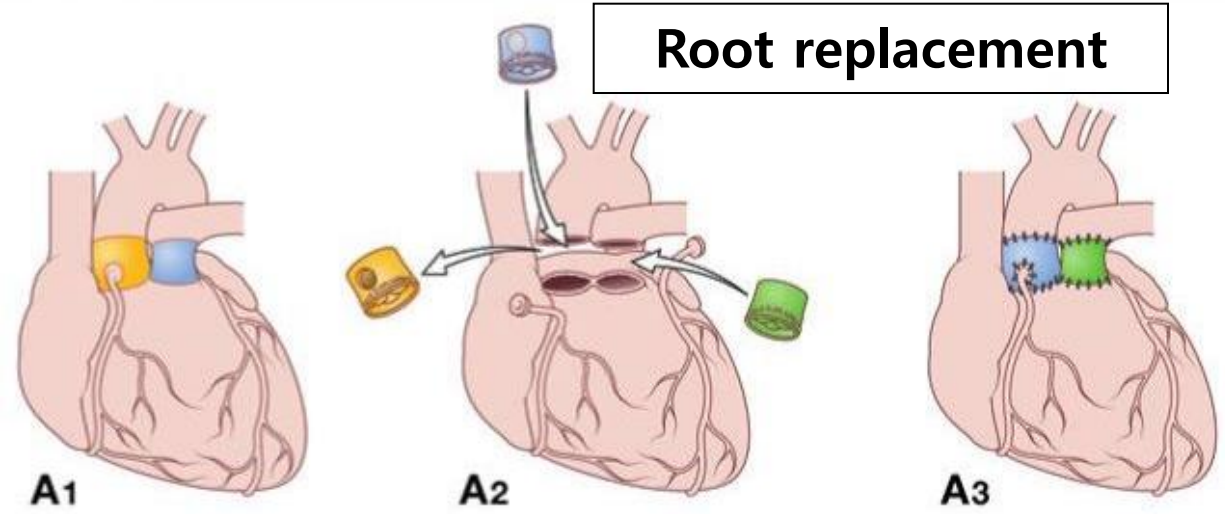
LVOTO

- Valvar -

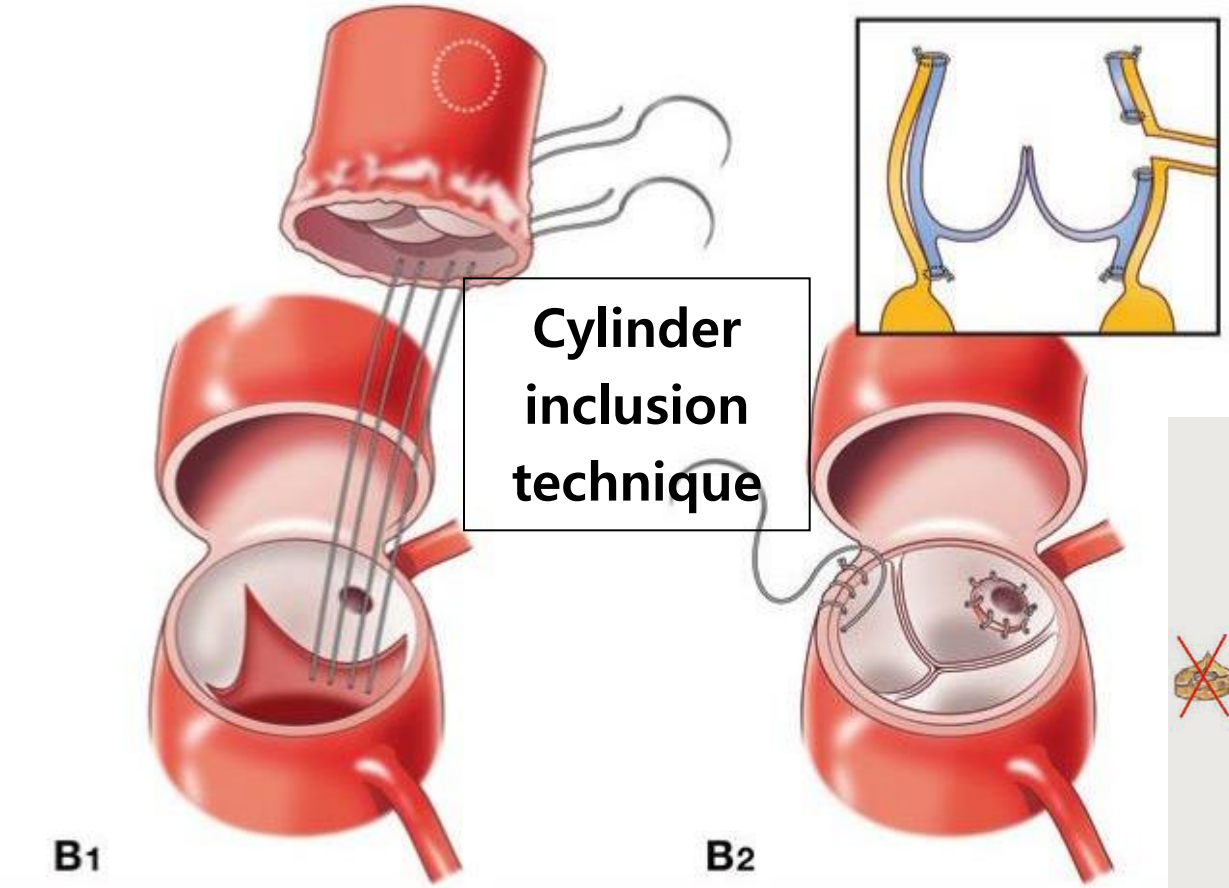


- **Ross operation**
  - ➢ **Safe autograft harvesting**
    - ✓ **Avoiding damaging leaflet**
    - ✓ **Sufficient RVOT m. cuff**
    - ✓ **Avoiding injury to 1st septal perforator**
  - ➢ **Not an ideal option**
    - ✓ **Autograft function : dilatation and dysfxn**
    - ✓ **Reoperation for pulmonary valve**

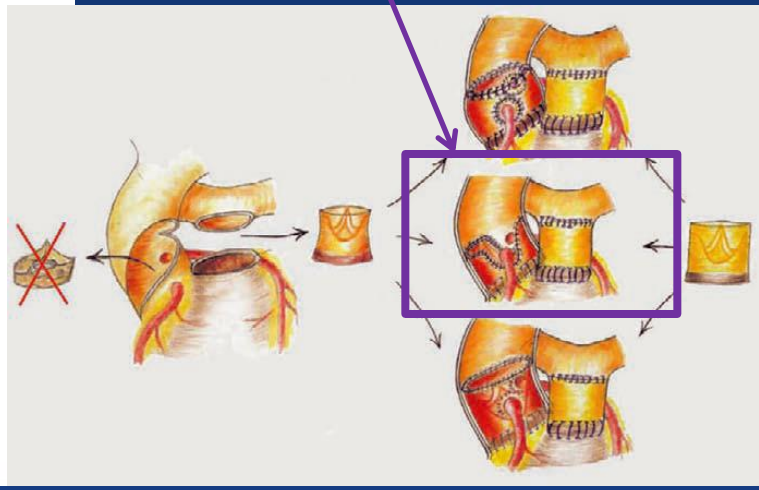
Root replacement



Cylinder inclusion technique

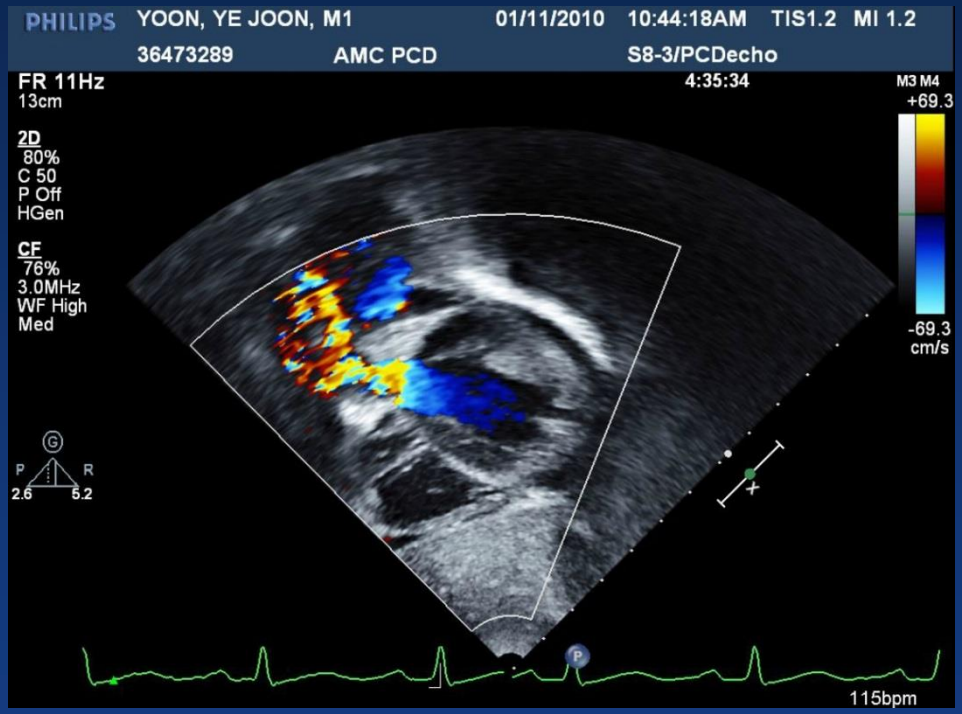


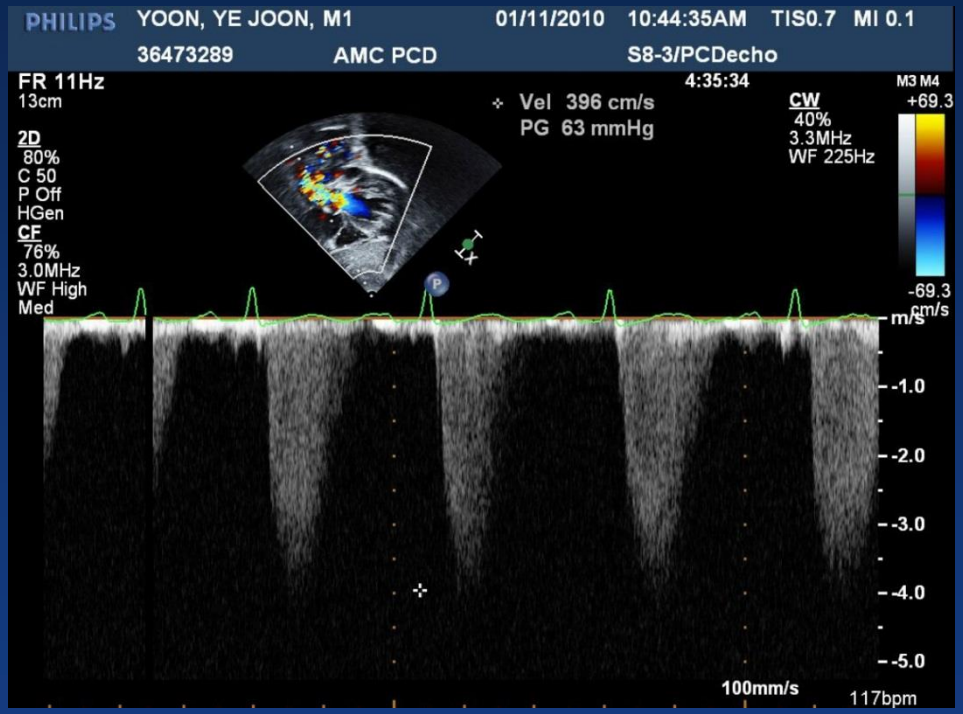
Subcoronary implantation



Case

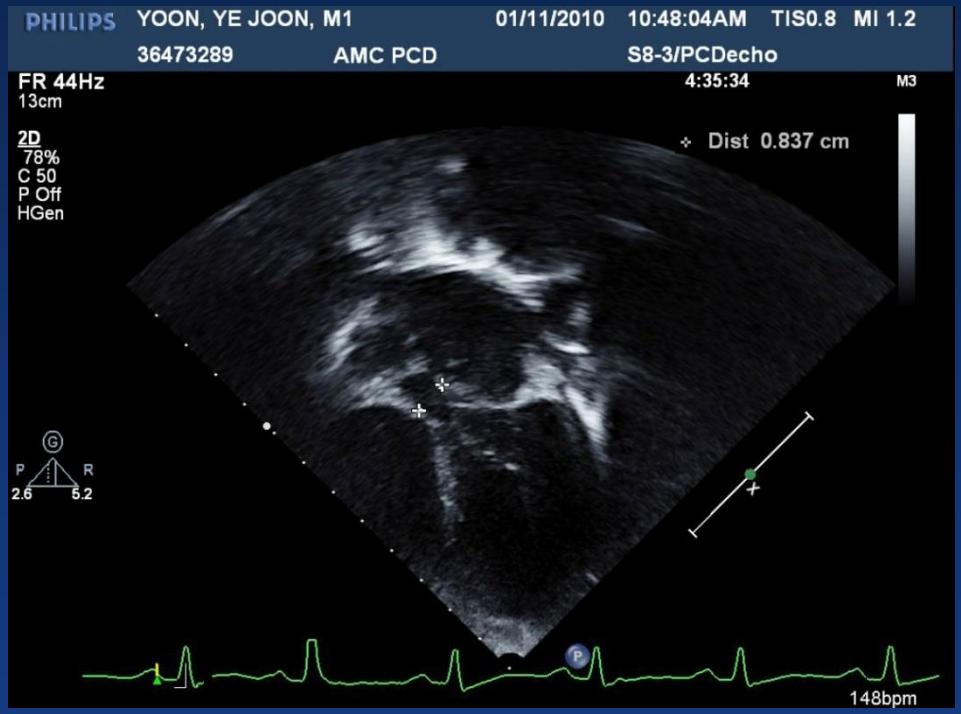
- M/1Y, s/p balloon AVP -

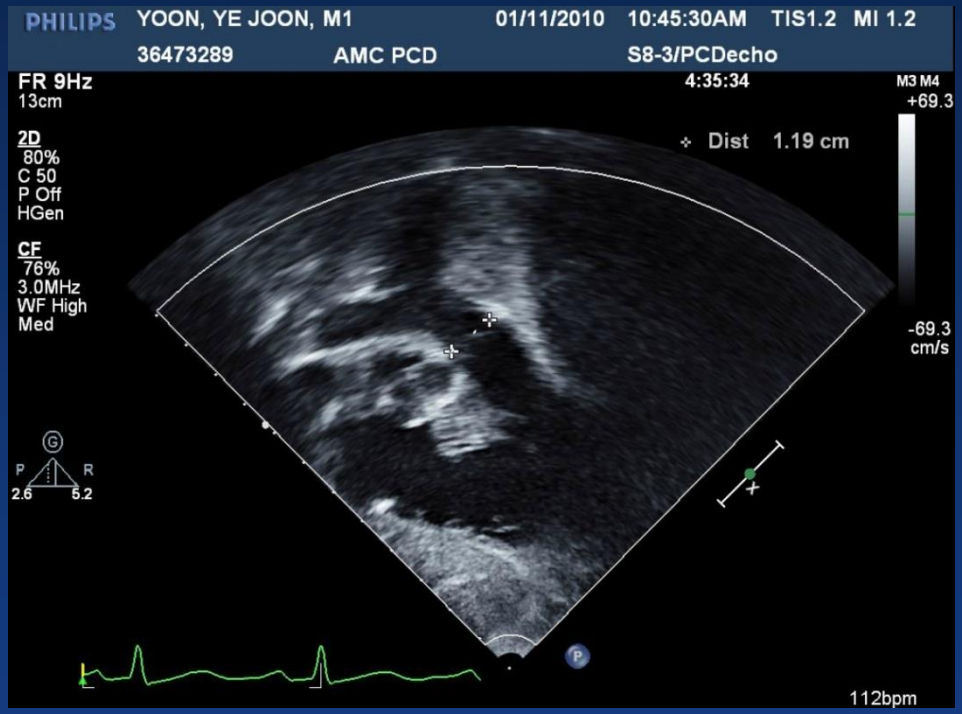




Case

- M/1Y, s/p balloon AVP -





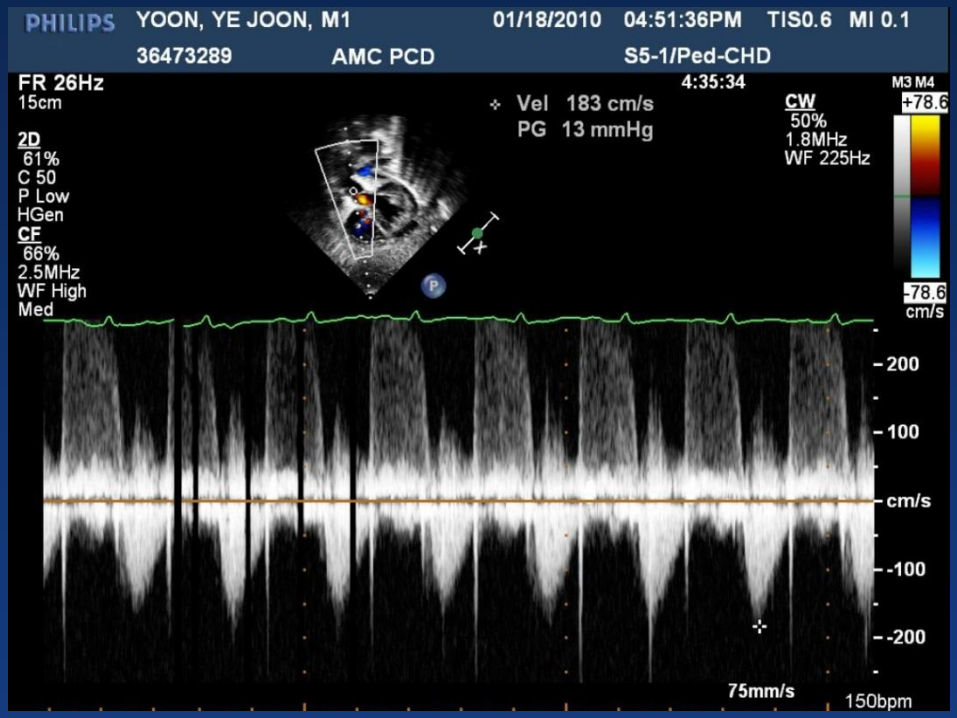
Case

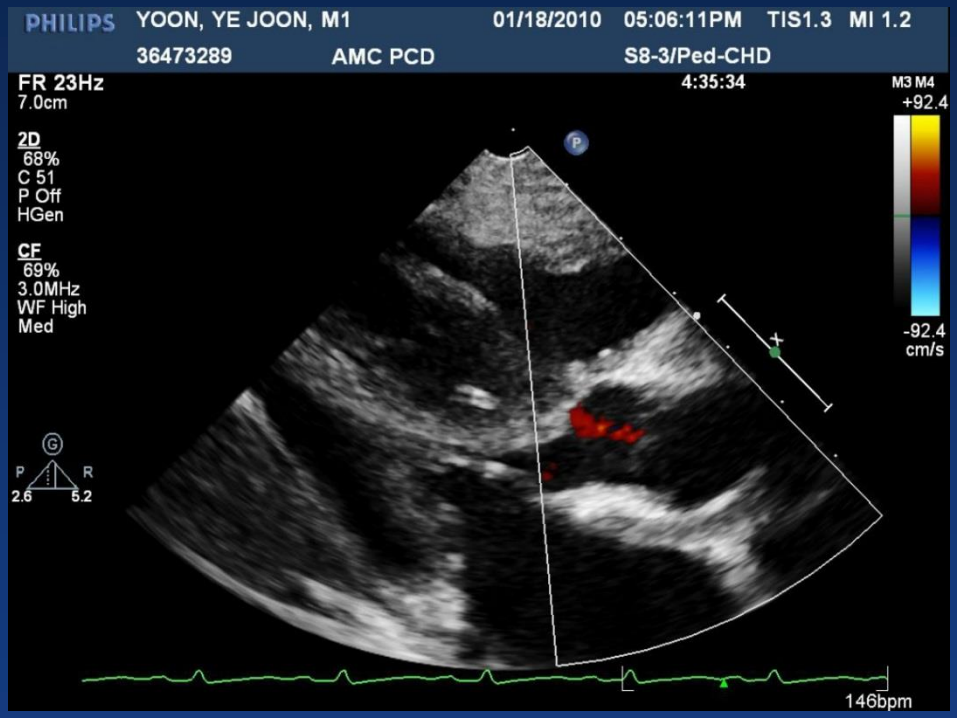
- M/1Y, s/p balloon AVP -

- Op name: Ross operation (root replacement), RVOT reconstruction with aortic homograft

Case

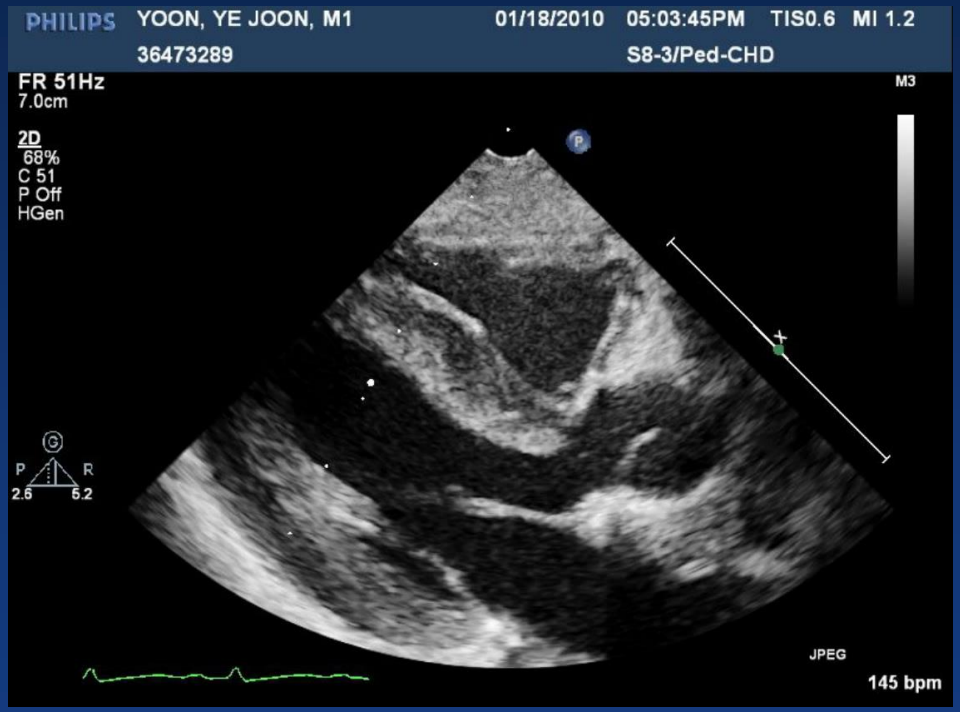
- M/1Y, s/p balloon AVP -





Case

- M/1Y, s/p balloon AVP -



LVOTO

- Valvar -

- **AS in older children**
  - **Outcomes**
    - ✓ **AVR (biologic/ mechanical)**
      - **Excellent (survival and durability)**
    - ✓ **Ross**
      - **Excellent early outcomes**
      - **Autograft failure (20~30% at 20 years)**
        - **Initial passive dilatation → late growth**
      - **RV-PA failure (20% at 20 years)**

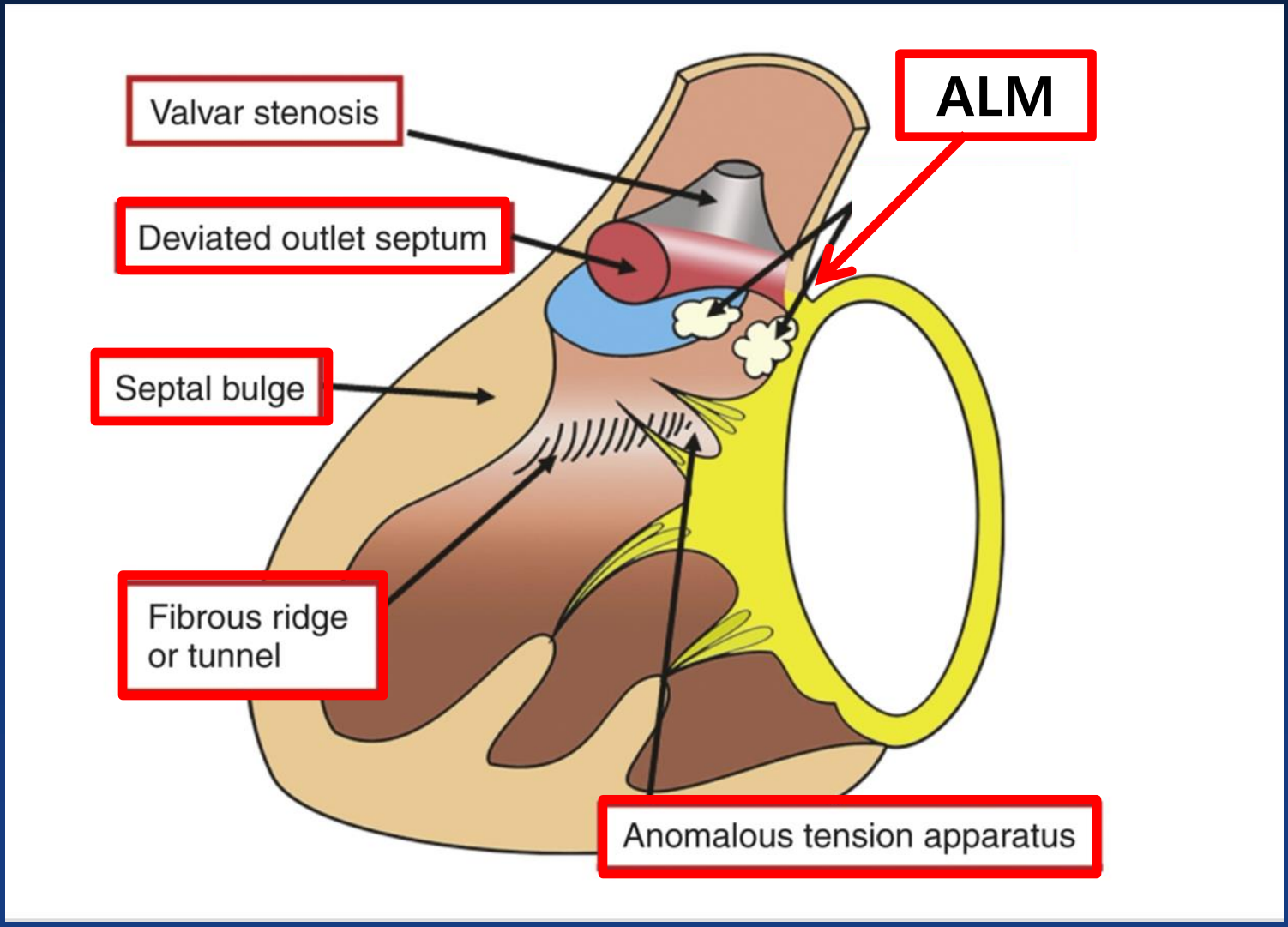
LVOTO

- Subvalvar -

- 15~20% of LVOTO
- Classification
  - Fixed SAS (70%)
    - ✓ Discrete
      - Membranous vs. fibromuscular
    - ✓ Diffuse
  - Hypertrophic CMP
  - Other SAS: MV mechanism associated, ALM hypertrophy, posteriorly deviated IS

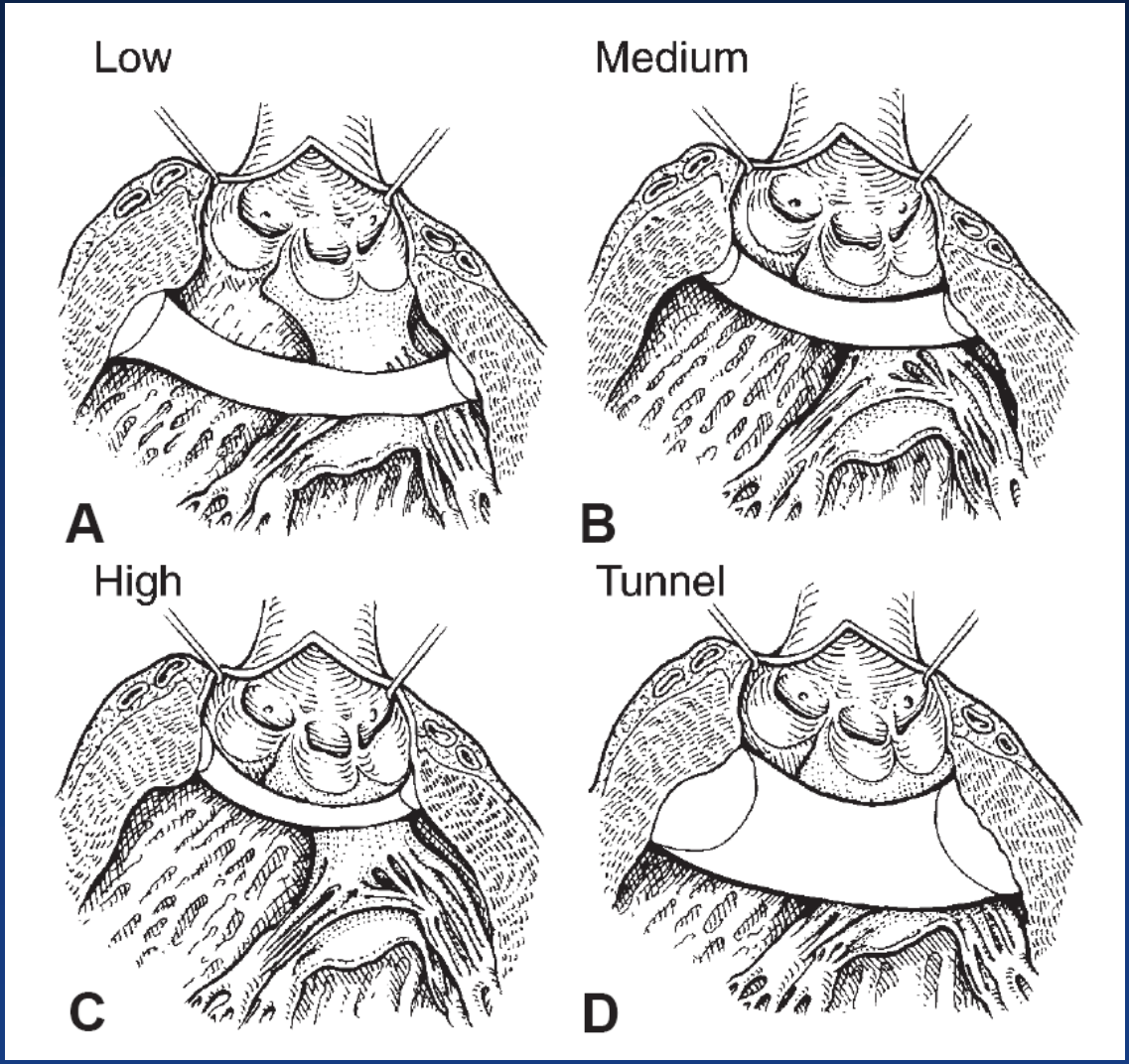
LVOTO

- Subvalvar -



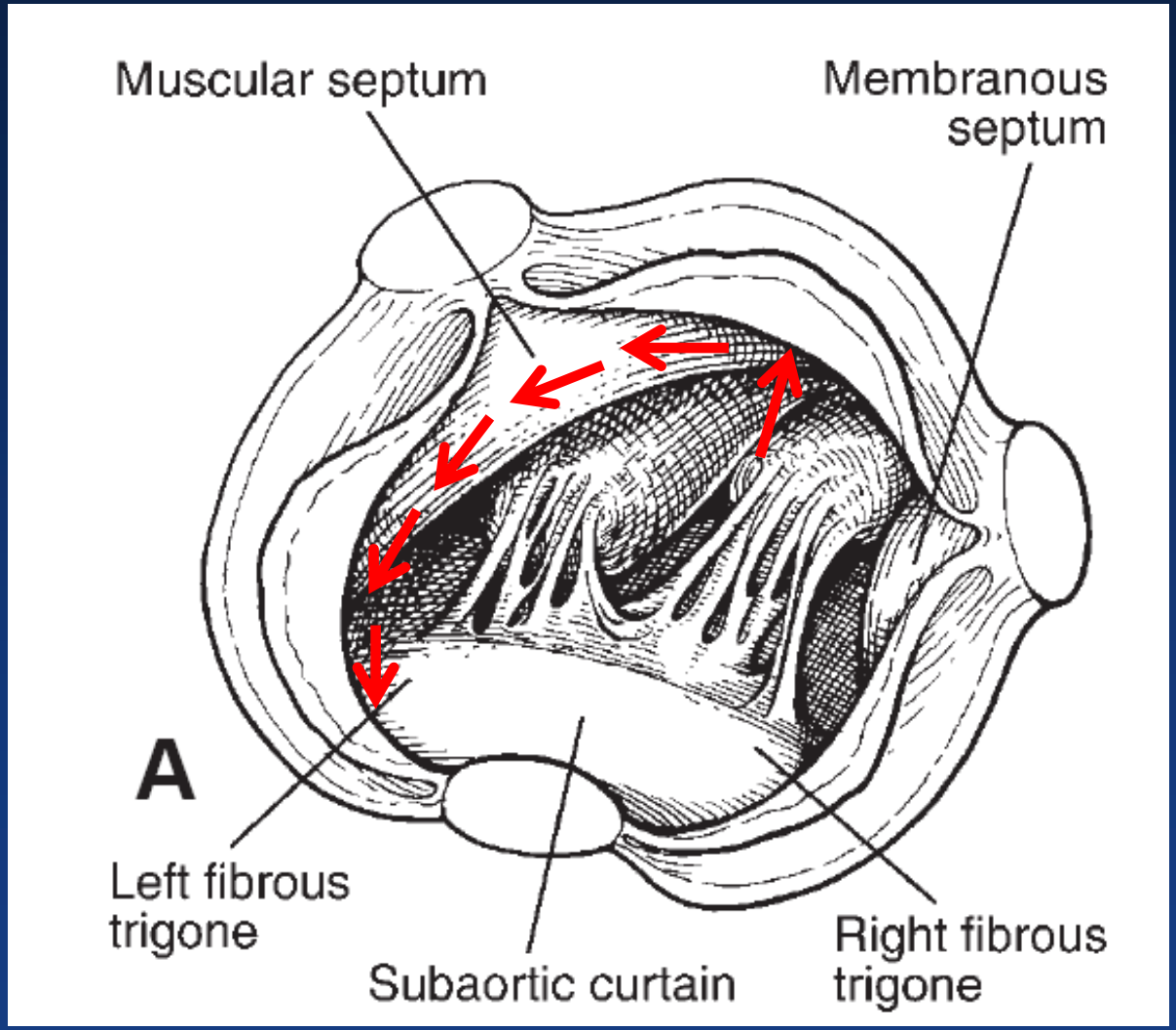
LVOTO

- Subvalvar -



LVOTO

- Subvalvar -



LVOTO

- Fixed SAS -

- Pathophysiology: similar to valvar AS
- Aortic valve in SAS
  - Smaller than normal but usually normal morphology
  - Turbulence and jet damaging leaflets

→ ↑ thickening or AoV

→ ↓ mobility of AoV

} ↑ AR

→ susceptible to infection

LVOTO

- Fixed SAS -

- **Indication for surgery**
  - **Symptoms: shortness of breath, angina, syncope, exercise intolerance**
  - **Progressive decompensation**
  - **In asymptomatic patients**
    - ✓ **Peak PG >30mmHg in discrete SAS**
    - ✓ **Peak PG >50mmHg in diffuse SAS**
  - **Aortic insufficiency**

LVOTO

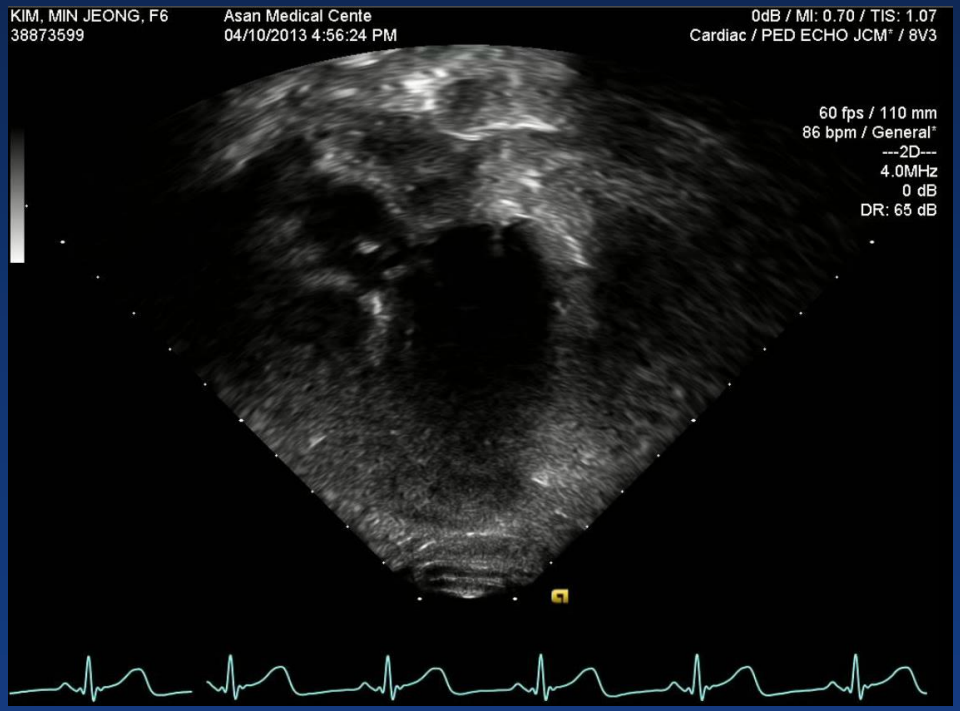
- Fixed SAS -

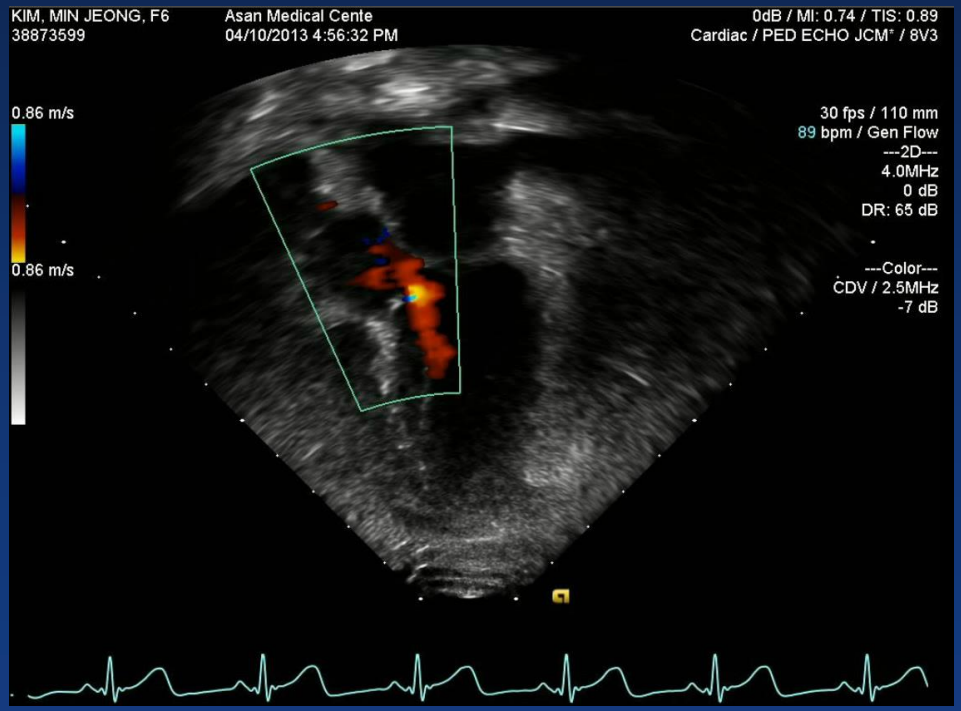
- **Surgery for fixed SAS**
  - **Discrete SAS**

    → **Membrane or ridge excision ± myectomy**

Case

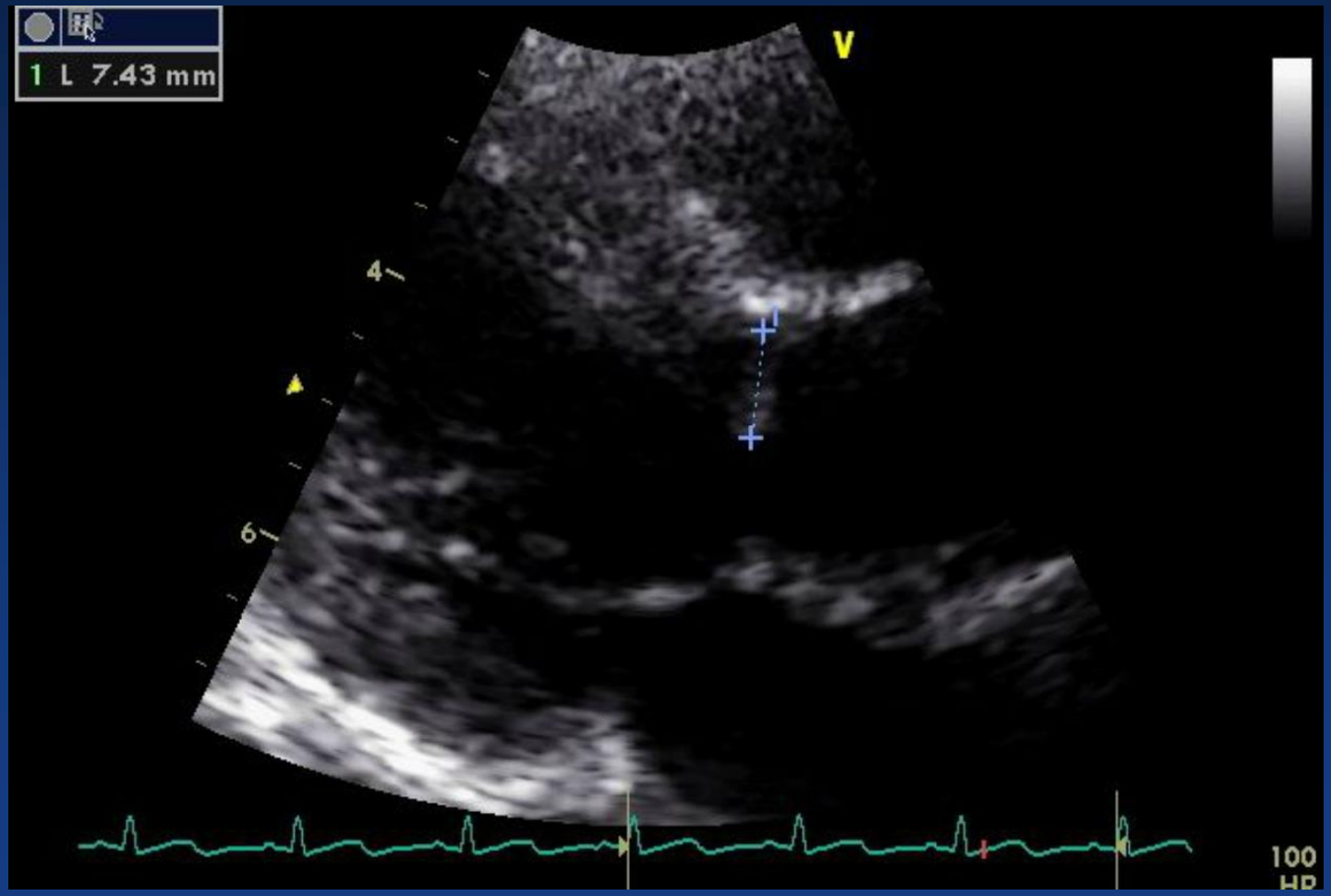
- F/7Y, s/p LVOTR (congenital SAS) -





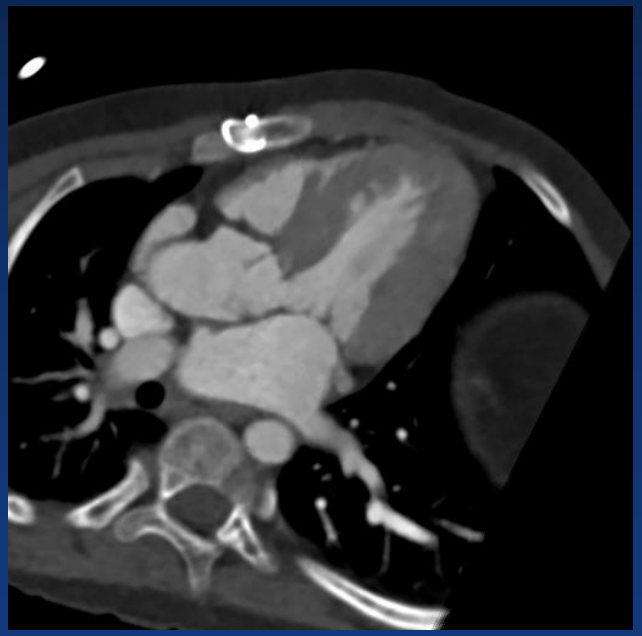
Case

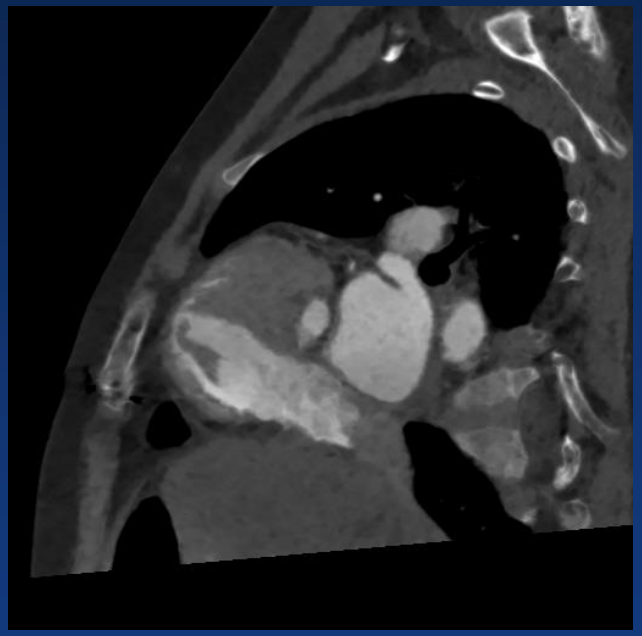
- F/7Y, s/p LVOTR (congenital SAS) -



Case

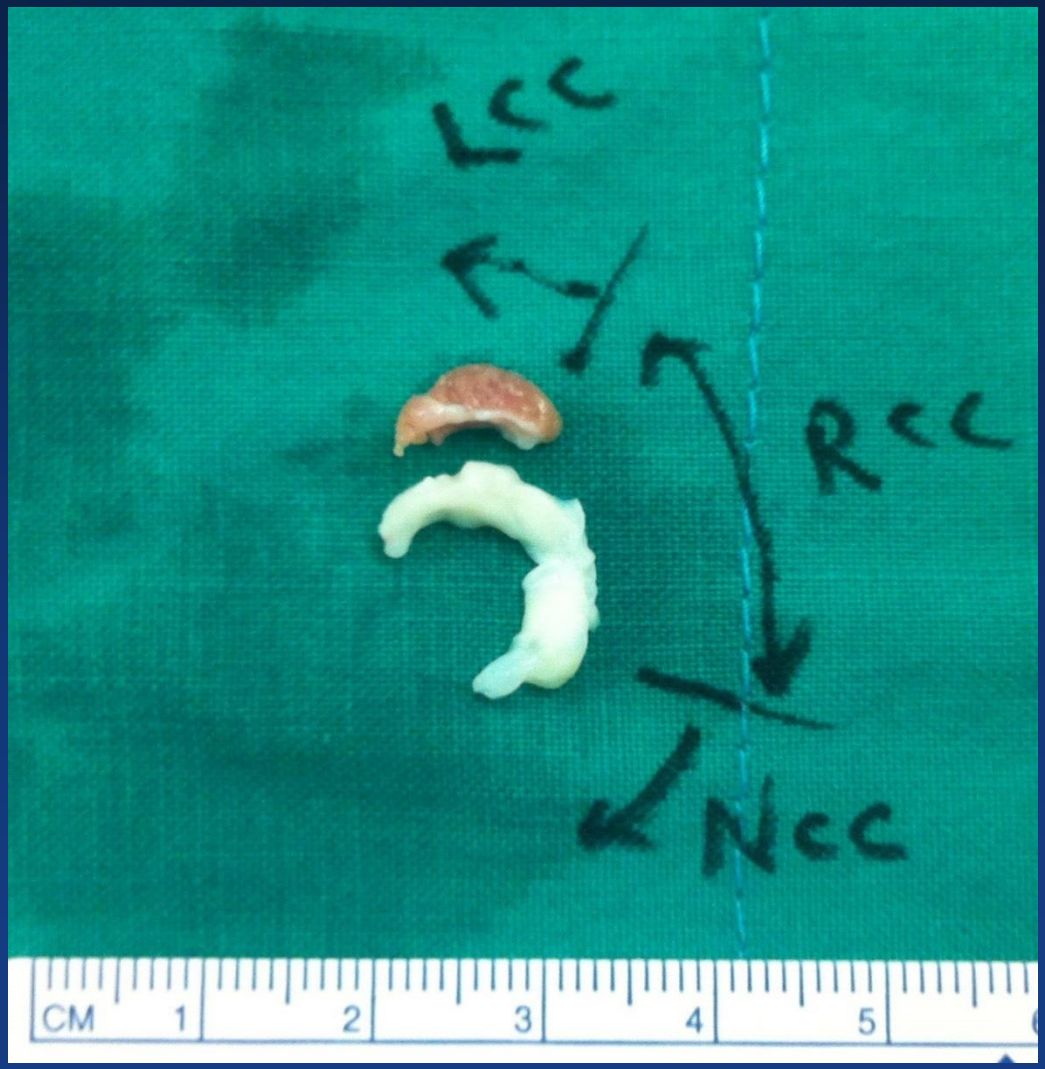
- F/7Y, s/p LVOTR (congenital SAS) -





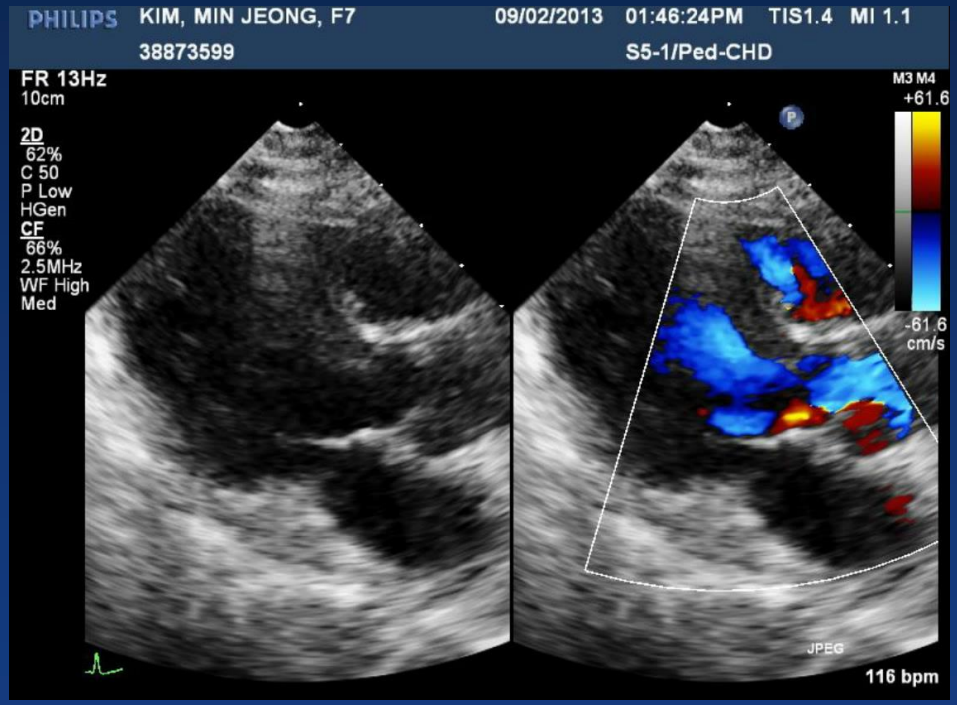
Case

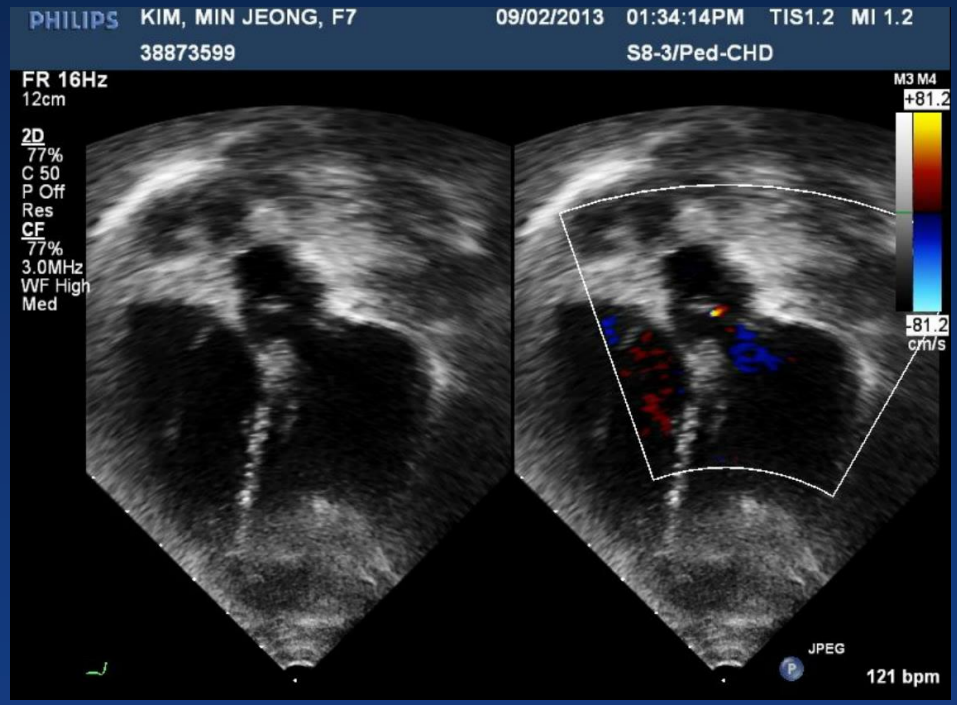
- F/7Y, s/p LVOTR (congenital SAS) -



Case

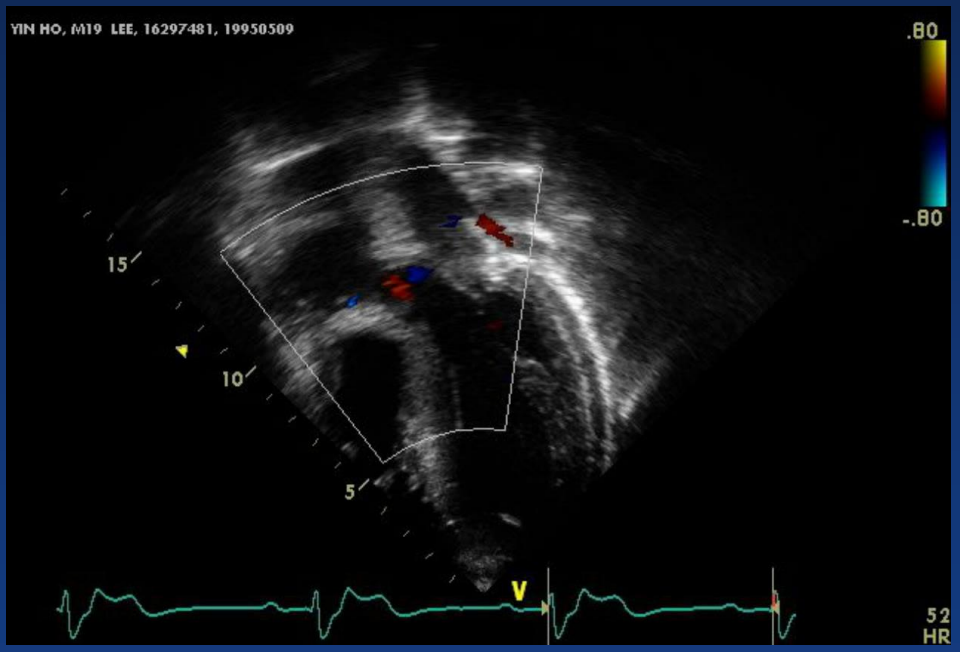
- F/7Y, s/p LVOTR (congenital SAS) -

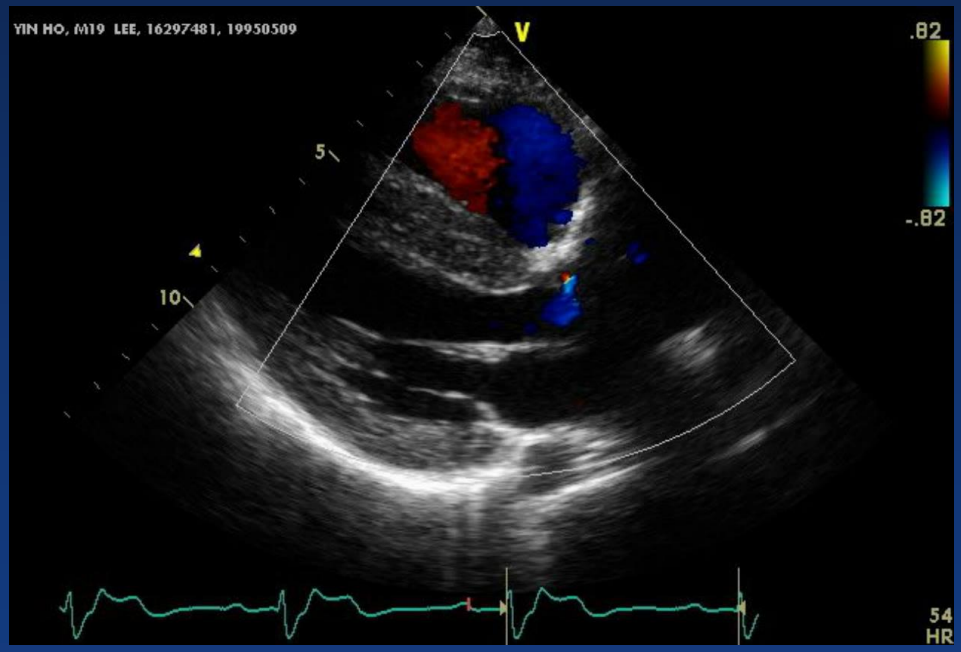




Case

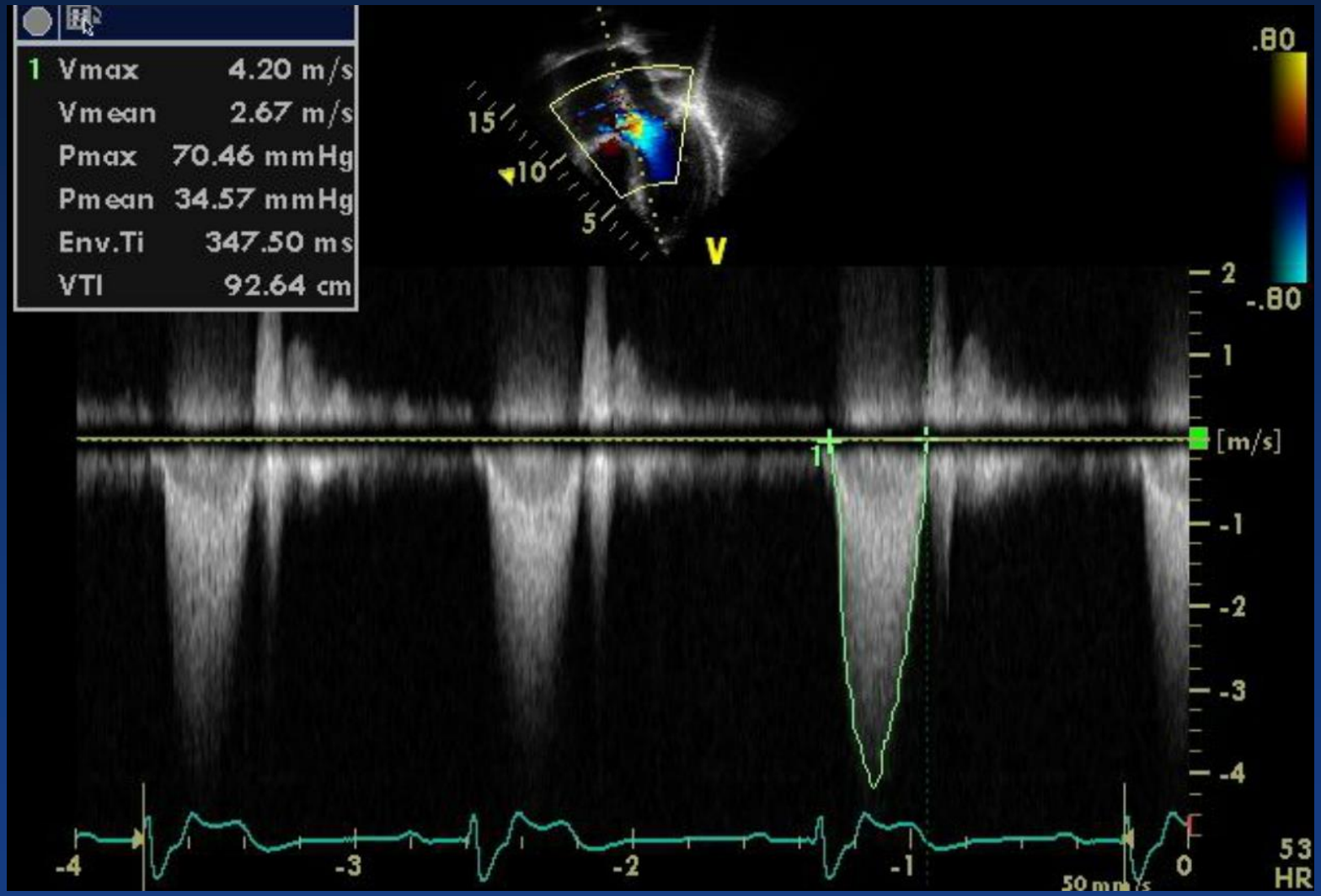
- M/19Y, s/p Fallot type DORV correction -





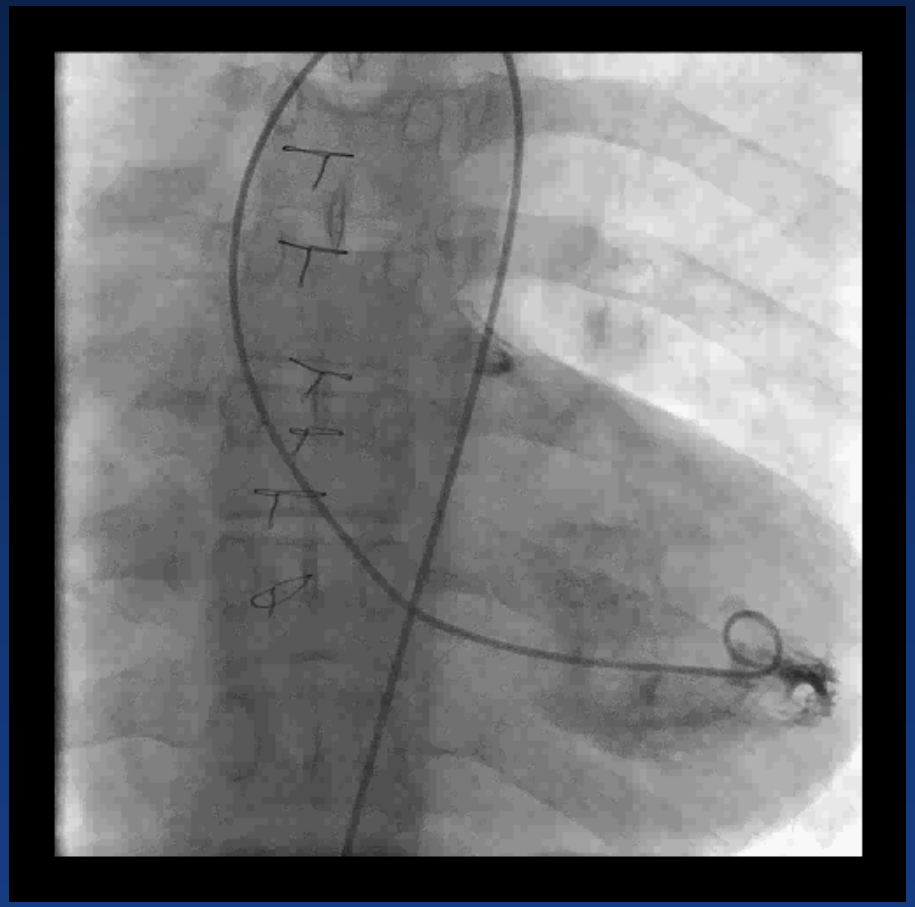
Case

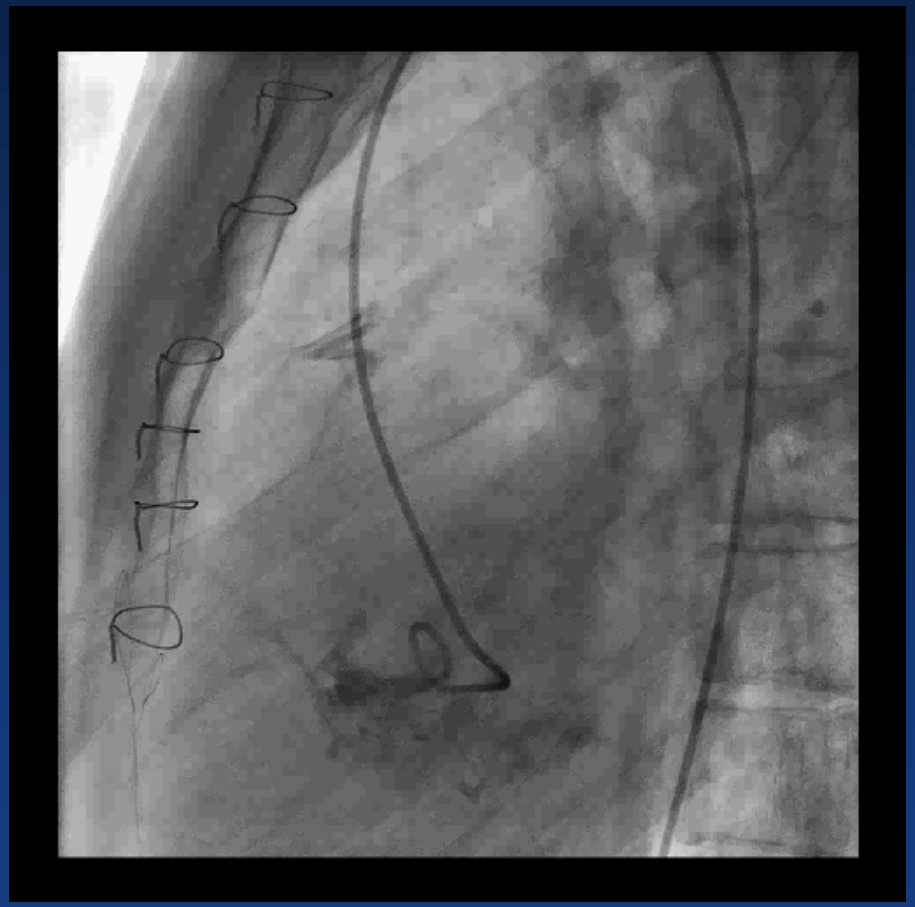
- M/19Y, s/p Fallot type DORV correction -



Case

- M/19Y, s/p Fallot type DORV correction -

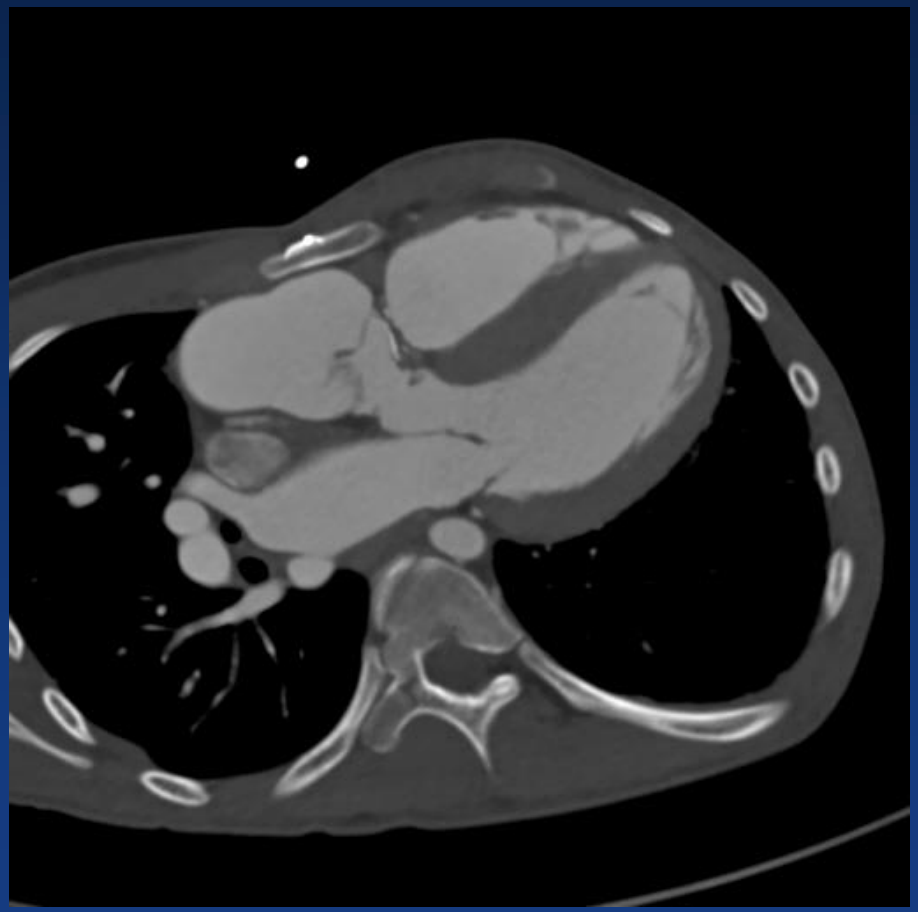


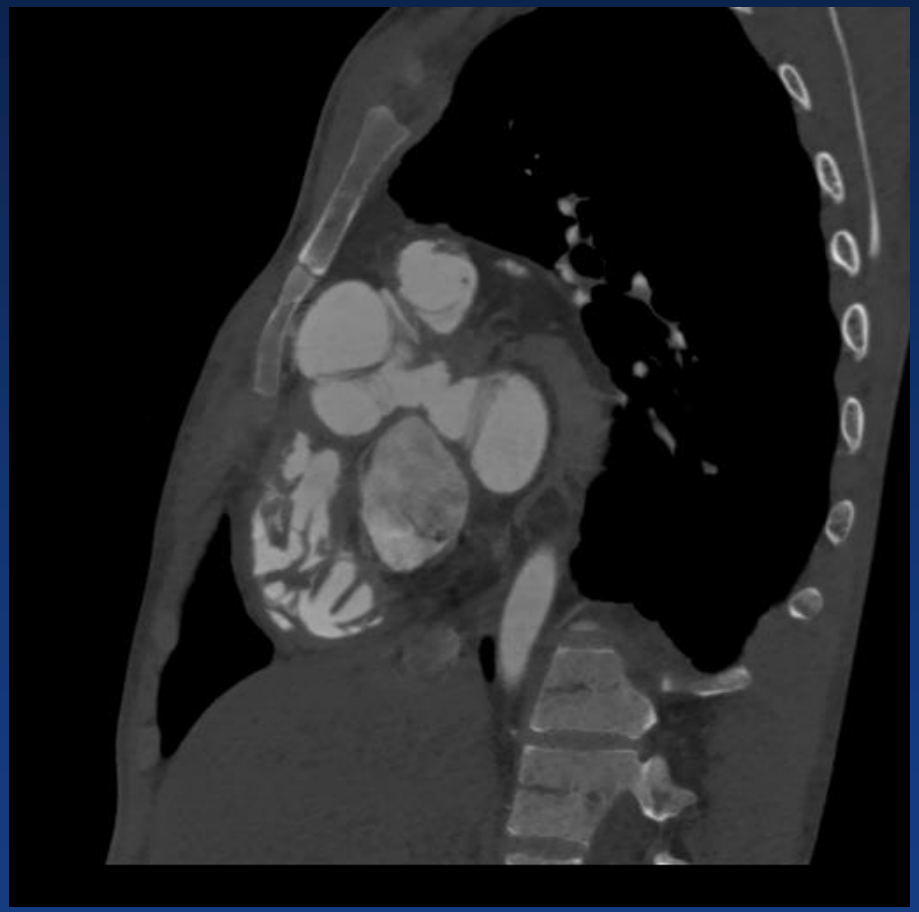


*** Peak-to-peak PG 80mmHg**

Case

- M/19Y, s/p Fallot type DORV correction -



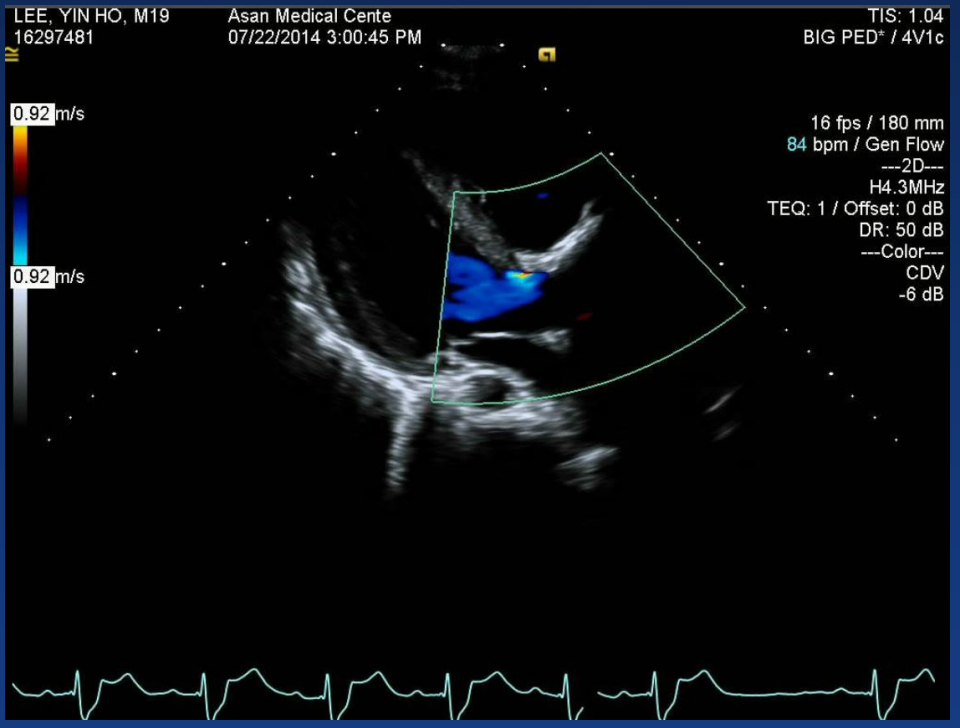


Case

- M/19Y, s/p Fallot type DORV correction -

Case

- M/19Y, s/p Fallot type DORV correction -



Case

- M/19Y, s/p Fallot type DORV correction -

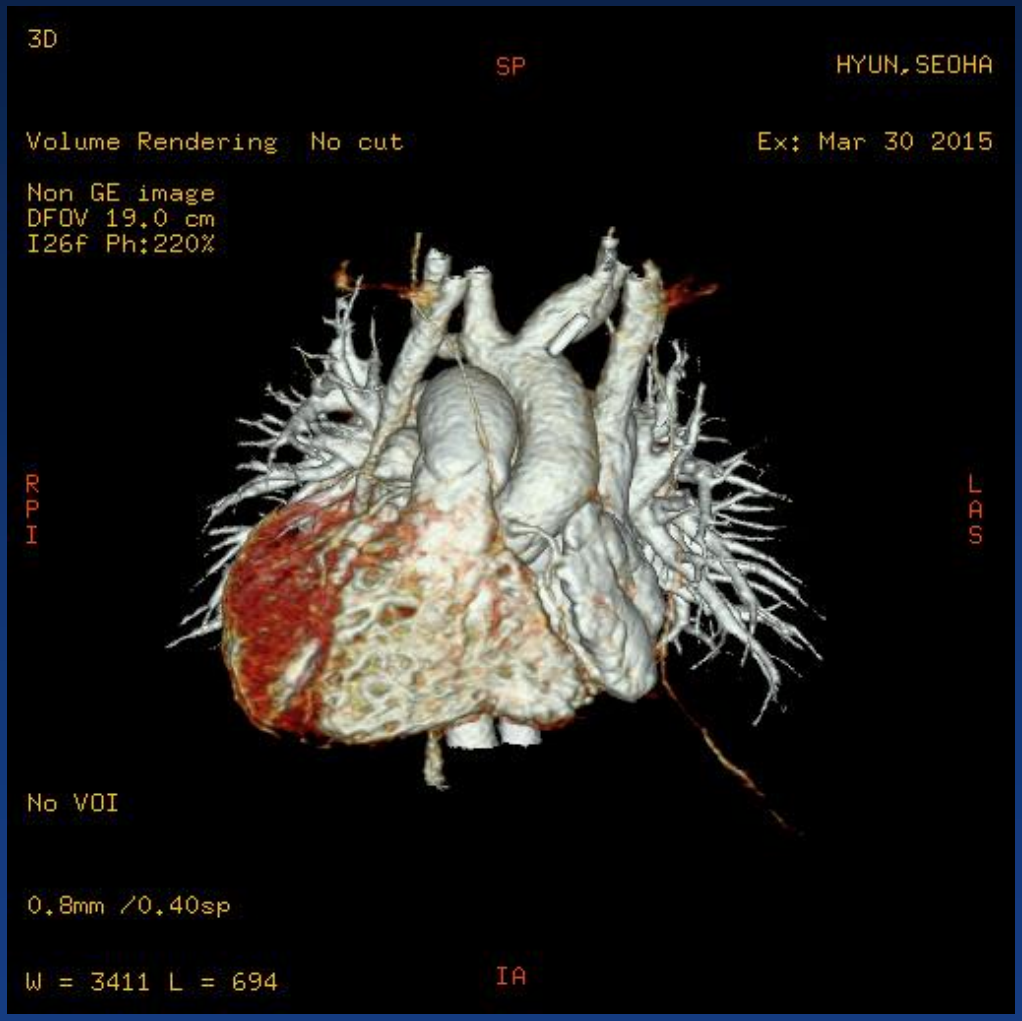
LVOTO

- Fixed SAS -

- **Surgery for fixed SAS**
  - ➢ **Diffuse SAS (the adequacy of AoV)**
    - ✓ **Adequate AoV**
      - → **Modified Konno (septal ventriculoplasty)**
    - ✓ **Small or damaged AoV**
      - → **Konno (aortoventriculoplasty) + AVR**

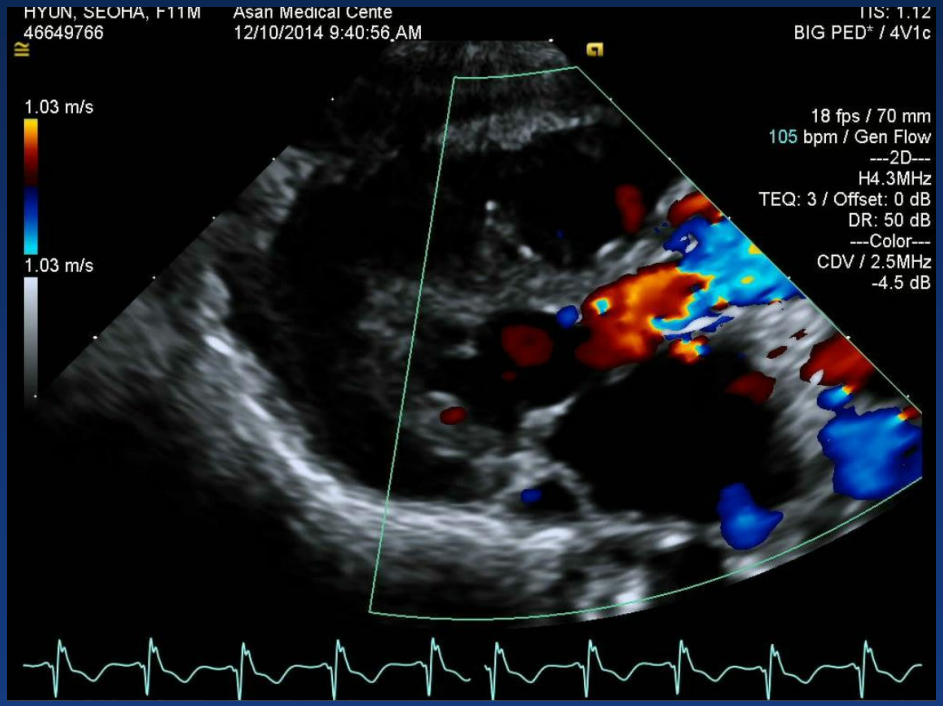
Case

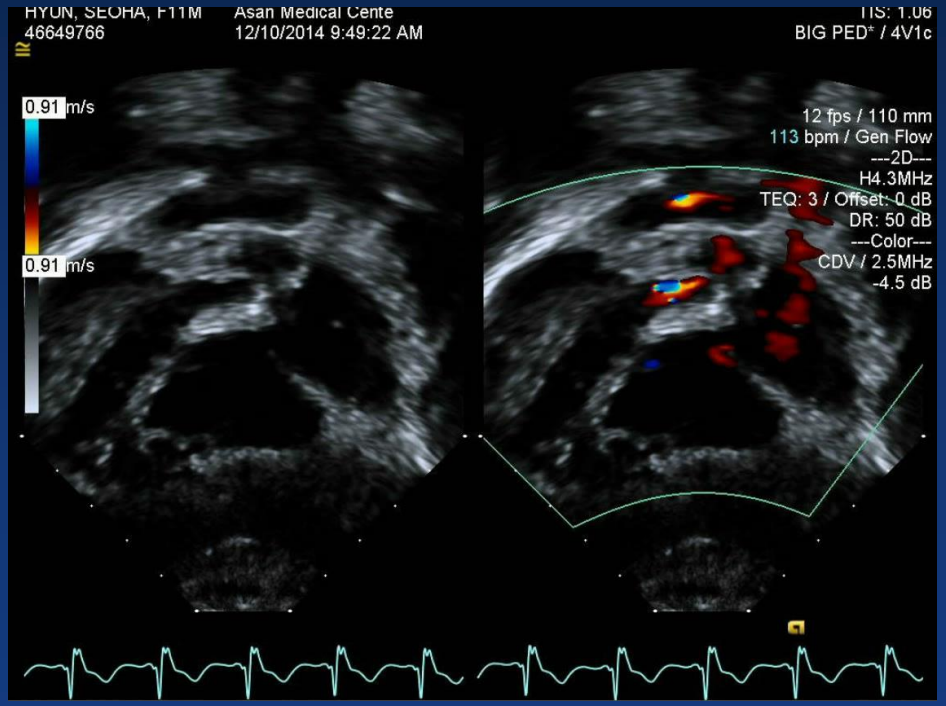
- F/1Y, s/p Total repair (CoA with VSD) -



Case

- F/1Y, s/p Total repair (CoA with VSD) -

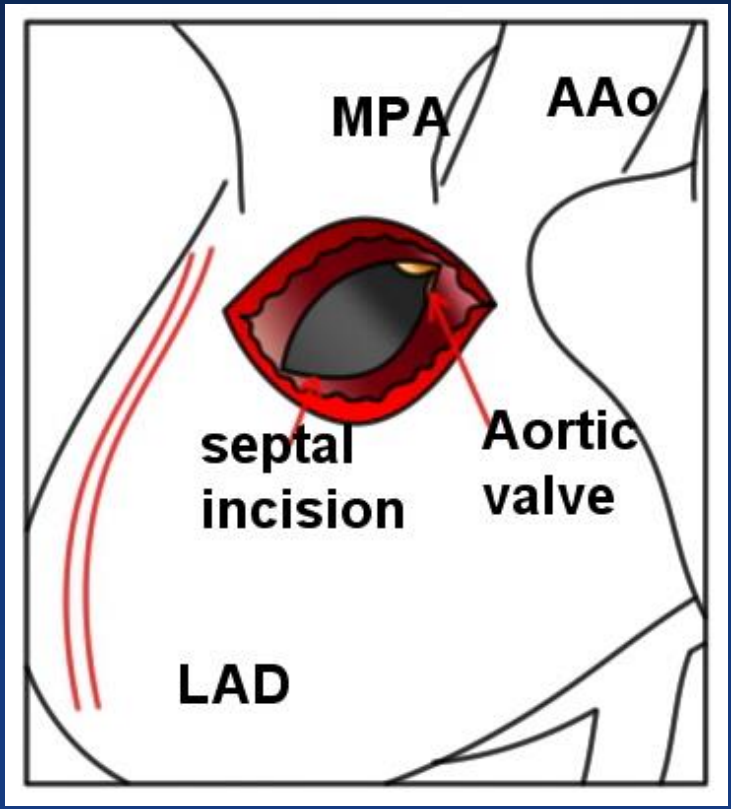




Case

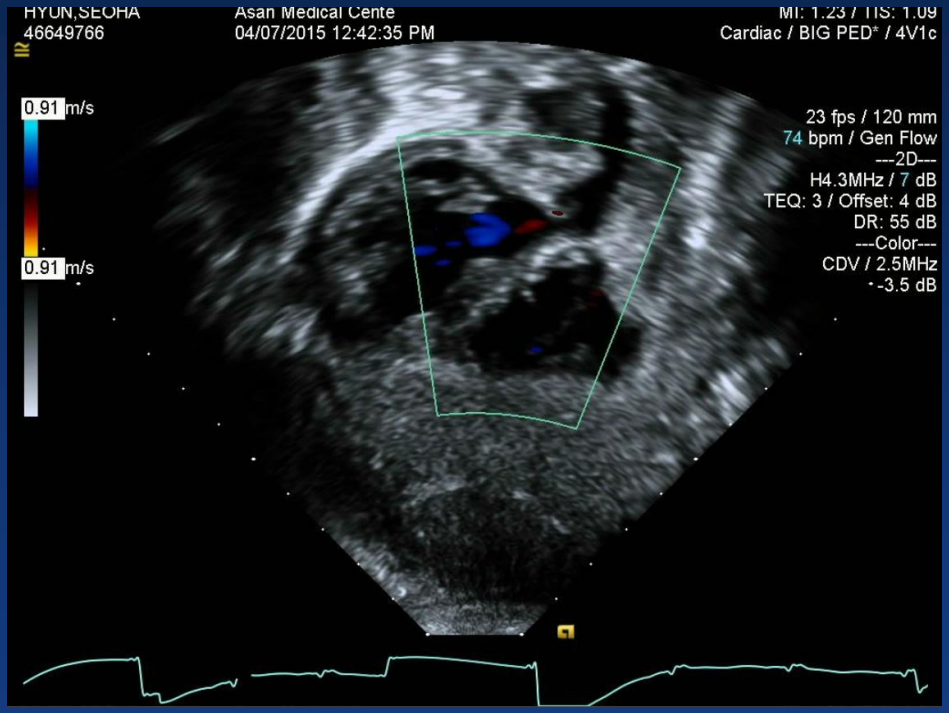
- F/1Y, s/p Total repair (CoA with VSD) -

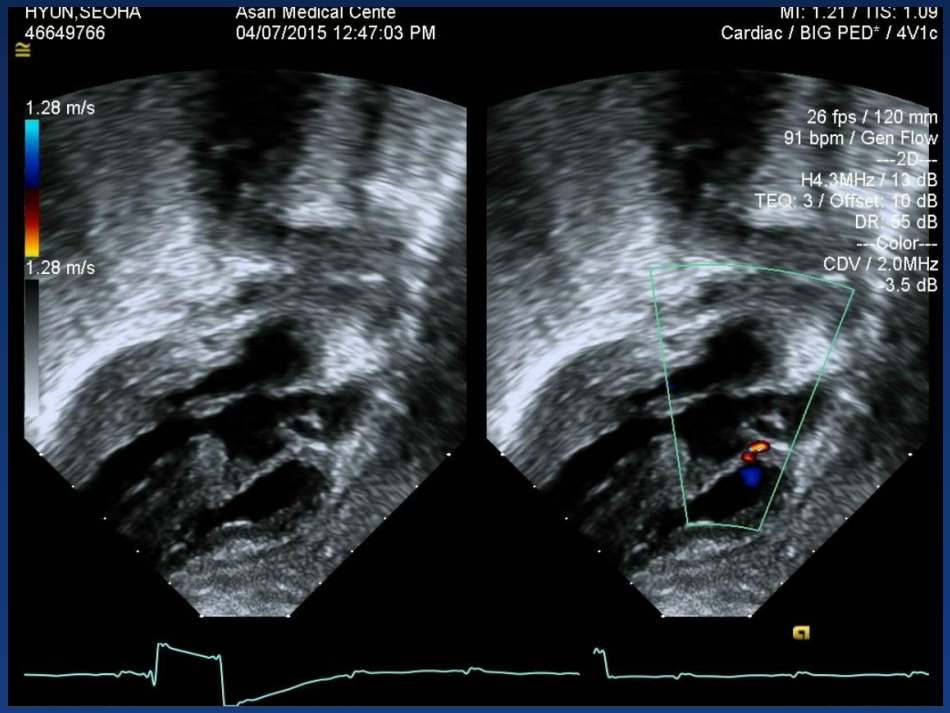
- **Modified-Konno operation**



Case

- F/1Y, s/p Total repair (CoA with VSD) -





LVOTO

- HCMP -

- **Idiopathic Hypertrophic Subaortic Stenosis (IHSS)**
  - **Asymmetric ventricular septal hypertrophy**
    - ✓ **Hypertrophic septum: fixed stenosis**
    - ✓ **SAM of anterior mitral leaflet: dyanamic stenosis**
  - **Septal myectomy**

    *** Konno, MVR, …**

LVOTO

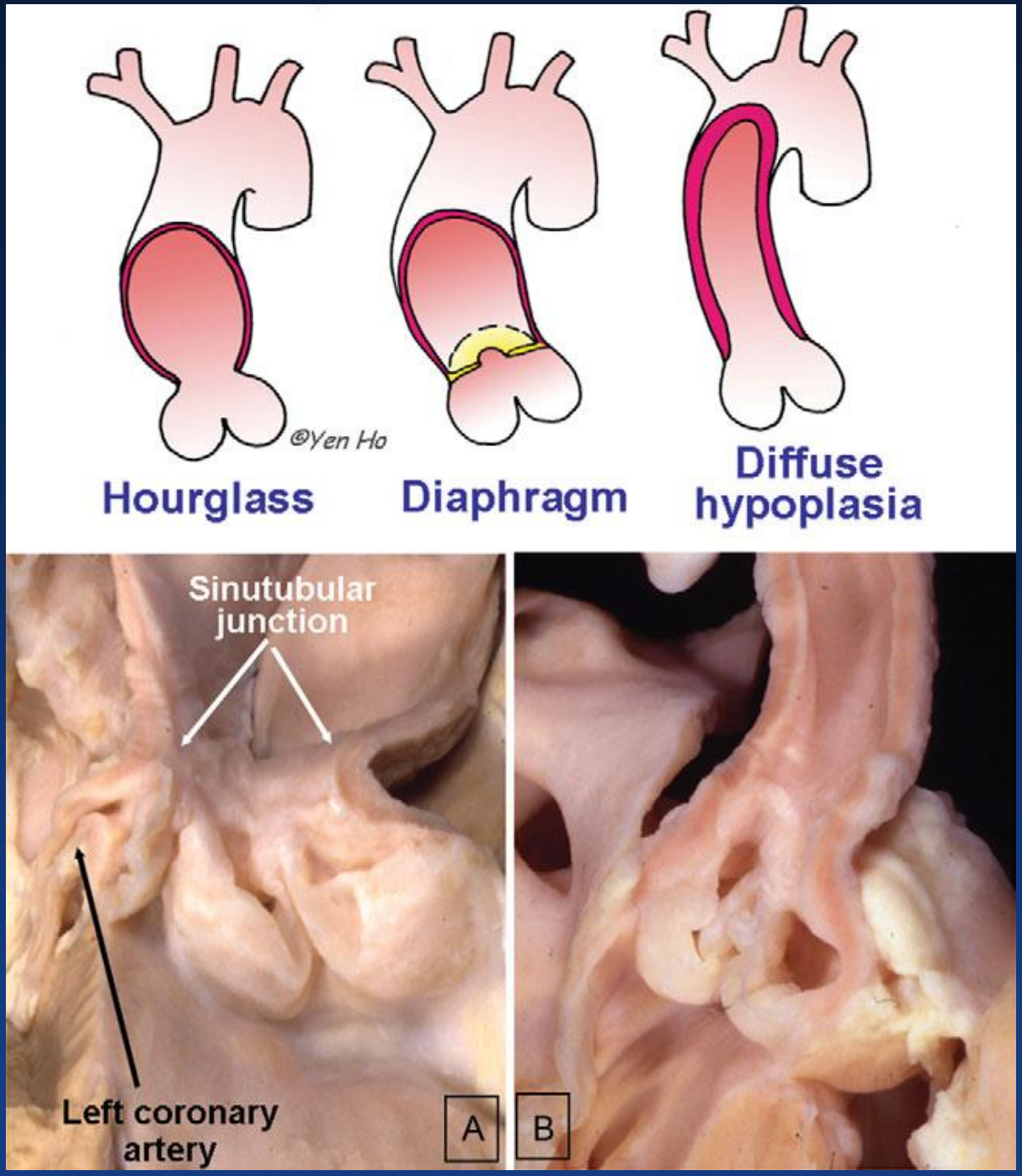
- Supravalvar -

- Narrowing of the aortic lumen above the aortic valve
- 5~10% of the LVOTO
- Often part of Williams' syndrome
- Frequently branch PA involvement

LVOTO

- Supravalvar -

- **Pathologic features**
  - **Great arteries**
    - ✓ **Localized (80%) vs. diffuse (20%)**
    - ✓ **Intimal hyperplasia, medial dysplasia,…**
    - ✓ **Pulmonary arterial involvement: lesser degree**


©Yen Ho
Hourglass
Diaphragm
Diffuse hypoplasia
Sinutubular junction
Left coronary artery
A
B

LVOTO

- Supravalvar -

- **Pathologic features**
  - **Coronary arteries**
    - ✓ Markely dilated and tortous (↑ root pressure)
    - ✓ ST ridge thickening → coronary ostial narrowing
  - **Valve abnormalities**
    - ✓ AoV: morphologically normal in most cases
    - ✓ MV and subV apparatus: fibrous thickening

LVOTO

- Supravalvar -

- **Pathophysiology**
  - **Difference from the other forms of LVOTO**
    - **coronary artery exposed to high systolic pressure**
    - **Premature coronary artery disease**

→ Myocardial hypoperfusion → Vf and sudden death

LVOTO

- Supravalvar -

- **Diagnosis**
  - ➢ **Williams' syndrome**
    - ✓ **Deletion on chromosome 7q11.23**
    - → **Affect the elastin gene**
  - ➢ **Non williams supravalvar AS**
    - ✓ **Loss-of-function of point mutation of the same elastin gene**

LVOTO

- Supravalvar -

- **Diagnosis**
  - **Echocardiography**
  - **Cardiac CT or MRI**
  - **Cardiac cath. and angiography**

LVOTO

- Supravalvar -

- **Indication for surgery**
  - **Symptoms**
  - **LVOT gradient >40~50mmHg**
  - **Early surgery**
    - ✓ **Progressive nature of disease**
    - ✓ **Detrimental effect on LV, aortic valve and coronary artery + myocardial hypoperfusion**

LVOTO

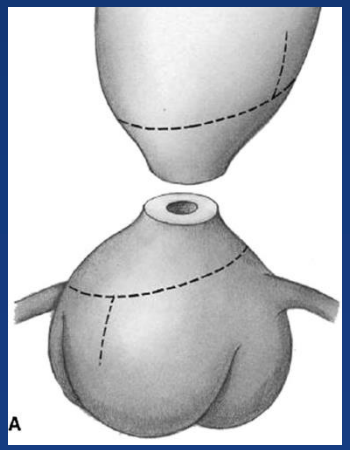
- Supravalvar -

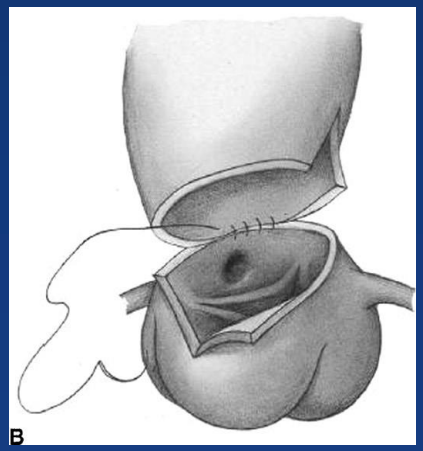
- **Surgical treatment**
  - **Widening of obstructive segment**
    - ✓ **With vs. without patch**
  - **Adressing coronary arteries**
    - ✓ **Complete resection of abnormal tissue around coronary os.**
    - ✓ **Patch angioplasty or CABG**

LVOTO

- Supravalvar -

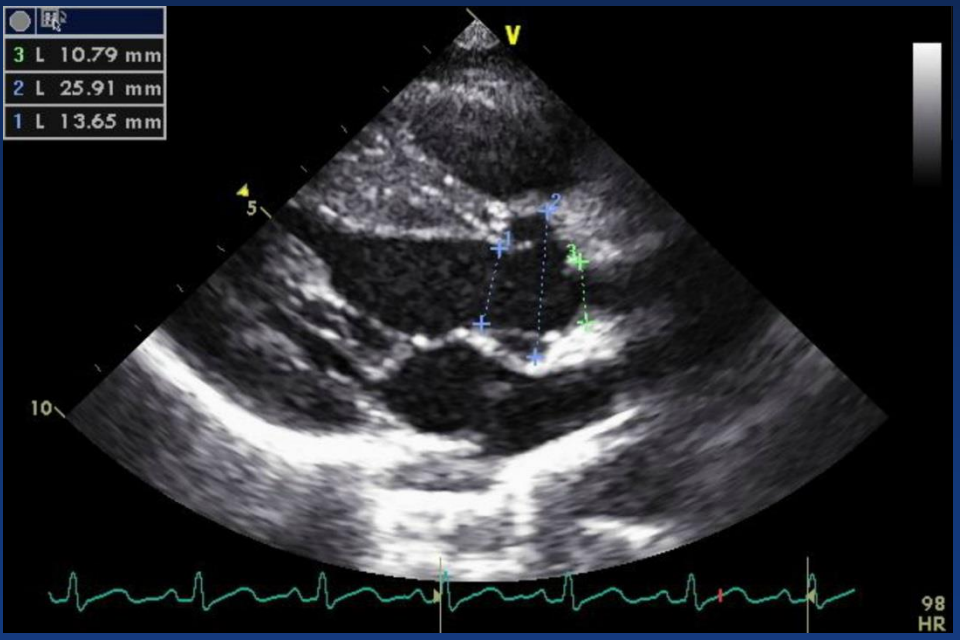
- **Surgery for localized disease**
  - ➢ **Single patch**
    - ✓ **One sinus vs. two sinuses**
  - ➢ **Separate multiple patches: Brom**
  - ➢ **No patch**
    - ✓ **Myer**
    - ✓ **Simple sliding**

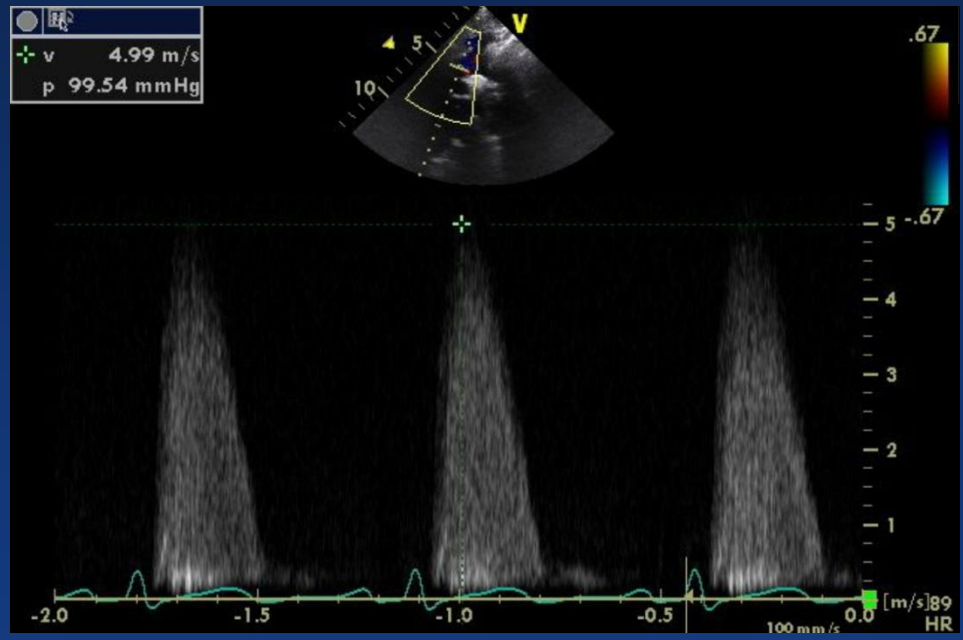




Case

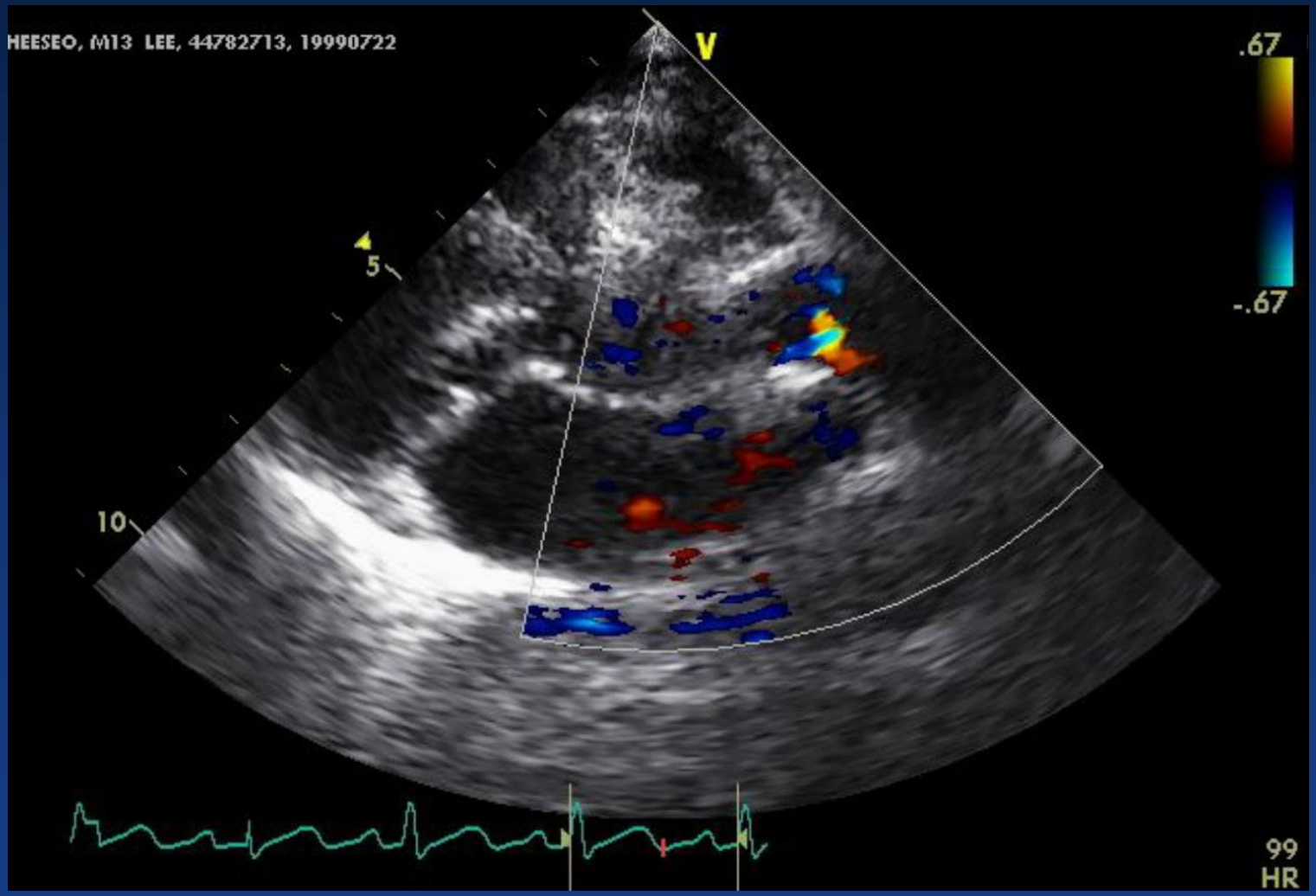
- M/13Y, SVAS(localized) -





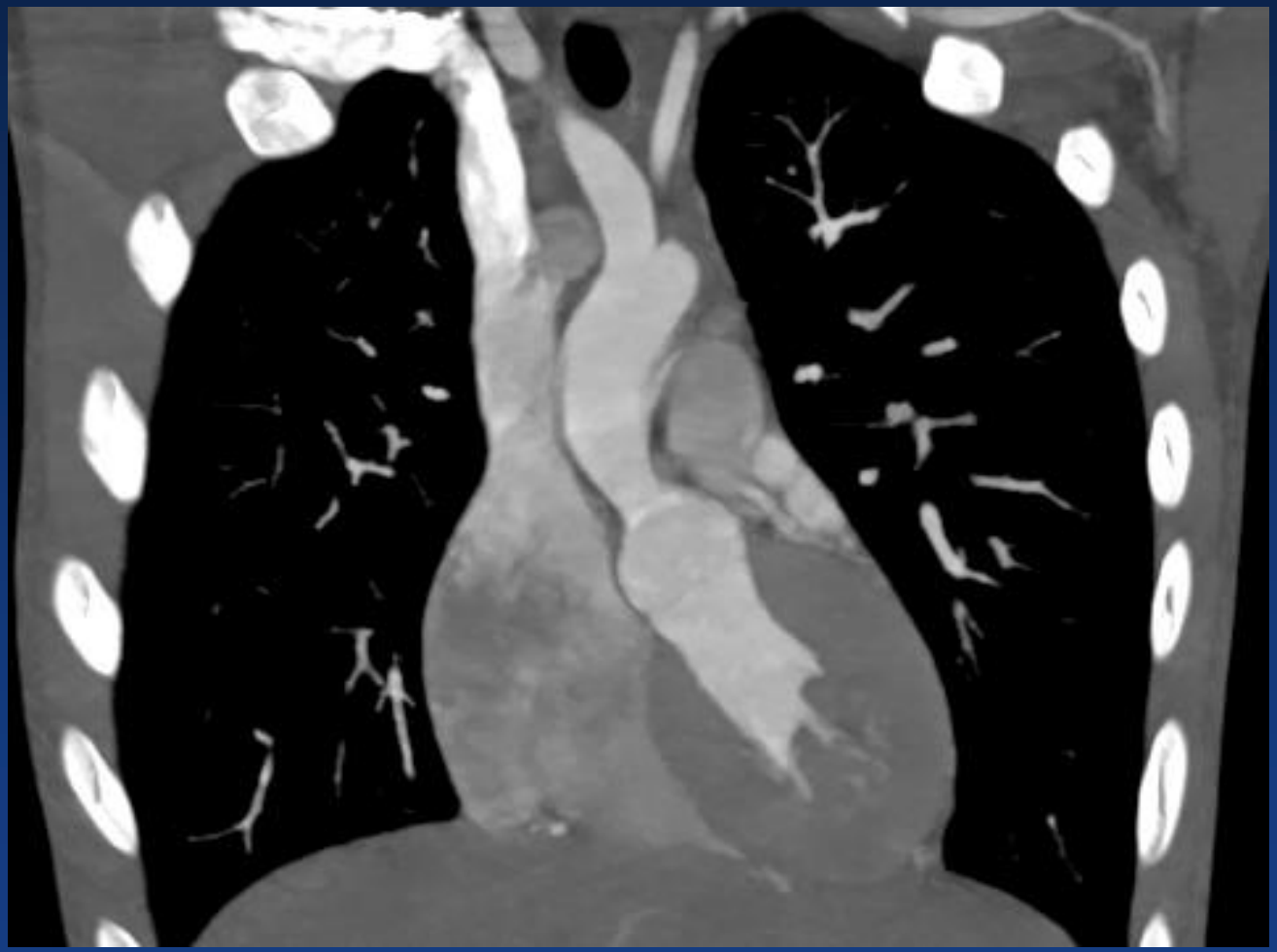
Case

- M/13Y, SVAS(localized) -



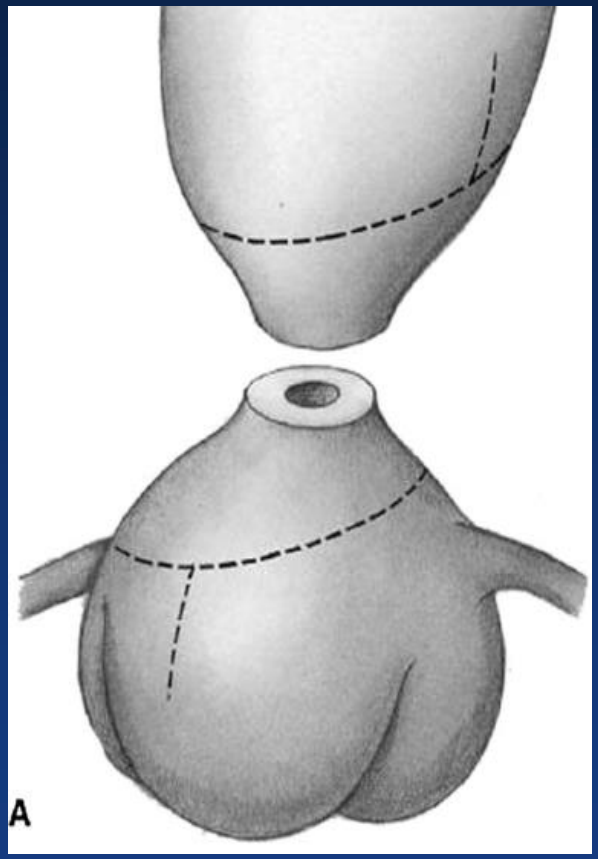
Case

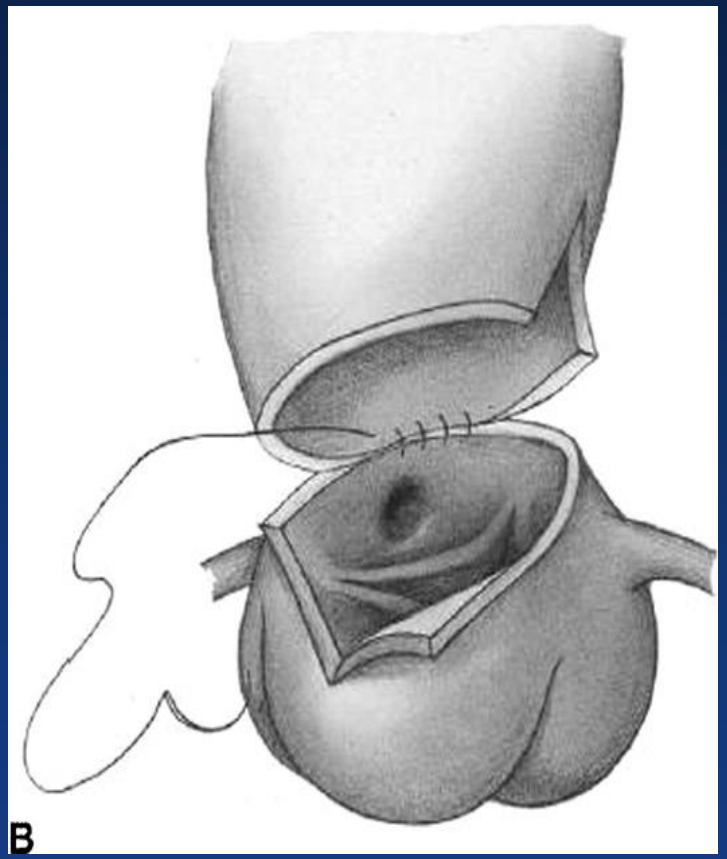
- M/13Y, SVAS(localized) -



Case

- M/13Y, SVAS(localized) -

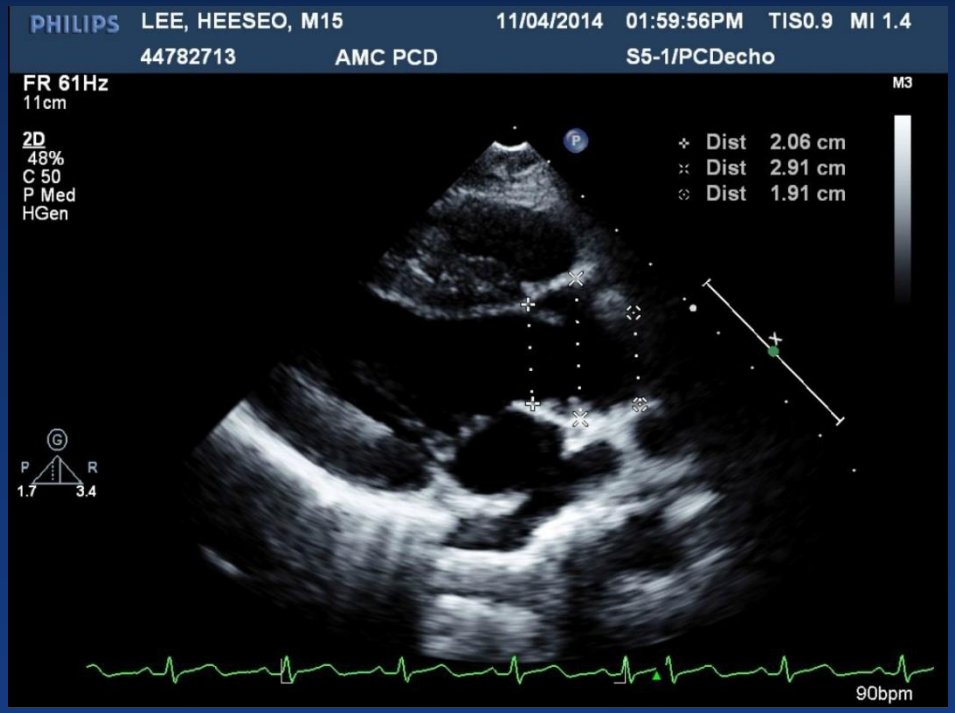


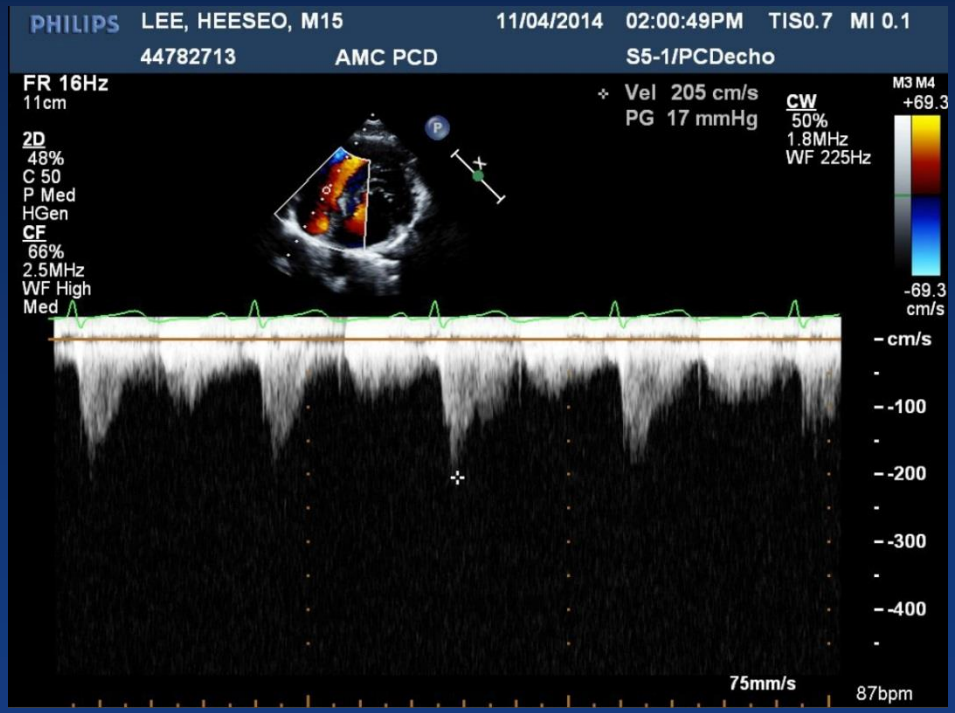


- **Simple sliding aortoplasty**

Case

- M/13Y, SVAS(localized) -





Case

- M/13Y, SVAS(localized) -

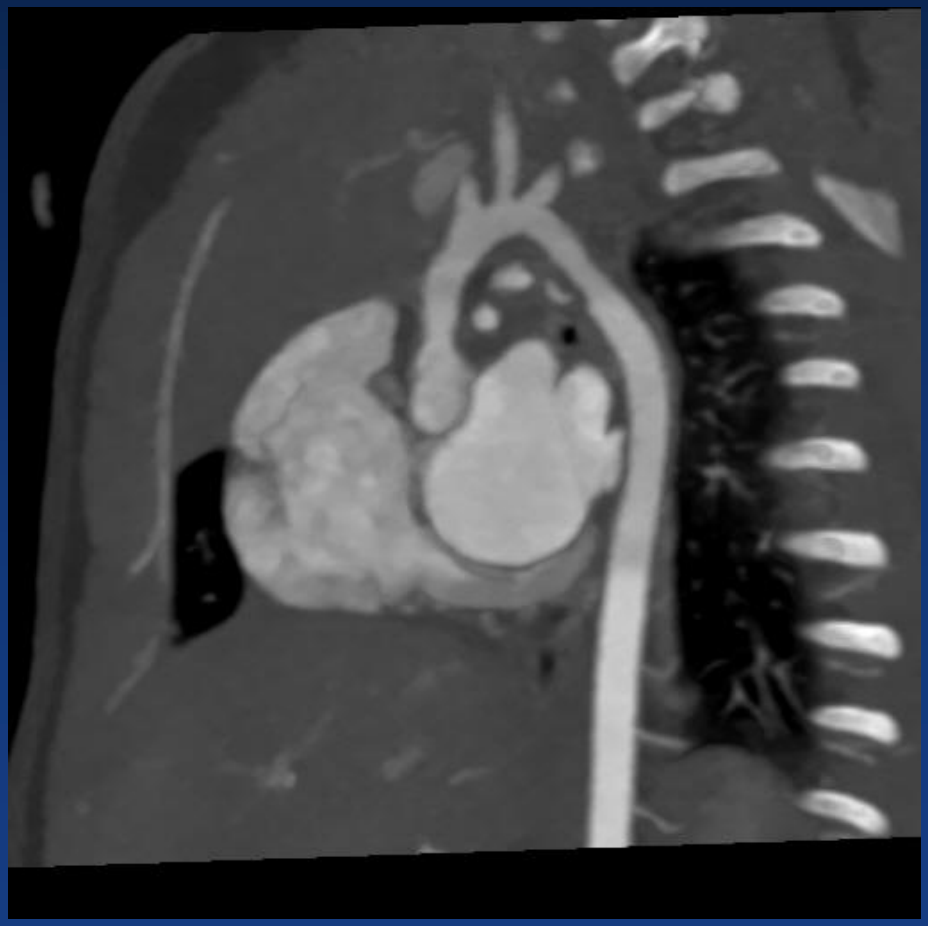
LVOTO

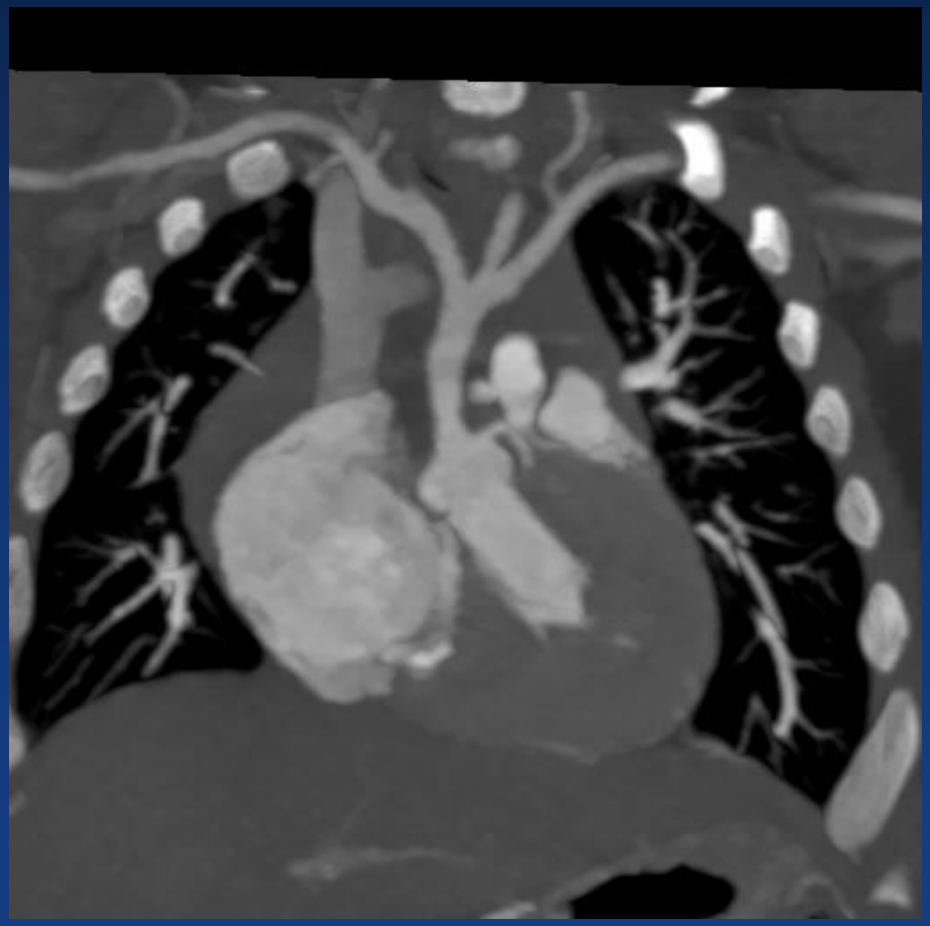
- Supravalvar -

- **Surgery for diffuse disease**
  - **Extended patch aortoplasty**
  - **Perfusion strategy**

Case

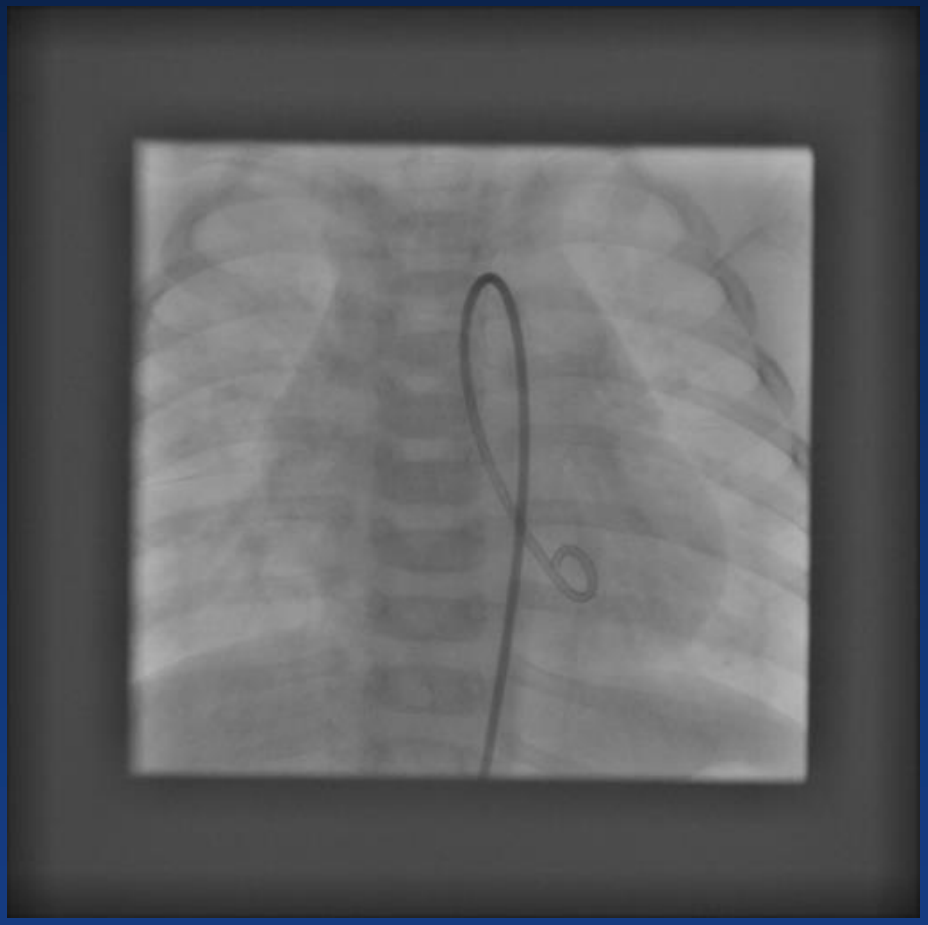
- M/8m, SVAS(diffuse) -

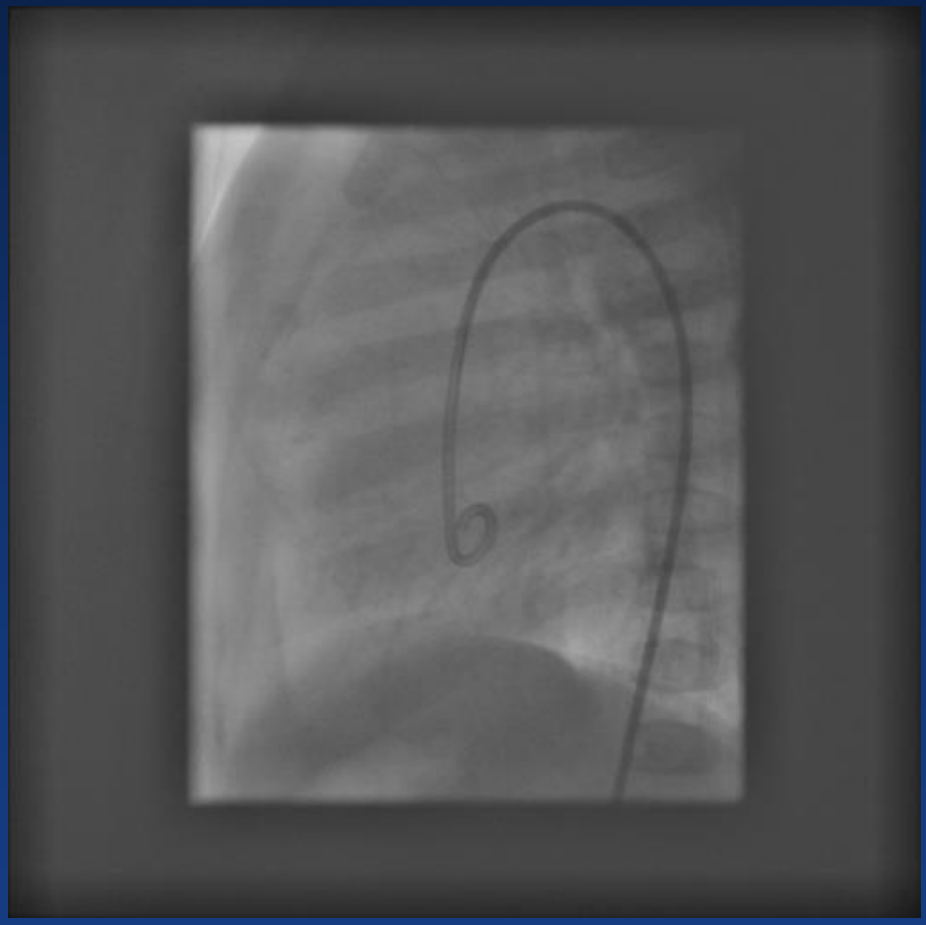




Case

- M/8m, SVAS(diffuse) -





Case

- M/8m, SVAS(diffuse) -

- **Simple sliding aortoplasty**
  + patch aortoplasty

Case

- M/8m, SVAS(diffuse) -