



Childrens Heart Center Nevada

continuous care for the fetus, child & adult with congenital heart disease

Neonatal Critical Aortic Stenosis

Prenatal to Adolescence

Group Discussion

21st Annual Heart Conference

October 28, 2017



Childrens Heart Center Nevada

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Neonatal Critical Aortic Stenosis

Prenatal Detection

2003-2006 Rate: 36%

Prenatal Detection of Congenital Heart Disease in Southern Nevada

The Need for Universal Fetal Cardiac Evaluation

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Objective. Congenital cardiac malformations are common developmental anomalies. In the United States, congenital heart disease is the number one cause of infant mortality from congenital malformations. Prenatal diagnosis of congenital heart defects aids treatment coordination. Our aim was to evaluate prenatal detection of serious congenital heart malformations in Clark County, Nevada. **Methods.** We electronically searched our research department-maintained computer database for patients with serious congenital heart disease born in Clark County between May 2003 and April 2006. We excluded patients that did not have at least 1 local prenatal ultrasound examination. All prenatal ultrasound studies were performed in obstetric offices, radiology imaging centers, or maternal-fetal medicine specialty practices. Fetal echocardiography was performed in maternal-fetal medicine specialists' offices under the supervision of a fetal cardiologist. Pediatric cardiologists performed all postnatal echocardiographic examinations. **Results.** During the study period, we diagnosed serious congenital heart malformations in 161 patients among a total estimated 77,000 births (2/1000). Of the 161 patients, 58 (36%) had a prenatal diagnosis, and 103 (64%) had an exclusively postnatal diagnosis. **Conclusions.** Standard prenatal ultrasound fails to show congenital heart disease in most fetuses. **Key words:** congenital heart disease; fetal echocardiography; prenatal diagnosis; prenatal ultrasound.

Abbreviations
AF, atrioventricular

Received July 11, 2007, from the Children's Heart Center Las Vegas, Nevada USA (R.J.A., W.N.E., C.F.L., R.R., K.T.K., J.C.C., H.R.); Departments of Pediatrics (R.J.A., W.N.E., C.F.L., R.R., K.T.K., H.R.) and Obstetrics and Gynecology (J.A., D.R., A.J.S.), University of Nevada, School of Medicine (J.C.C.), Las Vegas, Nevada USA; Desert Perinatal Associates, Las Vegas, Nevada USA (J.A.); Center for Maternal-Fetal Medicine, Las Vegas, Nevada USA (B.K.I.); Department of Obstetrics and Gynecology, University Medical Center, Las Vegas, Nevada USA (D.R.); and Alan J. Sacks Maternal-Fetal Medicine, Las Vegas, Nevada USA (A.J.S.). Revision requested August 7, 2007. Revised manuscript accepted for publication August 21, 2007.

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Article includes CME test

2012-2014 Rate: 71%

Moving Towards Universal Prenatal Detection of Critical Congenital Heart Disease in Southern Nevada: A Community-Wide Program

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Abstract This study compares the current, prenatal detection rate for critical congenital heart disease in Southern Nevada with the previously reported rate, after developing and expanding a comprehensive, community-wide fetal cardiology program. For the current-period analysis, we inquired our database and electronic health records for patients born in Clark County, Nevada, with critical congenital heart disease between May 2012 and April 2014, and we compared the results with the previous period between May 2003 and April 2006. The major components of the community-wide program include fetal congenital heart disease screening via general obstetric ultrasound studies performed in obstetrician's offices, radiology imaging centers, or maternal-fetal medicine specialty practices; subsequent referral for comprehensive fetal echocardiography performed in maternal-fetal medicine offices under the on-site supervision by fetal cardiologists; and recurring community educational programs teaching the 5-axial plane, fetal echocardiographic

screening protocol to general obstetric sonographers and instructing perinatal sonographers in advanced imaging topics. For the current period, the prenatal detection rate for critical congenital heart disease in Southern Nevada was 71 versus 36 % for the previous period ($p < 0.001$). The temporal improvement in prenatal detection of critical congenital heart disease may be related to our expanded decentralized, community-wide fetal cardiology program, and our experiences may be applicable to other metropolitan areas.

Keywords Congenital heart disease · Fetal echocardiography · Prenatal diagnosis · Prenatal ultrasound

Introduction

In contrast to postnatal physical exam and screening newborn oximetry, prenatal fetal echocardiography is both sensitive and highly specific for the anatomical details of most critical congenital heart disease (CCHD) malformations [24, 27, 34]. Prenatal detection allows for coordinated, cardiac center care for both fetus and neonate with significant cardiac abnormalities [5]. Lack of prenatal detection may result in costly, risky, emergent neonatal transfers from under-equipped delivery locations to congenital heart centers, unexpected postnatal parental stress, or even neonatal death from missed CCHD [6, 14, 23].

Nevertheless, prenatal detection rates for CCHD remain low for general populations in the USA, in part, because fetal echocardiography is most often reserved for women with risk factors for fetal heart disease; yet, most newborns with CCHD are born to mothers without risk factors [7, 29]. Women without risk factors have usually undergone fetal cardiac imaging primarily limited to a 4-chamber

The submission is with the full knowledge and approval of the listed co-authors.

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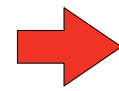
Published online: 08 August 2014

CHCN Fetal Cardiac Program

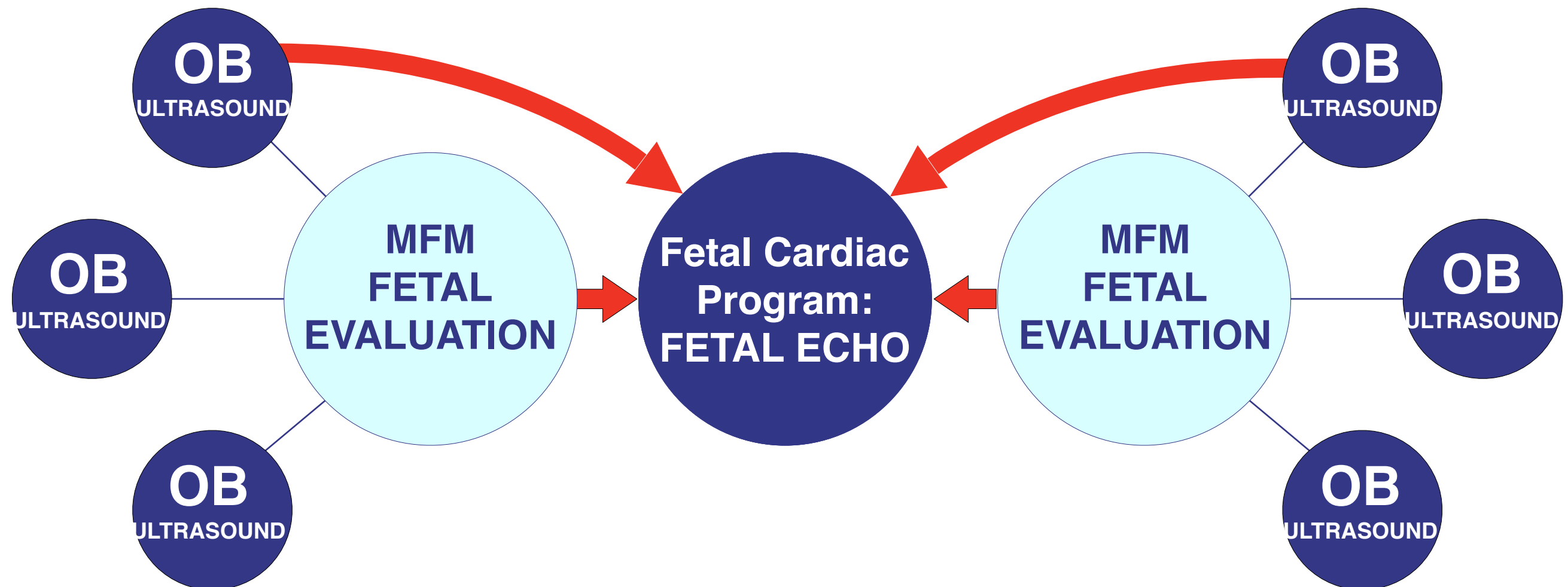
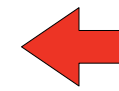
2 Fundamental Principles

- 1) Fetal cardiologists are present onsite daily in MFM offices
- 2) Ongoing education for the community's sonographers
 - Multiplanar screening for general OB/imaging center techs
 - Advanced courses for perinatal sonographers
 - All courses free & include CEUs

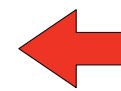
Usual Fetal Cardiac Program



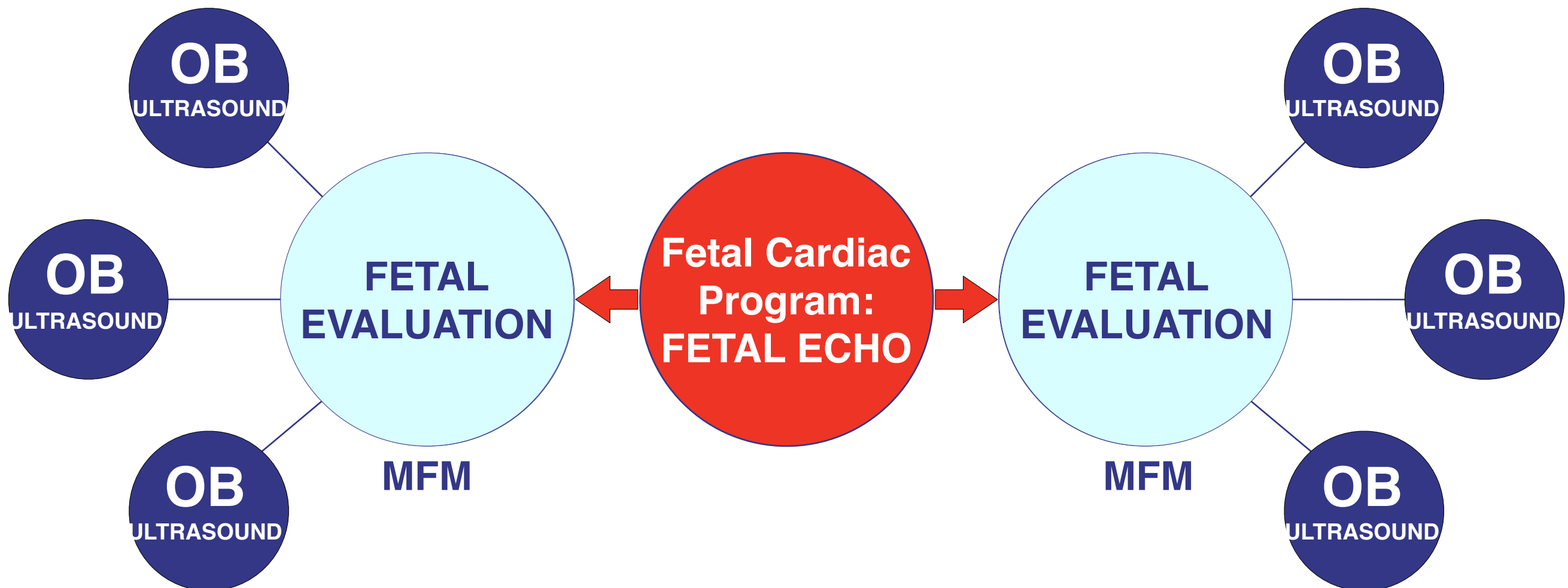
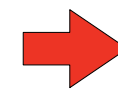
Centralized

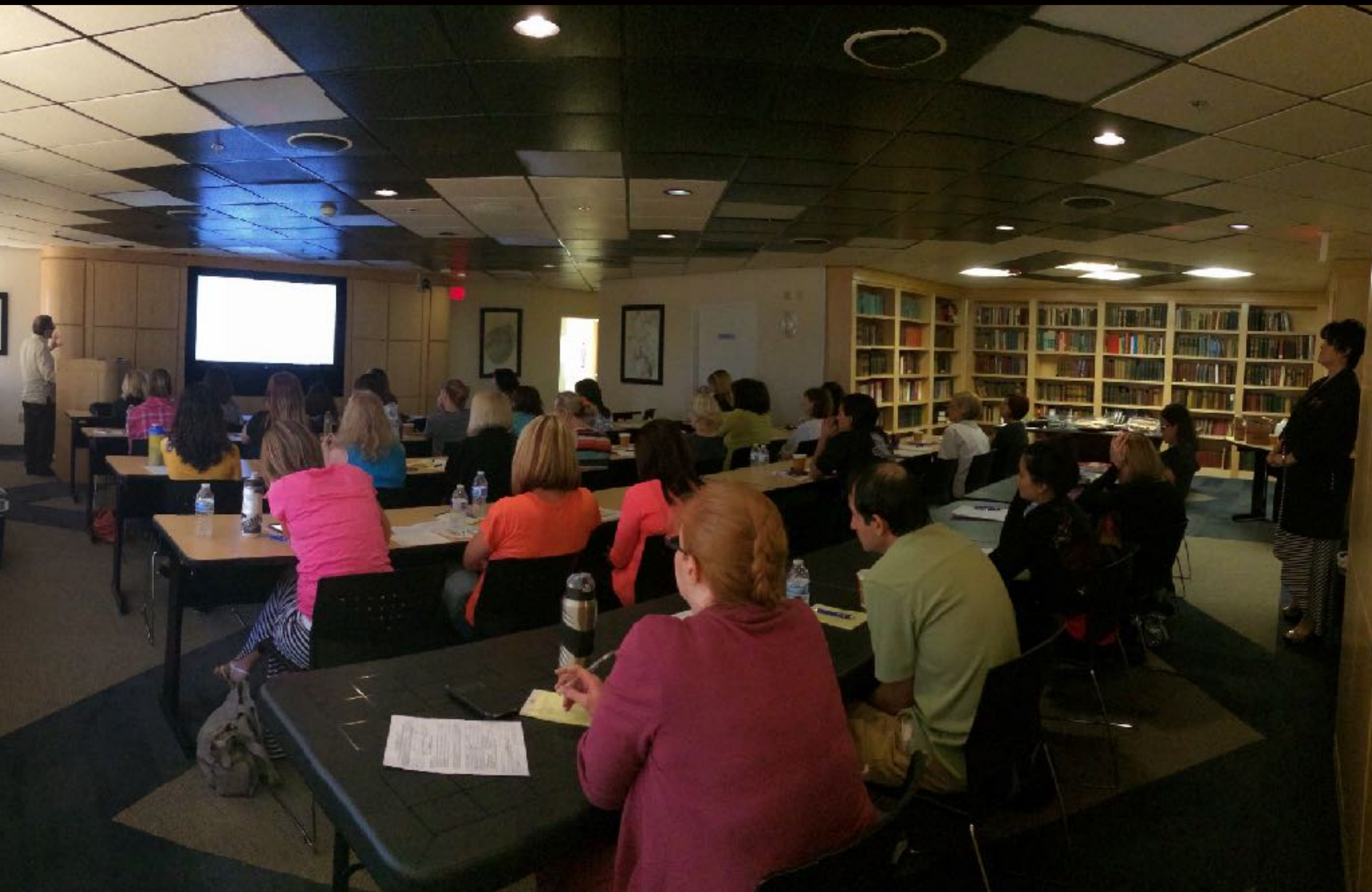


CHCN Fetal Cardiac Program

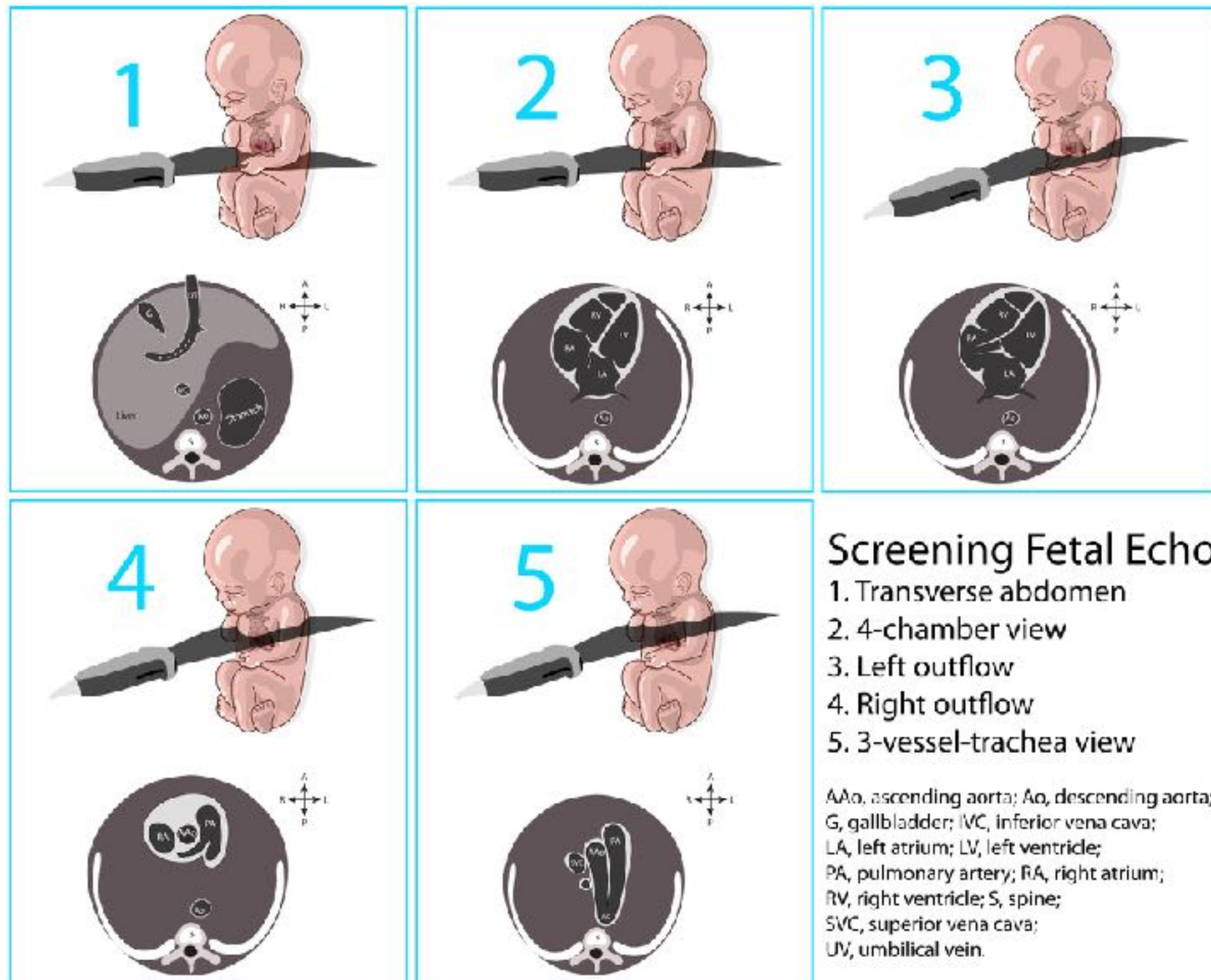


Decentralized





Yagel et al (2001) Ultrasound Obstet Gynecol





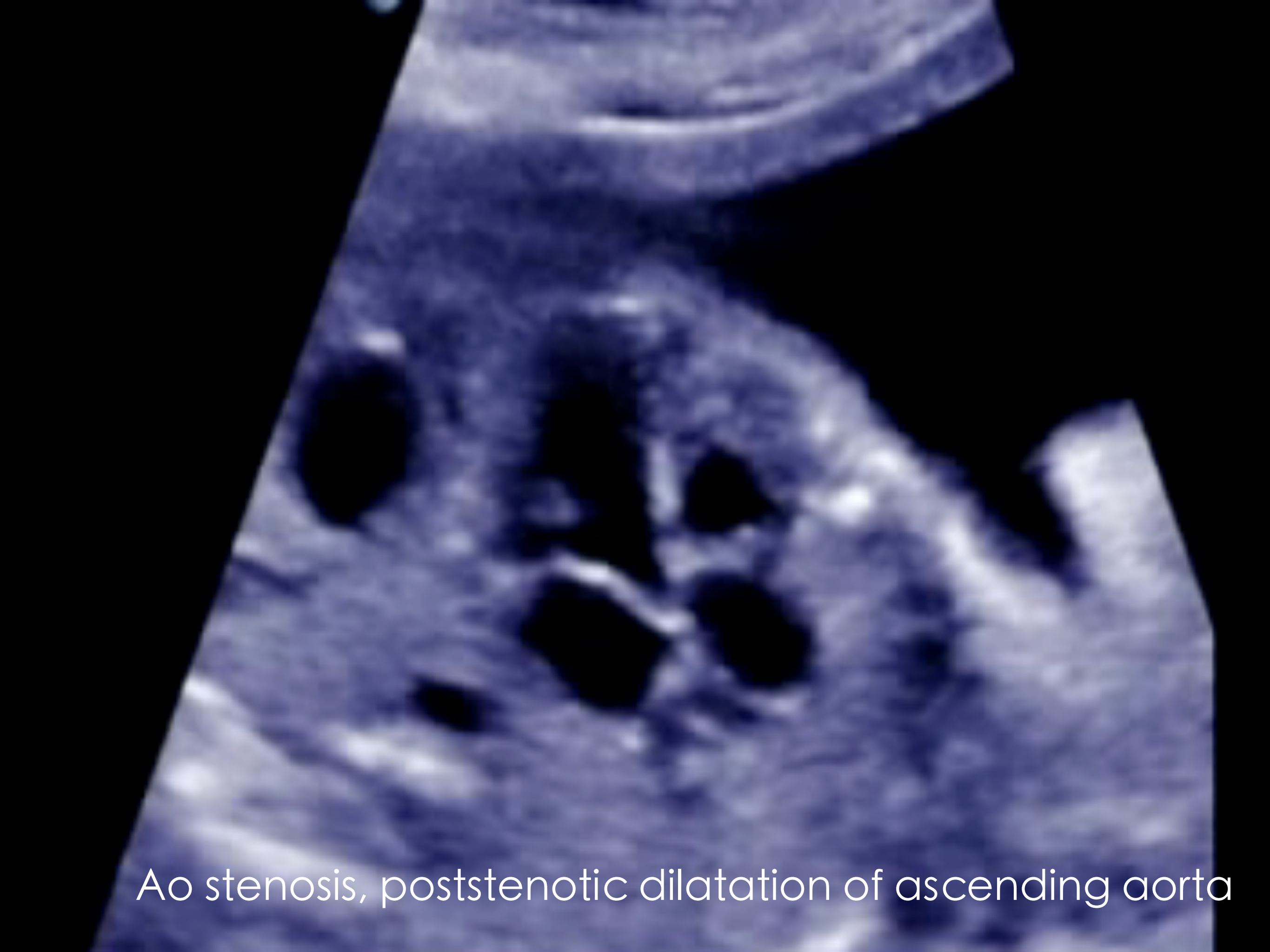


CHCN Congenital Cardiac Program

Prenatal Detection 2016-2017

- 124 patients needing medical/interventional/surgical RX
- 104 (84%) detected prenatally
- Estimate 3/1000 born with significant CHD
- NV: 40,000 births = ~120/year with significant CHD





Ao stenosis, poststenotic dilatation of ascending aorta



Decreased LV function



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Neonatal Critical Aortic Stenosis

Fetal Intervention

CLINICAL HISTORY

- L. K.
- EGA 22 WEEKS, 4 DAYS
- Dedicated fetal cardiac exam by echo
 - Severe aortic stenosis with velocity 3 m/sec
 - Moderate mitral regurgitation with velocity 3 m/sec
 - Good size ventricles left ventricular systolic function preserved “at this point”



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CHART NOTE

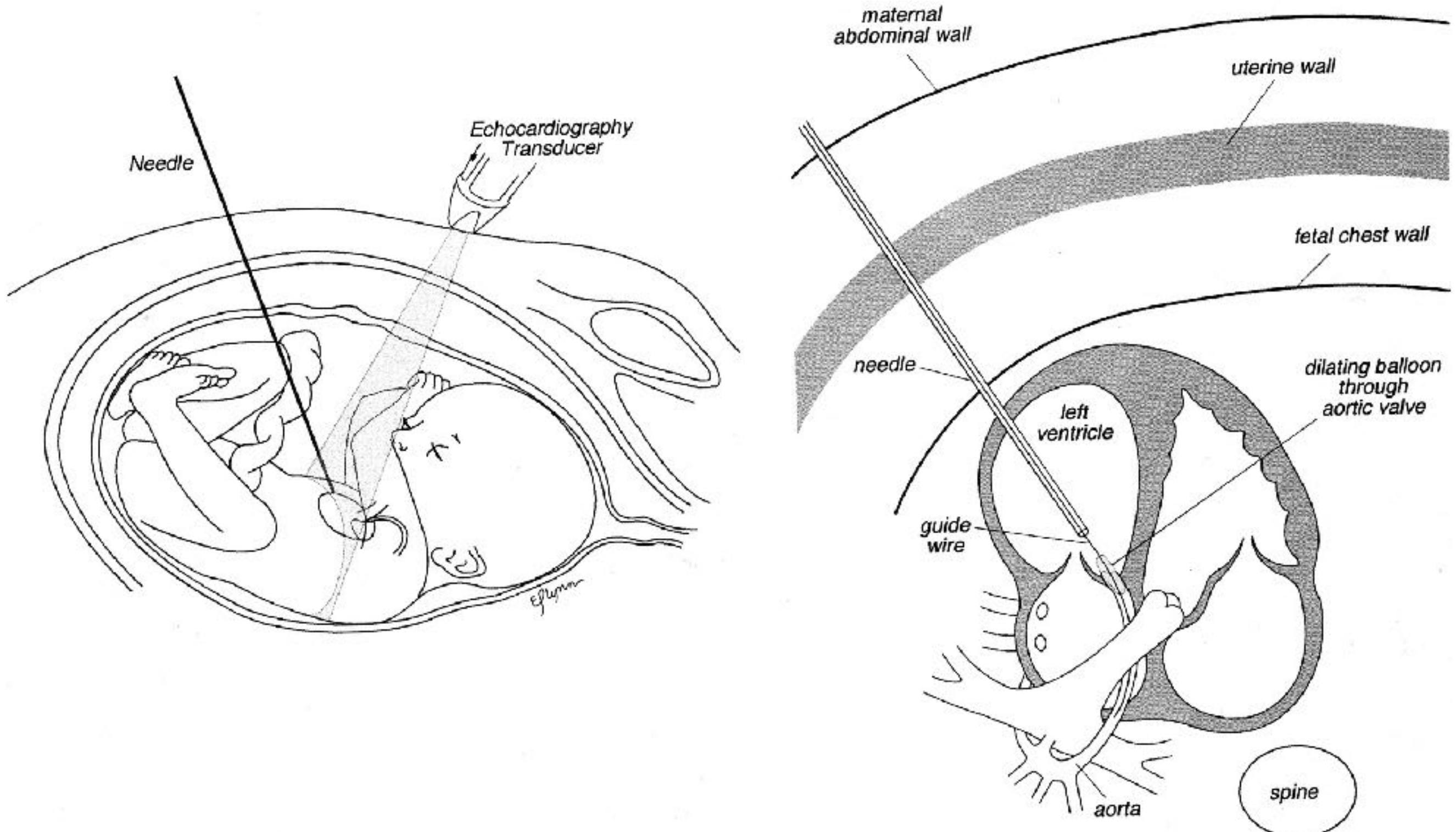
DATE: March 23, 2011
DOB: 10/04/74
EDD: 07/03/11
EGA: 25 weeks 3 days



DISCUSSION: I met with Mrs. [redacted] and her husband at the Childrens Heart Center – Nevada offices today. She is carrying a fetus with critical valvar aortic stenosis and evidence of retrograde flow in the aorta and at the level of the atria. It was proposed to Mr. and Mrs. [redacted] that a fetal aortic valvuloplasty be attempted. They have agreed with the procedure and came in to discuss the risks, benefits, and alternatives. I went over the procedure itself and answered their questions. The procedure has been approved by the IRB of Sunrise Hospital. It is scheduled to be performed next week. The mother was provided with the consent form to take home and study. She will return it in two days when she will be reevaluated by my colleague Dr. Acherman.

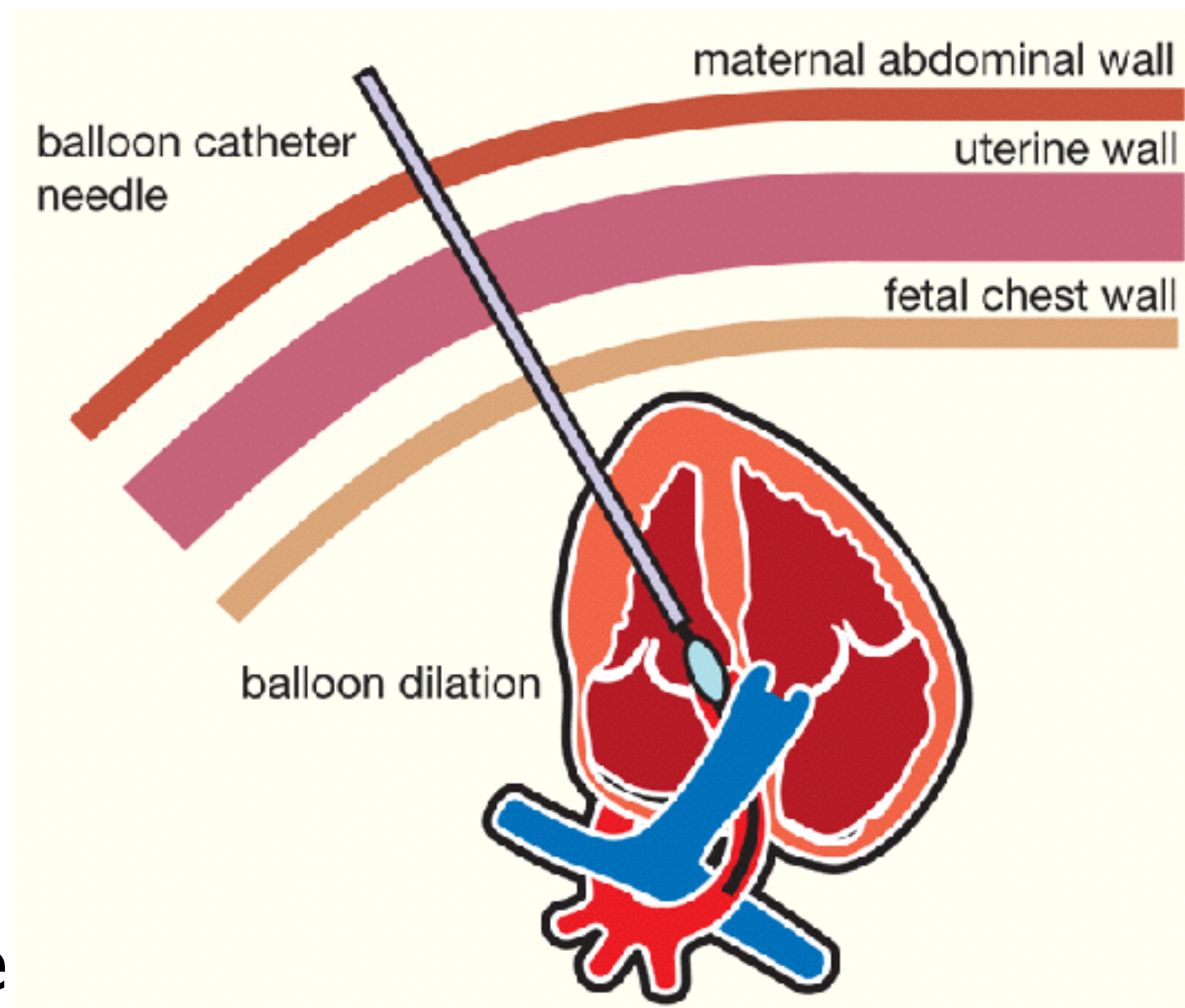
Abraham Rothman, M.D.

Fetal Intervention



CLINICAL HISTORY

- EGA 27 Weeks, 2 Days
- Dedicated fetal cardiac exam
 - Peak systolic velocity 1.8 m/sec
 - Mod-severe mitral valve regurgitation with severe LA enlargement
 - Moderately depressed LV function with EFE
 - Aortic arch flow bidirectional, equal retrograde and antegrade





Needle in LV



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Neonatal Critical Aortic Stenosis

Neonatal Ross-Konno

BIRTH DATA

Weight 3.6 Kg

2D echo

Good LV function

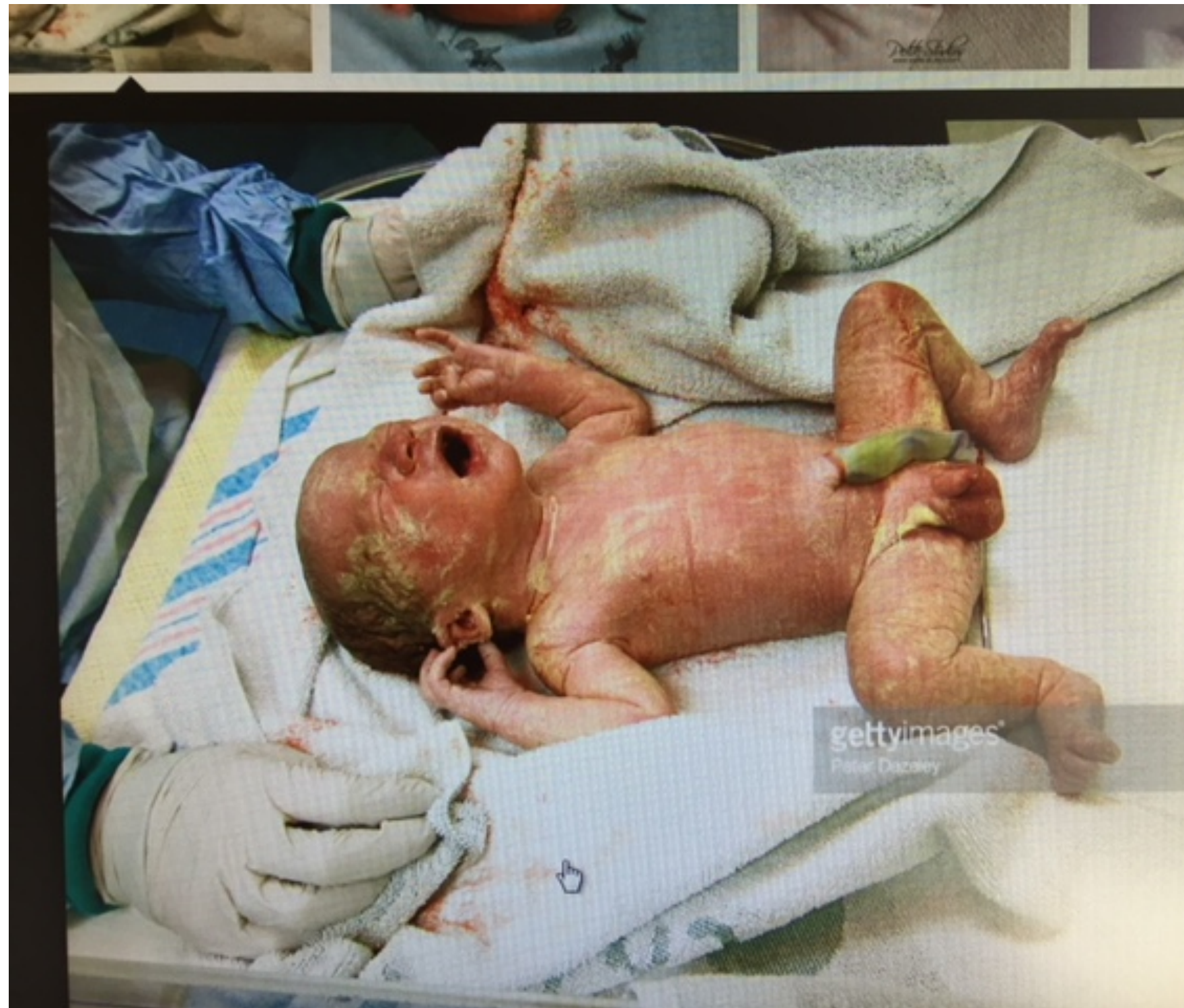
EFE

3.5 -4.0 mm Aortic
annulus

Unicommissural

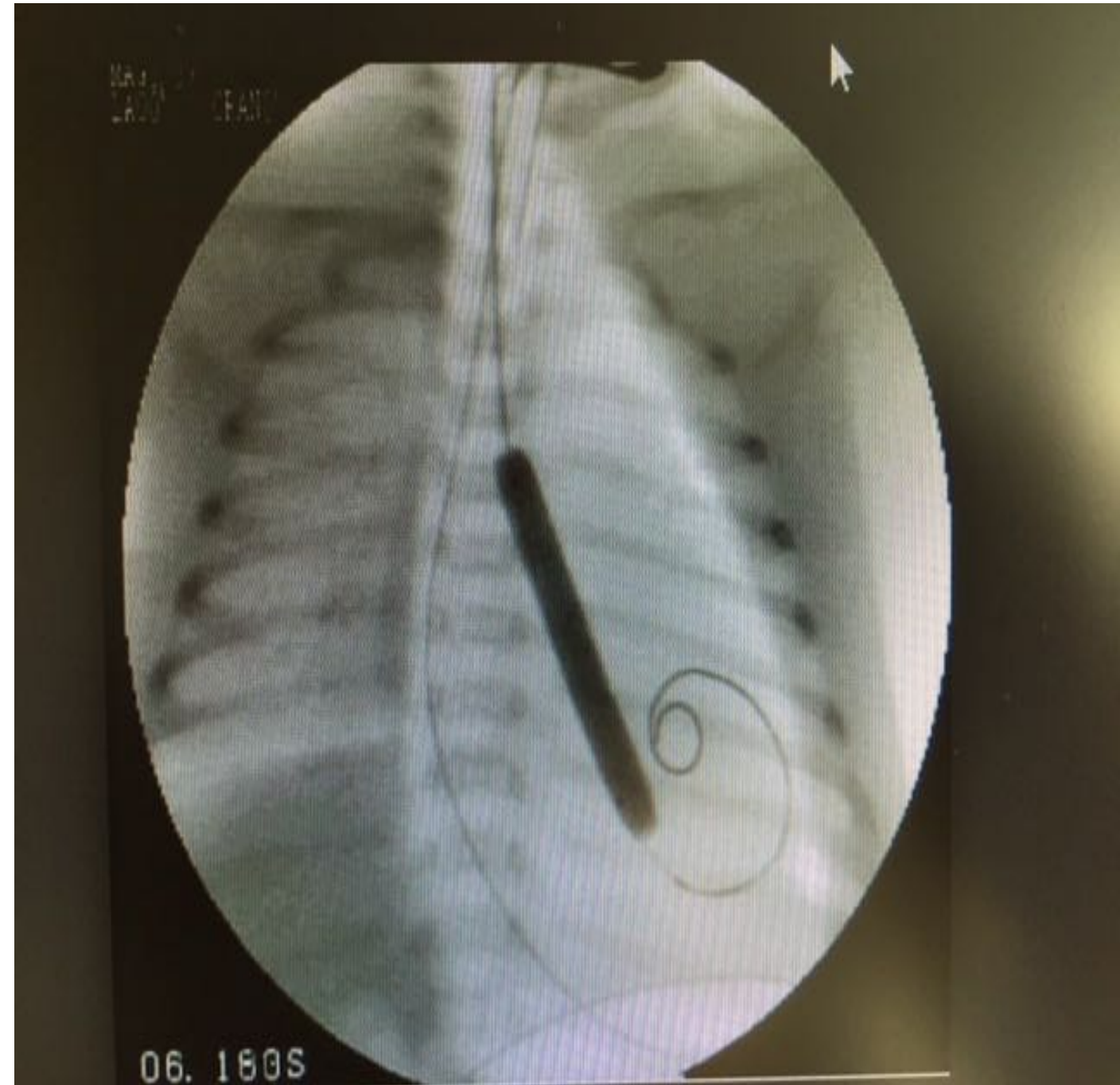
Extremely displastic

Severe MR



CLINICAL HISTORY

- EGA 36 Weeks
- C-section scheduled for 38 weeks GA
- Severe AS
- Severe MR
- LV function mildly depressed and dilated
- Plan cath and aortic valvuloplasty after postnatal exam







CORRECTION OF PULMONARY ATRESIA WITH A HOMOGRAFT AORTIC VALVE

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M.D. Lond., M.R.C.P.

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IN any large series of patients with Fallot's tetralogy, a few will be found with a complete obstruction of the right ventricle at the pulmonary valve or infundibular level. Lillehei et al. (1955) first described the management of such a case by reimplanting the distal pulmonary artery into the right ventricle, and subsequently the practice has been to reconstruct the outflow tract with plastic material (Lillehei et al. 1964, Sabiston et al. 1964). Although these

Electrocardiogram
wave in V₁
Cardiac catheterization

Femoral artery
Superior vena cava
Right ventricle
Right atrium

Angiocardiogram
Pulmonary artery
from a large
Operation

Exploratory
sternal incision
in diameter,

An attempt
by probing,
ventricle and
bypass was

REPLACEMENT OF AORTIC AND MITRAL VALVES WITH A PULMONARY AUTOGRAFT

DONALD N. ROSS

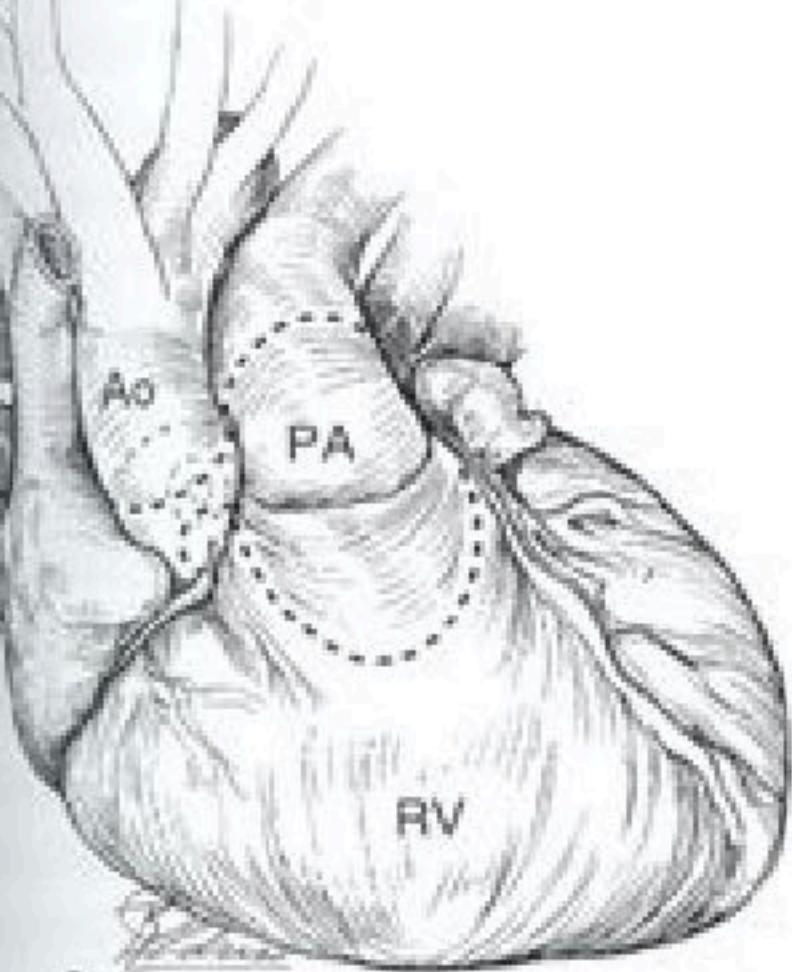
M.B., B.Sc. Cape Town, F.R.C.S.

CONSULTANT THORACIC SURGEON, GUY'S HOSPITAL, LONDON S.E.1, AND
NATIONAL HEART HOSPITAL, LONDON W.1

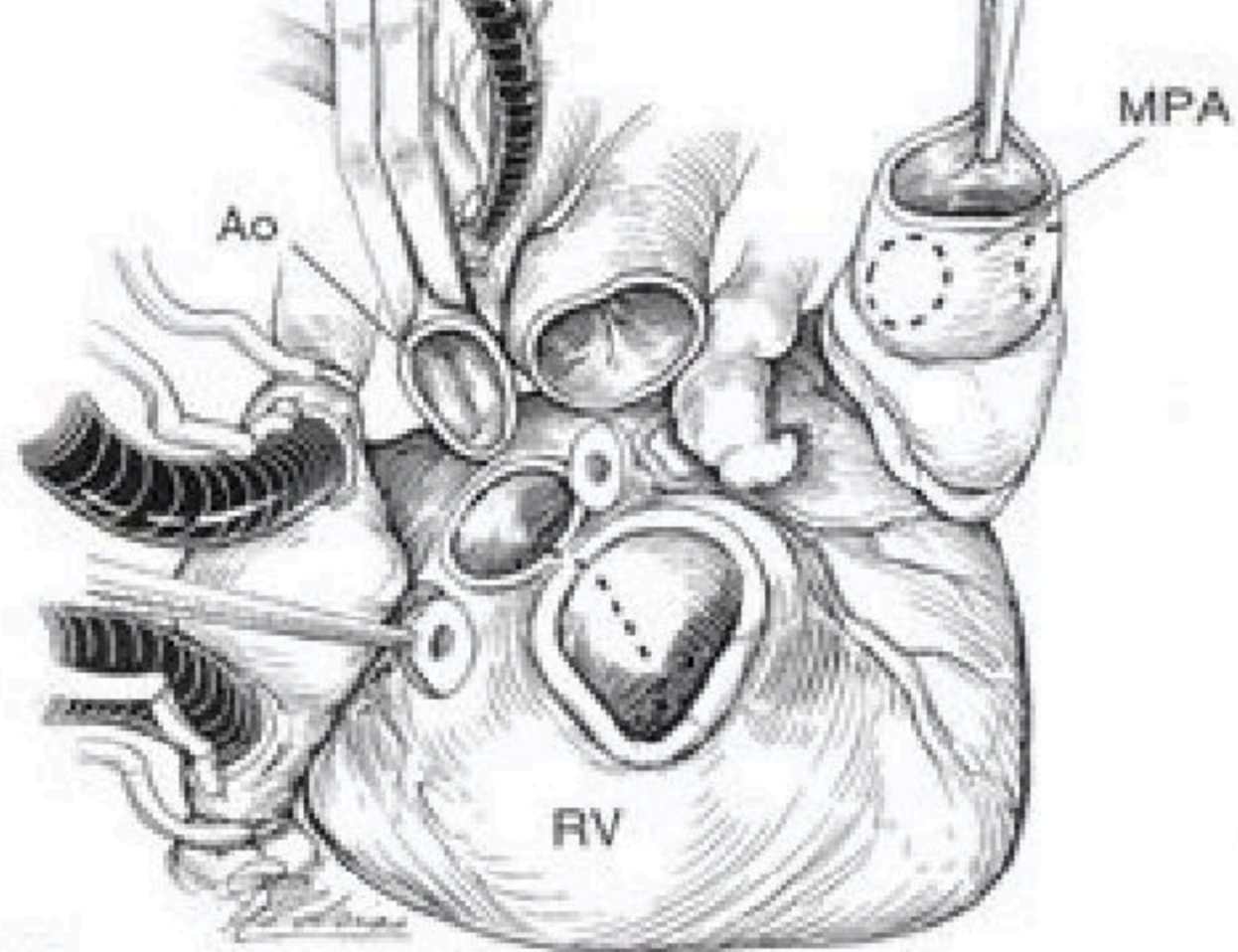
Summary The finding of late degenerative changes in a proportion of aortic homografts prompted the use of the patient's own pulmonary valve as a replacement. As a living autograft, the transplanted pulmonary valve has the prospect of long-term or permanent survival, whilst retaining the advantages of an aortic homograft. Similarly it can be used to replace the mitral valve. The excised pulmonary valve is replaced with a homograft.

INTRAOPERATIVE FINDINGS

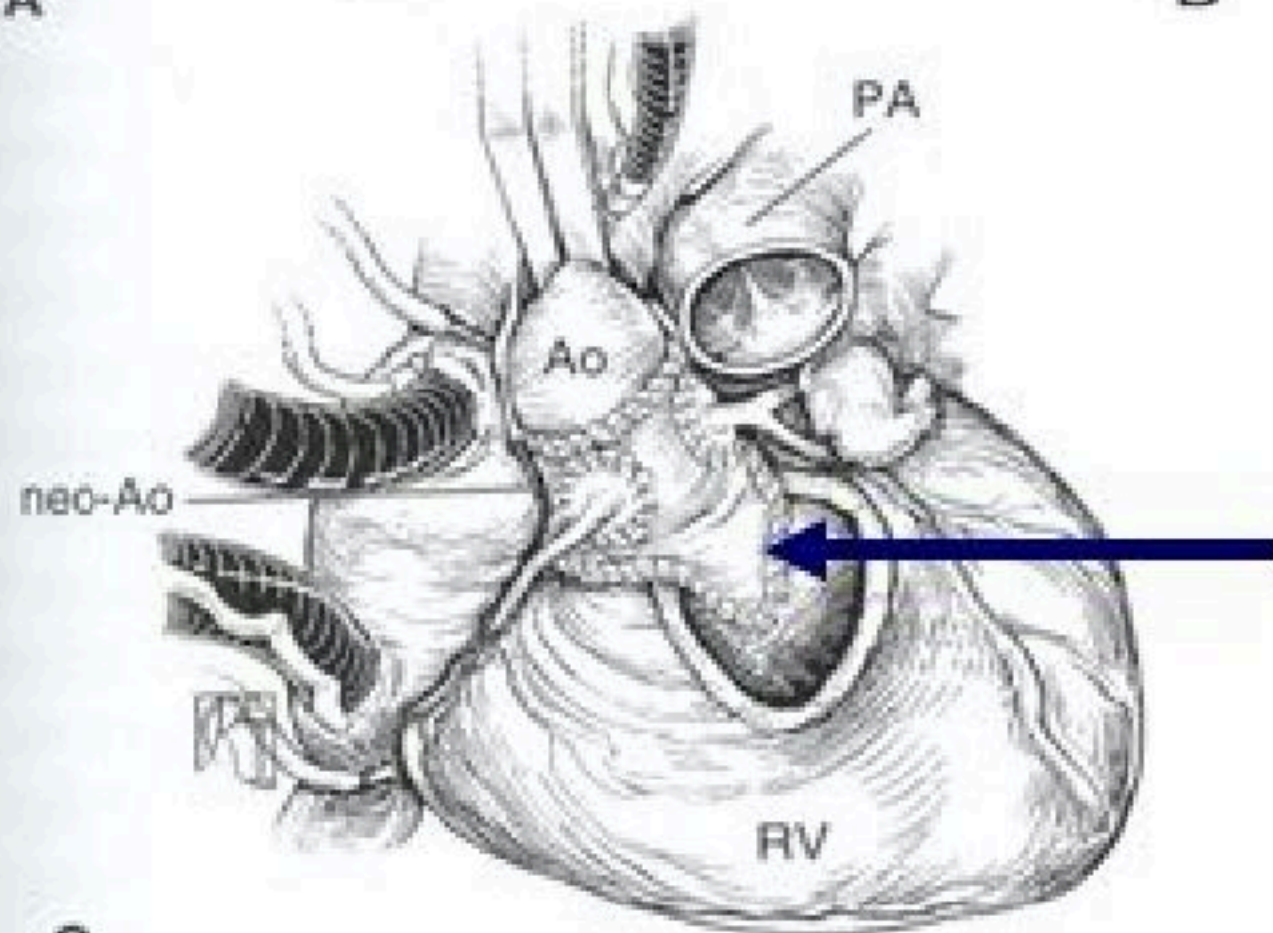
- 3+ MR under anesthesia (mod-severe)
- Aortic valve bicuspid without fusion and would take 3 mm probe but not 3.5 mm
- Autograft 10.5 mm and LVOT opened to 9 mm after myomectomy and resection of EFE
- Mitral regurgitation central and treated with Kay-Wooler annuloplasty in each commissure
- Chest left open



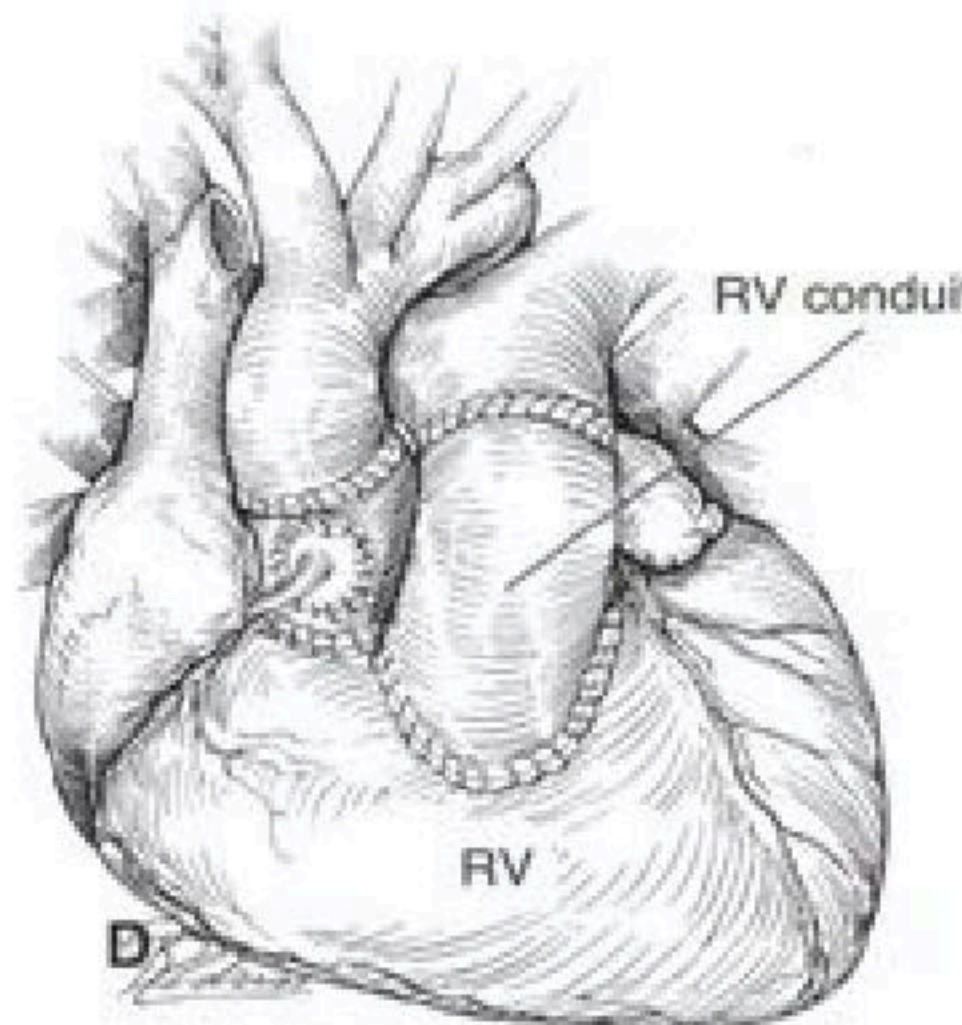
A



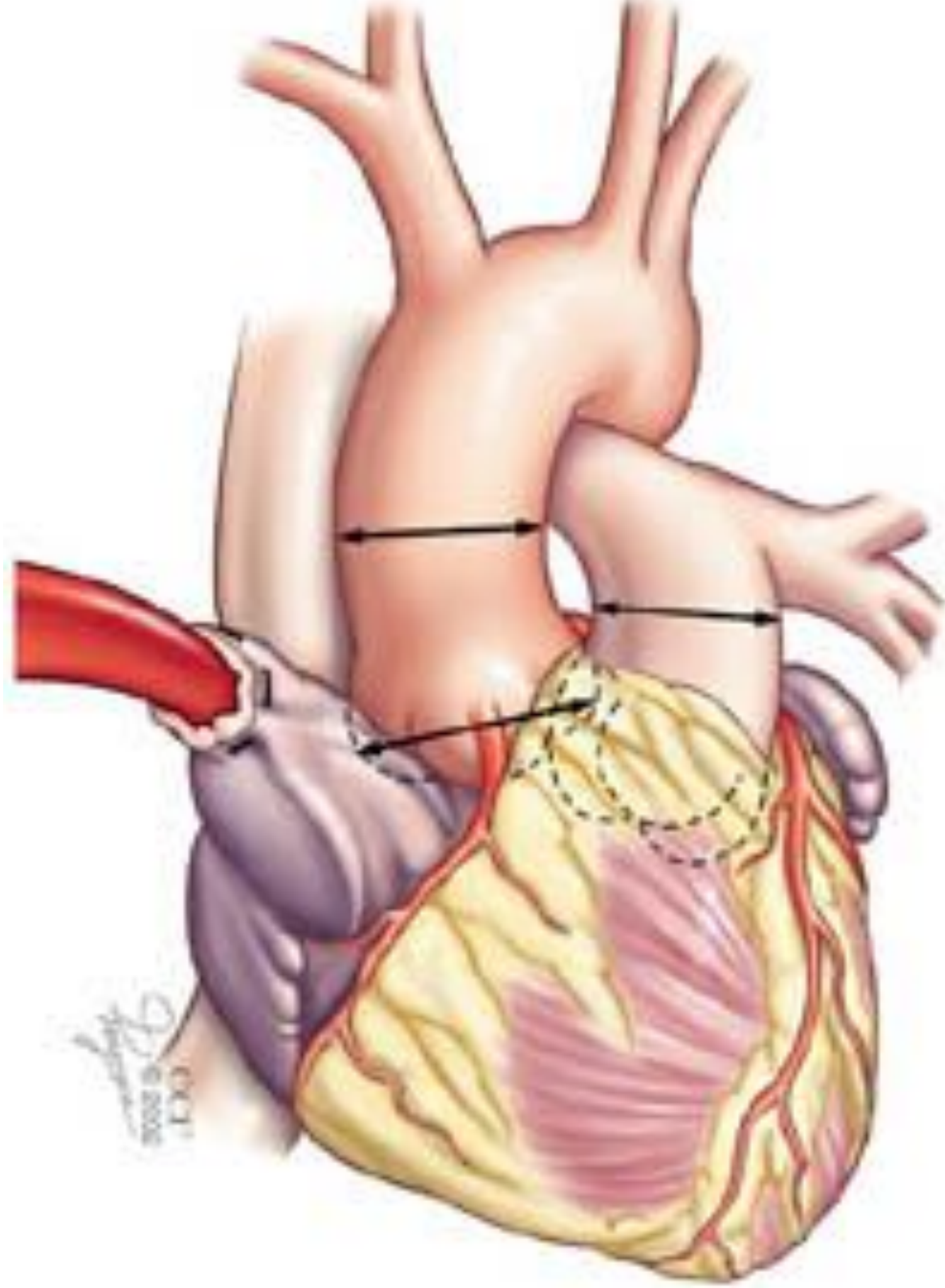
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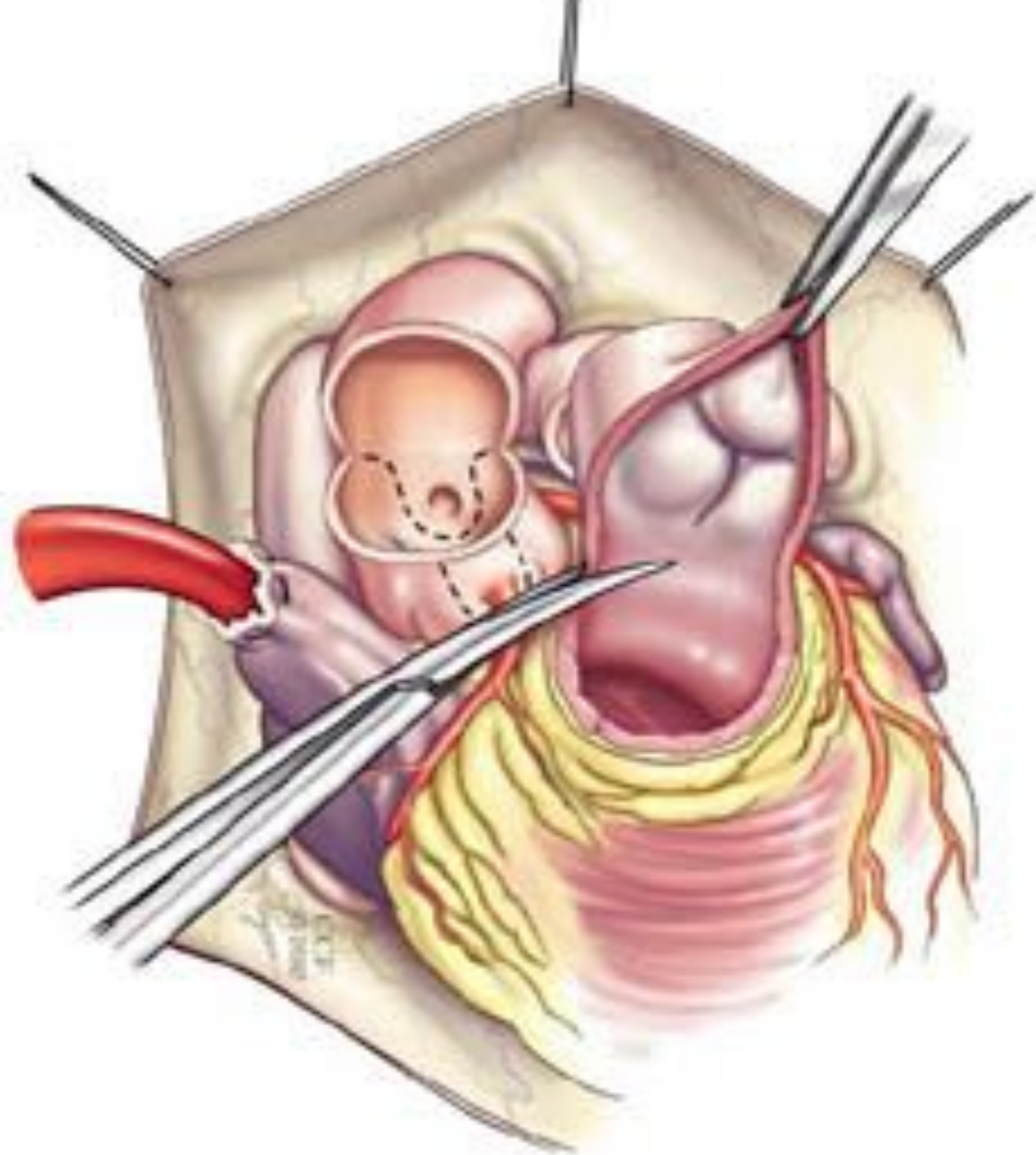


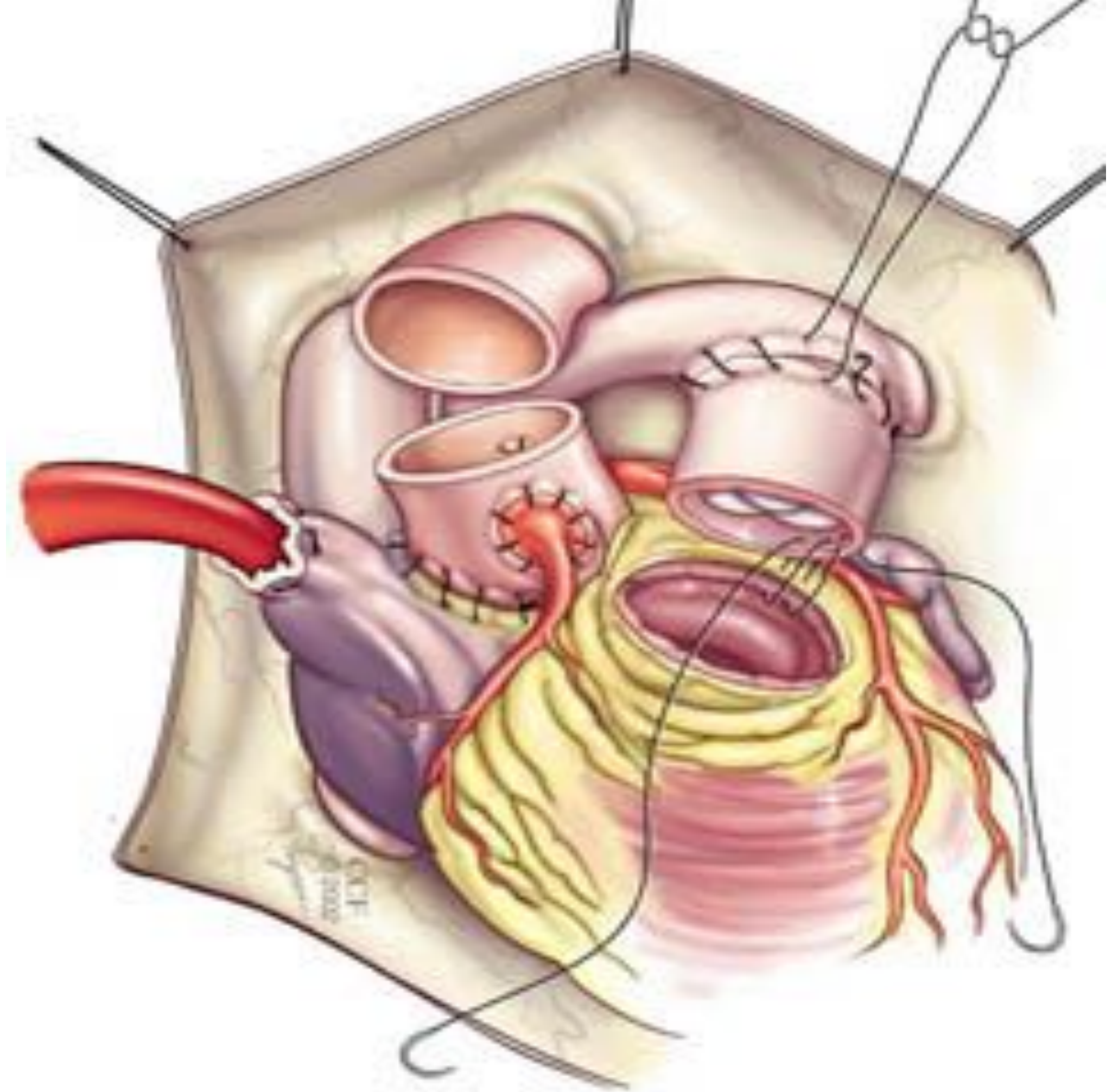
C



D









POSTOPERATIVE STATUS

Postop Transesophageal Echo

- Unobstructed LVOTO
- Trace AI, intermittent, 7 mm
- Mild plus-moderate MR
- Preserved biventricular function
- 9 mm PV allograft and RVOT unobstructed
- Normal sinus rhythm

Postop recovery

- Chest closure postop day 3
- Extubated 48 hours later
- Discharged day 33



Successful Immediate Newborn Ross-Konno and Mitral Valve Repair Following Fetal Aortic Valvuloplasty

World Journal for Pediatric and
Congenital Heart Surgery

3(2) 264-266

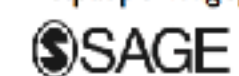
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DOI: 10.1177/2150135111433471

<http://pch.sagepub.com>



**Michael L. Ciccolo, MD^{1,2}, Abraham Rothman, MD^{3,4},
Alvaro Galindo, MD^{3,4}, Ruben J. Acherman, MD^{3,4}, and
William N. Evans, MD^{3,4}**

Abstract

Perinatal management of severe mitral regurgitation (MR) and aortic stenosis (AS) is difficult; mortality is high, and there are few reports of successful postnatal biventricular repairs. We report a patient with severe MR and AS, diagnosed prenatally, that underwent a fetal aortic valvuloplasty and a successful modified Ross-Konno procedure with concomitant mitral valve repair shortly after birth.





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Neonatal Critical Aortic Stenosis

Out-Patient Follow-Up

SUNRISE ECHO

FR 78Hz

8.1cm

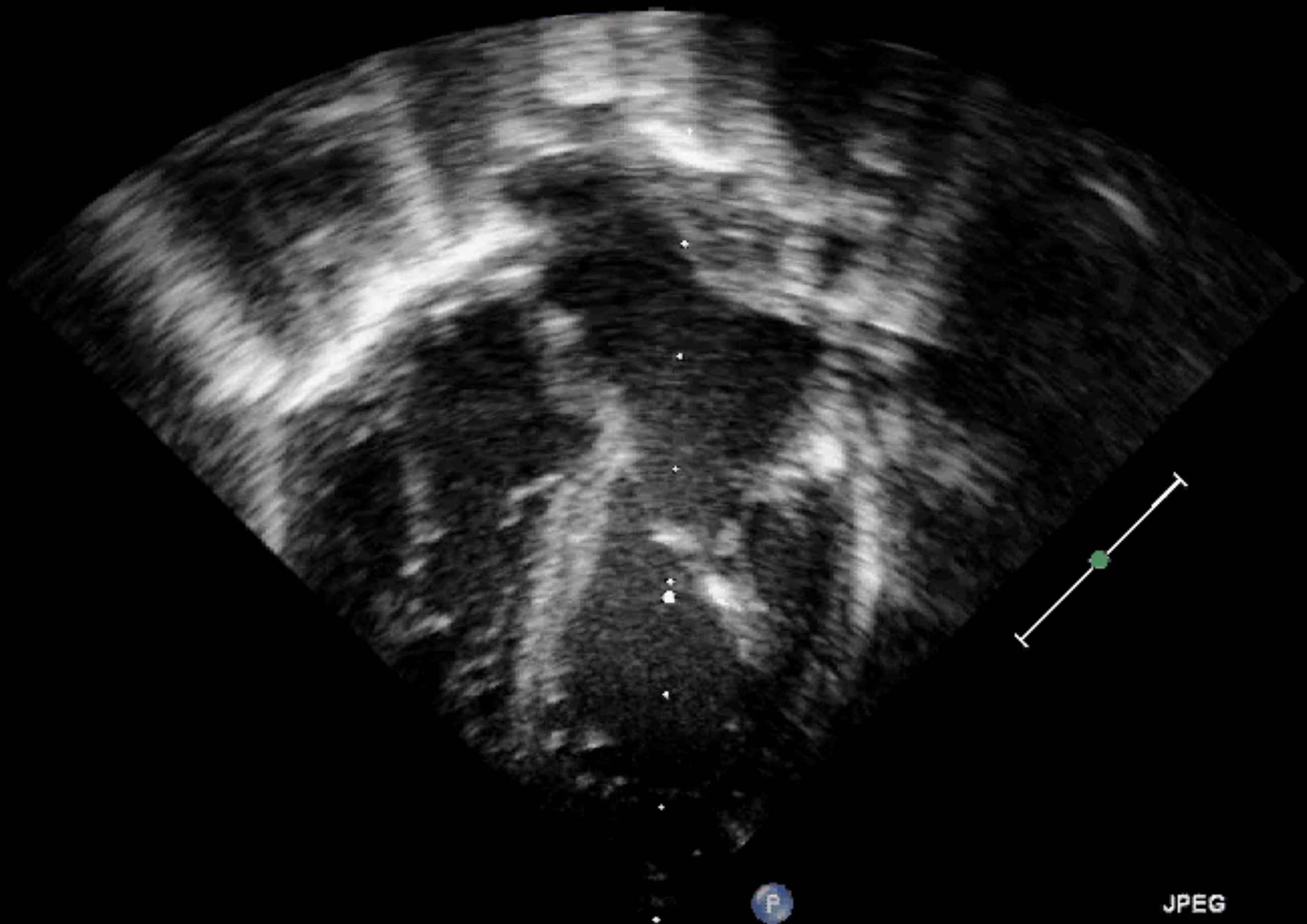
2D

87%

C 50

P Off

Gen

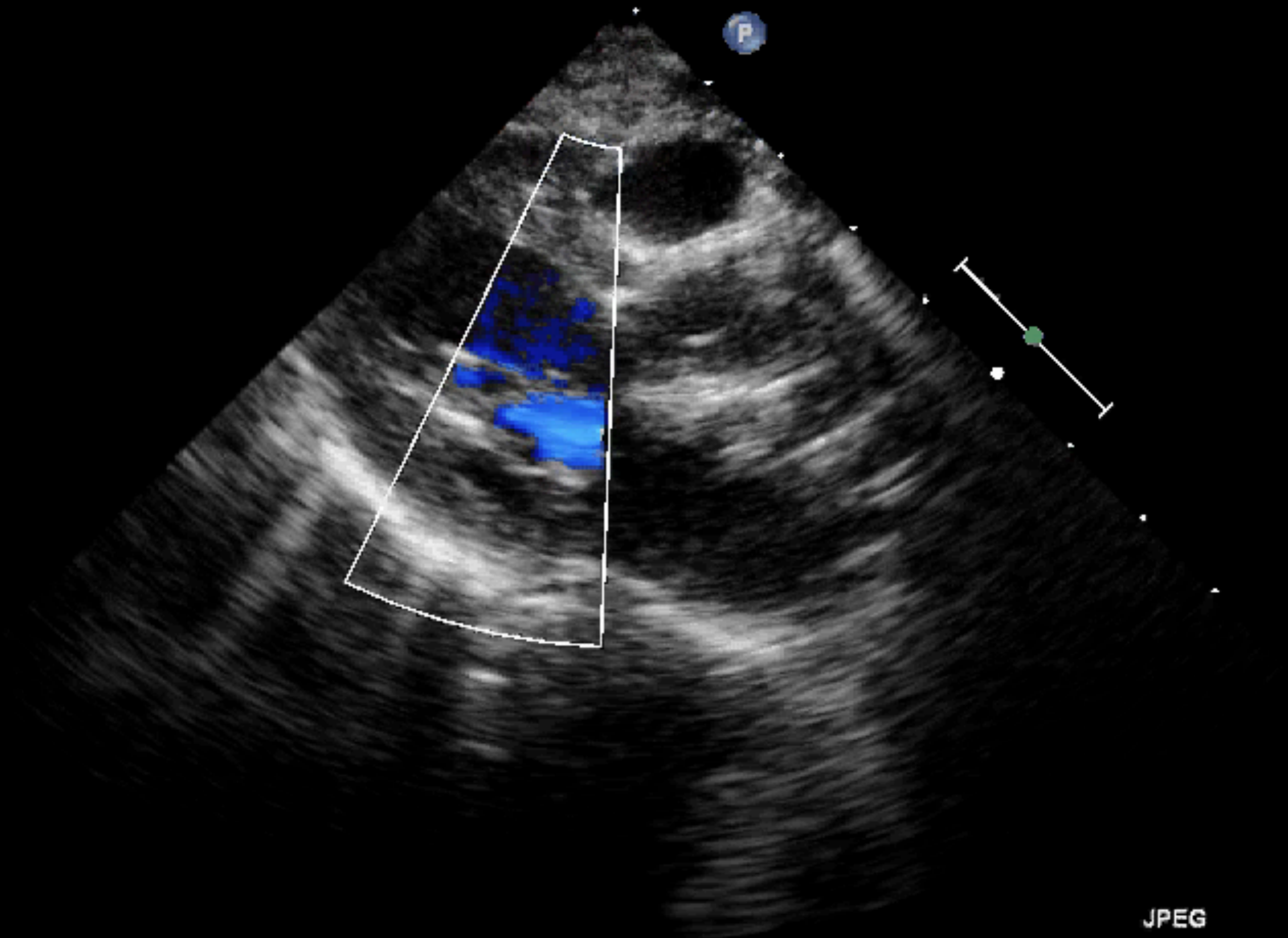


JPEG

UNRISE ECHO
PR 44HZ
9.0cm

2D
90%
C 50
P Off
Gen

CF
77%
5.0MHz
WF High
Med



UNRISE ECHO

R 44Hz

1.0cm

D

90%

C 50

P Off

Gen

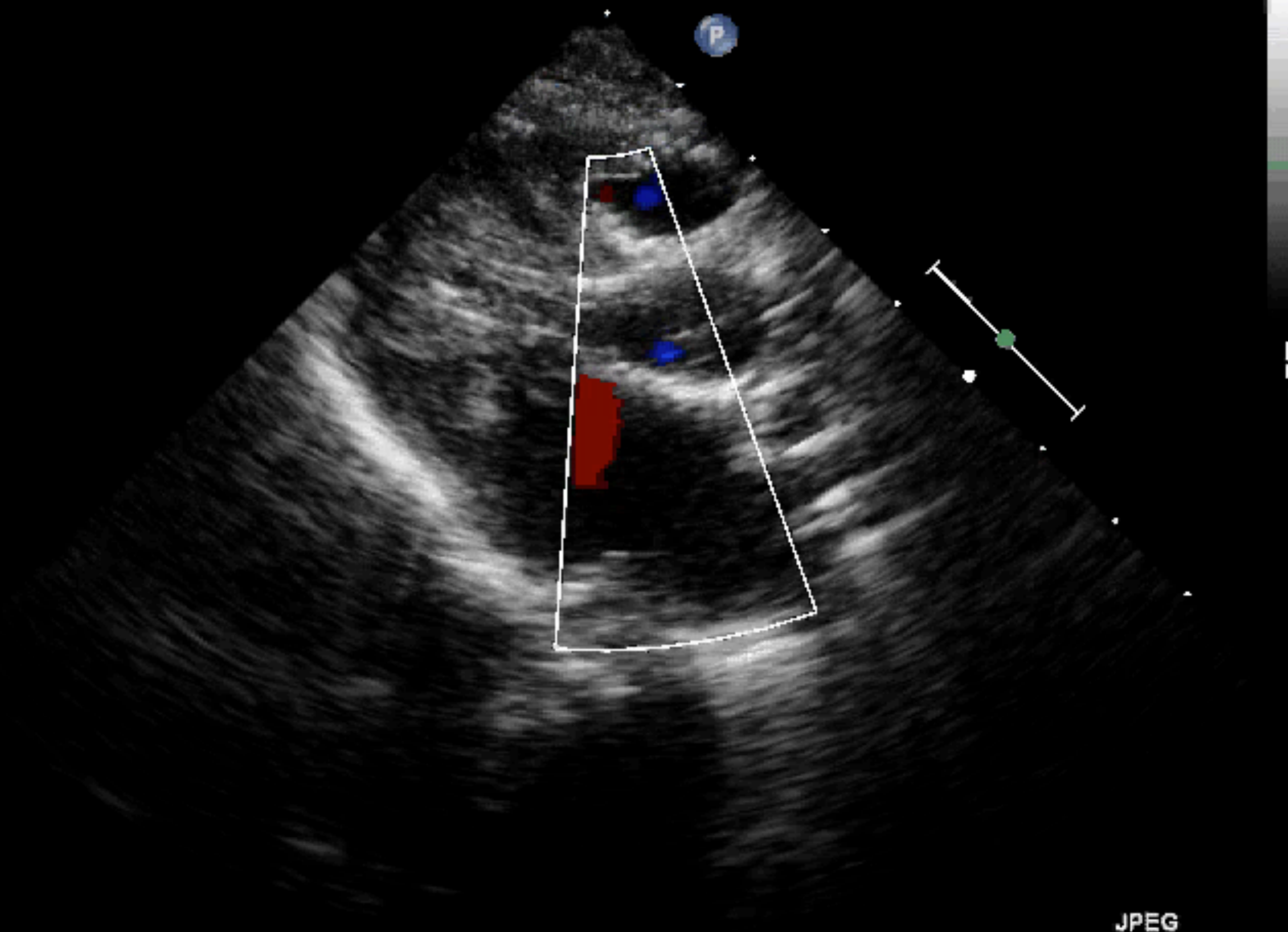
CF

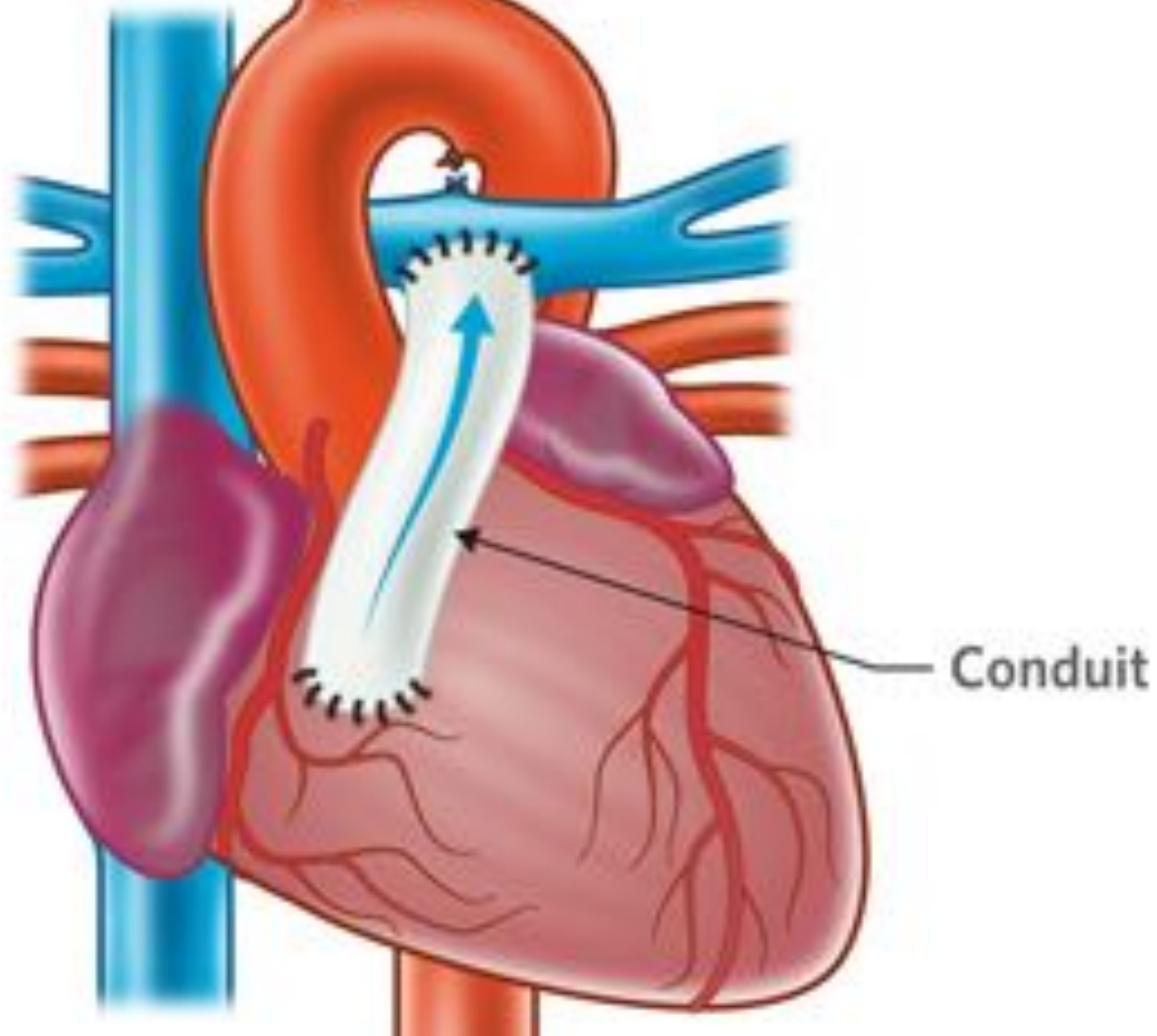
77%

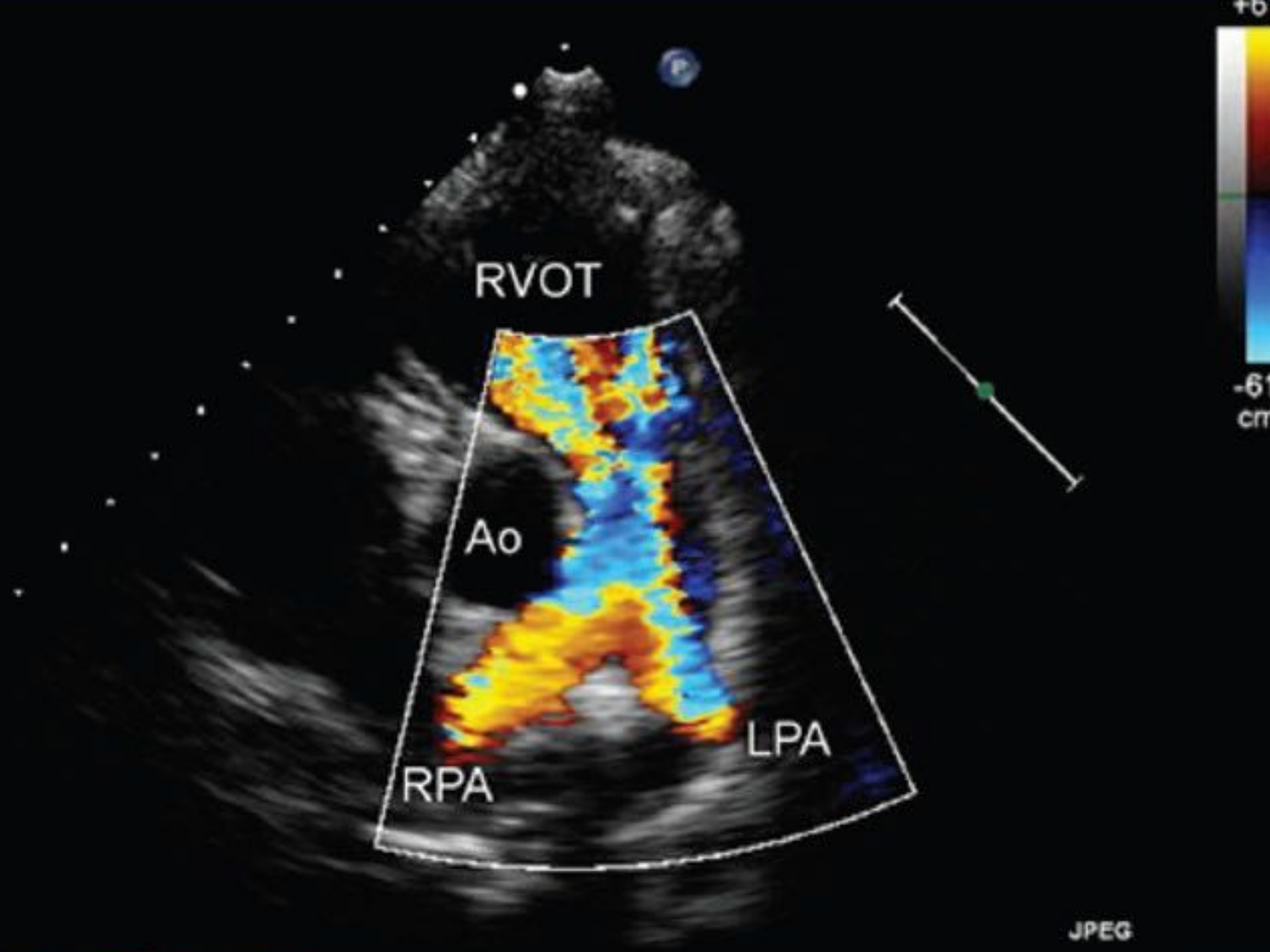
5.0MHz

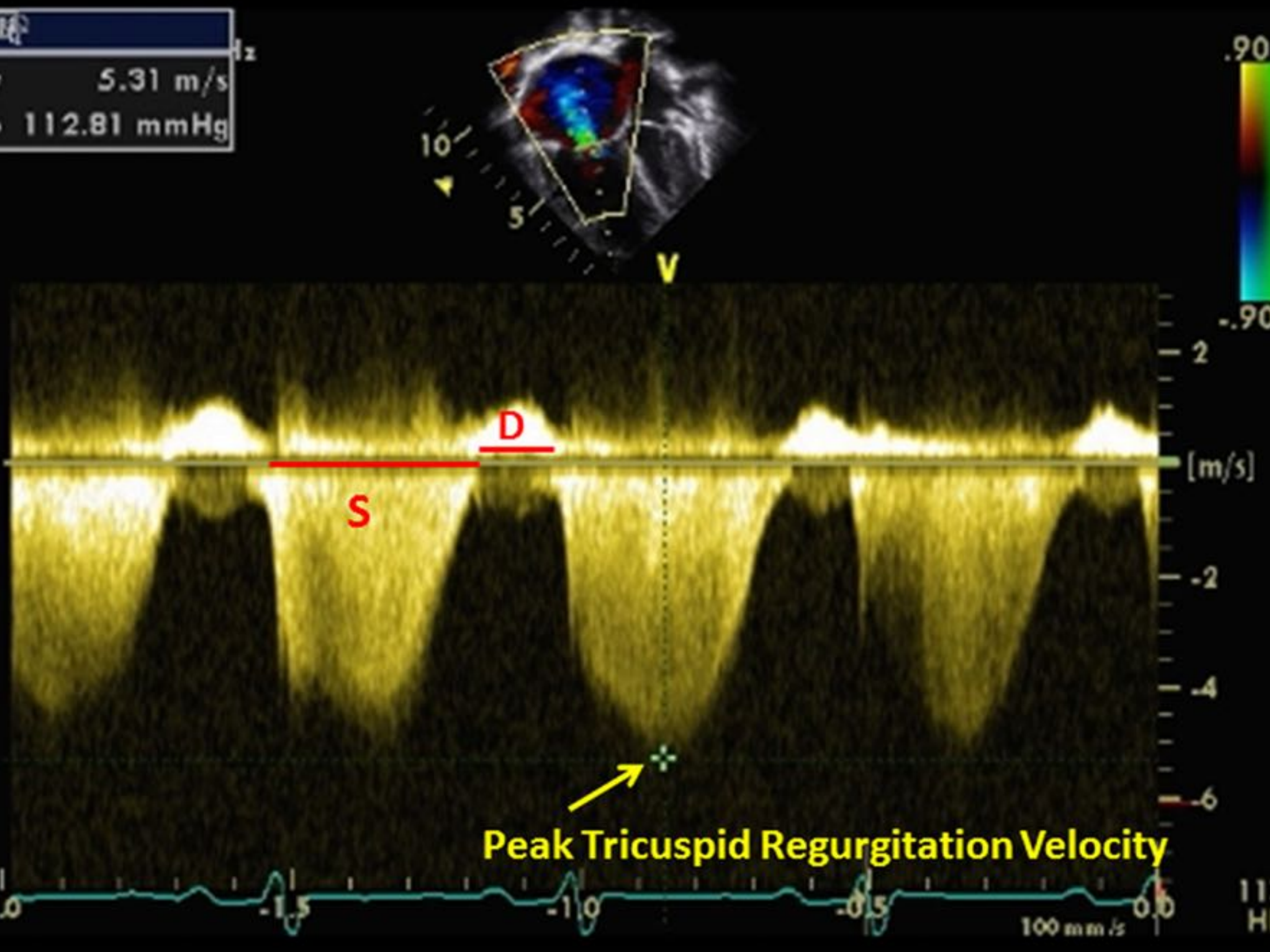
VF High

Med











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Neonatal Critical Aortic Stenosis

RV-PA Conduit Replacement

RV-to-PA continuity

- Conduit types
- Durability
- Indications for intervention

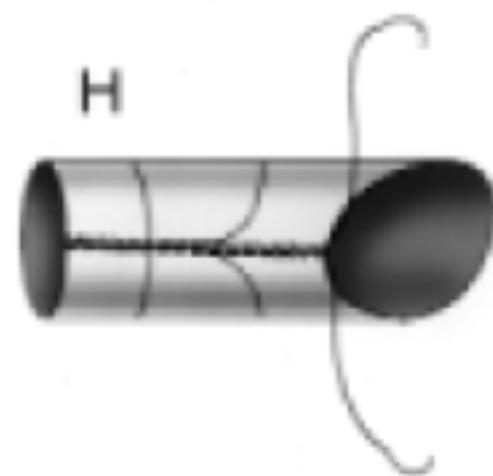
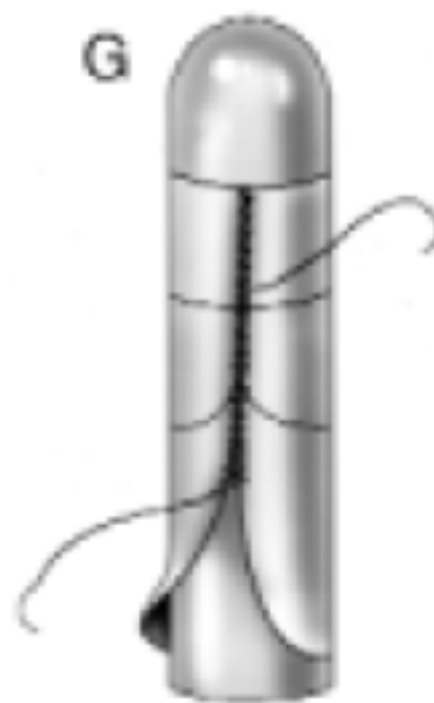
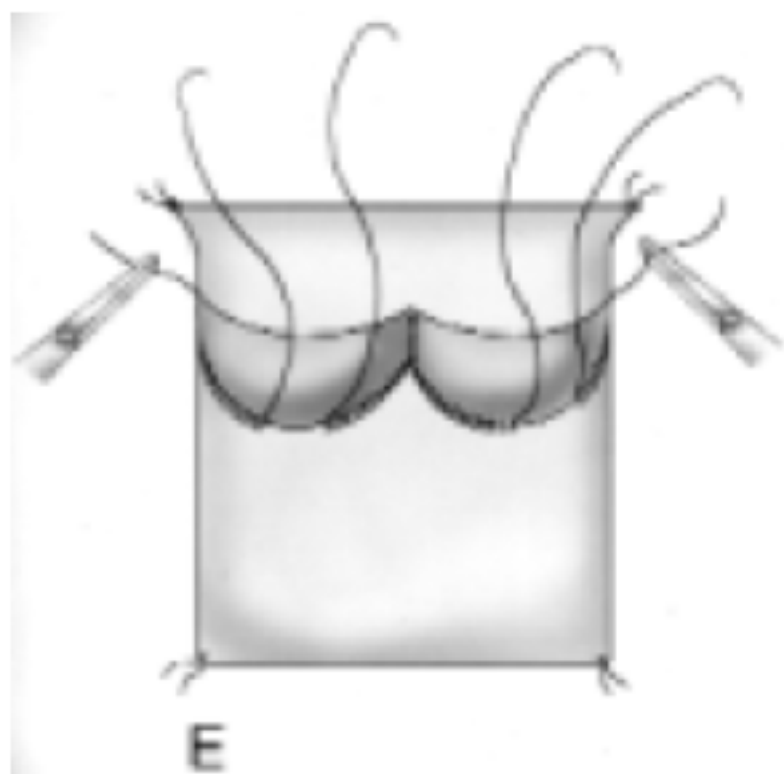
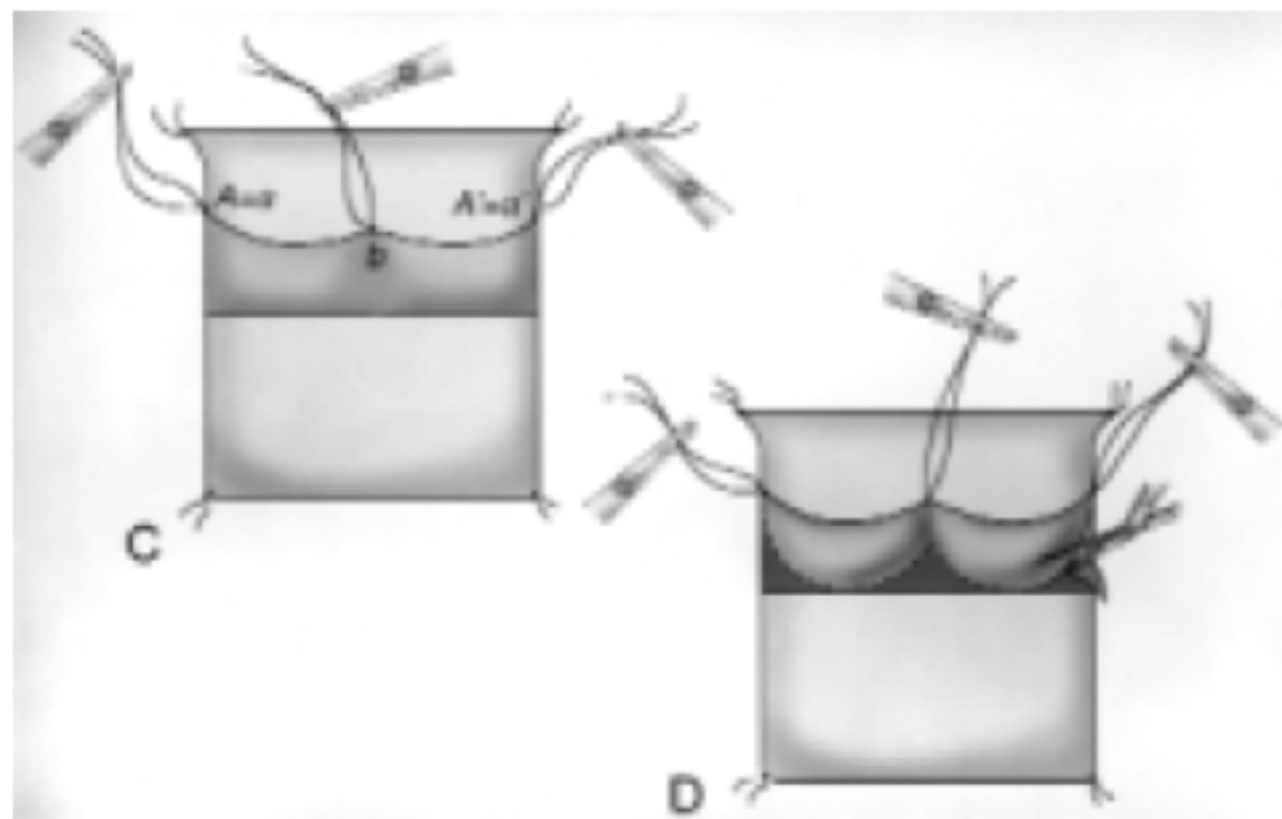
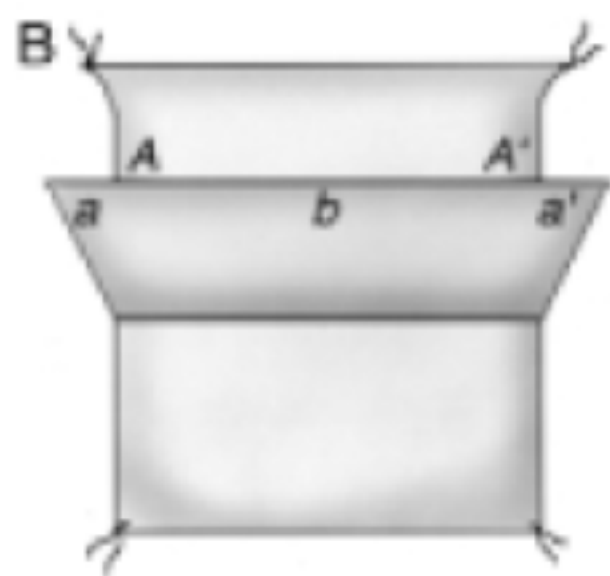
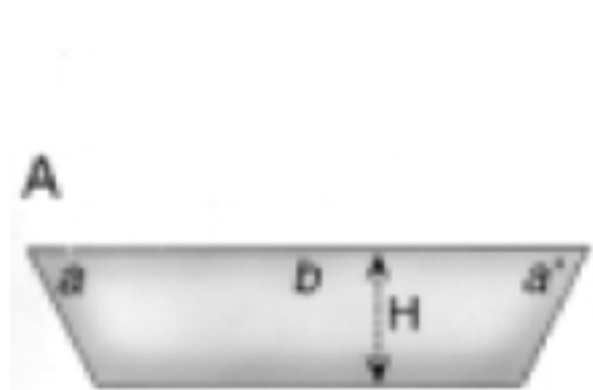
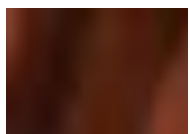


Table 3: Comparison of the different right ventricular outflow tract grafts after the Ross procedure

	PA	dt XG	P-value	gt XG	P-value	AR XG	P-value	PTFE ^a	P-value
Patients, n	175	375	-	74	-	112	-	5	-
Age (years)	39.6 ± 11.1	52.8 ± 10.6	<0.001	47.4 ± 12.8	<0.001	42.0 ± 13.5	0.112	42.0 (41.0; 48.0)	0.383
Conduit size (mm)	26.1 ± 3.0	26.4 ± 0.9	0.321	26.5 ± 1.2	0.131	26.5 ± 1.3	0.323	25.0 (25.0; 27.0)	0.223
Follow-up (years)	5.1 ± 1.8	5.2 ± 1.7	0.121	5.3 ± 1.9	0.243	7.7 ± 2.1	0.012	1.0 (1.0; 2.0)	0.014
Dysfunction, n (%)	0	3 (0.8)	0.555	10 (13.5)	<0.001	15 (13.4)	<0.001	0	-
Freedom from reoperation ^b									
At 5 years	100%	98.2 (91.4-99.5)	0.231	82.7 (69.8-90.2)	<0.001	93.3 (86.5-96.8)	0.049	100%	-
At 10 years		94.4 (74.0-98.4)		82.7 (69.8-90.2)		80.6 (66.3-88.0)			
RVOT peak gradient (mmHg)									
At discharge	8.1 ± 3.7	11.4 ± 3.7	0.013	14.9 ± 6.1	<0.001	17.9 ± 7.4	<0.001	12.8 (11.5; 16.0)	0.144
At follow-up	15.4 ± 4.6	22.7 ± 6.1	0.002	36.7 ± 15.2	<0.001	38.8 ± 21.3	<0.001	21.5 (20.0; 23.5)	0.053
P-value*	0.022	0.003	-	<0.001	-	<0.001	-	0.044	

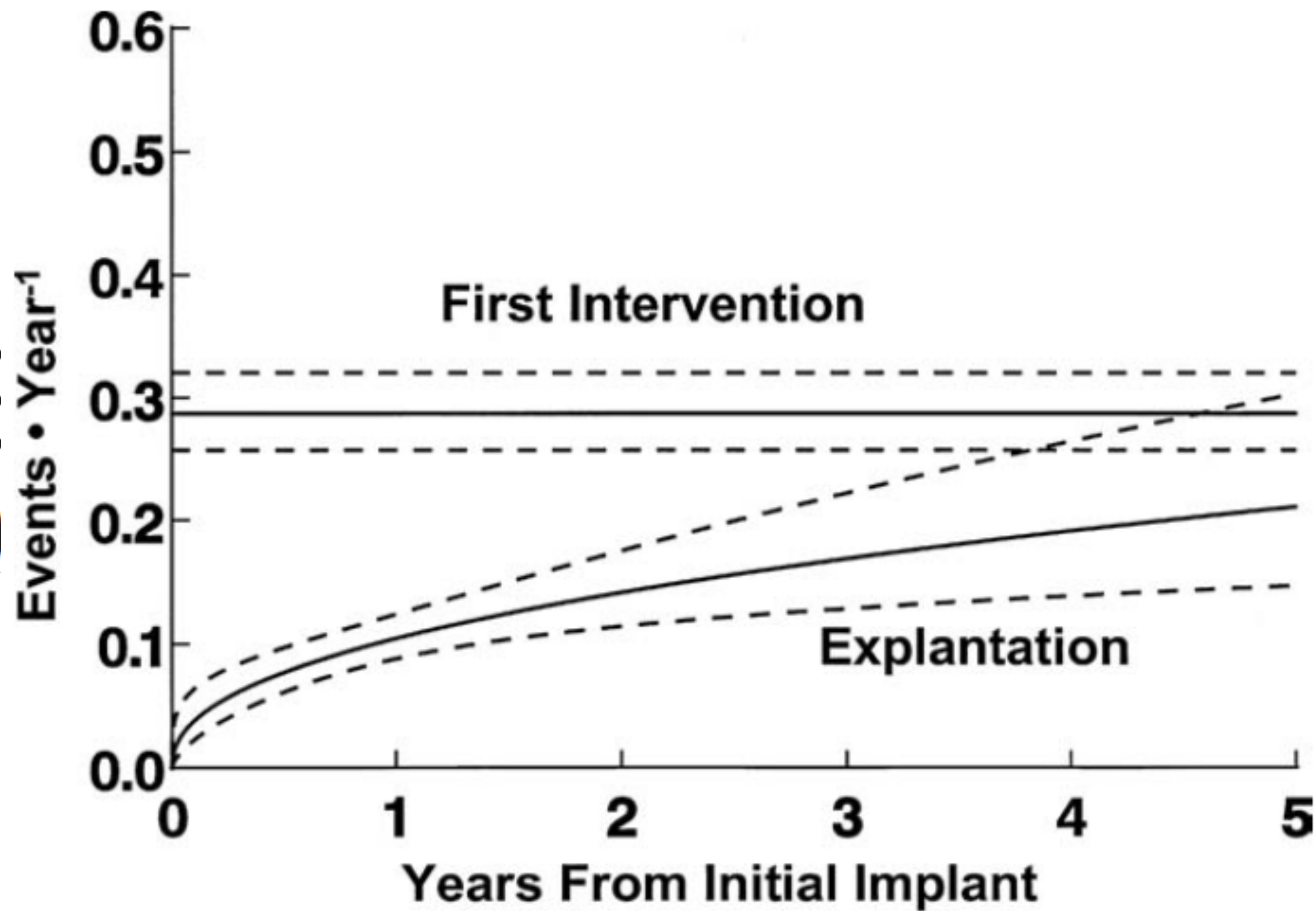
PA: pulmonary allograft; dt XG: diepoxide-treated xenograft; gt XG: glutaraldehyde-treated xenograft; AR XG: aortic root xenograft; PTFE: polytetrafluoroethylene conduit; RVOT: right ventricular outflow tract.

^aData are presented as median (25th; 75th percentile).

^bData are presented as percentages (95% confidence interval).

P-values: comparison with the findings for the allograft group; P-values*: comparison of gradients at follow-up with gradients at discharge.

Copy



16 NEONATAL

RV-PA CONDUITS

1ST COND	AGE 1st	2ND COND	AGE 2nd
10mm (8-13)	0	18mm (12-24)	28 months (6-84)

Indications for intervention

- Stenosis
- Insufficiency
- Endocarditis
- Aneurysm, others



Conduit Surgeries 2012-2017

- 120
- 2 (1.6%) deaths
- STAT score average 3.2, benchmark mortality STS ~3%
- Our results are 2X better than the national average



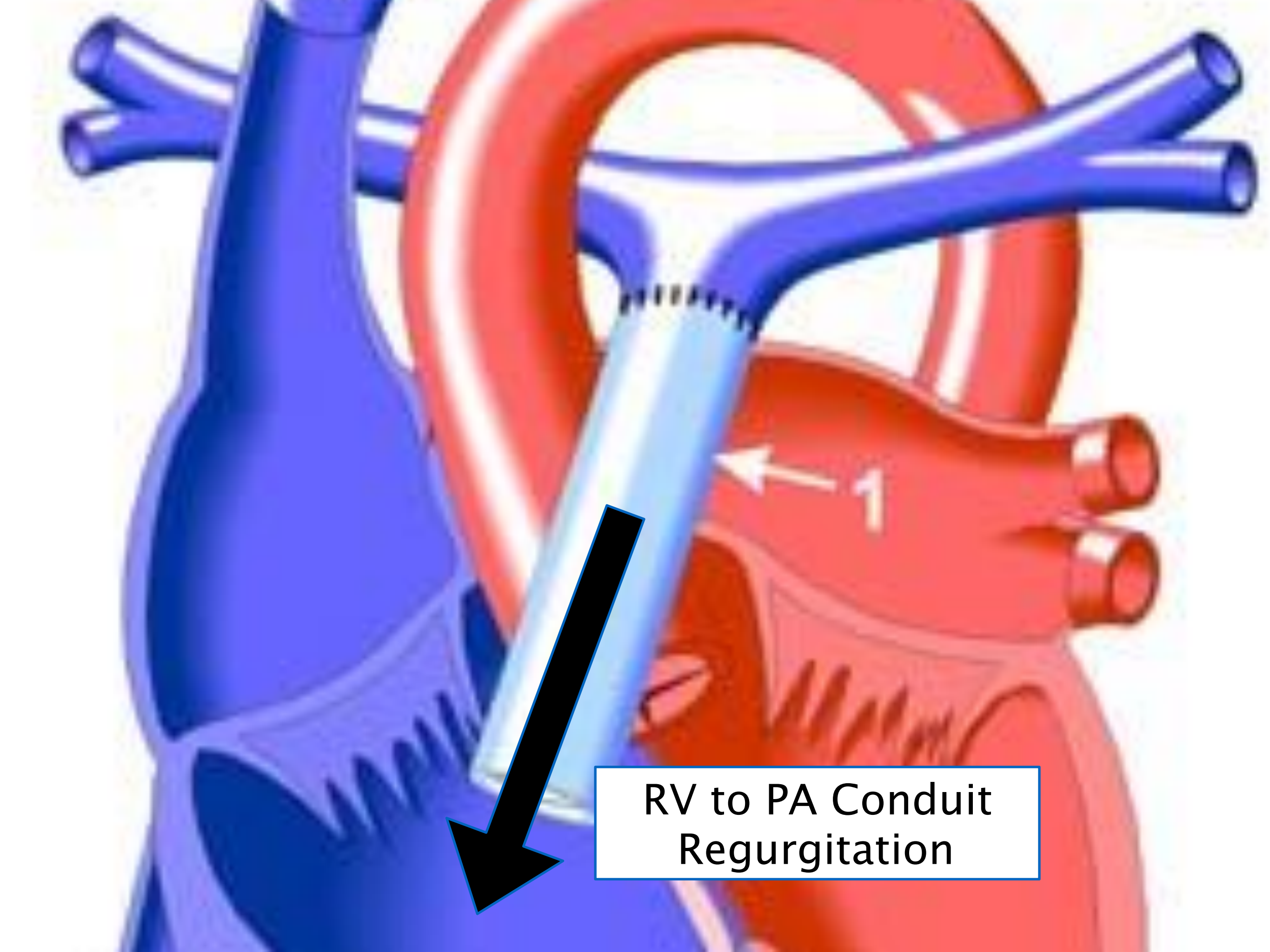


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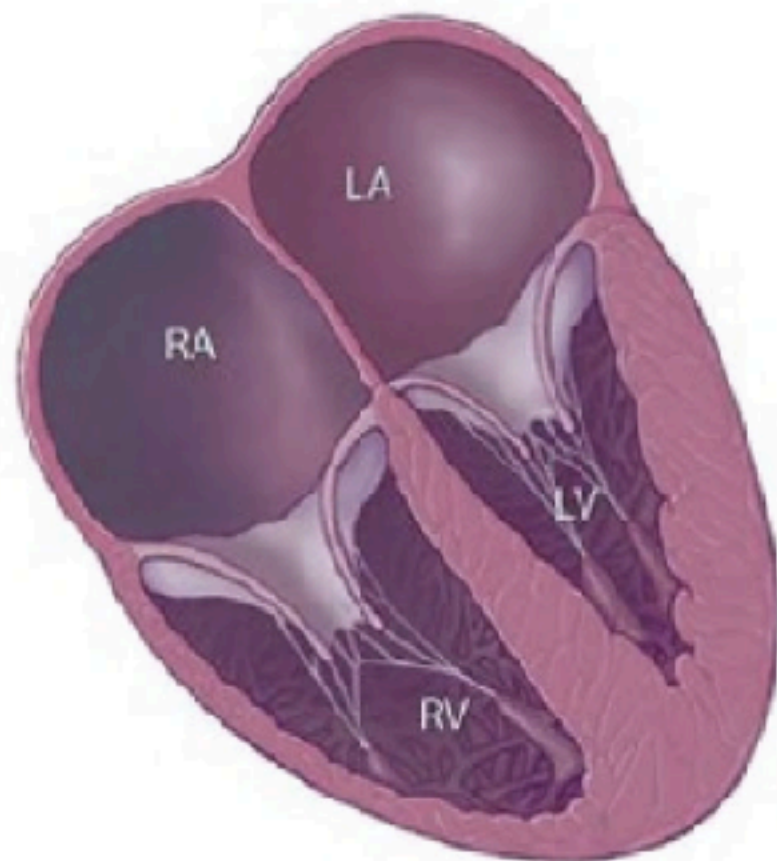
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Neonatal Critical Aortic Stenosis

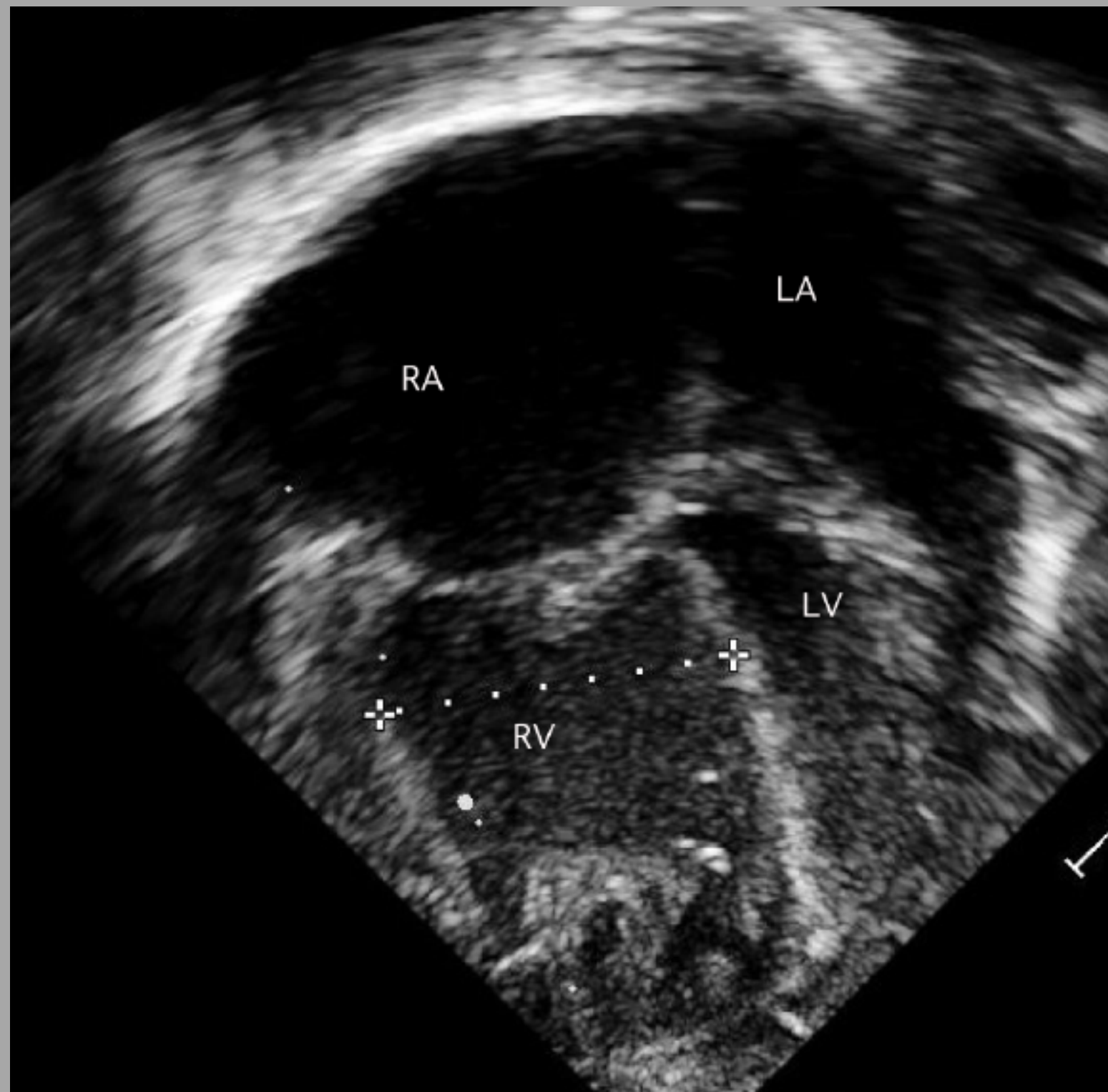
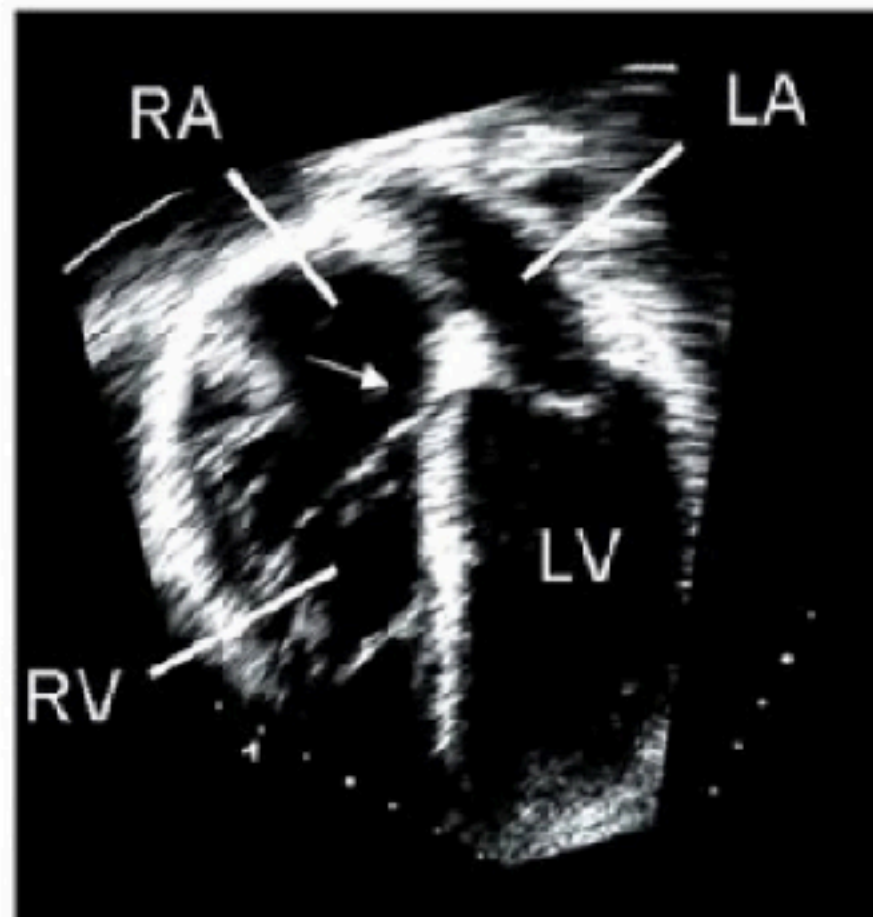
Trans-catheter Pulmonary Valve



RV to PA Conduit
Regurgitation



Normal

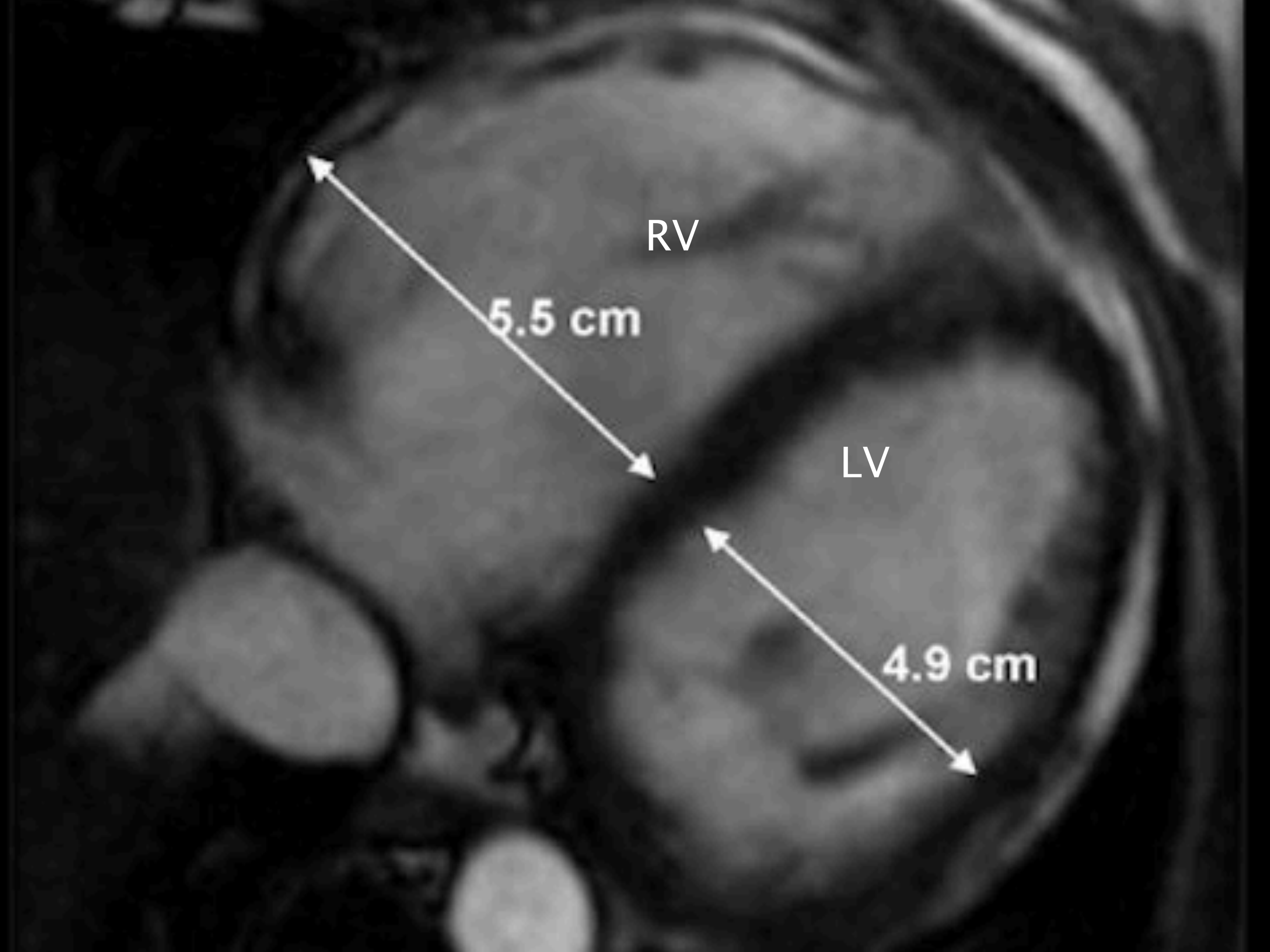


NORMAL

RV

LV





RV

5.5 cm

LV

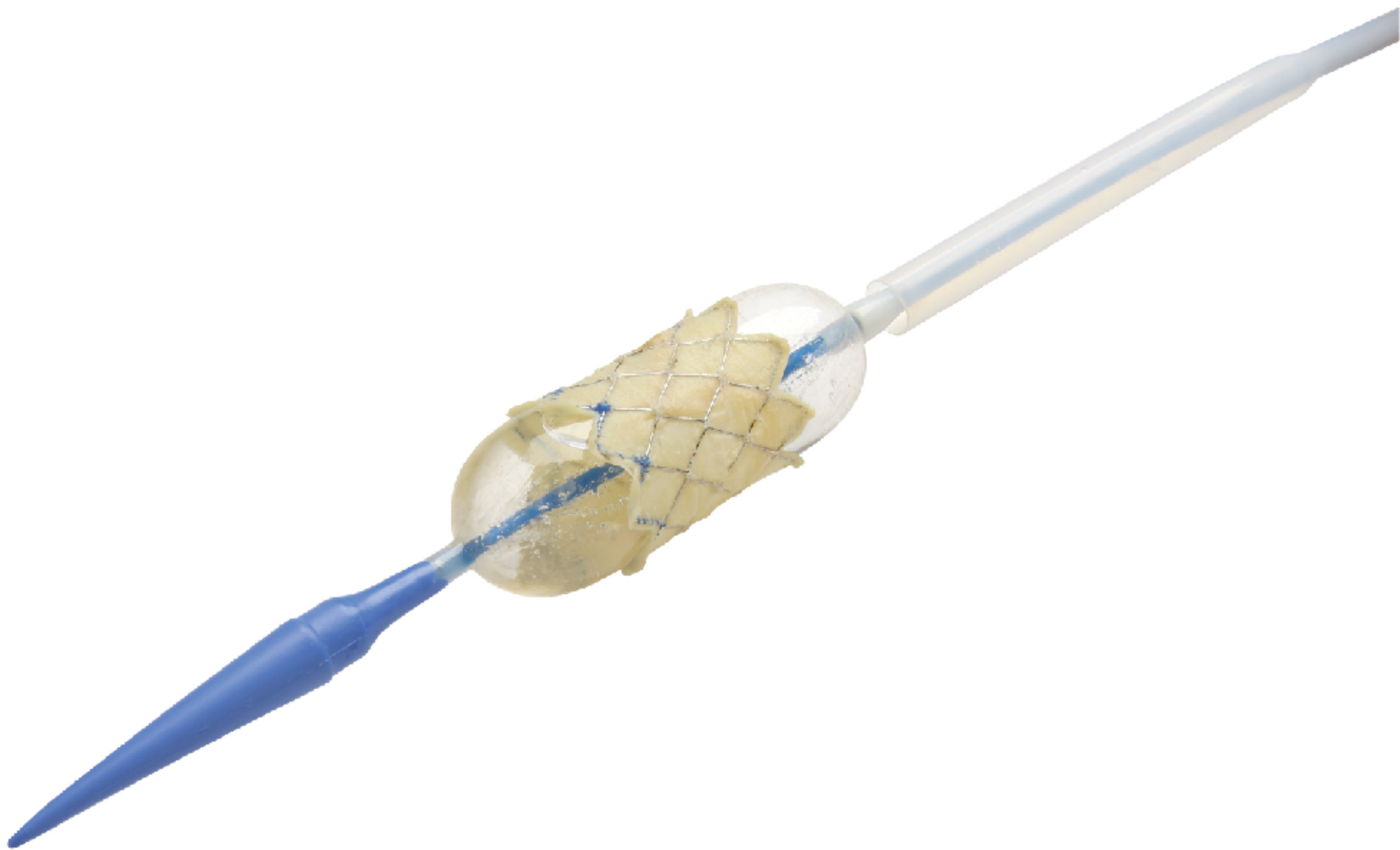
4.9 cm

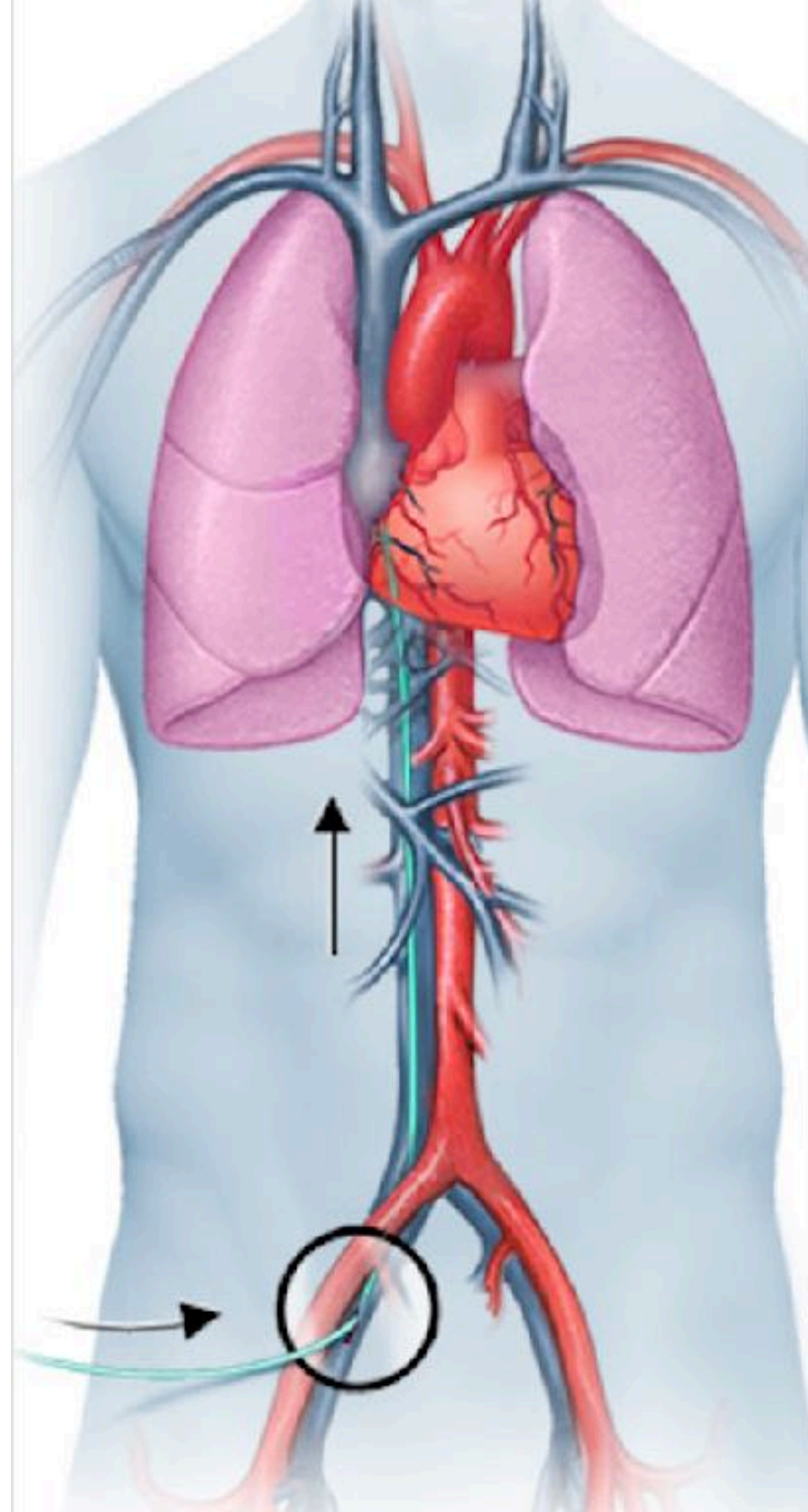
Bovine Pericardial Tissue
Leaflets with Carpentier-Edwards
ThermaFix Process*

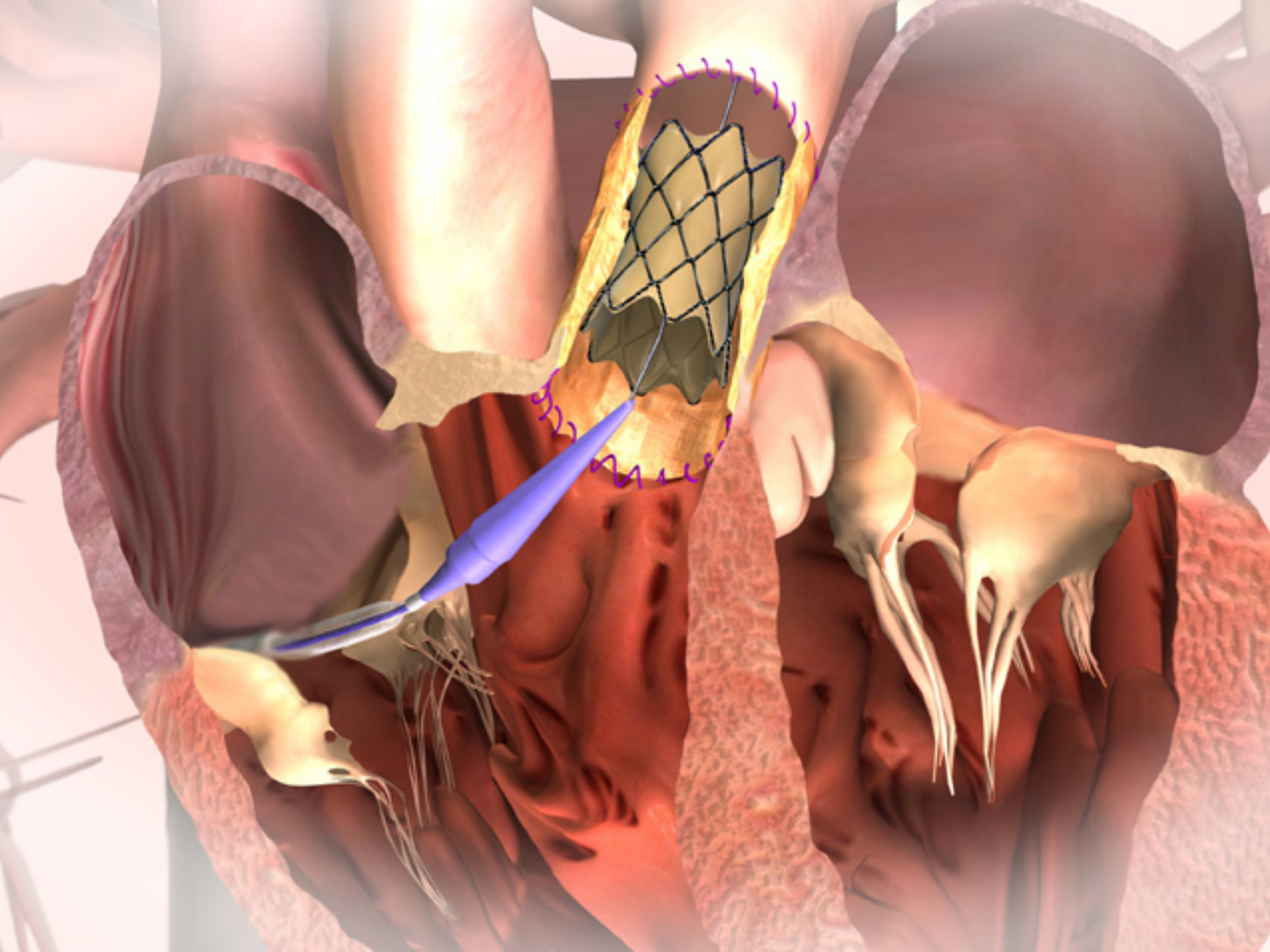
Balloon-expandable
Stainless Steel Frame

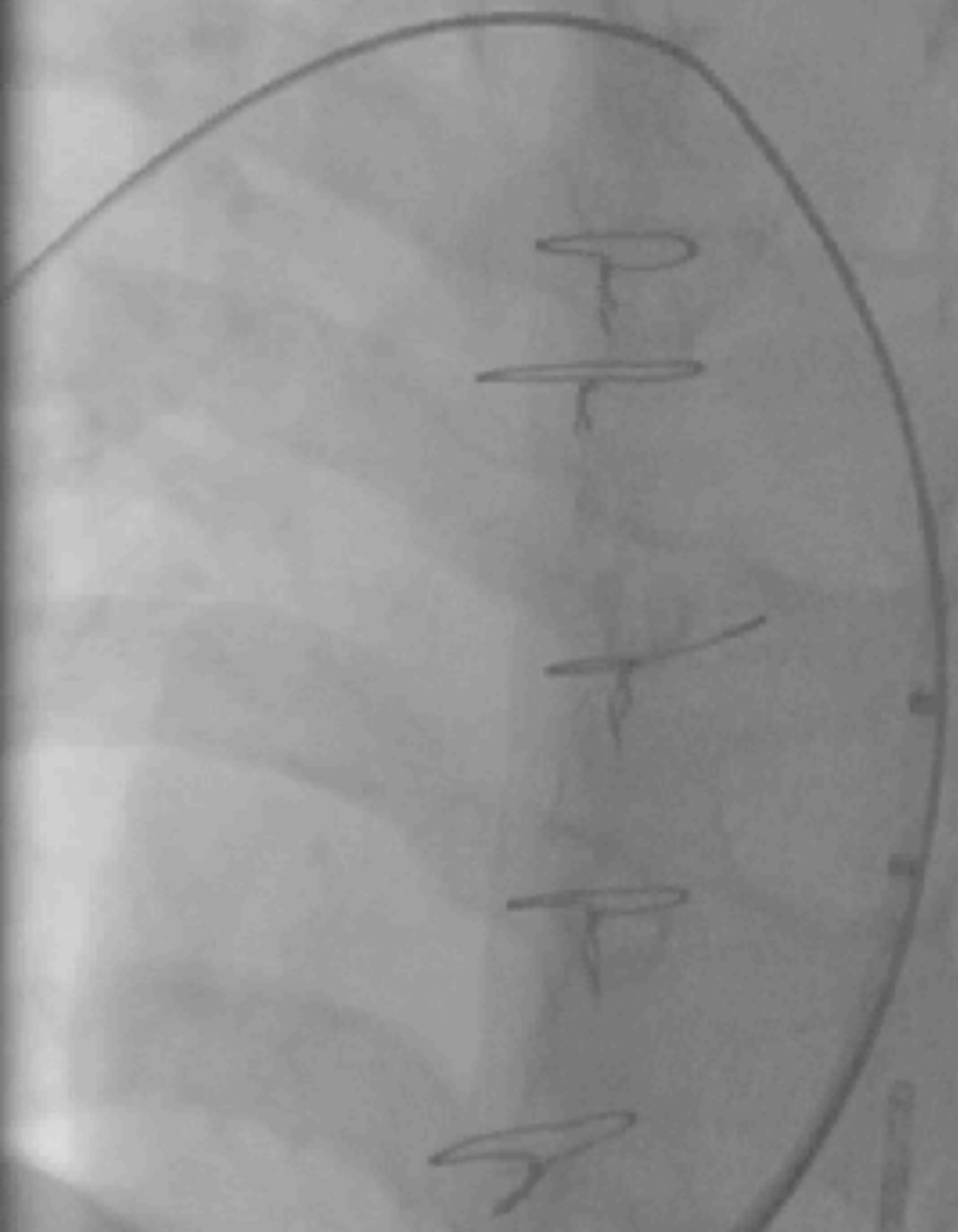
Polyethylene Terephthalate (PET) Skirt

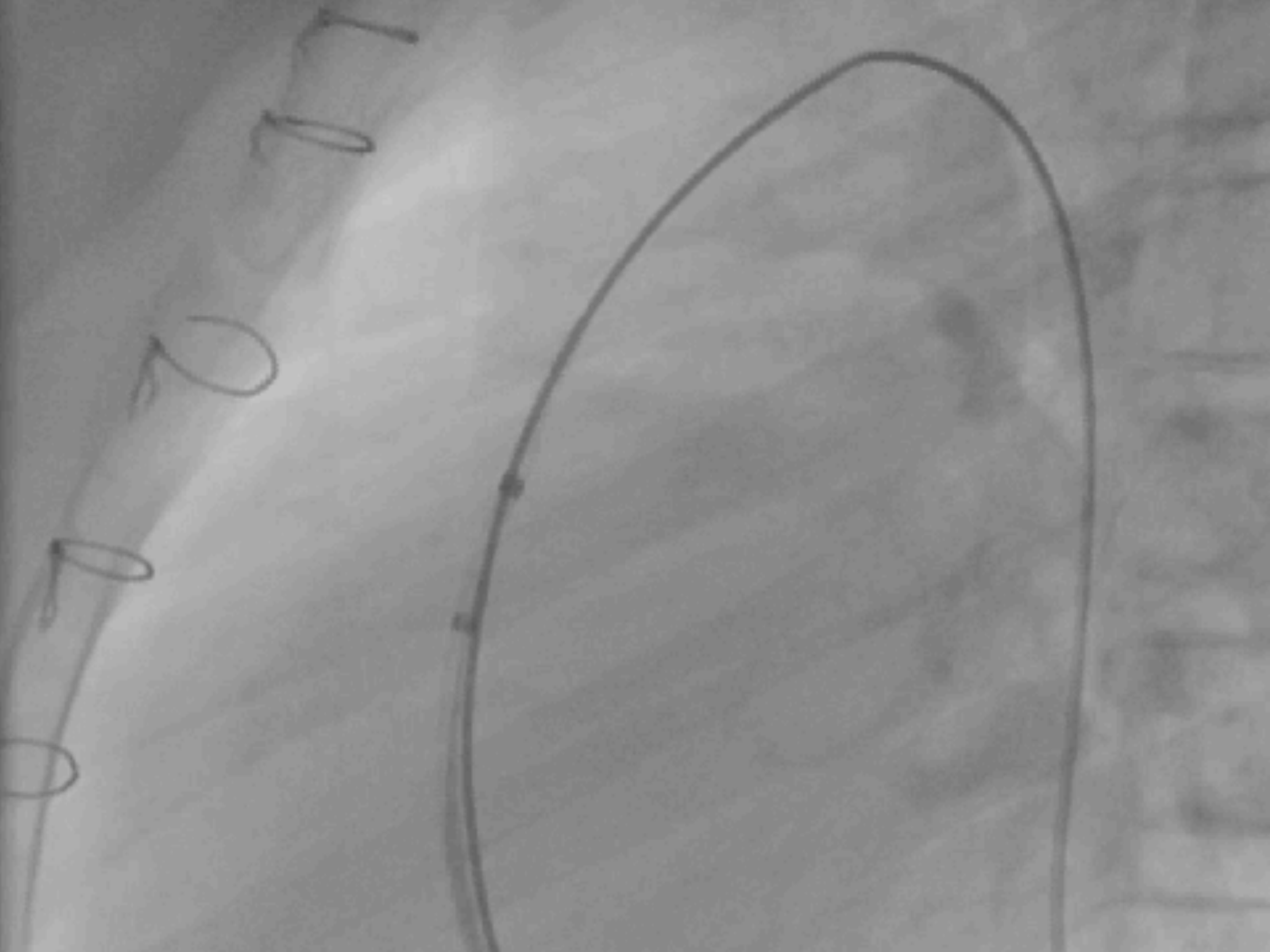


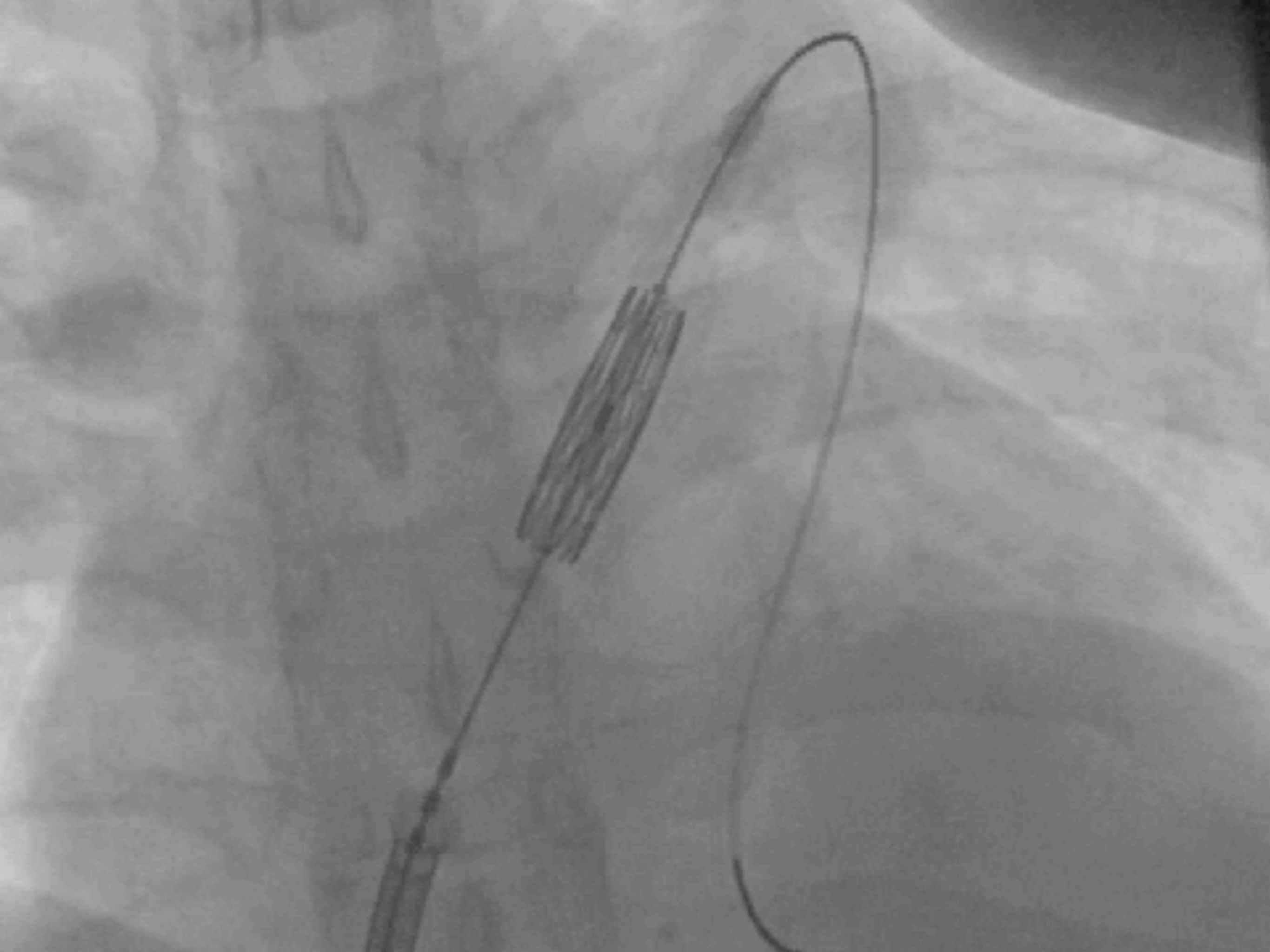


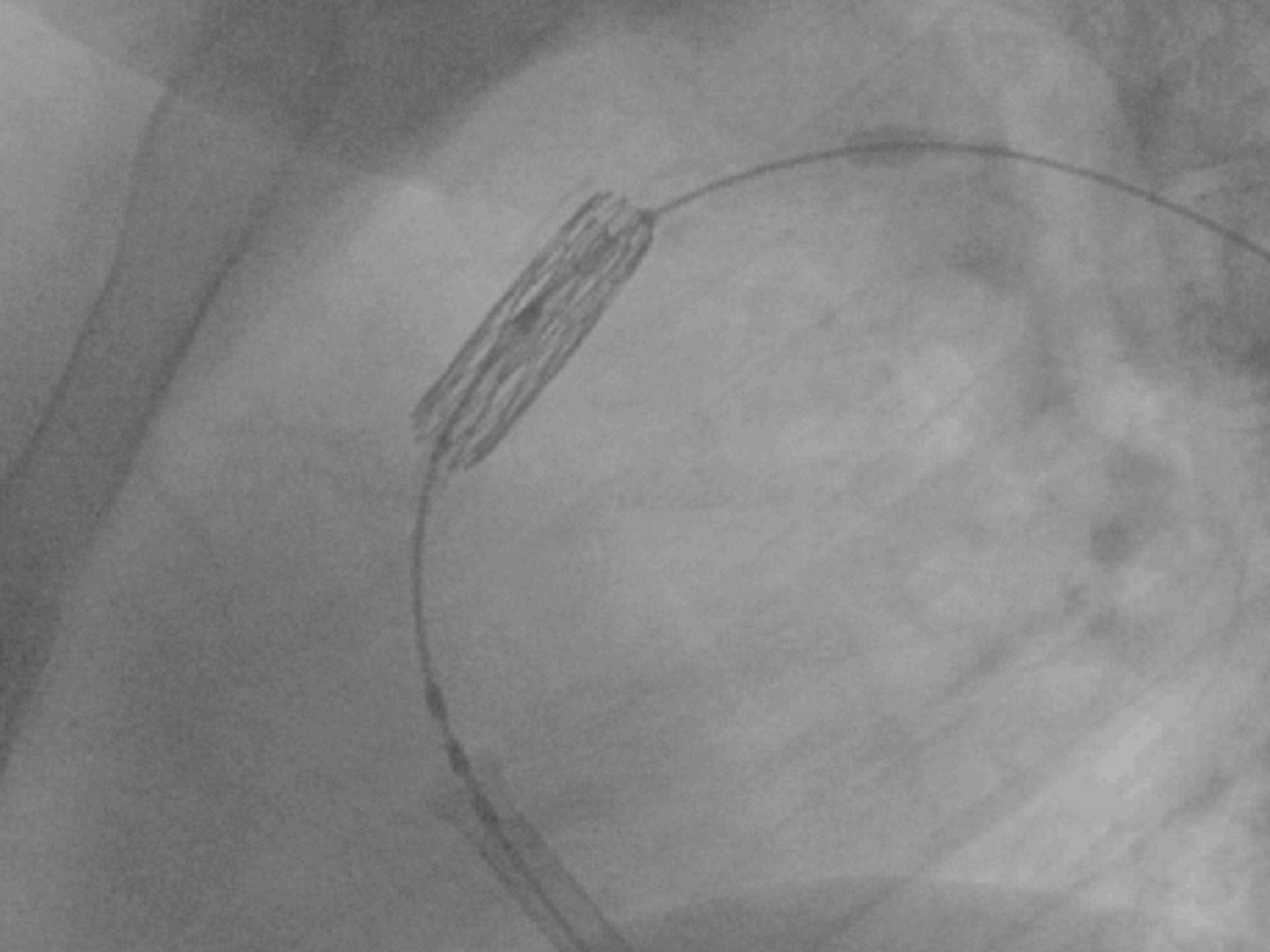


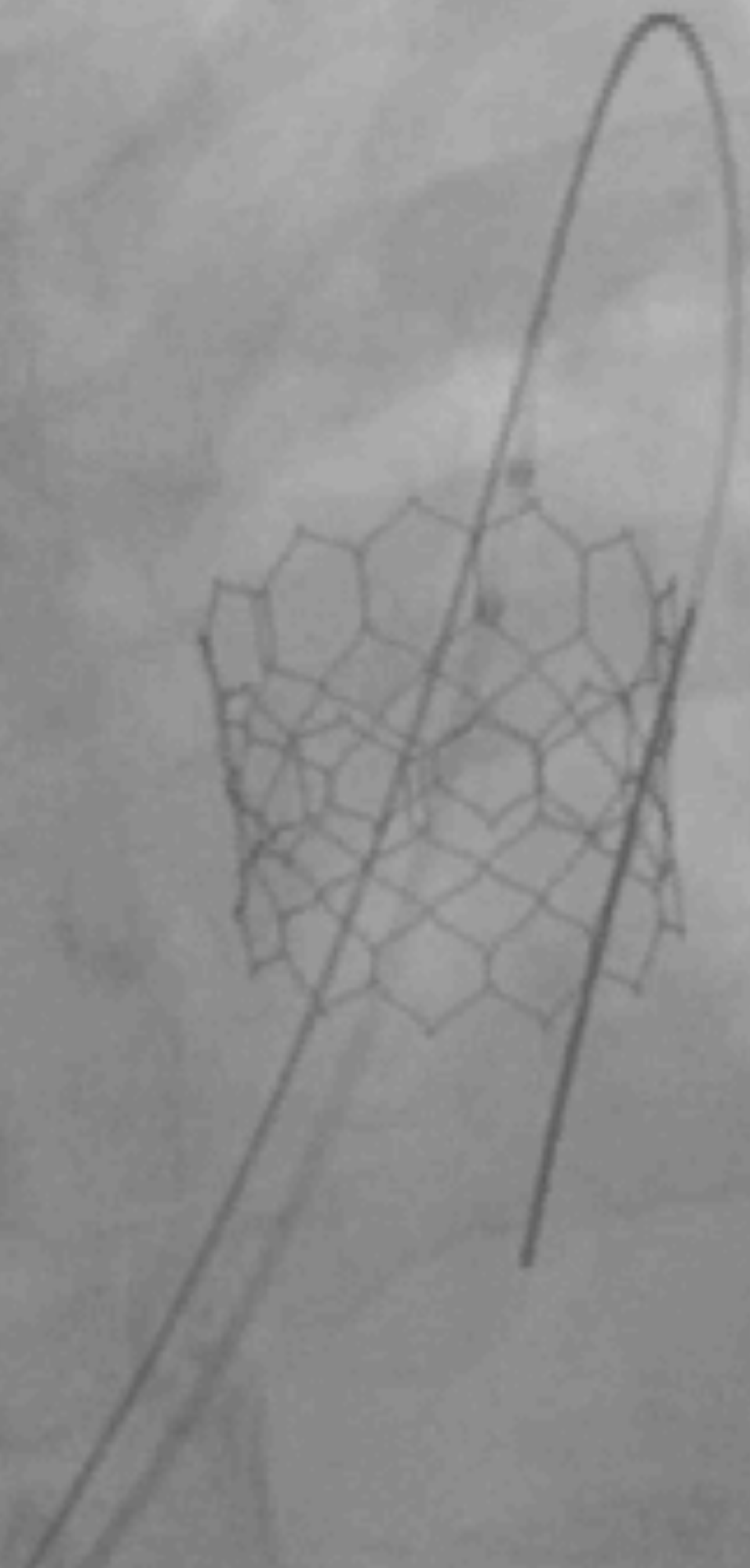


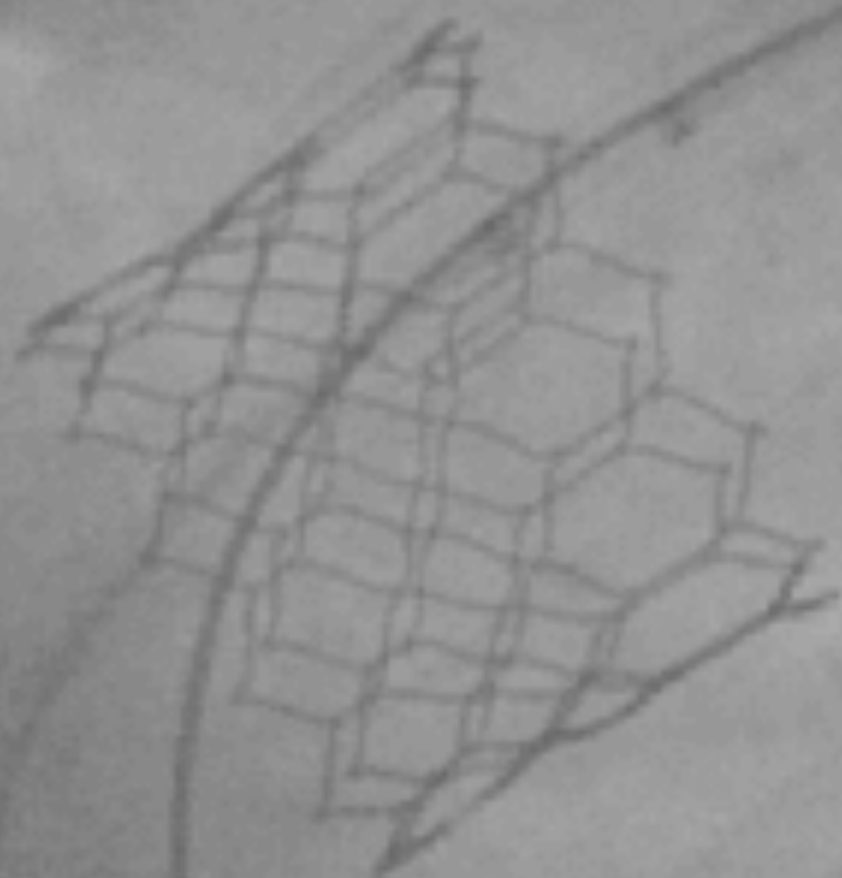














Childrens Heart Center Nevada

continuous care for the fetus, child & adult with congenital heart disease

Thank You !

