Thursday

Findings in Repaired Congenital Disease: *Expected Findings or a Problem?*

Raymond F. Stainback, MD, FASE
President, American Society of Echocardiography
Chief, Non-invasive Cardiology
Texas Heart Institute at
Baylor St. Luke's Medical Center
Assoc. Prof., Baylor College of Medicine
Houston, TX

ECHO HAWAII™ 31st ANNUAL

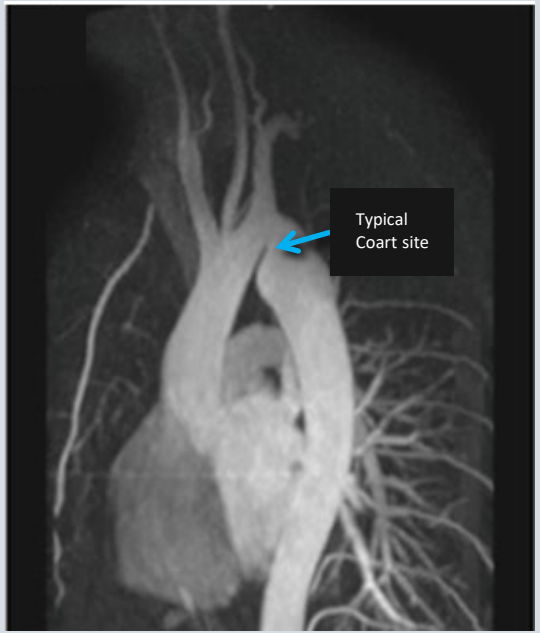
January 20, 2022

Disclosures: *None*

Case - 1

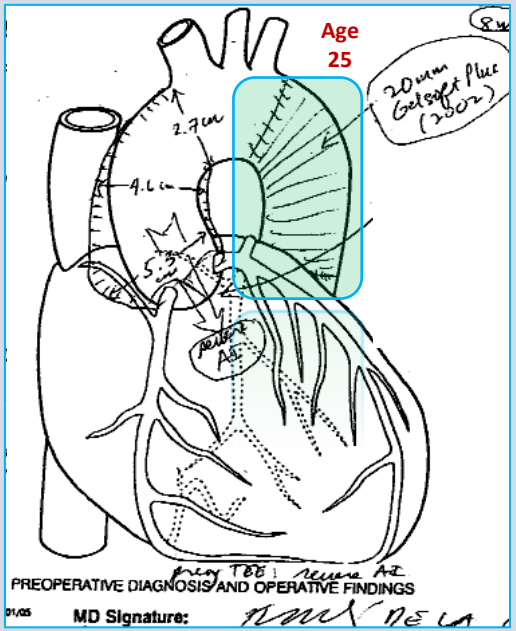
37 yr m. Swiss professional

- **Age 25**
  - Aortic Coarctation repair (right)
  - Dilated Aortic Root 5.0 cm (stable)
  - Moderate Aortic Regurgitation
  - Survillance Echo (annually)
  - CTA – periodically
- **Age 37– working in US**
  - AR now Severe
  - LV and Aorta enlarging
  - Ao Sinus of Valsalva - 5.3 cm
  - Why is there Aortic Regurgitation?



Diagnosis, imaging and clinical management of aortic coarctation

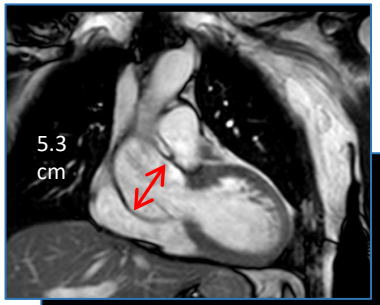
Dijkema EJ, *et al*. *Heart* 2017;**103**:1148–1155.

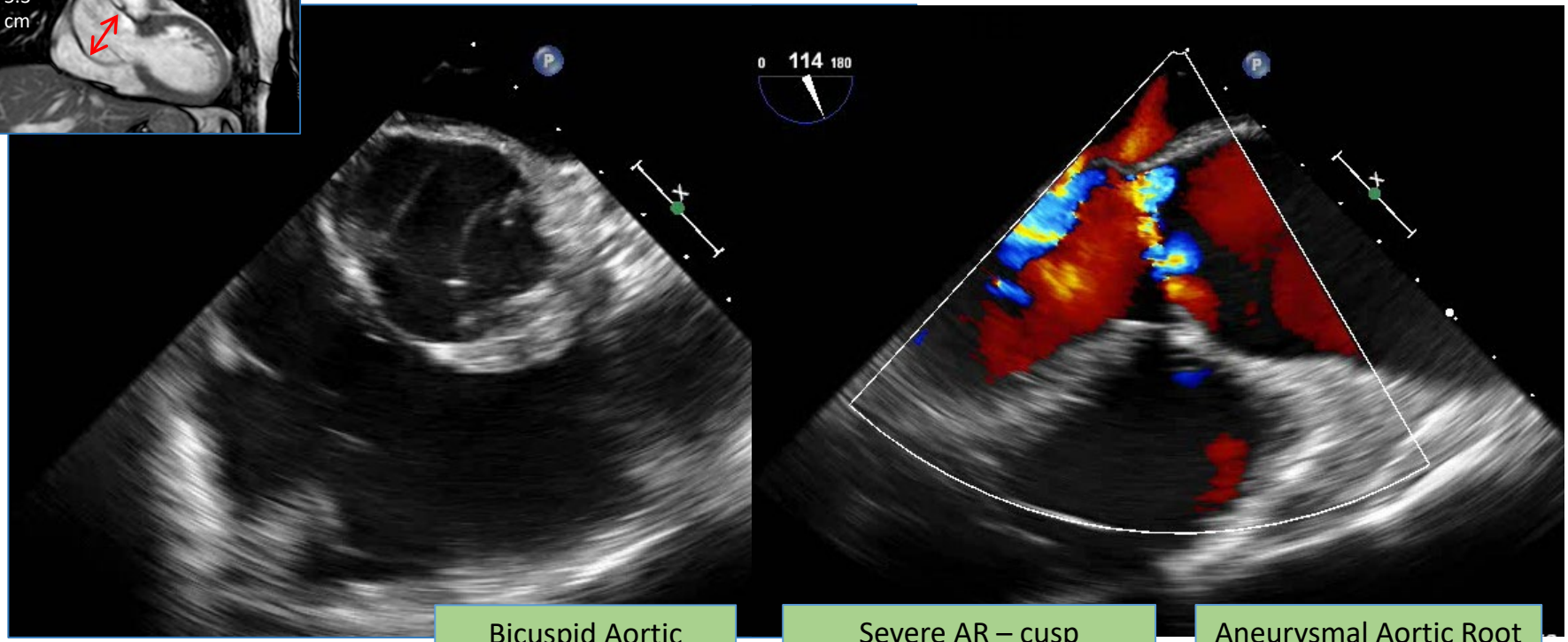


Postop

Case - 1

Intra-operative





Bicuspid Aortic Valve

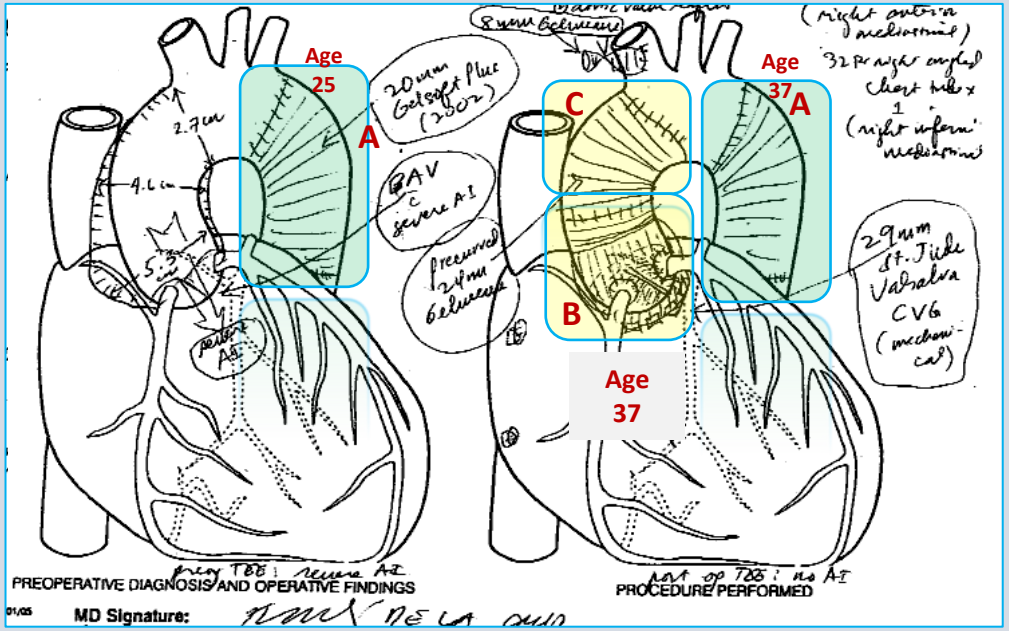
Severe AR – cusp prolapse

Aneurysmal Aortic Root

Case - 1

37 yr ♂ architect

- **Age 25**
  - Aortic Coarctation repair (A)
  - Dilated Aortic Root 5.0 cm (stable)
  - Moderate AR from Bicuspid AoV
  - Survillance Echo (annually)
  - CTA – periodically
- **Age 37**
  - AR now severe
  - LV and Aorta enlarging (Ao 5.3 cm S of Val)
  - Bental procedure (B)
  - Asc. Ao -Hemi-arch repair (C)



Preop Postop

Diagrams from Dr. Joseph Coselli

Case - 1

Coarctation of the Aorta

+

Bicuspid Aortic Valve

?

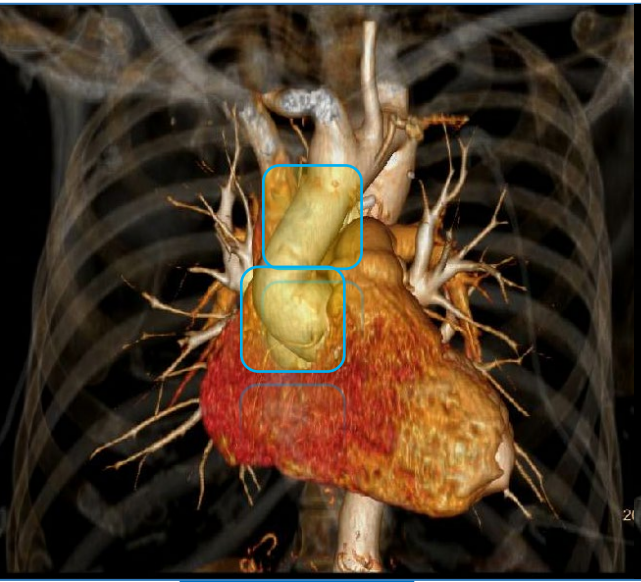
expected or *unexpected ?*

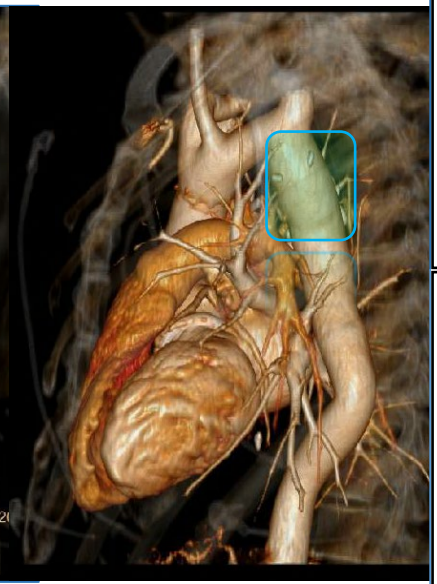
Case - 2

Coarctation of the Aorta		Bicuspid Aortic Valve
0.3-4 of 1000 births (♂ predom)		1-2 of 100 births (♂ predom)
Bicuspid aortic Valve (50-75%)		Coarctation of the aorta (~7%)
VSD - ASD		Aortopathy (more common in BAV + CoA)
PDA		Shone's complex
Shone's complex		
Cerebral aneurysm (2.5-10%)		

expected or *unexpected*

Case - 1



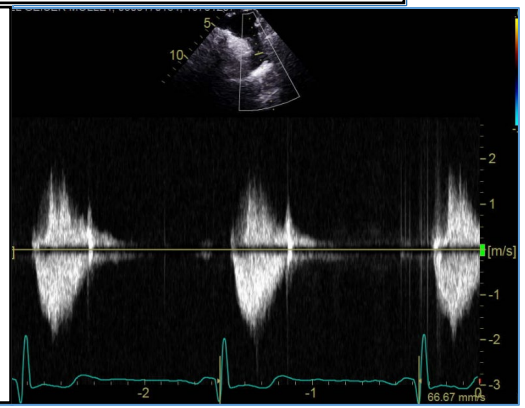


Age 37

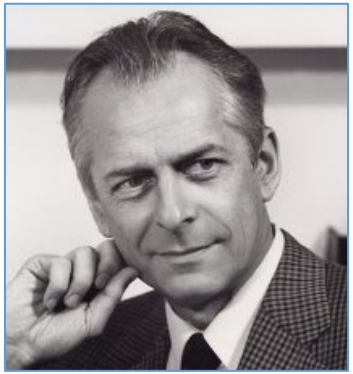
hemi-arch graft

Bental - root graft

Age 25 – coarct repair



Suprasternal notch view – systolic velocity 1.8 m/sec (acceptable / expected)



Marko Turina, MD

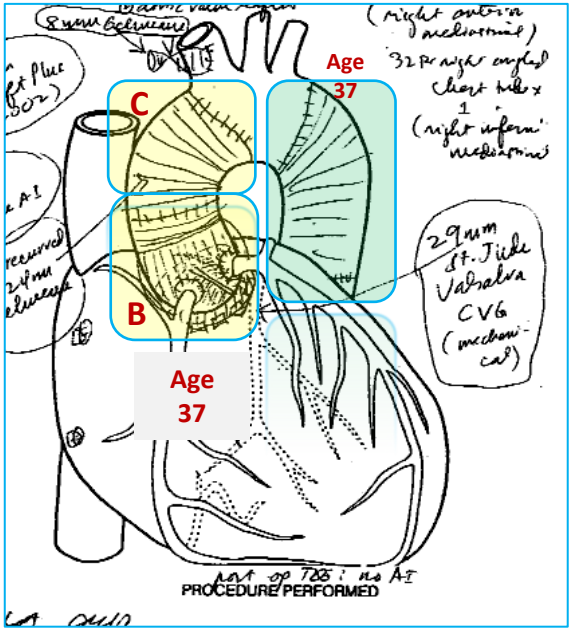
Zurich

- Director Clinic for Cardiovascular Surgery Chairman, Department of Surgery **Univ Hospital Zurich**
- b. 1937, Yugoslavia
- 1964 - University hospital of Zürich under Swedish surgeon Dr. Ake Senning (Karolinska Inst.) [Senning for TGA & first PPM implant 1958].
- 1985 chief of Center for Heart and Blood Vessels Diseases.
- Retired in 2004

Joseph Coselli, MD, FACS

Houston

- **BCM – Chief, Cardiothoracic Surgery**
- **THI – Chief, Adult Cardiac Surgery**
- **Mentor:** Aortic Surgery Pioneer **Stanley Crawford, MD**
- **Outcomes of 3309 thoracoabdominal aortic aneurysm repairs *J Thor & Cardiovasc Surg. 2015 (May) – a single practice experience*.**
- performed more than 7,500 surgical repairs of the aorta and over 3,300 open repairs of the thoracoabdominal aorta - world's most experienced surgeon



Postop

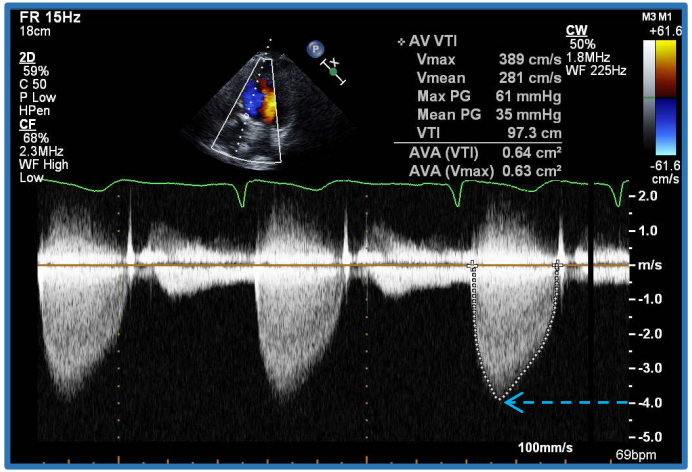
Case - 2

67 y. law enforcement admin.

- 1967 - Age 13: Aortic Coarctation repair

  (Dr. Stanley Crawford)

- Mild aortic stenosis –adulthood surveillance
- 2016 - Age 62 easy fatigue and DOE
  - Harsh systolic murmur
  - Echo – Severe Aortic Stenosis



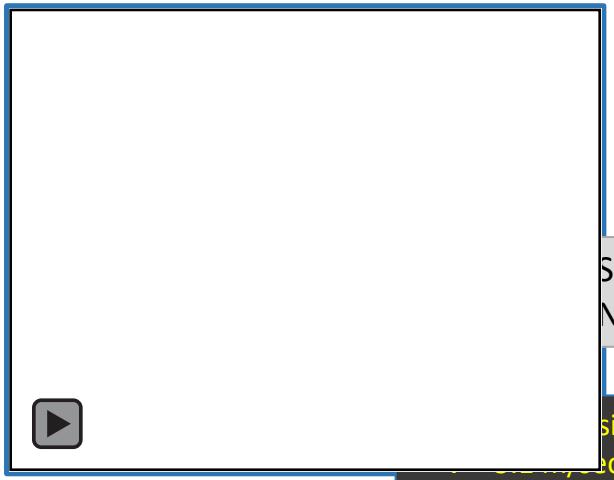
AoV Area – 0.6 cm^2

Case - 2

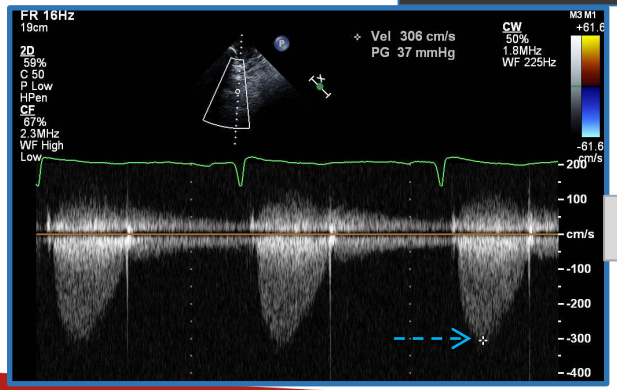
67 y. law enforcement admin.

- 1967 - Age 13: Aortic Coarctation repair (Dr. Stanley Crawford)
- Mild aortic stenosis –adulthood surveillance
- 2016 - Age 62 easy fatigue and DOE
  - echo shows severe Aortic Stenosis
  - Echo SSN view: V ~ 3.1 m/sec Ao isthmus

- Is residual coarct gradient **37 mm Hg** ($4V^2$)?
- Does modified Bernoulli equation apply?
- Should we intervene?

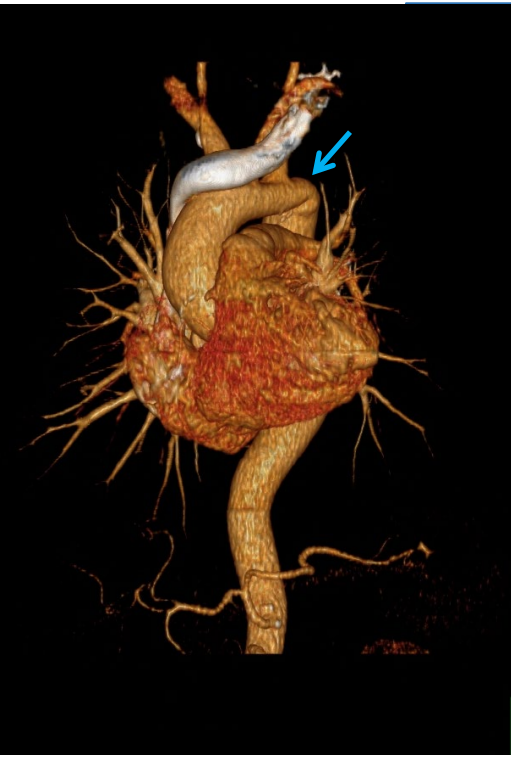


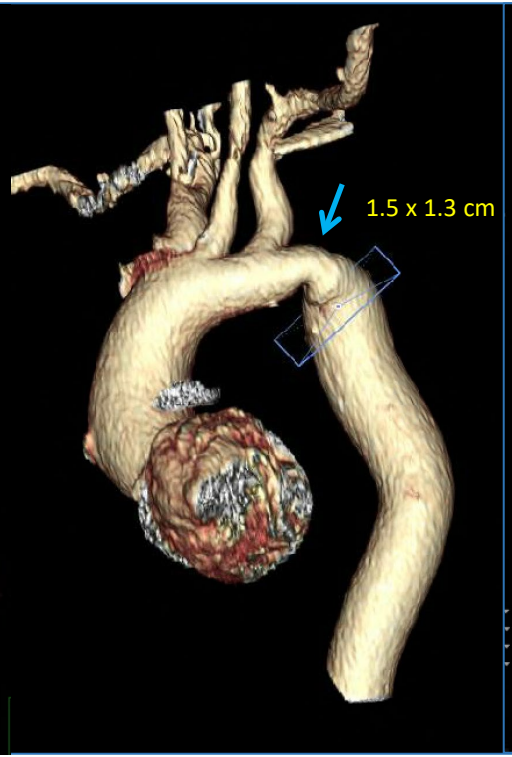
Suprasternal Notch

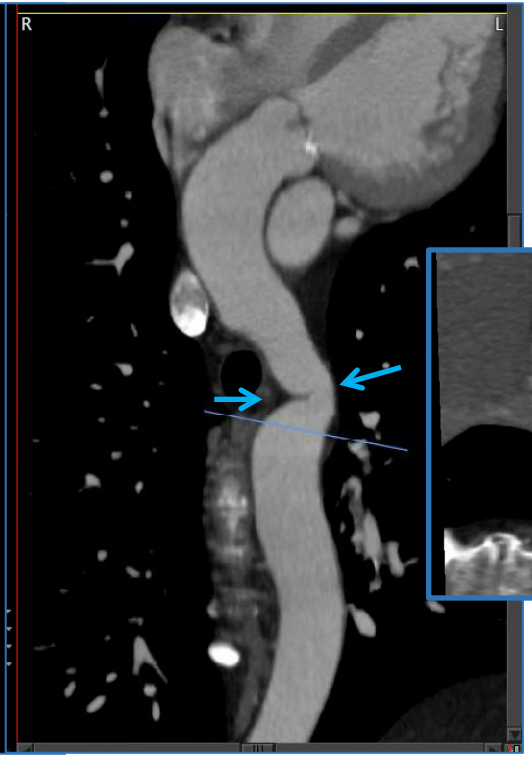


Expected?

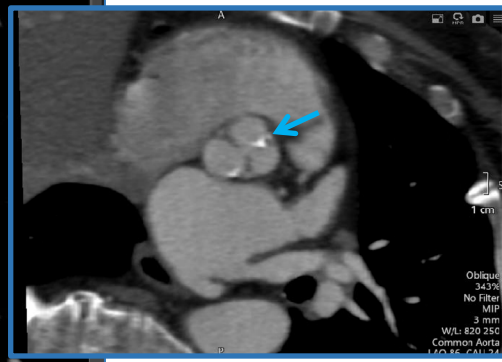
Narrowing – prior aortic Coarctation repair







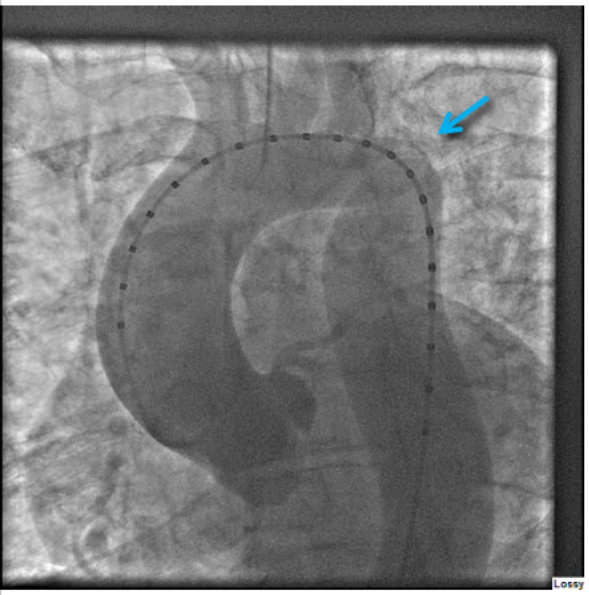
Bicuspid Aortic Valve
Fusion Raphe (Ca++)



CCT 2016

Case - 2

2016 –preop Cath

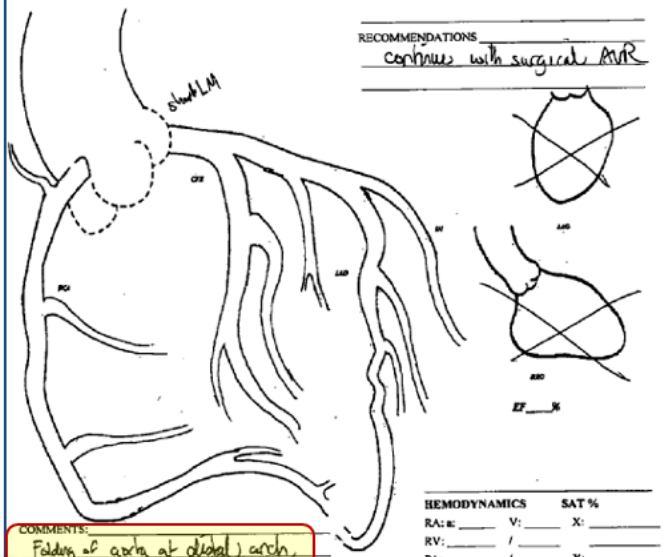


CHI St. Luke's Health

CORONARY ANGIOGRAPHY

DATE OF CATH: 6-16-2016 INDICATIONS: Coarctation s/p repair, severe AS

RECOMMENDATIONS: continues with surgical AVR



COMMENTS: Folding of aorta at distal arch, <10mmHg gradient

CARDIOLOGIST SIGNATURE: DATE: 6/16/16 TIME: 14:00

FELLOW SIGNATURE: DATE: 6/16/16 TIME: 14:00

PS-2313

HEMODYNAMICS **SAT %**

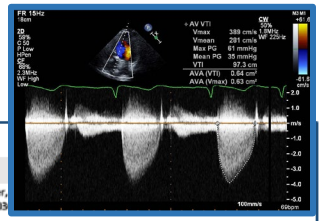
RA: a:	V:	X:
RV: /	/	
PA: /	/	X:
PCP: a:	V	X:
AO: 110	/	
LV: 160	/ EDP 23	
CI:	BSA	

Page 1 of 1

"folding of aorta at distal arch, < 10 mm Hg gradient" 2016

Expected?

SSN > 3 m/sec



Post mechanical AVR – 2021 f/u

Baylor College of Medicine

St. Luke's Medical Tower, Houston TX, 7703

LOWER EXTREMITY ARTERIAL DUPLEX EXAM

Name:
MRN:
History: Lower extremity claudication. Coarctation of the aorta (repaired), AS with AVR. History of HTN, DM, and hypertrieglyceridemia.

RIGHT:	PSV
CIA:	90cm/s
EIA:	142cm/s
CFA:	99cm/s
PFA:	42cm/s
Prox SFA:	88cm/s
Mid SFA:	70cm/s
Dist SFA:	65cm/s
Prox POP:	52cm/s
Dist POP:	56cm/s
Prox PTA:	60cm/s
Mid PTA:	59cm/s
Dist PTA:	59cm/s
Prox Pero:	42cm/s
Dist Pero:	36cm/s
Prox ATA:	46cm/s
Mid ATA:	37cm/s
Dist ATA:	39cm/s
DPA:	29cm/s

Blood Pressures

Brachial 141mmHg

Brachial 135mmHg

DPA 140mmHg
PTA 138mmHg
DP ABI 0.99
PT ABI 0.97

DPA 138mmHg
PTA 136mmHg
DP ABI 0.97
PT ABI 0.96

Left:	PSV
CIA:	92cm/s
EIA:	110cm/s
CFA:	76cm/s
PFA:	52cm/s
Prox SFA:	78cm/s
Mid SFA:	68cm/s
Dist SFA:	70cm/s
Prox POP:	61cm/s
Dist POP:	57cm/s
Prox PTA:	56cm/s
Mid PTA:	50cm/s
Dist PTA:	46cm/s
Prox Pero:	30cm/s
Dist Pero:	35cm/s
Prox ATA:	42cm/s
Mid ATA:	49cm/s
Dist ATA:	50cm/s
DPA:	28cm/s

Comments: Post coarctation repair of the aorta, there is no significant differences between the right and left brachial pressures with normal right and left ankle/brachial indices.

Patent iliac arteries with mild calcific

Patent femoropopliteal arteries with

Patent three vessel runoff to the ank

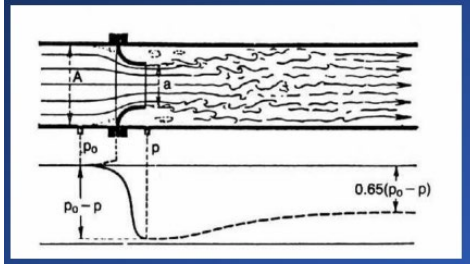
Performed By: Calvin Ng, RVT

Date: 8/26/2021

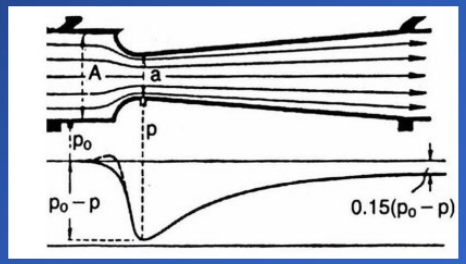
"No significant differences between R. & L. Brachial pressures with normal R. & L. ankle/brachial indices" 2021

Pressure Recovery

Outlet geometry allowing gradual expansion of streamlines eliminates flow separation and prevents turbulence (pressure recovery)



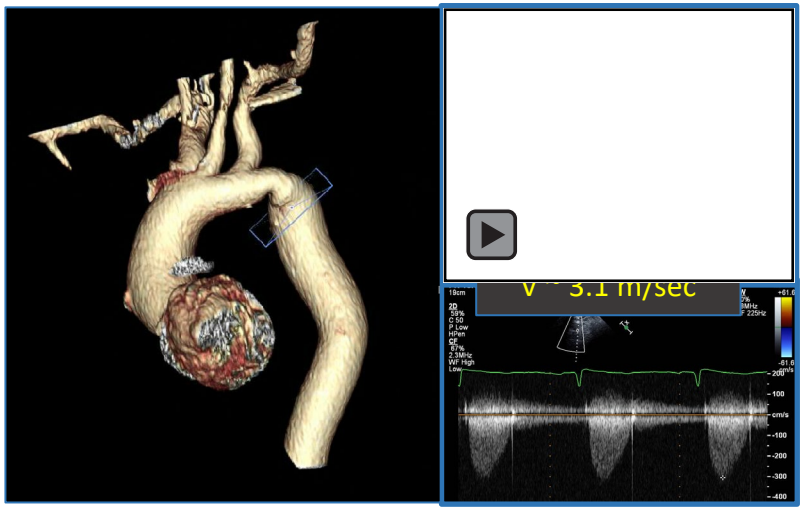
Abrupt outflow (nozzle): turbulence, head loss



Gradually expanding outflow: eliminates flow separation and recovers the pressure drop

http://assets.escardio.org/Assets/Presentations/OTHER2010/EAE-clinical-application-echo/15.Schwammenthal-hemodynamic-assessment-aortic-stenosis.pdf

Prandtl L, Tietjens O: Applied Hydro- and Aeromechanics, New York, Dover 1957



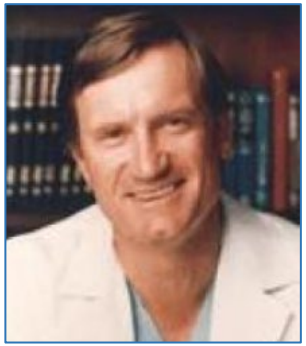
congenital AS, very small patient, Asc Ao < 3.0 cm, bi-leaflet mechanical Valves, congenital conduit repairs / tubular structures (Coarct)

Stanley Crawford, MD

Houston

UAB 1943 - Harvard Med 1946
MGH – Chief Surg Resident
US Navy _ Micheal E. Debakey recruit
Baylor College of Med 1966 – d. 1992
"Pioneer of Aortic Surgery"
Textbook: *Diseases of the Aorta - 1984*

Coarct Repair 1967 – age 13

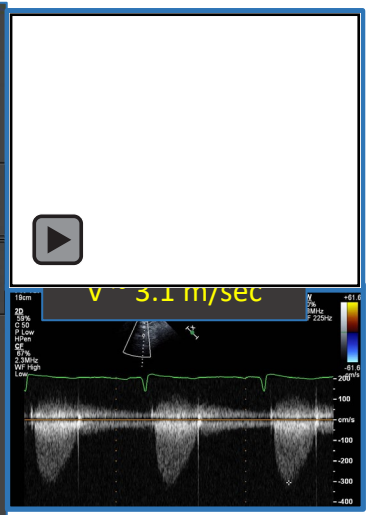


David Ott, MD

Houston

- THI professional staff for 36 years
- **Surgeon in Chief, Texas Heart Institute - succeeded Dr. Denton A. Cooley**, who held the position for 50 years
- DeBakey & Cooley mentee
- 1978 – 2015 > 20,000 Cardiac repairs
- Many hundreds of congenital heart and aortic repairs. - ret 2017

Mech AVR 2016 – age 62



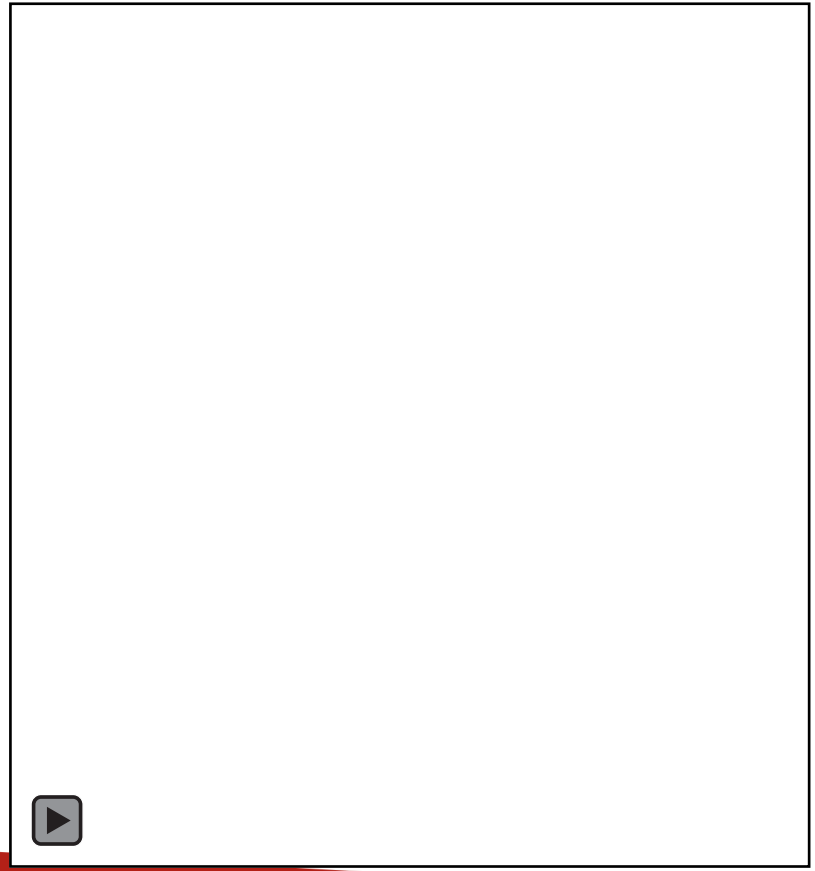
Case - 3

37 yr female with freq palpitations

-TTE → RA & RV enlargement

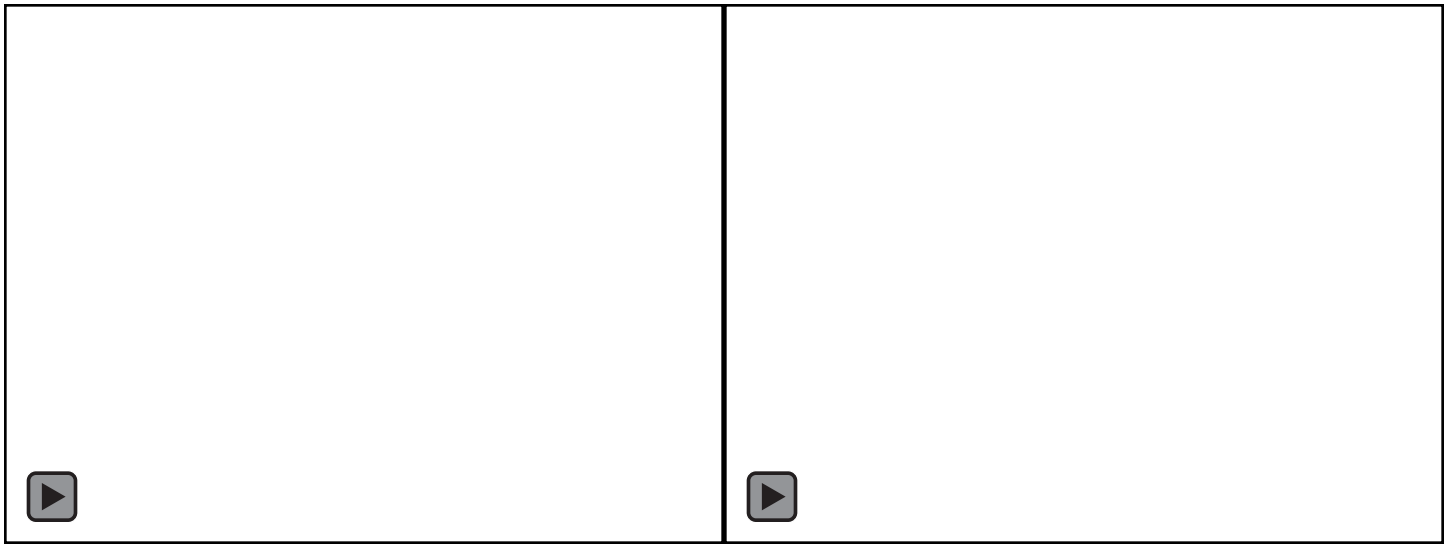
- IV Saline Study + L →R shunt

- TEE was requested



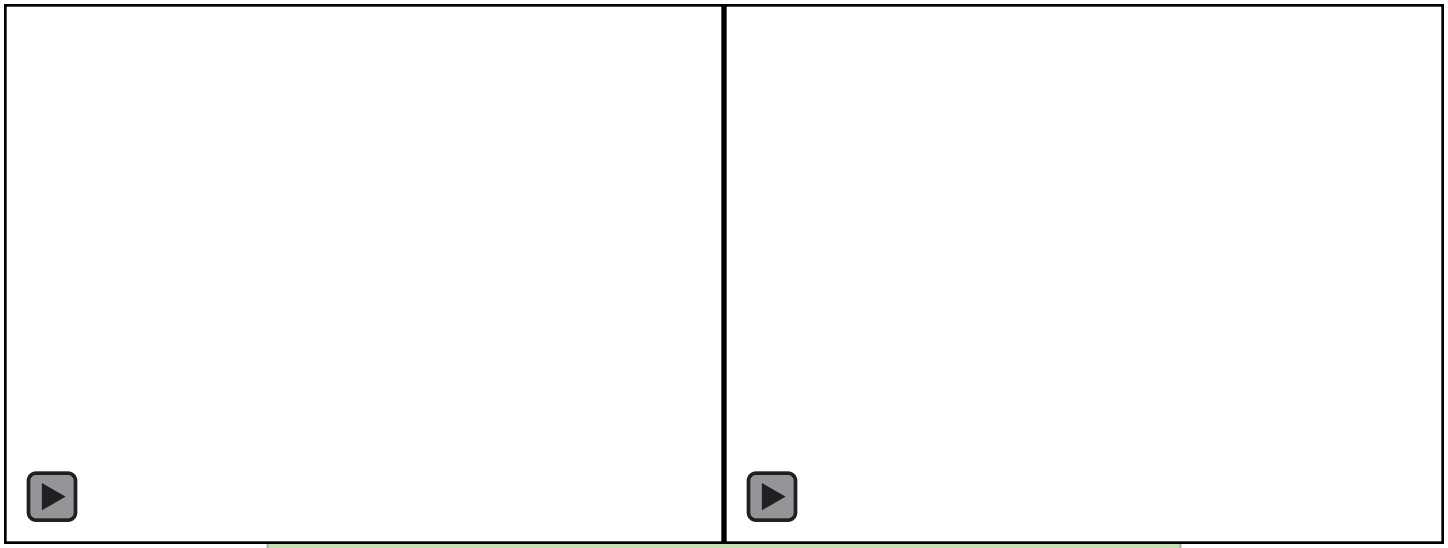
Case - 3

TEE: bicaval view



Case - 3

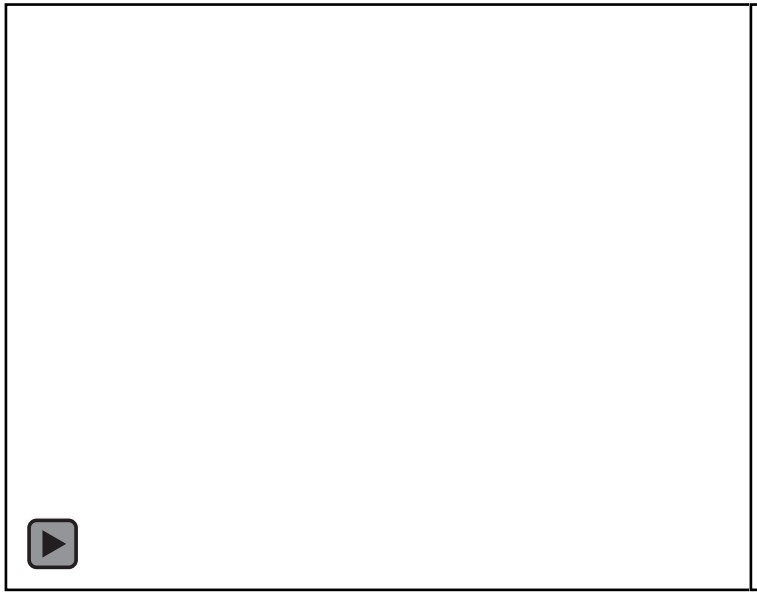
TEE: bicaval view

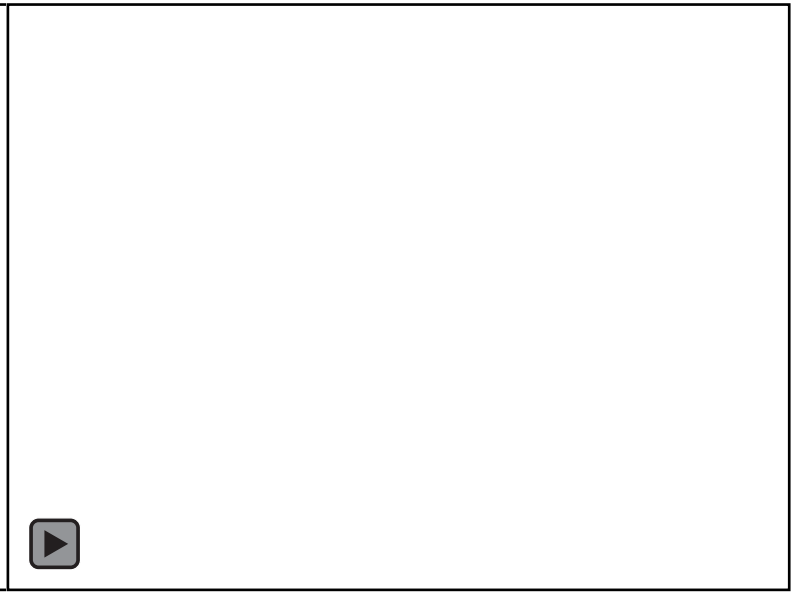


Superior Sinus Venosus ASD

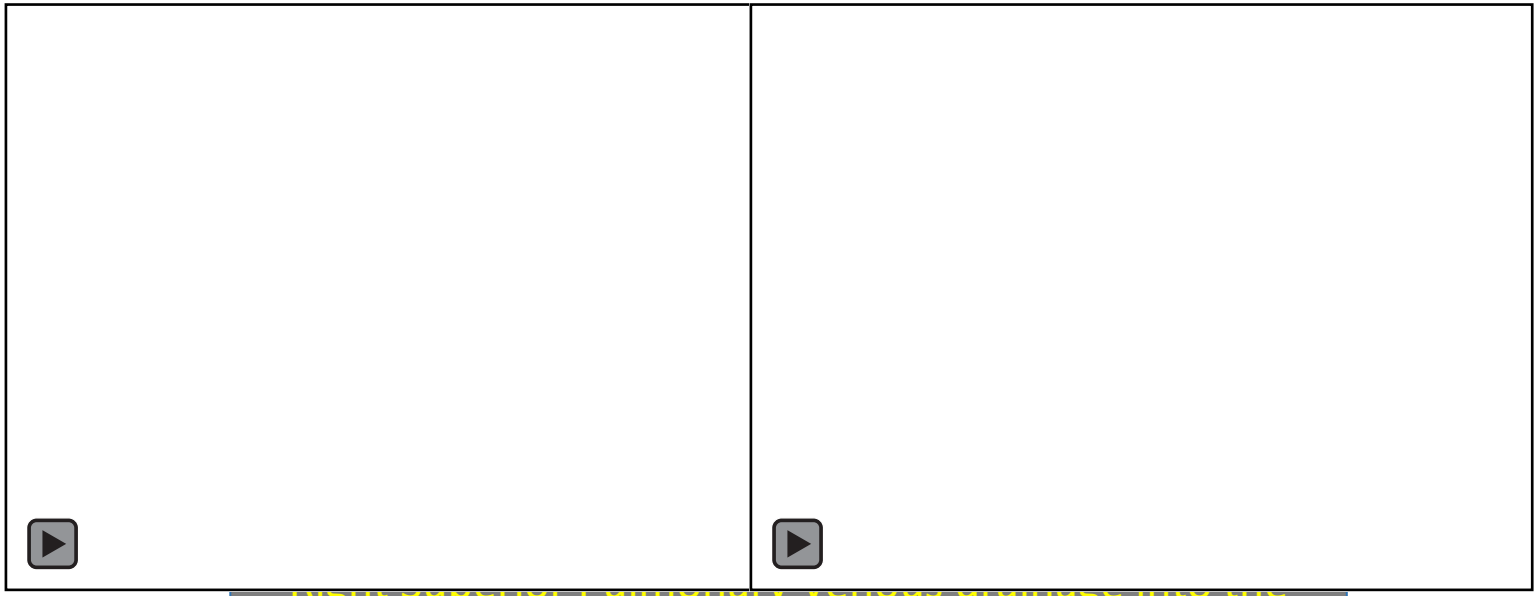
- TEE bi-caval view
- Superior to the fossa ovalis
- RA-LA communication at RA-SVC junction

Case - 3





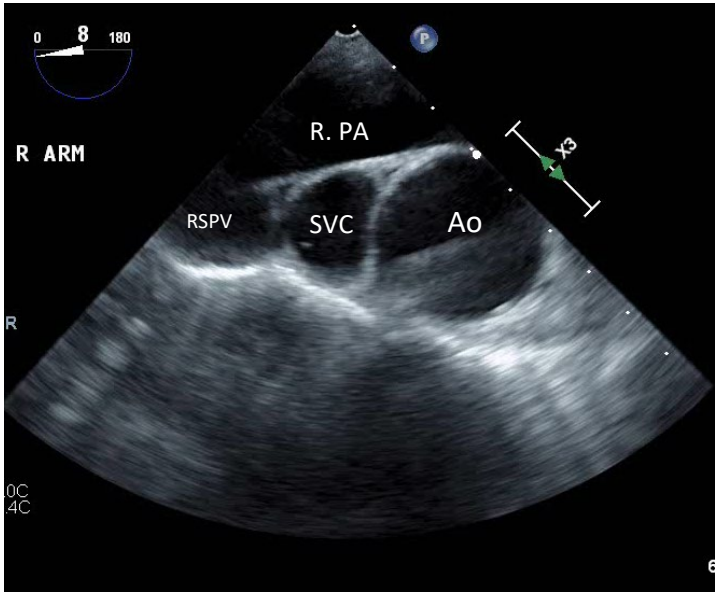
Case - 3



Right Superior Pulmonary Venous drainage into the SVC (RSPV – to – SVC)

RSPV blood shown mixing with SVC blood by IV saline contrast injection (SVC→ R. PA → aorta saline contrast)

Case - 3



Normal – 3 circles in a row

One circle (Ao) and a teardrop “Teardrop Sign”

Ammash NM et al. Partial anomalous pulmonary venous connection: diagnosis by TEE. JACC Vol.29, May 1997

Case - 3

Superior Sinus Venosus ASD

+

RSPV – to – SVC

?

expected or *unexpected ?*

Case - 3

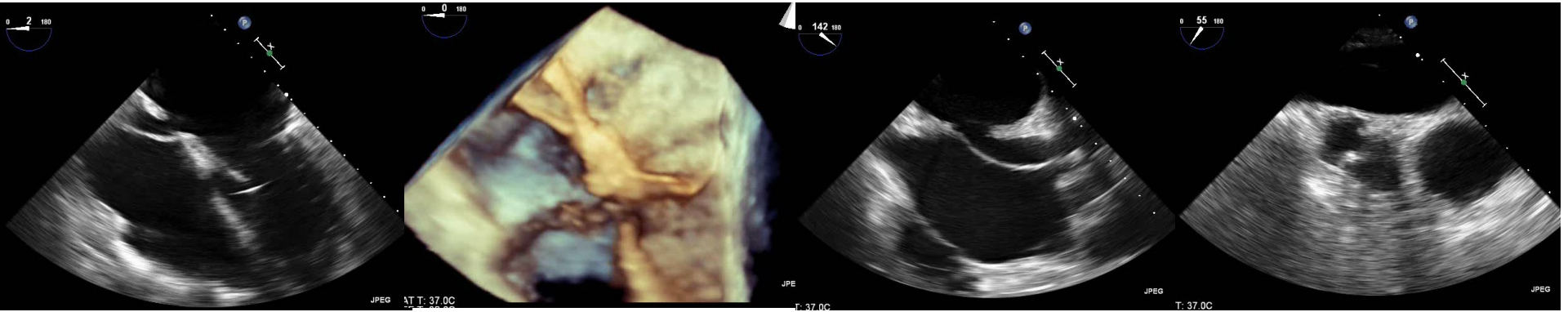
Superior Sinus Venosus ASD	+	RSPV – to – SVC
Uncommon 5-10% of ASDs > 90% partial anomalous PV connection		Uncommon defect Isolated defect (high SVC) rare
RSPV-to-SVC most cases		Superior Sinus Venosus ASD

expected or *unexpected*

Attenhofer Jost CH, Connolly HM, Danielson GK, et al.
Sinus venosus ASD . . . Circulation. 2005;112:1953-1958

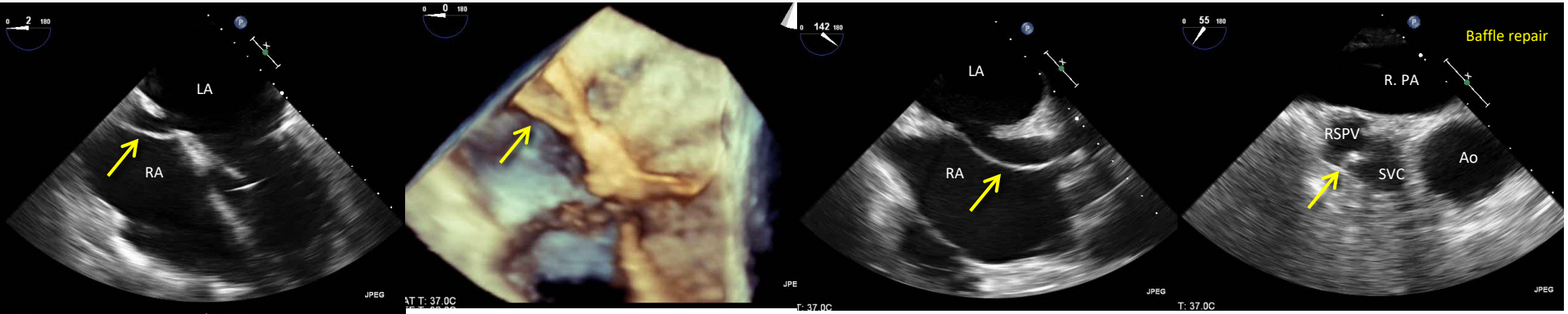
Case – 3A

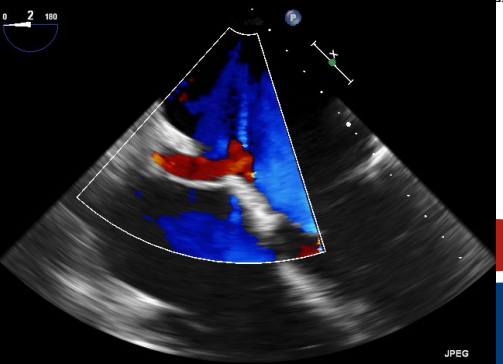
What are we looking at here ?!

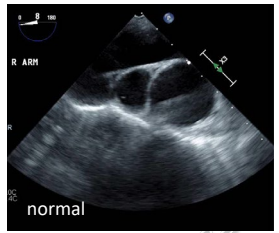


Case – 3A

Superior Sinus Venosus ASD post baffle Repair - Expected







Case - 4

54 yr asymptomatic normally active female

Pentalogy of Fallot (VSD + ASD)

1965 complete repair – age 3 (DACmd)

- louder systolic murmur "RVOT "infundibular resection"

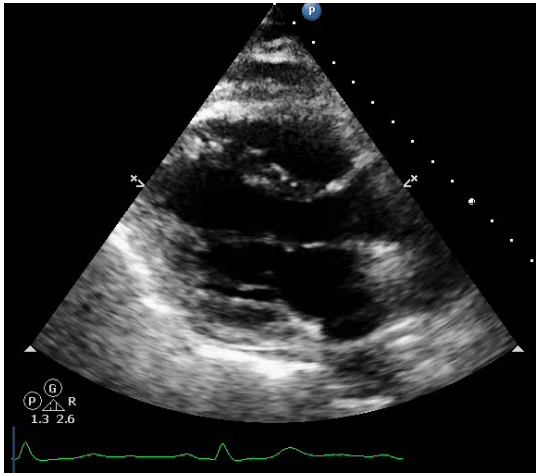
TTE – min Ao override, RVOT turbulent flow

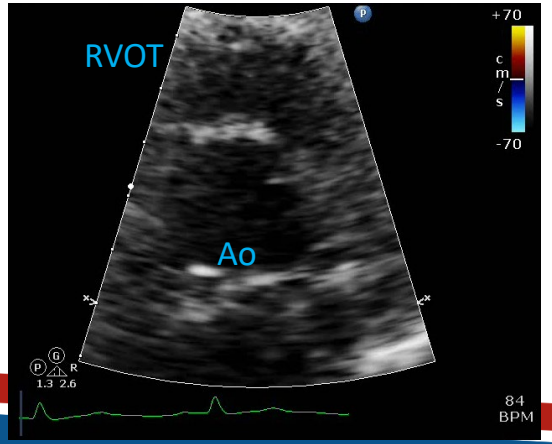
MRI – 4 yr prior – RVOT "redundant VSD patch"

MRI – recent – RVOT "redundant tissue"

R & L Heart Cath – unusual contrast in RCC

TEE Performed



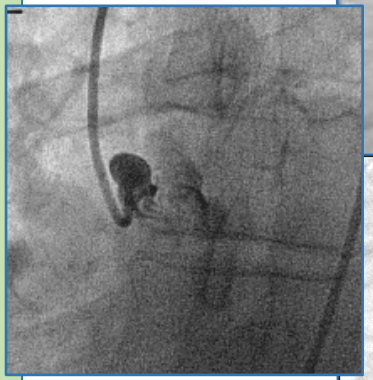


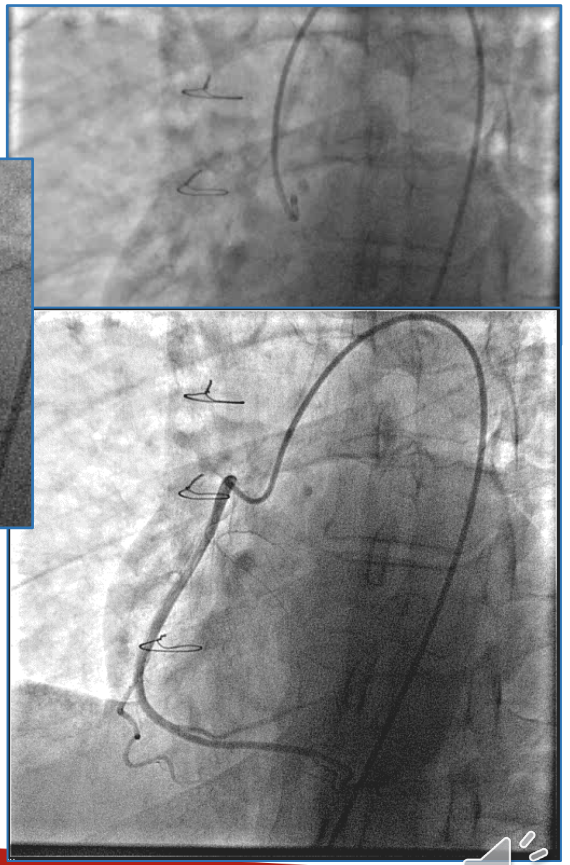
Case - 4

54 yr female

Pentalogy of Fallot (ASD)
Complete repair age 3 (DACmd)
Adulthood: louder systolic murmur
TTE – minimal Ao override, RVOT turbulent flow
MRI – 2011 – RVOT redundant VSD patch material
MRI – 2014 – RVOT "redundant tissue"
R & L Heart Cath – unusual contrast in RCC pouch
TEE Performed

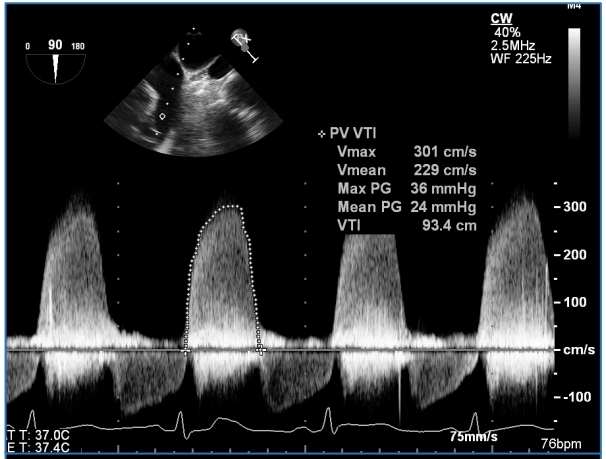
Age 3 repair
- VSD
- ASD
- RVOT infundibular resection
- small PV intact

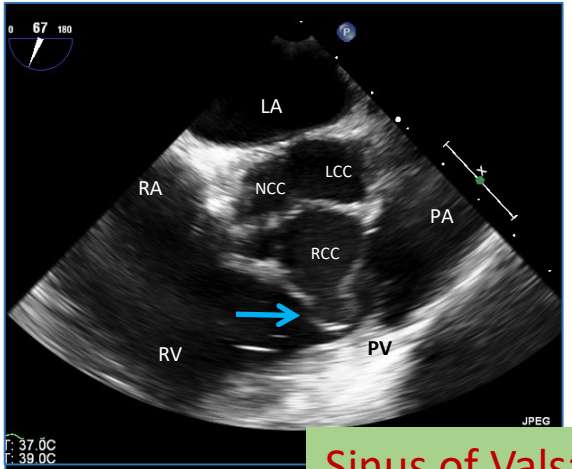


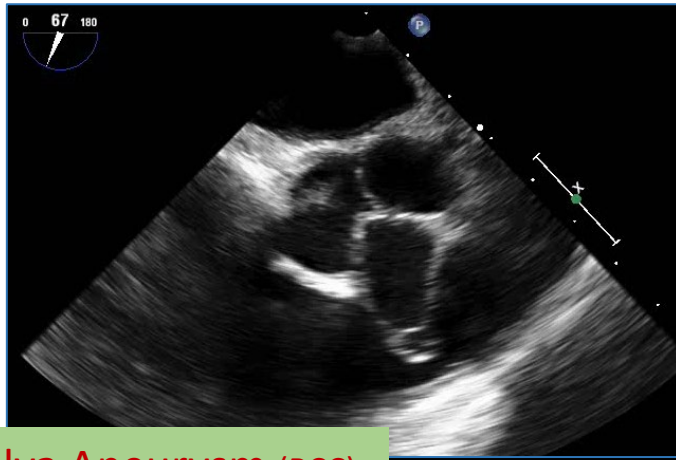


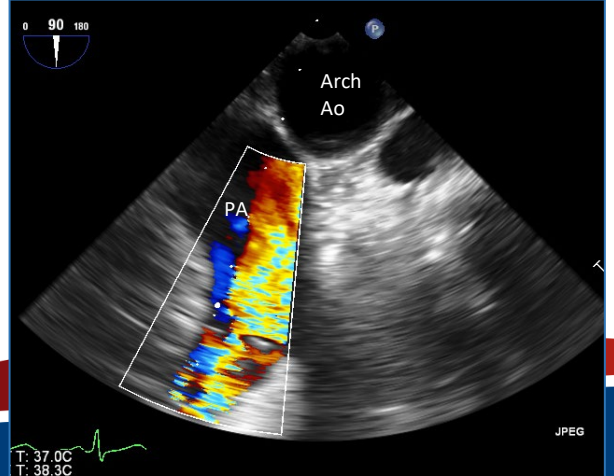
Case - 4

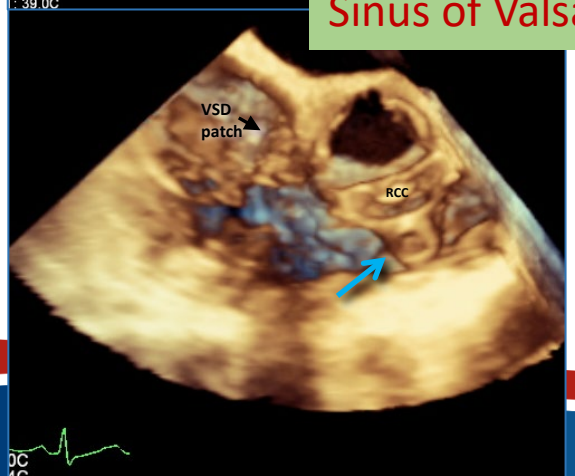
RVOT grad = 36 / 24 mm Hg (arch view)
(50 mm Hg gastric view)











Sinus of Valsalva Aneurysm (RCC)

Case - 4

Pentalogy / Tetralogy of Fallot

+

Sinus of Valsalva Aneurysm

?

expected or *unexpected*

Case - 4

Tetralogy of Fallot

1. RVOT obstruction
2. Overriding Aorta
3. **Ventricular Septal Defect**
4. RV Hypertrophy
5. ASD* / PDA

- right-sided Aorta
- coronary artery anomalies
- distal pulmonary stenosis / atresia
- AV canal defects ; other valve disease

+

Sinus of Valsalva Aneurysm

Rare (0.2-2% CV surg pts) ♂ 4:1

RCC (80%) > NCC (20%) >> LCC

Ventricular Septal Defect

Bicuspid AoV

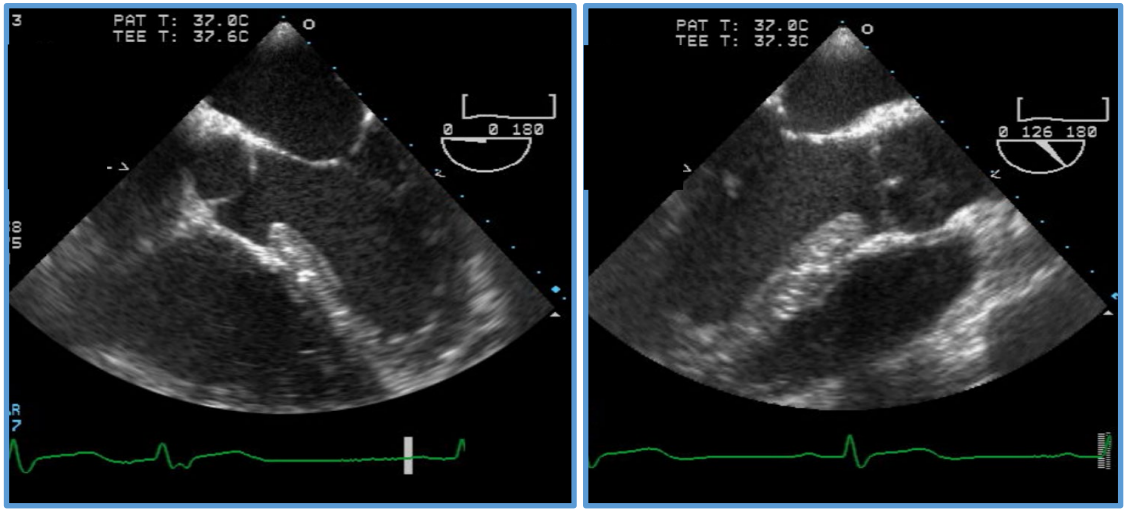
Coactation of Aorta

most are Isolated defects

expected or *unexpected*

Case 5

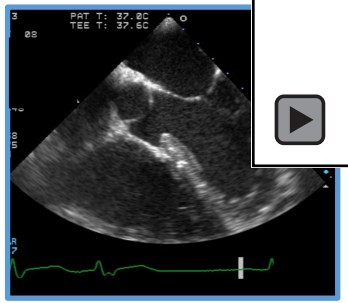
42 yr male, remote complex congenital repair – TEE to rule out VSD patch leak.

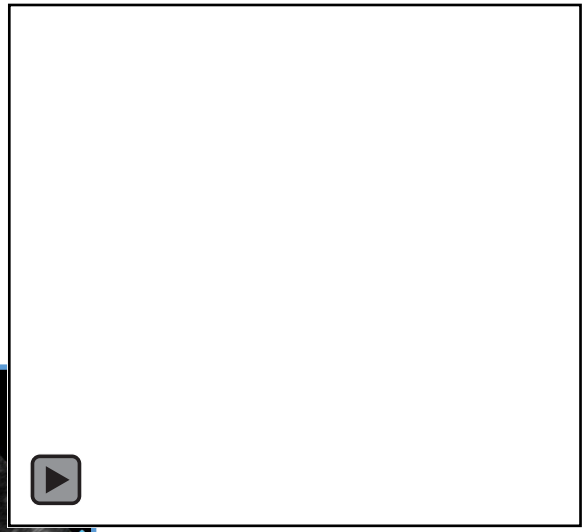


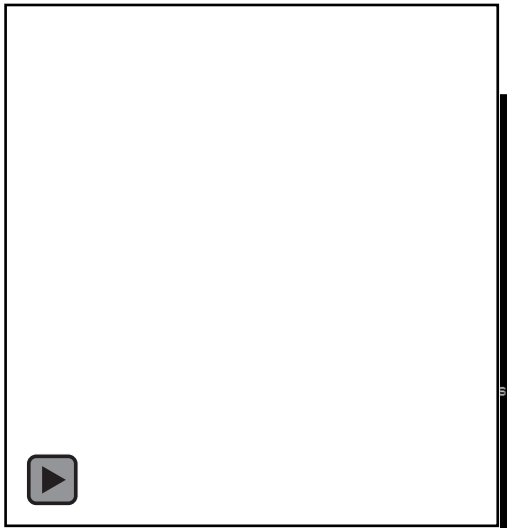
A. What is the likely congenital defect?
B. What other valve disease is most likely? - TV or PV

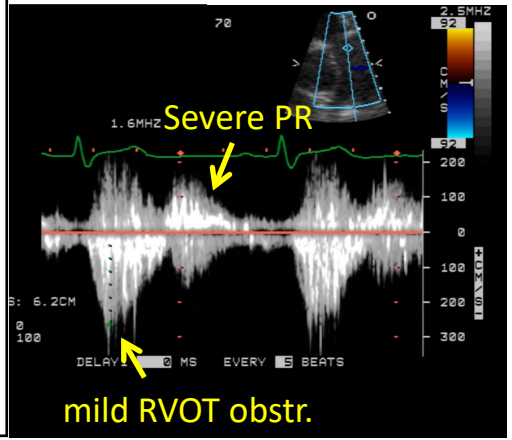
Case 5

Overriding aorta









A. What is the likely congenital defect? **TOF**
B. What other valve disease is most likely? - TV or **PV**

No residual VSD – AR striking perimembranous VSD patch
Overriding aorta – **EXPECTED repaired TOF findings**

Thank you!

stainbac@bcm.edu