

Interventions in Adult Congenital Heart Disease: Role of CV Imaging

Sangeeta Shah MD, FACC, FASE
Associate Professor, Ochsner Clinical School of Medicine
Advanced CV Imaging and Adult Congenital Heart Disease
New Orleans, LA



Adult Congenital Heart Disease

Heterogeneity Anatomy and Complications

CV Imaging

Percutaneous/Surgical

CV imaging and Interventions

- Atrial septal defect
- Tetralogy of Fallot
- D- TGA with atrial switch

Atrial Septal Defect: Recommendation for Closure

CLASS I
Closure is indicated for RA or RV enlargement with or without symptoms
CLASS IIa
Paradoxical embolism
Documented orthodeoxia-platypnea

ACC/AHA 2008 Guidelines for Management of Adults with CHD

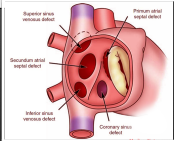
PERCUTANEOUS	SURGERY
Secundum	All other types
	Fenestrated ASD

ASE GUIDELINES & STANDARDS

Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: From the American Society of Echocardiography and Society for Cardiac Angiography and Interventions

Frank E. Silver, MD, FASE, Chair; Meryl S. Cohen, MD, FASE, Co-Chair; Laurie B. Armbray, MD, FSCAI, Nina J. Brunck, MD, DM, FASE, Craig E. Frickman, MD, FASE; Ziyad M. Hijazi, MD, MPH, MSCAI, Roberto M. Lang, MD, FASE; Jonathan J. Rome, MD, and Yin Wang, RDCS, Philadelphia, Pennsylvania; Portland, Oregon; Thane, India; Orlando, Florida; Doha, Qatar; and Chicago, Illinois

(J Am Soc Echocardiogr 2015;28:910-68)



Secundum: Percutaneous

- Size
- Location
- Adequate Rims: >5mm

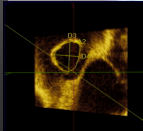
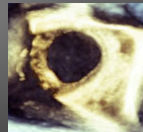
Size



- Amplatzer Septal Occluder
- up to 38mm diameter

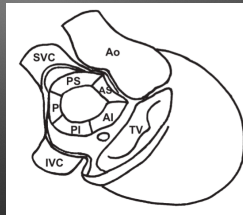


- Helex Occluder
- up to 18mm diameter

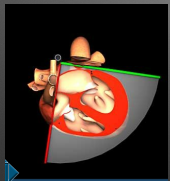


Rims

- At least 75% Rim
- > 5mm lip in all views

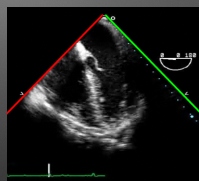


Rims and Views



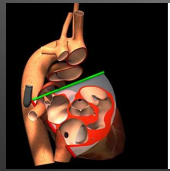
Mid-esophageal 4 chamber
0°

<https://pie.med.utoronto.ca/TEE/>

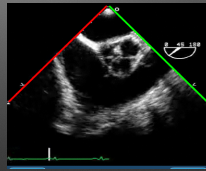


Anterior Inferior +
Posterior

Rims and View

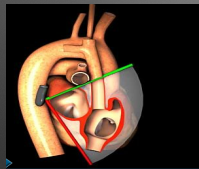


Mid- esophageal SAX 45°

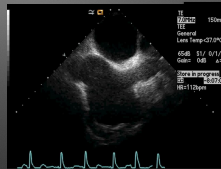


Superior Anterior +
Posterior

Rims and Views

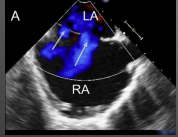


Bicaaval 90°



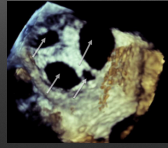
Posterior Superior +
Posterior Inferior Rims

Fenestrations



TEE with color flow Doppler:
Swiss Cheese

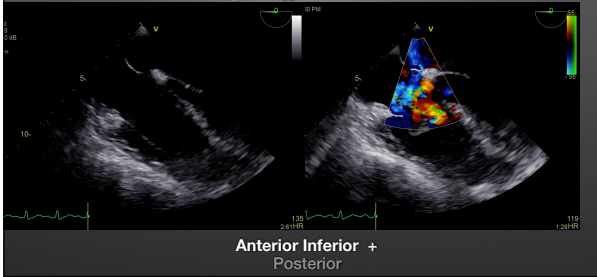
3D: Swiss Cheese



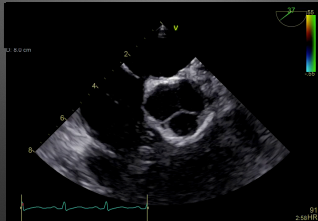
Case

- 36 y.o. woman with increased dyspnea on exertion noted to have an enlarged heart on Echo
- TEE performed for assessment of ASD prior to percutaneous closure

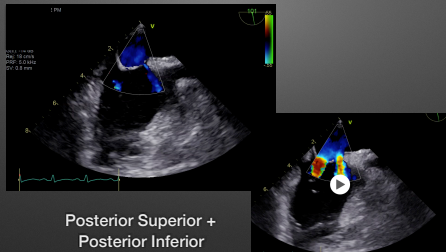
Mid-Esophageal 4 chamber



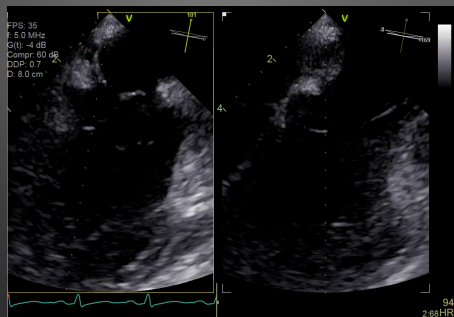
Mid- esophageal SAX 45°

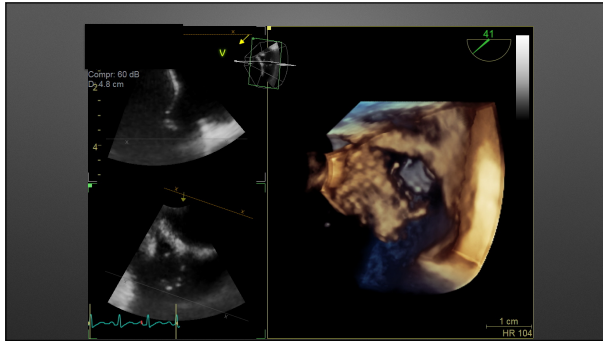


Bicaval 90°



Posterior Superior +
Posterior Inferior





Diagnosis and Plan

- 36 y.o. woman with increased dyspnea on exertion noted to have an enlarged Echo

TEE: Fenestrated ASD with an atrial septal aneurysm

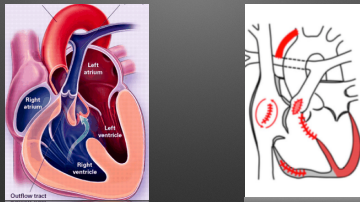


Surgical Closure

ARS Question

- ASD

Tetralogy of Fallot



Complications

RIGHT -SIDE	LEFT-SIDED
Significant PI	LV dysfunction
Right heart dilation with dysfunction	Aortic aneurysm
RV hypertension secondary to pulmonary artery stenosis	Left side arrhythmia
Right-side arrhythmias	

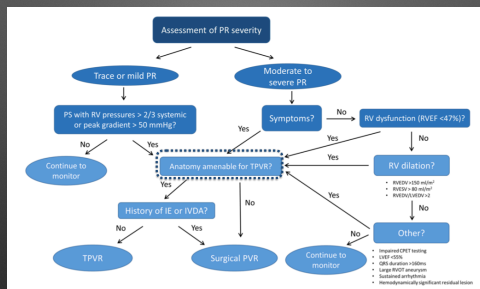
Sudden Cardiac Death of 6%

GUIDELINES AND STANDARDS

Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

Anna Maria Valente, MD, FASE, Co-Chair, Stephen Cook, MD, Douglas Stone, MD, H. Hiram Kim, BSc, BDMSc, BDCS, FASE, Rachel Kulkarni, MD, Andrew H. Taylor, MD, Carlos A. Wesson, MD, Jacqueline Krueger, MD, and Tal Geva, MD, FASE, Co-Chair, Boston, Massachusetts; Pittsburgh, Pennsylvania; Mainz, Italy; New York, New York; Houston, Texas, London, United Kingdom; Rochester, Minnesota

J Am Soc Echocardiogr 2014;27:111-413

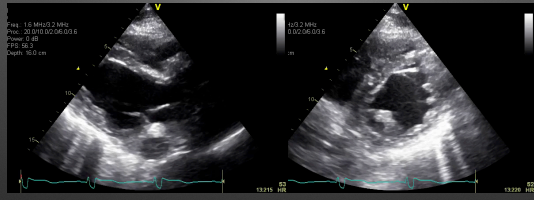


Kim Y. Heart 2016;102:1520

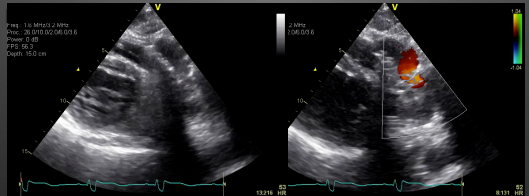
Case

- 32 y.o. male with worsening symptoms of exercise intolerance
- PE:BP 110/70 with HR of 77 bpm; height 70in; weight 200 lbs; BSA 2.1 m²
- Right thoracotomy scar and mid sternal scar
- 3/6 systolic EM at RUSB with a 1/6 early peaking diastolic murmur

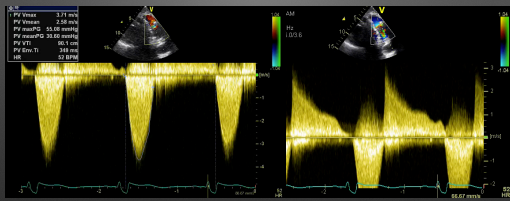
Tetralogy of Fallot



RVOT View- Conduit

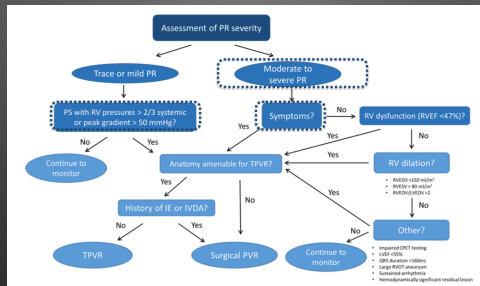


CW Doppler



3.7 m/sec; mean 31 mmHg;
peak 55mmHg

Moderate-severe PI
peak 55mmHg



Kim Y. Heart 2016;102:1520

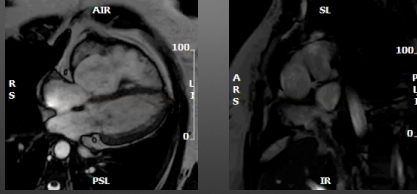
Case

- Repaired Tetralogy of Fallot
- Initial Right subclavian artery to Pulmonary artery shunt
- Subsequent full repair - takedown of shunt, closure of VSD, RVOT tranannular patch reconstruction and pulmonary valvulotomy
- Most recently about 7 years ago underwent a RV to PA conduit
- Management?

Cardiac MRI

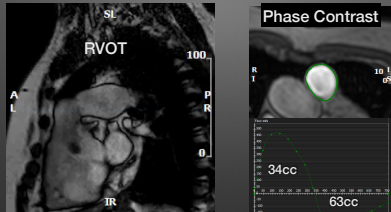
- Gold standard
- Left and Right ventricular volume and ejection fraction
- MRA
 - conduit
 - pulmonary artery
 - aorta

Cardiac MRI

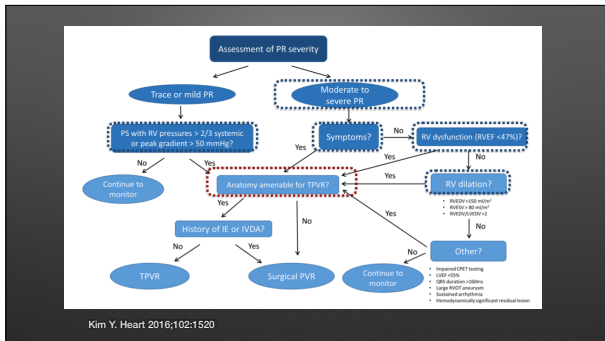
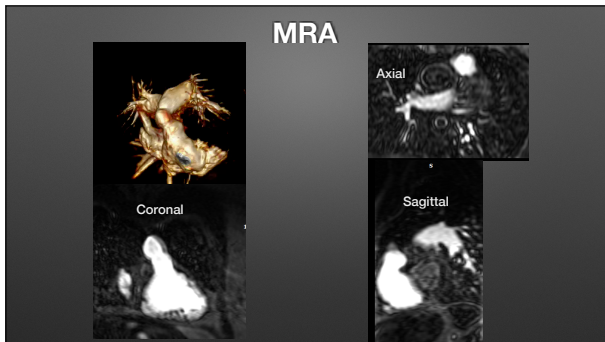


- Ventricular function
- LVEDV 54cc/m2 LVESV 24cc/m2; EF 56%
- RVEDV 121cc/m2 RVESV 68cc/m2 EF 44%

Cardiac MRI



Peak velocity 3.2m/sec



Percutaneous

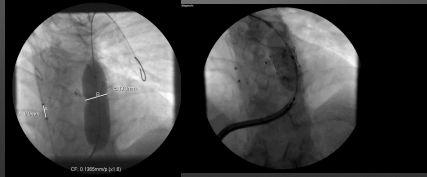


VALVE	TYPE	APPROVED USE	EXPANDABLE DIAMETER
MELODY	Bovine jugular venous valve in covered stent	RVOT conduits >16mm	20,22
SAPIEN	Bovine pericardial valve on stainless steel stent	Conduit >21mm	23,26
SAPIEN XT	Bovine pericardial valve on stainless steel stent	Aortic, mitral	20,23,26,29
NATIVE OUTFLOW DEVICE	Porcine pericardial valve on nitinol stent	Investigational	
VENUS-P	Porcine pericardial valve in covered self expanding stent	Investigational	

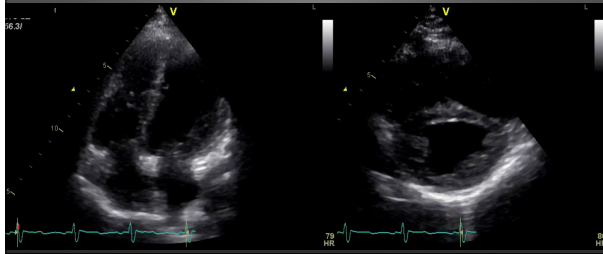
Diagnosis and Plan

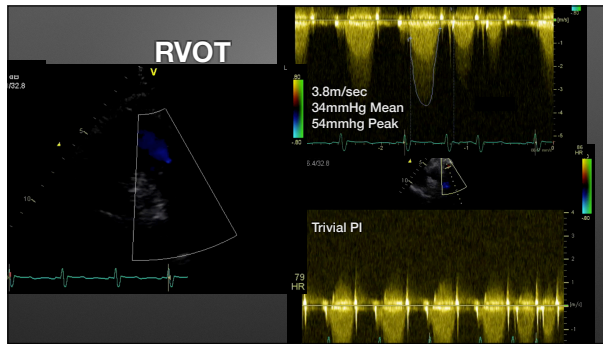
- Tetralogy of Fallot with RV to PA conduit dysfunction
- Peak systolic conduit gradient 55mmhg
- Moderate to severe PI
- No pulmonary artery stenosis
- TPVR

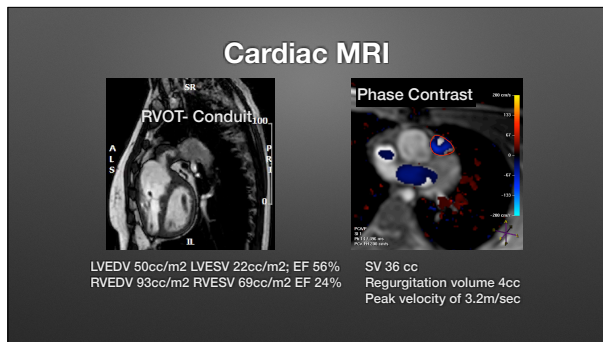
Melody Valve



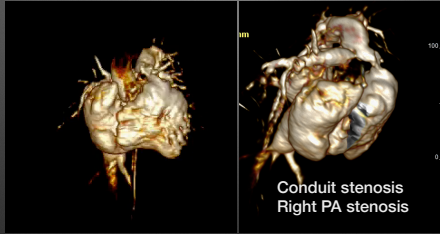
27 y.o. female with history of repaired Tetralogy of Fallot with right BT shunt (15 months) followed by complete repair (3 yo) and subsequent RV to PA conduit (18 yo)

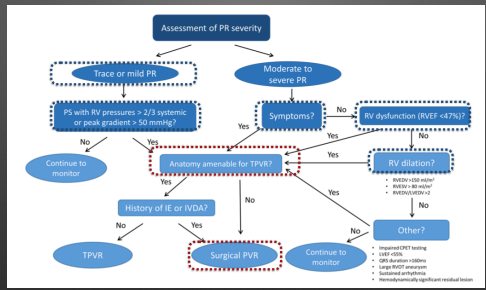






MRA



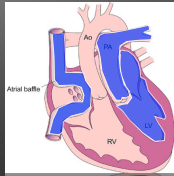
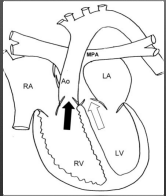


Kim Y. Heart 2016;102:1520

Diagnosis and Plan

- Tetralogy of Fallot with RV to pulmonary conduit dysfunction
Surgical repair
- RVEF by MRI 24%
- Conduit- peak gradient of 55mmHg no PI
- Severe conduit narrowing
- Surgical placement of 25mm St Jude bioprosthesis and right PA augmentation

D-TGA -Atrial Switch

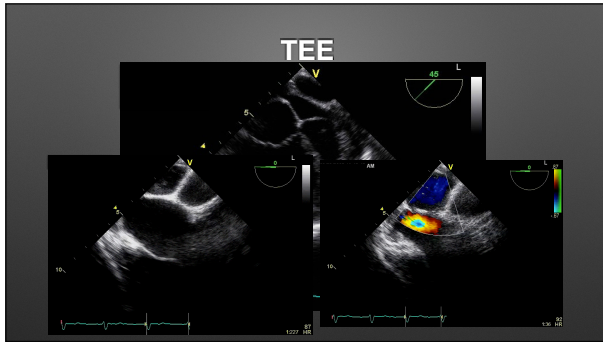


Complications and Interventions

- Systemic RV failure
- Tricuspid Regurgitation
- Baffle leaks/obstructions
- Pulmonary hypertension

Case D-TGA- Atrial switch

- 32 y.o. male
- Viral syndrome for one week; treated with antibiotics & prednisone; still with dyspnea on exertion and fatigue
- Vitals: BP 110/70; HR of 150 bpm; Pox 92%
- Cardiovascular exam: RRR with systolic murmur; JVP of 10cm; HJR; +1 LE edema

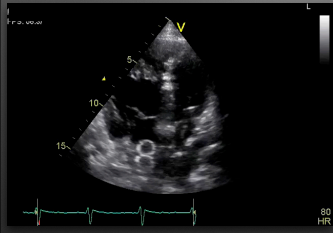


Baffle Leak

- Saturations poor sensitivity for detection of baffle leak
- Incidence of baffle leak of 50-75%
- Especially important if desaturation with exercise or cardiac lead placement
- Agitated Saline injection superior to MRI for evaluation of baffle leak

Wilhelm. Echocardiography 2016;33:437

Diagnosis and Plan

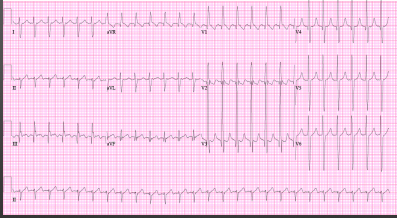


- D-TGA with atrial switch with baffle leak
- Percutaneous Amplatzer Closure

Case

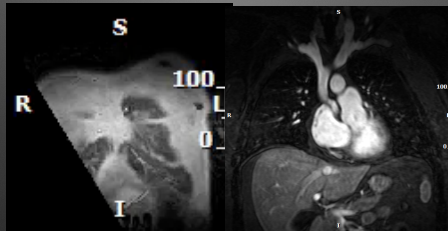
- 35 y.o. male with D-TGA with atrial switch with symptoms of palpitations associated with decreased exercise tolerance

Intra-atrial Tachycardia



Has a TEE and undergoes DCCV
Develops 4-6 second sinus pauses

MRA



Thank you