

Multimodality Imaging of Septal Defects

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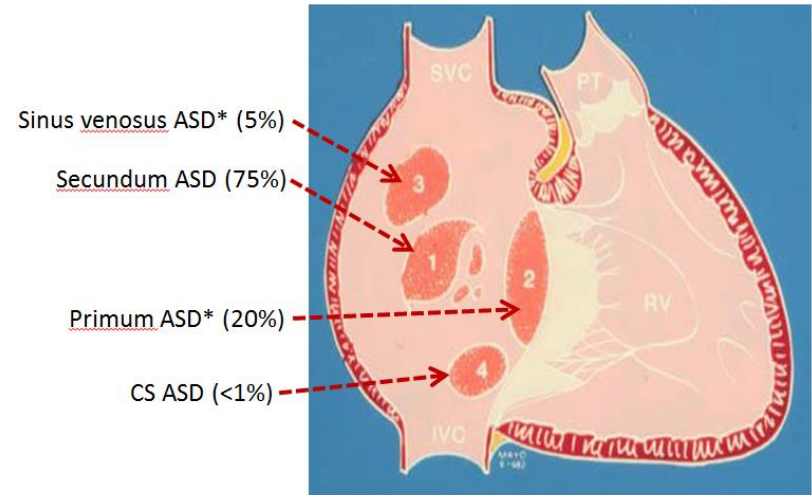


Outline (Learning Objectives)

- Describe the different types of septal defects
 - Isolated ASD and VSD
 - Septal defects in complex congenital heart defects
- Review of available modalities to image septal defects
 - Echocardiogram, CT and MRI
- Determine when to use each modality
 - Example of cases to show added value of different modalities

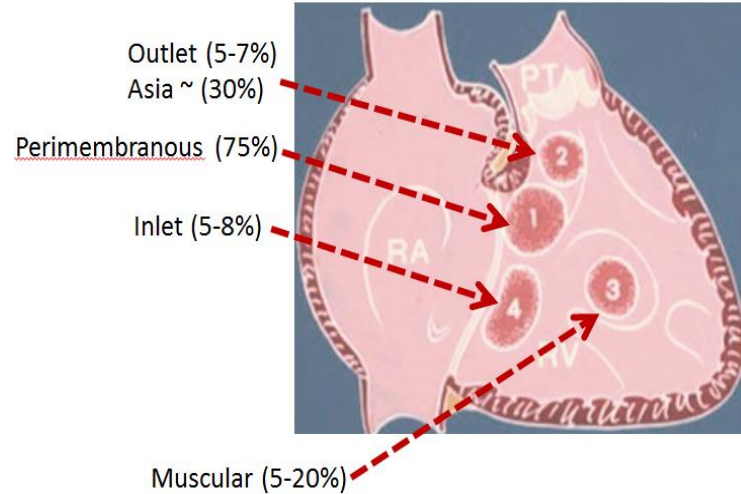
Background: Septal Defects

- Atrial Septal Defects (ASD)
 - 7-10% of all congenital defects
 - 100/100,000 live births
- Frequently associated with complex CHD
- ASD shunt crucial in:
 - **Hypoplastic left heart syndrome**
 - D-transposition of the great arteries
 - Tricuspid atresia
 - Total anomalous pulmonary venous return



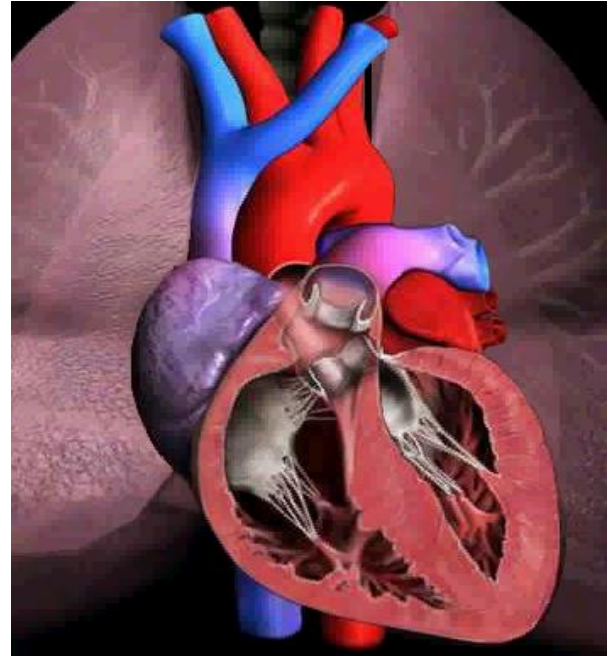
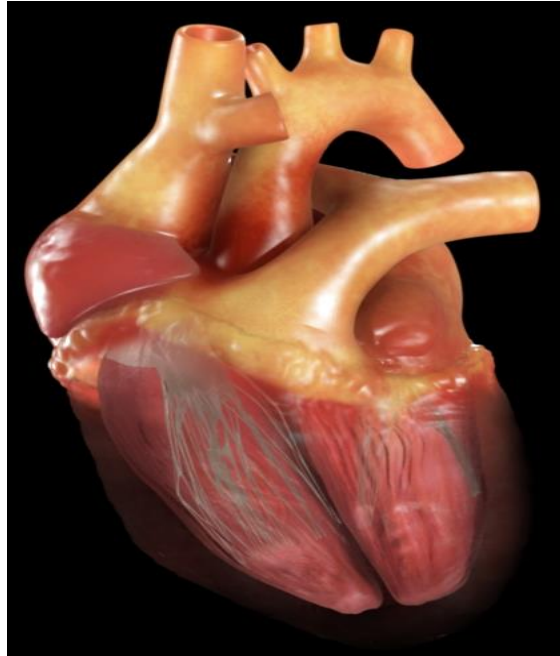
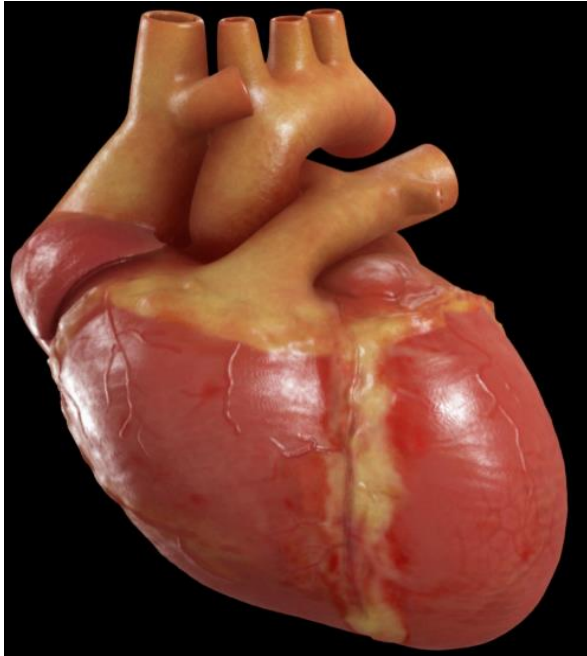
Background: Septal Defects

- Ventricular Septal Defects (VSD)
 - 20% of all congenital defects
 - 5-50/1,000 live births
- Frequently associated with complex CHD
- Most common lesion in chromosomal anomalies
- >95% of VSDs do not have associated chromosomal anomaly



Background: Imaging Modalities

- Images of the heart will be obtained to evaluate structure and function

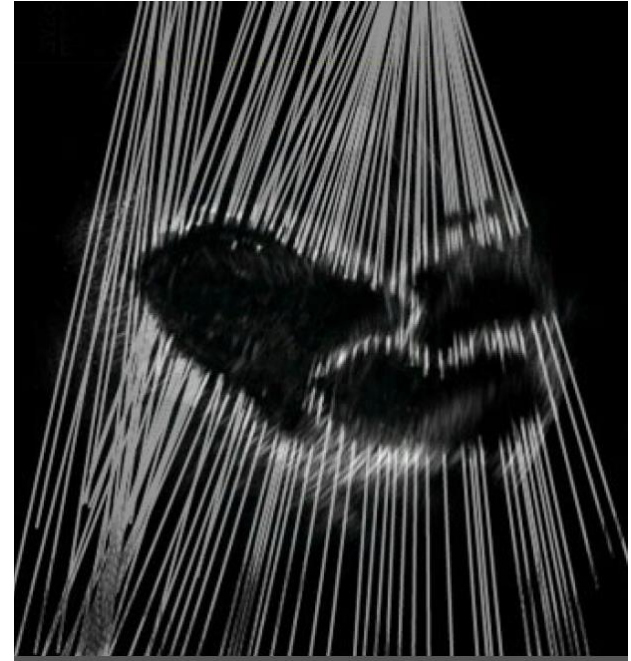


Background: Imaging Modalities

- Echocardiography
- Cardiac Magnetic Resonance Imaging
- Cardiac CT Angiography

Background: Imaging Modalities

- Echocardiography
 - Transthoracic (TTE)
 - Transesophageal (TEE)
 - 3-Dimension (3D TTE or TEE)
- Workhorse of most imaging lab



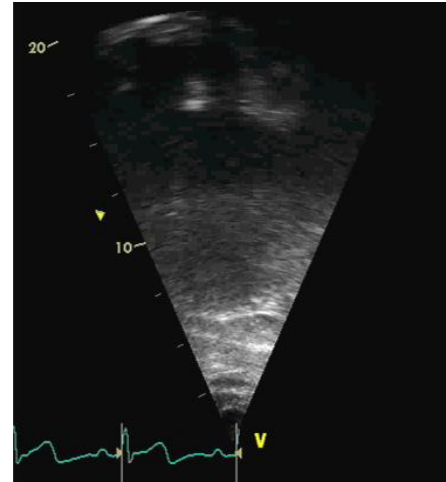
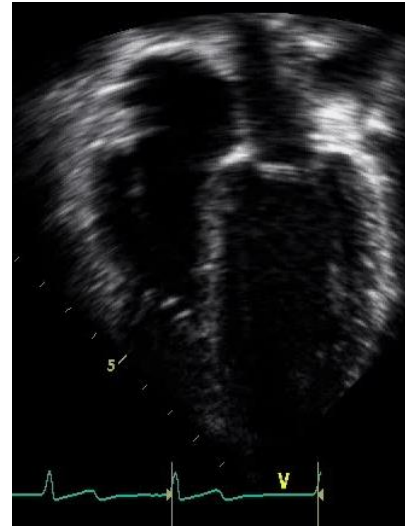
Background: Echocardiography

- Advantages

- Readily available
- Can be done quickly
- Great first line tool
- Doppler assessment*

- Disadvantages

- Difficult windows
- Extra cardiac vessels can be difficult to visualize
- Not able to measure flow or saturation accurately



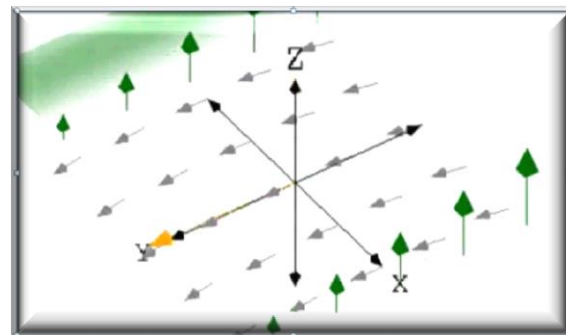
Background: Cardiac MRI

■ Advantages

- Accurate measurements
 - Anatomy, function, chamber, flow
 - Tissue characterization and other newer techniques

■ Disadvantages

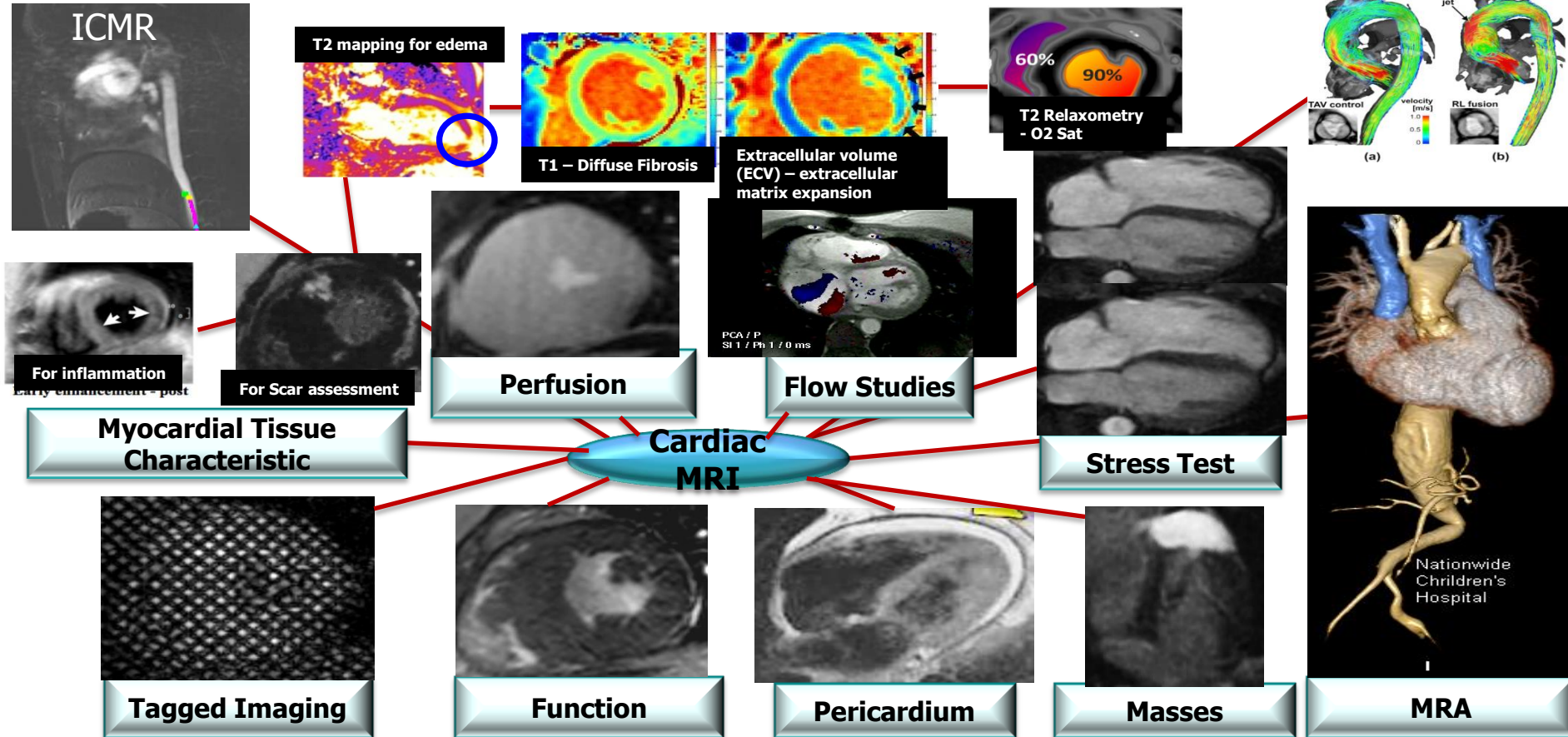
- IV placement for scar and angiography
- May take longer study - improving
- Have to lay still
- Not as readily available-changing



Background: Cardiac MRI

- CMR - is not just one test, it is many tests rolled into one modality (one stop shop)
- Similar to CT angiography but without radiation exposure
- Similar to echocardiogram but without acoustic window limitations
- Cardiac MRI is a one stop shop

Background: Cardiac MRI



Background: CMR vs CTA

- Cardiac MRI – comprehensive diagnostic test
- But there are times when we should consider a CT instead:
 - Risk of heating up
 - Risk of movement
 - Interruption of device function
 - Artifacts from device, stents or coils
 - Sedation issues/risk

Background: CTA

- When to use CT Angiography over Cardiac MRI?
 - The need for speed
 - CTA takes 0.35 to 0.5 seconds
 - MRA 15-20 seconds per dynamic
 - CT has better spatial resolution (0.5 mm)
 - Avoid or minimize sedation
 - **Basis:** in some patients initiation of anesthesia adds significant risk (Williams Syndrome, pulmonary hypertension, unstable patients)

Cases Scenarios and Best Modality?

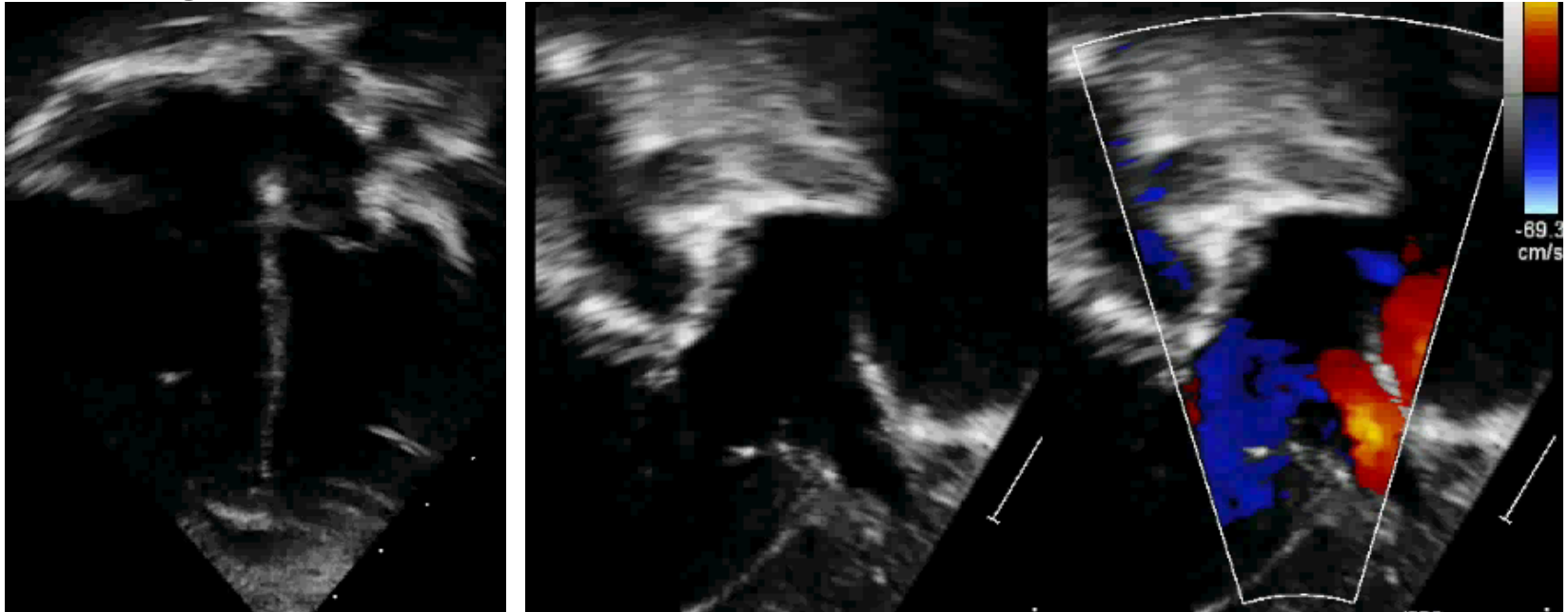
- Isolated secundum ASD in pediatrics
- Dilated right heart in older patients (? ASD)
- Other ASDs (superior sinus venosus ASD)
- ASD associated with complex CHD (non-heterotaxy)
 - Complete Atrioventricular canal
 - Hypoplastic left heart syndrome
- Isolated perimembranous VSD in pediatrics VSD frequently associated with complex CHD
- Unusually position large muscular VSD
- TOF/PA VSD with multiple aortopulmonary collateral vessels (MAPCAs)
- Double outlet right ventricle (DORV)
- A case of post-operative blue.....

Best Modality?

- 3 year male with isolated large secundum ASD with right sided enlargement
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed without sedation
 - Define ASD size, rims and chamber sizes
 - **CT Angiography**: limited to no value in this setting even with low dose new generation multi-detector scanners
 - **Cardiac MRI** – limited use in this situation at this time
 - Issues including need for sedation (may change with faster sequences and bundling of patient)
 - Additional value of Qp/Qs does not alter management

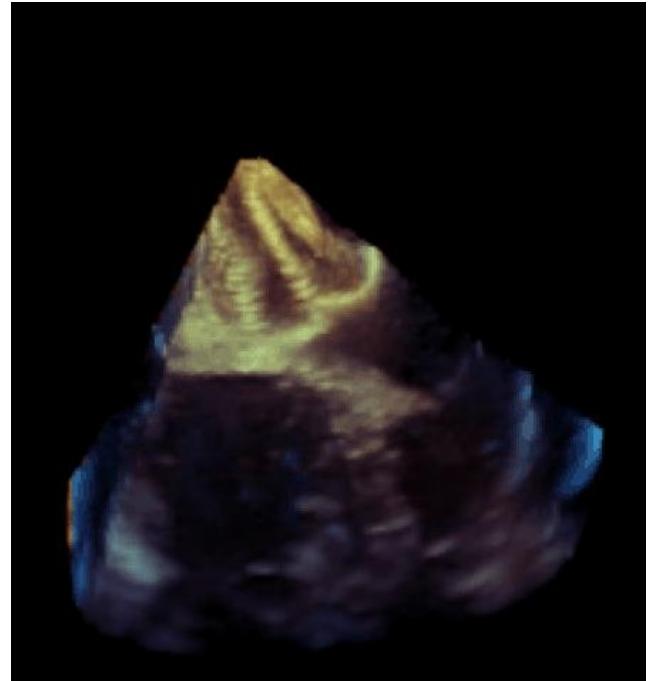
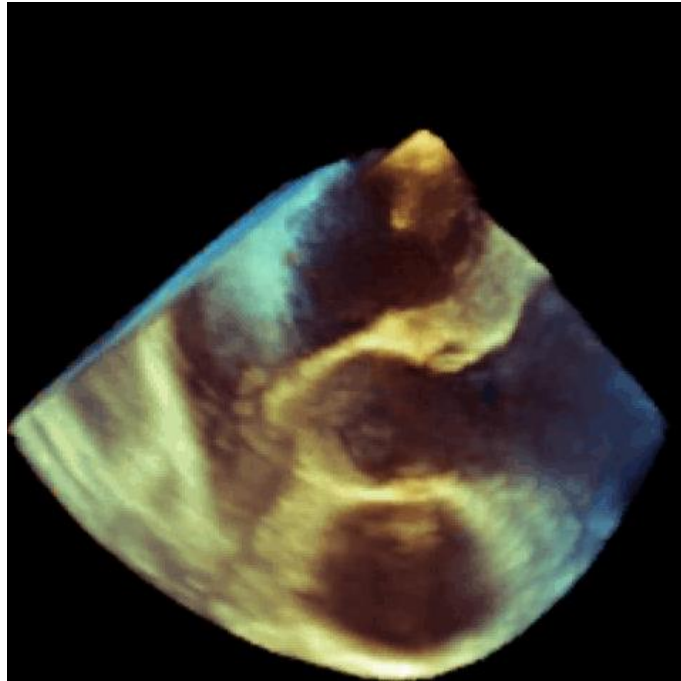
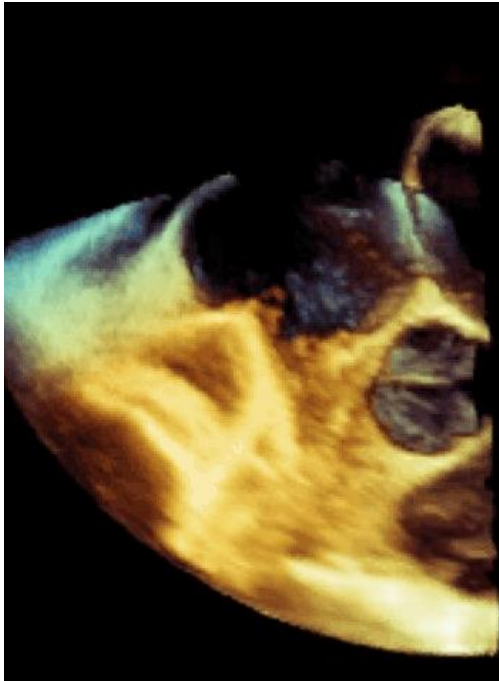
ECHO Best Modality: Isolated ASD

- 3 year male with murmur and fix split S2.
- Echo below with isolated large secundum ASD with right sided enlargement



ECHO Best Modality: Isolated ASD

- 3D Echo – not vital but can help visualize septum in 3D space

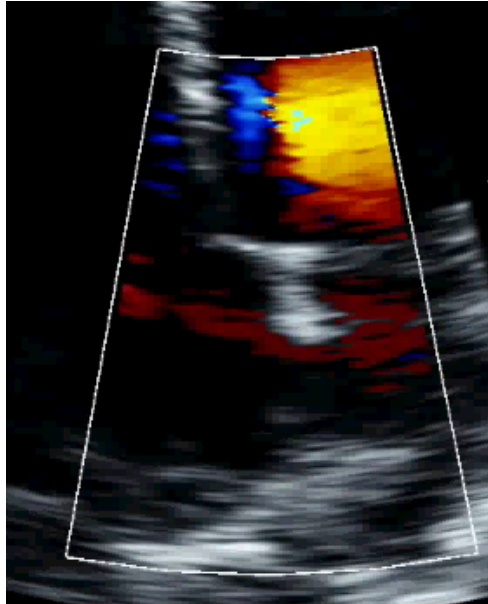


Best Modality?

- 50 y/o male with murmur since childhood and RVH on ECG.
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk but in larger patients the acoustic windows is limited
 - Can we define ASD size, rims and chamber sizes?
 - Identify other causes of right sided chamber enlargement?
 - **CT Angiography**: limited to no value in this setting even with low dose new generation multi-detector scanners – lack ability to show physiologic data
 - **Cardiac MRI** – Augment Echo finding with no risk if echo limited
 - Confirm right chamber enlargement
 - Additional value of Qp/Qs does can alter management
 - Additional causes of right chamber enlargement can alter management

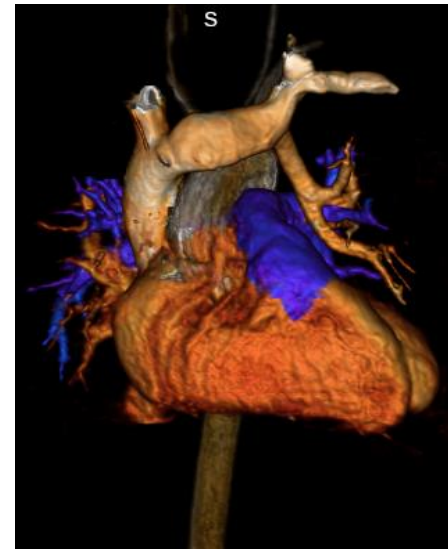
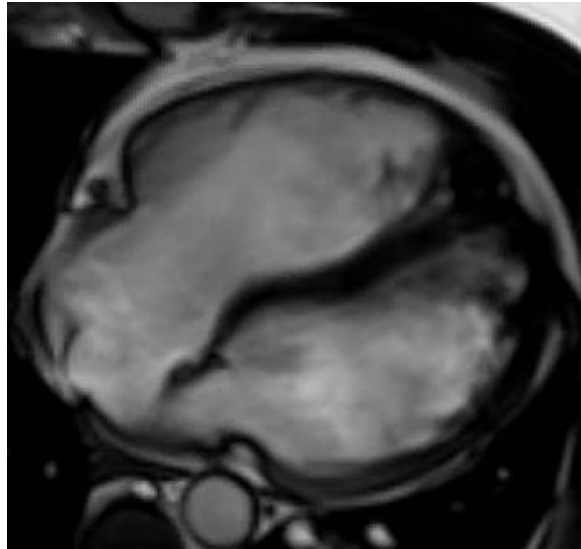
Best Modality? ECHO Enough?

- 50 y/o male murmur since childhood.
 - Initial echo: possible drop out in atrial septum with some color jet noted with right heart enlargement
 - Possible small ASD – poor imaging windows



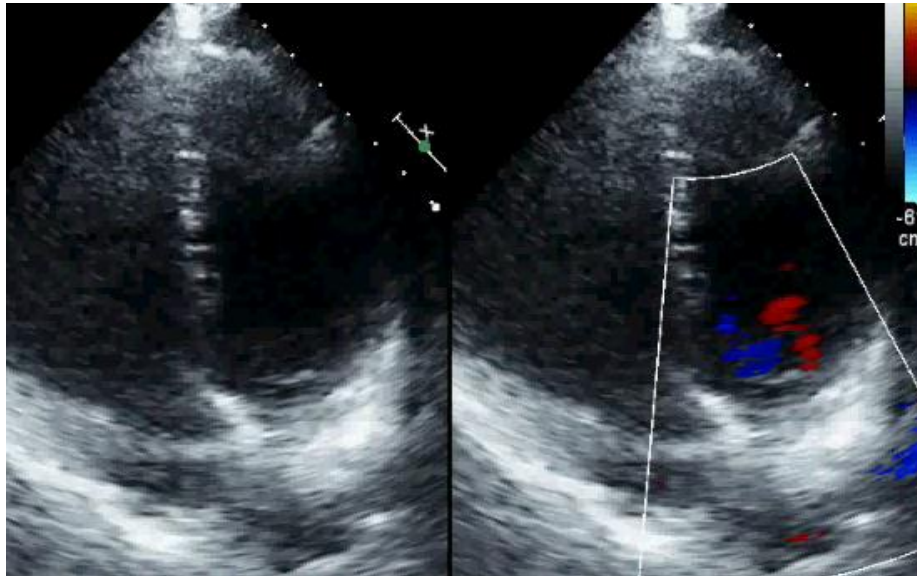
ECHO + CMR Best Modality

- 50 y/o male murmur since childhood.
 - CMR: confirmed small ASD but found partial anomalous venous return of the LUPV and lingula with RVEDVi 118 ml/m² and Qp/Qs of 1.7 to 1 (mostly from PAPVR)
 - Surgical repair of PAPVR only
 - ASD left open (very small shunt)
 - CMR better delineate disease in this setting altered management



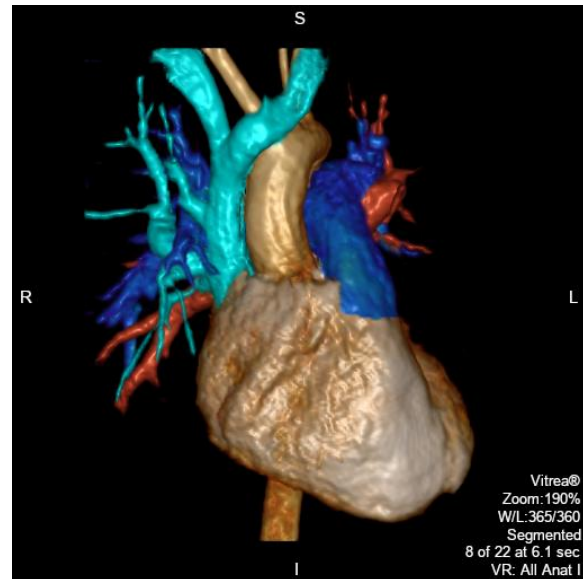
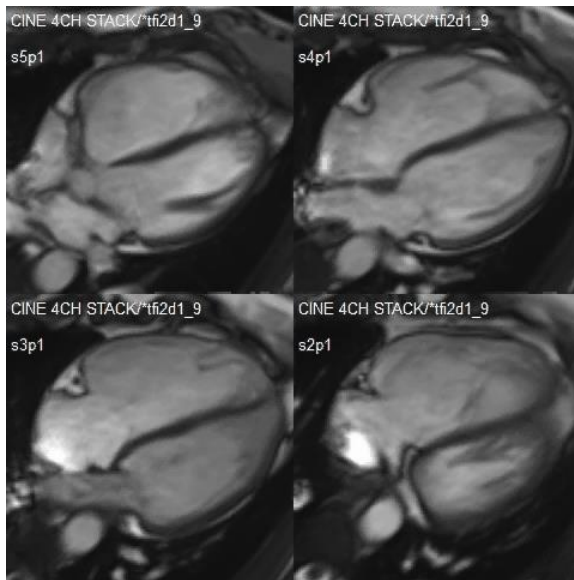
Best Modality? Superior Sinus Venosus ASD?

- 37 y/o with SOB and fatigue with history of murmur.
 - Initial echo: drop out in superior atrial septum with some color jet noted with right heart enlargement.



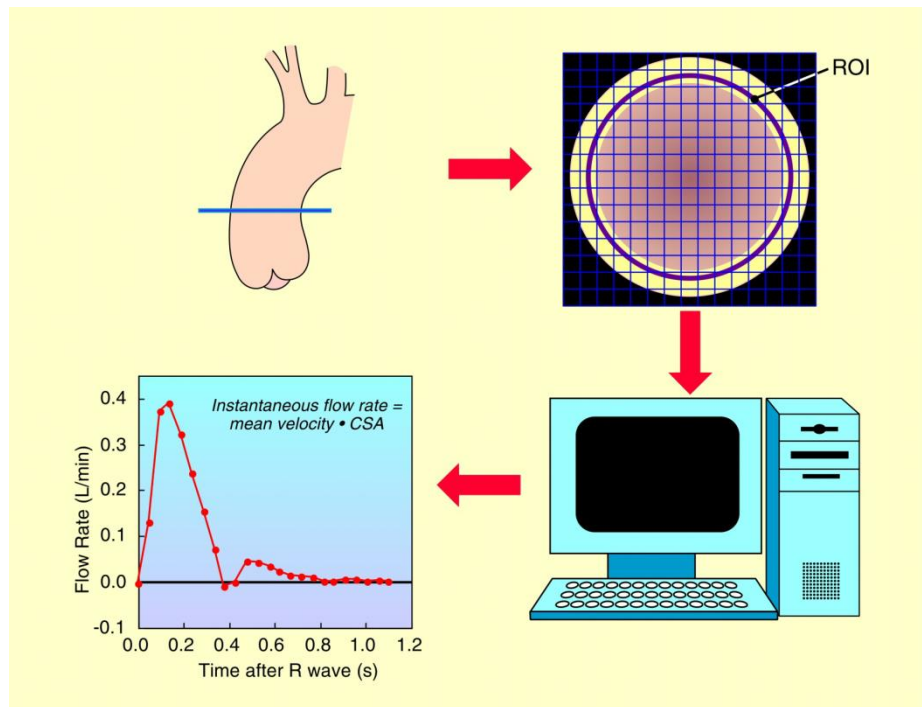
CMR Best Modality: Beyond isolated ASD

- 37 y/o with SOB and fatigue with history of murmur.
 - CMR: confirmed superior sinus ASD with RUPV, RMPV and likely part of RLPV with RVEDVi 129 ml/m² and Qp/Qs of 1.8 to 1
 - Surgical repair ASD and PAPVR
 - CMR better delineate disease in this setting
 - CT adds no additional value with risk of radiation exposure



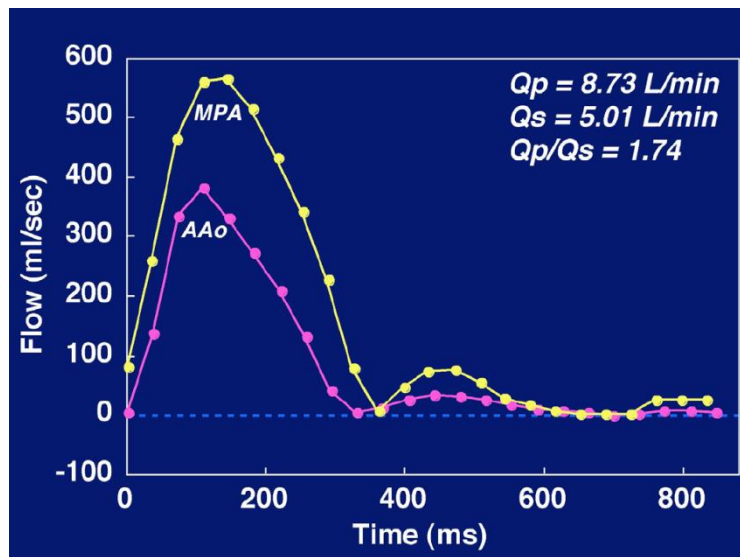
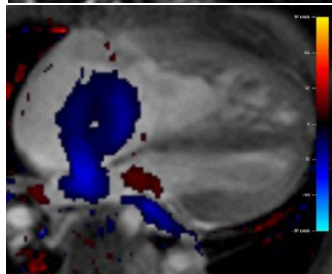
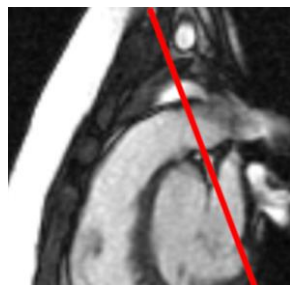
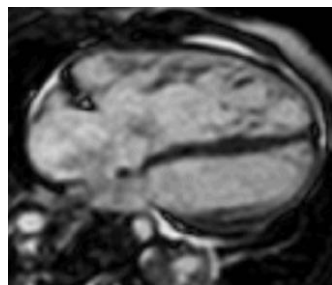
The Added Value of Cardiac MR

- Velocity Mapping for Flow Quantification
 - In setting of septal defect use to assess Qp/Qs
 - Augments anatomic, functional and volumetric data



The Added Value of Cardiac MR

- Quantification of volume and function
- Velocity Mapping for Flow Quantification: Q_p/Q_s
 - In setting of septal defect (example of ASD)
 - MPA flow (Q_p) and Aortic flow (Q_s)



Best Modality?

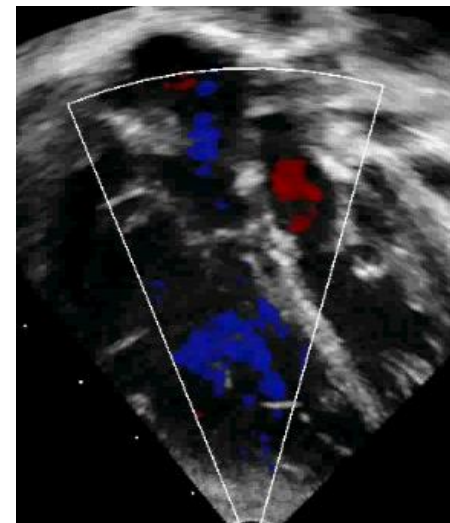
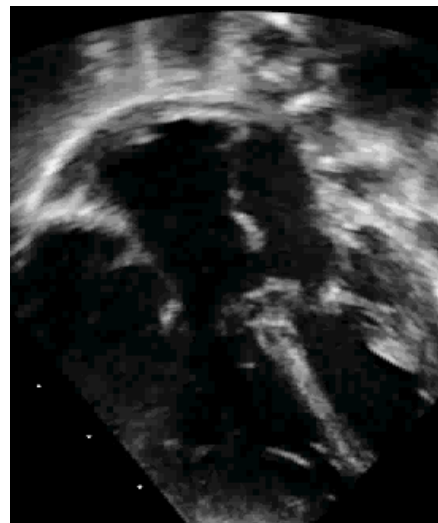
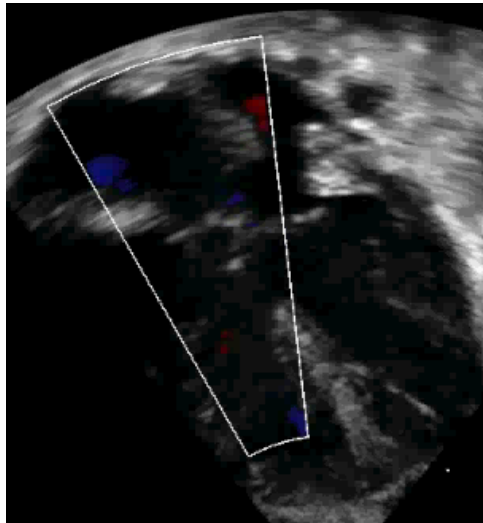
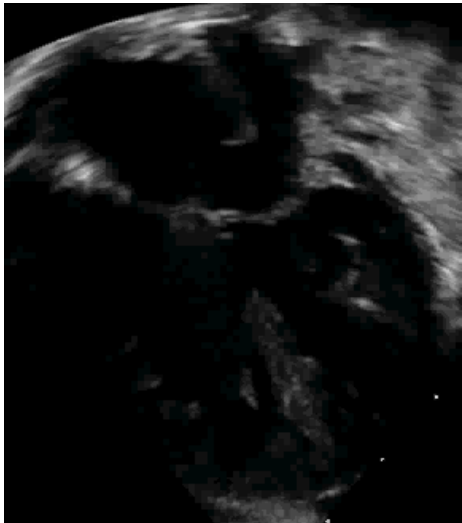
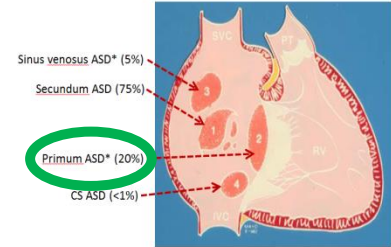
- ASD frequently associated with complex CHD (non-heterotaxy)
 - **Complete atrioventricular canal defect**
- ASD shunt crucial in:
 - **Hypoplastic left heart syndrome (obligate left to right shunt)**
 - D-transposition of the great arteries (mixing)
 - Tricuspid atresia (obligate right to left shunt)
 - Total anomalous pulmonary venous return (obligate right to left shunt)
- Atrial shunt must be non-restrictive

Best Modality? ASD in Complex CHD

- 1 week day old with Trisomy 21 with CAVC
- 2 day old with hypoplastic left heart syndrome
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed without sedation
 - Define intra- and extracardiac anatomy well in most instances
 - **CT Angiography**: limited to no value in this setting even with low dose new generation multi-detector scanners
 - **Cardiac MRI** – limited use in this situation at this time
 - Issues including need for sedation with complex long scan time
 - Additional value of Qp/Qs does not alter management
 - Exception: associated with heterotaxy syndrome

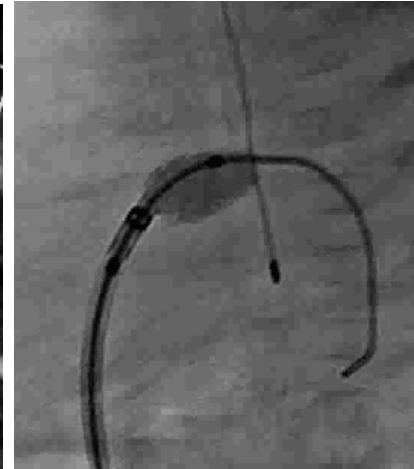
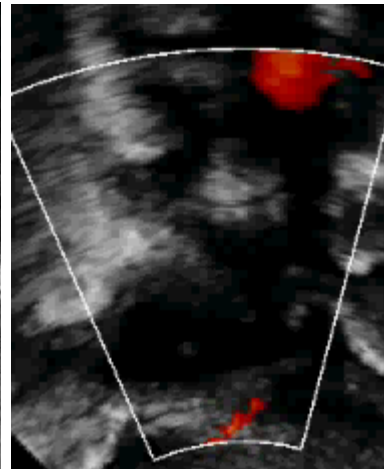
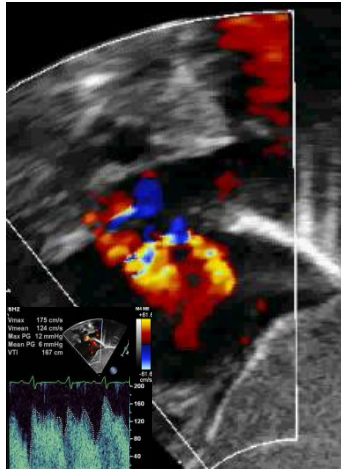
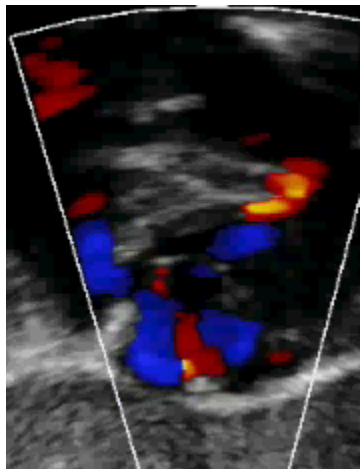
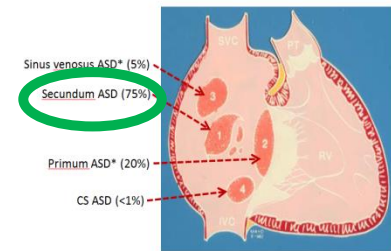
ECHO is Best Modality: ASD with CAVC

- ASD in setting of atrioventricular canal defect (AVC)
 - Spectrum of AV canal defect from complete AVC or isolated primum ASD with or without cleft MV
 - Echo is the best modality (no benefit of CT or CMR in my cases)



ECHO is Best Modality: ASD in HLHS

- ASD shunt crucial in:
 - Hypoplastic left heart syndrome (obligate left to right shunt)
 - Atrial restriction requires emergent balloon septostomy and/or stent
 - Atrial restriction associated with high mortality
 - Echo is the best modality (no benefit of CT or CMR in many cases, exception would be complex CHD with heterotaxy)

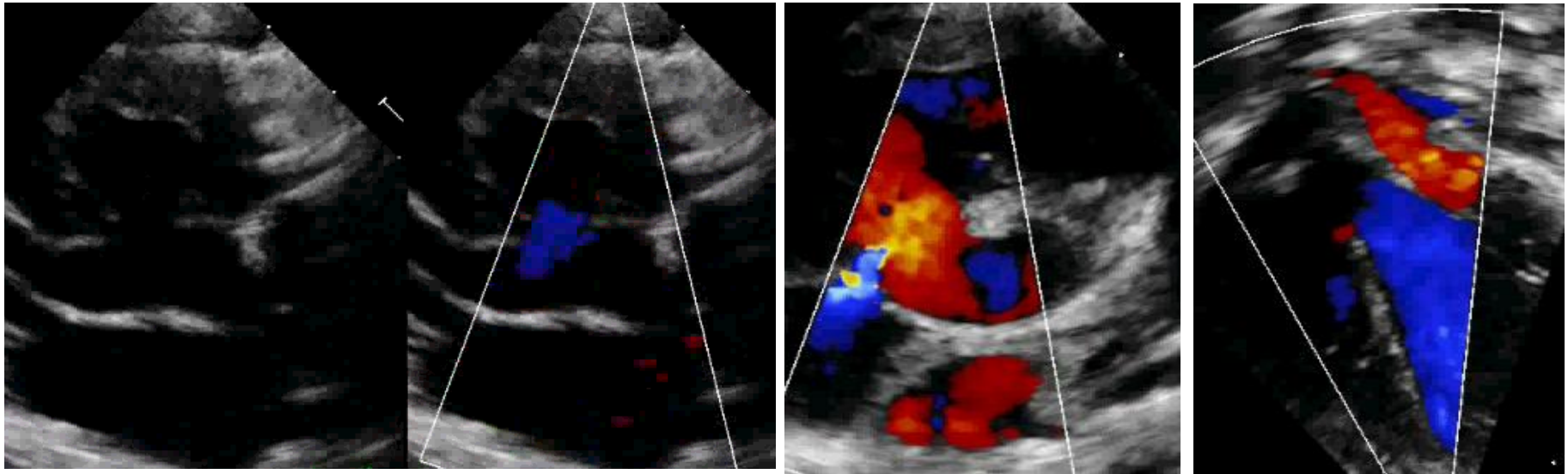


Best Modality?

- 3 month old with **isolated** large ventricular septal defect with increase work of breathing
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed with or without sedation
 - Define intra- and extracardiac anatomy well in most instances
 - **CT Angiography**: Need outweighs benefit – CT for 3D modeling and printing can be performed without sedation and low dose (1 msv)
 - **Cardiac MRI** – limited use in this situation at this time

ECHO Best Modality: Isolated VSD

- 3 month old with large perimembranous VSD with inlet extension
 - Repaired at 6 months with no complications
 - No added value of CT or Cardiac MRI (risk outweighs benefit)

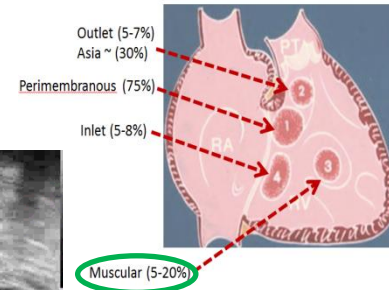
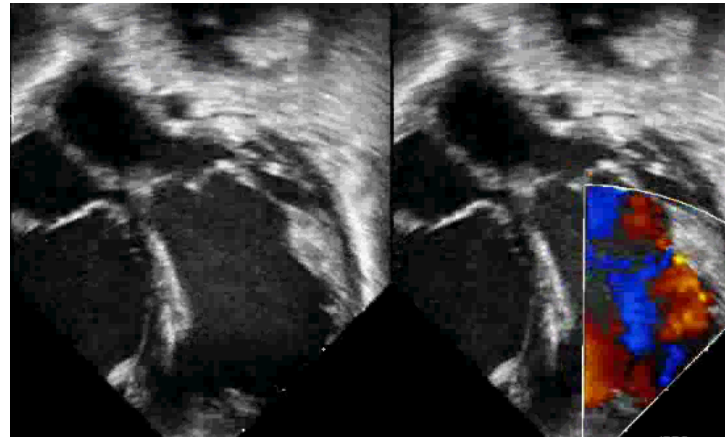
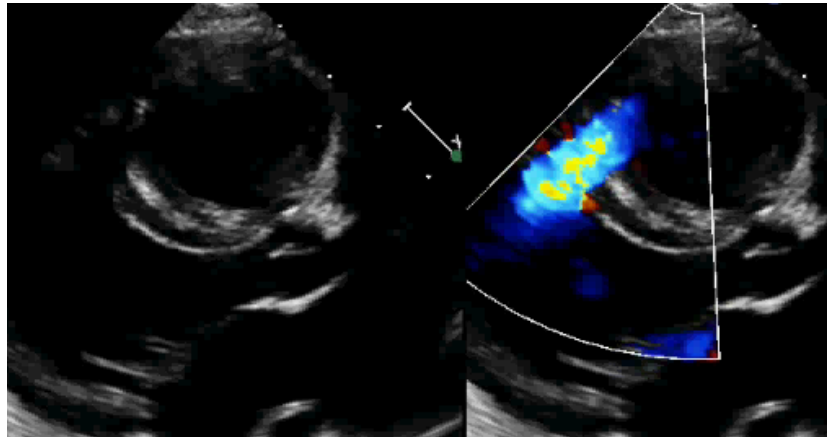


Best Modality?

- 3 month old with large **unusually position** apical muscular VSD (felt not to be a straight forward surgical candidate)
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed without sedation
 - Define intra- and extracardiac anatomy well but challenging for 3D modeling and road mapping
 - **CT Angiography**: Need outweighs benefit – CT for 3D modeling and printing can be performed without sedation and low dose (1 msv)
 - **Cardiac MRI** – limited use in this situation at this time
 - Compared to CT (need for sedation and low spatial resolution) results in risks outweighing benefit

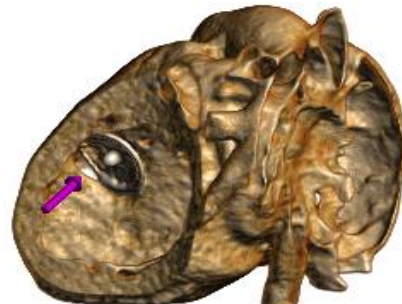
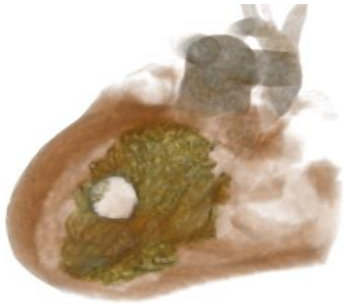
ECHO Best Modality?

- 3 month old with isolated large unusually position apical muscular VSD
 - Felt not to be a straight forward surgical candidate
 - Rare occasion CT can complement echo for 3D printing and modeling



ECHO + CTA Best Modality

- 3 month old with large unusually position apical muscular VSD (felt not to be a straight forward surgical candidate)
- **Added value of CTA:** 3D dataset for virtual assessment, 3D printing for device selection, post-device assessment on model and implantation
- Trans-atrial approach by modeling with prediction of small residual defect

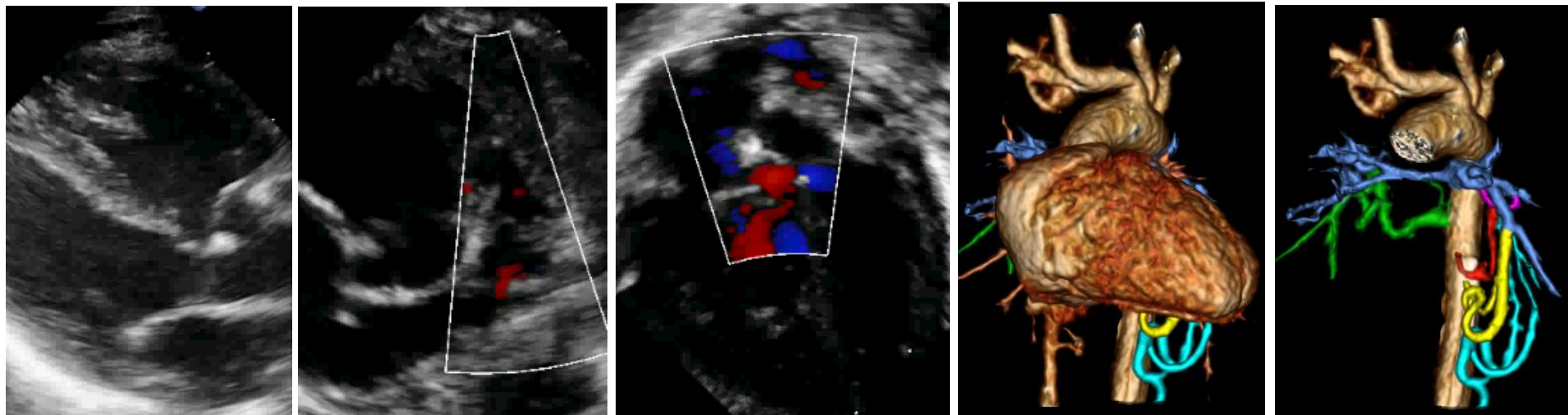


Best Modality? VSD in Complex CHD

- VSD frequently associated with complex CHD
 - Pulmonary atresia VSD with multiple aortopulmonary collateral vessels (MAPCAs)
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed without sedation
 - Challenging to define extracardiac anatomy in MAPCAs
 - **CT Angiography**: Need outweighs benefit – CT for to define collaterals and can potentially delay cardiac catheterization can be performed without sedation and low dose (1 mSv)
 - **Cardiac MRI** – limited use in this situation at this time
 - Compared to CT (need for sedation and low spatial resolution) results in risks outweighing benefit

ECHO + CTA = Best Modality

- 1 day with Tetralogy of Fallot with near pulmonary atresia with MAPCAs
 - Echocardiogram: Define intracardiac anatomy very well
 - Non-sedated CT Angiography augmented Echo with road-mapping of the MAPCA
 - Allows cardiac catheterization to be delayed until closer to surgical date



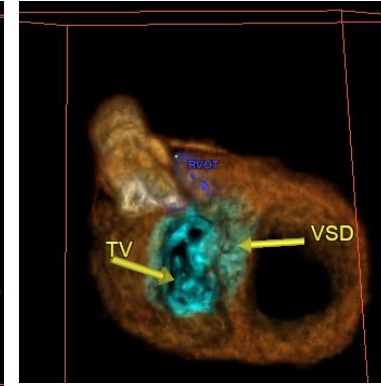
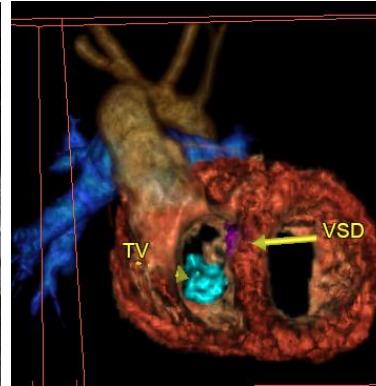
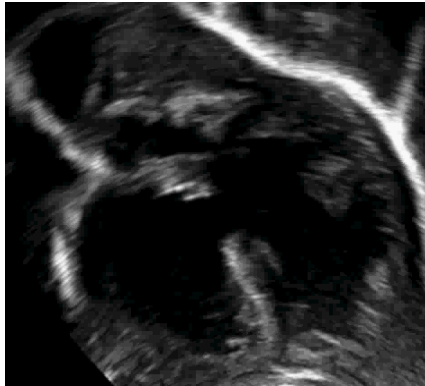
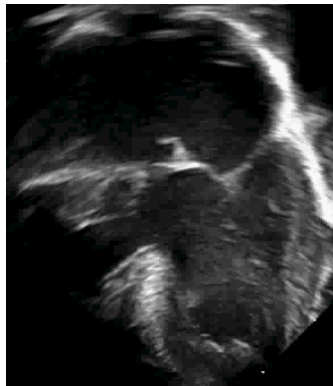
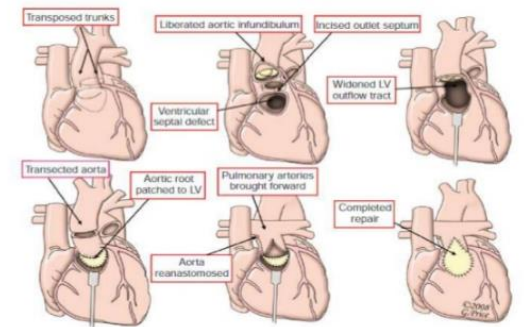
Best Modality? VSD in Complex CHD

- VSD frequently associated with complex CHD
 - Double outlet right ventricle (defining relationship of VSD to aorta)
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk and can be performed without sedation
 - Challenging to define relationship of VSD to Aorta in some cases
 - **CT Angiography**: Need outweighs benefit – CT for 3D modeling and printing can be performed without sedation and low dose (1 msev)
 - **Cardiac MRI** – limited use in this situation at this time
 - Compared to CT (need for sedation and low spatial resolution) results in risks outweighing benefit

ECHO + CTA = Best Modality

- Double outlet right ventricle inlet VSD with concern of AV chords affecting VSD to Aortic baffle
 - Echocardiogram: Define intracardiac anatomy well but challenging to roadmap VSD to Aorta
 - Non-sedated CT with low dose radiation performed for virtual and 3D modeling to determine that VSD can be baffle to the aorta
 - Underwent biventricular repair with a Nikaidoh procedure with reconstruction of his right ventricular outflow tract (16 mm Contegra valved conduit)

Nikaidoh Procedure



Added Value of CT Angiography

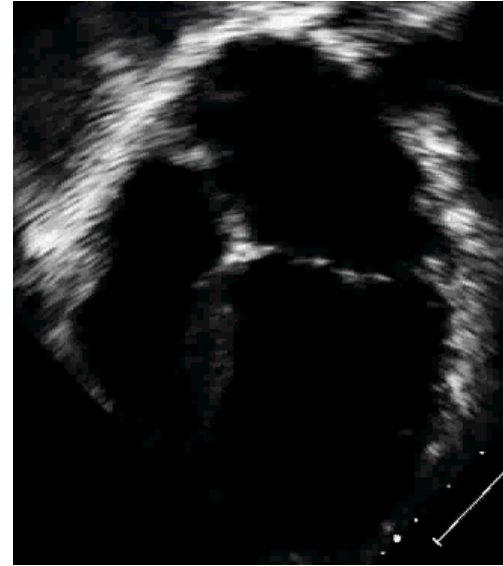
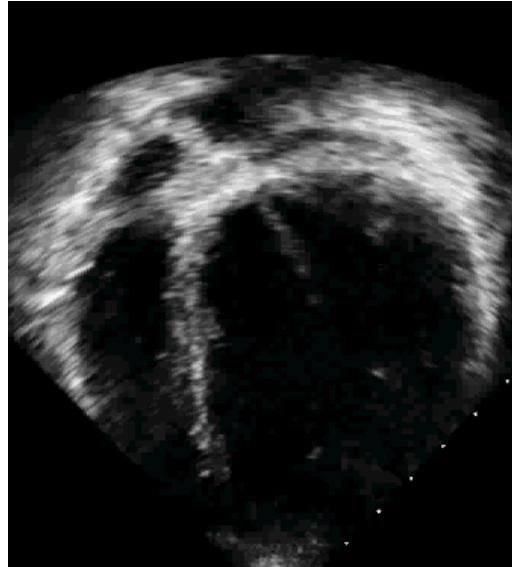
- VSD frequently associated with complex CHD
 - **Pulmonary atresia VSD with multiple aortopulmonary collateral vessels (MAPCAs)**
 - **Double outlet right ventricle (DORV)**
- Echocardiogram is still first line tool
- CMR and CT augments echocardiogram
 - Extracardiac vasculature with roadmapping of the MAPCAs.
 - CT preference over Cardiac MRI with use of low dose radiation and no sedation (anatomy over physiology)

Best Modality? Post-Operative Blues

- 27 year old with history of ASD repair with desaturation on exercise testing
- ECHO only modality needed?
- Basis
 - **Echocardiogram** has no risk should be first line
 - Challenging in adults, agitated saline study (must understand defect)
 - **CT Angiography**: limited to no value in this setting – lack ability to show physiologic data
 - **Cardiac MRI** – Augment Echo finding if echo limited or questions remain
 - Confirm any echo finding
 - Additional value of Qp/Qs does can alter management
 - Improve ability to assess challenging problems can alter management

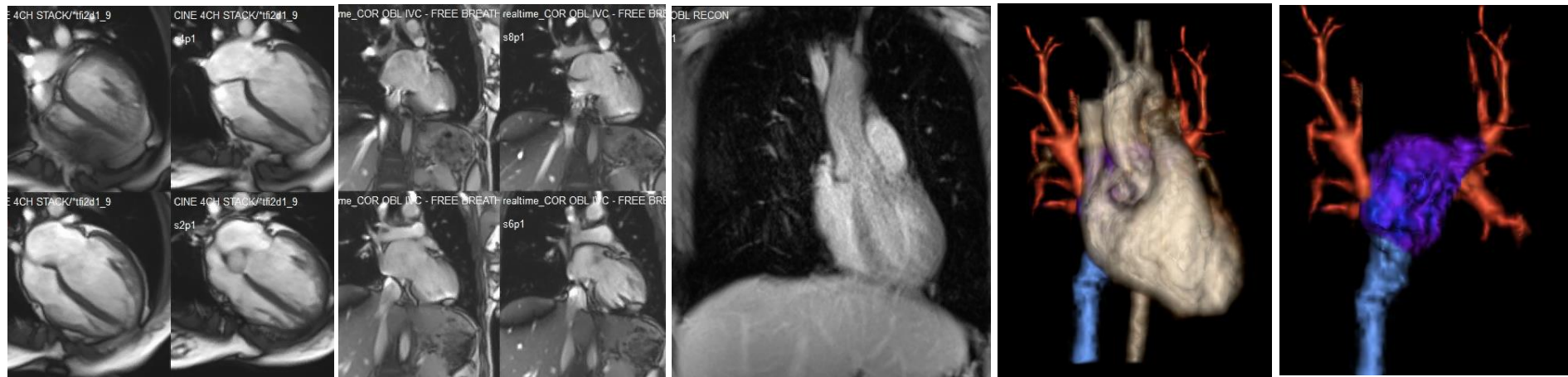
Best Modality?

- 27 year old with history of ASD repair with desaturation on exercise testing
 - ECHO: Structurally normal heart with left heart enlargement
 - Intravenous injection of agitated saline contrast with no shunt.
 - Further testing needed?



CMR = Best Modality

- 27 year old with history of ASD repair with desaturation on exercise testing
 - Cardiac MRI: Sinus venosus defect and PAPVR s/p repair with no residual ASD but inferior vena cava baffle to left atrium (pt had inferior sinus venosus ASD with PAPVR)



The Added Value of Cardiac MR

- First issue: agitated saline contrast was performed on upper extremity. Given history of inferior ASD and cyanosis a LLE IV should have been placed
- Added value of CMR: Anatomically confirmed the surgical mis-hap and demonstrated the issues needed
 - Alter management course
 - Patient underwent surgical correction
 - Patient feels great and has no desaturation with exercise

Multimodality Imaging of Septal Defects

- What is the best modality?
 - ECHO, CT and Cardiac MRI are complementary tests
- Depends on what you are looking for
 - Echo should always be first line tool
- Add CT or Cardiac MRI when Echo has limitations
 - Difficult windows
 - Extra cardiac vessels that are not well visualized
 - When your questions have not been answered

Multimodality Imaging of Septal Defects

- What is the best modality?
- Use low risk test first
 - Avoid radiation if possible
 - In children balance risk benefit of radiation against sedation
- Determine what your goal is:
 - Intracardiac anatomy: Echo is best
 - Extra-cardiac anatomy: CT is best
 - Anatomy and detail physiology: Cardiac MR is best



NATIONWIDE CHILDREN'S

When your child needs a hospital, everything matters.SM



NATIONWIDE CHILDREN'S

When your child needs a hospital, everything matters.SM



THE OHIO STATE UNIVERSITY
COLLEGE OF MEDICINE



Ohio
CHAPTER

Thank you!



Questions?

Background: CMR vs CTA

■ CT Scan

- Radiation exposure: 2-10 mSv
= 3-5 yr back ground radiation
- Cost \$1200-3000
- Time \leq seconds
- Application: vascular, bone and lungs
- Better spatial resolution
- Scan in systole HR >90, diastole HR < 90 or whole cardiac cycle

■ MRI

- None - MRI does not emit ionizing radiation
- Cost \$1200-5500
- Time = minutes to hours
- Application: vascular and soft tissue
- Better temporal resolution
- MRA is collected over many heart beats and average of systole and diastole