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
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

http://www.youtube.com/watch?feature=player_detailpage&v=7MRm5mD2YxQ
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
ICMR

T2 mapping for edema

T1 – Diffuse Fibrosis

Extracellular volume (ECV) – extracellular matrix expansion

60% 90%

T2 Relaxometry - O2 Sat

Perfusion

Flow Studies

Myocardial Tissue Characteristic

Cardiac MRI

For inflammation

Early enhancement post

For Scar assessment

Tagged Imaging

Function

Pericardium

Stress Test

Masses

MRA

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)

Han et al, JCCT, 2013
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......
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: Qp/Qs
  - In setting of septal defect (example of ASD)
  - MPA flow (Qp) and Aortic flow (Qs)

\[
\text{Qp} = 8.73 \text{ L/min} \\
\text{Qs} = 5.01 \text{ L/min} \\
\text{Qp/Qs} = 1.74
\]
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 msev)
  - **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 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 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 straightforward 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 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

- 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
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)

**ECHO + CTA = Best Modality**
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?
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)

CMR = Best Modality
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.
Thank you!

Questions?
Background: CMR vs CTA

- **CT Scan**
  - Radiation exposure: 2-10 mSv = 3-5 yr background radiation
  - Cost: $1200-3000
  - Time: ≤ 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