Total Anomalous Pulmonary Venous Connections: Anatomy and Diagnostic Imaging

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March 12, 2015
Congenital Cardiac Anesthesia Society Annual Meeting
Disclosures

• I will discuss the use of a medical device/drug (Gadolinium) that is classified by the Food and Drug Administration as investigational for the intended use.

• I do not have relationships with financial interests.
Goals

- TAPVR anatomy and subtypes
- Spectrum of presentations
- Key echocardiographic features
- Alternative diagnostic imaging options (and when to use them)
“There’s a new blue baby coming to the NICU... they say the heart was normal on the outside echo but we’ll need you to take a look”
Acutely cyanotic infant in shock

- 2 hour old neonate
- Term delivery
- APGARs 61 55
- Deeply cyanotic in the DR
- Decreased perfusion
Acutely cyanotic infant with shock

- “Normal” prenatal care
- Outside hospital placed lines and performed a “quick echo” which was interpreted as normal
What you want to see...
What you do see...
Our Neonate

- HR 195, on ventilator, BP 45/25, SaO2 40’s (pre and post-ductal)
- Poor perfusion
- Gallop rhythm, no murmurs
Differential diagnosis?

- Meconium aspiration / CDH / HMD
- Tetralogy of Fallot
- Transposition of the Great Arteries
- Tricuspid Atresia
- Truncus Arteriosus
- “Terrible” HLHS
- Total Anomalous Pulmonary Venous Connection
What you want to see...
Key Features of Our Echo

Small appearing left heart
Key Features of Our Echo

Pure R -> L shunt at PFO
Key Features of Our Echo

P Veins to confluence
Key Features of Our Echo
TAPVR and Its Subtypes
Definition

• All pulmonary venous blood flow returns anomalously to the systemic veins or directly to the right atrium

• Prevalence estimated at 1 in 10,000

Embryology

- Early gestation lung buds drain to the common cardinal systems (right forms SVC and azygous, left forms LSVC and CS) and the umbilicovitelline system (forms the IVC, ductus venosus, and portal system)
- Day 27-29 the common pulmonary vein (CPV) begins to develop, and by day 30 connects with pulmonary venous plexus
Embryology

- Failure of the CPV to connect to pulmonary venous plexus leads to persistence of one of the primitive connections
- Failure of the septum primum to normally form can lead to pulmonary veins connecting directly to the RA
- Failure to incorporate the common pulmonary vein leads to cor triatriatum
Anatomic Subtypes

- **Type 1: Supracardiac** (43-50%)
- **Type 2: Cardiac** (18-20%)
- **Type 3: Infracardiac** (20-27%)
- **Type 4: Mixed** (10-12%)
- **Non-obstructed vs Obstructed** (for all types)

http://uttamsmedicalnotes.blogspot.com/2013/02/total-anomalous-pulmonary-venous-return.html
https://apps.childrenshospital.org/clinical/mml/index.cfm?CAT=media&MEDIA_ID=1612
Type 1: Supracardiac

- Vertical Vein most often drains to LIV

Type 1: Supracardiac

- Vertical Vein most often drains to LIV
- Can course between LPA and left mainstem bronchus - > VICE
- May present obstructed (around 50%)

Type 2: Cardiac

- Typically to the Coronary Sinus
- Least likely to be obstructed
- Can present later (few months)
Type 3: Infracardiac (Infradiaphragmatic)

- Descending vein to portal vein, IVC, hepatic vein, or ductus venosus (umbiicovitelline venous system)
- Classic teaching is they are ALWAYS obstructed -> present at birth

The Importance of the PFO/ASD

- ALL preload to the LV (aka systemic cardiac output) is supplied by right to left shunting across the atrial communication
Confluence
Visualize All Pulmonary Veins
Type 1 to the LIV
Type 2 – to the CS
Type 3 - Infradiaphragmatic
Type 3 – Infradiaphragmatic Unobstructed?
Type 3 – Infradiaphragmatic Obstructed
Type 2 Again?
Type 4 – Mixed
All types: Doppler to Assess for Obstruction
Small Left Heart
“Pulmonary Hypertension”
Atrial Communication
Atrial Communication
PDA
Alternative Imaging Options
Very Challenging to Diagnose Prenatally

- Estimated that pulmonary veins carry only ~5% of pulmonary blood flow in utero
In Utero Diagnosis
Cardiac Catheterization
Cardiac Catheterization – Stenting VV
Cardiac Catheterization – CPV Atresia
Cardiac Catheterization – CPV Atresia
TEE

- Not utilized much as can obstruct the confluence / drainage
CT Angiogram

Images courtesy of Dr. Tony Hlavacek, MUSC
Cardiac MRI
Follow up Cardiac MRI
Back to Our Patient
Emergent Surgery

• More to come in the next talk…
Clinical Course

- ICU for 16 days, total LOS 20 days
- Discharged on room air, full oral feeds, on twice daily diuretics as only medication
Clinical Course

- Now 3 years old
- Some mild developmental delays
- Clinically asymptomatic
- Mild flow acceleration at pulmonary venous anastomosis, no PH, normal function
Conclusions

• TTE remains the mainstay in diagnosis of TAPVR

• Venous flow away from the heart is BAD!
  – aka Red venous flow signals on either suprasternal notch or abdominal imaging planes are BAD!

• Visualization of the individual veins, confluence, and course of drainage are critical to accurate diagnosis
Conclusions

- Always assess the PFO, pure right to left shunting is BAD!
- Supracardiac TAPVR is the most common, and roughly 50% present obstructed
- Infracardiac are nearly all obstructed at birth
- For mixed TAPVR or if the diagnosis is unclear, CTA and Cardiac MRI can be helpful
- Cath is primarily reserved for rare cases where pre-operative intervention is needed
Thank You