Evaluation of the Right Ventricle and Risk Stratification for Sudden Cardiac Death

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Unique Forms of Heart Failure in the Congenital Heart Disease Population

- Single ventricle status post Fontan
- Right ventricular failure
  - Systolic failure of the systemic right ventricle
  - Systolic failure of the sub-pulmonary right ventricle
  - Diastolic dysfunction
  - Secondary LV dysfunction
Peak VO2 of Heart Failure Patients versus CHD Patients

Age Matched Controls

NYHA I

NYHA II

NYHA III

Peak VO2 mL/kg/min

Diller et al. Circ 2005; 112:828-835
VO2 Predicts Clinical Outcome

Diller et al. Circ 2005; 112:828-845
26 Year old Woman

- Diagnosed with tetralogy of Fallot in the neonatal period
- Operative repair at age 2
- Sporadic follow up after age 12
- Presented with progressive decline in stamina
- Elsewhere underwent mitral valve replacement with a tissue prosthesis for “mitral valve prolapse” and regurgitation
- Presents 4 weeks after surgery with intractable pleural effusions, fatigue and high grade AV block
Doppler Recognition of Severe Pulmonary Valve Regurgitation
ECHO Report

- Severe RV enlargement, moderate-severe decrease in function, RVSP 39 mmHg.
- Severe (free) pulmonary regurgitation
- LV EF 30% - 35%
- Abnormal hepatic vein Doppler related to junction rhythm
- Normal mitral tissue prosthesis

Patient medically optimized then referred for PVR
Consequences of Pulmonary Valve Regurgitation

- Exercise intolerance
- Right ventricular dilatation
- Right ventricular dysfunction
- Increased risk of ventricular tachycardia
- Increased risk of atrial arrhythmia
- Left ventricular dysfunction
Assessment of Right Ventricular Function

• **Quantitative M-mode & 2D Echo**
  - RV EF %
  - RV FAC
  - TAPSE

• **Doppler Echocardiography**
  - TV dP / dt
  - RV MPI
  - TV TDI
  - RV strain
Assessment of RV Function
Myocardial Performance Index

• Potential Advantages of MPI
  • Obtained by routine Doppler techniques
  • Non-geometric
    • LV function
    • RV function & single ventricle function
  • Quantitative
  • Global ventricular function
    • Systolic (ICT, ET) & diastolic (IRT) components
Myocardial Performance Index

Normal RV Values

- **Fetal**: 0.35 +/- 0.05
- **Peds**: 0.32 +/- 0.03
- **Adult**: 0.28 +/- 0.04
Assessment of RV Function
Myocardial Performance Index by Doppler

ET
TCO
MPI Correlates to MRI RVEF

RV Free Wall Longitudinal Peak Systolic Strain

Abnormal: Less Negative than -25%

• Angle independent
• Not prone to translational motion
Assessment of RV Diastolic Function

Hepatic Vein Doppler

Prominent atrial reversals indicate RV non-compliance.
Assessment of RV Diastolic Function

RVOT Doppler

Forward flow with atrial systole indicates poor RV compliance
Which Ventrices Normalize?

- RV end-systolic volume < 100 mL/m²
- Corrected RV ejection fraction > 20%
- RV end-diastolic volume < 160 ml/m²
- RV end-systolic volume < 82 mL/m²


Arrhythmias in Adults with Repaired TOF

• Sustained tachyarrhythmia: 29.9%
• VT: 14.2%
• IART: 11.5%
• AF: 7.4%
• Other: 6.7%
• VF: 0.5%

Risk Factors for Ventricular Tachycardia

- Inducible sustained ventricular tachycardia
- Non-sustained ventricular tachycardia
- Ventriculotomy incision
- LV systolic dysfunction
- LV diastolic dysfunction
- RV dysfunction
- QRS duration $\geq 180$ ms
Risk of VT/VF Related to LV Filling Pressure

ICD Therapies in Patients with TOF

Questions?