Value of Cardiovascular Magnetic Resonance Imaging in Noninvasive Risk Stratification in Tetralogy of Fallot.
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Take Home Points:

- In patients with repaired tetralogy of Fallot, RV EF <30% and LV EF <45% by cardiac MRI can help predict major clinical outcomes, particularly when combined to the non-invasive component of the Khairy et al risk score (prior palliative shunt, ventriculotomy incision, non-sustained ventricular tachycardia, and QRS duration ≥180 ms)

Commentary from Dr. Maan Jokhadar (Atlanta), section editor of ACHD Journal Watch: Due to the increased risk of ventricular arrhythmia and sudden death in repaired tetralogy of Fallot (TOF) patients, many require implantable cardiac defibrillators (ICD) for primary or secondary prevention. In 2008, Dr. Khairy and colleagues published a landmark study describing a risk score for appropriate ICD shocks in repaired TOF patients, which included prior palliative shunt, ventriculotomy incision, non-sustained ventricular tachycardia (NSVT), QRS duration ≥180 ms, in addition to 2 invasive parameters that include inducible ventricular tachycardia (VT), and left ventricle and diastolic pressure (LVEDP) ≥ 12 mmHg. (Circulation. 2008; 117:363 – 370)

Dr. Bokma and colleagues from the Netherlands performed a multi-center, retrospective cohort study that included 575 patients with repaired TOF who had cardiac MRI. The mean age was 31 and the mean follow-up 7.1 years. This study describes a noninvasive risk model that includes MRI derived LV and RV ejection fraction in place of the invasive parameters used in the Khairy et al risk model.

Of 575 patients, 35 (6.1%) had the primary composite endpoint, which included 13 deaths, 8 cardiac arrest/defibrillation, 4 sustained VT, and 11 recurrent and symptomatic NSVT. The 13 deaths included 1 sudden cardiac death, 5 heart failure, 1 renal failure, 1 preoperative bleeding, and 5 non-cardiac or unknown).

The C statistic of the noninvasive components of the Khairy et al score was 0.64 (95% CI, 0.54-0.73). The C statistic of the final noninvasive point-based risk model, including RV EF < 30% and LV EF < 45%, was 0.75 (95% CI, 0.63-0.85).

Bokma et al risk score:
- Prior palliative shunt: 2 points
- Ventriculotomy incision: 2 points
- QRS duration ≥180 ms: 1 point
- Previous VT: 2 points
- LV EF <45%: 2 points
RV EF <30%: 3 points
Total points: 0 to 12
*High risk: 7 to 12 points*

These thresholds appear to be applicable in patients with or without pulmonary valve replacement.

This noninvasive risk model could be used to help identify patients at risk for ventricular arrhythmia. Though additional studies are needed to determine if this score can help guide ICD implantation, it appears that this noninvasive score can may help identify patients who could benefit from invasive testing such as cardiac catheterization or electrophysiology testing. This study did not include invasive or exercise parameters, echocardiographic parameters (strain, diastolic function, or atrial size), and did not include other MRI parameters (tricuspid regurgitation, ventricular mass, or scar burden). This study does confirm the predictive importance of myocardial dysfunction.