Abstract: 2395

LGE CMR predicts sudden death and VT in adults with repaired tetralogy of Fallot- a prospective study with 3500 patient follow up years

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Background: Adults with repaired tetralogy of Fallot (rtoF) are at risk of ventricular arrhythmia and sudden cardiac death (SCD). Cross-sectional data suggest association of late gadolinium enhancement (LGE) cardiovascular magnetic resonance imaging (CMR) with adverse clinical risk factors

Purpose: We sought to determine prognosis related to LGE CMR.

Methods: In this prospective cohort study the primary composite outcome comprised the first of cardiovascular death (SCD or heart failure-related), aborted SCD (successfully resuscitated cardiac arrest or appropriate AICD shock for ventricular fibrillation), and clinical sustained ventricular tachycardia (VT>30 seconds duration).

Results: In 531 rtoF patients (median age 32;23-42, 296(56%) male, NYHA=II 17%) followed up after LGE CMR for median 5 (1.7-8.9) years, there were 39 primary composite outcomes: 10 SCD, 11 heart failure related deaths (2 perioperative RV failure), 2 aborted SCD and 16 clinical sustained VT events. At study end, there were 28 ventricular arrhythmic events in 28 rtoF patients (10 SCD, 16 clinical sustained VT, 2 aborted VF) that were significantly predicted by RV LGE extent (HR 1.45 CI:1.3-1.6;P<0.001).

Univariable predictors of the primary outcome were RV LGE score; HR:1.44 (1.31-1.57;p<0.001), (Figure) together with older age; HR:1.05 (1.02-1.07;p<0.001), late repair; HR:1.04 (1.02-1.07;p<0.001), lower RV ejection fraction; HR:0.92 (0.89-0.95;p<0.001), larger RVOT akinetic length; HR:1.04 (1.02-1.06;p<0.001) larger right atrial area;HR:1.2 (1.12-1.29;p<0.001); higher BNP levels; HR:1.01 (1-1.02; p<0.001), lower peak VO2;HR: 0.89(0.83-0.96;p=0.001), prior atrial arrhythmia; HR:5.3 (2.8-10.07;p<0.001), and non-sustained VT;HR:4.1 (2.1-7.7;p<0.001). Inducible VT did not predict the primary outcome; HR: 2.1 (0.57-8;p=0.25)

In multivariable analysis both RV LGE score and indexed right atrial area (RAAi) only, remained predictive of the primary outcome (HR 1.29 CI:1.12-1.49;p<0.001 and HR 1.1 CI:1.02-1.12;p=0.01,respectively). Patients could accordingly be stratified such that supramedian RV LGE score (=5) and RAAi =16cm²/m² had 5-year event free survival 84% vs 94% for supramedian RV LGE score (=5) and RAAi < 16cm²/m² or 98% for inframedian RV LGE score with RAAi< 16cm²/m². Figure.

Conclusions: For every unit increase in CMR defined RV fibrosis score there is a 44% increased risk of sudden cardiac death and VT. LGE CMR and maximal right atrial area should therefore be incorporated into risk stratification for sudden death in adults with rTOF.
LGE CMR predicts sudden death and VT in adults with repaired tetralogy of Fallot—a prospective study with 3500 patient follow-up years

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Background: Adults with repaired tetralogy of Fallot (rTOF) are at risk of ventricular arrhythmia and sudden cardiac death (SCD). Cross-sectional data suggest association of late gadolinium enhancement (LGE) cardiovascular magnetic resonance imaging (CMR) with adverse clinical risk factors.

Purpose: We sought to determine prognosis related to LGE CMR.

Methods: In this prospective cohort study, the primary composite outcome comprised the first of cardiovascular death (SCD or heart failure-related), aborted SCD (successfully resuscitated cardiac arrest or appropriate AICD shock for ventricular fibrillation), and clinical sustained ventricular tachycardia (VT > 30 seconds duration).

Results: In 531 rTOF patients (median age 32.2; range 23–42, 296 (56%) male, NYHA = II 17%) followed up after LGE CMR for median 5 (1.7–8.9) years, there were 39 primary composite outcomes: 10 SCD, 11 heart failure-related deaths (2 perioperative RV failure), 2 aborted SCD, and 16 clinical sustained VT events. At study end, there were 28 ventricular arrhythmic events in 28 rTOF patients (10 SCD, 16 clinical sustained VT, 2 aborted VF) that were significantly predicted by RV LGE extent (HR 1.45 CI: 1.3–1.6; P < 0.001).

Univariable predictors of the primary outcome were RV LGE score; HR: 1.44 (1.31–1.57; P < 0.001), together with older age; HR: 1.05 (1.02–1.07; P < 0.001), late repair; HR: 1.04 (1.02–1.07; P < 0.001), lower RV ejection fraction; HR: 0.92 (0.89–0.95; P < 0.001), larger RVOT akinetic length; HR: 1.04 (1.02–1.06; P < 0.001), larger right atrial area; HR: 1.2 (1.12–1.29; P < 0.001); higher BNP levels; HR: 1.01 (1–1.02; P < 0.001), lower peak VO2; HR: 0.89 (0.83–0.96; P = 0.001), prior atrial arrhythmia; HR: 5.3 (2.8–10.07; P < 0.001), and non-sustained VT; HR: 4.1 (2.1–7.7; P < 0.001). Inducible VT did not predict the primary outcome; HR: 2.1 (0.57–8; P = 0.25).

In multivariable analysis, neither RV LGE score nor indexed right atrial area (RAAi) were predictive of the primary outcome (HR 1.12 CI: 1.02–1.21; P = 0.01, respectively). Patients could accordingly be stratified such that supramedian RV LGE score (= 5) and RAAi = 16cm²/m² had 5-year event free survival 84% vs 94% for supramedian RV LGE score (= 5) and RAAir < 16cm²/m² or 98% for inframedian RV LGE extent and RAAir < 16cm²/m².

Conclusions: For every unit increase in CMR defined RV fibrosis score there is a 44% increased risk of sudden cardiac death and VT. LGE CMR and maximal right atrial area should therefore be incorporated into risk stratification for sudden death in adults with rTOF.

Figure: Kaplan Meier event free survival curves for reaching the combined clinical endpoint. A, RV LGE score quintiles: scores <3, 3–4, 5–6, 7–8 and ≥9. B, supramedian LGE extent and indexed right atrial area (RAAi) ≥16cm²/m², supramedian LGE extent and RAAi <16cm²/m², inframedian LGE extent and RAAi <16cm²/m².