Methods: Eighty-seven ToF patients (54% male, age 33±10 years, 97% in NYHA I) and 56 healthy controls were investigated with STE. Additionally, 91 patients underwent cardiac magnetic resonance imaging (CMR). Using STE, we analyzed the LV rotation curves at apex and base and calculated LV twist. Peak apical rotation (AR) > 4 degrees was defined as normal.

Results: In 63 (72%) ToF patients and in all healthy controls a normal AR was found (9.9±4.2 vs 10.7±3.8 degrees, p=0.3). However, in these patients the basal rotation (BR) was impaired compared to controls (-4.8±3.7 vs -7.2±3.4 degrees, p=0.001). In 24 (28%) patients with reduced AR, a strong relation was seen with enlarged LV dimensions and decreased systolic LV and RV function compared to ToF patients with normal AR. Multivariate regression analysis in the 51 patients who underwent CMR, showed a positive relation with LV twist and LV and RV ejection fraction (beta 0.62, p<0.001 and beta 0.38, p=0.007).

Conclusion: Patients with congenital PAH exhibit a different right heart geometry, a modified right-to-left interaction and RV systolic function with better radial RV function compared to patients with other causes of PAH, chronic thromboembolic PH and PH associated with respiratory disease. This is possibly due to the preservation of RV hypertrophy from birth in CHD and to chronic adaptation to elevated RV afterload.

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Arrhythmic risk in adult patients after repaired tetralogy of Fallot: cardiovascular magnetic resonance predictors
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Introduction: Tetralogy of Fallot (TOF) is the most common type of cyanotic congenital heart disease and adult survivors after TOF repair is growing. Chronic right ventricular volume overload due to severe pulmonary regurgitation (PR) as been pointed as the main risk factor for adverse outcome. Fibrosis evaluation by cardiovascular magnetic resonance (CMR) seems to correlate with adverse clinical outcomes. We aim to evaluate CMR predictors of arrhythmic risk in patients (pts) after TOF repair.

Methods: We included 47 adult pts who performed CMR from March 2011 till November 2012 (mean age 32.8±9.6 years; 59.5% female). Arrhythmic risk was defined as sustained supraventricular or ventricular arrhythmias clinically proved or identified by 24 hours Holter monitoring. The CMR protocol included evaluation of ventricular systolic function, ventricular and atrial volumes, pulmonary regurgitation fraction (PRF) and fibrosis by late gadolinium enhancement (LGE).

Results: 41 pts (87.2%) were asymptomatic; mean follow-up time since TOF repair 23.5±7.3 years. Right ventricle (RV) end-diastolic volume index was 133.8±44.3 ml/m², RV end-systolic volume index was 73.7±31.1 ml/m², mean RV ejection fraction was 46.1±6.3%, left ventricle (LV) end-diastolic volume (LVEDV) index was 76.7±13.9 ml/m², LV end-systolic volume index 76.7±13.9 ml/m², mean LV ejection fraction was 58.0±7.5%, LV mass index was 55.1±10.4 gr/m². Significant PR (PRF ≥30%) was present in 25 pts (62.5%), Right atrial systolic area was 23.2±7.5 cm². LGE was present in all pts at the RV outflow tract and at LV in 10 pts (25%). Arrhythmias were documented in 10 pts (21%). Male gender (p=0.032), NYHA functional class (p=0.009), right atrial systolic area (p=0.034), LVEDV (p=0.010) and LV mass (p=0.028) correlated with the arrhythmic risk. We didn’t find a correlation between the arrhythmic risk and other classic risk factors, namely QRS duration, RV volumes, PR or right LGE score. In multivariable analysis NYHA class (odds ratio 18.743, 95% confidence interval 1.305-269.196; p=0.031), right atrial systolic area (odds ratio 1.425, 95% confidence interval 1.033-1.967; p=0.031) and LVEDV (odds ratio 1.045, 95% confidence interval 1.005-1.086; p=0.027) were independent predictors for arrhythmias, after adjusting for gender and age.

Conclusions: Right atrial area and left ventricular volumes evaluated by CMR in adult pts after TOF repair correlate with the arrhythmic risk defined in this study. LGE score didn’t correlate with arrhythmias. Accurate evaluation of left ventricle seems to be an important tool in the risk stratification of this population.

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Correlation between twist and EF

Conclusion: The majority of adults with corrected ToF have a normal AR, but an impaired BR compared to controls as assessed with STE. In more than one quarter of the ToF patients an abnormal AR is observed, which is related to a decreased systolic LV and RV function. These findings suggest that an abnormal AR may be helpful for early detection of LV dysfunction.