External validation of survival predicting score in repaired tetralogy of Fallot: an opportunity to improve

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Introduction: In recent years, integrating both better diagnosis and treatment allowed most adults with repaired tetralogy of Fallot (rTOF) to expect a longer life expectancy. However, a minority is at higher risk for premature cardiovascular death. Recently, 2 scores identified the subgroup of rTOF patients (pts) who are at high annual risk of death and ventricular arrhythmias (VA). We aimed to implement both in our population.

Methods: Retrospective single center review was done on rTOF pts who undergone cardiovascular magnetic resonance with gadolinium, blood sampling for B-type natriuretic peptide (BNP), echocardiography and cardiopulmonary exercise testing. The risk scores were calculated with weighted independent predictors, namely presence of late gadolinium enhancement in left ventricle (LV) and right ventricle (RV), akinetic RV outflow length, LV and RV ejection fraction (EF), RV systolic pressure, peak oxygen uptake (pVO2), BNP, atrial arrhythmias and age. Low mortality risk was defined as 0-20 points, intermediate as 21-50 and high ≥51. In terms of VA prediction, low risk was defined as 0-20 points, intermediate as 21-39 and high as ≥51.

Results: From a total of 240 rTOF pts, 61 (56% male, mean age 34±9 years) had all the necessary data. RV dysfunction was present in 52.5%, RV systolic pressure was high in 12 and LVEF ≤35% in 18% pts. BNP level was elevated in 27 and pVO2 ≤17mL/kg/m2 in 14.8% pts. The mean mortality risk score was 15.1±11.2, being that 68.9% had a lower and 31.1% an intermediate risk. Regarding VA score, the mean was 14.7±10.8, where the majority (80.3%) were in lower risk. 29.5% had a sustain atrial arrhythmia (mainly atrial fibrillation) and 14.8% a VA. There were 3 cardiac related deaths (4.9%). Analysis of time to event data showed that mortality and VA scores were not predictors of overall mortality (M) or VA events, respectively. However, considering 1-year M, LVEF ≤35% (p= 0.002), RVEF ≤35% (p=0.002), pVO2 ≤17mL/kg/m2 (p<0.001), and both scores: M (p=0.013) and VA (p<0.001) were predictors of that endpoint. In multivariate analysis, VA risk score (p=0.035) and low pVO2 (p= 0.006) were independent 1-year M predictors.

Conclusion: In our population, ventricular arrhythmia score and low peak oxygen uptake were independent predictors of 1-year mortality. However, there is probably a selection bias. Since the scores were recently proposed, patients were not recruited prospectively, many ended up not having all the needed or recent data, thus were not included. There is the need to validate and improve available scores to truly assess the risk of mortality and arrhythmias, as well as defined protocols and longer follow-up for this purpose.