Long-term follow-up of patients with tetralogy of Fallot and implantable cardioverter defibrillator

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Topic(s):
Congenital Heart Disease – Epidemiology, Prognosis, Outcome

Citation:
Background
Ventricular arrhythmias and sudden death are potential late complications in patients with tetralogy of Fallot. Data regarding the value of implantable cardioverter defibrillators (ICD) are scarce in this population.

Purpose
To assess long-term rates of appropriate ICD therapies and ICD-related complications in a large registry of ICD recipients with tetralogy of Fallot.

Methods
The DAI-T4F study is an ongoing national French registry including all patients with tetralogy of Fallot and ICD (NCT03837574). Information have been collected prospectively since 2010 with annual update. Baseline patient characteristics and clinical events during follow-up were analyzed with central adjudication. Cox proportional hazard models were used to identify factors associated with appropriate ICD therapies and complications.

Results
A total of 134 patients (median age 41.7 years, 70.7% males) were enrolled. The median (IQR) follow-up duration was 6.1 (2.7-10.2) years. ICDs were implanted for primary prevention in 47 (35.1%) patients and for secondary prevention in 87 (64.9%) patients. Overall, 14 (29.8%) and 45 (51.7%) patients received at least one appropriate ICD therapy in primary and secondary prevention, respectively, giving annual incidences of 5.5% and 7.1% (p = 0.06). Patients with altered left ventricle ejection fraction (LVEF) at inclusion =35% experienced less appropriate ICD therapies (HR=0.31, 95% CI: 0.11-0.86, p=0.02), whereas a history of sustained or non-sustained ventricular arrhythmia (HR=2.7, 95% CI: 1.2-3.9, p=0.03) was positively associated with appropriate therapies. Fifty-seven (42.5%) patients had ICD-related complications, including 32 (24.2%) inappropriate ICD shocks, 22 (16.4%) significant lead dysfunction, 14 (10.4%) device infection, and 5 (3.7%) generator dysfunction/recall. History of supraventricular arrhythmias (HR=2.2, 95% CI: 1.2-3.7, p=0.01) and congestive heart failure (HR=2.0, 95% CI: 1.2-3.6, p=0.01) were both associated with a higher
risk of complications. During follow-up, 7 (5.2%) patients underwent cardiac transplantation and 12 (9.0%) patients died, mainly from progressive heart failure (n = 5). Only one sudden death due to electrical storm was recorded.

Conclusions

Appropriate therapies are frequent in patients with tetralogy of Fallot and ICDs, including in primary prevention. The relatively important proportion of ICD-related complication highlights the need for improving risk stratification in this population, considering associated conditions in the individual benefit-risk equation.