

Extracardiac conduit Fontan procedure has a similar arrhythmogenicity to classic Fontan procedure at long term follow-up of patients with univentricular physiology without heterotaxia syndrome

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Introduction. Arrhythmias are a common comorbidity in subjects who underwent Fontan procedure (FP). Many studies reported that the extra-cardiac conduit (ECC) FP have a lower arrhythmia burden, as compared to the atriopulmonary FP. However, the ECC cohorts described in the literature have a significantly shorter follow-up (FU) than the atriopulmonary FP cohorts. Our aim was to investigate the prevalence and time of onset of arrhythmic complications in patients who underwent ECC with long-term FU at a single institution.

Patients and methods. We retrospectively evaluated 348 patients who underwent ECC at our institution (1987-2013). Patients with heterotaxy were excluded for their intrinsic arrhythmic risk; other exclusion criteria were previous non-ECC FP, ECC take-down and loss to FU (overall 119 patients). Seven patients (3%) died during FU (only 1 sudden death). The remaining 222 patients (60.4% males) had a maximum FU after ECC of 30 years. Mean age at FU was 19.2 years, while at surgery - 4 years. Native cardiovascular anomalies were mainly univentricular hearts of left ventricular type (60%).

Results. Four patients experienced complete atrioventricular block before surgery, thus postoperative arrhythmic FU was performed on 218 subjects. Overall arrhythmia burden was 28.5% (62 patients): 4.6% early postoperative arrhythmias and 23.8% late postoperative arrhythmias, with a mean length of time (Δt) from ECC to development of arrhythmia of 9 years. All the early-onset arrhythmias were bradyarrhythmias. Among the late-onset arrhythmias 21.5% (47 patients) were bradyarrhythmias (Δt from ECC 8 years), 13.5% (29 patients) were tachyarrhythmias (Δt from ECC: 11.5 years for supraventricular tachycardias and 13.5 years for ventricular tachyarrhythmias). Fourteen (6.4%) patients had both a bradi- and a tachy-arrhythmia. 28 subjects (12.8%) required permanent pacemaker implantation. No implantable cardioverter-defibrillators were placed. The incidence of arrhythmia increased with age – mean age at FU of the arrhythmia group being 22 years compared with 18.2 years in the no arrhythmia group ($p=0.0026$).

Conclusions. This study demonstrates that arrhythmias are a frequent complication also in patients with ECC Fontan without heterotaxy, suggesting that the length of time from surgery, rather than the type of FP, is the main risk factor for development of arrhythmia.