Long-term follow up of bradi-arrhythmias after surgical correction of atrioventricular septal defects in a single-center registry of 551 children: Down syndrome is a risk factor or not?

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Background: Atrioventricular septal defects (AVSD) represent nearly 7% of all congenital heart diseases and have been described often associated to Down Syndrome (DS). Our aim was to explore the incidence of atrioventricular block (AVB) and sinus node dysfunction (SND) onset after cardiac surgery, requiring pacemaker (PMK) implantation in a large single center cohort during a maximum follow-up of 34 years and to determine the impact of DS.

Methods: Demographic information, cardiac diagnosis, age at first surgical repair, surgical procedure, age at PMK implantation of 522 partial and complete AVSD who underwent intracardiac repair from 1982 to 2016 were reviewed from the system database of Our Institution.

Results: DS was present in 202 cases (38.7% of overall population). Thirty-eight patients (7.3%) underwent to permanent PMK implantation for early or late AVB or SND. AVB was present in 26 patients (5%), of which 20 (3.8%) with early onset and 6 (1.2%) with late onset (median 5 years [IQ 4-9.5 years] after surgery), while 12 patients (2.3%) had SND, of which 10 (1.9%) with late onset (median 10.5 years [3.5-15.2 years IQ] after surgery). Children who undergone surgical repair for a partial AVSD had more frequently an early AVB than those with complete AVSD (p=0.019), while no difference was found for late AVB onset (p=0.68). Meanwhile, late SND onset was significantly more frequent in patients with complete AVSD than in partial AVSD (p=0.017). At Kaplan Meier survival curve, the presence of DS was significantly associated with late onset of bradi-arrhythmic disorders (occurring at a medium of 5,7 ± 4,4 years in non-DS patients compared to 11,6 ± 5,8 years in DS patients). Finally, we have clearly noted that the incidence of AVB has gradually declined over the years, while the DNS has maintained the same percentage.

Conclusions: Our findings recommend a close follow up to early identify potential life-threatening bradi-arrhythmias in this high risk patients. Furthermore, Down syndrome seems to not increase the risk for bradiarrhythmia disorders but to be related with a late timing onset of PMK implantation.