Long Term Evaluation of Patients with Congenital Heart Disease and Congenital Arrhythmias with Pacemakers

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Introduction: Congenital heart diseases (CHD) and congenital arrhythmia or bradycardia syndromes represent a common cause of both morbidity and mortality, and may be associated with both structural and electrophysiological concerns. Many CHD patients undergo pacemaker implantation for reasons including heart block, sinus node dysfunction, and arrhythmias and those with congenital arrhythmia substrate may have a surgical placement of a pacemaker at a young age. This study evaluates the long-term outcomes, need for repeat intervention, and longevity of pacemaker generators and leads in patients treated with pacemakers who were diagnosed with a congenital heart disease, congenital arrhythmia.

Methods: IRB approval was obtained. Surgical records, patient charts, pacemaker interrogations, and echocardiographic data were collected from the University of Iowa pacemaker, medical records and surgical databases. Patients were included if they had a pacemaker and were diagnosed CHD, congenital heart block, sinus node dysfunction or other congenital arrhythmia substrate. All patients who underwent pacemaker implantation before May 30th 2014 were included. A REDCap database utilized.

Results: The study population is made up of 239 patients, who combined have had 484 generators and 599 leads implanted since 31 March 1975. The median age at first pacemaker implant was 6 years. Of the 239 patients, 68.2% (163) utilized epicardial leads for their first pacemaker, 31% (74) endocardial leads, and 0.8% (2) a mixture of lead types. Patients with structural CHD represented 71.1% (170) of the population and included 10.9% (26) with dextro-TGA and 12.6% (30) with levo-TGA. Surgical heart block was an indication in 38.5% (92) patients, sinus node dysfunction in 15.5% (37) and for arrhythmia treatment in 14.6% (35) of patients. The most common indication for patients without structural heart disease was congenital complete heart block (24.3%). DDD (40.6%) and VVI (23.8%) were the most common pacing setting at the time of first implant. On average, the first generator was replaced 5.8 years after implantation (N=121). The mean longevities did not differ significantly for the first four generators in an individual (p=0.745). 15.1% of the population was still using their first generator 10 years after initial implantation by Kaplan-Meier. By this same time mark, approximately 70% had undergone a lead revision or lead addition. For patients requiring lead addition or revision, the mean time to this surgery was 5.78 years (N=100). Echocardiographic analysis revealed no substantial change in ejection fraction and a slight decrease in shortening fraction over a 30 year timeframe.

Discussion: This study evaluated pacemaker implants over the last four decades in the congenital heart disease and congenital arrhythmia patients. In this population, initial pacemaker implantation frequently occurs in childhood, and epicardial leads were consequently the most common initial lead type. Many patients required follow-up pacemaker surgeries within 10 years of implantation for generator depletion, lead revision, or device upgrade. Despite the need for lifelong care in this population, systolic function did not appear to suffer substantially in the long-term.