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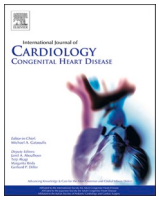

Cardiac devices in the adult congenital population; A blessing and a curse

Cardiac implantable electronic device (CIED) provision in the adult congenital heart disease (ACHD) population is a complex affair. CIED requirement can be conceptually divided into bradycardic and tachycardic indications, though the two indications frequently overlap. As medical and surgical care of ACHD patients has advanced over recent decades CIED implantation has increased, in part due to improved life expectancy [1]. More recently our understanding of the risk of sudden cardiac death in the ACHD population has increased and has resulted in improved discrimination when it comes to patient selection for primary prevention implantable cardioverter-defibrillators (ICDs) [2,3]. In part, motivation for better discrimination stems from a desire to minimise the known adverse events associated with CIEDs including 'physical' complications (lead fracture, infection, vascular occlusion, etc.) and psychological complications (inappropriate therapy, anxiety, depression, etc.) [4,5].

The appreciation of the extent of pacing-mediated ventricular dysfunction in the ACHD population has driven the bradycardia treatment of ACHD patients towards more complex devices that mitigate this risk in the form of bi-ventricular cardiac re-synchronisation therapy (CRT) [6]. CRT provides improved functional class, ventricular size and function compared to traditional pacing [7]. The advantage of this approach is offset by its complexity requiring an additional lead in the, often not-accessible, coronary sinus.

The increase in technology and advent of new techniques is all in aid of mastering the adage of 'right patient, right device, right time'. Balancing these advances is the complication rate associated with CIEDs in the ACHD population; a poorly characterised outcome. The study by Chami et al. addresses this knowledge gap and provides important insights into the outcomes of this patient group. The authors make use of their state-wide database of ACHD patients identifying 133 pacemaker patients and 78 ICD patients (47 primary prevention and 31 secondary prevention) with a median age of 39 years. The database used is comprehensive due to the Australian Medicare coverage providing for all patients and furthermore the centralisation of all ACHD care at a single centre re-enforces the robustness of the dataset. The population was representative of typical ACHD cohorts including Tetralogy of Fallot, transposition of the great arteries (palliated and congenitally corrected), Fontan circulations and a variety of other anatomies.

The findings highlight the complexity of appropriate device selection in that even in this quaternary centre of excellence one fifth of pacemaker patients required subsequent device upgrade to CRT or ICD. Excluding inappropriate therapy, approximately one third of both ICD and pacemaker patients experienced a complication. The authors rightly highlight that this represents a risk profile of 2-3x that of the non-ACHD population. This is likely to be because of the younger age of patients (longer time 'at-risk) who are more physically active (with associated accelerated lead fracture/failure) and also as a result of the anatomical and electrophysiological challenges that one needs to overcome [8]. Chami et al. confirm and expand on prior data demonstrating that a higher complexity of ACHD lesion is associated with a significant increase in complication rates (40% vs 27% in pacemaker patients and 47% vs 33% in ICD patients) [4].

Inappropriate therapy is a huge source of psychological distress, anxiety and morbidity [5]; Chami et al. reveal the rate of inappropriate therapy over median follow-up of 4.6 years to be high at 17%, very similar to the rate of appropriate therapy (19%). Inappropriate therapy was predominantly due to atrial arrhythmias; given the aforementioned detrimental psychological effects of inappropriate shocks, but also the clinical deterioration in many ACHD patients with the onset of atrial arrhythmia this data highlights the need for close collaboration between ACHD Cardiologists and ACHD Electrophysiologists. This high rate of both appropriate and inappropriate therapy cement the need for thorough and effective pre-implantation counselling of patients based on contemporary evidence of device outcomes.

CIED technology continues to rapidly evolve, for example conduction system pacing (CSP) has recently emerged as a single lead alternative to CRT with equivalent (or superior) improvement in functional class, QRS duration, LV size and systolic function in the ACHD population [9,10]. It requires the lead to be implanted either directly into the His bundle (His bundle pacing) or deep into the ventricular septum in the case of left bundle pacing. CSP is feasible in even the most complex of ACHD anatomies and the learning curve is relatively short leading to relatively rapid uptake of the technique [11–13]. Leadless pacemakers are also commercially available and may have a growing role in the ACHD population [14]. They can overcome a number of infection and access-related issues, but come with their own anatomical challenges and possible complications [15]. Finally, the entirely subcutaneous ICD (S-ICD) has entered mainstream use and has an expanding uptake in the ACHD population due to the absence of intra-vascular complications, ease of extraction and lower infective risk profile [16]. It is imperative that registries including that which Chami et al. have created and reported on are maintained to provide up to date data on these new technologies as they enter clinical practice in ACHD populations.

The authors acknowledge the usual limitation of ACHD studies of scale and highlight the need for wider-scale collaboration. The authors should be applauded for their plan to establish an (Inter-)National Registry of the Congenital Heart Alliance of Australia and New Zealand (CHAANZ). It is collaborative projects such as this that will provide the next generation of data elevating the level of evidence available for ACHD management. Moore et al. have shown that true global

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collaboration is possible in a specific study; one hopes that these networks continue to grow for the benefit of ACHD patients around the world [9].

Overall, the study by Chami et al. provides great insight into the outcomes of CIEDs in the ACHD population and the data is a great addition to our knowledge and ability to facilitate informed consent for our patients and to appropriately prepare them for their future with a device.

Declaration of interests

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Associate editor of CHD-IJC. If there are other authors, they declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

CRediT authorship contribution statement

Matthew O'Connor: Writing – original draft, Writing – review & editing. **Tom Wong:** Conceptualization, Writing – review & editing.

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