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Handling SCD risk in adult congenital heart disease: Should we InVEST in the WCD?

Advancements in both surgical and catheter-based interventional techniques for congenital heart disease (CHD) have resulted in an increasing pool of patients surviving to adulthood who can present with long-term sequelae related to the underlying condition or the corrective surgery performed. Ventricular arrhythmias constitute an area of particular concern, with an attendant risk of sudden cardiac death (SCD) in this population [1]. The implantable cardioverter defibrillator (ICD) is undoubtedly an important tool to combat SCD in these subjects; however, implanting a lifelong device in this relatively young patient population comes with a significant risk of long-term complications, both in terms of inappropriate shocks and lead-related problems [2]. Thus, careful patient selection is crucial, with permanent device implantation being reserved for those who definitively need it. Decision-making is not always straightforward in this regard; often one needs to assess the impact of surgical or other interventions on ventricular function and arrhythmia risk which may entail a period of observation. At other times, immediate ICD implantation may be precluded by the clinical situation of the patient such as the presence of active infection. However, deferring ICD placement potentially leaves the patient vulnerable for SCD during this phase. In this context, the wearable cardioverter defibrillator (WCD) has been proposed as an alternative to the ICD to manage this transient risk situation.

The WCD had been demonstrated to reliably diagnose and terminate life-threatening arrhythmias, presenting a feasible alternative to an invasive, permanent implant for a short period of time [3]. However, studies assessing the utility of the WCD in the adult CHD population are relatively lacking. Sarubbi and colleagues, in this issue of IPEJ present a single center experience of using the WCD in adults with complex CHD [4]. They report the results of WCD use in eight consecutive patients over an average duration of 4 months. Several noteworthy findings emerge from this study. Firstly, none of the patients experienced any appropriate therapies for ventricular arrhythmia. This needs to be interpreted with caution due to the limited sample size; it is likely that studies with greater numbers would reveal a fair proportion of appropriate therapies, given the significant arrhythmia risk in complex CHD substrates [1,5]. However studies encompassing adequate number of CHD patients are difficult to perform; in a nation-wide Swiss WCD registry, only 3.3% had CHD, with no appropriate therapies recorded during the study [6]. In another WCD study confined to CHD and inherited arrhythmia syndromes, again no appropriate therapies were seen in the 43 CHD patients studied; however the

mean follow-up was only 27 days [7]. Hence more studies in greater numbers of CHD patients with adequate follow-up, assessing the utility of the WCD, are clearly needed. Encouragingly, there were no inappropriate therapies in the present study. Again, although the small number of patients studied precludes firm conclusions, other observational studies have also shown low rates of inappropriate shocks from the WCD [6,8]. A useful feature of the WCD is the occurrence of an audible alarm on arrhythmia detection and a patient response button which allows a shock to be aborted in case of a well-tolerated arrhythmia or inappropriate detection, thus affording patients a sense of confidence and control. In a large French study, this feature allowed shock to be aborted in 95.4% of inappropriate detections, with a resultant inappropriate shock rate of <1% [8], although it is not clear whether the response button was used by any patient in the present study.

A key point in the present study was that good compliance was seen with a fairly high median daily wear time of 21 hours. In the landmark VEST trial, there was no significant reduction in arrhythmic death in the WCD arm; however compliance with WCD was quite poor in the trial overall, with a median wear time of only 18 hours per day. Furthermore, in that trial, 75% of patients who died in the WCD arm were not wearing the WCD at the time of death, obviously precluding any benefit from the device [9]. Subsequent as-treated and per-protocol analysis of the VEST trial showed a significant reduction in arrhythmic mortality [10]. Thus ensuring high wear time is crucial, especially in the relatively younger CHD population, where compliance may potentially be an issue [8]; in this regard the findings of this study are reassuring. This is also in line with findings from large real-world WCD registries where median daily wear times exceeding 23 hours have been documented, underlining the importance of proper patient education and counseling in order to derive benefit from the WCD [8,11]. Remote monitoring is also useful in this context with the opportunity to promptly provide telephonic reminders in case of non-compliance. In the current study too, daily remote monitoring was done which may have contributed to high compliance.

The paper by Sarubbi et al. also suggests the feasibility of extended use of the WCD beyond three months. While a duration of three months has been put forth as a putative time for WCD use, based on guidelines framed for ischemic or non-ischemic cardiomyopathies to assess response of left ventricular function to optimal medical therapy or coronary revascularization, this is rather arbitrary [12]. Indeed, in a large German registry, one-third of patients with WCD use >90 days further improved their EF and ICD implantation was avoided [13]. Furthermore, SCD substrates are often complex in the CHD population, with a combination of scar and hemodynamic stresses both contributing to SCD risk.

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Thus, decision-making is more individualized and certainly a subgroup of patients can benefit from prolonged assessment beyond three months. An analysis of the individual cases presented in Table 1 of the present paper is very illustrative, with only 50% of them finally needing an ICD due to clinical improvement for varied reasons. The information in Table 1 also highlights typical scenarios in adult CHD wherein the WCD could be potentially valuable while awaiting definitive decision-making for ICD implantation.

Can the WCD be routinely advocated in the adult CHD population and what is its potential role in Low-Middle Income countries such as India, given significant cost constraints? Firstly, as mentioned earlier, more data is needed in this unique population subset to accurately assess the extent to which patients would benefit in terms of actual SCD prevention with the WCD and also to reconfirm safety. Large multicenter collaborations are needed to be able to gather data from a sufficient number of patients with adequately long follow-up. Secondly robust assessments of cost-effectiveness are necessary especially in the context of developing countries. Western studies evaluating the cost-effectiveness of the WCD thus far have come up with varying cost-effectiveness figures ranging from 20,000 to 50,000 US dollars (USD) per quality-adjusted life year gained [14,15]. However, many of the studies have been performed in the context of an ICD being temporarily explanted due to an infection which is a very different scenario from the issue of upfront use of a WCD for primary prevention. A key factor, clearly, will be the pricing of the WCD by the company. A Chinese study demonstrated that cost-effectiveness sharply declined when WCD costs exceeded 70 USD per day [16]. In resource-limited settings, if the cost of using a WCD for three months equals or exceeds the cost of an ICD, this could be a potential deterrent for uptake of this therapy. Other barriers for WCD use may be encountered in the context of developing countries. Ensuring compliance could be challenging given varying educational levels and patient insight. Social acceptance may also be difficult, especially in women.

The above considerations notwithstanding, the WCD is an important tool which provides clinicians with an option to handle the transient risk of SCD in selected situations in patients with adult CHD. Sarubbi and colleagues' efforts to provide much-needed data in this challenging patient subset need to be appreciated. Ongoing and future studies will hopefully throw better light on the right scenarios in which one would consider investing in the WCD with maximal benefit to the patient.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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