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EDITORIAL COMMENT

Arrhythmic Risk in Shone Complex

Lumping the Heterogeneity Together*

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In this issue of *JACC: Advances*, Bhalla et al[1] present a retrospective single-center observational study on the arrhythmic risk of patients with Shone complex. As Shone complex is a very rare congenital abnormality compromising only approximately 0.6% of all congenital heart malformations, few centers overview cohorts of patients or are regularly faced with arrhythmic risk estimation. Bhalla et al. reported on 73 patients with a Shone complex over a period of 21 years, present the largest observation to date and therefore have to be congratulated for their efforts. Prediction of arrhythmic risk in adults with congenital heart disease (CHD) is of major interest and clinical relevance, as life expectancy in CHD has fortunately increased and arrhythmias are a common long-term complication.[2,3] As stated by the authors, the retrospective and observational design is a limitation of their study; however, the long follow-up was necessary due to the rare occurrence of this condition.

One may question whether the heterogeneity of Shone Complex can be analyzed together in terms of arrhythmogenic risk stratification. Different morphologies bear noncomparable functional impairments. Furthermore, differences in surgical strategies lead to a variety of arrhythmic substrates and subsequent risks for atrial and ventricular tachyarrhythmias. There is no more fertile substrate for arrhythmias than that of surgically corrected CHD. Heterogeneity and the long inclusion period are associated with changes in operative techniques over time with probable alterations in arrhythmogenic risk. The impact of the surgical approach on long-term occurrence of arrhythmias has been described previously in CHD cohorts (eg, tetralogy of Fallot). In general, surgery aims for fewer operations at younger ages with as little as possible alteration of valve and ventricular function.[4] Nevertheless, the above-mentioned applies to most CHD, and heterogeneity should not deter investigators from studying the risk of tachyarrhythmias. Counseling adults with CHD in terms of arrhythmic risk usually implies dealing with huge heterogeneity.

In terms of atrial tachyarrhythmias in patients with Shone complex, the risk is high with 33% of patients experiencing a variety of atrial tachyarrhythmias. Comparably high rates have been reported in isolated mitral valve surgery.[5] Atrial fibrillation has the highest prevalence in Shone complex patients followed by typical and atypical atrial flutter. One important finding from the Bhalla et al. study is that risk factors for atrial tachyarrhythmias in Shone complex do not differ substantially from known risk factors: higher atrial volumes, more prior operations, mitral valve surgery, and impaired hemodynamical status. Most atrial tachyarrhythmias occur as early as the third decade of life. The occurrence of supraventricular arrhythmias in patients with CHD is known to occur approximately 30 years earlier than in patients without CHD.[6] In addition, poor tolerance of atrial tachyarrhythmias in Shone complex patients is comparable to other forms of CHD and necessitates prompt intervention. Unfortunately, results for rhythm control are disappointing with an 80% relapse after surgical or catheter-based ablation. However, as discussed above, a heterogeneous population over a long period is presented, and improvements in catheter and surgical ablation techniques may have ameliorated success rates over time.

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Regarding ventricular tachyarrhythmias in patients with Shone complex, the study by Bhalla et al. is reassuring. First, the risk for ventricular arrhythmias is moderate. Second, the majority of patients with an implantable cardioverter defibrillator (ICD) and appropriate intervention had either a reduced ejection fraction or a substrate based on coronary occlusion during operation (n = 1). Overall, only 2 patients of the cohort presenting with broad complex tachycardias remain without known risk factors for ventricular arrhythmias. These 2 patients are interesting to look at thoroughly to reveal specific risk factors uniquely applicable to Shone complex patients. Again, heterogeneity comes into play as aortic coarctation seems to be the only similarity despite rather different clinical settings in the 2 patients. Nevertheless, the fact that clinicians can rely on left ventricular function as a risk predictor for ventricular tachyarrhythmias, even in complex CHD, is reassuring. Closer characterization of myocardium and scar burden by cardiac magnetic resonance imaging would be of interest especially in Shone complex patients with multilevel left-sided obstruction (eg, scar burden). As with atrial tachyarrhythmias, the rare prevalence of Shone complex and long follow-up are associated with changes in management over time that may impact study findings.

The rate of ICD interventions in patients with reduced left ventricular function and a primary preventive ICD is astonishingly high with 75% experiencing appropriate ICD interventions during a mean follow-up of 7.3 years. Further studies in patients with CHD will have to clarify the role of subcutaneous ICD systems in these patients.[7] The rate of ventricular arrhythmia burden contrasts with primary preventive ICD indications in ischemic and nonischemic heart diseases but has also been reported for other CHDs earlier. In a mixed cohort mainly compromising tetralogy of Fallot patients, appropriate ICD intervention rate was 22% in a 3.3-year follow-up.[8] Whether the impaired left ventricular function in Shone complex is the final path of profoundly restricted hemodynamics remains unclear. Nevertheless, a key clinical message in terms of sudden cardiac death in patients with Shone complex is the disproportionally high risk in patients with reduced left ventricular systolic function with a high benefit of primary preventive ICD implantation.

In conclusion, irrespective of heterogeneity in CHD, collecting observational evidence is helpful in counseling patients. Retrospective and observational data should be seen as incentives to establish registries and conduct prospective studies. Thanks to the detailed work by Bhalla et al., we know more about Shone complex, and their important efforts will further optimize patient care in CHD.

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