Pulmonary atresia with intact ventricular septum: how good is a biventricular repair?

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In this issue of EJCTS, Romeih et al. [1] present some interesting and provocative data regarding long-term outcome for biventricular repair of pulmonary atresia with intact ventricular septum (PAIVS). Debate relative to timing, technique and surgical indications has been ongoing for decades, and major centres still employ diverse management strategies for this relatively uncommon malformation [2]. The current cross-sectional study is very well conceived and designed, employing state-of-the-art imaging tools to assess exercise capacity and pharmacological stress response in a small group of (asymptomatic) long-term survivors of biventricular repair of PAIVS. Of note, all patients had a tricuspid Z-value > 2.5 at birth, with 2 or more right ventricular (RV) segments present. This set of qualifications suggests that the study group represents the more favourable end of the spectrum of PAIVS as understood in the era of operation. Whatever the actual right ventricular outflow tract strategy employed, all patients had important pulmonary insufficiency at late followup, despite the universal presence of age-related RV diastolic dysfunction. Reduction in exercise capacity and limitation of response to pharmacological stress were also age-related. From clinical experience, the carefully documented current findings are not unexpected, and, of course, this cohort represents only children qualifying for an early biventricular repair. There is a growing body of data, some cited by the authors, suggesting that Fontan patients (with varying anatomy, and without arrhythmias) can achieve or surpass the exercise capacity of patients undergoing biventricular repair [3]. Of course, this is only a part of the Fontan story, but an important observation nonetheless, and the one which invites comparative studies using the same techniques. Effects of age at initial repair (as opposed to follow-up interval) and the risk-benefit analysis of valved repair strategies would also be of interest. The authors must be commended on this excellent study which does address some important questions relating to late follow-up for biventricular repair of PAIVS.

REFERENCES

