126. Impact of right ventricular dominance and AVV surgery in patients with Fontan circulation and AVSD

Objective: Outcomes in patients with atrioventricular septal defect (AVSD) who survive to Fontan completion are better than previously thought. However, the impact of ventricular dominance and atrioventricular valve (AVV) surgery remains unclear. We sought to determine the impact of ventricular dominance and AVV surgery on survival and AVV function in patients with AVSD and Fontan circulation.

Methods: We conducted a retrospective study of 1703 patients in the Australia and New Zealand Fontan Registry, who survived Fontan operation between 1987 and 2021.

Results: Of 174 patients with AVSD, 60% (105/174) had right ventricular (RV) dominance and 40% (69/174) had left ventricular (LV) dominance. The median follow-up time after Fontan operation was 12 years (interquartile range 10-13 years). A total of 24% (41/174) of patients underwent AVV surgery, and there was no significant difference in the proportion of patients undergoing AVV surgery with LV dominance (26%, 18/69), compared to those with RV dominance (22%, 23/105, p=0.5). The cumulative incidence of new moderate AVV regurgitation 25-years after Fontan operation in patients with LV dominance was 46% (95% confidence interval [CI] 26%-64%), compared to 53% (95% CI 38%-66%) in patients with RV dominance (p=0.31, Figure 1A). Twelve percent (21/174) of patients died or underwent heart transplantation. Transplant-free survival at 25-years after Fontan operation in patients with LV dominance was 94% (95% CI 86%-100%), compared to 67% (95% CI 52%-87%) in patients with RV dominance (p=0.007, Figure 1B). RV dominance (hazard ratio 5.9, 95% CI 1.4-25.4) was the only risk factor for death or transplantation after Fontan operation. There was no significant difference in the cumulative incidence of new moderate AVV regurgitation after Fontan operation in patients who underwent AVV surgery before or at Fontan completion compared to those who did not (10-years: 37%, 95% CI 19%-55% vs 22% 95% CI 15%-30%, p=0.26). Of note, transplant-free survival after Fontan operation was not significantly different in patients who underwent AVV surgery before or at Fontan completion compared to those who did not (15 years: 81%, 95% CI 62%-100% vs 88%, 95% CI 81%-95%, p=0.33).

Conclusion: Ventricular dominance does not impact the rate of AVV failure in patients with AVSD and Fontan circulation. RV dominance, rather than AVSD itself, is the dominant risk factor for death or transplantation after Fontan operation. Patients with AVSD who undergo AVV surgery before or at Fontan completion are not at increased risk of death or transplantation compared to those without AVV surgery before Fontan completion.

Greg King (1), Ajay Iyengar (2), Rachael Cordina (3), Nelson Alphonso (4), Yves d'Udekem (5), Igor Konstantinov (1), Edward Buratto (6), (1) Royal Children's Hospital, Melbourne, Victoria, (2) Starship Children's Health, Auckland, Auckland, (3) Royal Prince Alfred Hospital, Sydney, New South Wales, (4) Queensland Children's Hospital, Brisbane, Queensland, (5) Children's National Hospital, Washington, DC, (6) Royal Children's Hospital, Melbourne, Parkville, Victoria