Long-term Outcomes after Surgery

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The Australian and New Zealand Fontan Registry collects all data of patients who have undergone Fontan surgery living in the region. With 1300 Fontan patients alive it is the largest database of Fontan patients. It is has allowed to specify outcomes for the Fontan population. The survival is better than expected, with 76% of the patients with an atrial pulmonary connection Fontan alive at 25 years and 97% of those with a lateral tunnel and an extra cardiac conduit alive at 15 years. These results clearly show us that the majority of Fontan patients will survive three decades after their Fontan operations. We have already demonstrated that their physical capacity was better than expected, and that their quality of life was absolutely normal.

We are now facing the challenge to support this population as they age into adulthood. There is doubt about the best medications to support them. We have demonstrated that Warfarin is of no benefit compared to Aspirin. No anti-hypertensive agents bring any benefits. The only therapy that has been proven effective in increasing cardiac output and exercise capacity is regular exercise training. The possibility of a large proportion of these patients facing liver and renal failure is looming. The majority of these patients present mild abnormalities but their real impact is still unclear.

The therapeutic options for patients facing a failure of their circulation are being explored. It has become clear that Fontan conversion of atrio-pulmonary connection should occur early if we want the patients to benefit from this intervention. Supporting the circulation with assist devices met its first success’. Centralized system for heart transplantation for these complex patients is mandatory to achieve favourable outcomes.