

Research with heart

Trainee Dr Ajay Iyengar hopes to improve long-term outcomes from childhood surgery

HIGHLIGHTS

2013: RACS Foundation for Surgery Eric Bishop Scholarship

profile

2013: NHMRC Grant "Detection of Liver and Renal Function Abnormalities in the Australian & New Zealand Population of Fontan Patients"

2012: RACS Foundation for Surgery Catherine Marie Enright Kelly Scholarship

2011: RACS First Part Surgical Examinations (Basic Surgical Sciences/ Cardiothoracic Specialty-Specific/ Clinical Examination)

RACS Clinical Examination Committee Prize

2007: Co-Investigator and National Heart Foundation Summer Scholar: "The Australia and New Zealand Heart Research Group Fontan Database: An international, multicentric experience"

> rants provided by the College, the NHMRC and the National Heart Foundation have allowed Melbourne cardiothoracic Trainee Dr Ajay Iyengar to analyse the outcomes, failure rates and risk factors facing children in Australia and New Zealand who have undergone the Fontan Procedure to treat complex congenital heart malformations.

Only offered in New Zealand since 1975 and Australia since 1980, the Fontan Procedure is used to treat children born with only one ventricle and involves connecting the caval veins directly to the pulmonary arteries allowing blood to pass passively through the lungs without going through the heart (Figure 1).

Dr Iyengar said, however, that as the Fontan population aged, long-term risk factors and physiological effects now needed to be analysed and understood.

He is now conducting that population-based analysis of Australian and New Zealand Fontan patients using data generated through the Australia and New Zealand Fontan Registry, a



The Fontan procedure. Three modifications of the Fontan procedure have been performed in Australia and New Zealand since 1975.

multi-centre, bi-national registry established in 2008, the world's first such Fontan registry.

In particular, Dr Iyengar is seeking to analyse early patient outcomes following the surgery, the long-term rate of failure, death, transplantation, arrhythmia and thromboembolic events and to examine the effects of warfarin and aspirin, the two main anticoagulation regimes given to Fontan patients.

"Fontan patients obviously undergo a significant physiological change and now that the population is growing steadily (Figure 2) and aging, we are in a better position to understand the long-term effects of that change," Dr Iyengar said.

"For instance, we know their veins are under much higher pressure, that the blood passing through the lungs isn't pulsatile and that cardiac output is fixed even during strenuous exercise.

"There is also enormous inter-centre variation in the practices of anticoagulation and fenestration and we need to know if there is a significant difference between regimes to determine optimal long-term treatment.

"We are also now seeking to determine the incidence of subclinical chronic liver and renal disease amongst Fontan patients, long-term complications that are only now emerging."

Dr Iyengar, who helped establish the Fontan Registry as a medical student and intern, said information was gathered from the six centres conducting the surgery and the further six centres following up adults with congenital heart disease in Australia and New Zealand.

He said that approximately 65 Fontan procedures were conducted in Australia and New Zealand per annum and said the information collected via the registry was invaluable given the heterogeneous nature of the Fontan cohort.

"Some of these children are missing the left ventricle and some the right, and while we do not know the exact cause of these defects, the usual risk factors for congenital heart disease including genetic and chromosomal abnormalities and parental exposure to environmental factors have been implicated," he said. "These children, however, are at the worst end of the congenital heart disease spectrum, and many have already undergone multiple surgeries before having the Fontan Procedure which is now usually offered at around four to five years of age.

"We now also have a subset of patients with hypoplastic left heart syndrome, children who have only begun to survive since the development of a very large and complex operation known as the Norwood Procedure in the last decade.

"Long-term outcomes for all Fontan patients have significantly improved; however, the increase in the proportion of patients with hypoplastic left-heart syndrome has led to high rates of re-intervention and long-term failure and we need to understand this so that we can offer the best possible advice to the parents of such very sick babies."

Dr Iyengar is conducting his PhD research through the University of Melbourne, the Department of Cardiac Surgery at the Royal Children's Hospital and the Heart Research Group, Murdoch Children's Research Institute.

His work is being supervised by Associate Professor Yves d'Udekem, Department of Cardiac Surgery at the Royal Children's Hospital and Heart Research Group at the Murdoch Children's Research Institute, Professor David S Celermajer, Department of Cardiology at Royal Prince Alfred Hospital and the Sydney Heart Research Institute, University of Sydney, and Professor John Hutson, Department of General Surgery, Royal Children's Hospital and Surgical Research at the Murdoch Children's Research Institute.

A helping hand

The College has supported Dr Iyengar through the Foundation for Surgery Catherine Marie Enright Kelly Scholarship for 2012 and the Eric Bishop Scholarship in 2013, with monies provided to top up funding provided through external funding agencies.



Steady growth of the Fontan population in Australia and New Zealand, with a recent exponential growth of the proportion with hypoplastic left heart syndrome (HLHS). Source: Australia and New Zealand Fontan Registry.