Australian Safety and Quality Goals for Health Care
Proposal for establishing a registry of outcomes after procedures after congenital heart disease.

Congenital heart disease, the most frequent cause of death in children. Whilst well-treated at birth, it exposes patients to late complications.

Congenital heart disease is one of the most frequent causes of death in children, accounting for 20% of perinatal deaths and 5,000 years of life lost. Around 50% of children born with congenital defects will require corrective surgery because without it the majority of these children would die in early childhood. The interventions to treat these conditions have become very successful to save the lives of these children. When this surgery was pioneered in the 1960s, the mortality was close to 20% even for simple conditions. However today, most of the procedures performed have a very low mortality of close to 1%. In 2009 in Australia, 2,322 of these procedures were performed in 1,556 patients.

In recent years, it has been shown that almost all patients receiving these treatments will be exposed to complications later in life, which often requires further interventions. In our recent work, we have shown that one in four patients undergoing Tetralogy of Fallot repair, the most frequent congenital heart condition to make children cyanosed or blue, will require a second operation after 15 years. Others suspect that after 30 years, half will require a reintervention. We are now realising that even the simplest conditions that we treat are leading to complications. For instance, it is now suspected that half of the patients undergoing repair for coarctation of the aorta will develop hypertension after the operation, and this is currently mostly going undetected.

It is becoming clear that congenital heart disease requires not only interventions at an early age, but also continuous follow-up and care of all patients throughout life.

Patients who should be closely followed up after cardiac interventions are not followed.

In our recent audits of outcomes of patients’ interventions, we are realising that an increasing number of patients are lost to follow-up as time goes by. The most dramatic drop in the follow-up of patients occurs after adolescence. We estimate that of patients with very severe conditions such as having only one pumping chamber, the rate of dropping from follow-up is 30%. For conditions considered more benign, such as coarctation of the aorta, this rate is close to 50%. In a recent audit organised by Heartkids, the parent association of children with heart conditions, the loss of patients to follow-up has been identified as one of the most important issues affecting this population of children. HeartKids estimate that up to 60,000 individuals may have been affected by congenital heart disease in Australia. This document was presented to the Australian Parliament on 21 February 2011.

Loss of patients to follow-up represents a significant burden, because patients suffering complications will be treated at a later stage, causing harm and increasing costs of care.
It is likely that a large number of the complications that will develop in these children can be prevented before they cause significant harm, if they are detected at an early stage. Hypertension can be treated. Surgical obstructions stemming from fixed structures not growing as the child grows can be relieved. Early intervention will prevent the heart from deteriorating as it fights against the increased load caused by obstructions or leaking valves. All of the complications occurring after congenital heart disease interventions are insidious, meaning they occur without the patient noticing symptoms. They develop over very long periods of time and, unlike their adult counterparts, children, adolescents and adults with congenital heart disease will complain of symptoms only when they are very severe. If we want to treat them promptly, we have to detect complications before they cause symptoms. Patients selected for intervention at an early stage will require less costly treatments than if left until they become symptomatic. The classical example frequently seen is the patient with Tetralogy of Fallot repaired in infancy who, rather than having a simple operation in adolescence, will ultimately require heart transplantation.

**An outcome registry will improve the quality of care of patients with congenital heart disease.**

It has now been demonstrated that monitoring outcomes results in improved quality of care, in particular in heart conditions. In the case of patients with congenital heart disease we expect to notice immediate effects, as we believe that an outcome registry will enable us to detect and track down patients who are being lost to follow-up. We intend to organise a recall system to closely encourage patients to come back for a visit if they neglect to present to their follow-up appointments. Early detection of the expected complications will result in a decreased rate of morbidity or death from these complications.

**The project is highly feasible. The groundwork has been completed.**

**ROCK database.** With initial support from Heartkids, The Children's Hospital at Westmead, Sydney and Royal Children's Hospital Melbourne, a registry of outcomes has been set up. The IT platform is complete and allows accumulation of identified data in State-based 'silos' but allowing manipulation and reporting of de-identified data on a National basis using internationally recognised coding systems. Secure token-based entry to the system has been implemented. The ROCK (Regional Outcomes for Cardiac Kids) is supported by all centres treating congenital heart disease in Australia and at this stage is used as a research tool for following cohorts of patient. The longer term plan is to extend this to all children having surgery and catheter based procedures however this will require coordination of ethics approval, formalisation of governance structures and employment of clinical data entry personnel at all sites.

**Fontan Registry.** The first outcome data that is now being entered into the ROCK database involves patients with the worst conditions, namely those born with a single pumping chamber. This project is so successful that within three years of conception it has become the largest database of these patients in the world. It demonstrates the excellence of the collaboration between the different Australian centres and the feasibility of the project.
With minimal funding, we will complete the work.
The project is currently at a standstill because no centres can find the resources to enter the data. A full-time data manager each for Victoria, New South Wales, Western Australia, Queensland, and South Australia would capture this whole population.

The project can be easily monitored and will lead to policy changes.

The success of the project will be easily monitored by (1) the rate of follow-up of patients and (2) the improvement in patients’ outcomes.

It is clear that government policies could improve the care of this patient population. For example, if we were to find that patients are not being referred to the appropriate specialists, health policies could be modified to ensure that patients are receiving appropriate follow-up.

Conclusion

Australia has the expertise and the resources to provide world-leading quality of care for patients with congenital heart defects. Whilst we currently offer all patients the very highest standard of early diagnosis, intervention and monitoring, there is still significant room for improvement when it comes to ensuring that these services are not just offered but also closely monitored and implemented in all patients as they grow up. A registry of outcomes would ensure closer monitoring and the early detection and treatment of complications as they occur.

1. Leggat, S. Childhood Heart Disease in Australia. 1–52 (2011).
5. Hilbert, J. E. et al. If you build a rare disease registry, will they enroll and will they use it? Methods and data from the National Registry of Myotonic Dystrophy (DM) and Facioscapulohumeral Muscular Dystrophy (FSHD). Contemp Clin Trials 33, 302–311 (2012).
The following representatives participate to this submission. This project has reached agreement of all centres involved in the care of congenital heart disease throughout Australia.

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