

Paediatric Cardiac Surgery: Progress and Challenges-Are We Getting There?

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Globally, defects in the heart are amongst the most common congenital diseases occurring at birth. Incidence of congenital heart defect (CHD) is about 8-9 per thousand live births and rate of disease is fairly stable across countries and populations. Considering the total projected live births of around 140 million in 2020, there could be close to 1.3 million children born with CHD this year alone! ⁽¹⁾ CHD is presenting as a significant health problem amongst children in developed, developing and under-developed nations, and there is a clear felt need for specialized centres with comprehensive programmes for treating these children. CHD's are the fourth leading cause of neonatal deaths. ⁽²⁾

Up to a third of babies born with CHD require some form of an intervention early in life. Even though number of percutaneous interventions done in cardiac catheterization lab have been steadily increasing over the last 2 decades, the vast majority of interventions are still cardiac surgeries. So, paediatric cardiac surgery is an indispensable component in curative and palliative services for CHD and a team of competent and highly trained professionals that include paediatric cardiac surgeons, paediatric cardiologists, intensivists, anaesthesiologists, nurses, and perfusion technologists form the essential human resource for any CHD programme. It would be inappropriate if any personnel with no or limited specialized training, deliver paediatric heart care, as happens in some centres where surgeons primarily doing adult cardiac surgeries perform surgeries in children with CHD also. Developing local expertise for treating children with heart disease has ripple effects on other health services in the organization too. Paediatric cardiac surgery interfaces with many other sub-specialties, so investments in paediatric cardiac surgery can, therefore, lead to parallel improvements in many departments of a hospital. ⁽³⁾

History of cardiac surgery dates back close to 8 decades and in the earliest era, operations on children and young adults with CHD accounted for lion's share of heart surgeries. Closure of patent ductus by Gross in Boston, MA (1938), repair of coarctation of the aorta by Craford in Stockholm, Sweden (1944), and the "blue baby" shunt procedure by Blalock et al in Baltimore (1944) were landmark achievements that signalled the birth of medical and surgical subspecialties of paediatric cardiology and congenital heart surgery. From these modest, albeit monumental beginnings, paediatric cardiac surgery has indeed come a long way with remarkable paradigm shift towards better results. These include innovation of cardio-pulmonary bypass (CPB) machines, miniaturization of CPB circuits, filtration techniques and better myocardial protection strategies. Surgical techniques are more refined now and are continuously evolving. Anaesthetic techniques have effectively kept up with these advances with increasing focus towards enhanced recovery after paediatric cardiac surgery (ERAS). Fast tracking and ultra-fast tracking in a few instances of even neonates/infants with complex CHD's, where they are extubated in

operation room (OR) itself or in the first few hours on ICU, was identified as one of the important international quality improvement initiatives, as well as a means for better use of resources. ⁽⁴⁾ In fact, it has been the author's observation and practice over the last few years that with proper planning and a motivated team it is possible to extubate majority of patients in the OR. We have followed this management plan even in neonates undergoing Norwood operation, which is considered to be the one of the most complex cardiac surgeries. This is probably the only such reported practice in the world. ⁽⁵⁾ Introduction of medications like Prostaglandin (PGE1) have made a sea change in salvaging new-borns with duct-dependant circulations to facilitate safe transition to cardiac surgery within the first few days to weeks of life.

One unmistakable realization regarding paediatric cardiac surgery is that it is quite a demanding specialty. One needs elements of technical skill, judgment, diagnostic acumen, physical stamina, and emotional intelligence to be effective and successful. It is not only hard to get to do, but it is indeed hard to do so, also. ⁽⁶⁾ When the great Dr. William Norwood remarked that congenital heart surgery may be the most difficult specialty in medicine, the author is more disposed to agree with him in all humility, even at the risk of sounding pompous. Over the last few decades, we have been operating on much younger and smaller patients and addressing more complex pathologies. At the same time, confidence of paediatric cardiologists when they refer cases and expectations of parents when their children get operated, of a successful outcome are not only increasing, but sometime border on irrational despite extensive counselling. All these make the task of a paediatric heart surgeon and the team not only more challenging and but also extremely stressful. Nevertheless, the field is one of the most gratifying specialties and presents lot of exciting work to do.

In the current era, across-the-board operative mortality for all patients in large international, multi-institutional congenital heart surgery registry databases is about 4%, or slightly less. However, there could still be a significant post-hospital morbidity burden after discharge in these patients despite 96% of them going home after treatment. Short-term and long-term outcomes, with respect to survival, have improved substantially and continue to get better for the most challenging forms of congenital heart disease. The break-through being early diagnosis in the intra-uterine period itself by foetal echocardiography and planning intervention within the first few days of life in a dedicated paediatric cardiac centre. A step-ahead, although in early phases, would be therapeutic interventions on the heart in the mother's womb itself! Anomalies such as transposition of the great arteries for which "anatomic repair", as early as first 3-5 days of life is indicated, offers a particularly good chance of restoring normal or near-normal cardiovascular physiology. ⁽⁷⁾ Initial reparative surgery for Tetralogy of Fallot is gaining popularity since it has an extremely high expectation of survival even though interventions might be

needed subsequently to preserve the function of the right ventricle. This has been made possible due to intense long-term follow-up and development of clear guidelines for interventions. ⁽⁸⁾ Finally, anomalies like interrupted aortic arch with ventricular septal defect, left ventricular outflow obstruction, and anomalies with univentricular physiology which form the spectrum of complex CHD's, reasonable palliation can be achieved without heart replacement even though they entail staged surgical interventions. Surgical results are dramatically better than those achieved a few decades ago but one has to acknowledge the distressing realities concerning durability of the surgical treatment and less than optimal quality of life. ⁽⁹⁾

To conclude, the focus of modern-day paediatric cardiac surgery should be on having a rational, evidence-based and a multi-disciplinary team approach to improve survival while optimizing functional status and quality of life. It is but an undeniable reality that, except for few simpler anomalies, majority of CHD are still chronic diseases with lifelong implications for a child's health and well-being, while adding burden on the healthcare systems. ⁽¹⁰⁾ A lot of work still needs to be done to safe-guard the life of these children and to add quality to their lives rather than just adding quantity.

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Front Row(L to R): Dr. Richie Jain, Dr. Rajnish Garg, Dr. Jameel Thareen Khan, Saeed (father of the baby) Dr. Roberto Di Donato, Dr. Anil Ramaiah, Dr. Munira Al Maazmi Second row(L to R): Dr. Fares Chedid, Monsy Sam, Dr. Christoph Fink, Abdul Ahad Khan

Innovation in surgery:
In a life saving complex and rare heart surgery a two-day old baby girl with a congenital heart defect that would have required a heart transplant, underwent a complicated procedure in Al Jalila Children's hospital. The team of surgeons were able to save her heart and also take her off the ventilator right after the surgery and establish her independent breathing. The surgery allowed the patient to have a much better chance of recovering from her serious condition, this is the first step of many to come on her way to full recovery.

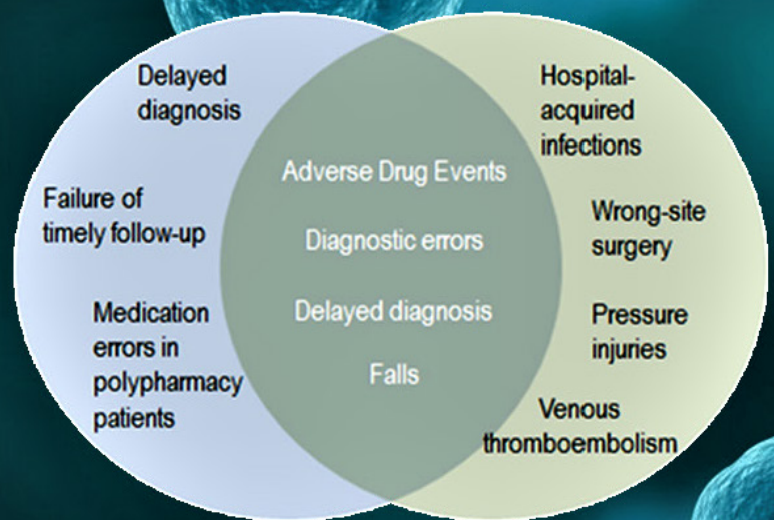


Improve patient safety by eliminating adverse events in health care settings

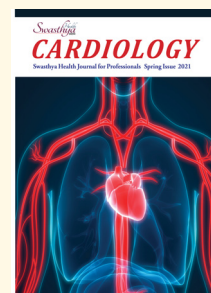
It is estimated that every year more than 300,000 patients acquire a healthcare associated infection (HCAI, HAI or nosocomial infection) as a result of care with in the NHS.

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