

New surgery for better outcomes: shaping the field of congenital heart disease

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Background: The field of congenital heart disease is constantly evolving through better understanding of the disease itself, albeit its history, prevalence, genetics, and follow-up. Concurrently surgical techniques and approaches have been developed, modified, and refined.

Data sources: The impact of interventional pediatric cardiology has been dramatic. The field of catheter-based therapies has exploded with the first pulmonary valve balloon angioplasty in 1982. With evolving stent technology, they are now used in multiple areas, including pulmonary arteries, vena cavae, aortic and arch and descending aorta for coarctation. The hybrid surgery concept involves a multidisciplinary team of interventional cardiologist and surgeon combining catheter-intervention and surgery in the surgical theater such as pulmonary artery stent implantation associated with pulmonary valve replacement. Furthermore, in selected cases, pulmonary valve device implantation is becoming an accepted approach to a surgical problem.

Results: Balloon angioplasties, stent implantations, hybrid surgeries and pulmonary valve device implantation are performed with a very low mortality and morbidity. The risks and benefits outweigh the ones associated to surgical procedures.

Conclusion: With fast developing interventional therapies, the work of pediatric cardiologists and cardiac surgeons is more intertwined than ever in search of better outcomes for the children with congenital heart disease.

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Introduction

The surgical practice of pediatric congenital heart disease is evolving. Technological advances are exerting a major impact on new less invasive surgical approaches. A multidisciplinary team work provides a more comprehensive surgical approach producing better outcomes with less risk to the patients. Basic science research is also progressing. The role of tissue engineering with the use of biological scaffolds to create valves and other tissue has great promise in developing new tissue and organs.

This paper reviews some of the technical advances in pediatric congenital heart disease, particularly hybrid surgery, catheter intervention, and implantable valve device.

Background

On May 6, 2003 we celebrated the 50th anniversary of the first successful open heart operation performed with the use of a heart-lung machine, pioneered by John H. Gibbon, as well as the first open heart surgery using the technique of cross circulation pioneered by C. Walton Lillehei.^[1,2] It was the beginning of many rich dramas that over the years changed and remarkably improved the outcomes of children with congenital heart disease. Concurrently evolution in pediatric cardiology led to many accomplishments including echocardiography, cardiac catheterization and angiography, pharmacological manipulation, and catheter intervention with creating an atrial septostomy for cyanotic babies by W. J. Raskind in 1966 and with balloon pulmonary valve dilatation in 1982, leading to an explosion of catheter-based therapy.^[3,4] The concept of hybrid surgery was just around the corner.

Hybrid surgery

Hybrid surgery involves the new concept of multidisciplinary surgery, whereby not only a surgeon is performing the surgery, but where a surgeon and an interventional pediatric cardiologist team up toward safer and better outcomes.^[5] Interventional pediatric cardiologists have been performing catheter interventions since the early 1980s, but have now brought their

expertise to the operating room to assist the surgeon.

Many children have early cardiac repairs, but must undergo subsequent operations to replace obstructed valves, and/or to repair stenosed pulmonary arteries. Such repairs are often complicated by difficult dissection of scar tissue around the pulmonary arteries. Under direct vision when the main pulmonary artery is opened, stents can be implanted in either or both pulmonary arteries, and dilated appropriately (Fig. 1).^[6] Our experience at BC Children's Hospital includes 28 patients with a mean age of 134 months at operation. The patients had a minimum of two previous operations, and one previous pulmonary valve implantation. The average stent size was 15 mm, and pulmonary valve size was 22 mm. The hospital stay was 7 days with no mortality. We encountered one complication; one stent was not fully deployed at surgery (Fig. 2), and will require further balloon angioplasty.

Hybrid surgery has become a new application in neonatal correction of hypoplastic left heart syndrome. In 1983, Norwood and colleagues^[7] published the first successful repair of hypoplastic left heart syndrome (Fig. 3). Over the last 20 years many refinements of the Norwood technique came to place with a decreased mortality from 40% to around 10%. As the greater risk of the Norwood operation comes from such extensive surgery in the newborn period, search was still on to improve outcomes. A clever hybrid approach was developed by no other than Norwood. It involves banding each pulmonary artery to limit pulmonary blood flow with a pre-measured band of 3.5 mm circumference and insertion of a 6 mm stent in the ductus arteriosus to provide blood supply to the upper and lower body (Fig. 4). This procedure is done in the catheterization laboratory without the use of cardiopulmonary bypass, therefore decreasing the time of postoperative ventilatory and cardiac support, and the risk of cerebral damage. The Norwood procedure is then postponed until 4-6 months of age. Although still requiring refinements, it may become accepted as the first stage repair of hypoplastic left heart syndrome.

Catheter intervention

The field of catheter intervention exploded following successful coronary angioplasty by Gruntzig in 1979, and the modern era of pulmonary valve balloon angioplasty in 1982.^[4,8] The stent technology has greatly evolved with a new delivery system, different stent design, and self expandable stent. These stents are used in multiple areas including pulmonary artery stenoses, superior and/or inferior vena cavae stenoses, aortic arch and descending aorta stenosis post coarctation repair, and carotid and renal arteries.

As an example of the application of stent

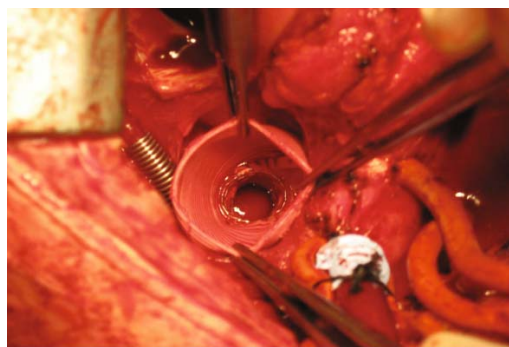


Fig. 1. A graft has been sewn to the distal main pulmonary artery and a metal stent is seen in the orifice of the left pulmonary artery.

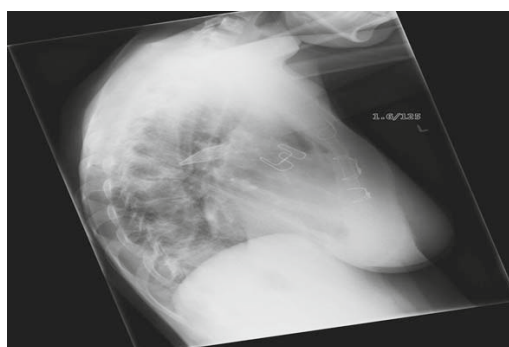


Fig. 2. Lateral chest X-ray showing a stent improperly deployed in the left pulmonary artery. The metal frame of the pulmonary valve bioprosthesis is seen.

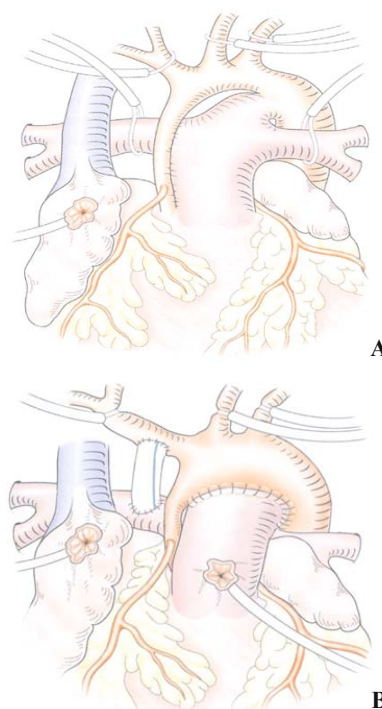


Fig. 3. Graphic representation of a Norwood surgical procedure. **A:** The main pulmonary artery is transected. **B:** The hypoplastic ascending and transverse aorta is opened; a neo-aorta is reconstructed, including the pulmonary valve as a new aortic valve, and a right Blalock Taussig shunt is constructed.

technology, this 32-year-old patient with a diagnosis of ventricular septal defect, pulmonary atresia and multiple aortopulmonary collateral arteries (Fig. 5) presented for increasing cyanosis and decreased exercise tolerance. She had never had surgery. Facing the prospect of multiple surgeries, she was palliated by inserting self-expandable stents in two collateral arteries to the left lung (Fig. 6) increasing her oxygen saturation to 88% and improving her exercise tolerance.

Repair of coarctation of the aorta is done in the neonatal period and in infancy. The surgical results are excellent, but some patients will develop re-coarctation or arch hypoplasia as they grow.^[9,10] As illustrated in Fig. 7, the top panel shows an area of re-coarctation dilated and stented successfully. The lower panel depicts an area of arch hypoplasia enlarged by a stent. Both patients avoided complex re-operations.

Because many infants undergo primary cardiac repair early in life, a certain number of them will require re-operation such as valve conduit replacement and Fontan completion. These re-operations are complicated by scar tissue formation and difficult dissection. Complications may occur during surgery and rescue with catheter intervention may be life-saving. Two children during a complex re-operation had injury to the coronary artery ostium. Despite surgical repair their coronary artery injuries were rescued by stent implantation (Fig. 8), keeping the right coronary in one child and the left in the other child, opened and patent.

Valve device implantation

The current generation of children with repaired congenital heart disease are exposed to increasing re-interventions, particularly those with right ventricular outflow tract problems, albeit obstruction and/or insufficiency. The timing of conduit replacement or pulmonary valve implantation is determined by right ventricular pressure and function, by dysrhythmia, and by the patient's exercise limitation. Currently, no "ideal" conduit or valve exists for reconstruction of the right ventricular outflow tract. All are susceptible to degeneration and loss of function, submitting the patient to multiple interventions.

Percutaneous pulmonary valve implantation was originally developed by Bonhoeffer et al.^[11] The implantable device consists of a bovine jugular vein with its valve stitched into a platinum-iridium stent. The valved stent is then crimped down and loaded on a delivery system, molding it around a balloon for deployment and dilatation (Fig. 9). The access to the right heart is through the femoral vein and under X-ray guidance. The loading delivery system including the valve is positioned appropriately in the right ventricular outflow tract, deployed and inflated fully with the inner

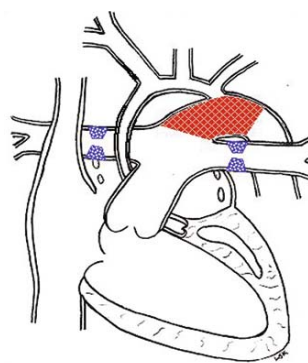


Fig. 4. Hybrid surgery for hypoplastic left heart syndrome. In red, stent in the ductus arteriosus, and in blue, pulmonary artery banding of the left and right pulmonary arteries.

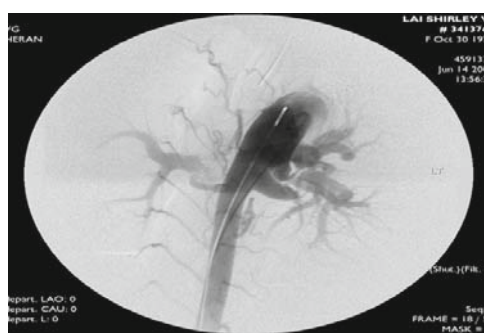


Fig. 5. Aortogram showing multiple aortopulmonary collateral arteries (2 on the left and 1 on the right).

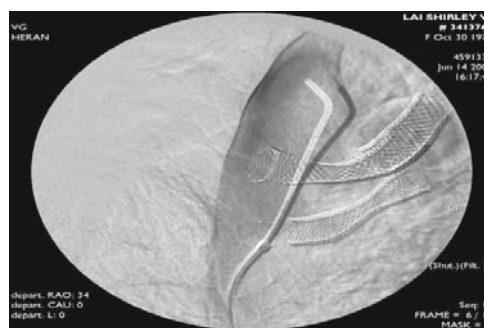


Fig. 6. Self-expandable stents in the aortopulmonary collateral arteries to the left lung.

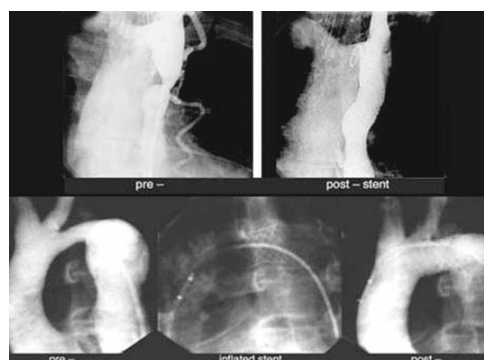


Fig. 7. The upper panel shows a residual coarctation pre- and post-stent placement. The lower panel shows an aortic arch hypoplasia pre- and post-stent placement.

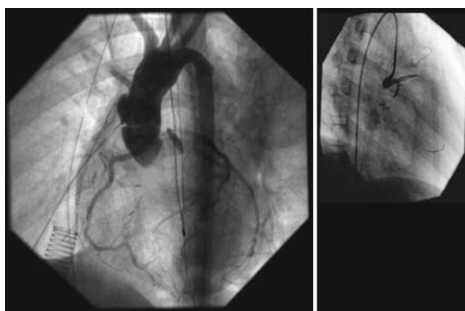


Fig. 8. The left panel shows the patent proximal right coronary stent. The right panel shows the patent proximal left main coronary artery through a selective injection in the left main orifice.



Fig. 9. Representation of a pulmonary valve device sewn inside a stent and mounted inside a delivery catheter before delivery and after full dilatation.

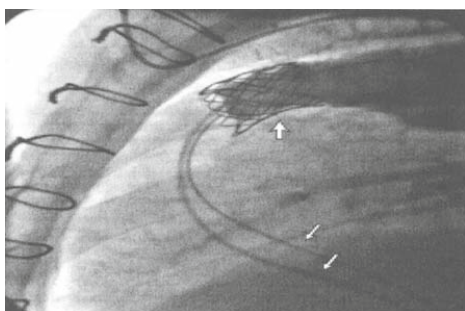


Fig. 10. Pulmonary cine angiogram following percutaneous pulmonary valve implantation. The stented valve device is shown with a large arrow. The 2 small arrows show the 2 delivery catheters through the right ventricle.

balloon. Angiography shows proper position and function of the implantable valve (Fig. 10).

The maximum available implantable valve is 22 mm. The indications are limited, but expanding with experience. The mid-term and long-term results are still missing, but current information from several series is promising.^[12] This technology of implantable pulmonary valve device will reduce the number of interventions required in children with conduit obstruction or pulmonary insufficiency. In fact, current work involves placing an implantable pulmonary valve device inside a previously surgically implanted conduit that has become stenotic. This approach increases the hope for teenagers and young adults that their conduits may not need

lifelong replacement, but will be kept functioning by inserting a pulmonary valve device whenever required.

Summary

These are examples of congenital heart disease, which illustrate our evolution in knowledge and therapy. The paradigm from bed-to-bench is well demonstrated by those ongoing ideas and innovations about better outcomes, safe surgical approaches, hybrid or not, and ultimately improved quality of life.

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References

- Cohn LH. Fifty years of open-heart surgery. *Circulation* 2003;107:2168-2170.
- Lillihei CW, Cohen M, Warden HE, Varco RL. The direct-vision intracardiac correction of congenital anomalies by controlled cross circulation; results in thirty-two patients with ventricular septal defects, tetralogy of Fallot, and atrioventricularis communis defects. *Surgery* 1955;38:11-29.
- Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy. A palliative approach to complete transposition of the great arteries. *JAMA* 1966;196:991-992.
- Kan JS, White RI Jr, Mitchell SE, Gardner TJ. Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary-valve stenosis. *N Engl J Med* 1982;307:540-542.
- Bacha EA, Hijazi ZM, Cao QL, Abdulla R, Starr JP, Quinones J, et al. Hybrid pediatric cardiac surgery. *Pediatr Cardiol* 2005;26:315-322.
- Bökenkamp R, Blom NA, De Wolf D, Francois K, Ottenkamp J, Hazekamp MG. Intraoperative stenting of pulmonary arteries. *Eur J Cardiothorac Surg* 2005;27:544-547.
- Norwood WI, Lang P, Hansen DD. Physiologic repair of aortic atresia-hypoplastic left heart syndrome. *N Engl J Med* 1983;308:23-26.
- Grüntzig A. Clinical experience with coronary intra-arterial dilatation using catheters. Results (author's transl). *Ann Cardiol Angeiol (Paris)* 1979;28:487-489.
- Slack MC. The role of stenting in coarctation of the aorta. *Prog Pediatr Cardiol* 2001;14:45-57.
- Ward KE. Complications of balloon coarctation angioplasty. *Prog Pediatr Cardiol* 2001;14:59-71.
- Bonhoeffer P, Boudjemline Y, Qureshi SA, Le Bidois J, Iserin L, Acar P, et al. Percutaneous insertion of the pulmonary valve. *J Am Coll Cardiol* 2002;39:1664-1669.
- Coats L, Tsang V, Khambadkone S, van Doorn C, Cullen S, Deanfield J, et al. The potential impact of percutaneous pulmonary valve stent implantation on right ventricular outflow tract re-intervention. *Eur J Cardiothorac Surg* 2005;27:536-543.

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