Advances in congenital heart surgery

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Purpose of review
We provide an overview of the past year's literature on congenital heart surgery.
Recent findings
This review focuses on selected disease entities, operative techniques, perioperative management strategies, and quality of care.
Summary
Congenital heart surgery is an evolving field.

Keywords
cardiac surgical procedures, congenital, heart defects, review

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Abbreviations

ASD	atrial septal defect
BTS	Blalock–Taussig shunt
IVS	intact ventricular septum
PFO	patent foramen ovale
RV-PA	right ventricular to pulmonary artery
VSD	ventricular septal defect

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Introduction

While congenital heart surgery continues to evolve and outcomes improve, little of our practice is evidence-based. The broad range of procedures performed and the relative infrequency of these procedures make conducting appropriately powered randomized controlled trials and large, timely cohort studies difficult. In addition, many important outcomes of congenital heart surgery are not manifest for many years necessitating costly, labor-intensive follow-up to understand the true impact of our work. As a result, most of the literature about congenital heart surgery consists of small series and retrospective reviews. Despite these shortcomings, much can be learned from the literature that exists. We have reviewed and summarized a selection of interesting contributions over the past year. Some of these articles may influence a change in current practice; others may serve as inspiration for more rigorous investigations.

The Norwood procedure

Management of patients who undergo the Norwood procedure continues to evolve. Postoperative stability, survival, and neurologic outcome for survivors remain major issues. Multiple authors reported strategies aimed at improving these outcomes.

Several authors investigated the use of right ventricular to pulmonary artery (RV-PA) conduits as an alternative to traditional systemic-to-pulmonary shunts. The suspected advantage of this technique is the elimination of diastolic runoff into the pulmonary circulation with concurrent unloading of the systemic right ventricle. Myocardial perfusion could improve by the higher diastolic pressure, lower aortic saturation, and decreased work volume.

Sano *et al.* [1••] reported a retrospective multi-institutional study describing the use of a nonvalved polytetrafluoroethylene shunt between a small right ventriculotomy and the distal stump of the main pulmonary artery as an alternative to a systemic-to-pulmonary shunt. Seventy-three infants at three centers received 4–6 mm RV-PA shunts as part of a modified Norwood procedure. Hospital survival for the cohort was 84% and actuarial survival at 2 years was 63%. In a small retrospective series ($n = 21$), Azakie *et al.* [2] reported lower heart rates 1 hour postoperatively and higher diastolic blood pressures 24 hours postoperatively in patients who underwent RV-PA conduits. Pizarro *et al.* [3•] compared 6-month mortality in 96 consecutive hospital survivors after stage 1 Norwood surgery. The source of pulmonary blood flow in 46 patients was a modified Blalock–Taussig shunt (BTS) and in the remaining 50 was a RV-PA conduit. Forty of 46 patients

(87%) with a BTS and 49 of 50 patients (98%) with a RV-PA conduit reached hemi-Fontan stage.

Questions remain about the long-term impact of a right ventriculotomy in a univentricular heart. Tanoue *et al.* [4] described mid-term follow-up of 21 patients who underwent either systemic-to-pulmonary artery shunt ($n = 11$) or RV-PA conduit ($n = 10$). Ventricular efficiency was comparable between the groups; however, after bidirectional Glenn procedure and total cavopulmonary connection, contractility in patients who had undergone a RV-PA conduit was inferior.

Other authors examined alternative strategies aimed at achieving the same results. Nakano *et al.* [5] reported a more stable postoperative course after Norwood with use of a low-resistance strategy. Intraoperative high flow and low-resistance cardiopulmonary bypass was accomplished with the use of chlorpromazine and the avoidance of circulatory arrest. In weaning from bypass, pulmonary vascular resistance was maximally decreased by using 100% oxygen, inhaled nitric oxide, and nitroglycerin. Pulmonary blood flow was determined by adjusting the systemic-to-pulmonary shunt. Postoperatively, chlorpromazine and nitroglycerin were continued while the pulmonary-to-systemic blood flow ratio was adjusted by regulating the inspired oxygen fraction and dose of inhaled nitric oxide.

Ungerleider *et al.* [6•] reported a different approach to achieving postoperative stability. Eighteen consecutive patients were placed on ventricular assist devices (VAD) in the operating room after modified ultrafiltration. VAD flows were maintained at 150–200 mL/kg/min and no attempt was made to balance the systemic and pulmonary circulations. The ventilator was adjusted to achieve a systemic PaO_2 of 30–45 mmHg and $PaCO_2$ of 35–45 mmHg. If evidence of hypoperfusion occurred, as measured by increasing lactic acid, VAD flows were increased. Average time of VAD support was 3.1 days and hospital survival was 89%. In addition, neurodevelopmental testing at 4–6 months was normal and improved compared with other reported neurologic outcomes after the Norwood procedure.

Bradley *et al.* [7] challenged the theory that the fraction of inspired oxygen should be minimized and ventilation should be tightly controlled in the immediate postoperative period. They reported that high levels of inspired oxygen improved mixed venous oxygen saturation and system oxygen delivery in a group of 14 neonates who underwent the Norwood procedure. In addition, hyperventilation did not change systemic or mixed venous saturation, arteriovenous saturation difference, oxygen excess factor, or blood pressure.

Cerebral blood flow

Neurologic outcomes are a key measure of success of congenital heart surgery. More work is needed to link elements of preoperative, intraoperative, and postoperative care to neurologic outcome. As part of these efforts, baseline neurologic function must be understood. Licht *et al.* [8] used pulsed arterial spin-label magnetic resonance imaging to show that preoperative cerebral blood flow was diminished in patients with a variety of congenital heart defects. Low cerebral blood flow values were associated with periventricular leukomalacia. Further work in this area may eventually help guide strategies to improve neurologic outcomes.

Complete atrioventricular septal defect and Down's syndrome

Formigari *et al.* [9] reviewed a two-institution retrospective series of 206 consecutive patients who underwent repair of complete atrioventricular septal defects to determine the impact of Down's syndrome on morbidity and mortality. They postulated that Down's syndrome patients despite, having higher pulmonary vascular resistance, abnormal airways, and abnormal lung parenchyma and thus requiring special care, would not have higher operative risk compared with patients with normal chromosomes. One hundred thirty-one patients (64%) had cytogenetic confirmation of Down's syndrome. The overall mortality rate was 7.7% (6% among Down's patients and 14% among non-Down's patients) and the only independent risk factor affecting survival was the presence of unbalanced ventricles. The need for the Norwood procedure was more frequent among non-Down's patients, as was the prevalence of pulmonary artery banding. Freedom from reoperation after successful biventricular repair or definitive univentricular palliation was lower in the group with normal karyotype because of the higher prevalence of anomalies of the mitral valve or left ventricular outflow tract. With improved process of care, Down's syndrome may no longer increase the risk of biventricular repair, and it should be reevaluated as a risk factor for surgical morbidity and mortality.

Valvular heart disease

Hraska *et al.* [10•] in a two-institution study reviewed the mid-term results of 66 patients who underwent aortic root replacement with pulmonary autograft. Twenty of these patients had left ventricular outflow tract obstruction defect requiring a Ross–Konno procedure. There were no early deaths, but one patient died at 3 months from bacterial endocarditis. Survival after a mean period of 2.4 years was 98.5%. The incidence of neo-aortic insufficiency was minimal: 3% mild, 53% trivial, and 44% none. There was a significant reduction in left ventricular diastolic diameter index and left ventricular mass index within 3–12 months after operation. Autograft replacement of the aortic root is an excellent choice for severe anomaly of the aortic valve with or without left ventricular outflow tract obstruction. Slater *et al.* [11] have suggested an interesting modification to the Ross procedure that may limit the likelihood of long-term autograft dilatation. Further follow

up of these patients will be needed to demonstrate the benefit of this modification.

Tetralogy of Fallot with absent pulmonary valve syndrome

Zucker *et al*. [12] presented the clinical presentation, natural history, and outcome of patients with Tetralogy of Fallot with absent pulmonary valve. In a retrospective review of 465 patients with a diagnosis of Tetralogy of Fallot or absent pulmonary valve syndrome, 18 consecutive patients had the combined syndrome. The overall mortality rate was 28% (5/18). Four patients with ventricular septal defect (VSD) died: two patients without surgery and two patients postoperatively due to sepsis and respiratory failure. One patient with intact ventricular septum (IVS) died at age of life of 1 hour. Surgical repair was performed in 73% of patients with VSD and 28% with IVS. The authors postulate that there are two populations of patients within this syndrome: those with ventricular septal defects and those with intact ventricular septa. Patients with VSD have a significant family history of congenital heart disease, and progress to develop the characteristic symptoms of Tetralogy of Fallot. These patients develop respiratory symptoms during infancy because of left-to-right shunting and airway obstruction. They are often cyanotic independent of respiratory function given high pulmonary vascular resistance in the presence of an incompetent pulmonary outflow tract and a VSD. In contrast, patients with IVS are often asymptomatic, present later in life, and have a relatively benign prognosis. Diagnosis was delayed in 43% of these patients. Symptomatic patients with IVS often have a patent ductus arteriosus or atrial septal defect leading to a similar left-to-right physiology. These two patient populations may be represent separate disease entities given their variable presentation and prognosis.

Surgical versus percutaneous repair of aortic coarctation

Walhout *et al*. [13•] compared traditional surgical repair to endovascular balloon angioplasty for treatment of coarctation of the aorta. A cohort of 46 patients (age >3 months) with native coarctation of the aorta was treated; 18 underwent standard surgical repair and 28 underwent balloon angioplasty. The patients were followed for up to 10 years (surgical group mean follow up 7.2 years, angioplasty group mean follow up 5.4 years). There was a significant age difference between the groups (mean surgical age: 0.63, mean angioplasty age 5.8, $P = 0.001$). Immediately after treatment, there was no significant difference between the groups in the decrease in pressure gradient across the coarctation. Hospital stay ranged from 6–20 days in the surgical group as compared with 48 hours in the angioplasty group. There were no mortalities. Anastomotic recoarctation occurred in one surgically treated patient; it was successfully treated with balloon angioplasty. Recoarctation occurred in two patients treated initially with balloon angioplasty and was treated with operative resection and end-to-end anastomosis. For appropriately selected older children with aortic coarctation, angioplasty compares favorably with surgical repair and should be the preferred treatment.

Surgical versus percutaneous repair of atrial septal defects

Bialkowski *et al*. [14] compared surgical to endovascular treatment of secundum ASD. In a series of 91 children found to have secundum ASD, 44 were treated with standard surgical repair and 47 with a percutaneous Amplatzer septal occluder. Records were reviewed for durability of closure and complication rates. Complications were classified as mild, moderate, or severe. All patients had significant hemodynamic lesions with a pulmonary-to-systemic flow rate of greater than 1.5:1 and right atrial and right ventricular dilation. Characteristics of the ASD that determined suitability for endovascular repair were the presence of an adequate rim of septum around the defect and a diameter that was not excessively large. Four patients were ineligible for interventional treatment because of ASD size. Surgery was performed in 41 patients who were not candidates for transcatheter closure and in three children whose parents preferred surgical repair. The mean ASD diameter was larger in the surgical group (mean 16 versus 10.8 mm, $P < 0.001$). Surgical repair was performed primarily in 38 patients and with pericardial patch in six patients. Mild complications were small pericardial effusions, headaches, first degree atrioventricular block, and rhythm disturbances. Moderate complications included pneumonia, atelectasis, paroxysmal atrial tachycardia, and atrioventricular junctional rhythm. Severe complications included hemorrhage requiring reoperation and transient neurologic events. No complications occurred in 31.8% of surgical patients and 93.8% of Amplatzer device patients. Mild complications occurred in 38.6% of surgical patients and 4.3% of endovascular device patients. Moderate complications occurred in 25% of surgical patients and 2.1% of endovascular patients. Severe complications arose in 4.5% of surgical patients and 0% of septal occluder patients. Overall, 68.2% of surgically closed ASD patients developed complications as compared with 6.4% of patients treated with the Amplatzer occluder device ($P < 0.05$). There were 95.5% of surgical patients and 97.9% of device-treated patients, who had complete closure of their ASD at discharge. In children with favorable anatomy, percutaneous ASD repair is preferred over surgical closure because of the shorter length of stay and lower rate of complications.

Percutaneous device closure of patent foramen ovale

El Said *et al*. [15] evaluated the effect of patent foramen ovale (PFO) and ASD morphology on the efficacy of

transcatheter closure. Of 47 patients who underwent percutaneous closure of a PFO, the morphology of the septum was flat in 33 and aneurysmal in 14. The PFO morphology was a simple flap in 20 patients and complex in 27 patients. Complex morphologies included a long tunnel in 15 patients, a coexistent small ASD in 5 patients, and an aneurysmal septum without a tunnel or ASD in 7 patients. Device placement was successful in all patients. In five patients with a long tunnel PFO, a trans-septal puncture of the foraminal flap was necessary. Effective closure on follow-up was seen in 95% of patients. In the remaining two patients, one had a simple and the other a complex PFO. At a median follow-up of 22 months, there were two patients with recurrence of neurologic events; however, neither patient had evidence of right-to-left shunting by transesophageal echocardiogram (TEE). Both were receiving antiplatelet medication at the time of recurrence. Variations in atrial septal and PFO morphology are frequently encountered; however, their presence does not preclude a successful outcome.

Robotic pediatric cardiac surgery

Suematsu and del Nido [16] compiled a review of current applications for robotically assisted pediatric cardiac surgery. Regarding extracardiac lesions, they used the Intuitive Surgical da Vinci System for the division of vascular rings in two patients. Length of stay was shorter than with standard thoracotomy; however, the total operative times were longer (180 and 165 minutes) because of robot arm placement and positioning of the surgical cart. Dissection of the aorta, subclavian artery and the ductus were safely performed using the EndoWrist instruments, which made tissue handling and dissection easy and accurate. Regarding intracardiac lesions, they used the da Vinci system for ASD repair in seven adult cases using the Heart Port-Access System for cardiopulmonary bypass. Operative times were longer than with conventional repair; however, the procedures were completed endoscopically and no perioperative complications were observed.

There remain limitations to robotic surgery for pediatric cardiac operations. Small patient size may make robotic surgery more difficult in complex operations. Five millimeter and smaller three-dimensional endoscopes have been developed; however, their use has been limited. Also, determination of optimal port placement is a significant issue as poor placement can lead to instrument conflicts and frequent unnecessary incisions. The absence of tactile feedback and the inability to regulate the force applied to the tissues also limit utility.

Current robotic technology has facilitated the use of thoracoscopic procedures for extracardiac lesions in the pediatric population. With technologic improvement, intracardiac robotic surgery may be one day possible. New technologies including real-time three-dimensional echocardiography may allow image guided surgical tasks using echocardiographic guidance alone. When evaluating this new technology, the advantages of minimally invasive surgery must always be weighed against the safety of traditional open surgical approaches.

Remote pump head system

Ando *et al.* [17••] described open-heart surgery in children without homologous blood transfusion using a miniaturized cardiopulmonary bypass circuit with a remote pump head. The small circulating blood volume of an infant or small child makes hemodilution inevitable when using standard cardiopulmonary bypass circuits unless homologous blood is transfused. This practice is not without risk as immune reaction to blood transfusion can lead to significant morbidity. A retrospective series of 158 patients weighing less than 5 kg undergoing ventricular septal defect repair were reviewed. Patients with ASD, patent ductus arteriosus, and mild right ventricular outflow tract obstruction that could be repaired through a similar cardiac incision were included. They were able to successfully commence cardiopulmonary bypass with crystalloid prime in 122 of 158 patients. The main reasons for blood prime in the other patients included low predicted bypass hematocrit or low body weight ($n = 20$), cardiac or respiratory failure or both ($n = 14$), or the presence of pulmonary vascular obstructive disease ($n = 2$). The success rate of completely bloodless surgery among 122 patients was 95.1%. One patient died on postoperative day 1 from acute pulmonary hypertensive crisis that developed after extubation; this patient had a thickened pulmonary artery media of about 33% of the entire wall thickness that should have been a contraindication for surgery. Four patients had ventricular dysfunction resulting from the placement of large ventricular septal patches. No neurologic deficits including seizure activity, delirium or delusion, or significant motor dysfunction were found by cardiologists or parents after operation. No mortality, reintervention, or neurologic sequelae occurred. Neurologic testing was performed in 43 patients (inclusion criteria were an interval of >6 months after operation, age of testing between 1–3 years, and no known chromosomal abnormality). Thirty-four patients completed the questionnaire. Four patients were found to have a borderline low result and these patients had a preoperative history of prolonged mechanical ventilation, and two had mild mental retardation that physicians had diagnosed prior to surgery. A low-developmental score was not associated with decreased hematocrit during surgery. A miniaturized cardiopulmonary bypass circuit may reduce the use of blood products and the surface contact of circulating blood and as a result reduce the overall inflammatory response.

Quality of care

There is widespread recognition that cardiothoracic surgery outcomes differ by surgeon and institution. Public

and private payers are using this variation to direct patients to high-performing providers. Patients and their families are becoming informed decision makers and are searching the Internet and media sources when choosing a healthcare provider. Despite this trend, the definition and measurement of quality in pediatric cardiac surgery remain nebulous. Kang *et al*. [18••] identified age at surgery, bypass time, and Risk Adjustment in Congenital Heart Surgery (RACHS-1) category as independent risk factors for mortality in a cohort of 1085 consecutive open-heart cases performed on patients less than 18 years of age. A risk model using these variables to discriminate between survival and mortality had good predictive value with an area under the receiver operating characteristic curve of 0.86. Checchia *et al*. [19] showed that institution volume might be a predictor of survival after Norwood procedure. Among a group of 29 hospitals from the Pediatric Health Information System database, those that performed more than one Norwood procedure per month (n = 4) had a survival rate of 78%; those performing less than one procedure per month had a survival rate of 59%. This finding is consistent with many other surgical procedures; however, it does not shed light on the system elements and process measures for which volume is a surrogate. Their finding that surgeon operative volume was not associated with mortality is expected. Care of the patient undergoing the Norwood procedure is complex and requires a dedicated, cohesive, communicating team of professionals with a variety of skills. Investigations into the function of these teams are likely to yield the true elements necessary for successful outcome after congenital heart surgery.

Conclusion

Congenital heart surgery continues to evolve and outcomes improve. Future investigations should emphasize quality study design and the linking of physiologic parameters to neurologic and functional outcomes. This will result in changes to our practice that will affect the outcomes important to our patients and their families.

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