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Selected 2017 Highlights in Congenital Cardiac Anesthesia

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This is the author's manuscript of the article published in final edited form as:
Nasr, V. G., Twite, M. D., Walker, S. G., Kussman, B. D., Motta, P., Mittnacht, A. J. C., & Mossad, E. (2018). Selected 2017 Highlights in Congenital Cardiac Anesthesia. Journal of Cardiothoracic and Vascular Anesthesia. https://doi.org/10.1053/j.jvca.2018.03.027

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Disclosures: None

Funding: Intramural Departmental Funding

Key words: congenital heart disease; pediatric, pediatric cardiac anesthesia, cerebral oximetry, cardiac catheterization, ventricular assist device

Introduction:

This article is a review of the highlights of pertinent literature published during the 12 months of 2017, which is of interest to the congenital cardiac anesthesiologist. Following a search of the US National Library of Medicine PubMed database, several topics emerged where significant contributions were made in 2017, and that the authors of this manuscript felt were noteworthy to be summarized in this review: Training in pediatric cardiac anesthesia, the costs of congenital heart disease (CHD), catheter versus surgical intervention for CHD, cerebral oxygen saturation in CHD, and mechanical circulatory support in children.

Training in pediatric cardiac anesthesia

In 2004, the Accreditation Council for Graduate Medical Education (ACGME) Next Accreditation system was implemented with the creation of milestones to all specialties.(1) Since 2007, for surgeons who operate on patients with CHD ranging from neonates to adults, a subspecialty certification in congenital

cardiac surgery is granted following completion of a one-year accredited residency and passing of a written examination and an oral examination.(2) The number of accredited surgical programs in the US has increased from 2 in 2006 to 12, and the number of graduates has increased from 2 in 2008 to 44 in 2014.(3,4) Along with the increase in structured training for congenital cardiac surgery, there has been an increase in the need of physicians with advanced knowledge and experience in pediatric cardiac intensive care. Several informal advanced training pathways have been suggested following surgical, anesthesia or intensive care trainings and were discussed at the 10th International Conference of the Pediatric Cardiac Intensive Care Society.(5) Similarly to surgical and intensive care specialties, pediatric cardiac anesthesiology has evolved as a subspecialty devoted to patients with CHD ranging from neonates to adults. While there is a consensus on the need to define standards for the pediatric cardiac anesthesia fellowship, there remains a wide variability in duration of training and clinical experience. Different training pathways have been proposed in the past few years and several recent publications are highlighted to outline the proposed future pathways.

In Germany, a working group representing the German Society for Anesthesiology and Intensive Care surveyed all pediatric cardiac centers with the intent to define "expertise in pediatric cardiac anesthesia".(6) Twenty-seven centers with an active pediatric cardiac surgery program participated in the survey with a response rate of 96.3%. The suggested length of training in pediatric cardiac anesthesia was 12-months in 42.3% of centers with a mean of 10.8-months to achieve sufficient experience. However, the length of training may not be reflective of the competence of the trainee.(7) Because, different learners with different knowledge or experience backgrounds progress at different rates, the time needed by a trainee to achieve competence may vary.(8,9) Recently, based on the ACGME Milestones Model and recognizing that the duration of training does not reflect competency, an expert panel of the Congenital Cardiac Anesthesia Society, a section of the Society of Pediatric Anesthesia defined 18-milestones as competency-based developmental outcomes for the pediatric cardiac anesthesia fellowship.(10) These 18 milestones cover all six-core competencies of the ACGME including patient

care, medical knowledge, systems-based practice, practice based learning and improvement, professionalism, and interpersonal and communication skills. They are listed in details in Table 1. In addition, recognizing the increase in adult patients with CHD and adult congenital heart centers as described by Kogon et al., these milestones address competency in anesthesiology for patients with CHD across the age spectrum from neonates to adults.(11) Pediatric cardiac anesthesiology is defined in the Consensus Statement as a subspecialty "devoted to caring for individuals with congenital heart disease ranging in age from neonates to adults." (10) Currently there are 17 programs in the US that offer 24 positions for pediatric cardiac anesthesia fellowship.(12) Each of these programs as well as international programs can use the recently described milestones as a template to define the objectives of the fellowship and develop a milestone-based curriculum.

The Cost of CHD

Adults living with CHD now outnumber children with CHD. In both children and adults, hospital admissions and associated costs may be the result of their primary CHD, or associated cardiac and non-cardiac comorbidities. Frequently, repair of a specific CHD requires multiple admissions (i.e. staged single ventricle palliation), or revisions later in life. Readmission rate after surgery is high, further increasing healthcare resource use and costs. It is obvious that the healthcare environment is undergoing major changes. Payers are increasingly reluctant to reimburse for quantity, and metrics of efficiency and quality are being established as the basis of reimbursement. It is therefore increasingly important to understand what drives costs in CHD. In 2017 several noteworthy contributions were made that help practitioners and administrators alike, to better understand the costs associated with CHD.

For example, Mackie and colleagues conducted a retrospective cohort study using the Canadian Institute for Health Information Discharge Abstract Database between 2004 and 2014.(13) Among 59,917 hospitalizations that included CHD as the primary or related diagnosis they found a 21.6% increase in

costs over the ten-year study period. Costs were higher for children compared with adults; however, the cost increase was greater in adults (4.5%/year) than in children (0.7%/year). Costs increased the most among adults with complex CHD (7.2%/year). The study investigators conclude that although children still account for greater inpatient CHD costs, adults with CHD are increasingly contributing to the overall cost burden of CHD on healthcare resources. Although the length of stay over the observed time period was unchanged in this study, it is without question that length of hospital stay is a major driver of inpatient hospital costs. Cedars and colleagues analyzed multiple State Inpatient Databases identifying 99,103 inpatient hospitalizations in adults with CHD.(14) They found that overall the variables most strongly associated with increased length of stay, regardless of underlying CHD anatomy, were acute renal failure, bacterial infection, complications from medical or surgical procedures and anemia. These potentially modifiable risk factors could be used to design targeted interventions to decrease length of stay and potentially cost in adult CHD inpatient care.

While many adult CHD hospitalizations are due to cardiac complications, there are a substantial proportion of admissions due to non-cardiac morbidities that are increasing as the population of adults with CHD ages. Seckeler and colleagues studied the cost of hospitalization for non-cardiac disease for adults with CHD.(15) They conducted a retrospective review of hospital discharge data from the University Health System Consortium Clinical Database from 2011 to 2013. They compared 9,169,700 adults without CHD to adults with differing severities of CHD. This study identified significantly increased hospitalization costs for adults with moderate and complex CHD who are admitted for non-cardiac medical conditions. Admissions with CHD diagnoses had higher ICU admission rates, longer lengths of stay, and higher mortality for most non-cardiac admission diagnoses. The study investigators point out that the creation of adult CHD centers of excellence in Quebec, Canada are associated with a decrease in all-cause mortality and cost in this patient population.(16) There may be a role for regionalization of adult CHD care in the USA as well, allowing highly specialized providers to focus on the general health and non-cardiac comorbidities that patients will experience as they age.

The presence of non-cardiac comorbidities is not unique to adult patients. Tuomela and colleagues studied non-cardiac comorbidities in infants with CHD and the impact on resource utilization.(17) They found non-cardiac comorbidities occurred in 55% of infants. Most commonly observed were gastrointestinal (the need for enteral tube feeds, malrotation), respiratory (vocal cord dysfunction, airway malacia) and genetic diseases (Trisomy 21 and DiGeorge syndrome). The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery (STAT) score and the presence of multiple comorbidities were associated with higher resource utilization following the index cardiac surgical procedure.

Although the association between genetic abnormalities and CHD is well known, its combined impact on resource use and cost has not been evaluated. Furlong-Dillard and colleagues studied the resource use and morbidities in infants with CHD and genetic conditions.(18) Using the Pediatric Health Information System Database, they identified 95,253 children under the age of 18 years undergoing congenital heart surgery. Of these, 84.6% had no genetic condition, trisomy 21 (9.9%), trisomy 13 or 18 (0.2%), DiGeorge syndrome (0.8%), Turner syndrome (0.4%) and 'other' genetic conditions (4.2%). Although complications varied by genetic condition, all groups with genetic abnormalities experienced increased length of stay and cost. Identifying genetic abnormalities in patients with CHD may thus, be helpful in deciding which children might benefit from a cardiac complex care program that partners families and providers to improve health and decrease healthcare costs. As an example Geddes and colleagues showed that implementing a standardized protocol with the help of a geneticist, increased the rate of diagnosis of genetic conditions and reduced cost per diagnosis significantly.(19)

Another important factor driving costs in CHD is hospital readmission rate, which is increasingly becoming a metric for quality in the current landscape of cost-effective medicine. Payers are viewing readmissions as preventable, often denying reimbursement for these hospitalizations. However, it is clear that infants with CHD are often readmitted to the hospital for both further planned procedures and unplanned reasons. In an Australian study the rate of cardiac surgical procedures in the first year of life was steady at 2.5 children per 1000 live births.(20) Excluding infants who had ligation of a patent ductus

arteriosus, 50% required readmission in the first year of life and over 20% of these infants had an additional non-cardiac congenital anomaly. Similarly, Sacks et al found a 20.5% readmission rate for infants with CHD not requiring surgery in a USA based study of 30-day readmission rates.(21) Patient factors associated with an increased likelihood of 30-day readmission were younger age, lower discharge weight and greater number of diagnoses and a longer length of initial stay. These results indicate that many common factors associated with readmissions are not easily modifiable. However, factors associated with readmissions may serve as important prognostic indicators, and a basis for counseling and potential cost savings.

Single ventricle pathology is perhaps the most complex form of CHD and increasing numbers of children with this condition now survive to adulthood. The single ventricle palliation pathway is associated with significant resource use and costs, as can be seen in a recent study by Huang and colleagues who studied 156 patients from the Fontan Registry Database in Australia and New Zealand, from birth to Fontan completion.(22) During the staged surgical treatment period, children had a mean 10 +/- 6 inpatient admissions and spent 85 +/- 64 days in hospital. Although cardiac surgeries were the major reason for resource use (77% of the total cost), non-cardiac admission costs were 18% of the total costs. Costs were higher in male and HLHS patients in the staged procedures period. Interestingly, over 40% of the pediatric hospital costs for Fontan patients occurred after the last planned surgery.(23)

Aside from a change in demographics (i.e. increasing number of adults with CHD), associated comorbidities and frequent readmissions, all contributing to the significant costs associated with CHD, care for CHD patients by itself is often more costly compared to other cardiac surgery populations. In a study using the 2013 Nationwide Inpatient Sample Database, Nasr and colleagues compared costs and outcomes for adult patients undergoing surgery for treatment of CHD to a reference population of adults undergoing coronary artery bypass graft (CABG).(24) This study showed that adult CHD patients undergoing cardiac surgery experience higher hospital costs and poorer outcomes than the reference population of adult CABG patients. Variation in the clinical management of patients with CHD may be

partially responsible. For example the off-label use of iNO is common in children undergoing cardiac surgery but the efficacy is unclear and the cost substantial. Gupta and colleagues studied the use of iNO after pediatric cardiac surgery, among centers of varying surgical volume.(25) Using the Pediatric Health Information System database, Gupta and colleagues identified 103,714 children from 41 hospitals of which 15.1% received iNO after cardiac surgery. They found higher iNO utilization among centers with lower surgical volume which also had worse outcomes (including mortality) compared to centers with higher surgical volume. There was wide variation in iNO utilization among centers and only 25% patients receiving iNO had an associated diagnosis of pulmonary hypertension. This may indicate that current indications for the use of iNO are poorly defined and/or poorly implemented and its arbitrary use significantly increases post-operative costs. There is clearly an opportunity to introduce a standardized protocol for the post-operative use of iNO to help guide clinical decisions and reduce costs. Rathod and colleagues undertook an excellent study in which they implement Standardized Clinical Assessment and Management Plans (SCAMPs) in children after the arterial switch operation (ASO).(26) Patient management that included SCAMPs resulted in significantly lower resource use and overall costs, and no difference in clinical outcomes between the historical and SCAMP cohorts. This study indicates that SCAMPs may be an excellent way forward for many institutions to improve resource utilization and reduce costs not only in ASO patients, while maintaining quality of care.

Catheter versus surgical intervention for CHD

In 1953 Rubio-Alvarez and colleagues reported the first successful cardiac catheter-based interventional technique for treatment of a congenital cardiac defect when they described cutting across a stenotic pulmonary valve using a “bow string” apparatus deployed at the catheter tip.(27) Little notice was taken, and it was more than a decade later that Rashkind and colleagues reported their results using balloon atrial septostomy to allow mixing of oxygenated and deoxygenated blood in cyanotic defects.(28) This was to become the first widely-used non-surgical technique for palliation of CHD, and it is still in use today.

Since that time, catheter-based interventions for treatment of CHD have become increasingly sophisticated and wide-spread. Indeed, with the increased utility of echocardiography and cardiac MRI as diagnostic techniques, the congenital cardiac catheterization laboratory has become more of a treatment center and less of a strictly diagnostic tool.

Current catheter-based interventional techniques are progressively expanding. Theoretical advantages of catheter-based interventional techniques over surgery are numerous and include absence of a surgical incision, avoidance of the effects of cardiopulmonary bypass, reduced infection rate, less pain, shorter procedure time, faster recovery time, and lower cost. At the same time, catheter-based interventions are not without risk. The Congenital Cardiac Interventional Study Consortium recently developed the Catheterization RISk Score for Pediatrics (CRISP) based on pre-intervention variables. The authors identified 41 reported significant adverse events (SAEs) in their study population involving 14,790 catheter-based interventional procedures. Many of these SAEs were specifically related to the interventional procedure, such as device migration, stent malposition, and wire retention.(29) Despite the expanding use of catheter-based interventional techniques in the management of CHD, important questions remain regarding their advantages over conventional surgery.

Several studies in 2017 analyzed the outcomes of both interventional and surgical treatment of various congenital heart lesions. Glatz and colleagues looked at the use of PDA stents versus modified Blalock-Taussig shunt (BTS) in patients with ductal-dependent pulmonary blood flow.(30) This was a multicenter comparison study by the Congenital Catheterization Research Collaborative, which includes investigators from four institutions. Patients were treated with either BTS or catheter-based PDA stent placement in all centers. There was no statistically significant difference in the composite primary outcome of death or re-intervention to treat cyanosis between the two groups, and the PDA stent group compared favorably in several secondary outcomes. For example, the PDA stent group had a lower ICU length of stay (5.3 vs. 9.19 days), was less likely to be on diuretics at discharge, and had fewer procedural complications (when adjusted for patient factors). When the PDA stent group returned later for either follow-up or definitive

surgical repair, they were found to have larger and more symmetric pulmonary arteries (PAs). This was postulated to be the result of a shunt source that was (1) central (rather than to a branch pulmonary artery), and (2) not associated with anastomotic vessel distortion (which is often seen with BTS). Perhaps as a result of this, the PDA stent group also tended to present for definitive surgery at an older age and after a longer interval since initial palliation (stent placement or BTS). However, the BTS group was no more likely to require pulmonary arterioplasty at the time of definitive surgical repair, nor did they require more subsequent PA interventions. In contrast, the PDA stent group did require more overall re-interventions, such as re-dilation of the stent by balloon angioplasty or pulmonary valvuloplasty. This likely reflects the unique nature of interventional stent placement and the opportunity to re-intervene through catheter-based techniques. Although catheter-based interventions present unique challenges, this first-of-its-kind large multicenter comparison indicates that PDA stent placement is a viable option versus BTS for patients with ductal-dependent lesions.

One lesion that has historically been palliated with BTS is Tetralogy of Fallot (TOF) associated with early severe cyanosis and/or ductal dependence. The Glatz study did not include TOF among the anatomic diagnoses of its subjects, perhaps because several major centers have moved away from this strategy, instead favoring either early complete repair or catheter-based palliation. An example is the Hospital for Sick Children in Toronto, where Wilder and colleagues recently reported the outcomes from their institution using either early (<3 months of age) primary repair (the EARLY group) or catheter-based palliation (the CATH group). Interventions used in the CATH group included right ventricular outflow tract stent, ductal stent, and right ventricular outflow tract balloon angioplasty. Patients who did not require early intervention but instead underwent elective repair after 3 months of age (the IDEAL group) served as a control group.(31) All three strategies (EARLY, CATH, and IDEAL) were associated with a low mortality rate, with 1 death in each group. The re-intervention rate was higher in the CATH group compared to the others, and the risk of reoperation was higher in the EARLY group. However, long-term outcomes including survival, somatic growth, right ventricular systolic pressure, branch pulmonary artery

dimensions, and prevalence of pulmonary insufficiency were not significantly different among groups at 8-10 year follow up. One limitation of this study is the selection bias inherent in a single-institution analysis – patients in the CATH group were more likely to be selected for catheter-based intervention if they were deemed less likely to tolerate early complete surgical repair based on systemic disease. Risk-adjustment was used to limit the impact of such patient-specific differences, but even if this was flawed by the relatively low patient numbers in the study the bias would tend to be unfavorable toward the outcomes in the CATH group. Despite this, the CATH group compared favorably to both the EARLY and IDEAL groups in the long term.

Recurrent coarctation of the aorta is a common problem after the Norwood operation for palliation of single ventricle lesions. Treatment options include surgical re-operation, balloon angioplasty, or catheter-based stent implantation at the coarctation site. Redo surgery is associated with good long-term results, but it entails the risks associated with repeat sternotomy and cardiopulmonary bypass. While balloon angioplasty has been commonly employed, it may not be effective for all lesions and is associated with a fairly high re-stenosis/re-intervention rate.(32) Theoretically, placing a stent at the coarctation site should provide a longer-term solution compared to ballooning alone. This was studied by Aldoss and colleagues, who reviewed outcomes for stent implantation for recurrent coarctation in the Norwood population in 33 patients at 8 centers.(33) The authors report excellent immediate results of both improvement of the aortic diameter and reduction in the coarctation pressure gradient with stenting. Twelve patients (36%) had SAEs. No patients died or required mechanical support. It was noted that a prograde catheter approach (percutaneous venous access with catheter course across the tricuspid and neo-aortic valves) was more commonly associated with hemodynamic compromise than a retrograde or other approach, likely related to iatrogenic insufficiency of one or both valves. Despite the good early results, over half the patients did require re-intervention to balloon the stent. Half of these were simply the result of somatic growth, but half were due to in-stent stenosis, residual arch obstruction, or stent fracture. The authors speculate that

future stents may be bioabsorbable, creating an option that would less likely require re-intervention, as the stent would absorb quickly enough to allow somatic vessel growth.

Another lesion that has long been managed with catheter-based intervention is persistent PDA. Traditionally, catheter-based intervention using coils or occluders has been undertaken mostly in children and adults, with limited application in infants < 1 year of age. This leaves open questions about the feasibility, safety and efficacy of this technique compared to surgical closure in infants. Backes and colleagues conducted a meta-analysis of the existing literature to investigate the technical success and safety of percutaneous PDA closure during infancy.(34) Thirty-eight studies representing 635 infants were included. Technical success, defined as the infant leaving the cath lab with a coil or device in the PDA, was high (92.2%). Procedural abandonment occurred in 40 patients, most commonly related to either device malposition or an adverse event (AE). If residual ductal shunting was observed following the procedure, it usually resolved within 24 hours, with only 8 cases reported as remaining patent at longest follow-up (3-36 months). The SAE rate was about 10%, with major or catastrophic events occurring in 1.6%. Over 92% of major or catastrophic events occurred among infants <6 kg. At least one death was reported; this was related to a cardiac perforation in a 1.5 kg premature infant. Publication bias, measured by the LFK index, was considered "major" (LFK index exceeds ±2) in many of the articles reviewed, indicating a tendency to report only clinically significant findings. This, and the void of randomized controlled trials investigating this technique, makes it difficult to draw conclusions about the superiority of this technique to surgery in the neonatal population. However, the preponderance of the available evidence would indicate that percutaneous PDA closure in the infant population is feasible and relatively safe, with few major or catastrophic adverse events (especially among infants >6 kg).

Cerebral oxygen saturation in CHD.

Near infrared spectroscopy (NIRS) is a non-invasive optical technology for measurement of tissue oxygenation. Commercialization of NIRS led to the development of tissue oximeters, which allow for continuous bedside monitoring of cerebral (ScO_2) and somatic (StO_2) tissue oxygen saturation.(35) The premise for cerebral oximetry is the identification of cerebral hypoxia, or hyperoxia (especially in premature neonates) to help guide therapy and improve clinical outcomes.(36, 37)

Treatment algorithms have been devised to guide practitioners in treating possible causes of cerebral desaturation or hyperoxia.(38) Interventions are triggered either by trend monitoring and deviation from baseline values, or absolute lower and upper thresholds beyond which the possibility for cerebral injury may increase. Establishing a normal range for ScO_2 in both pediatric and adult populations is therefore clinically important, but is beset with problems (beyond the scope of this article). In healthy children the normal range for ScO_2 is a mean (±SD) of 68% ± 10%.(39) However, what constitutes normal values in the diverse group of CHD patients has mostly been speculative and based on small population samples.(40) Because the difference (delta) between SaO_2 and ScO_2 in normal patients is ~30%, it is assumed that in cyanotic children the normal ScO_2 ranges from 40% to 60%.(41) However, in children with CHD, practitioners frequently observe a wide range of ScO_2 values which are attributed to the features of the presenting CHD.

In 2017, several noteworthy contributions in the field of NIRS monitoring were made, thereby enhancing our understanding of the use and interpretation of ScO_2 values specifically as it relates to neonates and infants with CHD. Kussman et al. evaluated ScO_2 in cyanotic ($SaO_2 < 90\%$) and acyanotic ($SaO_2 \geq 90\%$) children with CHD undergoing cardiac catheterization.(42) Although the mean (±SD) ScO_2 was statistically different between the acyanotic (74% ± 6%) and cyanotic (69% ± 6%) patients, the values in the cyanotic patients were within the range considered normal. The authors also estimated the cerebral oxygen extraction (COE) using the O_2 content difference between the arterial and jugular venous bulb blood. In their study the authors found no difference in COE between cyanotic and acyanotic patients

(P=.10). With cerebral oximetry monitoring the COE can be estimated noninvasively at the bedside by calculating the arterial-cerebral O_2 *saturation* difference (SaO_2-ScO_2), or fractional O_2 extraction (FOE = $[SaO_2-ScO_2]/SaO_2$). Using the cerebral oximetry-based method the COE was significantly different between the two diagnostic groups (P< .001). The implications of this study are (1) a low baseline ScO_2 value in the setting of chronic hypoxemia should not merely be attributed to a lower SaO_2; (2) an increased or increasing difference between the SaO_2 and ScO_2 in a cyanotic patient who is anemic or bleeding could lead to ScO_2 levels associated with brain injury in laboratory studies (43); and (3) the parameters used for estimation of COE at the bedside can be misleading.

These findings have important implications when interpreting cerebral oximetry values in children with CHD. For example, it has been suggested that a mixed venous oxygen saturation (SvO_2) <30% represents the anaerobic threshold in neonates after Stage I palliation for hypoplastic left heart syndrome (HLHS).(44) Goal-directed therapy with SvO_2 as an indicator of systemic oxygen delivery has been associated with excellent early survival and a low incidence of organ failure.(45) However, because SvO_2 monitoring is invasive, non-invasive ScO_2 and StO_2 monitoring have been suggested as substitutes for SvO_2. This substitution is not without pitfalls and limitations. Published studies have generally shown a statistically significant but modest correlation between ScO_2 and SvO_2, but with wide limits of agreement.(46, 47) In postoperative pediatric cardiac surgical patients, the two variables were not interchangeable and ScO_2 changed in the same direction as the SvO_2 only 64% of the time.(48)

In a recent study building on this concept, Rescoe et al. investigated the sensitivity of ScO_2 to screen for compromised tissue oxygen delivery (DO_2) defined as a SvO_2 <30% in neonates following stage 1 palliation for HLHS.(49) The main finding of this study was an insensitivity of cerebral oximetry in detecting a SvO_2 <30% in the ScO_2 range of clinical interest (i.e. ScO_2 30-50%). When SvO_2 was <30%, cerebral ScO_2 was <30% in less than 1%, <40% in less than 1%, and <50% in 45.7% of paired ScO_2 and SvO_2 cooximetry measurements. Put another way, 54.3% of the data points with a ScO_2 >50% were associated with a SvO_2 <30% (Figure 1). This is consistent with the physiology of preservation of CBF with decreasing cardiac output.

Aside from studies furthering our understanding of ScO_2interpretation, technological advances in NIRS-based tissue oxygenation monitoring are being investigated. Commercial oximeters use continuous wave (CW) spatially-resolved NIRS to measure blood hemoglobin-oxygen saturation in the tissue beneath the sensor. Because ScO_2 is not a direct measure of cerebral oxygen delivery or consumption ($CMRO_2$), inferences need to be made regarding cerebral oxygen balance. Frequency-domain near infrared spectroscopy (FD-NIRS) and diffuse correlation spectroscopy (DCS) are advanced optical technologies which allow for quantitative measurement of absolute cerebral oxygen saturation, oxyhemoglobin (HbO_2), deoxyhemoglobin (HbR), total hemoglobin (HbT) from which cerebral blood volume (CBV) is derived, cerebral blood flow (CBF), and a calculated estimate of $CMRO_2$.(50)

Ferradal et al. demonstrated the feasibility of a hybrid FD-NIRS/DCS system for intraoperative assessment of cerebral blood flow and oxygen metabolism in neonatal cardiac surgery.(51) Cerebral blood flow and $CMRO_2$ were quantified before, during, and after deep hypothermic cardiopulmonary bypass (CPB) in 9 neonates. The findings of a decrease in CBF and $CMRO_2$ with a change in coupling during deep hypothermia are consistent with the invasive techniques in the pioneering studies by Greeley et al.(52, 53) This study supports the concept of FD-NIRS/DCS to monitor cerebral physiology and provide the potential for individual optimization of surgical management.

Additionally, over the past decade there has been increasing recognition of the contribution of genetics, placental dysfunction, and direct trophic influences of abnormal cerebral perfusion and oxygenation to the adverse neurocognitive outcomes in children with CHD.(54, 55) Brain abnormalities before surgery in neonates with severe CHD include smaller global and regional cerebral volumes, delayed maturation (by about one month), white matter injury (periventricular leukomalacia), and abnormal metabolism. The study by Kelly et al. published in 2017 adds to the growing body of evidence that abnormal circulatory patterns result in decreased cerebral DO_2 and impaired brain growth.(56) In a comparison between 30 newborns with mixed CHD before surgery and 30 age-matched healthy controls, cerebral DO_2 was linearly related to cortical grey matter volume and gyrification index, with the greatest impairments associated with those lesions with the lowest cerebral DO_2. These findings are in line with the seminal

study by Limperopoulos et al. showing that the percentage of combined ventricular output through the aortic valve was independently associated with lower total brain volume in fetuses with CHD.(57)

Mechanical Circulatory Support in Children:

The prevalence of heart failure in children is steadily increasing, with a rate of 14.5-17/100,000 heart failure related admissions annually.(58) The most common comorbidities in these children are dilated cardiomyopathy, CHD , arrhythmia-induced heart failure and myocarditis. With aggressive medical care, the hospital length of stay for these patients (median of 19.5 days/admission) and mortality have continued to decrease.(59) However, there remains a subset of children with end-stage heart failure refractory to medical care. In those children, the expanded use of temporary mechanical circulatory support (MCS) (< 2 weeks) and long-term support with Ventricular Assist Devices (VAD) (>2 weeks of support) significantly decreased mortality and other outcome measures. The highlights and updates of short- and long-term MCS in 2017, especially issues pertinent to the practice of pediatric cardiac anesthesia are reviewed.

The Second Annual Pediatric Interagency Registry for Mechanical Circulatory Support (PediMacs) report reviewed the data on VAD implants in the period 2012-2016, showing a rise in the number of centers as well as device options, with more than 430 devices implanted in 364 patients during the study period.(60) Cardiomyopathy is the most common indication for VAD placement (60% of patients) followed by CHD (21%) in which the majority are single-ventricle (SV) patients (48/77). The most common age of children supported on VAD was the 11-18 year old (48% of patients), with neonates and infants forming 20% of the population. VAD implantation in CHD is more challenging than other in other indications due to anatomical abnormalities and communications and previous heart surgeries. The VAD use and duration of support as a bridge-to-transplant (BTT) increases in the adolescents. The most common support is L-VAD (80%), Bi-VAD 15% and Total Artificial Heart (Syncardia TAH, LLC, Tucson, AZ) (2%).

Complications such as bleeding, infection and stroke are common especially with the paracorporeal pulsatile devices (Berlin Excor, Berlin Heart AG, Berlin, Germany) significantly more than with the implantable continuous flow devices including the HeartWare HVAD (HeartWare Inc., Framingham, MA) and the HeartMate II (St Jude Medical Inc., St Paul, MN). The majority of complications occur within 3 month of implantation. Mortality is higher in younger and smaller patients possibly due to the more frequent CHD indications and the limited availability or increased size discrepancy with continuous flow devices.(60)

Temporary Mechanical Circulatory Support: The Role of Impella® in Pediatrics.

Cardiogenic shock remains a significant cause of morbidity and mortality in children with acute heart failure refractory to maximized medical treatment (e.g. high dose inotropes and mechanical ventilation). Temporary support has been available with extracorporeal membrane oxygenators (ECMO), and with percutaneous continuous flow devices including the Tandem Heart (CardiacAssist Technologies, Pittsburgh, PA) and most recently with Impella device (Abiomed Inc., Danvers, MA). Parekh and colleagues reported in a retrospective review the outcomes, implant techniques, complications, and hemodynamic data of 10 Impella® insertions at single pediatric heart center.(61) The diagnosis included 5 post-transplant rejection or allograft vasculopathy patients, 2 with myocarditis, and one patient with refractory ventricular tachycardia. Impella® was used as a solo support in 6 children and in 4 was used in addition to ECMO for left heart decompression. The device size (Impella 2.5/5.0 and CP) was chosen based on patient weight, vessel size and level of support required. Echocardiography was used to optimize device positioning. Impella® support was associated with reduction in pulmonary capillary wedge pressures ($p < 0.039$) and improved end-organ perfusion evidenced by increases in the cerebral and somatic near infrared spectroscopy ($p=0.039$). Complications were reported in 8/10 implantations most importantly device purge failure, insertion site bleeding and limb ischemia. All patients survived to discharge from the intensive care unit with one late death. A similar multicenter study of Impella device support of the systemic circulation in pediatric patients reported a median support duration of 45 hours,

survival of 85% at 7 days and 68% at 30 days following support. Most children experienced recovery of ventricular function with the temporary Impella support (41% of patients), or transition to a long term MCS (30%). Clearly, the use of these devices will increase both for procedural support in the catheterization laboratory as well as for temporary support to recovery or bridge to long-term devices.(62)

The support of a failing Fontan circulation is challenging. Morray et al published a multicenter, retrospective study of the Impella® to support the systemic ventricle in a cohort of Fontan patients with either ventricular failure (N=8) or high-risk electrophysiology (EP) procedures (N=2).(63) The median duration of support was 49 hr. (2.7–264 hr.) with a hospital survival of 80% (7 weaned off, 2 transitioned to long-term device, and 1death). Adverse events occurred in 4 patients (hemolysis, aortic valve insufficiency with implant and thrombus). These emerging evidence suggest that Impella® can provide temporary support in pediatric patients with cardiogenic shock including SV patients as a bridge to recovery or long-term support. Complications rate is high but might be due to the initial experience with this device in pediatrics.

Compared to a propensity-score matched cohort of children supported with ECMO as a bridge to transplant, patients with temporary MCS had a longer support duration (median 19 days vs 6 days, $p<0.001$) and longer overall survival before and after transplant.(64)

Long Term Devices: Advances in Pediatric Ventricular Devices.

Pediatric VADs implantation is following the adult trend of increased use of continuous-flow devices. These devices offer design simplicity, low complication rate and durability. This presents a paradigm shift in the type of devices used, the patient selection and the expected outcomes. In a review of the most important changes in VAD support in children, Adachi and colleagues highlights the findings form the most recent PediMacs report.(65) He outlines the emerging evidence of better outcomes with continuous flow implantable devices. He highlights the concern that continuous flow VADs were designed for adults and older children. However, its use in smaller children (< 25 kg) results in patient-device size mismatch

worsening the outcome. The future of continuous VAD support is in miniaturized devices including the axial-flow implantable infant Jarvik 2015 VAD (Jarvik Heart, Inc., New York, NY) which is in its final stages of experimental trials prior to human implants.(66)

One innovative approach is to "inactivate" the listing for transplant for at least 3 months after the continuous VAD insertion in children (grace period) with the rationale of improving the patient´s physiological status and increases suitability for transplantation and/or potentially recovery.(65)

In a 10 year review (2005-2015) of the influence of transplant center procedural volume on survival outcome of heart transplantation for children bridged with MCS, Hsieh et al showed that patients supported with ECMO until transplant have inferior survivals compared to those bridged and supported with VAD regardless of center volumes.(67) They demonstrated the significant advantage of long term MCS and argue for a reconsideration of the process of higher listing for children on ECMO support, where a bridge to VAD support for a period of time may be the better approach for improved transplant outcomes.

With the increasing number of children on temporary or long-term MCS, the American Heart Association recently published the guidelines for cardiopulmonary resuscitation for these patients while on support. Peberdy et al review the challenges of diagnosis of flow and perfusion impairment in these patients especially in continuous flow devices, where the standard monitoring and examination become difficult to interpret.(68) The statement recommends to assess perfusion using standard clinical exam to rule out non-VAD causes of arrest (e.g. hypoxia, hypoglycemia, drug overdose or stroke). Next is to investigate system alarms and the VAD hum to evaluate VAD function. Para-corporeal devices such as Berlin Heart EXCOR should be evaluated visually looking at diaphragm and valve opening/close. If the VAD is not functioning the device should be restarted after inspection of the power source and driveline. If the circulatory failure is non attributable to device failure, chest compressions should be started. Even though there is a risk of VAD dislodging with chest compression, the benefits outweigh the risk. The exception is with total artificial heart (TAH) patients, since chest compression, epinephrine and anti-arrhythmic drugs add no

benefit and may be detrimental (Table 2). Finally the scientific statement highlights the value of early rescue echocardiography for the evaluation of VAD performance. Echocardiography can easily evaluate cardiac filling, right ventricular function, major valve dysfunction, left ventricle (LV) unloading, VAD cannula obstruction and aortic valve opening in systole.

As the outcome of children with temporary and long-term MCS continues to improve, these patients will present to the anesthesiologist for non-cardiac surgical interventions as well as other procedures requiring various levels of sedation. Guidelines for the care of these patients will be necessary, as demonstrated by a report of 52 children on VAD support presenting for general surgery interventions. Anticoagulation status was not a factor in the success of the procedures. The perioperative management of these patients required knowledge and understanding of the device settings and a multidisciplinary team approach for an uneventful course.(69)

Figure Legend

Figure 1:

Raw data for rSO_2 and $ScvO_2$ with crude linear regression line.

Among all $ScvO_2$ <30, there were no rSO_2 values <30.

Modified from Rescoe et al. [49]

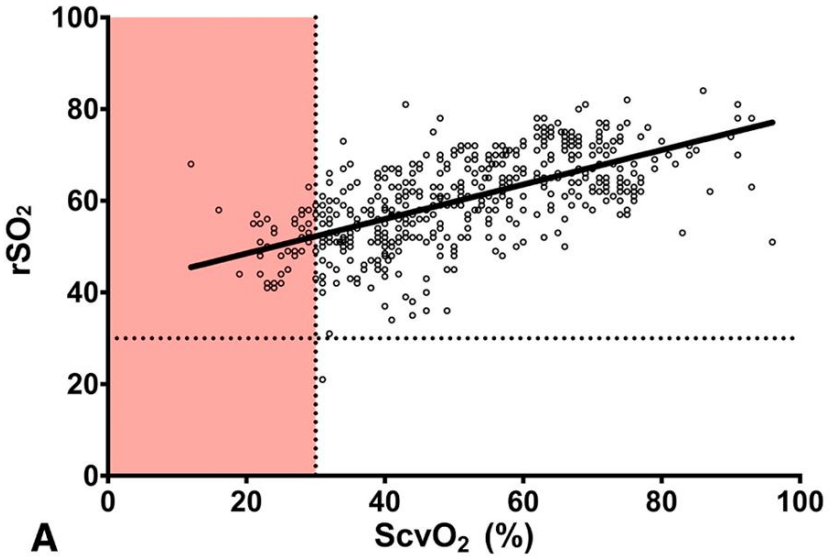

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Table 1:

Core Competencies and Milestones for the Pediatric Cardiac Anesthesia Fellowship. Modified from Nasr VG et al.[10]

Competency	Milestones
Patient Care (4)	• Perioperative assessment, planning and management • Technical/Procedural skills • Understanding cardiovascular surgical procedures • Understanding cardiac catheter-based interventional procedures and electrophysiologic studies
Medical knowledge (4)	• Congenital and acquired cardiovascular anatomy, physiology and pathophysiology • Pharmacology • Cardiopulmonary bypass, extracorporeal circulation, and mechanical assist device principles • Understanding cardiac diagnostic procedures
Systems-based practice (3)	• Coordination of care • Incorporation of patient safety and quality improvement into clinical practice • Understanding of health care economics; cost awareness and cost-benefit analysis
Practice-based learning and	• Self-directed learning and scholarly activity

improvement (2)	• Education of pediatric cardiac care team members and other health care providers
Professionalism (3)	• Commitment to institution, department and colleagues • Receiving and giving feedback • Personal responsibility to maintain emotional, physical and mental health
Interpersonal and Communication skills (2)	• Communication with patients and families • Interprofessional communication and transitions of care

Table 2

Cardiopulmonary resuscitation in unresponsive children with ventricular assist devices.

Perfusion & Capillary refill	LVAD	Echocardiography	Management
Good	No alarms- VAD hum present	Good LV unloading & RV function No valve insufficiency	Treat non-LVAD causes of altered mental status • Hypoxia • Hypoglycemia • Drug overdose • Stroke
Poor	Alarms present- VAD hum absent	Poor LV unloading RV dysfunction, Major valve insufficiency	1. Check: • Driveline • Power source 2. Restart VAD: • If perfusion improves => continue with ACLS protocol • No improvement in perfusion, MAP < 50 mmHg and/or $EtCO_2$ > 20 mmHg => start chest compressions*

Abbreviations: LVAD, left ventricular assist device; LV, left ventricle; RV, right ventricle; ACLS, advanced cardiac life support; MAP, mean arterial pressure; $EtCO_2$ end tidal CO_2

*TAH patient should not receive chest compression, epinephrine and/or anti-arrhythmic drugs