SCAI/CCAS/SPA Expert Consensus Statement for Anesthesia and Sedation Practice: Recommendations for Patients Undergoing Diagnostic and Therapeutic Procedures in the Pediatric and Congenital Cardiac Catheterization Laboratory

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Introduction

Over the last several decades, procedures performed in the pediatric and congenital cardiac catheterization laboratory (PCCCL) for children and adults with congenital heart disease (CHD) or children with acquired heart disease have transitioned from being primarily diagnostic to interventional and therapeutic. However, with the advanced therapeutic options for patients with CHD comes the increased risk of adverse events. Adverse events related to pediatric cardiac catheterization have been reported in 4%–10% of procedures. Although most adverse events are not associated with mortality, they can occur suddenly and unexpectedly [1–5]. Along with the increase in patient and procedure complexity, there has been a more detailed delineation of sedation policies both at the institutional and the national level (e.g., the American Academy of Pediatrics (AAP) and The Joint Commission). It is therefore imperative that personnel with the skill sets necessary to anticipate, prevent, and treat complications related to the catheterization procedure and sedation/anesthesia be in attendance.

Because the current practice of sedation and anesthesia for patients undergoing PCCCL procedures is known to vary among institutions, a multi-society expert panel with representatives from the Congenital Heart Disease Council of the Society for Cardiovascular Angiography and Interventions (SCAI), the Society for Pediatric Anesthesia (SPA) and the Congenital Cardiac Anesthesia Society (CCAS) was convened to evaluate the types of sedation and personnel necessary for procedures performed in the PCCCL. The goal of this panel was to provide practitioners and institutions performing these procedures with guidance consistent with national standards and to provide clinicians and institutions with consensus-based recommendations and the supporting references to encourage their application in quality improvement programs. This task is difficult because the patients cared for vary widely in age, ranging from premature infants to adults, from simple to complex in physiology, and ranging in cooperation ability: from those who cannot cooperate or are combative and might need general anesthesia to those who are fully cooperative and might need little or no sedation. Recommendations can neither encompass all clinical circumstances nor replace the judgment of individual clinicians in the management of each patient. The science of medicine is rooted in evidence, and the art of medicine is based on the application of this evidence to the individual patient. This expert consensus statement has adhered to these principles for optimal management of patients requiring sedation and anesthesia. What follows are recommendations for patient monitoring in the PCCCL regardless of whether minimal or no sedation is being used or general anesthesia is being provided by an anesthesiologist.

Complications and Risks

Factors contributing to adverse events in the PCCCL are multi-factorial in origin and can be patient, practitioner and/or procedure-related. Although it might not be possible to attribute an event to a specific cause, we will discuss procedural risks based on an overview of the current literature.

Anesthesia Risks in Patients with Cardiac Disease Undergoing Surgery and Procedures in the PCCCL

Studies concerning anesthesia-related morbidity and mortality in recent decades have demonstrated that pediatric patients and especially patients with CHD are at increased risk for adverse events and cardiac arrest during surgery [1–3]. Common complications in children undergoing sedation or general anesthesia include airway events (laryngospasm, bronchospasm, apnea and aspiration), cardiovascular events

(hypotension, arrhythmias and cardiac arrest) and postoperative issues such as nausea and vomiting, emergence agitation, hypoxemia and apnea. In a prospective quality assurance audit of 24,165 anesthetics in children undergoing surgery, Murat et al. showed that respiratory issues comprised 53% and cardiac issues 12.5% of intraoperative adverse events [4]. Respiratory events were more common in infants under one year of age, intubated patients and those who were American Society of Anesthesiologists Physical Status (ASA PS) 3 or 4. Cardiac events were also most common in those with ASA PS 3 or 4. Similarly, in 2006, Braz et al. studied 15,253 anesthetics in children and found the risk factors for adverse events to be an age <1 year, an ASA PS of 3 or 4, emergency procedures and preoperative intubation [5]. Patients with CHD undergoing cardiac catheterization typically have a higher ASA physical status and therefore are at increased risk even before the catheterization procedure has started.

Flick et al. found that in 92,881 anesthetics the cardiac arrest rate was 2.9/10,000 in children undergoing noncardiac surgery and 127/10,000 in cardiac surgical procedures, with a mortality rate of 1.6/10,000 [3]. Eighty-eight percent of those who experienced a cardiac arrest had CHD. The rate of cardiac arrest was highest in neonates undergoing cardiac surgery at 435/10,000 and mortality at 389/10,000.

Vitiello et al. found that patient age and interventional catheterization procedures were risk factors for morbidity and mortality in the PCCCL [6]. The specific risk factors for anesthesia and sedation were young age, low weight and need for intubation. Bennet et al. examined adverse events in the cardiac catheterization lab specifically from an anesthetic perspective and found in 4454 catheterizations an adverse event rate of 9.3% for diagnostic procedures and 11.6% for interventional procedures [7]. There were 90 incidents; 33 were respiratory, of which 20 were airway events, and 22 were cardiovascular events, of which 17 were transient arrhythmias. The mortality rate (4 deaths) was 0.08%. All the deaths were in patients under 18 months of age. Adverse events occurred most frequently in patients <1 year of age and in those having interventional procedures other than persistent ductus arteriosus (PDA) and atrial septal defect (ASD) closure. These rates of adverse events are similar to those published in the IMPACT (Improving Pediatric and Adult Congenital Treatment) Registry in nearly 20,000 patients, with adverse events occurring in 10% of diagnostic and 11.1% of interventional procedures [8].

The Pediatric Perioperative Cardiac Arrest (POCA) Registry collected data on 373 anesthesia-related cardiac arrests in children, 34% of whom had congenital or acquired heart disease [2]. Of the patients with heart disease, anesthesia-related cardiac arrests occurred 54% of the time in the general pediatric OR, 26% in the cardiac OR and 17% in the PCCCL. Fifty-nine percent of patients with uncorrected and 26% with palliated single-ventricle physiology had the highest risk of cardiac arrest, whereas patients with aortic stenosis or cardiomyopathy had the highest risk of mortality following a cardiac arrest, at 62% and 50%, respectively. These lesions accounted for more than 75% of all deaths reported to the POCA registry. Nearly half (47%) of cardiac arrests in children with heart disease occurred in those younger than 6 months of age.

Specific Cardiac Defects with Increased Anesthetic Risks

Several diagnoses merit mention due to their increased risks for complications during cardiac catheterization. The Risk Adjustment for Congenital Heart Surgery (RACHS) places stage I palliation of hypoplastic left heart syndrome (HLHS) in the highest risk category for perioperative complications following cardiac surgery [9]. Torres et al. reported a 19% mortality rate for non-cardiac surgery in children younger than 2 years of age with HLHS [10]. Induction instability was associated with

procedures performed prior to cavopulmonary anastomosis (Glenn procedure). Myocardial ischemia and cardiac arrest can occur suddenly in patients with single-ventricle physiology after induction of anesthesia and introduction of positive pressure ventilation. Reduced coronary perfusion pressure might be an important contributing factor and might be caused by a decrease in preload and a reduction in aortic root pressure if pulmonary vascular resistance decreases and pulmonary runoff increases, thereby decreasing systemic perfusion. Subendocardial perfusion is particularly tenuous if diastolic perfusion time is reduced by concomitant tachycardia. These patients also have a limited ability to increase coronary blood flow when myocardial oxygen demand is increased, for example with increased contractility or wall stress in response to an increased stimulus from an inadequate depth of anesthesia [11–13].

Pulmonary arterial hypertension is associated with an increased risk of perioperative cardiovascular complications. Carmosino et al. retrospectively reviewed children with pulmonary hypertension who underwent anesthesia for sedation for noncardiac surgery or cardiac catheterization [14]. Cardiac arrest and pulmonary hypertensive crises occurred in 4.5% of the children undergoing noncardiac surgery and 5.0% of the children undergoing cardiac catheterization. Major complications were predicted by suprasystemic pulmonary artery pressure, but were independent of patient age and the etiology of the pulmonary artery hypertension. A recent study on postoperative mortality in children identified 10 cases with preexisting medical conditions as a significant risk factor, with five of these patients having pulmonary hypertension [15]. Pulmonary hypertension causes right ventricle hypertension and hypertrophy, and induction of anesthesia with positive pressure ventilation further decreases the preload and increases the afterload on the right ventricle, causing the right ventricle to fall off the Starling curve and fail. These patients are almost impossible to resuscitate due to a lack of pulmonary blood flow secondary to increased pulmonary vascular resistance (PVR) and pulmonary artery pressure, leading to lack of venous return to the left heart and low cardiac output. In this patient population, it is important to maintain preload, potentially to start inotropic support prior to induction and to have inhaled nitric oxide (iNO) available to prevent or treat a pulmonary hypertensive crisis or cardiac arrest. Often, the baseline status of the patient is not achieved immediately following the procedure (particularly if the vascular resistance has been manipulated with iNO or other medications), and special attention to the patient post procedure is required until the patient returns to baseline.

Patients with left ventricular outflow tract (LVOT) obstruction have unique anesthetic implications. Williams-Beuren Syndrome patients with congenital supravalvular aortic stenosis with or without associated pulmonary stenosis and right ventricular pressure overload and hypertrophy are at increased risk for cardiac arrest when undergoing general anesthesia. These patients might develop coronary ischemia secondary to coronary artery abnormalities, including coronary ostial stenosis, despite being potentially asymptomatic. In patients with LVOT obstruction including aortic stenosis and hypertrophic cardiomyopathy with severe left ventricle hypertrophy, it is important to maintain diastolic pressure and coronary perfusion and to avoid tachycardia and ischemia [16]. Patients who have had a cardiac transplant are at increased risk of ventricular fibrillation during coronary angiography due to the risk of coronary artery disease in this population, as are patients with pulmonary atresia and RV-dependent coronary circulation.

Although most of these studies are limited by their retrospective nature, they identify significant risk factors for anesthesia in children, including an age younger than one year, a high ASA PS, a need for intubation and unrepaired or palliated cardiac lesions. Studies attempting to discern the causes of adverse

events have implicated anesthesia as a causative factor in a small percentage of cases. However, it is clear that anesthesia is associated with risks distinct from those related to the procedures themselves. The combination of these risks with the inherent risks of caring for pediatric patients with heart disease who might have severe alterations in cardiac physiology and function make it crucial to carefully consider the strategies and goals of management for each patient on an individual basis. Table 1 lists some of the common patient types and specific procedures at high risk for anesthesia-related complications.

Catheterization and Procedural Risks in the Catheterization Laboratory

The Congenital Cardiac Catheterization Outcomes Project (C3PO), using a strategy based on the congenital heart disease adjustment for risk method (CHARM), captured information from 8905 catheterization procedures over a three-year period from 2007 to 2010 at eight institutions with the goal of developing outcome assessment tools for cardiac catheterization procedures (17,18). The procedure risk types were grouped into four categories and diagnosis risk types were categorized into five categories [17]. Age younger than 1 year, recent cardiac surgery in the past 30 days, hemodynamic vulnerability, highest procedure risk group, transfer on ECMO support and longer case duration were risk factors for severe adverse events [18]. The risk of adverse events during cardiac catheterization of infants not only increases with lower age but also with weight less than 2 kg.

Odegard et al. retrospectively determined the risks of cardiac arrest in children with CHD in over 7289 cardiac catheterizations [19]. The risk of cardiac arrests was 0.96%, with a higher risk in children undergoing interventional procedures and in children younger than 1 year of age. The risk of cardiac arrests based on procedure type included device closure of a ventricular septal defect (11.9 per 100 procedures), intervention for intact atrial septum or restricted atrial septal flow (10.0 per 100 procedure), mitral valve balloon dilation (5.0 per 100 procedures), pulmonary vein dilations (3.6 per 100 procedures) and pulmonary artery balloon dilations (0.6 per 100 procedures). The higher risk of cardiac arrest in these patient populations is not surprising and should be anticipated. The risk is primarily related to the technical aspects of the procedure itself, including the pro-arrhythmogenic effect of stiff wires and catheters passing across the muscular septum and the low cardiac output resulting from stenting open semilunar and atrioventricular valves. There is also the risk of pulmonary reperfusion injury and pulmonary edema following pulmonary artery dilation. This complication might not be immediately apparent and careful post-procedure evaluation is necessary, in a similar manner as with the surveillance of pulmonary hypertension patients following their procedures. Newborns with single-ventricle physiology and a restrictive atrial communication can have a small left atrium, severe left atrial hypertension and can be in a low cardiac output state with severe hypoxemia at the time of the intervention. The risk of an inadvertent perforation causes these patients to be at increased risk for a cardiac arrest during the atrial septal opening procedure. Patients undergoing mitral valve balloon dilation to treat valvular stenosis also have left atrial hypertension and are at risk for arrhythmias and decreased cardiac output during balloon inflation, thus predisposing them to cardiac arrest.

Data from the IMPACT registry demonstrates that the rate of adverse events for diagnostic or interventional procedures is greatest in neonates (30.9% and 30.2% for diagnostic vs. interventional procedures, respectively) [8]. These data have not been broken down into weight categories, but we suspect that smaller and more premature newborns are likely at greater risk of complications than their

older and larger counterparts for any given procedure. Following neonates, infants (≥30 days, <1 year) have the next highest risk of adverse events (26.3% and 20.8%, respectively). Children (≥1 year, ≤18 years) have a risk of 5.5% and 7.3% for adverse events, respectively, whereas adult patients with congenital heart disease (>18 years) have a risk of 6.3% and 9.0%, respectively, for adverse events. When interventions are further stratified into one of six specific procedures captured in the IMPACT Registry (ASD and PDA occlusion, aortic and pulmonary valvuloplasty, angioplasty and stenting of aortic coarctation, and proximal pulmonary artery stenting), there are differences between groups with major adverse events (MAE) ranging from 0% in pulmonary valvuloplasty to 3.3% in aortic valvuloplasty and any adverse event occurring with a range of 4.7% in PDA occlusion, to 22.3% in balloon aortic valvuloplasty [20]. Holzer et al. reviewed the adverse event rate in the same cohort of patients undergoing interventions other than the six procedures cited above and found a similar spread of adverse events depending on the specific procedure. Pulmonary vein and Sano shunt interventions had the highest rate of adverse events at 25.3% and 37.5%, respectively [21].

A risk model is currently being applied to data from the IMPACT registry. After multivariable adjustment, eight variables were identified as critical for risk standardization: patient age, renal insufficiency, single-ventricle physiology, procedure-type risk group, low systemic saturation, low mixed venous saturation, elevated systemic ventricular end diastolic pressure and elevated main pulmonary artery mean pressure. The model had good discrimination (C-statistic of 0.70), confirmed by bootstrap validation (validation C-statistic of 0.69) [22].

It is clear that children with complex congenital heart disease are at increased risk during catheterization procedures of adverse events related not only to the procedure type but also to the underlying diagnosis and hemodynamic state. Although there are multiple publications addressing the management of children with CHD, there is no established methodology to address the magnitude of incremental risk conferred by the degree and severity and compensation of the heart disease. Identification of high-risk patient types presenting for cardiac catheterization must be extrapolated from retrospective studies across multiple disciplines, including cardiology, cardiac anesthesiology, pediatric anesthesiology and cardiac surgery.

Anesthesia

There is no specific anesthetic method that is appropriate for all patients with CHD in the PCCCL as long as the sedation provider or anesthesiologist understands the risks, the underlying pathophysiology and the impact of the sedation or anesthetic strategy on the hemodynamic status of the patient.

Volume Management

Attention to detail regarding the intravascular volume and hematocrit in patients with congenital heart disease is imperative during cardiac catheterizations, and physiologic alterations should be promptly addressed.

Hypovolemia might be present at the start of the procedure, particularly in small infants and children, secondary to dehydration occurring during prolonged periods of preoperative fasting (NPO). Hypovolemia is particularly important in very young, cyanotic, erythrocytotic or shunt-dependent patients. In these circumstances it is preferable to administer intravenous isotonic fluids to maintain hydration during the fasting period prior to catheterization. Hypovolemia can also occur acutely

secondary to massive blood loss. In that setting, normal saline, packed red blood cells, whole blood or 5% albumin can be administered to expand the intravascular blood volume while the cause of the bleeding is addressed. Careful attention to blood loss is particularly important in neonates who have a small blood volume and in cyanotic patients accustomed to an increased hematocrit.

Volume overload can occur during longer procedures, particularly those involving multiple angiograms, and is less tolerated in patients with congestive heart failure or shunt lesions. Acutely, this overload can lead to increased filling pressures, pulmonary edema and decreased ventricular function. Judicious use of flush solution administered to the patient with impaired reserves or to smaller patients is imperative to avoid iatrogenic hypervolemia. Any volume load (saline or blood transfusion) during a hemodynamic catheterization procedure needs to be performed with caution and communicated to the invasive cardiologist due to possible acute changes in filling pressures. The effect of changing hemoglobin concentrations on the calculations of cardiac output and vascular resistance must also be considered, as this affects hemodynamic measurements.

Some assessment of baseline hemoglobin or hematocrit should be performed either before or shortly after the start of the procedure the introduction IV or sheath. Erythrocytosis (HCT greater than 65%) occurs in cyanotic patients and is particularly common in the older cyanotic patient. While erythrocytosis increases oxygen carrying capacity, its higher viscosity reduces flow through the microcirculation, leading to decreasing cardiac output, increased pulmonary and systemic vascular resistance and a higher risk of thrombosis and emboli. If required, this erythrocytosis can be treated in the catheterization laboratory with phlebotomy, which includes replacing the blood withdrawn with either isotonic crystalloids or 5% albumin. At the other extreme, anemia decreases the oxygen carrying capacity of the blood, leading to increased cardiac output, elevation of trans-stenotic pressure gradients and exacerbation of congestive heart failure. Patients with cyanotic or single ventricular congenital heart disease, or those with moderate to severe myocardial dysfunction, benefit from the improved oxygen-carrying capacity of a hematocrit greater than 40%, whereas patients with excellent myocardial function can tolerate hematocrits as low as 25% without difficulty. Preexisting anemia is usually exacerbated during a cardiac catheterization by anticipated blood loss during sampling for oxygen saturations, blood gas determinations or clotting studies, as well as inadvertent blood loss occurring in vascular puncture sites, around catheters and wires and during sheath and catheter exchanges. Significant anemia should be identified and corrected prior to the catheterization procedure.

Effect of the Ventilation Strategy on Physiologic Measurements in the Cardiac Catheterization Laboratory

There is no preferred ventilation strategy in the cardiac catheterization laboratory. General anesthesia with positive pressure ventilation provides a secure airway and control of PaCO2, but increased intrathoracic pressure may alter hemodynamic measurements. Spontaneous ventilation might maintain more natural intrathoracic physiology and consequentially can result in the acquisition of more accurate hemodynamic data. However, oversedation can cause airway obstruction, hypoventilation and subsequent respiratory acidosis. This increases pulmonary vascular resistance and might alter shunt physiology and affect hemodynamic measurements [23,24].

During spontaneous ventilation, reduced intrathoracic pressure with inspiration facilitates venous return and right ventricular output, but afterload on the left ventricle is elevated, causing decreased left

ventricular cardiac output. The overall effect on cardiac output is minimal. During controlled ventilation, positive intrathoracic pressure causes IVC compression, reducing venous return and preload. The reduced preload causes a reduction in pulmonary blood flow and, subsequently, in cardiac output. These reductions are particularly pronounced in patients with right heart failure, hypovolemia, or Fontan physiology. Although this outcome should be taken into consideration, particularly in pulmonary hypertensive patients, studies have demonstrated no difference in complication rates between general anesthesia and sedation in pulmonary hypertension patients undergoing cardiac catheterization [25].

Children with CHD can also have alterations in lung function. Extrinsic compression of conducting airways by enlarged atria and pulmonary arteries can cause decreased lung compliance and increased airway resistance [28]. Controlled ventilation can have variable effects on these patients.

The individual patient and clinical situation should be considered when developing an anesthetic plan and selecting a ventilation technique. Regardless of the technique, patients having even minimal sedation should have their airway and EtCO2 monitored throughout the procedure.

Fostering an Environment of Partnership in Decision Making for Optimal Patient Care, Open Communication and Coordination Between Cardiologists and Anesthesiologists

First and foremost, patient care is about patient safety and achieving the desired outcome. In a multi-professional procedure, optimal outcomes are best accomplished by face-to-face/personal interactions between team members. Interventional cardiologists, anesthesiologists and surgeons (for hybrid procedures) must spend time discussing the procedure, risks and strategies prior to the procedure taking place, especially for high-risk procedures. It is important to consider all viewpoints, including whether the procedure is truly necessary before undertaking it. Anesthesiologists and cardiologists must have the ability to work effectively with each other as well as with the cardiac catheterization team, and when instituting ECMO or during hybrid procedures with the operating and surgical team. The development of small, expert teams that promote the optimization of individual performance and enhanced communication should be the ultimate goal. Cardiologists and anesthesiologists should value the importance of divergent perspectives because group expertise often trumps individual expertise. In so doing, leadership in the catheterization lab should be fluid. The decision maker can change during the case, depending on the patient's status and the situation requiring a decision. Finally, each specialty should appreciate the constraints of the other's work environment.

Periprocedural checklists, (similar to perioperative checklists used in surgery) have become increasingly routine, with the goal of improving outcomes and reducing adverse events. These checklists are meant to improve intraoperative management and communication and to facilitate postprocedural handoffs between units and providers. These checklists <u>might</u> include the lists below.

Preprocedure

- 1. Patient identification, diagnosis, intended procedure, documentation of a history and physical examination and a signed consent form.
- 2. Documentation of allergies, including drug, food, latex and contrast.

- 3. Airway assessment.
- 4. Verified NPO status.
- 5. Confirmed plan for patient monitoring.
- 6. Room setup including the need for transesophageal echocardiography (TEE), iNO, and rotational angiography.
- 7. Special medications required, including but not limited to antibiotics, antiarrhythmics, anticoagulants, vasopressors, stress-dose steroids.
- 8. Availability of implanted devices, including pacemakers and implantable cardioverter defibrillators.
 - 9. Bleeding risks, including the presence of a type and screen or crossmatch and the need for immediate blood in the catheterization laboratory.
- 10. Anesthesia plan (general anesthesia versus monitored anesthesia care [MAC] with spontaneous ventilation).
 - 11. Consideration of the need for ECMO support and surgical backup.
 - 12. Access plan, sheath size and previous access issues.
 - 13. Plan for postoperative recovery; home, regular floor or ICU.
 - 14. Pregnancy test as necessary.

Intraprocedure

- 1. Patient identification, diagnosis, and procedure.
- 2. Allergies, including drug, food, latex, and contrast.
- 3. Antibiotic requirements.
- 4. Heparin plan.
- 5. Blood availability.
- 6. Availability of vasopressor medications and resuscitation equipment.
- 7. Disposition postprocedure (inpatient unit, intermediate-care unit, or intensive-care unit).

Postprocedure

- 1. Adverse events.
- 2. Equipment issues.
- 3. Specimens labeled and sent.
- 3. Detailed sign-out to floor, intermediate-care unit, intensive-care unit, or PACU.
- 4. Necessary postprocedure tests, including echocardiogram, X-ray, or EKG.
- 5. Necessary postoperative medications, including but not limited to antiarrhythmics, vasopressors, iNO, and anticoagulation, antiemetics and intravenous fluid therapy.
- 6. Indwelling catheters and lines.
- 7. Last dose of sedation and neuromuscular blockade.
- 8. What could have gone better during the procedure.

Enhancing Awareness of the Congenital Cardiac Catheterization Lab as a Physiological Laboratory and a Unique High-Risk Environment

Although the PCCCL continues to transition from a diagnostic tool to an interventional and therapeutic theater, (much like an operating room), hemodynamic assessment continues to be of paramount importance in establishing a diagnosis and assessing the need for intervention (catheter-based or surgery). Awareness of the effect that anesthetic agents and ventilator strategies have on these parameters must be considered when planning the case. Therefore, direct communication between the cardiologist and anesthesiologist is mandatory and is best done prior to the patient arriving in the catheterization laboratory. This communication can reduce the procedural, anesthetic and radiation times, all of which are likely to improve patient outcomes.

The Level of Cardiology Expertise Appropriate for the Pediatric and Cardiac Catheterization Laboratory

There is no sub-specialty certification from the American Board of Pediatrics for pediatric cardiologists preforming catheterizations or for any other pediatric cardiology subspecialty, nor does the Accreditation Council for Graduate Medical Education (ACGME) recognize advanced training in catheterization. However, new core training requirements for pediatric cardiology recently published by the Society of Pediatric Cardiology Training Program Directors (SPCTPD) [26] and endorsed by the ACC, AHA, AAP and SCAI suggest additional training should be required for pediatric cardiologists performing these procedures. This suggestion is further detailed in SCAI's 2014 expert consensus statement, recommending that physicians intending to perform cardiac catheterization procedures have additional advanced training [27].

The Level of Anesthesia Expertise Appropriate for the Pediatric and Congenital Cardiac Catheterization Laboratory

As referenced above, children with CHD are at significant risk for morbidity and mortality in the PCCCL. This is particularly true for infants, for patients with specific cardiac pathology, for patients with ASA patient status 3 or above and for patients undergoing certain transcatheter interventions. Specific knowledge of congenital cardiac anomalies and physiology with an applicable skill set is necessary for prevention and management of hemodynamic compromise and cardiac arrest in the catheterization laboratory. Clinicians providing moderate-to-deep sedation for patients with CHD in the catheterization laboratory must be prepared to manage not only the airway, but must also understand that airway obstruction and/or hypoventilation affects the patient's unique physiology and could have catastrophic effects in patients with CHD. Clinicians must balance providing adequate sedation/anesthesia to the patient with the ability to anticipate, rapidly identify and appropriately respond to hemodynamic changes and deterioration that might require medical resuscitation, cardiopulmonary resuscitation (CPR), initiation of inhaled nitric oxide, treatment of massive hemorrhage and emergent cannulation into extracorporeal (ECMO) support.

In 1997, pediatric anesthesiology met the criteria for recognition as a subspecialty by the ACGME, and the Anesthesiology Residency Review Committee developed program requirements for a 12-month

subspecialty fellowship training program. In 2013, the first pediatric anesthesiology subspecialty examination was offered by the American Board of Anesthesiologists. As with the training of pediatric congenital cardiologists, there is no formal ACGME-accredited fellowship training or certification process in pediatric congenital cardiac anesthesiology. Pediatric cardiac anesthesiology has evolved on an institutional basis, leading to wide discrepancies in training. At the same time, there have been significant advances in surgical interventions, nonsurgical interventions and medical therapies for congenital heart disease. These changes, along with improved outcomes, have led to surgical and nonsurgical treatments being offered to higher-risk patients than in the past. Pediatric cardiac anesthesiologists have played a pivotal role in the advancement of this care. Many tertiary care centers have pediatric cardiac anesthesiology divisions with members who attained expertise through a variety of training pathways. Due to the need for standardized and regimented training pathways to ensure high-quality care and continued success with the treatment of CHD, a working group from the Congenital Cardiac Anesthesia Society (CCAS) published a proposal for formal training in pediatric cardiac anesthesiology was published in 2014 [29], but no formal training has yet been developed.

Because a limited number of academic medical centers offer advanced training or fellowship training in pediatric cardiac anesthesiology, there are an insufficient number of trained pediatric cardiac anesthesiologists to support every catheterization laboratory that cares for patients with CHD undergoing cardiac catheterization.

Given the complexity of these patients, the unique environment of the cardiac catheterization laboratory and the limited number of pediatric cardiac anesthesiologists, who then should provide sedation/anesthetic care for these patients? Current manpower limitations suggest that, in many institutions, having pediatric cardiac anesthesiologists provide care for all patients in cardiac catheterization laboratories is not feasible. Given this situation, it is recommended that anesthesiologists involved in pediatric cardiac catheterization procedures should, as is true for cardiologists, have sufficient subspecialty training and experience to provide expert care. Guidelines defining sufficient training are evolving. Our recommendation is that the expertise of those providing anesthetic care for these patients be appropriate to the level of risk associated with the procedure. For higher-risk patients, care should be provided, at a minimum, by anesthesiologists with advanced skills and knowledge relevant to the pathophysiology of CHD. This knowledge must include a comprehensive understanding of the effects of anesthetic drugs, inotropes and respiratory interventions on the physiology specific to each congenital heart lesion and surgical palliation. This understanding is critical to providing a stable hemodynamic state that allows the accurate measurement and interpretation of the hemodynamic parameters obtained in the catheterization laboratory. Examples include the effects of FiO2 and pCO2 on pulmonary vascular resistance, changes in cardiac output associated with controlled versus spontaneous ventilation and the myocardial and electrophysiologic effects of anesthetics.

Future Considerations

Central to addressing patient safety in the PCCCL is the ability to prospectively stratify patients and their procedures to accurately predict and thereby potentially prevent or treat an adverse event during

catheterization. This approach is critically important for optimal planning of the appropriate resources, levels of expertise, and types of anesthesia or sedation required for success. Data are emerging that identify both patient- and procedure-specific characteristics that currently affect adverse event risks and therefore allow for improved planning and resource deployment. Recently, a scoring system (CRISP Score) has been developed that predicts the risk of a serious adverse event based on specific patient and procedural characteristics [30]. This scoring system assigns risk based on demographics such as age and weight as well as the underlying diagnosis and other concurrent systemic illnesses. In addition, the system assesses the potential for hemodynamic compromise by considering the need for inotropic support and the presence or absence of specific physiologic parameters that place a patient at higher risk. Such a tool could have considerable application in planning for the appropriate availability of resources, including anesthesia expertise. The clinical impact of such tools remains to be seen, because resource utilization requires careful consideration of what is needed, but also of what might be excessive.



To this end, we foresee that programs might assign specific PCCCL patients and procedures to broad risk categories that would inform the level of resource, expertise and anesthesia or sedation strategy. For some select procedures and patients we can now quantify a very high potential risk for a major adverse event or even a catastrophic event. For example, the following patients are at increased risk: neonates who are candidates for PDA stent placement; neonates with single ventricular physiology; neonates with left-sided AV valve hypoplasia or atresia with a restrictive atrial septum, who are candidates for atrial balloon septostomy or stent placement; and older patients with severely calcified stenotic RV to PA conduits who are candidates for dilation and transcatheter valve placement. In this group of patients, maximal resources, expertise and an aggressive anesthesia and airway management strategy are indicated. Conversely, we know that some procedures have extremely low adverse event risks, such as noninfant ASD device closure, PDA closure >1 year or pulmonary valvuloplasty, suggesting a more modest allocation of resource support could be adequate.

Although seemingly intuitive, the majority of procedures and patients lie between the extremes of this continuum. A critical evaluation of each patient and procedure is paramount to assessing potential risks and planning appropriate resources and pharmacologic management for the sedated or anesthetized patient. Formal pre-catheterization risk assignment tools such as the CRISP scoring system have the potential to structure this assessment so that resource availability can be critically evaluated with respect to the impact on resource allocation and associated expenses. Table 2 outlines a scheme for resource allocation specific to provision of sedation/anesthesia based on the CRISP scoring system. Programmatic management dictates that resources for reasonably anticipated adverse events be available for every patient. Above all, patients, not their practitioners, take risks. The patients we serve deserve no less than a formal assessment of the resources needed to ensure their safety and a successful outcome.

Conclusion

The care of patients with complex congenital heart disease requires multiple professionals with differing but interdependent skill sets. Achieving the best possible outcome requires a team approach with mutual respect for all involved. Knowledge of patient anatomy and physiology, the goal of the procedure, an

understanding of the risks and a plan of action for potential complications are paramount for a successful outcome.

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Table 1: Anesthetic Risks for Specific Patients and Procedures

High-risk patients and procedures for anesthesia	Possible adverse events from anesthesia	
Williams-Beuren Syndrome	Hypotension, Coronary Ischemia, Cardiac Arrest	
Hypertrophic Cardiomyopathy	Coronary Ischemia, Cardiac Arrest	
Single-Ventricle Physiology	Coronary Ischemia, Cardiac Arrest	
	Low Cardiac Output (CO), Coronary Ischemia,	
Aortic Valve Stenosis	Cardiac Arrest	
Mitral Valve Stenosis	Low CO, Right Ventricular (RV) Failure	
	RV Failure, Pulmonary Hypertensive Crisis,	
Pulmonary Hypertension	Cardiac Arrest	
Pulmonary Vein Dilation	RV Failure, Pulmonary Edema, Hypoxia	
VSD Device Closure	Arrhythmia, Low CO, Cardiac Arrest	
Balloon Atrial Septostomy (newborns with single-		
ventricle physiology and left AV valve hypoplasia		
or atresia)	Hypoxia, Atrial Perforation, Cardiac Tamponade	
s/p Heart Transplant	Ventricular Fibrillation (VF), Myocardial Ischemia	

Table 2: Recommended anesthesia provider expertise based on CRISP score

	Minimum Level of Anesthesia Provider Expertise		
	Sedation Team ¹	Anesthesiologist with	Pediatric Cardiac
		Special Expertise in CHD	Anesthesiologist ²
CRISP Score			
0 to 1	X		
2 to 4		X	
5 or greater			X

 $^{^{1}}$ Sedation provided by non-anesthesiologists with training and certification in sedation practices

² Pediatric anesthesiologist with either advanced training or extensive experience in congenital cardiac anesthesia