Chapter 13
Sedation Considerations for Patients with Congenital and Acquired Heart Disease

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Introduction

Infants and children with congenital heart disease (CHD) often require deep sedation for procedures and imaging. The increased use of cross-sectional imaging for diagnostic purposes in congenital and acquired heart disease has also increased the need for deep sedation in this patient population. Patients with CHD and acquired heart disease are at increased risk for sedation-related complications. They often possess unique physiology as a result of their heart defects or palliative surgeries, which impacts their response to sedation. Among this group of patients, those who carry the highest risk of sedation-related complications include those younger than 2 years, single ventricle physiology, left ventricular outflow tract obstruction, impaired ventricular function, and pulmonary hypertension.

When considering sedation for the patient with heart disease it is imperative for the provider to understand anatomic variations, surgical history (if applicable), and the physiologic implications this may have on the individual patient. It is helpful to group the physiology of heart disease in children into major categories. This can assist the provider in preparing for potential consequences of providing sedation. Table 13.1 shows the preparation of a cardiac patient for procedural sedation.

The first grouping is to divide patients into congenital versus acquired heart disease. Within CHD there are several subcategories including: cyanotic versus acyanotic; repaired versus palliated; right-to-left intracardiac shunts versus left-to-right intracardiac shunts; and right and left ventricular outflow tract obstructions. The acquired heart disease category includes the cardiomyopathies (dilated, hypertrophic, restrictive), myocarditis, and arrhythmias. Understanding the general category

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Table 13.1 Preparation for sedation of the cardiac patient

Review most recent progress note from cardiology
Review most recent imaging studies including echocardiogram and ECG
Review all cardiac medications
Review baseline oxygen saturations in room air
Discuss patient with cardiology or cardiac anesthesia if any concerns arise

Table 13.1 is made by author Dr. Michael Wolf

Table 13.2 When referral to cardiac anesthesia is warranted

Unrepaired cyanotic congenital heart disease
Neonate with repaired complex congenital heart disease
Pulmonary hypertension
Single ventricle physiology
Shunt-dependent pulmonary blood flow
Left ventricular outflow tract obstruction
Cardiomyopathy with depressed systolic or diastolic ventricular function

Table 13.2 is made by author Dr. Michael Wolf

that a patient belongs to and what implications that will have on their response to sedation is the first step in preparation.

Consideration should also be given to common comorbidities associated with CHD and surgical repairs such as compromised lung compliance from congestive heart failure, genetic associations, phrenic or recurrent laryngeal nerve injury, pulmonary hypertension, or rhythm abnormalities. Table 13.2 shows patients with heart disease who should be referred to the anesthesia service.

Congenital Heart Disease

In order to simplify the grouping of patients with CHD, it is easiest to group them into three major anatomic categories: those with increased pulmonary blood flow, those with decreased pulmonary blood flow, and those with outflow tract obstruction. These anatomic subsets are not exclusive and certain patients may fit into one or more of these categories. In addition, those with single ventricle anatomy have a unique physiology that may swing from increased to decreased pulmonary blood flow with variations in their intrinsic vascular resistance.

Patients with increased pulmonary blood flow typically have CHD lesions that cause left-to-right shunting of blood and over circulation of the pulmonary vascular bed. This includes ventricular septal defects (VSD), atrial septal defects (ASD), and patent ductus arteriosus (PDA). Symptomatic patients in this category will present with symptoms of increased work of breathing, relative tachycardia, feeding intolerance, and difficulty gaining weight as a result of increased pulmonary blood flow at

the expense of systemic blood flow. The balance of pulmonary and systemic circulations is influenced by the relative resistances in each circuit: pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR).

Patients with decreased pulmonary blood flow typically have CHD lesions that cause right to left shunting of blood or decreased pulmonary blood flow. This includes tetralogy of Fallot, pulmonary stenosis, and Ebstein anomaly of the tricuspid valve. These patients present with cyanosis, respiratory difficulties, or exercise intolerance as a result of chronic hypoxia. Changes in lung compliance including from patient agitation can exacerbate the level of cyanosis and it can take time until saturations recover to their baseline. When an intracardiac communication is present (such as the VSD of tetralogy of Fallot) cardiac output is preserved even in the face of relative hypoxia.

Patients with left ventricular outflow tract obstructions can be in the category of CHD or acquired heart disease. This includes those with aortic stenosis (subvalvar, valvar, supravalvar), coarctation of the aorta, and hypertrophic obstructive cardiomyopathy. The downstream obstruction to outflow from the left ventricle increases the oxygen demand on that ventricle and comes at the expense of decreased cardiac output. Patients may develop symptoms related to decreased left ventricular output including respiratory difficulties from pulmonary edema, feeding intolerance from intestinal ischemia, and end-organ damage from decreased oxygen delivery. Maintaining SVR in this patient population can be an important factor in maintaining adequate cardiac output.

Patients with single ventricle physiology are generally grouped into their own physiologic category. As they move through the palliative stages of surgery, their baseline saturations, physiologic expectations, and balance of systemic and pulmonary blood flow can change drastically. Single ventricle patients with shunt-dependent pulmonary blood flow have the most tenuous physiology and their response to alterations in PVR and/or SVR can be unpredictable and dramatic. It is generally accepted that these patients, and even those who have undergone superior cavopulmonary anastomosis (Glenn operation) be referred primarily to cardiac anesthesia when sedation is needed. Following Fontan completion, there are scenarios in which these patients become acceptable candidates for deep sedation by a sedation service.

Acquired Heart Disease

Patients with cardiomyopathies are typically quite sensitive to changes in PVR and SVR and are thought to be poor sedation candidates. Dilated cardiomyopathy is usually associated with decreased left ventricular function; changes in SVR, PVR, venous return to the right side of the heart, and alterations in PVR can have devastating effects on these patients. Hypertrophic and restrictive cardiomyopathies are often present with preserved ventricular function. This should not reassure the sedation provider as both cardiomyopathies cause baseline alterations in cardiac physiology that is exquisitely sensitive to alterations in both SVR and PVR. It is generally

accepted that patients with myocardial disease should be referred primarily to cardiac anesthesia when sedation is needed.

Arrhythmias are a broad category of acquired heart disease that may impact sedation candidacy. Consideration must be given to the underlying arrhythmia itself as well as to the current therapy required to treat the abnormal rhythm. Supraventricular tachyarrhythmias are controlled with medications until such time as an ablation can be performed. Ventricular arrhythmias are controlled with medication, and cause significantly more distress to providers when considering the physiologic impact of triggering the arrhythmia. Bradyarrhythmias typically require pacemaker insertion when they cause symptoms. Genetic arrhythmias such as long QT syndrome (LQTS) can also influence sedation considerations. Medication selection must be undertaken with careful consideration for potential QTc prolongation. In addition to consideration of the arrhythmia itself, many antiarrhythmic medications cause myocardial depression. Careful attention should be paid to patients' medications as well as their most recent functional evaluation by echocardiogram prior to consideration for sedation in the setting of an arrhythmia.

Pulmonary Hypertension

Pulmonary hypertension is a category of heart disease that combines the spectrum of both congenital and acquired heart disease. Congenital lesions with unrepaired left-to-right shunts can develop Eisenmenger syndrome with fixed pulmonary vascular resistance and pulmonary hypertension. Pulmonary vein obstruction, whether following CHD such as total anomalous pulmonary venous connection (TAPVC) or in former premature infants with bronchopulmonary dysplasia, causes significant pulmonary hypertension. Lesions that cause elevation in left atrial pressure such as mitral stenosis can cause pulmonary hypertension as well. In the absence of structural heart disease, patients can present with primary pulmonary hypertension from a variety of causes including etiologies such as idiopathic, chronic lung disease, or chronic thromboembolic pulmonary hypertension. Pulmonary hypertensive crises typically involve severe refractory hypoxia and decreased cardiac output; triggers are variable for different patients, but sedation is extremely high risk in this patient population out of concern for triggering a crisis.

Noncardiac Considerations

Cardiac physiology and hemodynamics in isolation is not the sole determinant of sedation candidacy; patients with both CHD and acquired heart disease can develop several noncardiac consequences of their underlying cardiac pathology. Pulmonary compliance and mechanics can be altered by increased pulmonary blood flow or

chronic hypoxia. Attention should be paid to the presence of airway compression by vascular structures that may mimic tracheomalacia. Phrenic nerve injury can lead to impaired respiratory mechanics from diaphragm dysfunction, and recurrent laryngeal nerve injury may result in chronic aspiration and lung disease from vocal cord dysfunction.

Patients with chronic cyanosis will typically develop a compensatory polycythemia and resultant hyper viscosity syndrome. This can lead to neurologic sequalae including thrombotic strokes. The presence of chronic right to left shunts exposes patients to paradoxical emboli and resultant strokes or cerebral abscesses. Exposure to cardiopulmonary bypass in the neonatal period increases the risk of development of both attention deficit hyperactive disorder (ADHD), seizure disorder, and autism.

Several genetic syndromes carry strong association with CHD including: Trisomy 21, DiGeorge syndrome, William syndrome, Noonan syndrome, Mucopolysaccharidoses, Pompe disease, Alagille syndrome, to name a few. Sedation providers need to be aware that some of the genetic syndromes with CHD may have airways, which are associated with difficulty with bag-mask ventilation, laryngoscopy, and airway visualization.

Cardiac Sedation Physiology

When considering sedation for a patient with CHD there are several important hemodynamic principles to keep in mind. Patients with left-to-right intracardiac shunts (VSD, ASD, PDA) will tend to have some pulmonary edema and decreased pulmonary compliance from pulmonary vascular overload. The degree of left-to-right shunting will improve with decreased SVR; therefore, deep sedation in these patients is generally well tolerated assuming their pulmonary status is stable prior to induction of sedation.

The degree of hypoxia in patients with right to left shunts tends to improve with increased SVR; when sedated, these patients tend to have increased hypoxia as more blood flow is directed away from the pulmonary vascular bed. Supplemental oxygen and using medications that do not significantly lower SVR can be helpful in improving sedation tolerance. In addition, avoiding scenarios that increase PVR such as airway obstruction can help prevent further hypoxia. While sedation is generally well tolerated in this population, the variability in both saturations and response to supplemental oxygen tend to push them toward cardiac anesthesia referral.

Left ventricular outflow tract obstruction is exquisitely sensitive to alterations in SVR. Decreased SVR will exaggerate the gradient from the left ventricle to the systemic circulation. Intravascular volume status is equally important to maintain cardiac preload; and close attention must be paid when scheduling fasting (NPO) times prior to sedation. It is generally accepted that patients with left ventricular outflow tract obstruction require referral to cardiac anesthesia.

Contraindications to Sedation

There are no published guidelines for referral to cardiac anesthesia, but there are widely accepted principles that guide decision-making regarding sedating the cardiac patient. It is helpful to have a sedation provider who is oriented to both critical care and cardiology to review questionable cases and decide on referral necessity. Providers trained in both critical care and cardiology have a unique perspective and the ability to review echocardiographic imaging, cardiac physiology, and sedation response when formulating a plan for sedation candidacy. Residual lesions are fairly common that following pediatric cardiac surgery; having a provider who intuitively understands the physiologic impacts of those residual lesions is valuable.

Each center will have different thresholds for anesthesia referral. However, it is generally accepted that patients with the following issues are referred to cardiac anesthesia for sedation: unrepaired cyanotic CHD, neonates with repaired or unrepaired CHD, single ventricle physiology, all shunt-dependent infants, left ventricular outflow tract obstruction, cardiomyopathy with impaired systolic or diastolic ventricular function, and pulmonary hypertension. Patients with Williams syndrome (supravalvular aortic stenosis and coronary anomalies) are at inherent risk for myocardial ischemia during procedural sedation and are best referred to cardiac anesthesia. When in doubt a cardiology and/or cardiac anesthesia consult is advised to evaluate sedation candidacy.

Presedation Considerations

In addition to reviewing all medical history prior to standard sedation, there is some crucial data that should be reviewed carefully prior to sedating the pediatric patient with cardiac disease. Reviewing the most recent inpatient and/or outpatient note from a cardiologist is imperative. This should include information regarding candidacy for anesthesia and minor procedures as well as the need for endocarditis prophylaxis. The most recent echocardiogram, electrocardiogram (ECG), and cardiac catheterization (when applicable) should be reviewed. Having a cardiologist or cardiac intensivist review, this data can be extremely helpful when possible.

All medications should be reviewed with attention paid to diuretics, antihypertensives, and pulmonary vasodilators. Significant diuretic need can be a good indicator of the degree of potential respiratory compromise that may be anticipated during sedation. Lastly, reviewing the patient's baseline oxygen saturations in room air will guide the provider regarding what to expect during sedation. It is also important to understand whether oxygen may exacerbate certain conditions, such as in the presence of a left-to-right shunt.

Specific Medications

Propofol

Propofol is a potent hypnotic drug with sedative and amnestic properties. It decreases SVR while PVR remains unchanged. It can be used safely in patients with repaired CHD who have normal ventricular function. Decreasing SVR will lessen the degree of left-to-right shunting in those patients with unrepaired ASD, VSD, or PDA; this is usually well tolerated in this patient population as decreased pulmonary blood flow has the potential to decrease respiratory compromise.

Propofol has the opposite effect on those with limitation in their pulmonary blood flow such as tetralogy of Fallot or obligate right to left shunting. Decreasing SVR in this patient population will decrease pulmonary blood flow further potentiating hypoxia during sedation. Propofol's pronounce impact on SVR makes it a poor choice in patients whose pulmonary blood flow depends on balancing SVR and PVR.

Propofol is a potent myocardial depressant and should be avoided in patients with compromised ventricular function. Its effect on SVR will exacerbate left ventricular outflow tract gradients and should be avoided in any scenario that involved such obstructions including hypertrophic obstructive cardiomyopathy. It is considered safe in patients with singe right ventricles who have undergone Fontan completion and have normal systolic function. Consultation with a pediatric cardiologist is advised prior to using propofol in this population.

Propofol's effect on the QTc interval remains the subject of dispute. The literature is equivocal regarding whether QTc prolongation is an absolute contraindication for propofol use. Propofol should be avoided in patients with confirmed LQTS; and consideration should be given to avoiding its use in those with baseline prolonged QTc on ECG.

Ketamine

Ketamine is a mixed sedative and analgesic medication with a favorable hemodynamic side effect profile. It can be delivered via intramuscular injection when no intravenous line is present. It typically allows for maintenance of mean arterial pressure without meaningful changes in either SVR or PVR. It has positive effect on bronchospasm and increases upper airway tone making it an attractive choice to maintain spontaneous respiration. It is well tolerated in most patients with CHD including those with unrepaired cyanotic CHD and pulmonary hypertension. It should be used with caution in patients with airway issues because of its propensity to increase secretions.

Ketamine should be used with caution in patients with decompensated cardiogenic shock. It can potentiate circulatory collapse because of their

catecholamine-depleted state; ketamine's maintenance of SVR relies on its ability to cause release of intrinsic catecholamines.

Etomidate

Etomidate is a sedative hypnotic agent with a favorable hemodynamic side effect profile. It does not cause decrease in either SVR and PVR and has no negative effect on myocardial contractility. It is an excellent choice for deep sedation in patients with impaired ventricular function. It is typically well tolerated in this population. Side effects to be aware of include laryngospasm and myoclonus. Caution is advised in patients with infectious concerns given the potential for adrenal–pituitary suppression from etomidate. Etomidate has a relatively short half-life, which should be considered when using it for longer imaging studies that may necessitate additional doses or a continuous infusion.

Fentanyl

Fentanyl is a potent opioid analgesic with sedative properties and a favorable hemodynamic profile. It is typically well tolerated even in relatively large doses in patients with repaired and unrepaired CHD. It is also well tolerated in neonates and typically does not cause appreciable changes in SVR and PVR. It is an excellent choice for invasive or painful procedures and can be used together with a sedative to provide adequate sedation and analgesia. Care should be taken regarding bolus infusion rates in neonates because of the risk of rigid chest syndrome.

Midazolam

Midazolam is a potent benzodiazepine sedative that provides both anxiolysis and sedation. It can be delivered via the oral and intranasal route when intravenous access is not available. Midazolam has pronounced effects on decreasing SVR and, therefore, its effect on left-to-right and right-to-left shunting will be similar to propofol. Although not a direct myocardial depressant, it can lead to pronounced hemodynamic compromise because of its effects on mean arterial pressure and SVR. When possible, midazolam is avoided in patients with cardiac disease because of its hemodynamic effects. The oral and intranasal routes have less impact on patient hemodynamics and can be considered safer in this patient population.

Dexmedetomidine

Dexmedetomidine has sedative, anxiolytic, and mild analgesic properties with minimal respiratory depression and variable effects on hemodynamics. By virtue of its pharmacodynamics, dexmedetomidine can cause bradycardia and hypotension that are dose dependent and not necessarily consistent from one patient to the next. It can be delivered via the intranasal route when intravenous access is not available. It is considered a safe sedation medication in patients with both repaired and unrepaired CHD and is generally well tolerated in all ages including neonates. It should be avoided in patients with bradyarrhythmias and those on digoxin because of the potential to worsen bradycardia or cause intermittent heart block.

Sedation Recovery

Sedation recovery does not differ significantly for cardiac patients. For those with hemodynamically significant cardiac disease, continuous telemetry and pulse oximetry should be monitored during the recovery period. Minimizing NPO times is crucial for certain patient populations; consideration for intravenous hydration should be given to those patients with a longer recovery time to avoid hypovolemia. Discharge from sedation is appropriate once patients have returned to their neurologic baseline with stable cardiac and respiratory status.

Summary

Infants and children with congenital and acquired heart disease are at increased risk for sedation-related complications. With increasing use of cross-sectional imaging as well as other minor procedures the demand for sedating patients with heart disease continues to increase. Careful consideration should be given to each individual patient's candidacy for sedation and consultation with a cardiac intensivist or cardiologist is recommended. A low threshold for cardiac anesthesia consultation and referral should be maintained in those patients with unrepaired, hemodynamically significant CHD, hemodynamically significant acquired heart disease, or hemodynamically significant residual lesions following cardiac surgery.

Suggested Readings

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