The Pre-sedation Assessment and Implications on Management

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Abstract

In this chapter we will present the essentials of pre-sedation screening and risk stratification, discuss fasting guidelines, and review the most commonly encountered scenarios and comorbidities that impact sedation management and outcomes. Today's practice of pediatric sedation (PS) involves ever more complex patients whose care is coordinated with multidisciplinary teams. Technological advances have allowed for the development of various invasive and noninvasive pediatric procedures and imaging modalities, resulting in a tremendous demand for and growth in PS in children. Despite the increasing complexity and patient volume, sedation providers generally meet the child and his family only minutes before the scheduled (or unscheduled) procedure. The provider must assess the situation quickly and accurately to ensure safety and optimal effectiveness. Important data from all available resources should be gathered and synthesized before the procedure to formulate a successful sedation plan within the context of the urgency of the procedure.

Keywords

Pediatric sedation • Pre-sedation • Screening • History • Physical exam • Fasting guidelines • Asthma • Reactive airway disease • Autism spectrum disorders (ASD) • Developmental delay • Intellectual disability • Bronchopulmonary dysplasia (BPD) • Cerebral palsy • Congenital heart disease • Cystic fibrosis • Diabetes mellitus • Endocrinopathies • Mitochondrial disease • Allergies • Muscular dystrophy • Musculoskeletal disorders • Obstructive sleep apnea • Pregnancy • Premature infant • Psychiatric disorders • Behavioral disorders • Sickle cell disease • Syndromes • Trauma • Tuberous sclerosis • Upper respiratory tract infection • Food and Drug Administration (FDA) • Cyanotic heart disease • New York Heart Association classification (NYHA) • Congestive heart failure (CHF)

Introduction

In this chapter we will present the essentials of pre-sedation screening and risk stratification, discuss fasting guidelines, and review the most commonly encountered scenarios and comorbidities that impact sedation management and outcomes.

Today's practice of pediatric sedation (PS) involves ever more complex patients whose care is coordinated with multidisciplinary teams. Technological advances have allowed for the development of various invasive and noninvasive pediatric procedures and imaging modalities, resulting in a

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tremendous demand for and growth in PS in children. Despite the increasing complexity and patient volume, sedation providers generally meet the child and his family only minutes before the scheduled (or unscheduled) procedure. The provider must assess the situation quickly and accurately to ensure safety and optimal effectiveness. Important data from all available resources should be gathered and synthesized before the procedure to formulate a successful sedation plan within the context of the urgency of the procedure.

The saying "an ounce of prevention is worth a pound of cure" encapsulates the pre-sedation mindset. The main objective for the sedation provider during pre-procedural assessment is to answer the question: **Is this child optimized for the procedure or not?**

Components of a successful sedation plan include readily accessible medical records, a thorough medical history with review of systems and careful attention to red flags, pre-sedation tests, or consultation if indicated, a targeted physical exam, and a complete understanding of the procedure and its potential physiologic effects on the patient.

Pre-sedation Screening

All children scheduled for elective sedation should receive a prescreen telephone call before the scheduled invasive or noninvasive procedure. Last-minute cancellation due to new information surfacing on the day of the procedure can result in delay of care and economic loss for the parents and the institution. The telephone screening allows for review of the medical history, gives the opportunity to determine if there is some underlying medical issue that requires further investigation, confirms that the child has not been recently ill, and reinforces *nil per os* (NPO) instructions. Pertinent data points should be clearly documented and attached to a standardized, hospital-approved sedation assessment form.

Once the screening process is complete, an established triage system can help to determine whether the procedure is appropriate for non-anesthesiologist sedation or whether the expertise of an anesthesiologist is needed. In many centers there is a "point person" to whom non-anesthesiologists may direct questions regarding patient management issues in off-site venues. This coordinator should be familiar with the requirements, challenges, and needs of the individual specialists. In the case of an urgent or emergent (non-elective) procedure, the same logic applies: **Gather as much information as possible and reasonable for your setting to make the most informed decision regarding the timing and approach to the procedure**.

History

The process of constructing a successful sedation plan starts with a careful, targeted history focusing on a few critical domains. Ask about past problems or known abnormalities of the respiratory, cardiovascular, neurologic, gastrointestinal, and endocrine systems. Some parents may not be familiar with medical terminology or may assume that you are aware of the child's history; the provider can work around this by describing common problems and/or procedures, pursuing anything that "sounds familiar." Review any available medical records and contact the primary care provider if possible. Examine previous records in regard to previous problems with airway management, obtaining intravenous access, or prior adverse events related to sedatives-anesthetics.

Antenatal history should be reviewed, as maternal medical conditions or complications may affect the neonate adversely. Determine gestational age and conceptional age—premature infants may have pulmonary, cardiovascular, neurologic, gastrointestinal, and hematologic conditions that may lead to decompensation during sedation.

Elicit a history of prior sedation-anesthesia and any known adverse reactions, such as marked nausea, vomiting, increased or decreased sensitivity to sedatives or analgesics, and/or prior need for intervention during sedation or unexpected hospitalization after procedures. The complete list of current medications and allergies should be carefully documented.

Confirming NPO status is important: Children can never be trusted to have fasted. The child and parent should be carefully questioned about any recent intake by mouth, however trivial it may seem.

Physical Examination

The initial physical examination provides the sedation practitioner with an opportunity to become familiar with the patient's baseline physiologic status. **Perform a targeted physical examination, including airway assessment, respiratory status, and volume status**. Some children will present with a syndrome that the parents do not disclose, either because they assume you are aware or for personal reasons; in these cases, tactfully ask about any special needs. Specific syndromes may be recognized by unusual features, many of which appear as a constellation of associated findings. Inquire as to what extent the child is affected by the syndrome and his current functional status.

Fasting Guidelines and Sedation

Although the presence of gastric contents theoretically increases the risk of aspiration pneumonia, **there is no known gastric fluid volume (GFV) that places a particular patient at clinically relevant risk or that eliminates all risk** [1]. The traditional teaching is that the risk of aspiration increases with gastric acid volume greater than 0.4 mL/kg and a pH of less than 2.5 [2]. However, if these threshold values were applied, a great number of appropriately fasted patients would be classified as at risk for aspiration. **That is, the stomach is rarely completely empty**—even in the fasted state—given ongoing salivary (1 mL/kg/h) and gastric (0.6 mL/kg/h) secretions [3]. The provider may expect GFV to be minimal in most fasting patients, but some patients may have large residual GFV despite having followed traditional fasting guidelines (Fig. 4.1). Prolonged fasting in children is not entirely benign: The fasting child is always at risk for hypoglycemia and/or hypovolemia. Optimize your patient's volume and metabolic status before the procedure with the appropriate intravenous fluids if needed. Due to high metabolic needs, an infant should be offered clear fluids until 2 h before sedation.

There is a presumption that the relative risk of aspiration is lower during sedation than under general anesthesia, and that protective airway reflexes are retained fully during sedation. It is important to note that the progression from mild sedation or analgesia to general anesthesia represents a continuum not easily divided into discrete stages [4]. **Anyone receiving moderate or deep sedation should be treated similarly to those receiving general anesthesia because the sedation level can change rapidly and deepen subtly with subsequent impairment of airway reflexes.**

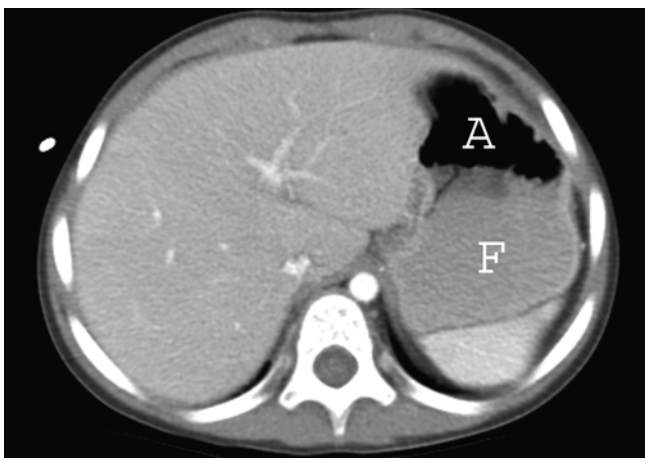

Fig. 4.1 CT of the abdomen without administration of oral contrast in a fasting 2-year-old child in supine position. CT shown in axial (A) plane. Note fluid (labeled "F") and air (Labeled "A") in distended stomach. Measured volume of fluid in stomach was 41.8 mL (3.3 mL/kg). Courtesy of Mohamed Mahmoud, MD

Although aspiration is a widely feared complication of general anesthesia, fortunately clinically relevant aspiration in modern anesthesia practice is exceptionally rare in pediatrics. The incidence is estimated to be 1 in 10,000 to 10 in 10,000, with the wide reported range likely due to variation in research methodologies, definitions, and reporting sensitivities [5]. In those undergoing general anesthesia, approximately two-thirds of aspiration occurs during manipulation of the airway (endotracheal tube placement and removal) [6]. The multicenter Pediatric Sedation Research Consortium collected data on 49,836 propofol sedations in children: Aspiration during sedation occurred four times (0.04 %) [7]. A retrospective study by Sanborn et al. of 16,467 sedations during imaging procedures in children using chloral hydrate, midazolam, fentanyl, or pentobarbital found 70 (0.4 %) respiratory incidents; only two patients of 16,467 aspirated (0.012 %) [8].

The low incidence of aspiration pneumonia with sedation and anesthesia may be attributed to the fact that the stomach is very distensible and can accommodate a large volume before resting intragastric pressure rises [9]. Intragastric pressure must exceed the barrier pressure of the lower esophageal sphincter (LES) for regurgitation to occur. The barrier pressure of the LES does not appear to be as easily overcome under general anesthesia as is widely believed [9].

The **American Society of Anesthesiology's (ASA) Task Force on Fasting** has published consensus guidelines for elective anesthesia: clear fluids, 2 h; breast milk, 4 h; formula, 6 h; and solids, 8 h [10]. These guidelines are intended for healthy patients of all ages undergoing elective procedures; they are not intended for patients with coexisting diseases or conditions that may delay gastric emptying such as diabetes, hiatal hernia, gastroesophageal reflux, or bowel obstruction. The ASA acknowledges that there is insufficient evidence to codify preoperative fasting times. In addition, the task force does not offer specific guidance for fasting times for emergency procedures.

When practitioners formulate a plan for sedation for emergency procedures in children who have not fasted, the risks of sedation and the possibility of aspiration must be balanced against the benefits of performing the procedure emergently. **The American College of Emergency Physicians (ACEP)** Clinical Policy on Sedation assesses risk based on the nature of last oral intake and the urgency of the procedure (Table 4.1) [11]. In this setting, aspiration has been found to be very rare among patients sedated in an emergency room setting for procedures, regardless of fasting status [12].

There is an ongoing debate regarding the **administration of oral contrast for Computerized Tomography (CT) prior to sedation**. The administration of oral contrast less

Table 4.1 Prudent limits of targeted depth of ED procedural sedation

STANDARD RISK				
ORAL INTAKE IN THE PRIOR 3 HOURS	**Urgency of the Procedure**			
	Emergent	***Urgent***	***Semi-Urgent***	***Non-Urgent***
Nothing	All levels of sedation	All levels of sedation	All levels of sedation	All levels of sedation
Clear liquids only	All levels of sedation	All levels of sedation	Up to and including brief deep sedation	Up to and including extended moderate sedation
Light snack	All levels of sedation	Up to and including brief deep sedation	Up to and including dissociative sedation; non-extended moderate sedation	Minimal sedation only
Heavy snack or meal	All levels of sedation	Up to and including extended moderate sedation	Minimal sedation only	Minimal sedation only
HIGHER RISK				
ORAL INTAKE IN THE PRIOR 3 HOURS	**Procedural Urgency**			
	Emergent Procedure	***Urgent Procedure***	***Semi-Urgent Procedure***	***Non-Urgent Procedure***
Nothing	All levels of sedation	All levels of sedation	All levels of sedation	All levels of sedation
Clear liquids only	All levels of sedation	Up to and including brief deep sedation	Up to and including extended moderate sedation	Minimal sedation only
Light snack	All levels of sedation	Up to and including dissociative sedation; non-extended moderate sedation	Minimal sedation only	Minimal sedation only
Heavy snack or meal	All levels of sedation	Up to and including dissociative sedation; non-extended moderate sedation	Minimal sedation only	Minimal sedation only
Procedural Sedation and Analgesia Targeted Depth and Duration				
← Increasing Potential Aspiration Risk ←	Minimal sedation only			
	Dissociative sedation; brief or intermediate-length moderate sedation			
	Extended moderate sedation			
	Brief deep sedation			
	Intermediate or extended-length deep sedation			

Modified with permission from Green SM, Roback MG, Miner JR, Burton JH, Krauss B. Fasting and Emergency Department Procedural Sedation and Analgesia: A Consensus-Based Clinical Practice Advisory. Ann Emerg Med. 2007; 49(4): 454–461
Brief: <10 min
Intermediate: 10–20 min
Extended: >20 min

than 2 h before sedation-anesthesia is at odds with elective NPO guidelines, and in theory would increase the risk of aspiration pneumonia. Sedation practitioners are asked to make an exception to the fasting guidelines and permit the use of enteric contrast material with CT in order to obtain an accurate study. There does not appear to be a perfect resolution to this issue, since waiting several hours after administration of contrast often results in inadequate opacification of the small bowel and a poor study [13].

Small bowel transit time can be as rapid as 15 min and on average is 1 h 24 min [14]. In one study, in 83 % of cases small bowel transit time was less than 2 h [14]. Inadequate opacification of the small bowel can lead to lack of distinction between small bowel loops and fluid collections or masses [13].

At one author's institution, administration of contrast begins 2 h before and ends 1 h prior to anesthesia-sedation. The challenge lies in balancing technical factors governing the image quality of the study with safety concerns related to sedating a child with a potentially full stomach for an elective CT. A recent retrospective chart review concluded that administering oral contrast material within 2 h of propofol sedation for abdominal CT in children appears to be relatively safe. The data sample, however, was small relative to the reported incidence of aspiration in the literature [15]. Currently we are not aware of any clear consensus among institutions that care for these patients. Some clinicians may choose to perform rapid sequence induction of general anesthesia with endotracheal intubation while others may choose deep sedation without definitive airway protection. Others may negotiate with radiologists to have the oral contrast given 2 h before the study or administered through an oral gastric tube after placement of an endotracheal tube [16, 17].

When Not to Proceed

Barring emergent or life-threatening circumstances, situations arise in which—despite pressure from consultants, providers, and/or families—the practitioner should forgo sedation outside of the operating room for a more opportune time, setting, or facility. Proper monitoring, rescue equipment, and sufficient staff should be in place. The provider should use sound clinical judgment before proceeding, informed by the patient's risk for complications and the urgency of the procedure, as well as practical concerns such as the ability to dedicate the necessary time, attention, and human resources to the endeavor. The following section is a broad overview that will address specific safety considerations and focused assessments in important special populations.

Preparation for and Considerations in Special Populations

Asthma and Reactive Airway Disease

The child who wheezes presents a common challenge to the sedation practitioner. *Transient wheezers* are infants whose symptoms are provoked by an active viral respiratory infection. These children typically "outgrow" their reactivity within the first few years of life. After the toddler and preschool period, *non-atopic wheezers* continue to experience wheezing with active viral illnesses, but are not likely to develop lifelong symptoms. Both transient and non-atopic wheezers tend to have mild reactions to the inciting event. *Atopic wheezers* are equally sensitive to viral illnesses, but often also suffer from allergy, allergic rhinitis, and atopic dermatitis. These children are at highest risk for severe and persistent symptoms exacerbated by a variety of infectious and/or environmental factors [18].

The diagnosis of asthma is difficult to make under the age of 6, since there is significant overlap with reactive airway disease and pulmonary function tests are problematic in young children. In those with an established diagnosis of asthma, the assessment of symptoms follows a step-wise approach (Table 4.2).

In addition to the assessment of severity of symptoms, confirm the overall control of symptoms and what level of therapy the child is currently receiving. It is also helpful to ascertain the responsiveness that the child has shown to previous exacerbations [19]. This is especially important in the planning of procedures that involve airway stimulation or those that would require frequent suctioning.

Children with a history of either reactive airway disease or diagnosed asthma are at risk for **bronchial hyperreactivity** (40 % of school-aged children with asthma) [20]. Bronchial hyperreactivity may persist for weeks after an exacerbation. For this reason, a careful history of recent illness, changes in medication, and history of hospitalization are important in all children with a history of wheezing. In general in children with stable and controlled asthma or reactive airway disease, the peri-procedural risk for bronchospasm is low and is not associated with a significant morbidity [21].

A recent prospective study found that patient factors (readily known on pre-procedural assessment) such as active respiratory symptoms, eczema, family history of asthma, rhinitis, or exposure to tobacco smoke were associated with an increased relative risk of peri-procedural respiratory adverse events such as airway obstruction, oxygen desaturation (<95 %), and severe or sustained cough [22]. In patients with active symptoms, the practitioner should determine the *severity of illness* and weigh this with *the urgency and importance* of

Table 4.2 Asthma severity assessment in children older than 5 years of age

Clinical features	Mild intermittent asthma	Mild persistent asthma	Moderate persistent asthma	Severe persistent asthma
A. Symptoms: wheezing, coughing, chest tightness	Symptoms ≤2 times/week	Symptoms >2 times/week but <1 time/day	Daily symptoms	Continual symptoms
	Asymptomatic between brief exacerbations		Exacerbations 2 or more time/week; may last days	Frequent exacerbations
B. Activity limitations	No activity limitations	Activity may cause exacerbations	Activity causes exacerbations	Limited physical activity
C. Nocturnal symptoms	≤2 times/month	>2 times/month	>1 time/week	Frequent nighttime symptoms
D. Lung function	PEF or $FEV_1 \geq 80$ % of predicted or personal best	PEF or $FEV_1 \geq 80$ % of predicted or personal best	PEF or $FEV_1 > 60$ % and <80 % of predicted or personal best	PEF or $FEV_1 \leq 60$ % of predicted or personal best

Modified from: National Heart, Lung, and Blood Institute: National Asthma Education and Prevention Program. *Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma*. 2007

the procedure. The actively wheezing patient should have his current illness addressed immediately, and if the procedure is to go forward, a plan for pre-, intra-, and post-procedure treatment should be formulated to anticipate and manage potential complications such as bronchospasm.

Autism, Developmental Delay, and Intellectual Disability

Autism spectrum disorders (ASD) are characterized by neurodevelopmental impairments in three major domains**: behavior, communication, and socialization** [23]. Although the rate of diagnosis of ASD has markedly increased recently, its pathogenesis is incompletely understood; the current consensus is that autism has a genetic basis with possible contributing environmental factors. Approximately 40–62 % of children with ASD demonstrate some learning disability [24].

Children with intellectual disability, developmental delay, or ASDs require a holistic view in preparation for sedation. Caretakers are typically very helpful in sharing the child's past reactions to the procedure, and may be vocal in their preferences in the timing, type, and route of administration of sedatives. The practitioner would do well to consider the caregivers' experience with their child and weigh this with the practicalities and requirements of the procedure at hand.

These children may exhibit challenging behavior, especially when anxious or stressed, such as punching/slapping/pulling (50 %) or kicking (24 %) [25]. Boys and adolescent males form the majority (66 %) of children with challenging behavior [26]. These behaviors may be exacerbated by frequent and sometimes unpleasant interactions with the health care system. **Observing the child while nonstressed during the pre-sedation assessment may help to reveal caregiver-patient dynamics as well as to inform the clinician of how best to keep him calm and cooperative**. Non-pharmacologic methods such as distraction, storytelling, watching videos, or playing games are particularly helpful in this setting and during the induction/pre-procedural period. (Refer to Chap. 34.)

Intellectual, developmental, and learning disabilities are not a specific medical condition, but rather manifestations of neurologic disease. It is important to note that **comorbidities are common**, such as epilepsy (44 %), psychiatric disorders (50 %), and gastroesophageal reflux (49 %) [24]. The pre-procedural assessment should include a review of medical conditions, frequency and control of symptoms, and current medications.

A small observational study found that as a group, children with developmental delay (given the prevalence of substantial neurologic comorbidities) may have a smaller airway diameter at the level of the soft palate when sedated for magnetic resonance imaging (MRI). The authors' findings were thought to be multifactorial: anatomic (different airway shape), physiologic (abnormal airway tone), and pharmacologic (increased susceptibility to sedative) [27]. In this light, concurrent illness such as viral respiratory symptoms should be considered carefully in these patients.

If the child requires pretreatment, one may start with noninvasive approaches such as the oral route for pre-sedation, the intranasal route to facilitate IV access if needed, and the intramuscular route if necessary. Nitrous oxide, if available, may be a good choice if the child sees the device as a novelty or game, rather than as a restraint. Close attention to risk factors for pre-procedural anxiety or behavioral challenges is important, as these are associated with post-procedural delirium and maladaptive behaviors, which complicate the feasibility of a successful outpatient visit [28].

Anticipating behavioral disruptions and having a ready plan for escalation of treatment are essential. Discussion with the caregiver before the procedure may help to decrease his or her anxiety, allowing for a capable, present, and calm ally in the endeavor. This includes the timing and threshold

for physical restraint if needed, based on the urgency and nature of the procedure. A brief pre-sedation "team huddle" with caregivers and staff to review the sedation plan may promote a smooth procedure and help to avoid injury to the patient, parents, practitioner, or staff.

Bronchopulmonary Dysplasia

Bronchopulmonary dysplasia (BPD) is the most common cause of chronic lung disease in infants. It affects premature infants who survive the acute phase of respiratory distress syndrome and is characterized by the need for supplemental oxygen beyond 4 weeks of life. BPD is thought to develop after prolonged periods of mechanical ventilation and exposure to high concentrations of inspired oxygen. Other proposed pathophysiologic mechanisms include initial volume overload, increased pulmonary blood flow, and generalized inflammation. These patients typically have decreased lung compliance, airway hyperactivity, lung hyperinflation, rapid respiration, wheezing, cough, and frequent episodes of fever, desaturation, hypercarbia, abnormal functional airway growth, and increased risk for bradycardia and congestive heart failure (CHF) [29].

Implications of BPD in sedation-anesthesia include tracheomalacia, tracheal granuloma, subglottic stenosis, increased airway reactivity and bronchospasm, and diuretic-induced electrolyte disorders. **Adequate pre-procedure preparation should focus on optimizing oxygenation, reducing airway hyperactivity, and correcting electrolyte abnormalities.** These children require special attention to fluid balance with careful titration of fluids during the procedure. A laryngeal mask airway (LMA) is less irritating to both the upper and lower airways; it may offer some advantage in reducing the incidence of post-procedural coughing, wheezing, and hoarseness compared to endotracheal intubation in these patients.

Cerebral Palsy

Cerebral palsy (CP), a nonprogressive, permanent disorder of motor function and posture, is the most common physical disability in childhood, occurring in 2–2.5 in 1,000 births [30]. The majority of cases are of unknown etiology. Known associations are multifactorial: prematurity (78 %), intrauterine growth restriction (34 %), intrauterine infection (28 %), antepartum hemorrhage (27 %), and maternal alcohol use (threefold increased risk) [31, 32]. One in four have epilepsy and one in five have a sleep disorder [33].

The spectrum of disease varies from mild focal weakness with normal intelligence to total body spasticity and severe intellectual disability. CP may be classified by the predominant motor component: **spasticity, ataxia, or dyskinesia** [34]. Medical therapy emphasizes control of spasticity with medications, injections, or surgery. In the pre-sedation assessment, the type, dosage, and route of medications are important especially if there will be prolonged fasting. The clinician should determine the presence (and recent setting changes) of an intrathecal pump. Although rarely an issue, children with recent Botulinum toxin type A injection (for local control of spasticity) if unwittingly overdosed may later experience relative respiratory muscle weakness, which may be exacerbated during sedation [35].

Common comorbidities such as scoliosis, gastroesophageal reflux, decubitus ulcers, and skin infections should be assessed for control of disease. This will help in planning for successful positioning (to optimize ventilation and comfort), IV access, and ready access to the airway if advanced measures are needed during the procedure. Children with CP often have considerable drooling due to difficulty in swallowing secretions; **plan for frequent suctioning**. Atropine or glycopyrrolate may be considered for their antisialagogue effect, but they may also thicken lung secretions and potentially increase the risk of lung infection in CP patients [34].

Part of the pre-sedation assessment is anticipating and avoiding pitfalls in the care of children with CP. **Chronic low fluid intake and relative malnutrition put the child at risk for pre-renal failure and the development of pressure ulcers**. Careful attention to fluid replacement (especially during prolonged fasting periods) and proper positioning of the patient during the procedure will help to attenuate these risks. Other common challenges are the presence of extremity casts that may obscure blood loss (from trauma or the procedure itself) or developing compartment syndrome from malpositioning.

Pain control in intellectually disabled children is an important issue. Clinician understanding of the analgesic needs of these children is changing, and there is evidence to suggest that they may, in fact, be more sensitive to pain than non-disabled children [36]. Unfortunately, these vulnerable children are often undertreated due to barriers in communication or misinterpretation of behaviors [37]. Children on chronic opioids may have 30–100 % higher dosage requirements than opioid-naïve children [38]. Control of symptoms should begin early in the visit to promote a successful procedure and post-procedure course.

Congenital Heart Disease

Congenital heart disease (CHD) occurs in approximately 8 in 1,000 live births [39]. The most common acyanotic

Table 4.3 Classification systems of heart failure [45, 46]

Class	NYHA classification	Ross classification
I	No symptoms	No limitations or symptoms
II	Symptoms with moderate exertion	Mild tachypnea or diaphoresis with feeding in infants; dyspnea on exertion in older children
III	Symptoms with mild exertion	Marked tachypnea or diaphoresis with feeding or exertion
IV	Symptoms at rest	Symptomatic at rest with tachypnea, retractions, grunting, or diaphoresis

Table 4.4 Heart failure staging for infants and children [43]

Stage	Interpretation
A	Increased risk of developing heart failure, but with normal cardiac function and size
B	Abnormal cardiac morphology or function, with no heart failure symptoms or history of symptoms in the past
C	Underlying structural or functional heart disease and heart failure symptoms past or present
D	End-stage heart failure

Table 4.5 Cardiac conditions associated with the highest risk of adverse outcome from endocarditis for which prophylaxis with dental procedures is reasonable [48]

Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
Previous infectious endocarditis
Unrepaired cyanotic CHD, including palliative shunts and conduits
Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure
Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
Cardiac transplantation recipients who develop cardiac valvulopathy

lesion is a ventricular septal defect; the most common cyanotic lesion is the tetralogy of Fallot. Although lesions may be classified as acyanotic or cyanotic and/or ductal dependent or not, the clinician may risk stratify based on whether the child has been fully repaired or whether his lesion involves palliation. That is, a child with a repaired ventricular septal defect and normal baseline oxygenation may have no long-term sequelae relevant to sedation while a child with single ventricle pathology, a palliative shunt (e.g., hypoplastic left heart syndrome status-post Fontan procedure), or baseline low oxygen saturation requires a more judicious approach.

Children with cyanotic disease with or without palliative surgery are very sensitive to changes in volume status, as many are pre-load dependent. In addition, certain lesions are more prone to dysrhythmias [40]. Their low baseline oxygen saturations offer little to no reserve in times of stress. For this reason and in general, **children with cyanotic heart disease are poor non-emergent outpatient candidates for sedation beyond mild anxiolysis** [40–42].

Although each lesion has a unique set of considerations in the pre-sedation assessment, current functional status is most informative of appropriateness for sedation outside of the operating room. Children with CHD (both cyanotic and acyanotic lesions) often develop some degree of CHF. The New York Heart Association (NYHA) classification was originally designed for adults, and is often applied to children (Table 4.3) [41]. The Ross classification was designed specifically for children and mirrors the NYHA classification [43]; recently a detailed age-specific modification to the Ross classification has been proposed [44].

Both the NYHA and the Ross classifications assess current symptoms; neither discriminates well in the early stages of disease. Since overt heart failure symptoms are a late sign in children (due to compensatory mechanisms), and the sedating clinician is interested in detecting subtle risk factors, an updated heart failure staging classification has been proposed (Table 4.4).

Stages A and B correspond to NYHA I, and stage C corresponds to NYHA II and III. Stage D patients typically require inotropic and/or ventilator support. In addition to the above, the assessment should include the child's general health and change in behavior, oral intake, or urine output. A recent cough or taking longer to feed may be subtle alerts to hypervolemia and poor control of CHF. On examination, infants may be in mild to moderate respiratory distress and/or have evidence of hepatic engorgement, a sign of right-sided heart failure (*N.B.* peripheral edema as seen in adults in CHF is rare in children).

Recent illnesses, especially upper respiratory tract infections (URIs), are especially important to note in these children, as airway reactivity and changes in pulmonary vascular resistance are not well tolerated in children with CHD. A thorough review of previous surgeries and complications, current medications, and drug allergies is required. Anticoagulants may need to be held for the procedure in consultation with the child's cardiologist. The presence of an implantable cardiac defibrillator or pacer should be determined and recent changes or complications noted [47].

Prophylaxis for bacterial endocarditis is recommended for all dental procedures only in children with high-risk historical features (Table 4.5). In eligible children, it is reasonable to give prophylaxis for procedures on the respiratory tract, infected skin, or musculoskeletal tissue. Prophylaxis is no longer recommended for gastrointestinal or genitourinary procedures.

Cystic Fibrosis

Cystic fibrosis (CF) is the most common fatal inherited disease in Caucasians, and exists in smaller frequencies in

other racial groups [49]. The basis of its pathophysiology is a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) protein, a chloride channel found in all exocrine tissues. As such, **CF is a multi-organ system disease**, involving impaired lung function, pancreatic insufficiency and diabetes mellitus, hepatobiliary disease and cirrhosis, bone disease, and genitourinary disease. Pulmonary complications account for over 90 % of the morbidity and mortality in CF patients [50].

CF demonstrates a spectrum not only in terms of organ systems involved but also in severity of disease burden in the individual patent [51]. For this reason, the pre-sedation assessment should include pointed questioning about the child's frequency of illness, strength of cough, amount of sputum produced, airway reactivity, and history of recovery from procedures and illnesses. A thorough review of current therapies and recent acceleration of treatment may reveal the child's current trajectory of disease.

Younger children with CF have more reactive airways, which may respond to β(beta)-agonists. **It is important to note, however, that older children may have worsening expiratory airflow with the use of bronchodilators**. This is due to progressive damage to cartilaginous support in the lower airways; bronchial muscle hypertrophy may in fact help to "stent" the airways open [52]. In these patients, bronchodilators may result in "floppy" lower airways, and impaired gas exchange. A careful history regarding response to β(beta)-agonists is important to anticipate and avoid intra-procedure complications.

In addition to acute exacerbations and worsening lung infections, children with CF are at risk for apical blebs (up to 3.4 %) that may cause spontaneous pneumothorax [50]. Planning for sedation of a child with CF should include preparation for the management of this complication, such as oxygen therapy, IV catheters for decompressive thoracostomy, and a plan for emergent definitive chest tube thoracostomy. Chronic lung disease may manifest in chronic hypoxia and hypercarbia with resulting increases in pulmonary vascular resistance and pulmonary hypertension. An electrocardiogram (ECG) with evidence of *cor pulmonale* is an ominous sign [53].

Control of diabetes mellitus, if present, should be addressed. The presence of liver disease should be noted, as hepatic clearance of medications may be enhanced in early disease and impaired with the onset of cirrhosis; liver function tests are unreliable in this context [54]. Older CF patients may develop distal intestinal obstruction syndrome (DIOS) in the colon and ileum, mimicking medical and surgical causes of nausea, vomiting, abdominal pain, and distention [55]. Volume depletion, chronic narcotics, and medication nonadherence put the patient at higher risk [50].

If possible, a review of the medications given during previous procedures may be helpful in planning for sedation. Patients with CF may have higher opioid and benzodiazepine requirements than patients without CF [56]. Plan to balance titrating to effect with possible impairment of overall oxygenation and ventilation during the procedure.

Diabetes Mellitus

Type 1 (insulin-dependent) diabetes mellitus (DM) accounts for over 90 % of DM cases in children [57]. Early onset of type 2 (non-insulin-dependent) DM is rising with obesity rates in children [57]. Other less common causes of DM in children include maturity onset diabetes of youth (MODY), insulin resistance syndromes (idiopathic), genetic syndromes (chromosomal abnormalities, congenital disorders of the pancreas), and secondary diabetes (e.g., drugs such as corticosteroids) [58].

The clinician should gain a general view of the patient's overall diabetes control and any recent change in regimen. A thorough account of the child's medications (e.g., insulin, oral hypoglycemic sulfonylureas, oral biguanide) and timing of the last dose should be reviewed. Patients may have taken a recent dose of medication, only to be unexpectedly fasting during the visit. Physical exam should pay close attention to volume status, as these children are at risk for hypovolemia. If an insulin pump is found, the silastic catheter may be removed before the procedure to ensure that ongoing insulin is not administered to the fasting child. A baseline fingerstick blood glucose will be helpful in the initial assessment.

Regardless of the type or current control of the patient's diabetes, **the overall goal during sedation is to avoid hypoglycemia and excessive hyperglycemia** [58, 59]. When appropriate, IV fluids may be given, and if the procedure is prolonged, supplemental glucose with frequent fingerstick blood glucose monitoring. Case reports demonstrate the importance of glucose monitoring in DM patients undergoing sedation: hypoglycemic coma may be confused for deep or prolonged sedation [60].

Endocrinopathies

Knowledge of the normal anatomy and physiology of the endocrine glands is essential in understanding their potential pathophysiologic effects relevant to procedural sedation. In this section we will outline the considerations for sedating a child with adrenal insufficiency, hypothyroidism, hyperthyroidism, or diabetes insipidus (DI).

The adrenal cortex synthesizes and secretes steroid hormones (glucocorticoids, mineralocorticoids, and sex steroids) that are essential to life. Glucocorticoids (especially cortisol) play a critical role in the body's response to stress and play an important role in maintaining vascular tone.

Causes of adrenal insufficiency can be classified as primary (adrenal gland dysfunction), secondary (the pituitary gland dysfunction), or tertiary (hypothalamic dysfunction). The most common cause of adrenal insufficiency is long-term administration of exogenous glucocorticoids via oral, intravenous, inhaled, intranasal, or topical routes. Even a short course (5 days) of prednisone mildly suppresses the hypothalamic–pituitary–adrenal axis for 5 days after discontinuation (usually without clinical sequelae in the healthy patient). Long-term glucocorticoid use produces adrenal cortical atrophy as a result of chronic suppression of ACTH production, requiring variable recovery times of up to 1 year [61].

The practice of providing perioperative glucocorticoid replacement therapy to patients with adrenal insufficiency is well established. Insufficient levels of cortisol can be produced in response to stress in these patients, posing the risk of acute adrenal crisis with hypotension and cardiovascular collapse.

Peri-procedural stress dosing depends on the duration and invasiveness of the procedure. Most elective minor procedures and noninvasive diagnostic studies do not warrant supplementation with additional glucocorticoids. A continuation of the current dose of corticosteroids is sufficient to maintain cardiovascular function in patients who receive long-term administration of exogenous glucocorticoids [62]. It is extremely important to note that **primary hypopituitarism is a condition that always requires peri-procedure steroid supplementation** regardless of the daily dose taken. Parenteral cortisol (e.g., Solu-Cortef) at a dose of 0.5–1 mg/kg every 6 h is recommended for perioperative, intensive care, or emergency department indications for up to 72 h [63].

Thyroid hormones are integral to the normal physiology of every organ system of the human body, playing a crucial role in regulating myocardial function, pulmonary ventilation, energy homeostasis, vascular tone, water and electrolyte balance, and normal function of the central nervous system. **The most important adverse effects of hypothyroidism include impaired cardiac contractility with decreased cardiac output, increased peripheral vascular resistance, and decreased blood volume and peripheral oxygen consumption.**

A detailed history should be obtained from the patient or the family about prior thyroid disease, thyroid surgery, radiation therapy (radioactive iodine or neck irradiation), treatment with any thyroid medications, or family history of thyroid disease. Physical examination is equally important. Dry skin, a slowed deep tendon reflex relaxation phase, bradycardia, and hypothermia are all signs of clinical hypothyroidism. Children with known hypothyroidism have increased sensitivity to anesthetic-sedative agents; these children should have documented normal thyroid function tests before elective procedures.

Hyperthyroidism is less common in children than hypothyroidism and is most commonly caused by Graves disease. The classical features of thyrotoxicosis include hyperactivity, weight loss, tremor, heat intolerance, dyspnea, insomnia, diarrhea, and nervousness. Cardiovascular effects of hyperthyroidism include palpitations, tachycardia, atrial fibrillation, and congestive cardiac failure. **Thyroid storm can be lethal.** Fortunately, it is rarely seen due to widespread use of antithyroid drugs. In an attempt to prevent this catastrophic complication, **these children should be euthyroid before the procedure**. Thyroid storm responds to symptomatic treatment including parenteral β(beta)-blockers and propylthiouracil.

The clearance and distribution volume of propofol are increased in hyperthyroid patients. When total intravenous anesthesia is used, propofol infusion rates should be increased to reach anesthetic blood concentrations [64].

Optimal anesthetic-sedative care of patients with history of DI requires an understanding of the complex pathophysiology of this disease. Arginine vasopressin (AVP) is produced within the hypothalamus, and it is normally stored for release in the posterior pituitary gland. After its release, AVP acts on V2 receptors in the collecting tubules of the nephron in order to allow for effective urine concentration.

DI is a syndrome manifested by high output urine, low urine specific gravity (<1.005), high plasma osmolality (>200 mOsm/L), and high plasma sodium (>150 mEq/L). **Nephrogenic DI** occurs when the kidney is unable to control plasma osmolality due to a defect in the action of AVP. Medications such as demeclocycline, lithium, amphotericin B, and fluoride [5], and electrolyte abnormalities such as hypokalemia and hypercalcemia [6] are known to cause or precipitate nephrogenic DI. **Central DI** occurs due to destruction of the posterior pituitary and eventually lack of AVP production or release. Without treatment, intravascular volume depletion occurs, cardiac stroke volume decreases, and eventually heart rate increases. These patients will have orthostatic hypotension, weak pulses, rapid breathing, and decreased level of consciousness. They may present with seizures if significant hypernatremia is present.

A child undergoing procedural sedation should receive his usual morning dose of desmopressin. The sedation provider should pay attention to fluid management in the patient on desmopressin therapy, as some degree of fluid restriction is required. Intravenous fluids (use 5 % dextrose-0.9 % saline) should total 1 L/m^2/24 h to approximate insensible losses and obligate urine output. Oral fluids may be offered once the child is awake.

Mitochondrial Disease

Mitochondrial disease (MD) is a group of disorders that arise from defects in the oxidative phosphorylation or electron transport chain involved in generation of ATP [65]. Primary

mitochondrial disorder is caused by deletions in nuclear DNA or mitochondrial DNA. Secondary disorders are due to mitochondria dysfunction caused by various drugs and by free radicals.

The ten most common syndromes associated with MD are: Kearns-Sayre syndrome; Leigh syndrome; mitochondrial DNA depletion syndrome; mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS); myoclonic epilepsy with ragged red fibers; neurogastrointestinal encephalomyopathy, neuropathy, ataxia and retinitis pigmentosa (NARP); and external ophthalmoplegia. There is no definitive treatment for MD, although some patients improve with specific therapies such as coenzyme Q10; those with seizures may respond to a ketogenic diet.

MD may present with any symptom in any organ at any age, but some symptoms and signs are more suggestive of a mitochondrial disorder than others. These red-flag features require the initiation of a diagnostic evaluation for mitochondrial disease (Table 4.6).

Sedating-anesthetizing children with MD may perplex many practitioners. Currently there is no clear evidence-based guidance in the literature regarding the anesthetic-sedative management of these patients. Complicating matters further is the risk of clinical deterioration related to the stress of the procedure itself, unrelated to nature of the anesthetic-sedative agents used. It is well known that **children with mitochondrial defects (MD) may have an increased risk for cardiorespiratory and neurological and metabolic complications from anesthesia-sedation**. Any organ may be affected in MD: meticulous individualized pre-sedation assessment is essential. Sedation providers should review and consider obtaining complete blood count, basic metabolic panel, liver function tests, thyroid function tests, sleep studies, and ECG and/or echocardiogram as indicated by the patient's condition and the associated syndrome.

Patients with MD often develop hypoglycemia and lactic acidosis, which can be exacerbated by the stress of the procedure. Hypoglycemia is common: diseased mitochondria cannot keep up with the body's energy requirements via fatty acid oxidation during stress, which leads to drawing on and rapid depletion of carbohydrate stores. Minimizing periods of fasting and routine use of lactate-free intravenous fluids (such as 5 % dextrose-0.9 % saline) in all patients with MD undergoing sedation-anesthesia is recommended. Prolonged procedure time requires lactate and blood glucose monitoring. This is especially important for infants, as glucose is the major energy supply to the myocardium, and hypoglycemia may contribute to myocardial depression.

The prevalence of cardiomyopathy in children with MD is reported to be 20 % [66, 67]. The severity of MD correlates with the severity of impairment of cardiac function. Cardiac impairment occurs in Barth syndrome, Kearns-Sayre syndrome, ocular myopathy, and MELAS. *A pre-procedure baseline ECG is strongly recommended* and can be extremely valuable; red flags in the ECG include any form of heart block or prolonged QT. If the screening ECG is abnormal, a cardiology consult is recommended before proceeding with elective sedation-anesthesia in these patients. For those with cardiomyopathy, an echocardiogram within the past year is recommended.

Table 4.6 Factors that warrant initiation of a diagnostic evaluation in mitochondrial disease

Possible indicators of mitochondrial disease
Neurologic
• Nonvascular pattern for cerebral stroke-like lesions
• Basal ganglia diseases
• Encephalopathy—either recurrent or induced by low or moderate dosing of valproate
• Neurodegeneration
• Epilepsia partialis continua (Kojevnikov's epilepsia)
• Myoclonus
• Ataxia
• Magnetic resonance imaging consistent with Leigh disease
• Characteristic magnetic resonance spectroscopy peaks:
– Lactate peak at 1.3 ppm TE (echo time) at 35 and 135 ms
– Succinate peak at 2.4 ppm
Cardiovascular
• Hypertrophic cardiomyopathy with rhythm disturbance
• In a child: unexplained heart block
• Cardiomyopathy combined with lactic acidosis (>5 mM)
• Dilated cardiomyopathy combined with muscle weakness
• Wolff-Parkinson-White arrhythmia
Ophthalmologic
• Retinal degeneration. May include:
– Decreased visual acuity
– Night blindness
– Deficits in color vision
– Pigmentary retinopathy
• Ophthalmoplegia/paresis
• Disconjugate movement of eyes
• Ptosis
• Sudden-onset or insidious-onset optic neuropathy or atrophy
Gastroenterologic
• Liver failure: unexplained or valproate-induced
• Severe dysmotility
• Pseudo-obstructive episodes
Other red flags
• Newborn, infant, or young child experiencing:
– Unexplained hypotonia
– Weakness
– Failure to thrive
– Metabolic acidosis (particularly lactic acidosis)
• Exercise intolerance disproportionate to weakness
• Hypersensitivity to general anesthesia
• Acute rhabdomyolysis

Adapted from [166]

There is no absolute contraindication to any particular anesthetic-sedative agent for patients with MD. Many anesthetic agents adversely affect mitochondrial function in vitro but adverse events in vivo are only sparsely reported. Furthermore, the anesthetic agents implicated in these cases have been used without incident in many other reports. Opioids, ketamine, midazolam, and dexmedetomidine do not appear to inhibit mitochondrial function. At the present time there is no need to avoid volatile agents in patients with MD; **inhalational anesthetics have been used without ill effects in these children.** Keep in mind that patients with MD may have impaired upper airway and respiratory response to hypoxia and hypercarbia. Sedative agents should be titrated carefully in order to avoid respiratory depression.

Patients with MD may be more susceptible to the effects of lipophilic agents such as propofol. Propofol uncouples oxidative phosphorylation in mitochondria and suppresses ATP production by interfering with the electron transport chain [68]. There are cases in which short-term use of propofol has resulted in propofol infusion syndrome (acute bradycardia resistant to treatment and progressing to asystole). These patients may have subclinical forms of mitochondrial disease that are uncovered by the infusion of propofol. Single dose propofol has been used safely in many patients, but the true risk associated with this practice and the safe total dose and duration of infusion is not established. **Since there are many sedative-anesthetic alternatives, it is reasonable to avoid the use of propofol infusion in these patients.**

As in any child with a known myopathy, children with MD are at risk at baseline for rhabdomyolysis. Further, due to abnormal neuromuscular endplates with the subsequent risk of hyperkalemia, a **depolarizing agent such as succinylcholine is contraindicated.** Note also that patients with MD also exhibit variable sensitivity to the non-depolarizing neuromuscular blocking agents. Many report mitochondrial patients' experiencing prolonged neuromuscular block with non-depolarizing neuromuscular blocking agents. Careful titration of neuromuscular blocking agents by twitch monitoring and consideration of administration of reversal agents are recommended.

To summarize, the most important anesthetic-sedative considerations in these patient are: **to maintain normoglycemia and normothermia, to avoid any period of hypoxia, to maintain normovolemia, and to avoid metabolic stresses that can lead to or worsen lactic acidosis.**

Multiple Allergies

The term "drug allergy" is often misused by clinicians and patients to describe any reaction (proven or perceived) to a medication. The preferred general term is *adverse drug reaction*, which encompasses the important subcategories. Three clinically relevant subcategories are: **drug allergy** (reaction resulting from an immunologic mechanism), **drug intolerance** (reaction resulting from non-immunologic and/or unknown reasons), and **pseudo-allergy** (reaction resembling allergy, but with a multifactorial, unknown, or idiosyncratic cause) [69].

It may not be feasible to differentiate the above in the pre-sedation assessment [70]. Allergists suggest referring to these events as **predictable reactions** (drug overdose, side effects, drug–drug interactions) and **unpredictable reactions** (allergy, intolerance, pseudo allergy). Predictable reactions are often benign, and account for approximately 80 % of adverse drug reactions. Unpredictable reactions account for the remaining 20 %, with allergic or pseudo-allergic reactions comprising 5–10 % of adverse drug reactions [69].

Confirming the diagnosis of a drug allergy is not the goal of the pre-sedation assessment; drug provocation testing performed in other settings remains the criterion standard. However, it is important to note that drug allergy is over-diagnosed in children [71]. Although it is prudent to avoid drugs that may have provoked some reaction in the past, when few alternatives remain the clinician should focus on determining the risk and potential severity of unpredictable reactions during sedation. **Type I** allergic reactions are immediate and due to drug-specific antibodies; they require prior exposure and sensitization to the drug. Clinical manifestations include urticaria, angioedema, bronchospasm, and/or anaphylaxis. **Type II** reactions (anti-tissue cytotoxic, e.g., hemolytic anemia or thrombocytopenia) and **Type III** reactions (immune complex, e.g., serum sickness) are readily identified by a history of severe illness or hospitalization. **Type IV** reactions (the most common) are delayed hypersensitivity reactions evolving over hours to days, and often present with maculopapular exanthems (but may also manifest as eczematous, pustular, or bullous lesions) [69].

Documenting the timing, course of the reaction, and likely inciting drug may help the clinician to understand the safety of the use of the proposed medication during the procedure. Electronic medical records may be a good source of information, as many include entries on when the drug was given and the nature of the reaction [72].

Multiple drug allergy syndrome (MDAS) describes a condition in which the patient experiences allergic or pseudo-allergic reactions to related and non-related drugs [73]. Most cases involve urticarial and/or angioedema; however, Stevens-Johnson syndrome and anaphylaxis have been reported. Interestingly, **skin testing in these patients may be negative, even after significant clinical manifestations have been documented**. These patients typically are older, most are adults, and many have multiple comorbidities and a

long past medical history (with many opportunities to become sensitized to many different types of drugs). Information about the pathophysiology of MDAS remains limited, as there is no criterion standard for diagnosis and prospective studies are lacking [70].

Multiple drug intolerance syndrome (MDIS) may be a separate entity from that which is described above. MDIS is defined as a hypersensitivity to three or more drugs that are "chemically, pharmacologically, and immunogenically unrelated, taken on three different occasions, and with negative allergy skin tests" [74, 75]. MDIS patients are also typically older, have anxiety, depressive and/or somatoform symptoms, and are typically convinced that they are allergic to all drugs. These patients often require allergy and psychiatric consultations as an outpatient [76].

In summary, the pre-sedation assessment should focus on true allergic or pseudo-allergic signs or symptoms associated with a particular drug and the severity of the presentation. When in doubt and feasible, the clinician in this setting may avoid the drug altogether. If there is a conflict or no acceptable alternative, a frank discussion about the risks, benefits, and other possible alternatives is needed.

Muscular Dystrophies

The muscular dystrophies (MD) are a group of progressive myopathic disorders characterized by muscle wasting and weakness. The most common are Duchenne and Becker MDs; other types present at different stages in life, with varying degrees of severity and involving different muscle groups: fascioscapulohumeral, limb-girdle, distal, oculopharyngeal, and Emery-Dreifuss [77]. The morbidity of the most common, Duchenne and Becker MDs, involves progressive respiratory failure with recurrent lung infections.

The disease is characterized by severe proximal muscle weakness, progressive degeneration, and fatty infiltration of the muscles. Symptoms typically appear at the age of 2–6 years; delayed walking beyond 15 months of age is a common initial sign. Affected children never run properly and have difficulty climbing stairs; only approximately 10 % manage to jump with both feet together. Many children require the use of a wheelchair by age 12, and may not live past their 20s [77]. **Most MDs involve some degree of cardiomyopathy and all are at risk for heart failure** [78]. Other manifestations include pseudohypertrophy of the calves and markedly elevated creatine kinase levels. The progressive nature of the disorder results in restrictive pulmonary disease, multiple contractures, and scoliosis. Due to advances in medical management, many of these patients may now be expected to live into adulthood.

The pre-procedure assessment should focus on the child's overall function (ambulatory or wheelchair) with careful attention to respiratory toilet. The child with disturbed sleep, nightmares, daytime drowsiness, or early morning headaches may have unrecognized nocturnal hypoventilation. This may be a clue to a recent worsening trajectory of illness and make the child more likely to benefit from noninvasive positive pressure ventilation during sleep or sedation. Worsening respiratory symptoms may preclude outpatient sedation.

Symptoms of dizziness, chest pain, intermittent increased shortness of breath, nausea, and decreased oral intake may be consistent with developing (or worsening) cardiomyopathy. A thorough cardiovascular exam with careful attention to signs of heart failure (hepatic congestion in infants and toddlers, facial and extremity edema in older children; presence of an S3 or precordial heave) is warranted. One-third of these patients have dilated cardiomyopathy by age 14, with nearly all patients developing some degree of cardiomyopathy by age 18. Due to the prevalence of cardiac disorders in these patients, the American Academy of Pediatrics recommends that children with DMD should undergo cardiac evaluation and optimization of cardiovascular status prior to elective anesthesia [79].

While it is important to investigate and optimize cardiovascular status before the elective procedure, these patients can develop complications despite the presence of reassuring pre-procedure tests. Unexplained tachycardia should raise the suspicion of cardiomyopathy. *A pre-procedure baseline ECG* and potentially an echocardiographic assessment (within a year from the date of the procedure) are recommended to optimize cardiac function and avoid a dysrhythmia. A child with **a pre-procedure echocardiogram showing good left ventricular function may not respond adequately to myocardial stress during the procedure**. Some children with particular MDs are at higher risk for dysrhythmias, and require a prophylactic implantable defibrillator [80]. **The severity and progression of skeletal muscular disease may be outpaced by worsening cardiac muscular disease, such as non-ischemic cardiomyopathy** [81].

Another important concern in these patients is careful evaluation of the airway and respiratory apparatus. These patients may have a difficult airway due to a combination of macroglossia, weak upper respiratory muscles, limited cervical spine mobility, and limited mandibular mobility. DMD is characterized by weakness of the diaphragm, intercostal muscles, and the accessory muscles of respiration, resulting in restrictive pulmonary impairment and a progressive decrease in total lung capacity and vital capacity. For patients with declining respiratory function, it may be necessary to prepare for noninvasive ventilation prior to the procedure.

During sedation, patients with MD are at risk for rhabdomyolysis, with subsequent acute renal failure or hyperkalemia. A careful review of the child's past procedures and outcomes is recommended. Ideally the child is euvolemic prior to the procedure; care should be taken for proper positioning and potentially adjusting positions during long procedures to discourage the development of rhabdomyolysis. Keep in mind that **children with MDs are often sensitive to small doses of opioids and sedatives, which may cause a sudden and prolonged apnea** [82]. Plan for minimum presedation and small titratable aliquots.

Controversy exists concerning the role of inhalational anesthetics and succinylcholine in "triggering" rhabdomyolysis or malignant hyperthermia [78, 83–85]. Some experts recommend against their use based on case reports. Many clinicians avoid their use altogether in children with MD. Propofol, dexmedetomidine, and ketamine (among others) have all been used with success in intravenous sedation in these children [78, 86–88]. Nitrous oxide may be considered in children with MD without significant cardiomyopathy or cardiac dysfunction [66].

Musculoskeletal Disorders

Children with musculoskeletal disorders may present repeatedly for diagnostic procedures. These children should be managed with sensitivity. Positioning for the procedure can be challenging, especially in those with limb deformities and contractures. Whenever possible, offer the child a position of comfort and minimize focal pressure during sedation.

Achondroplasia is the most common nonlethal skeletal dysplasia. There are two causes for this disorder: the child has either a de novo mutation of the fibroblast growth factor receptor 3 gene or inherits the disorder from his parents. These patients have midface hypoplasia, a depressed nasal base, small nasal airways, narrow oropharynx, and upper airway muscle hypotonia, which predispose them to development of obstructive sleep apnea (OSA) [89]. They tend to have a large head, a bell-shaped chest, cupping of the ribs, and short arms and legs.

Sedative-anesthetic risks in these patients include a challenging airway and increased sensitivity to sedative-anesthetic agents. Patients with severe kyphoscoliosis and restrictive lung disease may have baseline hypoxemia and low lung volumes, predisposing them to hypoxemia during sedation. Review of CT scans and MRI of the spine is helpful before sedating these children. Hyperextension of the neck should be avoided and special consideration should be taken before manipulating the neck due to the possibility of cervical cord compression [90].

The sedation practitioner must be aware of potential complications when sedating a patient with history of significant scoliosis. The primary aim of pre-procedure evaluation is to detect the presence and extent of cardiac or pulmonary compromise. The earlier the age of onset and the more immature the bone growth at the time the process begins, the worse the disease burden. Children with *idiopathic scoliosis* tend to have less pulmonary embarrassment than children with *neuromuscular scoliosis*, who may have abnormalities in the central control of breathing and impaired airway reflexes. Poor coordination of laryngeal and pharyngeal muscles may result in abnormal control of secretions and inadequate cough, increasing the risk of aspiration.

Respiratory function should be assessed by a thorough history, focusing on functional impairment (exercise tolerance). Physical examination should include a good understanding of vital capacity (review any pulmonary function tests that may be available). If pre-procedure vital capacity is less than 30–35 % of predicted, post-procedure ventilation is likely to be required. Cardiac dysfunction may occur in scoliosis from distortion of the mediastinum; patients may develop cor pulmonale from chronic hypoxemia and pulmonary hypertension. Cardiac studies (ECG, echocardiogram) may be performed as indicated.

Osteogenesis imperfecta (OI) is an inherited disorder of the connective tissue whose primary manifestation is an increased susceptibility to fractures. Patients usually present with growth retardation, multiple fractures, progressive kyphoscoliosis, vertebral compression, megalocephaly, macroglossia, blue sclera, dentinogenesis imperfecta, bleeding diathesis, and temperature dysregulation. Anesthetic-sedative challenges in OI include airway anomalies, chronic lung disease (due to kyphoscoliosis, rib fractures, intrinsic pulmonary hypoplasia, and defective lung collagen), coagulation dysfunction, hyperthyroidism, and an increased tendency to develop peri-procedure hyperthermia [91, 92]. Fractures occur from minor trauma and result in severe deformity of the extremities complicating intravenous access and blood pressure cuff placement [91, 92].

Obstructive Sleep Apnea

OSA is an increasingly recognized disorder in children that can present unique challenges to the sedationist and pose substantial morbidity to the patient. It belongs to the spectrum of anomalies known as sleep-related breathing disorders in which the airway may become completely (as in apnea) or partially (as in hypopnea) occluded despite respiratory effort. These abnormalities lead to abnormal gas exchange resulting in increasing hypoxemia, hypercapnia, and sleep fragmentation. Common clinical manifestations include snoring (pauses and gasps), disrupted sleep, daytime somnolence, and behavioral problems. Systemic manifestations in the cardiovascular, pulmonary, metabolic, and neurologic systems

Table 4.7 STOP-BANG scoring model[a]

S	Snoring: Do you snore loudly (louder than talking or loud enough to be heard through closed doors)?	Yes	No
T	Tired: Do you often feel tired, fatigued, or sleepy during the daytime?	Yes	No
O	Observed: Has anyone observed you stop breathing during your sleep?	Yes	No
P	Blood pressure: Do you have or are you being treated for high blood pressure?	Yes	No
B	BMI: BMI more than 35 kg/m^2	Yes	No
A	Age: Age over 50 years	Yes	No
N	Neck circumference: Neck circumference greater than 40 cm	Yes	No
G	Gender: Male	Yes	No

Reprinted with permission from Mehta PP, Kochhar G, Kalra S, Maurer W, Tetzlaff J, Singh G, et al. Can a validated sleep apnea scoring system predict cardiopulmonary events using propofol sedation for routine EGD or colonoscopy? A prospective cohort study. Gastrointest Endosc. 2013 Nov 9. pii: S0016-5107(13)02407-3

[a]High risk of obstructive sleep apnea: yes to ≥3 questions; low risk of obstructive sleep apnea: yes to <3 questions

occur secondary to recurrent hypoxemia, activation of the sympathetic nervous system, and sleep disruption. There is an **increased incidence of OSA among children with syndromes affecting the upper airway** such as Down syndrome, Treacher Collins syndrome, and Pierre Robin sequence.

A description of symptoms related to OSA, their severity, and provocative and palliative factors should be sought from the parents or caregiver. Ask about a history of snoring, as this is common in children with OSA. Further questioning for paradoxical breathing, episodes of apnea, mouth breathing, behavioral disturbances, and restless sleep alert the clinician to undiagnosed OSA. Observe for failure to thrive, obesity, micrognathia, midface hypoplasia, retrognathia, and macroglossia, all of which are associated with OSA. Interventions during sleep, such as supplemental oxygen, bilevel positive airway pressure (BiPAP), and special positioning aids should be noted. It is important to realize that tonsil size does not predict the presence or severity of OSA [93].

In cases of severe OSA, pulmonary hypertension can develop secondary to pulmonary vasoconstriction with subsequent right ventricular failure and cor pulmonale; fortunately this presentation in children is uncommon. High-risk features for cor pulmonale include signs of right ventricular failure and the presence of severe OSA: patients may experience episodes of desaturation to less than 70 %. These children should have an ECG, echocardiogram, and an evaluation by a cardiologist [94]. A complete metabolic panel helps to determine the degree of chronic hypercarbia, which manifests as a compensatory metabolic alkalosis.

Polysomnography (PSG) is the criterion ("gold") standard for diagnosis and quantification of OSA. PSG includes electroencephalography (EEG), electrooculography, chin-leg electromyography, transthoracic impedance, video recording, oral-nasal thermal sensors, nasal airflow pressure transducer, chest/abdomen plethysmography monitors, pulse oximeter, end tidal or transcutaneous CO_2, and snore microphone. OSA should be differentiated from primary snoring (snoring without hypopnea or apnea). **Central sleep apnea** is characterized by the absence of both airway flow and respiratory effort. Some patients, especially those with neuromuscular conditions, may display mixed sleep apnea (central and obstructive sleep apnea).

The sedation provider must identify which patients are most at risk and who can be managed as an outpatient. PSG provides clues to the severity of the airway obstruction during sleep by noting the lowest oxygen saturation observed, as well as the types of apnea (obstructive, central, or mixed) experienced and the frequency of apnea events. The apnea-hypopnea index (AHI) measures the number of hypopnea/apnea episodes per hour of sleep (the AHI does not take into account duration of the obstructive events). The American Society of Anesthesiologists Task Force on Perioperative Management of Patients with Obstructive Sleep Apnea defines OSA as: mild, with an AHI of 1–5; moderate, with an AHI of 5–10; and severe, with an AHI > 10 [95]. The respiratory disturbance index (RDI) is calculated from all respiratory events (including central apnea) occurring in 1 h. AHI and RDI are sometimes used interchangeably but the bottom line is that they may be used to risk-stratify OSA. Nocturnal oximetry assesses the severity of OSA. Isolated severe desaturation (<80 %) or clusters of desaturation (more than three desaturations <90 %) are considered abnormal.

In 2008, the STOP-BANG questionnaire was introduced and validated as a screening tool to identify OSA in adults (Table 4.7) [96]. This questionnaire consists of eight questions (yes/no answers) that together can total a score from 0 to 8. Chung et al. found that in adults, a high STOP-BANG score (5–8) was predictive of moderate and severe OSA [97]. Cote et al. found that in adults high STOP-BANG scores (3 or greater) were predictive of the need for airway intervention (chin lift, mask ventilation, nasal airway, endotracheal intubation) and oxygen desaturation to <90 % with propofol sedation [98]. This scoring tool has not been validated in

children. Although one question pertains mostly to adults (neck circumference greater than 40 cm), this screening tool may be relevant to predict OSA and sedation-related complications in children. Future studies are needed in order to determine whether there is a predictive application of this questionnaire to extrapolate outcomes and the presence of OSA in children.

Children with OSA are sensitive to respiratory depression by opioids, sedatives, and hypnotics; they are especially vulnerable to the development of upper airway obstruction during sedation-anesthesia [99]. Investigations on the effect of these drugs on airway morphology indicate the pharynx to be a primary site of obstruction during anesthesia [100]. Changes in airway patency in sedation and anesthesia mirror those associated with sleep disordered breathing: increased airway collapsibility due to an increase in closing pressure [101], loss of tonic activity in pharyngeal muscles [102], and failure of coordination of phasic activation of upper airway muscles with diaphragmatic activity [103]. Residual effects of sedatives/anesthetics can lead to similar changes in airway dynamics resulting in significant post-procedure airway obstruction. Recurrent episodes of apnea, hypopnea, desaturation, and hypercarbia that occur during the pre-procedure sleep state are expected to occur in the recovery room, on the ward, and at home.

Sedatives (such as diazepam and midazolam) relax the pharyngeal musculature, causing a reduction of the pharyngeal space [104]. Propofol, barbiturates, opioid analgesics, and sub-anesthetic concentrations of inhalational agents similarly exacerbate upper airway obstruction and increase the risk of respiratory depression and/or apnea [99]. In contrast to other sedatives, dexmedetomidine induces a state that mimics non-rapid eye movement sleep, without significant respiratory depression. These properties make dexmedetomidine an attractive agent for noninvasive procedural sedation in children with OSA [105]. Increasing doses of dexmedetomidine in children without OSA have minimal effect on the upper airway and are not associated with clinical signs of airway obstruction. However, the effect of high doses of dexmedetomidine in children with OSA is unknown [106]. Ketamine is a good alternative: it has been shown to preserve hypopharyngeal caliber in adults [107].

Examination of patterns of dynamic airway collapse in patients with OSA during sleep permits identification of anatomic causes of airway obstruction and facilitates planning for treatments required to relieve airway obstruction. MRI sleep studies demonstrate airway motion abnormalities that are related to OSA [108]. The most common challenge faced during sleep MR airway imaging studies is the inability of the child breathing via the native airway to tolerate an adequate level of sedation or anesthesia without experiencing significant oxygen desaturation. There is no strict consensus among sedation providers as to when to interrupt airway imaging for interventions to improve oxygenation. Absolute lower limits of oxygen saturation below which artificial airway adjuncts are required may differ from patient to patient depending on the benefits to be gained from the imaging study and the severity of the patient's condition. It is helpful to review overnight PSG reports, noting in particular the severity of oxygen desaturations during natural sleep, as a guide to acceptable minimal arterial oxygen saturations for a particular patient. Dexmedetomidine provides an acceptable level of sedation-anesthesia for MRI sleep studies in children with OSA and makes it possible to complete the study successfully in the majority of children without resorting to the use of artificial airways [109].

A recent study using an electronic survey of national and international members of the Society of Pediatric Anesthesia and a closed claims database (from 1990 to 2011) focused on OSA and reported all deaths and neurologic injury in relation to apnea. Closed claims involving death or neurologic injury after tonsillectomy due to apparent apnea in children suggest that at least 16 children out of 86 may have been rescued had respiratory monitoring been continued throughout first- and second-stage recovery, as well as on the ward during the first postoperative night. The authors recommended a validated pediatric-specific risk assessment scoring system to identify children at risk for OSA [110]. Another recent review of the LexisNexis "MEGATM Jury Verdicts and Settlements" database reported that sleep apnea was inculpated in 17 fatal malpractice claims related to post-tonsillectomy management [111].

An essential duty of the sedationist is to determine which patients are at risk for post-procedure respiratory adverse events and which can be managed as an outpatient. Currently we are not aware of any consensus among institutions that care for these patients as to clear post-procedure discharge criteria. The most recent literature is insufficient to offer definitive guidance regarding which patients with OSA can be safely managed as an outpatient, who should be admitted, and the appropriate time for discharge of these patients from the facility [112].

The ASA's Practice Guidelines for the Perioperative Management of Patients with Obstructive Sleep Apnea recommend **the following factors to be considered in determining whether outpatient care is appropriate** or not. These factors include: (1) sleep apnea status, (2) anatomical and physiologic abnormalities, (3) status of coexisting diseases, (4) nature of the surgery, (5) type of anesthesia, (6) need for postoperative opioids, (7) patient age, (8) adequacy of post-discharge observation, and (9) capabilities of the outpatient facility [112].

The authors approach these patients in the following way: at the end of the pre-procedure evaluation, we perform a risk assessment based on the presence and severity of symptoms, invasiveness of the procedure, associated comorbidities,

physical examination, and, if available, the results of PSG. We have a very low threshold to admit children with OSA after procedural sedation who have any of the following comorbidities: craniofacial anomalies, obesity, history of prematurity, neuromuscular diseases, cardiac manifestations of OSA (e.g., right ventricular hypertrophy), Down syndrome, chronic lung disease, and sickle cell anemia. The decision to admit the child with whose OSA severity is yet undetermined is more challenging. If the patient develops significant episodes of obstruction during the procedure, we admit overnight with continuous monitoring for observation. OSA patients who are on home apnea monitoring or receive CPAP or BiPAP should be closely monitored in the hospital setting after the procedure to minimize respiratory complications. Patients with severe OSA undergoing lengthy procedures associated with the use of high doses of opioids require admission to the ICU.

Pregnancy

Although teenage pregnancy rates are currently in a steady decline, the pregnant teenager presenting with the need for an urgent or emergent procedure is not uncommon [108, 109, 113]. Girls of child-bearing age should have a screening pregnancy test done before procedural sedation. **Any elective procedure involving sedation-anesthesia in pregnancy is best postponed until after delivery**. In the urgent or emergent setting, the clinician must stratify risk and minimize harm to the mother and fetus.

The pregnant woman or girl experiences anatomic and physiologic changes throughout the pregnancy, many of which are important considerations in the pre-sedation assessment (Table 4.8) [114]. In general, there is increased oxygen consumption, decreased vascular resistance, increased edema of the upper airway, decreased vital lung capacity, decreased gastroesophageal motility, and decreased lower esophageal tone. Individually and in combination, these normal findings in pregnancy increase the risk of an adverse event during sedation. Screen for symptoms of heart failure, uncontrolled gastroesophageal reflux, frequent or painful uterine contractions, and vaginal bleeding.

It is important to verify the relative safety of the planned agents (and alternatives) prior to starting the procedure. In the United States, the Food and Drug Administration (FDA) has classified the relative risks of medications to the fetus into five categories (Table 4.9) [115, 116].

The clinician should always consult the most recent references for a given drug. It is important to note that sources may vary in classification of risk in pregnancy; **the timing, context, and chronicity of administration will affect the category** [117, 118]. Know and follow your institutional protocols and guidelines.

Premature Infant

Neonates are at high risk for the development of postoperative apnea after sedation-anesthesia. Infants at highest risk are those born prematurely (before the 37th week of gestation), or those with multiple congenital anomalies, a history of apnea and bradycardia, or chronic lung disease. Apneas occur postoperatively at rates of 5–49 % with spinal and general anesthesia [119]. The large variation is mainly due to the use of variable anesthetic and monitoring techniques as well as to the different study populations. The most significant risk factor of apnea in premature infants is conceptional age; the lower the conceptional age, the greater the risk of delayed apnea, with the incidence of postoperative apnea in the micropremie greater than 50 %. The frequency and duration of apnea decrease between 1 and 20 weeks postnatal age [120].

The etiology of apnea is likely multifactorial. Premature infants have decreased ventilatory control and response to hypoxia and hypercarbia—chemoreceptor responses are blunted in these babies. The normal response to hypoxemia (hyperventilation, followed by hypoventilation or apnea) is replaced by apnea only. This lack of physiologic response

Table 4.8 Anatomic and physiologic changes in pregnancy [114]

System	Anatomy	Physiology
Cardiovascular	Uterine obstruction of inferior vena cava → supine hypotensive syndrome	↑ Plasma volume ↑ Cardiac output ↓ SVR
Respiratory	Elevation of diaphragm Airway edema ↓ Upper airway caliber	↑ Minute volume ↑ Oxygen consumption ↓ $PaCO_2$
CNS		↓ Effective distribution of sedatives and hypnotics
Gastrointestinal	↓ Lower esophageal sphincter tone	↑ Gastric volume and acidity Delayed gastric motility
Hematologic		↑ Activity of coagulation factors

Table 4.9 United States FDA pharmaceutical pregnancy categories [115, 116]

Pregnancy Category A	Controlled studies in women fail to demonstrate a risk to the fetus in the first trimester (and there is no evidence of a risk in later trimesters), and the possibility of fetal harm appears remote
Pregnancy Category B	Either animal reproduction studies have not demonstrated fetal risk (but no controlled studies in pregnant women have been reported), or animal reproduction studies have shown an adverse effect (other than a decrease in fertility) that was not confirmed in controlled studies in women in the first trimester (and there is no evidence of risk in later trimesters)
Pregnancy Category C	Either studies in animals have revealed adverse effects on the fetus (teratogenic, embryocidal, or other) but no controlled studies in women have been reported, or studies in women and animals are not available. Drugs should be given only if the potential benefit justifies the potential risk to the fetus
Pregnancy Category D	Positive evidence of human fetal risk exists, but the benefits from use in pregnant women may be acceptable despite the risk (e.g., if the drug is needed for a life-threatening condition or for a serious disease for which safer drugs cannot be used or are ineffective).
Pregnancy Category X	Studies in animals or human beings have demonstrated fetal abnormalities or evidence exists of fetal risk based on human experience, or both, and the risk in pregnant women clearly outweighs any possible benefit. The drug is contraindicated in women who are or may become pregnant

may be worsened by sedative agents. Postoperative apnea can occur after surgery with inhalational-based anesthetics or even after surgery for which a regional anesthetic was used and no anesthetic drugs were utilized [121]. Apneas are frequent in the first 12 h and can continue until 48–72 h.

Kurth et al. studied the breathing patterns of 47 preterm infants less than 60 weeks postconception with pneumocardiograms before and after general inhalational anesthesia. The study found that 18 infants (37 %) had prolonged apnea (>15 s) and an additional 7 infants (14 %) had short apnea (6–15 s) postoperatively [122]. **The authors conclude that preterm infants younger than 60 postconceptional weeks of age should be monitored continuously for at least 12 h postoperatively in order to prevent apnea-related complications.**

The best evidence basis is found in a 1995 meta-analysis of eight prospective studies examining 254 premature infants undergoing general anesthesia for inguinal hernia repair; apnea was strongly inversely related to both gestational age and conceptional age [123]. Anemia (<10 g/dL) and apnea at home were also risk factors. Based on this data, many institutions adopted the study's **recommendation that all infants born <37 weeks gestational age and less than 60 weeks conceptional age be monitored for postoperative apnea for a minimum of an overnight stay in an ICU setting**.

The appropriate discharge time frame of these patients remains controversial. The cutoff for outpatient surgery in infants born before 37 weeks may be 50–52 weeks conceptional age, provided there is no anemia, prior apnea, or coexisting disease. **The most conservative approach is to admit all premature infants (for monitored 24-h observation) younger than 60 weeks conceptional age, regardless of the anesthetic used** [122]. Certainly this should be the case for any high-risk infant, such as those using a home apnea monitor or taking methylxanthine drugs.

There is considerable institutional variability in practice and hospitals have different age-based guidelines for admission. Some institutions feel comfortable performing elective outpatient procedures if the infant is born full term. Other centers prefer to wait until the infant is 2–4 weeks of age to ensure the resolution of physiologic jaundice, decreased pulmonary vascular resistance, and to give sufficient time for the ductus arteriosus to close. Still in other settings, such as the emergency department, full-term infants less than 3 months of age undergoing significant sedation for an emergent procedure are rarely discharged home on the same day. Options are limited in this high-risk population, as otherwise "safe" agents such as ketamine are contraindicated in these very young infants (<3 months of age in a full-term infant).

Regardless of the timing or setting, premature infants should have both pulse oximetry and apnea monitoring, since standard impedance pneumatography can fail to detect episodes that result in serious desaturation [123]. Although there is limited evidence that prophylactic caffeine or theophylline reduces the rate of post-procedure apnea, if the infant experiences any irregular breathing after the procedure, caffeine should be given without delay.

In-depth understanding of the preterm neonatal physiology is vital to the sedation provider. For example, in patients who have a patent ductus arteriosus, one pulse oximetry probe should be placed on the right hand (pre-ductal) and the other on a lower limb (post-ductal). In the premature infant, fetal hemoglobin persists. For example, a premature infant at first glance may have a reassuring hemoglobin concentration of 13–15 g/dL; however, 70–80 % may be fetal Hb, which is known to have a reduced ability to release oxygen to the tissues.

Another important concern in these babies is the immaturity of the renal and hepatic systems. **Preterm infants do not maintain fluids and electrolyte balance well, requiring**

care in the administration of the IV fluids and electrolytes. Liver immaturity (both in synthetic and metabolic capacity) may lead to longer duration of action of sedative agents.

Sedation providers should make every effort to **avoid hypothermia during the procedure**. Preterm infants have a high surface-area-to-body-weight ratio and decreased brown fat stores, rendering them very susceptible to heat loss. Heat loss is a major potential stress in premature babies and hypothermia-induced stress can lead to hypoglycemia, apnea, and metabolic acidosis.

In summary, sedating-anesthetizing a preterm neonate requires in-depth understanding of neonatal physiology, constant vigilance, rapid recognition of any adverse event, and rapid intervention.

Psychiatric and Behavioral Disorders

It is estimated that one in ten children meets criteria for a serious emotional disturbance, defined as "a mental health problem that has drastic impact on the child's ability to function socially, academically, and emotionally" [124, 125]. Due to changing diagnostic criteria ("diagnosis shifting") and worldwide variation, exact estimates of the prevalence of individual disorders are problematic; nonetheless, increased awareness and diagnosis are commonly seen in practice [126].

Mood disorders in children include anxiety disorders (8 %), major depression (4 %), and bipolar disorder (1 %) [125]. The pre-procedural assessment in these children should include a brief review of the child's general health, control of mood disorder, recent additions or changes to medications, and history of previous procedures and adverse drug reactions (especially to psychotropic medications). These children are at risk for eating disorders and substance abuse, and may present with hypothermia, hypokalemia, hypomagnesemia, and/or hypokalemia [127]. If an eating disorder such as anorexia or bulimia is suspected, a screening ECG or chemistry profile should be performed prior to sedation [128, 129].

Behavior disorders are multifactorial in nature, and rates vary greatly by criteria used, population studied, and survey conducted. Attention deficit hyperactivity disorder (ADHD) involves inattention, impulsivity, and hyperactivity. The National Health and Nutrition Examination Survey reveals an overall prevalence of ADHD in children 8–15 to be 8.7 % [130]. Conduct disorder (CD) and oppositional defiant disorder (ODD) are characterized by a pattern of disobedient, hostile, and defiant behavior toward authority figures [131]. As a group, rates of CD and ODD are reported to be as high as 5.5 % in recent US studies, but the rate varies greatly by country and subpopulation [125, 132]. Children with behavior disorders are often prescribed stimulant or other psychotropic medications; they may have an altered reaction to premedication (such as decreased response to benzodiazepines), increased risk of post-procedure nausea and vomiting, and a decreased seizure threshold [133]. Although the literature is inconclusive regarding the need for a special approach to the sedation of these children, the clinician may use this information especially when considering pre-procedural fasting requirements.

Substance abuse disorders in older children and adolescents are estimated to have a prevalence of approximately 5 %, with a wide range of 1–24 % [125]. **There is a significant overlap in behavior and mood disorders in this population.** Although the long-term effects of substance abuse (cardiac, pulmonary, hepatic, renal, immune) may not be evident in children, a good general history and physical examination should reveal red flags in the pre-sedation assessment. *Marijuana* use may cause relaxation and a decreased sedation requirement; however, patients may also present with tachycardia and anxiety from recent use. A mild abstinence syndrome has been reported; conversely, overuse can result in intractable nausea, as in *cannabinoid hyperemesis syndrome. Cocaine* is highly addictive and may cause dysrhythmias, ischemia, and heart failure. These patients often have altered pain perception. Concomitant cocaine use and β(beta)-blocker administration may precipitate hypertensive crisis, due to unopposed α(alpha)-adrenergic stimulation. *Opioid abuse* may present with altered pain tolerance, increased requirements during sedation, and acute withdrawal, depending on the timing of last ingestion. *Alcohol abuse* may present with increased sedative requirements [134].

Designer drugs (also called "club" or "party" drugs) include 3,4-methylene-dioxymethamphetamine (MDMA) or "ecstasy," phencyclidine (PCP), ketamine, inhalants, rohypnol, γ(gamma)-hydroxybutyrate, and bath salts, among others. The clinician will undoubtedly recognize an acutely intoxicated child or adolescent on presentation. However, the non-intoxicated patient with regular use of these substances may not be apparent without a focused history; many have considerable anxiety in the pre-procedure assessment. During sedation, these patients are at risk for **autonomic dysregulation with wide swings in blood pressure and heart rate**, with case reports of non-hemorrhagic cerebral vascular accidents and myocardial ischemia and infarction [135].

During the pre-sedation assessment, the clinician should screen for risk factors for pre- and post-procedural combativeness, such as previous negative experiences with procedures, sedation, or anesthesia; preoperative anxiety; parental anxiety; and other baseline emotional problems [136–138]. In children at risk for combativeness or lack of cooperation, early involvement of supportive family

members, play therapists, and/or nursing staff with distraction techniques may be helpful, as well as the use of noninvasive oral premedication [139].

Sickle Cell Disease

The term sickle cell disease (SCD) includes all hemoglobinopathies that result in sickling of red blood cells (HbSS, HbSC, sickle-cell thalassemias, and other variants). SCD is characterized by hemolytic anemia and vaso-occlusive phenomena, causing painful episodes and a variety of crises affecting virtually every organ system. Although the sickle cell trait originated in West Africa, it is now estimated that more than 250,000 children worldwide are born each year with SCD [140].

Sickling occurs due to deoxygenation stress on HbS polymers, resulting in a process called *gelation*—red blood cells subsequently become less able to deform normally as they pass through capillary beds, which may result in vaso-occlusion and infarction [141]. Even fully oxygenated blood in a child in SCD is more viscous than in non-affected individuals. Volume depletion or dehydration accentuates their baseline hyperviscosity and promotes vascular stasis. For this reason, the pre-sedation assessment should carefully consider the child's volume status. Recent intake, number of wet diapers or frequency of urination, and recent illness should be assessed.

Take a careful history of past sickle-cell crises (e.g., acute chest syndrome, splenic sequestration, hemolytic crises, stroke, priapism, cardiomyopathy, renal disease, avascular necrosis of bones) and the severity of the course of illness. It is important to note whether the child is currently controlled with medications or requires intensive treatment such as red blood cell exchange transfusions [142]. Common medications in SCD include penicillin prophylaxis, hydroxyurea, and folic acid. Transfusion therapy lowers the percentage of HbS in the blood and is used to treat vaso-occlusive crises acutely or to prevent stroke or pain crisis [143]. It is helpful to know the child's recent hematocrit; if there is history of recent illness or complaint consistent with a hemolytic crisis, obtain a CBC and reticulocyte count and address the patient's current complaint and volume status before sedation.

Ask about recent illness, including any fever or atypical pain. If possible, ascertain what medications have helped to relieve pain in the past. Children with SCD typically have high opioid requirements, thought to be due to a variety of reasons, including severe pain, tolerance, and altered plasma clearance of opioids [144]. Certain medications should be avoided in the sedation or analgesia of SCD children, such as meperidine. Multiple doses of meperidine may cause an accumulation of its metabolite, associated with central toxicity such as myoclonus and seizures [145]. Expert opinion varies on the use of nitrous oxide in children with SCD, but it is generally considered safe [146–148].

When possible and appropriate, **consider liberal use of intranasal, oral, and intramuscular medications if intravenous access is not otherwise required**. Children with SCD often have limited reliable vascular access due to frequent venipuncture; be judicious with their remaining usable peripheral veins if feasible.

Syndromes

There is a vast array of pediatric genetic syndromes, each with its particular considerations and challenges in general and acute care. Syndromes may be classified by morphology into four broad categories: **malformation** (poor formation of tissue), **deformation** (unusual forces on normal tissue), **disruption** (breakdown of normal tissue), or **dysplasia** (abnormal organization of tissues). Keep in mind the variance of expression in most syndromes—some children may be mildly affected while others may be severely affected [149].

The pre-sedation assessment should focus on children with abnormal airway anatomy, as airway reflexes may be affected during sedation, and a contingency plan for airway rescue must be ready before the procedure. Ask about previous procedures, previous or current tracheostomies, problems with oral intake or reflux, snoring, or easy choking or fatigue. Some syndromes are associated with specific metabolic issues, such as frequent hypoglycemia (e.g., Beckwith-Wiedemann, pituitary dwarfism). Perform a careful review of the child's medications and ask how the child responds to and recovers from illness and stress (i.e., history of decompensation or requiring medication supplementation). Perform a careful assessment of the size and shape of the mouth and tongue, the ability to open the mouth wide, and identify the Mallampati classification of pharyngeal structures (Fig. 4.2, Table 4.10) [149–153]. **It is important to palpate the distance from the anterior ramus of the mandible to the hyoid bone**. In infants, it should measure at least one finger breadth (of the adult examiner); in children at least two finger breadths; and in adolescents at least three finger breadths. **A decreased distance correlates with a more difficult rescue airway** [150].

Down syndrome is the most common chromosomal abnormality, with an overall incidence of as high as 1 in 700 live births, varying by region and maternal age. The sedation practitioner must be familiar with its associated multisystem abnormalities including OSA, CHD (endocardial cushion defect, VSD), atlantoaxial instability, obesity, and subglottic stenosis.

Predisposing factors for OSA in these children include midfacial and mandibular hypoplasia, glossoptosis, adenoidal encroachment, increased secretions, and an increased

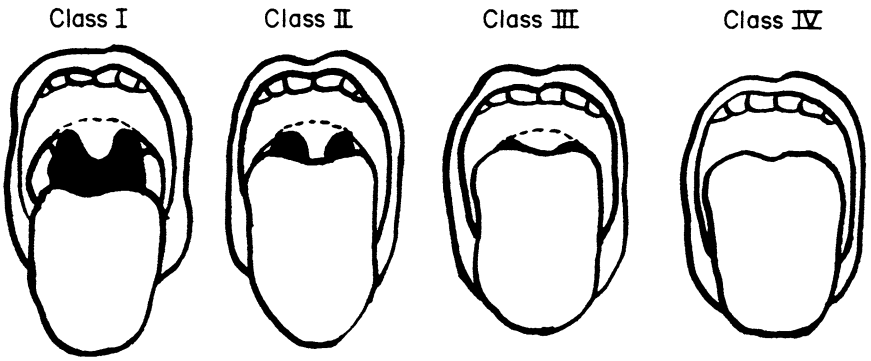

Fig. 4.2 Mallampati classification of pharyngeal structures. Reprinted with permission from Samsoon GL, Young JRB. Difficult tracheal intubation: a retrospective study. Anaesthesia. 1987;42:487–90

Table 4.10 Anatomic considerations in common syndromes [149–153]

Anatomic consideration		Associated syndromes
Alanto-occipital joint abnormalities	• Short neck	***Down syndrome*** (Trisomy 21)
	• Limited mobility	***Goldenhar syndrome*** (incomplete development of the ear, nose, palate, and mandible)
	• Instability	***Juvenile Rheumatoid Arthritis*** (JRA)
		Klippel-Feil syndrome (short neck, restricted upper spine mobility)
Abnormal airway anatomy	• Mandibular hypoplasia	***Airway mass/tumor***
	• High arched/narrow palate	***Arteriovenous malformation*** (AVM)
	• Macroglossia	***Arthrogryposis*** (congenital multiple contractures)
		Beckwith-Wiedemann syndrome (exomphalos, macroglossia, gigantism)
		Cornelia de Lange syndrome (microcephaly, dwarfism, cleft palate)
		Cri du chat (microcephaly, clinodactyly)
		Crouzon syndrome (cranial synostosis, hypotelorism, hypoplastic maxilla)
		DiGeorge syndrome (velo-pharyngeal insufficiency, hypothyroidism)
		Dwarfism (various)
		Goldenhar syndrome (incomplete development of the ear, nose, palate, and mandible)
		Mucopolysaccharidosis (various)
		Pierre Robin sequence (micrognathia, upper airway obstruction)
		Treacher Collins syndrome (micrognathia, hearing loss)
		Trisomies (especially 18, 21, 22)
Midface abnormalities	• Maxillary hypoplasia	***Apert syndrome*** (hypertelorism, craniosynostosis, hydrocephalus)
	• Nasal or choanal stenosis	***Down syndrome*** (Trisomy 21)

incidence of lower respiratory tract anomalies, obesity, and generalized hypotonia. **These children are sensitive to respiratory depression by opioids, sedatives, and hypnotics**; they are especially vulnerable to the development of upper airway obstruction during sedation-anesthesia. A smaller than normal endotracheal tube should be placed if indicated and the head should remain in neutral position during intubation.

The most common sedation-anesthesia-related complication in these patients is **bradycardia**, especially during induction. This may occur even in the absence of heart disease. Borland et al. reported the incidence of severe bradycardia associated with inhaled anesthetic induction (halothane or isoflurane) in children with Down syndrome to be 3.7 % [154]. Recently Kraemer et al. examined the incidence of bradycardia in 209 children with Down syndrome and 268 healthy control patients who had inhaled induction of anesthesia with sevoflurane over an 8-year period. On univariate analysis Down syndrome, low ASA physical status, CHD, and mean sevoflurane concentrations were factors associated with bradycardia. However, multivariate analysis showed that only Down syndrome and low ASA physical status remained as independent factors associated with bradycardia [155].

Table 4.11 ATLS hemorrhagic shock classification [156]

	Class I	Class II	Class III	Class IV
Percent blood loss (%)	Up to 15 %	15–30	30–40	>40
Heart rate	Normal	Mild tachycardia	Moderate tachycardia	Severe tachycardia
Blood pressure	Normal	Normal to decreased	Decreased	Decreased
Respiratory Rate	Normal	Mild tachypnea	Moderate tachypnea	Severe tachypnea
Urine Output	Normal	0.5–1 mL/kg/h (minimum goal)	0.25–0.5 mL/kg/h (markedly decreased)	Negligible
Mental status	Slightly anxious	Mildly anxious	Anxious/confused	Confused/lethargic
Fluid replacement	Crystalloid	Crystalloid	Crystalloid and blood	Crystalloid and blood

Cardiac output is dependent on heart rate, especially in neonates and infants, and bradycardia can have a significant effect on the patient's hemodynamic stability. Some practitioners routinely use intramuscular prophylactic atropine to prevent bradycardia before anesthesia induction. It is important to recognize that atropine will not prevent or reverse the negative inotropic effect of an inhalational anesthetic, but it may maintain heart rate. Gradual titration of the volatile agent concentration and close monitoring of blood pressure and heart rate are recommended during inhalational induction of patients with Down syndrome. If bradycardia occurs and an IV is not in place, intramuscular atropine should be administered if there is sustained bradycardia or if hemodynamic instability develops.

Trauma

The acutely injured child poses a particular challenge to the clinician performing sedation. The child may present immediately after trauma or subacutely. Only after primary and secondary advanced trauma life support (ATLS) surveys are completed and injuries addressed and stabilized is the child a candidate for sedation outside of the operating room.

In addition to the injury-specific brief history and physical examination, the pre-sedation assessment will include last intake by mouth, allergies, medications, and prior sedation or anesthesia. The urgency of procedural sedation will match the urgency of the presenting condition, such as neurovascular compromise; this will affect the clinician's decision in the amount of fasting time allowed (Table 4.1).

Keep in mind that a child with one injury is at risk for other obvious or occult injuries, due to the pliable thorax and underdeveloped musculature of the pediatric abdomen. ATLS describes four classes of hemorrhagic shock, initially developed for adults (Table 4.11) [156]. Children will compensate well with tachycardia (compensated shock) until a precipitous fall in blood pressure is noted (decompensated shock), and ominous sign [157].

Medication given during sedation may affect vital signs that would otherwise serve as an early warning sign of ongoing occult hemorrhage. For example, ketamine administered for orthopedic reduction invariably causes an increase in heart rate, which makes the recognition of compensated shock difficult. Similarly, propofol, opioids, and benzodiazepines may cause a small drop in blood pressure that may mask an underlying decompensated shock. Meticulous history and physical examination to screen for occult injuries is imperative before the urgent or elective sedation. During sedation, consideration of developing shock should always be at the forefront of the clinician's mind. Consider strategies such as peripheral nerve blocks and mild anxiolysis in these patients.

Tuberous Sclerosis

Tuberous sclerosis (TS) is one of the commonest autosomal dominant genetic disorders, displaying high genetic penetrance in affected families. TS is a neurocutaneous disorder characterized by a classic triad of epilepsy, fibroangiomas, and mental retardation. TS causes hamartomas in multiple organs, including the brain, skin, heart, kidneys, lungs, and liver. Awareness of the signs, symptoms, and organs affected is critical to reduce the risk of a life-threatening complication.

A **baseline cardiac evaluation** (regardless of presence or absence of symptoms) is an essential part of the pre-procedure work-up to determine whether the procedure is appropriate for non-anesthesiologist sedation or whether the expertise of an anesthesiologist is needed. Cardiovascular manifestations, seen in more than 50 % of affected individuals, can have major anesthetic-sedative implications. Rhabdomyomas are the most common benign cardiac tumors associated with tuberous sclerosis [158]. They tend to regress spontaneously and are not usually excised unless they become obstructive or cause severe arrhythmias. A pre-procedure ECG is recommended to exclude dysrhythmia or conduction defects. Abdominal aortic aneurysms

have been reported as well as narrowing of major arteries in patients with TS.

Airway management can be challenging in these patients due to the presence of oropharyngeal or laryngeal tumors, fibromata, or papillomata. Pulmonary involvement is rare (<1 %). However, hamartomatous growths may involve the lungs or pleura and there have been a number of reports of spontaneous pneumothorax in patients with undiagnosed pulmonary manifestations of the disease. A **pre-procedure chest radiograph (X-ray)** is recommended to exclude silent pulmonary or mediastinal masses.

Renal function should also be assessed before the procedure because renal angiomyolipomas are present in 50–80 % of affected individuals [159]. Although possibly initially clinically silent, these patients are known to progress to renal failure. Anticonvulsants should be optimized and continued until the morning of surgery and should be resumed as soon as possible in order to prevent seizures [160].

Upper Respiratory Tract Infection

There is no consensus regarding the optimal management of children with URI who require sedation for an elective procedure. The economic and emotional consequences of cancelling a procedure are significant for the family and the institution. Studies showed that anywhere from 3 to 33 % of children coming for anesthesia and surgery present with an active URI [161]. Children with URIs who present for procedural sedation pose a perplexing clinical dilemma for sedation providers. Currently there is little agreement between individual providers and institutions on which children with respiratory tract infections (RTIs) should be sedated-anesthetized and under what circumstances. **Inflammation from a URI may persist for up to 6 weeks after apparent resolution of symptoms**.

An active URI may put the child at risk for laryngospasm, bronchospasm, severe coughing, major oxygen desaturations (<90 %) airway obstruction, pneumonia, and unanticipated admission. These complications are disturbing, but fortunately can be addressed with medications that should be readily available during any procedure, such as inhaled β(beta)-agonists for bronchospasm, succinylcholine followed by advanced airway management for sustained laryngospasm not amenable to positive-pressure ventilation, and supplemental oxygen for desaturation [162].

Sedation practitioners need to differentiate allergic rhinitis from URI and uncomplicated URIs from other illnesses. Typical symptoms of uncomplicated URI include low-grade fever, rhinorrhea, congestion, sneezing, sore throat, and laryngitis. If the child has a disproportionally high fever or shows signs of lower respiratory tract symptoms such as increased work of breathing, wheezing, or mucopurulent secretions, the pathology may have extended beyond the upper respiratory tract.

Many children with recurrent URIs have a very small window of opportunity to provide sedation in the symptom-free period. It is inevitable that the sedation provider will need to look to decision tools to help to disentangle this dilemma. Parnis et al. used logistic regression to determine which variables were predictors of perioperative anesthetic adverse events in 2,051 children. The analysis showed that 22.3 % of children had symptoms of an RTI on the day of surgery, and 45.8 % had a "cold" in the preceding 6 weeks [163]. Important independent preoperative predictors of anesthetic adverse events were: parental report of the child's having a "cold" on the day of surgery, nasal congestion, history of snoring, history of second-hand smoking, and cough productive of sputum. The study concluded that surgery requiring endotracheal intubation increases the probability of anesthetic complications, but when the airway is managed with a laryngeal mask or face mask the probability of complications is decreased. An interesting finding worth noting was that the identification of a viral pathogen did not help to identify individuals at risk for adverse events.

The never-ending question of what to do with a child with a URI will always be with us. In the absence of evidence-based clear criteria, the sedation practitioner should be especially aware of active signs and symptoms. A clinical algorithm has been proposed (Fig. 4.3) to guide the assessment and management of these children [164]. Most practitioners would agree that children with mild uncomplicated URIs undergoing procedures that do not involve airway manipulation can be safely anesthetized-sedated without any increase in risk [165].

Conclusion

The prepared provider should be as informed about the patient as he is about the procedure to be performed. Eliciting red flags in history and physical examination is the basis for safe sedation practice. When faced with a less-than-ideally prepared patient or situation, the provider should work to optimize the patient's status and anticipate complications before the procedure takes place.

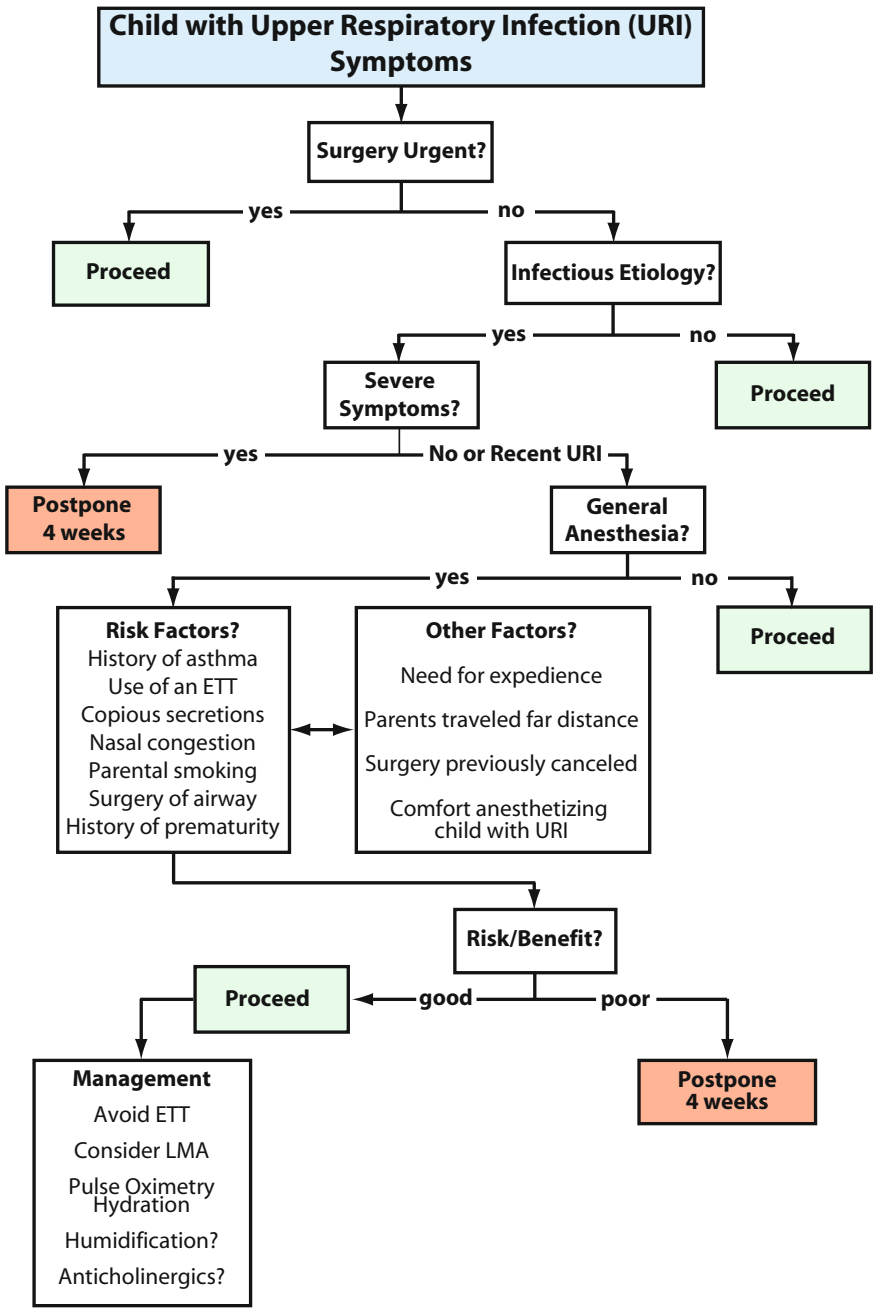

Fig. 4.3 Suggested algorithm and management of a child with upper respiratory infection. Modified with permission from Tait ATR, Malviya S. Anesthesia for the child with an upper respiratory tract infection: still a dilemma? Anesth Analg 2005;100:59–65

Case Studies in Pre-sedation Assessment

Case 1: Just Another URI?

A 4-year-old girl with a history of seizures is scheduled for magnetoencephalography (MEG) scan. She has a 4-day history of isolated clear rhinorrhea. Her lungs are clear to auscultation and she is afebrile. Her mother reported that her activity level and appetite have been unchanged since onset of rhinorrhea.

The main considerations for this child will be the pre-procedure URI and understanding the needs and requirements for MEG scan. This child appears to have an uncomplicated URI. Based on the information provided in this clinical scenario, proceeding with the scan is the most appropriate decision. Understanding the nature and demands of MEG is important to decide on the appropriate sedative agent. MEG scan records

(continued)

magnetic fields induced by the brain's electrical activity and recently is increasingly used in presurgical evaluation of epileptic children. Compared with the standard electroencephalogram (EEG), the MEG allows for a better spatial resolution in the localization of epileptogenic foci. MEG exams are conducted in magnetically shielded chambers to minimize interference of magnetic fields induced by other electric and electronic appliances. Our experience with dexmedetomidine-based technique (2 μg/kg loading dose followed by 2 μg/kg/h infusion) provides adequate depth of sedation required to prevent motion artifacts. Compared with propofol at higher doses dexmedetomidine does not appear to negatively affect inter-ictal activity and thereby does not interfere with spike identification.

Case 2: Snoring Away

A 2-year-old 16 kg boy born at 33 weeks gestation is scheduled for high resolution CT. The CT is being done as part of the work-up for recurrent aspiration pneumonias. On pre-imaging evaluation, the child's exam reveals micrognathia and a cleft palate. His mother reports that he "snores a lot" and seems to obstruct his upper airway at night. A look through the medical records shows that the patient recently underwent an overnight sleep study (PSG) that demonstrated a moderate degree of OSA with a minimum oxygen saturation of 86 %.

The considerations in this case are: difficult airway, OSA, and an imaging study requires controlled ventilation in off-site environment. A thoughtful, carefully implemented plan is essential to ensure safety and high-quality imaging study for this patient. In an ideal world this family should have been contacted prior to scheduling to ensure a proper consultation with an anesthesiologist who can guide the safest plan for sedating this infant.

It is important to evaluate the airway carefully prior to beginning anesthesia or sedation. Evaluation of the pediatric airway can be challenging as the patient may be uncooperative and the history given by parents may be misleading. The overnight PSG provides clues to the severity of the airway obstruction during sleep by providing the lowest oxygen saturation observed, as well as the types of apnea (obstructive, central, or mixed) and the frequency of apnea events. The combination of micrognathia and significant OSA in an off-site location would contraindicate non-anesthesiologist delivered sedation. This patient should be managed by an anesthesiologist who is trained in and prepared for the difficult airway. The anesthetic management is detailed below.

Before inducing this infant, the authors would manage this case as follows:

1. Discuss the benefits and risks of the study with the family and ordering physician and make arrangements for post-procedure admission if required.
2. Review previous anesthetic/sedative records and documentations for previous airway management.
3. Confirm that advanced airway management instruments are available including different sizes of face masks, endotracheal tubes, laryngoscope blades and handles, appropriate size LMA fiberoptic equipment, video laryngoscope, and the difficult airway cart.
4. Proceed with an inhalational induction with sevoflurane with maintenance of spontaneous ventilation followed by placement of LMA when it is established that the patient can be ventilated.

Help in the case of an emergency may be less readily available than in the operating room environment. A more conservative approach in this clinical scenario is to start the anesthetic in the more controlled environment of the operating room, secure the airway with an endotracheal tube, and then transport the patient to radiology. The operating room provides a safe, secure, and familiar environment in which the anesthesiologist has access to emergency airway equipment and assistance from colleagues who can assist with airway management.

Case 3: It's All in Your Head

A 5-year-old boy with developmental delay and autism is hit by a baseball on the left temporal aspect of his head. His GCS is 14, and he has a large scalp hematoma. The decision is made to perform a CT of his head. He is intermittently sleepy and agitated, but consolable by his mother.

The main questions for this potentially uncooperative patient are: (1) Is the procedure painful? (2) How long will the procedure take? (3) Will non-pharmacologic methods be appropriate?

This is an emergent study, but the provider has time to review any medical comorbidities, as well as any history of previous sedation and the outcome. In the proper context, with a calm and reassuring caregiver, a tablet computer or smart phone may be employed to distract the child for the very brief study.

(continued)

This would avoid any complication of sedation, allow the provider to watch his mental status more closely, and potentially ensure a more expedient discharge if the work-up is negative.

If this child is to be sedated, the less invasive the technique the better: Intranasal medications, such as combined midazolam and fentanyl, may give just enough sedation to accomplish this non-painful, non-distressing procedure. If this fails, the intravenous route offers a wide array of options. Rarely in children does the provider have to intubate and sedate in order to obtain advanced emergent imaging.

Case 4: Broken Heart, Broken Bone

A 7-year-old boy with hypoplastic left heart syndrome who is doing well as an outpatient falls off a slide and sustains a right femur fracture. His vital signs are his normal baseline, and he has no other evidence of trauma. On radiograph, his right femur shows a mid-shaft fracture with shortening of the thigh; he is neurovascularly intact distally. He requires emergent placement of a Steinmann pin and traction in anticipation for the operating room when it becomes available.

The urgency of this boy's condition requires action. Take a brief, focused history of previous cardiac surgeries, complications, and other comorbidities. Obtain a cardiology consultation with a pediatric cardiologist, if available, to discuss the patient's physiology and management option and concerns. Collaborate with or transfer this patient's care to an anesthesiologist, if possible. This child has had palliative surgery for his cyanotic heart disease; he has undergone a Fontan procedure, and therefore his cardiac output is pre-load dependent. His volume status should be optimized prior to the procedure. Small boluses of 10 mL/kg of normal saline may be given carefully to ensure euvolemia (with careful attention not to cause volume overload). Prior to proceeding, emergency medications and vasopressors should be immediately available for administration.

This child may be best served with a femoral nerve or fascia iliaca block, to avoid the potential problems with volume and oxygenation status. If this is not possible, a medication that preserves systemic vascular resistance, such as ketamine, would be a good option. Although short acting, a medication such as propofol would not be ideal in this child; propofol is a myocardial depressant and causes transient hypotension. In the otherwise healthy child, this is not an issue. In this child with CHD and low reserve, it is best avoided.

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