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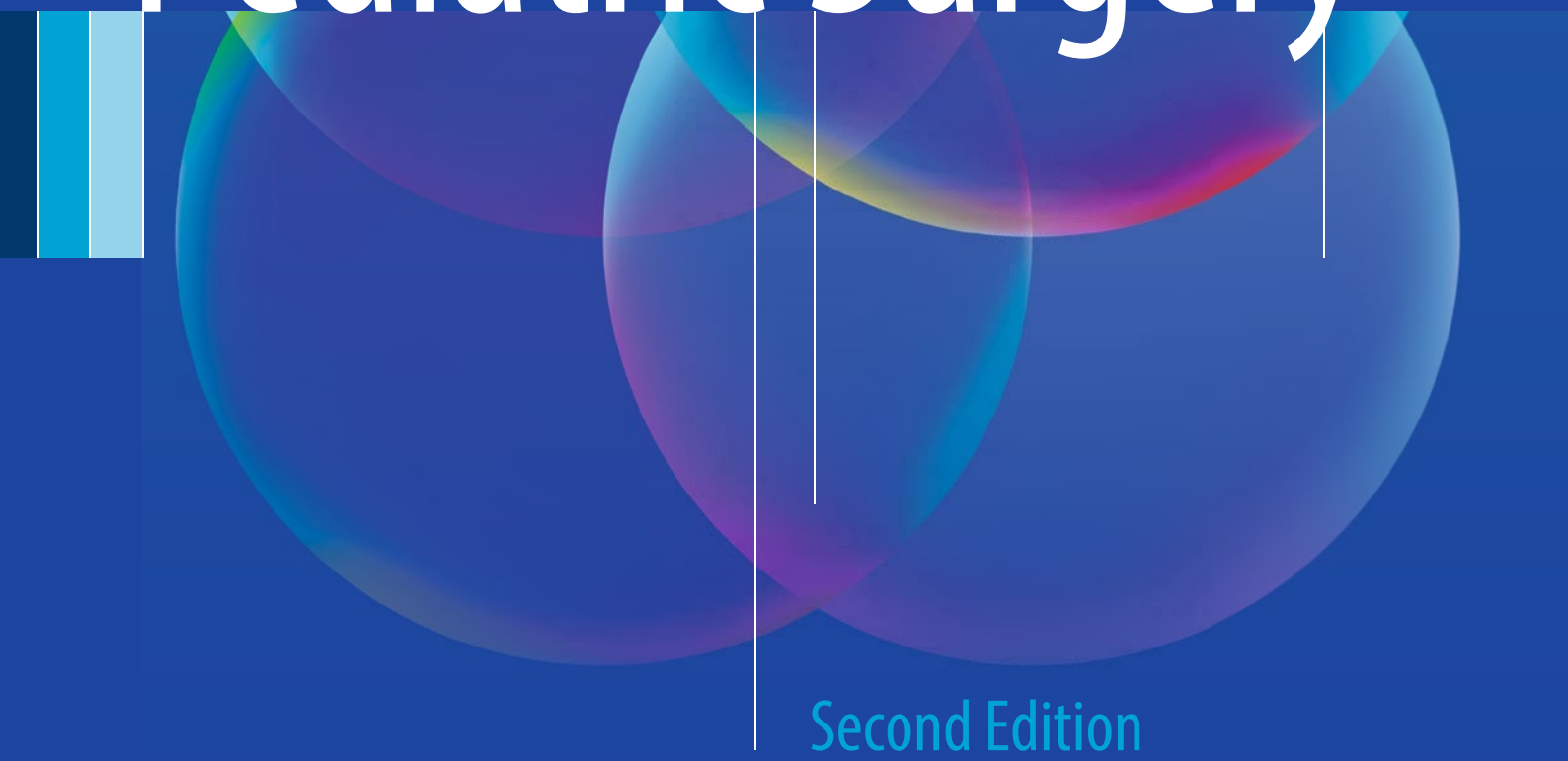
Fundamentals of Pediatric Surgery



Second Edition

Springer

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ISBN 978-3-319-27441-6 ISBN 978-3-319-27443-0 (eBook)
DOI 10.1007/978-3-319-27443-0

Library of Congress Control Number: 2016944232

Printed on acid-free paper

This Springer imprint is published by Springer Nature
The registered company is Springer International Publishing AG Switzerland

To Kim, Gina, Peter, Joey, and Michael, without whose love and support nothing would be possible or worthwhile.

–P.M.

To my Wife Maria and my sons Alessandro and Federico through whom all good things come.

–P.F.N.

To my wife and children for their constant support.

–M.D.R.

To my patients and their families, who entrust me with their most precious possession.
To my mentors, whose teachings motivate me to achieve excellence and precision.
To my family, whose love, support, and understanding provide the inspiration for the path I've chosen.
I am humbled and grateful.

–C.S.M.

Preface

Fundamentals of Pediatric Surgery, Second Edition is meant for pediatric and general surgeons, pediatric surgery fellows, surgery residents, and other advanced practitioners and intended to be a reliable source of up-to-date information regarding the everyday care of children with a surgical condition. Each chapter is written by an experienced authority in the field and addresses a specific aspect of clinical pediatric surgery, carefully edited to maintain a continuity of style and format while preserving the distinctive voice of the author. The goal is to provide practical and clinically relevant information in an accessible and straightforward presentation. The new edition features updates in every topic, many new authors, and three new associate editors. Every chapter begins with an abstract that highlights important themes and is written in a this-is-how-I-do-it narrative style that the reader ought to find familiar—more like an amiable conversation with a trusted mentor and friend rather than a dry or sterile lecture. Finally, nearly every chapter is followed by an additional comment written by the editors and intended to provide pearls, more in-depth analysis, or additional useful information.

In addition to providing a useful reference for pediatric surgeons and general surgeons in clinical practice, *Fundamentals of Pediatric Surgery, Second Edition* is also designed to be used by general surgical residents rotating in pediatric surgery and chief residents in Pediatric Surgery fellowship programs. The American Board of Surgery and the Accreditation Council for Graduate Medical Education (ACGME) consider experience in the clinical aspects of pediatric surgery a necessary and important aspect of the education and training of the general surgeon and most General Surgery residents are still expected to rotate on a Pediatric Surgery service. These brief rotations can be quite busy, with little time to read any of the excellent comprehensive pediatric surgery textbooks available, especially when what one really needs is a practical guide to the everyday care of the pediatric surgical patient. Enter *Fundamentals of Pediatric Surgery, Second Edition,* a concise easy-to-read textbook filled with detailed and relevant information that can help the resident care for the patient they are seeing in the clinic or in the hospital. The goal is to provide at least one reasonable and proven approach, recommended by a recognized expert, and presented in a context that includes a discussion of the underlying principles of care and essential clinical issues to be considered.

Finally, it is hoped that Pediatric Surgery fellows will find this book to be a rich and up-to-date source of pertinent information related to the actual day-to-day care of the surgical child and provide the foundation for what will be an exciting and lifelong education in the complexities of the surgical care of children. It was originally intended to be a valuable resource and study guide for preparation for the written and oral American Board of Surgery certifying examinations in Pediatric Surgery. It is our sincere hope that *Fundamentals of Pediatric Surgery*, *Second Edition*, designed with the more advanced practitioner in mind, will prove to be a useful and valuable complement to the many excellent pediatric surgical texts currently available.

Philadelphia, PA, USA
November 2015

Peter Mattei

Acknowledgements

This book is the result of a great deal of time and effort on the part of dedicated contributing authors, all of whom are exceptionally busy taking care of patients but who felt they had something important to pass along to current and future generations of pediatric and general surgeons and therefore volunteered to contribute their chapters. We also have many people to thank in our home departments including our partners and associates, administrative assistants, and, of course, our patients' families, who give us the opportunity and privilege of caring for their children every day. We would be remiss in not mentioning the help and inspiration provided by the generations of surgery residents and fellows, brave folks who allow us the privilege of teaching and training them to be the best surgeons they can be. Finally, we want to thank our Developmental Editor, Portia Levasseur, without whom this textbook would not have been possible.

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Part I

Perioperative Care

Preoperative Assessment and Preparation

1

Ari Y. Weintraub and Lynne G. Maxwell

The goals of the preoperative evaluation are to identify active medical issues and to ensure that the management of these conditions is optimized prior to anesthesia and surgery. Unresolved medical issues are sometimes significant enough to warrant cancelation of procedures for further diagnostic workup or treatment. It is in the best interest of all involved to avoid this.

Risks of Anesthesia

The risk of dying from general anesthesia can only be extrapolated from large series and appears to be as low as 1 in 250,000 in healthy patients. To put this in perspective for parents, the risk of a motor vehicle collision on the way to the hospital or surgery center is greater than the risk of death under anesthesia. Common minor adverse effects including discomfort from airway management and postoperative nausea and vomiting (PONV) should be discussed, along with assurances that everything will be done to prevent and treat these relatively common complaints.

The American Society of Anesthesiologists (ASA) physical status score is a means of describing the physical condition of the patient. The physical status score was never intended to represent a measure of operative risk but instead serves primarily as a means of communication among care providers (Table 1.1). In addition, certain information is essential and should be included in the preoperative assessment of every patient: weight, blood pressure, oxygen saturation (SpO_2) by pulse oximetry in room air (and with supplemental O_2, if applicable), allergies, medications, cardiac and murmur history, and previous subspecialty encounters.

Patients who have previously undergone general anesthesia should be asked specifically regarding a history of adverse effects: emergence delirium, PONV, difficult intubation, or difficult intravenous access. Keep in mind that patients and their parents are often very anxious about recurrence of these events. The family history should also be reviewed for pseudocholinesterase deficiency (prolonged paralysis after succinylcholine) or any first-degree relative who experienced malignant hyperthermia.

Airway/Respiratory System

Many congenital syndromes are associated with craniofacial abnormalities that may complicate or even preclude routine airway management techniques (Table 1.2). In addition to a detailed physical examination, a history of past intubations and details of the methods needed to secure the airway are even more useful in planning an anesthetic. Some patients are given a "difficult airway letter" by an anesthesiologist, and this information should be shared with the anesthesia care team in advance of the scheduled operation. In the absence of such information, prior anesthetic records should be obtained and reviewed to guide airway management.

Asthma (reactive airway disease) is one of the most common chronic diseases in children, and the disease can be exacerbated by perioperative procedures, including anesthetic induction and emergence or endotracheal intubation. As with all chronic conditions, asthma should be optimally medically managed prior to an operation or general anesthesia. In addition to the regular appropriate use of "controller medications" (inhaled corticosteroids, intermediate-acting bronchodilators, leukotriene modifiers), to minimize perioperative bronchospasm, we typically recommend that patients

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P. Mattei et al. (eds.), *Fundamentals of Pediatric Surgery*, DOI 10.1007/978-3-319-27443-0_1

Table 1.1 American Society of Anesthesiology (ASA) physical status (PS) classifications

Classification	Definition	Example
PS 1	Normal healthy person	
PS 2	Mild systemic disease without functional limitations	Well-controlled asthma
PS 3	Severe systemic disease	Acute lymphocytic leukemia
PS 4	Severe systemic disease that is a constant threat to life	Extreme prematurity
PS 5	Moribund patient, unexpected to survive without the procedure	Congenital heart disease for initiation of ECMO
PS 6	Brain-dead patient for organ procurement	
E	Suffix added for emergent procedures	

Table 1.2 Syndromes and craniofacial abnormalities associated with difficult ventilation or intubation

Syndrome	Associated airway features
Apert	Craniosynostosis, midface hypoplasia
Beckwith–Wiedemann syndrome	Macroglossia
Crouzon	Craniosynostosis, midface hypoplasia
Freeman–Sheldon (whistling face) syndrome	Microstomia
Goldenhar syndrome	Hemifacial microsomia, mandibular hypoplasia (uni- or bilateral)
Klippel–Feil syndrome	Limited cervical mobility
Mucopolysaccharide storage disorders	Redundant facial, pharyngeal, and supraglottic soft tissue; neck immobility
Pierre Robin sequence	Micrognathia, glossoptosis, cleft palate
Treacher Collins syndrome	Maxillary/mandibular hypoplasia
Trisomy 21 (Down syndrome)	Macroglossia, subglottic stenosis, midface hypoplasia

with asthma use their bronchodilators every 6 h for 48 h prior to anesthesia. A history of a recent flare requiring oral corticosteroids suggests poorly controlled disease and might warrant delay of an elective procedure until better control is achieved. Some recommend waiting 4–6 weeks after an acute exacerbation for the usual airway hyperreactivity to return to baseline. Patients with persistent poorly controlled reactive airway disease should be referred to their primary healthcare provider or pulmonologist for strategies to improve their status. These strategies sometimes include the administration of oral corticosteroids.

Children often have loose teeth as they transition from their primary to secondary dentition or due to poor oral hygiene or an underlying disorder such as osteogenesis imperfecta or ectodermal dysplasia. Because there is a significant risk of aspirating a tooth that is accidentally displaced during orotracheal intubation, loose teeth should be electively removed at induction. In some cases it is best to recommend a preoperative visit to a dentist.

Obstructive sleep apnea is seen commonly in patients with adenotonsillar hypertrophy, obesity, and some syndromes. Symptoms (snoring, daytime somnolence), results of sleep studies, and the need for noninvasive ventilation (CPAP, BIPAP) should be included in the preoperative assessment as airway obstruction is expected and should be anticipated in the postoperative period, often making inpatient observation and monitoring necessary.

A very common question is whether an anesthesiologist should cancel a procedure because of an upper respiratory infection. This can be a vexing problem for all parties involved and the decision is sometimes difficult to make with confidence. The patient with a current or recent URI undergoing general anesthesia is theoretically at increased risk of a postoperative respiratory complication, including laryngospasm, bronchospasm, hypoxia, and apnea, with the patients under 2 years of age at greatest risk. However, anesthetic management can also be tailored to reduce stimulation of a potentially hyperreactive airway. In addition, cancelation of a procedure can impose an emotional or economic burden on the patient, family, physician, and hospital or ambulatory surgical facility. Unless the patient is acutely ill, it is often acceptable to proceed with the procedure as planned. Patients with high fever, wheezing, or a productive cough may actually have a lower respiratory tract infection, and surgery is more likely to be canceled. Our approach is to discuss the urgency of the planned procedure with the surgeon and to review the risks and benefits of proceeding or rescheduling with the parents, including the possibility that the child may

have another URI at the time of the rescheduled procedure. Allowing the parents to participate in the decision-making process when appropriate usually leads to mutual satisfaction among all parties involved.

The patient with a difficult airway might require advanced airway management techniques, which often necessitates additional OR time and, in some cases, a planned period of postoperative mechanical ventilation or ICU stay.

The laryngeal mask airway is used routinely for general anesthesia. This technique allows the patient to breathe spontaneously, with or without pressure support from the anesthesia machine, and, in most cases, neuromuscular blocking agents are not used. Therefore, it is usually used for cases where skeletal muscle relaxation is not needed for safe conduct of the operation. Any requirement for muscle relaxation should be discussed in advance with the anesthesiologist.

Cardiovascular

At the time of the presurgical evaluation, up to 90 % of children are found to have an "innocent" murmur, probably due to turbulent flow at the aortic or pulmonary roots or in the subclavian or pulmonary arteries. Most of these children do not require a cardiology consultation and can be safely observed. These murmurs are frequently episodic and are associated with a normally split second heart sound, normal exercise tolerance, and normal electrocardiogram. Concomitant medical problems such as anemia and fever augment audibility of innocent murmurs because they increase cardiac output.

Nevertheless, a thorough history and physical examination will occasionally reveal findings that raise greater concern in a child with a murmur: an infant with failure to thrive or diaphoresis or tachypnea during feedings or the older child with dyspnea, tachypnea, exercise intolerance, or syncope. These findings warrant further evaluation, including an electrocardiogram, chest X-ray, consultation with a pediatric cardiologist, and, in some cases, an echocardiogram.

Children with congenital heart disease frequently require a surgical procedure. Assessment of the child's current health status includes a full history and physical examination and recent evaluation by the child's cardiologist. This communication should include a full description of the original lesion, documentation of any procedures performed for palliation or repair, residual abnormalities such as an intracardiac shunt or valve abnormality, current functional status, and results of the most recent echocardiogram.

Knowledge of the child's cardiac anatomy is essential to assess the risk of paradoxical emboli and endocarditis. The American Heart Association has published revised recommendations for antibiotic prophylaxis that are substantially different from those promulgated over the past 50 years (Table 1.3). Specifically, genitourinary and gastrointestinal procedures have been eliminated from those requiring prophylaxis, and prophylaxis for dental and respiratory tract procedures is restricted to patients with (1) unrepaired cyanotic congenital heart disease, (2) congenital heart defect repaired with prosthetic material within the prior 6 months, (3) cardiac transplantation, or (4) a history of endocarditis. Endotracheal intubation itself is not an indication for antibiotic prophylaxis. Patients with hemodynamically insignificant lesions such as bicuspid aortic valve or mitral valve prolapse no longer require prophylaxis for any procedure. Patients with congenital heart disease repaired with prosthetic material require prophylaxis only for the first 6 months after repair because endothelialization will have occurred. This is true for VSD and ASD repairs as long as there is no residual defect. Patients with prosthetic valves and those palliated with shunts or conduits require prophylaxis. Some cardiologists differ with these new guidelines. It is therefore advisable to request a recommendation from the cardiologist based on the child's condition and planned procedure.

Table 1.3 Cardiac conditions for which prophylaxis with dental or respiratory tract procedures is recommended

Congenital heart disease (CHD)[a]
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[b]
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
Prosthetic cardiac valves
Previous infective endocarditis

[a]Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD
[b]Prophylaxis is recommended because endothelialization of prosthetic material occurs within 6 months of the procedure
Source: Data from Wilson W, Taubert KA, Gewitz M et al. Prevention of infective endocarditis. Guidelines from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation* 116(15):1736–54, 2007

Although antibiotic prophylaxis is frequently administered orally to adults, it is usually given intravenously in children. When indicated, our practice is to give the antibiotic intravenously at induction of anesthesia, because the surgical preparation time generally allows sufficient time to achieve adequate blood levels before the incision is made. Starting an intravenous catheter in an awake child solely to administer antibiotics for antibiotics is rarely if ever necessary.

Surgical patients with long QT syndrome (LQTS), in which ion channels involved in repolarization function abnormally due either to a congenital defect or drug effect, are at risk for torsades de pointes, a potentially life-threatening ventricular tachycardia. Congenital LQTS occurs in 1 in 5000 individuals and can present at any age with syncope, seizures, or sudden cardiac death, usually after an increase in sympathetic activity such as exercise or emotional stress. Because volatile anesthetic agents and surgical stress increase the risk of developing ventricular tachycardia, a preoperative electrocardiogram should be obtained in patients who are symptomatic, have a family history of sudden death, or are taking drugs, which predispose to the condition (http://www.azcert.org/medical-pros/drug-lists/drug-lists.cfm). A QTc of more than 470 ms in males and 480 ms in females is diagnostic of LQTS. Since preoperative medical treatment is nearly always necessary, cardiology consultation should be obtained.

Any patient with congenital heart disease, cardiomyopathy, arrhythmia, or unexplained syncope requires a thorough cardiology evaluation before having an elective surgical procedure, especially one that requires a general anesthetic. In fact, anesthetists at most institutions will require that a letter of cardiology clearance be included in the medical record before the day of surgery. This letter is written by the consulting cardiologist and should include a detailed discussion of the anatomy of the defect, the current medical regimen, and specific recommendations regarding the perioperative care of the patient.

Gastroesophageal Reflux Disease

The majority of infants and a significant number of children have some degree of gastroesophageal reflux and the diagnosis of gastroesophageal reflux disease is increasing. Symptoms of GERD in infants and children differ substantially from those seen in adults and are often primarily respiratory in nature: cough, wheezing, or pneumonitis. Yet, despite a theoretical increase in the risk of aspiration of gastric contents during the induction of anesthesia, children with a history of GERD do not have an increased incidence of pulmonary aspiration as long as fasting guidelines have been followed. Unless there is a history of aspiration when fasting, an intravenous rapid sequence induction is not usually indicated. Patient with GERD should be taking appropriate chemoprophylaxis (H_2 blocker or proton pump inhibitor) as prescribed by their primary physician or gastroenterologist.

Obesity

Obesity is an increasing problem in children, with a recent estimated incidence of 15 %. As in adults, obese children have an increased incidence of obstructive sleep apnea, which can be associated with adverse respiratory events in the perioperative period. Problems during induction include difficult mask ventilation. Preoperative evaluation of children with a body mass index of 30 or greater should include a careful history of snoring and daytime somnolence. Patients with suspected obstructive sleep apnea should be referred to a pulmonologist for a sleep study and considered for therapy with a positive-pressure breathing device. In addition to airway and respiratory complications, obese patients have been found to have an increased incidence of postoperative complications such as infection, wound complications, and deep venous thrombosis when compared to children of normal weight.

Diabetes

Approximately 1 in 500 people under age 20 has diabetes; however, complications requiring surgical intervention, such as cardiovascular disease, are extremely rare in this age group. Nevertheless, patients with diabetes present for routine and emergent surgery with the same frequency as non-diabetic patients and their underlying diabetes must be addressed. As with any other chronic illness, the medical management of diabetes should be optimized before elective surgery, and a plan for perioperative glucose and insulin management should be formulated jointly by the endocrinologist and anesthesiologist. The stresses of surgery and its effects on a regular schedule can wreak havoc on normally well-controlled diabetes if not properly managed. The goal of perioperative management is no longer merely avoiding life-threatening hypoglycemia and severe hyperglycemia but to maintain euglycemia to the extent possible.

Regimens of multiple injections of long- and short-acting insulin are still common, but many patients with diabetes have insulin pumps that deliver a continuous subcutaneous infusion with on-demand boluses for carbohydrate intake or correction of hyperglycemia. Typical management includes the usual preoperative fast with clear liquids up to 2 h before the operation. Whenever possible, it is usually best to schedule the diabetic patient as the first case of the day. After consultation with the patient's endocrinologist, the insulin dosage regimen most often includes reduction of the long- or

moderate-acting insulin dose with a reduced or skipped short-acting insulin dose on the morning of surgery. Insulin pump infusions may be continued up until the time of surgery. Blood sugar should be checked upon arrival. Hypoglycemia requires intervention but oral treatment might require delaying the procedure due to fasting guidelines. Hyperglycemia (>250 mg/dL) should be treated with subcutaneous insulin or a bolus via the insulin pump. The presence of urine ketones will usually lead to cancelation or delay of an elective procedure.

An increasing number of institutions are allowing, and often advocating, continued use of the insulin pump throughout the perioperative period, although some institutions still consider insulin pumps unauthorized medical devices and prohibit their use. As long as the infusion set connecting the pump to the patient is not in the surgical field, there is generally no contraindication to continuing the insulin infusion via the pump. Although most pump manufacturers still recommend disconnecting the insulin pump in the setting of electrocautery use, there have been no credible reports of damage to the insulin pump or interrupted insulin delivery due to electrocautery, and we recommend continuing the use of the pump with placement of the grounding pad as close as possible to the surgical site (closer than to the infusion set). In institutions where insulin pump use is forbidden, for short procedures of less than 2-h duration, it is often sufficient to simply disconnect the insulin pump immediately before incision with monitoring of blood sugar by finger sticks regularly during the course of the anesthetic and administration of subcutaneous or intravenous insulin to correct hyperglycemia, using a sliding scale agreed upon in advance with the child's endocrinologist, with intravenous dextrose as needed for hypoglycemia. Longer procedures, or those requiring postoperative admission, sometimes require continuous intravenous insulin infusion along with dextrose-containing fluids in order to maintain glucose homeostasis. This might require a longer preoperative preparation time for obtaining intravenous access and initiating the infusions. The best glycemic control will generally be afforded by resuming the patient's normal management regimen as soon as possible. Involving an endocrinologist preoperatively to participate in planning for intra- and postoperative care is recommended.

Thyroid Disease

Thyroid disease is uncommon in childhood but is associated with certain pediatric conditions, including prematurity and trisomy 21. Hypothyroidism can lead to myocardial depression, arrhythmias, hypotension, hypothermia, or delayed gastric emptying, while hyperthyroidism can manifest as hyperthermia, tachycardia, hypertension, palpitations, or dysrhythmias. In addition, patients with very large goiters sometimes require imaging to exclude airway involvement. Both hypo- and hyperthyroidism have anesthetic and cardiovascular implications, and, whenever possible, patients should be euthyroid prior to an elective procedure.

Corticosteroids

Although there is little evidence to support the practice, many textbooks and practitioners advocate steroid supplementation during the perioperative period for patients receiving steroid therapy. Theoretically, chronic corticosteroid administration might suppress the hypothalamic–pituitary–adrenal (HPA) axis to the degree that an adrenal crisis is precipitated by the physiologic stress of surgery and anesthesia. In practice, patients who receive a short "pulse" of steroids (<14 days), for example, for treatment of an acute asthma exacerbation, generally do not require supplementation. The administration of "stress-dose" steroids is sometimes recommended for patients who have received supraphysiologic doses, multiple short courses of steroids, or chronic steroids. Adrenal suppression diminishes with time from completion of steroid therapy. In addition, the need for steroid supplementation and recommended doses and duration are also dependent on the degree of surgical stress. Patients exposed to minor surgical stress (hernia repair, extremity surgery) might require at most a single dose of hydrocortisone or methylprednisolone, whereas those who undergo a major operation (laparotomy or thoracotomy with blood loss requiring transfusion) might need multiple doses during the 2–3 day period of maximal physiologic stress. Consultation with an endocrinologist should be sought in these situations.

Anemia

The normal hemoglobin level varies with age. Term infants have a hemoglobin level between 14 and 18 g/dL, which, due to rapid weight gain and expansion of blood volume in the face of relatively low levels of erythropoietin, normally decreases to physiologic nadir of 9 or 10 g/dL by the age of 2–3 months. Preterm infants start with a lower hemoglobin level and have an even lower nadir of between 7 and 9 g/dL.

Hemoglobin is the most commonly requested preoperative laboratory test. Because the incidence of previously undetected anemia in healthy children undergoing elective surgery is extremely low (approximately 0.3 %), routine determination of hematocrit and hemoglobin is not necessary if the results of studies performed previously as part of well-child care have been normal. A selective hemoglobin determination should be performed in children with a chronic medical illness, those with acute blood loss (trauma, GI

bleeding), and those about to undergo procedures with the potential for significant blood loss. Infants younger than 6 months should have hemoglobin measured because of the nadir. In addition, in premature infants, hemoglobin levels of less than 10 g/dL have been associated with an increased incidence of postoperative apnea. Children of African ethnicity who have not been screened for sickle cell disease and have not had a hemoglobin determination after 6 months of age should have such measurements performed before undergoing a major surgical procedure.

Anemia results in a decrease in oxygen-carrying capacity and an increase in cardiac output. Most children with chronic anemia are in a well-compensated state. However, intraoperative blood loss can lead to decompensation in the face of surgical stress, systemic vasodilation, and myocardial depression caused by anesthetic agents. Further, the child with preoperative anemia is more likely to require a transfusion in the setting of moderate blood loss than children without anemia. Although the hemoglobin value at which individual anesthesiologists choose to transfuse varies greatly, most anesthesiologists allow a healthy child's hemoglobin to decline to the range of 7–8 g/dL before recommending a blood transfusion.

Sickle Cell Disease

Sickle cell anemia results from a single-base mutation in the β-globin gene. Under conditions of hypoxia, acidosis, dehydration, hypothermia, or the use of a tourniquet, HgbS can polymerize, causing sickling of red blood cells, resulting in microvascular occlusion, tissue ischemia, pain ("crisis"), and, when it occurs in the lung, impaired pulmonary function (acute chest syndrome). This is most common in children homozygous for the mutation but can also occur with one HgbS gene combined with another abnormal gene such as $HgbO_{Arab}$ or HgbC. The optimum hemoglobin level in patients with sickle cell disease is unknown, but recent studies indicate that simple transfusion to 10 g/dL is associated with morbidity no greater than that in patients treated with aggressive exchange transfusion to reduce the HgbS concentration to less than 30 %, which was the standard recommendation for many years. That is not to say that the rate of morbidity is low; in fact, it is around 20–30 % in both groups. These patients require (1) pre- and postoperative hydration, (2) careful attention to maintenance of normothermia, (3) avoidance of tourniquets whenever possible, (4) supplemental oxygen to avoid hypoxemia, and (5) good analgesia. Patients with sickle cell trait (Hgb AS) have no apparent perioperative risk of sickling or acute chest syndrome, except rarely in conditions associated with extreme dehydration and electrolyte depletion such as uncorrected GI losses from bowel obstruction.

Coagulation Disorders

Von Willebrand disease (vWD) is the most common congenital bleeding disorder. Most patients with vWD have type I disease, which is a quantitative deficiency of von Willebrand factor (vWF). Ninety percent of patients with type I vWD will respond to DDAVP with a two- to threefold increase in vWF. The DDAVP is administered intravenously, intranasally, or subcutaneously 30 min before the procedure. Because 10 % of patients with type 1 vWD do not respond to DDAVP, advance determination of the quality of the response is fundamental to the preoperative evaluation of a patient with vWD. Type 1 nonresponders, as well as patients with type 2 and type 3 vWD, require preoperative administration of plasma-derived factor VIII concentrate (Humate-P), which has a high concentration of vWF. All patients with vWD undergoing major surgical procedures require factor replacement preoperatively.

Hemophilia A, B, and C are inherited deficiencies of factors VIII, IX, and XI, respectively. Perioperative management of these patients depends on the procedure planned. Patients undergoing major surgical procedures require factor VIII and factor IX levels that approximate 100 % of normal from 30 min before the procedure through the first postoperative week. Factor administered to patients with hemophilia A can be plasma derived or recombinant, and the regimen should be discussed with the child's hematologist ahead of time. Recombinant factor VIII products have become available but are not necessarily associated with a lower rate of inhibitor or antibody formation. Patients undergoing minor procedures are usually fine with factor levels that are 50 % of normal for the first 2–3 postoperative days. Some patients with mild hemophilia A have a sufficient response to DDAVP to provide adequate protection for minor procedures. The coagulopathy of patients with hemophilia C does not directly correlate with factor levels. The need for fresh-frozen plasma transfusion in these patients should be determined by a pediatric hematologist.

Malignancy

Children with cancer frequently receive medications that have the potential to cause profound perianesthetic complications. Some receive prolonged doses of corticosteroids as part of their chemotherapy, which places them at risk for adrenal suppression. The anthracycline drugs, doxorubicin and daunorubicin, can cause myocardial dysfunction, whereas mithramycin, carmustine (BCNU), and bleomycin can cause pulmonary fibrosis, especially when combined with radiation therapy. The fact that this pulmonary damage can be exacerbated by supplemental oxygen is of concern to the anesthesiologist. The effects of these drugs are not always

apparent at the time of treatment and can present later in life or are unmasked by the additive effects of anesthetic agents (myocardial dysfunction) or oxygen exposure. As many protocols include serial echocardiographic evaluations, the most recent echocardiographic report should be included in the preoperative evaluation.

In addition to complications from chemotherapy and radiation, these children and their families frequently have psychological sequelae from prolonged treatment and the side effects associated with malignancy and bone marrow transplantation. They deserve careful evaluation and gentle treatment in the perioperative environment.

Anterior Mediastinal Mass

Patients presenting with an anterior mediastinal mass (especially lymphoma) are at particularly high risk of airway compromise and cardiovascular collapse with the induction of general anesthesia due to compression of the trachea or great vessels when intrinsic muscle tone is lost and spontaneous respiration ceases. Preoperative evaluation should begin with a careful history to elicit any respiratory symptoms, including dyspnea, orthopnea, stridor, or wheezing. A chest X-ray and complete echocardiogram must be performed, including evaluation of the great vessels with respect to compression of inflow or outflow tracts, the pericardium for direct infiltration or effusion, and the atria and ventricles with attention to degree of filling and the presence of atrial diastolic collapse. If it can be done safely, computed tomography should be obtained to assess the degree of tracheal and bronchial compression. Pulmonary function studies do not predict outcome or help to guide management and are no longer considered useful. Whenever possible, percutaneous biopsy of the mass or surgical cervical lymph node biopsy using local anesthesia with minimal sedation is preferred over a procedure performed under general anesthesia because it poses the least risk to the patient. If general anesthesia is required and airway or vascular compression exists, having rigid bronchoscopy or even ECMO capability on standby is strongly recommended.

Cerebral Palsy

Cerebral palsy is a polymorphic set of motor disorders with a wide spectrum of severity. Children with CP frequently require surgery to treat GERD or orthopedic problems. Many have increased oral secretions, dysfunctional swallowing, and chronic pulmonary aspiration of both oral and gastric contents. Together with an ineffective gag and inadequate cough, these commonly result in the development of reactive airway disease and recurrent pneumonitis. Up to one third of children with CP also have a seizure disorder. They are often taking several medications, including anticonvulsants, muscle relaxants, proton pump inhibitors, or H_2 blockers and drugs for reactive airway disease. Communication is important so that these essential medications are continued in the perioperative period. Confirmation of recent determination of adequate anticonvulsant blood level within the previous 6 months is helpful, although some patients have poorly controlled seizures and are expected to have seizures in the perioperative period despite adequate blood levels.

Preoperative assessment should include evaluation of room air oxygen saturation and the degree of underlying reactive airway disease, as well as the presence of snoring and other obstructive symptoms suggestive of inadequate airway tone. In the most severely affected patients, scheduling elective procedures between episodic exacerbations of reactive airway disease and aspiration pneumonia is challenging. Since many of these children have ongoing increased airway reactivity, preoperative evaluation and preparation should be directed to ensuring that the child's pulmonary status is as good as it can be. Chest radiographs are helpful in the child who has had frequent pneumonitis.

Hypotonia

Children with generalized hypotonia often present for definitive diagnosis by muscle biopsy under general anesthesia and should be considered at risk for malignant hyperthermia. Malignant hyperthermia precautions are commonly taken, consisting of avoidance of succinylcholine and potent volatile anesthetics. Patients with muscular dystrophy or myotonia are also at risk for MH and MH-like events with exposure to triggering agents. Succinylcholine should always be avoided in patients with Duchenne muscular dystrophy due to the risk of rhabdomyolysis.

Developmental Disorders

An increasing number of children are receiving pharmacotherapy with stimulant medications for attention deficit disorder. Although the American Heart Association recommends that an electrocardiogram be performed prior to initiation of stimulant therapy to identify significant cardiac conditions (LQTS, hypertrophic cardiomyopathy, Wolff–Parkinson–White syndrome), the American Academy of Pediatrics does not agree with this recommendation. There is no evidence to suggest that patients with these diagnoses are at higher risk of sudden cardiac death with stimulant medications than the general population. Therefore, in the absence of a personal history, family history, or physical exam findings suggestive of cardiac disease, no additional testing or evaluation is required prior to anesthesia and surgery.