Introduction

Williams–Beuren syndrome, commonly known as Williams syndrome (WS), results from a deletion on the long arm of chromosome 7q11.23, which code for the elastin gene (ELN). The ELN gene encodes tropoelastin, which is involved in elastic vascular wall fiber assembly. Reduced elastin formation results in widespread arteriopathy (1,2). In addition to supravalvar aortic stenosis (SVAS), patients with WS can develop supravalvar pulmonic stenosis, obstructive coronary disease, and diffuse aortic stenosis.

Several cases of cardiac arrest in patients with WS in the setting of sedation and anesthesia have been described. Burch described a series of 19 pediatric patients, all of whom, treated with various anesthetic and sedative techniques in multiple settings, suffered cardiac arrest. The patients presented with varying degrees of cardiac disease, including coronary artery disease, supravalvar pulmonic stenosis, and SVAS. The cause of cardiac arrest in these 19 cases was suspected to be myocardial ischemia caused by reduced coronary artery blood flow (2).

Even though patients with WS have a known risk of adverse events with sedative and anesthetic drugs, they often need anesthesia for various procedures throughout their lives. Many patients with WS have some central nervous system involvement and have an average IQ of 41–80, developmental delay, and significant procedural anxiety, which can sometimes make even painless procedures (e.g., transthoracic echocardiograms, computed tomography scans, dental procedures) unattainable without sedation (3,4). In addition, the workup for surgical management of severe cardiac disease often requires more invasive imaging techniques such as cardiac MRI or cardiac catheterization, which usually require prolonged sedation or general anesthesia for pediatric patients. Therefore, thorough preoperative workup and risk assessment are essential for proper management to reduce risk. In this article, the authors (a multidisciplinary team of cardiologists, cardiac anesthesiologists, and cardiac intensivists) review the clinical
manifestations of WS, propose a consensus, expert-informed method to estimate anesthetic risk based on the current literature, and provide recommendations for periprocedural management of patients with WS.

Clinical characteristics of WS with anesthetic implications

Facial appearance and airway

Patients with WS have distinctive facial features, highlighted by a broad forehead, epicanthal folds, short nose with a broad tip, long philtrum, and wide mouth with a thick vermillion of the lips. They often have mandibular hypoplasia and wide-spaced teeth with malocclusion. The small mandible and dental anomalies may make tracheal intubation difficult, though many of the documented cases of difficult airway in patients with WS are in adults (5,6).

Cognitive and developmental traits

Significant developmental delay with average IQs of 40–80 is typical for this patient population (3), with the largest deficit in the area of visuospatial functioning. Hypotonia and joint laxity often result in delays in achieving developmental milestones. The gregarious ‘cocktail party’ personality without social anxiety commonly associated with these patients can quickly evolve into anxiety and agitation in the setting of an invasive medical procedure such as anesthetic induction. Anxiety is a common feature of WS, and specific phobias have been reported in 36–56% of individuals (3,7). Procedural anxiety, often related to mask induction and IV placement, is also common.

Gastrointestinal disease

Feeding difficulties and failure to thrive occur in 70% of infants with WS (7). Gastrointestinal reflux is common, and the presence of hypercalcemia may worsen abdominal symptoms such as abdominal pain, feeding intolerance, and constipation. The poor weight gain and feeding problems often necessitate medical and sometimes surgical intervention.

Endocrine disease

Hypercalcemia occurs in approximately 15–50% of patients with WS (7,8), usually in infancy, with resolution by age 4; the cause is unknown. It frequently presents as irritability in infants, to such a degree that imaging procedures can be difficult without sedation (9), and it often leads to hypercalcuria, nephrocalcinosis, and abnormalities on electrocardiogram (ECG). Hypothyroidism is also seen in 10% of patients with WS (7).

Cardiac disease

Approximately 80% of patients with WS have cardiovascular disease, and 40% of those cases require an intervention or surgery (10–12). Most forms (93%) of cardiovascular disease in patients with WS are discovered by age 1 year (13). Although some lesions can progress with time, most patients with WS who require surgery for cardiovascular disease will need treatment in their first decade of life (10,13). The most common forms of cardiovascular disease in this patient population are discussed below.

Supravalvar aortic stenosis

SVAS is an aortic narrowing at the level of the sinotubular junction (Figure 1). It occurs in 45–75% of patients with WS (14,15), and most present before adulthood. Because of a microdeletion on chromosome 7q11.23, reduced elastin leads to arteries with reduced elasticity. Over time, the vessels develop smooth muscle hypertrophy and thickening of the vascular media (16–19). Often, a ridge of tissue forms at the sinotubular junction. The stiff, noncompliant aorta predisposes the aortic valve leaflets to early degeneration, and these leaflets then become adherent to the formed sinotubular ridge (1,14). SVAS can progress with time in patients with moderate to severe stenosis, but it remains stable in most patients (14).

Pulmonary artery stenosis

Pulmonary artery stenosis is present in 83% of patients with WS (14,15,20). Although peripheral pulmonary stenosis is most frequently seen, central pulmonary artery stenosis may also occur. Supravalvar pulmonary stenosis often occurs in conjunction with SVAS, and the presence of both defects is thought to increase perianesthetic risk, as shown in both case series by Burch and Gupta (2,18,21). Many cases of pulmonary artery stenosis tend to improve with time (13).

Coronary arteriopathy

Coronary anomalies occur in 45% of patients with SVAS (15). Coronary artery disease can occur in isolation or in conjunction with SVAS (Figure 2). It is estimated to occur in 5–9% of patients with WS (13). Commonly, the left coronary ostium can develop a silt-like stenosis caused by adhesion of the left aortic valvar leaflet to the sinotubular ridge that has formed. In addition, the sinotubular ridge may proliferate to such a degree that it impairs blood flow to the coronary
In the presence of SVAS, the coronary arteries are exposed to high prestenotic pressure that can also lead to coronary dilation and early atherosclerosis (14,16,20). It should be noted that the literature contains case reports of pediatric patients with WS who have coronary disease in the absence of SVAS (11,22).

**Left ventricular hypertrophy**

Left ventricular hypertrophy (LVH) is found on ECG in 40% of patients and commonly correlates with the degree of obstructive lesion (23). In a study of 15 patients with WS, LVH was also found on echocardiogram to occur in the absence of SVAS and hypertension (23,24). LVH may progress with time and may vary in its severity. The presence of LVH leads to altered myocardial geometry, abnormal contractility, and reduced left ventricular diastolic function.

**Systemic hypertension**

Systemic hypertension can occur in up to 55–60% of patients with WS (25). The etiology is still unclear; although it is often related to renal artery stenosis (60%), other hypotheses include vascular lesions caused by widespread arteriopathy associated with elastin deficiency, hypercalcemia, and increased sympathetic activity (9,25,26).

**Extracardiac vascular anomalies**

Renal artery stenosis is seen in up to 60% of patients with WS (25) and in 60% of patients with WS and hypertension (26). Thoracic aortic stenosis, or middle aortic syndrome, which is estimated to occur in up to 30% of patients with WS (27), involves aortic narrowing from the aortic root to the lumbar aorta. It represents a severe arteriopathy that results from an elastin defect.
and often occurs in conjunction with coronary artery stenosis. Other vascular stenosis is also common, including mesenteric, carotid, and peripheral arterial stenosis.

**Electrophysiologic abnormalities**
A prolonged corrected QT (QTc) is reported in 14% of patients with WS (28) and is thought to be caused by myocardial ischemia. The QT prolongation tends to worsen with increased heart rate, suggesting that microvascular perfusion defects may contribute to the prolonged QT in patients with WS (29), which is similar to the mechanism for QT prolongation caused by ischemia seen in patients with hypertrophic cardiomyopathy and coronary artery disease (30).

**Mechanism of ischemia in Williams–Beuren syndrome**
The presence of SVAS by itself is a risk factor for coronary ischemia because of both increased oxygen consumption and reduced oxygen supply. Significant obstruction of the left ventricular outflow tract leads to the development of LVH, increasing ventricular wall tension, and myocardial oxygen consumption. In adults with valvar aortic stenosis, ischemia and angina pectoris are often present despite the absence of coronary artery disease (31). Several theories exist as to the mechanism of ischemia in patients with obstruction of the left ventricular outflow tract, including (a) a decrease in coronary reserve to the hypertrophied left ventricle (32) and (b) abnormal hypertrophy with small coronary development in the presence of obstruction (31).

Beyond the ischemia caused by aortic stenosis, the reduction in oxygen delivery to the myocardium in patients with WS occurs via several mechanisms. The reduced elastin production leads to noncompliant vessels, which reduces diastolic aortic recoil, which in turn, reduces coronary blood flow. The presence of (i) coronary arteriopathy, (ii) ostial stenosis caused by adherence of the valvar leaflet or a thickened sinus wall, or (iii) both further impedes coronary blood flow to the already threatened myocardium. The presence of LVH further increases the risk of subendocardial ischemia, owing to increased myocardial wall tension, reduced subendocardial perfusion, and abnormal coronary autoregulation (33).

Given the high likelihood of subendocardial ischemia in certain patients with WS, any medications that further impair coronary blood flow or lead to increased oxygen consumption will increase the risk of cardiac arrest. Several anesthetic agents are known to cause direct myocardial depression, reduced systemic vascular resistance (SVR), or increased myocardial oxygen consumption, any of which can cause ischemia in this already vulnerable population. In the reported cases of cardiac arrest in patients with WS, drugs that altered the balance of myocardial supply and demand by reducing SVR or significantly increasing heart rate were often cited as the cause of cardiac arrest (2,20). A review of the cardiovascular mechanisms of commonly used anesthetic drugs is provided in Table 1.

In addition to the physiologic effects of anesthetic agents, both induction and emergence often are associated with brief periods of increased sympathetic activity, which may increase heart rate and blood pressure, thus increasing oxygen consumption (34,35).

**Preoperative workup and estimation of anesthetic risk**
A thorough preanesthetic history and physical examination should focus on all clinical manifestations of WS, with an emphasis on cardiovascular disease. Guidelines from both the Williams Syndrome Foundation and the American Academy of Pediatrics recommend a preoperative assessment—including an assessment of the degree of developmental delay and preprocedural anxiety—1 to

<table>
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<tr>
<th>Drug</th>
<th>SVR</th>
<th>Myocardial depression</th>
<th>Tachycardia and increased myocardial oxygen consumption</th>
<th>Dyshrhythmias</th>
<th>Respiratory depression with possible reduced oxygen delivery</th>
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<td>Propofol</td>
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<td>Inhalational anesthetics</td>
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<tr>
<td>(sevoflurane, isoflurane, desflurane)</td>
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<tr>
<td>Barbituates (thiopental, pentobarbital)</td>
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<td>Ketamine</td>
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<td>Chloral hydrate</td>
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<td>Benzodiazepines (midazolam, lorazepam)</td>
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<td>Etoimide</td>
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2 weeks prior to any planned anesthetic (26,36). An airway assessment should screen for mandibular hypoplasia and dental anomalies, which might make intubation and airway management difficult. Testing for hypercalcemia is recommended every 2 years by the American Academy of Pediatrics (26), and these results should be obtained prior to administering anesthesia. A baseline thyroid test should be available and repeated if signs of hypothyroidism are present. Cardiac assessment in all patients with WS should screen for symptoms of myocardial ischemia. Perspiration with feeds or dyspnea with feeding, both of which lead to failure to thrive, may be present in infants with active ischemia. An ECG should be performed to rule out LVH, ST-T wave abnormalities, and prolonged QT, all of which might indicate latent ischemia. An echocardiogram should be obtained to assess the degree of LVH, the severity of supravalvular aortic or pulmonary stenosis, the patency of coronary orifices, and of course, the cardiac function to specifically identify abnormalities in wall motion. Systemic hypertension may help to identify those at increased risk, and its occurrence in the absence of renal artery stenosis is thought to result from widespread arteriopathy caused by elastin deficiency (25). The more severe the deficiency of elastin, the less compliant the vasculature, and one could hypothesize an increased risk of anesthesia-related events, though no evidence supports this hypothesis. Thoracic aortic stenosis may be a risk factor for coronary ostial stenosis; one report found a 19% incidence of coronary stenosis in patients with stenosis of the thoracic aorta (27). All patients with moderate-to-severe SVAS, biventricular outflow tract disease, documented coronary anomalies, or a combination of any of the three should be treated as high risk. Those patients with WS and QT prolongation should also be considered at high risk for ischemia and ventricular dysrhythmias. Patients with repaired congenital heart disease should be assessed based on the type of defect corrected, the degree of residual defects, and the timing of the repair, with recent repairs classified as higher risk.

As with all patients, the risk of the surgical procedure should also be considered when determining preoperative risk. Although all patients with WS might be considered at higher risk, the perioperative management for those with significant cardiac disease differs greatly from that for those with mild cardiovascular disease. A classification of clinical and imaging data based on preoperative risk is provided in Table 2. Even though no large, prospective studies have been published that allow practitioners to estimate anesthetic risk in this patient population, the assumption of risk is an expert-derived consensus based on known physiologic risk factors for myocardial ischemia. In addition, anatomical anomalies known to be associated with more severe disease, such as stenosis of the thoracic aorta and disease of the biventricular outflow tract, are included. The total picture of a patient’s risk factors should be considered. Those patients with obstruction of the biventricular outflow tract should be considered at highest risk. In the published case series of patients who suffered cardiac arrest under anesthesia, most patients had moderate-to-severe SVAS, often with obstruction of the biventricular outflow tract (2,20).

Management of anesthesia

Regardless of preoperative risk, careful planning of the anesthetic technique to reduce the incidence of myocardial ischemia is essential in all patients with WS. Anesthetic goals in those patients, summarized by Burch, include preservation of sinus rhythm; maintenance of preload, contractility, and SVR; avoidance of anesthetic drugs that cause physiologic changes that may worsen ischemia; and avoidance of increased pulmonary vascular resistance (2) (Table 3).

Table 2 Classification of risks of Williams syndrome

<table>
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<tr>
<th>Low risk</th>
<th>Moderate risk</th>
<th>High risk</th>
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<tr>
<td>Normal ECG</td>
<td>Mild stenosis of a branch of the pulmonary artery</td>
<td>Severe SVAS (&gt;40 mmHg)</td>
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<tr>
<td>Normal Echocardiogram</td>
<td>Hypertension</td>
<td>Symptoms or ECG signs consistent with ischemia</td>
</tr>
<tr>
<td>Minimal extracardiac anomalies</td>
<td>Mild-to-moderate SVAS (&lt;40 mmHg)</td>
<td>Coronary disease demonstrated in imaging</td>
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<td>Other mild cardiac anomalies (e.g., ventricular septal defect)</td>
<td>Severe left ventricular hypertrophy</td>
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<td></td>
<td>Repaired SVAS or SVPS without residual gradients</td>
<td>Biventricular outflow tract disease</td>
</tr>
<tr>
<td></td>
<td>Mild left ventricular hypertrophy</td>
<td>Prolonged QTc on ECG</td>
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<td></td>
<td>Mild to moderate SVPS in isolation</td>
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<td></td>
<td>Significant extracardiac disease such as</td>
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<td></td>
<td>difficult airway or severe gastroesophageal reflux</td>
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ECG, electrocardiogram; SVAS, supravalvar aortic stenosis; SVPS, supravalvar pulmonary stenosis.
### Table 3 Anesthetic management of (a) low-risk, (b) moderate-risk, and (c) high-risk patients with WS

<table>
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<th>Preanesthesia planning</th>
<th>Induction and maintenance of anesthesia</th>
<th>Anesthesia emergence and disposition</th>
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<tr>
<td><em>(a)</em> Schedule preanesthesia visit 1–2 weeks prior to planned surgery focusing on airway, development, cardiac, gastrointestinal, renal disease Continue beta-blockers on day of anesthesia Induce anesthesia in an area suitable for resuscitation (operating room, catheterization lab, ICU) Have resuscitation drugs, including vasopressors, immediately available</td>
<td>Minimize npo (nothing by mouth) time and liberalize preoperative oral fluids May need oral premedication prior to induction with midazolam, pentobarbital, ketamine Use full ASA monitors, including 5-lead ECG Use slow, titrated mask induction or IV induction with ketamine, etomidate, narcotics, or small doses of propofol Employ balanced anesthetic with drugs that reduce myocardial oxygen consumption and minimize reductions in SVR Treat ST changes with vasopressors</td>
<td>Monitor for emergence tachycardia and ECG changes Aggressively manage postoperative pain and shivering to reduce oxygen consumption Monitor after anesthesia for at least 2 h</td>
</tr>
<tr>
<td><em>(b)</em> Consider transfer to center with pediatric cardiology and pediatric anesthesia for all nonurgent anesthesia Schedule preanesthesia visit 1–2 weeks prior to planned surgery, focusing on airway, development, cardiac, gastrointestinal, renal disease Continue beta-blockers on day of anesthesia Induce anesthesia in an area suitable for resuscitation (operating room, catheterization lab, ICU) Have resuscitation drugs, including vasopressors, immediately available</td>
<td>Minimize npo time and liberalize preoperative oral fluids May need oral premedication prior to induction with midazolam, pentobarbital, ketamine Use full ASA monitors, including 5-lead ECG Use slow, titrated mask induction or IV induction with ketamine, etomidate, narcotics, or small doses of propofol Employ balanced anesthetic with drugs that reduce myocardial oxygen consumption and minimize reductions in SVR Treat ST changes with vasopressors</td>
<td>Monitor for emergence tachycardia and ECG changes Aggressively manage postoperative pain and shivering to reduce oxygen consumption Recover in high-acuity setting with capability for resuscitation Monitor after anesthesia for at least 6 h, and preferably overnight, prior to discharge</td>
</tr>
<tr>
<td><em>(c)</em> Pediatric or cardiac anesthesia team involved in care should have experience in managing patients with WS Transfer to facility with ECMO availability Continue beta-blockers on day of anesthesia Induce anesthesia in an area suitable for resuscitation (operating room, catheterization lab, ICU) Ensure that ECMO team is aware and clear-primed ECMO circuit is nearby Have resuscitation drugs, including vasopressors, immediately available</td>
<td>Place IV for prehydration May need oral premedication prior to IV placement, with small doses of midazolam, ketamine, or pentobarbital Use full ASA monitors, including 5-lead ECG Induce via IV with etomidate, fentanyl, ketamine Use slow, titrated mask induction or IV induction with ketamine, etomidate, narcotics, or small doses of propofol Employ balanced anesthetic with drugs that reduce myocardial oxygen consumption and minimize reductions in SVR Treat ST changes with vasopressors Consider direct monitoring of myocardial contractility with echocardiogram in more complicated or prolonged cases Treat ST changes with vasopressors Monitor status of resuscitation with echocardiogram to evaluate contractility Deploy ECMO early if standard resuscitation measures are unsuccessful</td>
<td>Monitor for emergence tachycardia and ECG changes Aggressively manage postoperative pain and shivering to reduce oxygen consumption Recover in high-acuity setting with capability for resuscitation and ECMO deployment Monitor after anesthesia for at least 6 h, and preferably overnight, prior to discharge</td>
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A pediatric anesthesiologist should ideally be involved in the preoperative and intraoperative care of all children with WS up to 3–4 years of age (26,36). All sedation and anesthesia should be performed with 5-lead ECG monitoring to detect ischemia, in an environment with both space and equipment available for resuscitation.

Administering of anesthesia for moderate-risk patients with WS should occur in centers with intensive care capabilities by a clinician with significant experience in caring for such patients. Centers without such provisions should consider transferring moderate- and high-risk patients to institutions with pediatric cardiac facilities for nonurgent procedures. If the urgency of the procedure precludes transfer, the anesthesiologist should prepare to respond rapidly to even small physiologic changes that may quickly develop into cardiac arrest.

High-risk patients should be anesthetized for procedures that are necessary for the diagnosis and treatment of their cardiovascular disease only in a setting with the capability for deployment of extracorporeal membrane oxygenation (ECMO). All possible steps should be taken to hydrate patients preoperatively to augment left ventricular filling pressures, including liberalizing fluid intake and avoiding prolonged fasting times. If possible, patients should have an intravenous catheter placed for prehydration before induction of anesthesia. In addition to standard resuscitation medications, vasopressors such as phenylephrine and vasopressin should be immediately available and used as first-line agents to treat hypotension and ST-T wave changes indicative of myocardial ischemia. Joffe et al. (37) recommended using transthoracic echocardiogram to direct resuscitation (by assessing for wall motion abnormalities and worsening outflow tract obstruction) and to guide fluid and drug therapy.

Optimal care of the patient with WS requires a thorough understanding of the physiologic effects of anesthesia in the setting of the patient’s cardiac disease. It has been reported that in high-risk patients, even low and incremental increases in sevoflurane concentrations have led to cardiac arrest (2). In situations in which IV access is difficult, mask induction with high concentrations of sevoflurane should be limited to low- and moderate-risk patients. Intramuscular ketamine is a good option for providing sedation to obtain IV access in a resistant patient with significant heart disease. Ketamine maintains contractility and SVR while it increases heart rate. Ketamine may produce tachycardia that increases myocardial oxygen consumption; however, it has been used successfully in high-risk patients, both at our institution and elsewhere (38,39). Several options exist for intravenous induction with a drug that maintains contractility and coronary perfusion while it ensures intravenous access to give vasopressors rapidly. Etomidate has been used with success at our institution and elsewhere in such patients (2,38,40,41). High-dose opiate inductions have long been used safely in patients at risk for myocardial ischemia (39). Dexmedetomidine has several potential physiologic benefits in patients at risk for myocardial ischemia, including a reduction in heart rate and initial increase in SVR. It has been used with success in high-risk patients by the authors in combination with etomidate and ketamine as part of a balanced anesthetic. Regardless of the anesthetic technique, high-risk patients may still become ischemic despite the best efforts of the anesthesia team; therefore, vigilance, quick response to physiologic changes, and planning for resuscitation that includes ECMO availability is of utmost importance. Prior to administration of any anesthetics to high-risk patients with WS, a member of the care team should ensure that the patient’s family understands the risk of cardiac arrest, the possible need for ECMO, and the importance of reviewing all advance directives.

The anesthetic management of patients with prolonged QT is beyond the scope of this review. However, in those 14% of patients with WS and prolongation of the QT interval, it might be best to avoid anesthetic drugs that significantly increase the QT interval, such as 5HT3 inhibitors (ondansetron) (42). Patients are often treated with beta-blockers preoperatively to reduce tachycardia, myocardial oxygen demand, and risk of ventricular dysrhythmias. Treatment should be continued on the day of surgery to avoid rebound tachycardia. Patients who are being treated adequately with beta-blockers may not respond to standard doses of beta-adrenergic drugs such as epinephrine and may require higher doses to increase contractility (43). The postanesthesia disposition of same-day surgical patients with WS is controversial. The return of a patient’s physiology to baseline after an anesthetic depends on several factors, including the drugs used for anesthesia, the anesthetic technique, and the procedure performed (34). Cardiac arrest can occur in patients with WS on the day after anesthesia (2,21), and the possibility of undiagnosed coronary disease may exist in all patients with WS (22). Although low-risk patients may be appropriate for discharge on the day of surgery, postoperative monitoring, including cardiovascular monitoring with ECG, is essential in all patients with WS. Moderate-risk patients should be admitted for prolonged observation to a location with capabilities for continuous monitoring and resuscitation, with the possibility of overnight observation. High-risk patients should recover from all procedures that require sedation.
and anesthesia in a monitored setting with capabilities for ECMO deployment, such as an intensive care unit. The care team should discuss postanesthesia disposition prior to all procedures that require sedation and anesthesia, and should be determined by a clinician with significant experience in caring for patients with WS.

**Future research**

Currently, several studies are being conducted to address the genetics of WS and the cognitive and behavioral characteristics of the disease. Researchers at our institution are developing a multisite registry of patients with WS and their anesthesia profiles, with the goals of characterizing the clinical factors that might identify those at high risk for anesthesia and determining the safest anesthetic technique to use in caring for these patients.

**Conclusions**

Patients with WS continue to challenge anesthesiologists. Optimal management involves multidisciplinary cooperation between anesthesiologists, critical care physicians, cardiologists, and surgeons, as well as thorough perioperative preparation for all procedures that involve anesthesia.

**Acknowledgments**

The authors thank Tzipora Sofare, MA, for her valuable editorial assistance.

**Ethics approval**

Ethics approval was unnecessary for this review.

**Funding**

The study was funded by departmental resources.

**Conflict of interest**

All authors have no conflicts of interest to report.

**References**


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