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SURAVALVAR AORTIC STENOSIS REPAIR IN CHILDREN WITH WILLIAM'S SYNDROME

Surgical Specialty:	Cardiac Surgery
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Background:

- General Considerations
    - Williams Syndrome (WS) is a multisystem disorder caused by a chromosome 7 microdeletion which results in cardiovascular, endocrine, and neurodevelopmental abnormalities[1]
    - Patients with WS often require several anesthetics starting in the first year of life for a range of procedures from noninvasive imaging to full aortic repair on cardiopulmonary bypass
    - Sudden cardiovascular collapse is a well-documented complication of patients with WS, and the risk of major cardiovascular morbidity and mortality is as high as 9% in the perioperative period[2]
- Patient Considerations
    - Cardiovascular abnormalities are present in 80% of patients with WS and are the most common cause of morbidity and mortality[3]
    - Elastin gene defects lead to widespread arteriopathy, most notably causing supravalvar aortic stenosis (SVAS), pulmonary artery stenosis (PAS), and coronary artery anomalies[4]
    - Increased risk for QT prolongation
    - Extracardiac manifestations of WS[1]
        - Endocrine: glucose intolerance, diabetes mellitus, hypothyroidism, hypercalcemia
        - Neurodevelopmental: global cognitive impairment, hyperacusis, highly social & empathetic personality, anticipatory anxiety

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Outpatient anticipatory guidance for patients with WS[5]
        - Cardiology evaluation including 3-limb blood pressure measurements and echo every 3 months until age 1, yearly until age 5, and every 2 years thereafter
        - EKG yearly

        - Thyroid function tests yearly until age 3, every 2 years thereafter
        - Serum calcium concentration every 4 months until age 1, every 4-6 months until age 2, every 2 years thereafter
    - Additional labs/tests indicated during work-up
        - Recent echo, EKG, and BMP per the above guidelines
        - CT/MRA are often performed to evaluate the involvement of other vessels
        - CBC, type & crossmatch
        - If diabetic: blood glucose morning of the procedure

Table 1. Risk stratification and anesthetic considerations for patients with WS[4,6].

	Low risk	**Moderate risk**	**High risk**
Classification	Age >20 years, no cardiac involvement greater than mild SVAS or branch PAS, normal EKG, no renal artery involvement	HTN, mild-moderate SVAS (mean gradient <40mmHg), mild PAS, mild bilateral outflow obstruction, repaired SVAS or PAS without residual gradients, renal artery stenosis, renal dysfunction, QTc >450ms but <500ms, airway abnormalities, lung disease, or severe gastroesophageal reflux	Age <3 years, history of an adverse cardiovascular event, preprocedural arrhythmia, bilateral outflow tract obstruction of ≥ moderate severity, severe SVAS (mean gradient ≥ 40mmHg), coronary artery involvement, diffuse stenosis of the thoracic aorta, ≥ moderate LVH or RVH, symptoms or EKG signs of ischemia, QTc ≥ 500ms
Preanesthestetic considerations	Preanesthesia visit 1-2 weeks before the scheduled surgery, continue beta-blockers, induce in an area suitable for resuscitation (OR, ICU), and have resuscitation drugs immediately available	Consider transfer to a center with pediatric cardiology and pediatric anesthesia	Transfer to center with ECMO availability, pediatric or cardiac anesthesia team with experience in managing patients with WS, ensure that ECMO team is aware and clear-primed ECMO circuit is nearby, place preoperative IV for prehydration

(SVAS = supravalvar aortic stenosis, PAS = pulmonary artery stenosis, EKG = electrocardiogram, HTN = hypertension, LVH = left ventricular hypertrophy, RVH = right ventricular hypertophy)

    - Discussions to have with surgeon/family
        - Preoperative fluid status: Minimize NPO time, liberalize preoperative oral fluids, consider preadmission for hydration in high-risk patients
        - ECMO candidacy
        - Code status
- Specific or Unique Room Set-up Requirements
    - Airway
        - Standard airway equipment for patient age/weight
        - Dental disease and facial dysmorphisms may increase airway difficulty; consider video laryngoscope or fiberoptic scope
    - Drugs/Infusions
        - May benefit from premedication with midazolam or dexmedetomidine to help avoid abrupt increase in heart rate pre-induction
        - Vasopressor boluses and infusions should be available for induction including phenylephrine and vasopressin which offset decrease in SVR caused by anesthetic and elicit a baroreceptor reflex so decrease heart rate in a favorable manner
        - Antifibrinolytics: Tranexamic acid, aminocaproic acid
        - Post-bypass vasopressors/inotropes: Dopamine, epinephrine, milrinone

- Monitors
  - Standard ASA monitors
  - 5-lead EKG
  - Arterial and central venous pressure monitoring
  - Transesophageal echocardiography
  - NIRS
  - Cutaneous defibrillator pads for high risk patients given risk of significant arrthymia
- Blood Availability
  - Red blood cells should be available before incision
  - After separating from bypass: Platelets, cryoprecipitate, and other products as needed
- PICU Bed Availability: Cardiac ICU for recovery

Intraoperative Considerations:

- General
  - While catheterization lab procedures may be performed in the setting of isolated PAS, SVAS with a mean gradient of >40-50mmHg will require surgical intervention[7]
  - Simple aortic repair may be appropriate in the setting of isolated SVAS, though extended ascending and aortic arch patch plasty is often required
  - Care should be performed by providers with specific pediatric cardiac training and experience with patients with WS
- Induction[4]
  - Low or moderate risk: Slow, titrated mask induction with nitrous oxide and sevoflurane may be safe with IV placement as soon as possible
  - High risk: Preoperative IV placement and hydration; induce with IV etomidate, fentanyl, ketamine. Some institutions have policies of two pediatric cardiac faculty in attendance for the induction of high-risk patients.
  - Have vasopressor bolus doses and infusions ready, treat EKG changes quickly and aggressively
  - Post-induction placement of arterial and central venous catheters
    - Discuss with the surgeon whether innominate cannulation for antegrade cerebral perfusion is planned to determine optimal arterial catheter placement
    - If available, refer to preoperative imaging for vascular anomalies or known stenotic vessels
- Positioning
  - Supine with shoulder roll
- Maintenance
  - Judicious use of inhaled anesthetic
  - Optimize use of opiates, benzodiazepines, ketamine, and dexmedetomidine
  - Avoid vagolytic medications (atropine, glycopyrrolate) to mitigate tachycardia
- Hemodynamic/Physiologic goals
  - Maintain preload, afterload, and contractility
  - Avoid tachycardia
- Surgical Considerations
  - Potential for early blood loss if the patient has had a previous sternotomy
  - Monitor for and treat arrhythmias and ST changes during surgical manipulation
  - Muscle relaxation is required throughout the operation
  - Deep hypothermic cardiac arrest is used if the aortic arch requires intervention

  - Post-bypass considerations: Balance of avoiding hypertension given aortic suture lines vs. possible preexisting LVH requiring higher blood pressure
- Emergence/Disposition
  - Treat emergence tachycardia, pain, and shivering aggressively to reduce oxygen consumption
  - Recovery in pediatric cardiac ICU
- Post-op Pain
  - Continuous opiate infusion while intubated
  - Consider long-acting opiate at end of procedure if hemodynamics permit
  - Regional nerve or field block can be considered as an adjunct for analgesia

Case-Specific Complications/Pitfalls

- Perioperative cardiac arrest is a known risk factor for patients with WS undergoing any procedure, particularly cardiac surgery
- Patients with WS should be followed closely as outpatients with scheduled evaluations as above, with recent cardiac and laboratory workups available before any procedure
- Major adverse cardiac events are most common after combined LVOT/RVOT procedures and occur more frequently in patients with lower age and weight at the time of surgery, preoperative complications, and longer cross-clamp times[2]
- Careful planning, preparation, and emergency backup should minimize the risk of perioperative complications

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