Anesthesia for Patients with Williams Syndrome

Shahin H. Ismail (Author) and Matthew Knight, M.M.Sc., CAA (Advisor)
Nova Southeastern University, Department of Health Science, Jacksonville, FL

Introduction and Background

Williams Syndrome (WS) is a rare genetic disorder posing significant perioperative risks due to its cardiovascular, craniofacial, and behavioral challenges. **Supravalvular Aortic Stenosis (SVAS)**, the most common anomaly, increases risks of ischemia and instability. Craniofacial abnormalities like mandibular hypoplasia and subglottic narrowing complicate airway management, requiring advanced tools. Behavioral traits such as heightened anxiety and hypersensitivity further complicate preoperative preparation and recovery.

Dental abnormalities often necessitate general anesthesia, adding complexity to care. A multidisciplinary approach involving thorough preoperative assessment, advanced intraoperative techniques, and vigilant postoperative monitoring is essential for optimizing safety and outcomes.

While medical advancements and collaborative care models have improved outcomes, inconsistencies in tailored protocols and limited access to specialized equipment in resource-limited settings remain significant challenges.

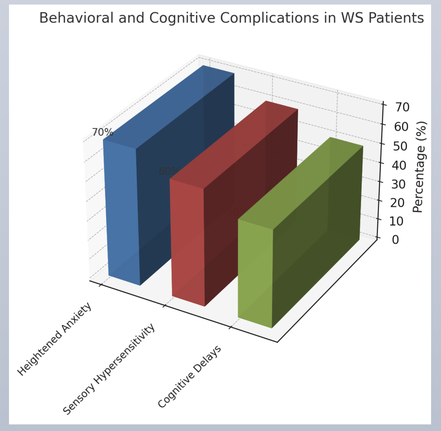

Fig. 1. Behavioral and cognitive complications in WS patients. Data adapted from Collins RT (2020) and Pober BR (2021)

Clinical Findings

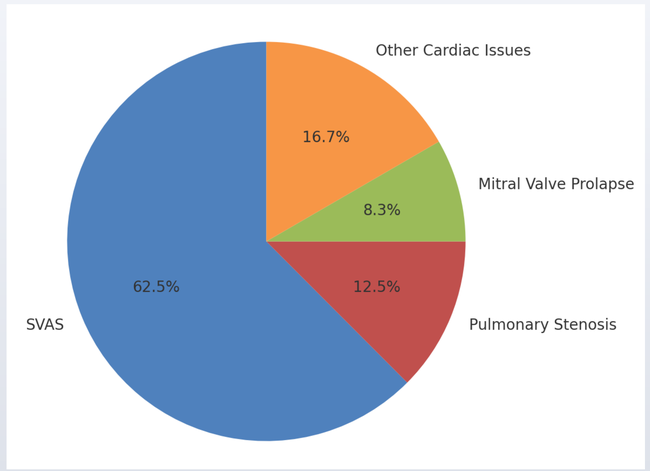

Fig. 2. Distribution of cardiovascular complications in WS patients. Data adapted from Collins RT (2020) and Pober BR (2021)

Supravalvular Aortic Stenosis (SVAS) affects 75% of Williams Syndrome (WS) patients, posing significant perioperative risks. Other cardiovascular anomalies, such as pulmonary stenosis (15%) and mitral valve prolapse (10%), add to the complexity, highlighting the need for thorough preoperative cardiac evaluations. Craniofacial anomalies, including mandibular hypoplasia and subglottic narrowing, make intubation challenging and necessitate the use of advanced tools like video laryngoscopy and fiberoptic intubation. Additionally, heightened anxiety, sensory hypersensitivity, and cognitive delays further complicate perioperative care, requiring preoperative sedation and a calm, controlled environment to reduce stress and improve cooperation.

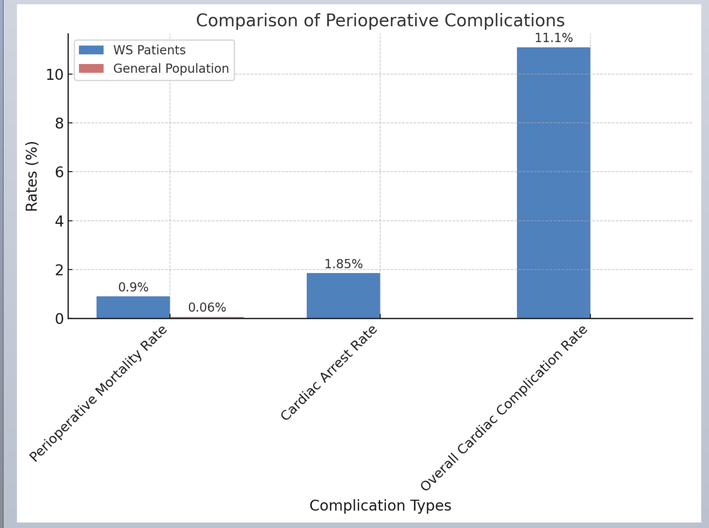

Fig. 3. Perioperative complications in WS patients compared to the general population (Olsen et al., 2014; Dencker et al., 2021)

Complications

Patients with Williams Syndrome face significantly higher perioperative risks compared to the general population due to cardiovascular, airway, and behavioral challenges. A perioperative mortality rate of 0.9% and cardiac arrest rate of 1.85% underscore the importance of preoperative cardiac evaluations, particularly for conditions like supravalvular aortic stenosis (SVAS). Craniofacial abnormalities increase the risk of difficult intubation, indirectly contributing to higher complication rates. Additionally, heightened anxiety and sensory hypersensitivity further complicate anesthetic management, emphasizing the need for meticulous planning and multidisciplinary approaches to minimize risks.

Discussion

Managing Williams Syndrome (WS) patients during anesthesia is challenging due to inconsistent protocols, limited access to advanced tools, and variability in collaboration among specialties. Comprehensive preoperative evaluations, advanced airway techniques, and behavioral support are essential but often underutilized. Simplified, evidence-based guidelines focusing on preoperative imaging, advanced airway devices, and continuity of care are needed. Streamlined protocols and ongoing education will enhance safety and promote consistent, patient-centered approaches across care teams.

Conclusion

Williams Syndrome (WS) poses unique perioperative challenges due to cardiovascular anomalies, craniofacial abnormalities, and behavioral issues. Comprehensive preoperative evaluations, including cardiac and airway assessments, are critical to mitigate risks like supravalvular aortic stenosis (SVAS) and difficult intubation. Tailored anesthetic strategies, such as advanced airway tools and preoperative sedation, enhance safety and cooperation. Multidisciplinary collaboration among anesthesiologists, cardiologists, and behavioral specialists is essential for improving outcomes.

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