**Anesthetic Management for Craniosynostosis Repair in a Child with Williams Syndrome**

Michael Davis MD, Mohamed Mahmoud MD, Rajeev Subramanyam MD, Ali Kandil DO, MPH
Cincinnati Children’s Hospital Medical Center, Department of Anesthesia, – Cincinnati, Ohio

**CASE REPORT/BACKGROUND**

17 month-old female with Williams Syndrome and sagittal craniosynostosis presents for posterior vault remodeling.

Pertinent Medical History:
- Moderate Supravalvar Aortic/Pulmonic Stenosis
- Micrognathia-glossophsis syndrome
- Tracheostomy/Ventilator-dependence
- Global developmental delay

There have been only 2 published case reports on children with Williams Syndrome and craniosynostosis (1).

This presentation highlights anesthetic management and considerations in this complex patient.

**ANESTHETIC MANAGEMENT**

- Gentle inhalational induction
- Exchange uncuffed tracheostomy tube for cuffed tracheostomy tube
- Two 22g saphenous intravenous catheters placed
- Ultrasound-guided placement of radial arterial line for tight blood pressure monitoring
- 1 MAC of Sevoflurane with 70% FiO2
- Remifentanil infusion at 0.1 - 0.3 mcg/kg/min
- Low-dose phenylephrine infusion at 0.1mcg/kg/min
- In-line tracheal suctioning available throughout case
- Transport to pediatric ICU postoperatively

**DISCUSSION**

Williams syndrome is a well-known genetic syndrome caused by a microdeletion on chromosome 7q11.23 encompassing the elastin gene. It is characterized by distinctive facial, congenital cardiovascular malformations, intellectual disabilities, and various other manifestations (2).

In children with Williams Syndrome, the risk of sudden death raises many anesthetic considerations for perioperative management.

Various types of acquired and congenital heart defects have been reported in patients with Williams Syndrome:
1) supravalvar aortic and pulmonic stenosis,
2) coronary artery lesions,
3) mitral valve prolapse,
4) coarctation of the aorta,
5) patent ductus arteriosus,
6) peripheral arterial abnormalities, and
7) intra-cardiac lesions including ventricular septal defect and tetralogy of Fallot.

These patients not only have an increased risk of sudden death, but in many cases, the cardiac arrests have been refractory to standard resuscitative algorithms. Coronary artery stenosis and severe biventricular outflow obstruction are most commonly implicated. Fatalities in these patients most commonly occur either immediately after incision or after surgical completion (3).

**REFERENCES**