

Difficult Anesthetics for Neonates with Chiari II Malformation

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Introduction: Myelomeningocele is an in-utero neural tube development defect. Eighty percent of the patients may develop hydrocephaly associated with Arnold-Chiari type II malformations.¹ Patients with Chiari II malformation may require urgent posterior fossa decompression, cervical laminectomy, or ventricular drainage procedure due to herniation of the cerebellum, medulla oblongata kinking and / or hydrocephaly. These patients may develop respiratory depression, apneic spells, retraction, swallowing difficulties and vocal cord paralysis and stridor and urologic and orthopedic anomalies.² We present two safe anesthetics on two consecutive days on a patient with a Chiari II malformation managed safely with general anesthesia (intravenous and inhalational anesthesia) followed by a cardiac arrest six hours later by an inadvertent manipulation of the neck airway while the patient was on the pediatric ward.

Case Report: A one day old patient with spina bifida presented for a meningomyelocele repair. Past medical history included having been born cyanotic and with respiratory distress, meconium staining and presented on a 50% oxygen hood. The anesthetic was uneventful. He was discharged home on day eight. Four days later, he presented at a local emergency room with apnea, difficult breathing and was therefore transferred to our institution to rule out sepsis and meningitis. The anesthetic for the MRI was administered with a 1 and ½ LMA and propofol drip at 70-80 mcg/kg/min and two doses of midazolam 0.4 and 0.2 mg with plans of not manipulating the neck. On several occasions, the heart rate would increase to 200 bpm or decrease to 80 bpm without any major manipulation and the patient would occasionally become apneic with Cheyne Stokes breathing. The MRI showed marked ventriculomegaly with proteinaceous or blood layering in the ventricles. The posterior fossa was small with herniation of the cerebellar tonsils through the foramen magnum to C5 consistent with the syrinx in Chiari II malformation.

A second anesthetic was required the following day for central line placement for antibiotic administration. Again, a 1 and ½ LMA was placed after an induction with fentanyl 1.5 mcg and propofol 6 mg and anesthesia was maintained with oxygen, air and Sevoflurane. Heart rate again increased occasionally to about 200 bpm and decrease to 90 bpm without any significant stimulation with periods of apnea. The patient was taken to PACU after an uneventful anesthetic. Six hours later while on the ward, the patient was turned to the side to check on the meningomyelocele site and the patient arrested. His trachea was then intubated and the patient transferred to the PICU.



Discussion: Patients with Chiari II malformation are a risk during anesthesia: airway management and postoperative apneic episodes. We present a patient with a syrinx and cerebellar herniation to C5 with ventriculomegaly secondary to accumulated blood or material to whom we administered two anesthetics without airway manipulation and successfully discharged the patient from PACU. Six hours later after the second anesthetic, while checking the meningomyelocele site, the patient was turned on the side and arrested and was then transferred to the PICU. We emphasize on the importance of neck manipulation on patients with Chiari malformation presenting with syrinx and acknowledge that some may require cervical protection and postoperative PICU admission.

References:

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