A 3 year old male, 8.4 kg, presented for palatoplasty, hearing test and bilateral myringotomy tube placement. On examination he had distinctive facial features of Wolf-Hirschhorn Syndrome (WHS) that included cleft palate, prominent glabella, microcephaly, micrognathia, microsomia, and generalized hypotonia. He is developmentally delayed and has a history of seizures that is controlled with Keppra. He has an ASD and no other cardiac abnormality. He was born at 39 weeks and had a three-month NICU stay for congenital anomalies.

His past surgical history includes laparoscopic gastrostomy tube placement, cleft lip repair, and CT Scan of the brain under anesthesia.

On this occasion, he received midazolam PO for premedication and then an inhalational induction with sevoflurane was performed. The larynx was visualized and an oral RAE tube was placed. A smaller than expected, for his age, laryngoscope blade and endotracheal tube were used, Miller #1 and a 4.0 uncuffed oral RAE ETT. However, there was no leak and the ETT was changed to a 3.5 uncuffed oral RAE tube.

Although there have been some publications in the anesthesia literature regarding management of WHS patients, anesthesiologists are not overly familiar with the condition and how to appropriately care for these patients.

Preoperatively, the assessment of these patients should include a review of neurological symptoms, including seizures and current anti-epileptics, pulmonary function, GERD, and associated cardiac defects. A physical exam noticing characteristic facial features, developmental delay, hypotonia, skin changes, and possible cardiac murmurs should also be performed.

As pulmonary function may be compromised in these patients, a chest x-ray or blood gas may be necessary to assess the condition as pulmonary function tests are likely to be difficult to ascertain, secondary to these patients inability to cooperate (1).

Micrognathia, microsomia, and microcephaly may result in difficult direct laryngoscopy and the presence of oro-facial clefts may make securing the endotracheal tube in place difficult as well (2). As with our patient, smaller endotracheal tubes and airway equipment may be necessary and smaller doses of muscle relaxants used when there is associated hypotonia.

As with most patients with seizures, anti-epileptics should be continued and the pharmacologic effects of these medications are relevant especially with regard to effects on drug metabolism and coagulation.

The coagulation effects seen from some anti-convulsants are also relevant when a WHS patient presents for a surgical procedure that could benefit from a neuraxial block. In addition to coagulation abnormalities, it is important to note that these patients may have kyphoscoliosis further complicating epidural placement (1).

In conclusion, WHS patients present a challenge to the anesthesiologist not only in airway management but also with regard to the associated neurological, pulmonary, and cardiac features.

References: