Anesthesia for Children with Wolf-Hirshhorn Syndrome: A Report and Literature Review
Shireen Mohiuddin, James Mayhew, Mohammad Y Qasim
Department of Anesthesiology, Division of Pediatric Anesthesiology, University of Arkansas for Medical Sciences

Introduction: Wolf-Hirshhorn is a rare syndrome due to deletion of the short arm of chromosome four. This syndrome is also referred to as the 4p-syndrome and was first described by Cooper and Hirshhorn in 1961 (1). We present two cases of children with Wolf-Hirshhorn syndrome who presented for surgery at our institution, and discuss their anesthetic management and literature review.

Results: Case 1 This nine month-old male infant presented for open reduction of the left hip. Past history revealed a diagnosis of Wolf-Hirshhorn syndrome. He was born full term with a birth weight of 2.5 kg. He had significant hypotonia with spasticity. At one month of age he was diagnosed with laryngomalacia. He had mild developmental delay, severe gastroesophageal reflux, mild pulmonary stenosis and hearing loss. His past surgical history included an epiglottoplasty and an esophagastroduodenoscopy. At eight months of age an open reduction of his hip was attempted, but difficulty with intravenous access, an attempted caudal block and inability to place a Foley catheter by a pediatric urologist resulted in postponement of the case. There was no reported difficulty with his airway.

In the operating room he was premedicated with subcutaneous midazolam HCl and oral subacute bacterial endocarditis prophylaxis was given. The patient was monitored with pulse oximetry, non-invasive blood pressure, electrocardiogram, and precordial stethoscope. Anesthesia was induced using oxygen, nitrous oxide and sevoflurane. Due to his history of difficult intravenous access, a surgical consult had been obtained to accomplish central line placement at time of this surgery. A caudal block was then achieved without difficulty. Since a Foley catheter could not be placed at the previous attempt at surgery, a Texas catheter was placed by the surgeon. The patient’s trachea was intubated without difficulty and the surgical procedure went uneventfully, although surgical time was approximately four times the original estimate.

Upon completion of the surgery and anesthetic, the trachea was extubated when the patient was awake and responding. His recovery was uneventful.

Case 2 This 8 month-old was a full term infant delivered by spontaneous vaginal delivery at 39 weeks gestation. He had severe intrauterine growth retardation, weighing 1698g at birth. Chromosomal studies proved that he had Wolf-Hirshhorn syndrome. He had bilaterally small kidneys which were otherwise normal. His echocardiogram at birth was normal. On exam he had hypertelorism, broad forehead, small nose and low-set ears. He had mild gastroesophageal reflux, and suffered from motor skill delay.

He presented to the operating room for bilateral myringotomy tubes and auditory-evoked potentials because he failed hearing tests on three occasions.

After placing the standard ASA monitors, anesthesia was induced with 70% nitrous oxide and 8% sevoflurane. Bilateral myringotomy tubes were placed by the otolaryngologist, and the patient was then taken to the post-anesthesia care unit (PACU). His stay in the PACU was uneventful. He received 120 mg of acetaminophen. He was feeding without difficulty and was sent home.

Discussion: Children diagnosed with Wolf-Hirshhorn syndrome are born with intrauterine growth retardation and are thus small for gestational age, as were our patients who were born at term by date, but weighed only 2.5 kg and 1.698 kg, respectively. Their characteristics included microcephaly with mental retardation. They may also have epilepsy. Cardiac lesions included pulmonary stenosis, ventricular septal defects, atrial septal defects and patent ductus arteriosus. Skeletal anomalies include absent pubic rami, congenital hip dislocation and scoliosis (2).

Review of the literature reveals only one case report of anesthesia in a child with Wolf-Hirshhorn syndrome (3). In this report the authors suspected that their patient had malignant hyperthermia. Although this particular patient had definite evidence of malignant hyperthermia and was treated appropriately, our experience would suggest that patients with the Wolf-Hirshhorn syndrome are probably not at high risk for malignant hyperthermia. However, as the authors of that report suggest, continual vigilance is needed when anesthetizing these children.

References: