Cri du Chat (CdC) is a syndrome from partial deletion of chromosome 5 that presents unique challenges to the anesthesiologist. Anomalies of the airway such as micrognathia, abnormal epiglottis, hypotonia, and alterations in the hard and soft palate create potential for a difficult airway and the characteristic cry of the cat phonation of newborns. Severe mental retardation, can make pain management and perioperative cooperation a challenge. Other manifestations may be cardiac, neurologic and/or renal which may present additional challenges for the anesthesiologist. 1,2

Case: An 18 year-old, 40kg, female with CdC and ESRD presented for a cadaveric renal allotransplantation. Her manifestations included characteristic changes in her voice, mental retardation, mild hyptonia and micrognathia. She also had potassium of 5.2 mEq/L and history of hypertension and a parathyroidectomy. Her family reported no history of problems with anesthesia. Airway exam revealed a MP Class I airway, normal neck extension and micrognathia (see image).

An IV induction was performed and after ventilation was successful with the assistance of an oral airway, cisatracurium was administered. Laryngoscopy with both Miller and MAC blades revealed a Grade 4 by two different skilled anesthesiologists. A video laryngoscope provided a Grade 2 view, but the tube was unable to be passed due to the acute angle needed to reach the anterior positioned larynx. An attempt with the fiberoptic bronchoscope failed due to secretions. The video laryngoscope was then used to visualize the vocal cords and the fiberoptic bronchoscope was used as a flexible stylette for successful intubation. Subsequently, an arterial line and additional IV access was obtained. Unilateral transversus abdominis plane (TAP) block was performed for post-operative analgesia. Real-time ultrasound in an in-plane approach using a 22 gauge 50mm block needle was utilized and 2.5mL aliquots of 0.25% bupivacaine were injected to a total of 12.5mL.

Summary:

CdC is not a disease of just young children--the anatomical and behavioral challenges persist into adulthood. Special attention must be accorded to the airway, even if the exam is relatively normal. If hypotonia is present preoperative medication and intraoperative neuromuscular blockers should probably be avoided, as there is a potential for airway compromise. Furthermore, postoperative pain control is difficult in patients with limited ability to understand and/or cooperate, and non-narcotic analgesia can be extremely helpful. In our case, both a TAP block and narcotics kept the patient comfortable throughout the recovery room stay, and into the first postoperative day. This case highlights multiple perioperative challenges in a patient for whom extra caution cannot be understated.
