Diastrophic Dwarf with a Difficult Airway and Malignant Hyperthermia for Urgent Cesarean Section

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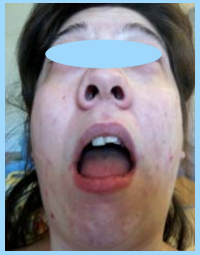
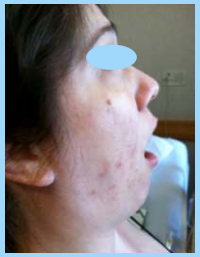
Introduction

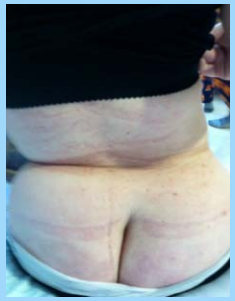
Diastrophic dysplasia (DD) is an autosomal recessive disorder characterized by an abnormality of the cartilage matrix. The mutation leads to many anomalies including short stature, spinal deformities, severe progressive scoliosis, cleft palate, retrognathic mandible, laryngeal and tracheal stenosis. Cervical instability can occur secondary to malformed vertebrae and kyphosis of the cervical spine. Patients with this disease are at increased risk for difficult intubation. Diastrophic dysplasia is not associated with a higher rate of malignant hyperthermia.





Case

A 31 year old G1P0 presented at 39 weeks gestation with breech presentation contracting. She had DD with a height of 4'9" (after 5" limb lengthening). She had severe lumbar scoliosis but her cervical spine did not appear deformed. She had a Mallampati IV/IV airway with a high arched palate, large upper teeth and tongue. Her vital signs were within normal limits including a SaO_2. She was NPO except for clears 6 hours prior to presentation. The patient had an extensive surgical history. In all nine procedures in which she was put to sleep she had an easy mask airway but intubation was difficult. Direct laryngoscopy with Mac 3s and Miller 2s gave grade IV views each time. Airway adjuncts including Eschmann stylet, Bullard scope, fiberoptic bronchoscope and LMAs had been used to assist intubation. All her anesthetics had been performed with malignant hyperthermia precautions since a "febrile reaction" to general anesthesia as a 2 year old. The patient refused regional anesthesia secondary to a severe and persistent paresthesia after a lumbar puncture. An awake intubation with topical local anesthesia and sedation (propofol 200mg, midazolam 4mg) using a fiberoptic bronchoscope was attempted. After multiple failed attempts, an intubating LMA #4 was place but the ETT could not be passed. The procedure proceeded with the LMA. Anesthesia was maintained with a propofol drip and nitrous oxide. The baby had APGAR scores of 1 and 8 at 1 and 5 minutes. Mother and baby suffered no complications.



Discussion

Patients with DD pose many challenges to the anesthesiologists. Cervical instability must be considered and precautions must be taken during manipulation of the airway. There is at least one case report of a patient with severe cervical kyphosis becoming quadriplegia during anesthesia.[1] Significant scoliosis may lead to restrictive lung disease and even pulmonary hypertension. Respiratory compromise in the mother may force delivery of the neonate prior to term.[2] Difficulty with intubation is common secondary to high incidence of cleft palate, posteriorly positioned upper and lower jaws and larygno-tracheal stenosis [3] Neuraxial anesthesia is not contraindicated but may be difficult. Optimal dosing of the spinal and/or epidural is not known so small doses with titration should be chosen.[4]

References

1.Poussa et al. Spine 1991;16:881-7
2.Ayoubi et al. Lancet 2001;358:1778
3. Karlstedt et al. Am J Med Genetics 1997;72:266-2-74
4.Porter et al. Int J Ob Anes. 2007;16:145-8