POST-OPERATIVE RESPIRATORY COMPLICATIONS AFTER PALATOPLASTY IN A 19 MONTH OLD FEMALE WITH CORNELIA DE LANGE SYNDROME

- A Case Report -

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Abstract

Cornelia De Lange syndrome is a rare genetically heterogeneous and sporadic syndrome, with an estimated prevalence of 1 in 10,000 to 30,000. The disorder may present many complications during anesthesia due to cardiac, gastrointestinal and airway anomalies. We report a case of an ex premature toddler presenting for repair of a cleft palate. Postoperatively she had respiratory distress, successfully treated by the anesthetic care team Causes for the complication are discussed.

Case Report

A 19 month old former 34-week gestational age female with a history of Cornelia De Lange Syndrome presented for closure of a cleft palate. Additional past medical history was significant for reflux disease treated by Nissen fundoplication, failure to thrive managed with G-tube placement, and hearing loss for which she had undergone left myringotomy tube placement. Previous anesthetic records indicated she was easily ventilated with an oral airway but a Grade 3 view was achieved during prior intubation attempts with a Miller 1 blade. On examination, with a deep bite, her lower front incisors bit into her malformed palate.

Induction of anesthesia was achieved with sevoflurane and nitrous oxide in oxygen. A WIS blade 1.0 revealed an anterior airway with a grade 3 view which improved to a grade 2 view with cricoid pressure. The first attempt to pass a 4.0 endotracheal tube failed. The next attempt with a 3.5 cuffed tube was successful. Intravenous access was secured. Anesthesia was maintained with isoflurane in oxygen plus intermittent fentanyl. At the conclusion of surgery, the patient's mouth and gastric tube were suctioned; she was moving spontaneously. The trachea was extubated and she was given blow-by oxygen. Her oxygen saturation remained satisfactory for approximately five minutes. She was then moved to her hospital bed at which time acute desaturation to the 60s occurred. The patient was moved back to the OR table and ventilated with 100% inspired oxygen. She remained otherwise hemodynamically stable. Dexamethasone 4 mg and glycopyrrolate 30 mcg were given in addition to albuterol nebulization. An oral airway was placed with the surgeon's approval and mask ventilation with continuous positive pressure reestablished. No upper airway obstruction or swelling could be observed, but auscultation of her lungs revealed rhonchi bilaterally. After approximately one hour, the inspired oxygen was decreased from 100% with assisted ventilation to blow-by oxygen. At time of transport, she remained hemodynamically stable

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with oxygen saturations above 92%. Over the next few hours, the oxygen saturation improved to 99%. There were no further episodes of desaturation. Chest X-ray was clear. She was discharged to home after 3 days.

Discussion

Cornelia de Lange syndrome (CdLS) was first documented by Brachman in 1916[1] and later by a Dutch pediatrician, Cornelia de Lange in 1933[2,3]. The syndrome consists of multiple congenital anomalies resulting in a distinctive facial appearance, prenatal and postnatal growth deficiency (95%), feeding difficulties (reflux in 90%), psychomotor delay, behavioral problems, and associated malformations that mainly involve the upper extremities (80% have clinodactyly, bradydactyly or olidactylyl). Congenital heart disease occurs in 20-30% and hearing loss in 60%[4,5]. The most diagnostic aspect of the disease are the facial characteristics which include: confluent eyebrows, long curly eyelashes, low anterior and posterior hairline, underdeveloped orbital arches, well-defined arched eyebrows (as though they had been penciled), long philtrum, thin lip, depressed nasal bridge, high arched/cleft palate, and micrognathia (Fig. 1). A gene responsible for CdLS-NIPBL on chromosome 5-was discovered in 2004 jointly by researchers at the Children's Hospital of Philadelphia, USA and researchers at Newcastle University, UK[6,7]. In 2006, a second gene-SMCIA on the X chromosone-was found by Italian scientists. A third gene discovery was announced in 2007. The gene SMC3 is on chromosone 10 and was also discovered by the research team in Philadelphia. The latter two genes seem to correlate with a milder form of the syndrome. NIPBL and SMC3 have autosomal dominance and SMCIA, an X linked dominant pattern of inheritance. All genes are involved in sister chromatid cohesion. There is no known cure but the syndrome can be managed by treating associated clinical symptoms. Sixty six percent of CdLS individuals die during the first year of life. Mortality occurs primarily from aspiration in infancy and from infection and bowel obstruction there after[4,5]. However, some children live until adulthood.The vast majority of cases are due to spontaneous mutations, although the defected gene can be inherited from either parent, making it autosomal dominant.

Anesthetic concerns include high risk of aspiration, upper airway obstruction and difficult intubation, unpredictable response to drugs due to endocrine disorders and renal failure, and adverse effects of pulmonary hypertension or congenital cardiac anomalies. Pulmonary hypoplasia also predisposes the patient to respiratory infection post operatively.

The etiology of our patient's postoperative respiratory distress was probably multifactorial. She may have had undiagnosed sleep apnea and thus was extremely susceptible to sedatives. There may also have been residual effects of prematurity as she remained very small for her age at the time of surgery. Opioid-induced respiratory depression and residual inhalational anesthetic effects are other possibilities. As prolonged surgery was performed in the area, postoperative edema of the oropharynx may also have had some additive effect. Most likely, it was a combination of these factors. The importance of close communication between all members of the perioperative team and careful monitoring of vital signs, especially oxygenation, is emphasized. These patients should not be candidates for day care surgery or anesthetic management in an out patient facility, including a dental office.

In an attempt to identify patients and learn more about the syndrome, the Cornelia de Lange Syndrome (CdLS) Foundation was established as a nonprofit, family support organization based in Connecticut, USA. It provides materials for public education and information. In addition to Reaching Out, a bi-monthly newsletter, the Foundation produces and distributes

several other publications on the syndrome, as well as a video The foudation can be reached at info@cdlsusa.org Ph:001 860-676-8166 860-676-8255 Toll-Free Support Lines: 800-753-2357 (United States)

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