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An Infant with Crouzon Syndrome Presenting with Reversible Chronic Airway Obstruction

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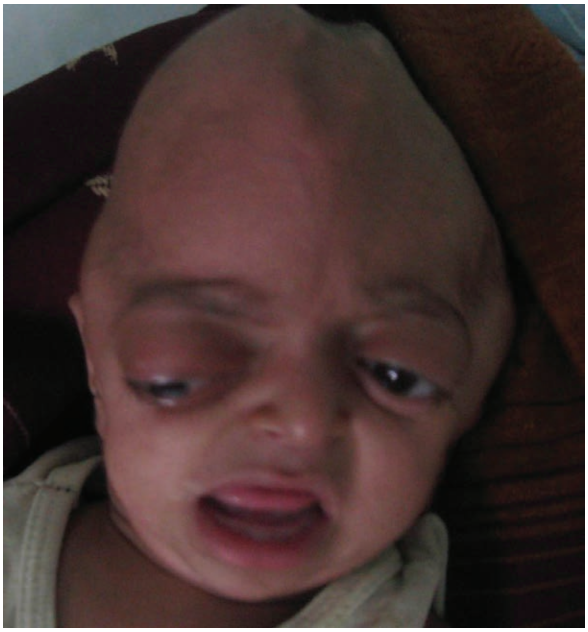

Crouzon syndrome is a craniofacial dysostosis characterized by premature closure of calvarial, cranial base, orbital, and maxillary sutures resulting in a cloverleaf-shaped skull with midface hypoplasia, exophthalmose, hypertelorism, and beaked nose, as shown in figure 1 and Supplemental Digital Content, video 1 (http://links.lww.com/ALN/C346). Diagnosis is suggested by such phenotypic features that are often present at birth. Genetic testing is confirmatory. Children with Crouzon syndrome may develop respiratory distress at any time during their growth, including shortly after birth. Reduced nasopharyngeal dimensions with some degree of choanal atresia often result in obligate mouth breathing.[1,2] Soft palate obstruction, relative macroglossia, glossoptosis, and vocal cord palsy secondary to acquired Chiari malformation can cause stridulous breathing, obstructive apnea, and even sudden death. Symptoms may improve after ventriculoperitoneal shunt surgery (Supplemental Digital Content, video 2, http://links.lww.com/ALN/C347). Emergency airway management in a child with Crouzon syndrome is challenging. Personnel and equipment to enable immediate tracheostomy in the event of failed oxygenation should be on standby. Midfacial hypoplasia and proptosis can make a facemask seal difficult. Gently holding the mouth open and pressing down the mask may improve the seal while overcoming oropharyngeal obstruction. Alternatively, spraying the mouth with topical lidocaine before induction will allow placement of an oral airway at light depths of anesthesia.[2] A smaller than expected endotracheal tube is recommended. A nasopharyngeal airway fitted with a 15-mm endotracheal tube adaptor that permits connection to an anesthesia circuit can be a rescue option to maintain airway patency and spontaneous ventilation, while the level of anesthesia sufficient for laryngeal mask airway insertion or fiberoptic-guided tracheal intubation is reached.[3]

Competing Interests

The authors declare no competing interests.

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Supplemental Digital Content is available for this article. Direct URL citations appear in the printed text and are available in both the HTML and PDF versions of this article. Links to the digital files are provided in the HTML text of this article on the Journal's Web site (www.anesthesiology.org). Portions of this work were presented previously at the Difficult Airway Society Meeting 2014, Stratford-upon-Avon, United Kingdom, November 12–14, 2014.

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