Airway management of a child with frontometaphyseal dysplasia (Gorlin Cohen syndrome)

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Abstract
Frontometaphyseal dysplasia (FMD), also called Gorlin-Cohen syndrome, is a rare hereditary X-linked dominant craniofacial bone disorder. The presentation describes the airway management of a 2-year-old child suffering from FMD with significant retrognathia, posted for major long bone corrective osteotomy. Induction with a combination of dexmedetomidine and ketamine preceded a successful endotracheal intubation under spontaneous ventilation.

Key words: Dexmedetomidine, difficult airway, frontometaphyseal dysplasia, ketamine

Introduction
Frontometaphyseal dysplasia (FMD), also called Gorlin Cohen syndrome, is a hereditary X-linked dominant syndrome described in 1969 with less than 30 cases described in the literature.[1,2] This case report of a child with FMD is presented owing to the rarity of the syndrome and the anticipated difficult airway, which was successfully managed by using a combination of dexmedetomidine and ketamine while preserving spontaneous ventilation.

Case Report
A 2-year-old female child, a known case of FMD, presented for open reduction of the left hip with osteotomy of femur. Physical examination revealed a slender undernourished girl of 8 kg with prominent supraorbital ridges, ocular hypertelorism, low set ears and a wide bridge nose with prominent eyes. Airway examination revealed a Mallampatti score of III with significant retrognathia, high arched palate with malocclusion of teeth. In addition, she had dorsolumbar scoliosis, pectus carinatum, bowing of long bones with distal phalangeal hypoplasia and multiple joint dislocations [Figure 1].

Pre-operative blood investigations, echocardiography and chest X-ray were within physiological limits. Oral midazolam 4 mg was selected for premedication. On arrival to the operation theatre, monitors were connected and child pre-oxygenated for 5 minutes.

Injection dexmedetomidine 1 μg/kg was administered for 10 min and then a continuous infusion at 1 μg/kg/h was set for the duration of the remaining procedure. Ketamine was administered in increments of 5 mg up to 12 mg until there

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**Figure 1:** X-ray lateral view
Discussed was no response to jaw thrust while ensuring spontaneous respiration. Just before direct laryngoscopy intravenous lignocaine 12 mg was administered. Rigid laryngoscopy with Miller 1 straight blade offered a Grade IV Cormack and Lehan view. An additional dose of ketamine 5 mg was administered. After optimal external laryngeal manipulation, the visible glottic chink was sprayed with topical lignocaine and tracheal intubation was successfully performed using an uncuffed 4 sized endotracheal tube. Anesthesia was continued with N₂O in 40% O₂ along with a continuous Dexmedetomidine and atracurium infusion. Adequate padding was provided at pressure points and extreme caution was exercised during positioning. At the end of the surgery tracheal extubation was successful.

Discussion

FMD belongs to the otopalatodigital spectrum syndromes that includes four phenotypically related conditions, otopalatodigital syndrome Types 1 and 2, FMD and Melnick-Needles syndrome.[1,2] The most common manifestations include supraorbital hyperostosis, hypertelorism, down-slanting palpebral fissures, broad nasal bridge and micrognathia with anomalies of teeth and generalized skeletal dysplasia. Congenital heart disease, subglottic tracheal narrowing and genitourinary anomalies, muscular hypotonia.[3,4]

Micrognathia, microstomia and malocclusion of teeth may make direct laryngoscopy impossible; therefore, a well-planned airway strategy is mandatory.

Ketamine combined with dexmedetomidine was selected for induction of anesthesia. Ketamine was preferred in our case of anticipated difficult airway due to it’s inherent sympathomimetic actions devoid of respiratory depression alongwith provision of excellent analgesia and amnesia.[5,7] Dexmedetomidine a specific and selective α₂-adrenoceptor agonist known for its sedative, anxiolytic, analgesic properties was used to complement ketamine. At the same time dexmedetomidine offsets the sympathomimetic effects of ketamine, also decreases the sialorrhea and emergence delirium associated with ketamine.[8,9] This unique pharmacological combination in the present case preserved the respiratory drive, allowed maintenance of a patent airway and provided sufficient sedation, analgesia and anesthesia to allow successful airway control. In addition, topical lignocaine was used as per recommendation of Aroni et al.[5] which states that ketamine does not depress coughing or swallowing reflexes.

Available literature describes the use of combination of both these drugs in children during procedural anesthesia and not as a complete anesthesia protocol in a challenging case.[10] The present experience of using this combination successfully paves the way to emerging new solutions for management of a difficult pediatric airway.

Hence safety profile, rapid onset of action with adequate sedation and analgesia provided by the ketamine and dexmedetomidine make them a distinctive drug combination in the pediatric difficult airway situation in a child with FMD.

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