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Successful airway management with use of a laryngeal mask airway in a patient with CHARGE syndrome

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Abstract
CHARGE syndrome refers to an autosomal dominant disorder in patients with coloboma (C) of the eye, heart disease (H), atresia of choanae (A), retarded growth (R), genital hypoplasia (G), and ear anomalies (E). In addition to these typical features, airway abnormalities, including retrognathia, glossoptosis, and laryngeal paralysis, have been reported in patients with this syndrome. This report describes a case of CHARGE syndrome observed in a 6-year-old male patient with a difficult airway, in whom anesthesia was managed successfully using a laryngeal mask airway.

Key words CHARGE syndrome · Airway management · Laryngeal mask airway · Difficult airway

CHARGE syndrome manifests as multiple malformations [1,2]. This syndrome comprises coloboma of the eye, heart disease, atresia of choanae, retardation (of growth or development), genital hypoplasia, and ear anomalies (abnormal pinnae or hearing loss). This report describes a case of CHARGE syndrome in a patient with a difficult airway, in whom anesthesia was successfully managed by using a laryngeal mask airway (LMA).

A 6-year-old male patient with perceptive deafness was admitted to Kyoto University Hospital, for left cochlear implantation. The patient had a height of 100 cm and a body weight of 13.6 kg. Examination revealed characteristic features, including coloboma of the eye, heart disease (e.g., patent ductus arteriosus), retarded growth, genital hypoplasia, ear anomalies, and hearing loss, thus leading to a diagnosis of CHARGE syndrome. At 2 years of age, the patient had undergone cryptorchidopexy and percutaneous occlusion of patent ductus arteriosus with a controlled-release coil, under general anesthesia with mask ventilation. At 5 years of age, the patient had been scheduled to have cochlea implantation at this hospital. After the induction of general anesthesia, two well-experienced anesthesiologists unsuccessfully tried to insert an endotracheal tube. Direct laryngoscopy showed a Cormack Lehane score of IV with the aid of the backward-upward-rightward pressure (BURP) maneuver. The surgery was canceled and anesthesia was aborted due to the failure to secure airway control. Subsequently, the patient was also diagnosed to have sleep apnea syndrome (SAS), and continuous positive airway pressure (CPAP) therapy was indicated.

One year after the canceled surgery, artificial auris internal implantation was again scheduled, due to progressive hearing loss. Routine preoperative laboratory tests showed no abnormalities. At the preoperative examination, the patient's Mallampati score was 4. Because of the history of difficult airway, we devised a strategy to secure the airway as follows: firstly, assessment of the anatomy using a fiberoptic scope; secondly, intubation of the trachea with an endotracheal tube (ETT) under bronchofiberscopic guidance; thirdly, insertion of a ProSeal LMA (The Laryngeal Mask, Jersey, UK); and finally, securing of the airway via a tracheostomy. General anesthesia was induced with sevoflurane in oxygen under spontaneous ventilation. In order to prevent saliva secretion, 0.1 mg atropine was administered intravenously. Direct laryngoscopy showed a Cormack Lehane score of IV with the aid of the BURP maneuver. We tried to observe the vocal cords by passing a bronchofiberscope (BFS) through the nasopharynx. Due to osseous choanal atresia, a flexible BFS of 2-mm outer diameter could not be passed down the nasal cavity. Thereafter, we tried to observe the patient's epiglottis using the flexible BFS via the mouth. The tip of the epiglottis was observed, but neither the vocal cords nor the arytenoid cartilage was observed due to a reduction in the oxygen saturation

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(S_{PO_2}) during the observation period. Tracheal intubation using an ETT was thus abandoned. We then decided to use the ProSeal LMA to secure the airway. A size 2.5 ProSeal LMA was selected. It was impossible to advance the mask when its tip had reached the back of the throat. Then, after the unsuccessful use of a size 2.0 ProSeal LMA, a size 1.5 ProSeal LMA was finally inserted smoothly without the aid of any muscle relaxant. After the LMA insertion, the position of the LMA was evaluated using the oropharyngeal leak pressure and a fiberoptic scoring system [3]. A gastric tube was inserted via the drain port of the LMA. The oropharyngeal leak was determined by mechanical ventilation with a peak airway pressure of 20 cmH_2O while monitoring for any gas leakage into the mouth [4]. No gas leakage was detected. For fiberoptic scoring, a BFS was passed to the position proximal to the mask aperture bars, and the vocal cords and anterior epiglottis were observed. Anesthesia was maintained by the use of sevoflurane and 60% nitrogen in oxygen, with intermittent supplementation of fentanyl without muscle relaxant under spontaneous respiration with intermittent mandatory manual ventilation. But the procedure was aborted 6 h after the induction of anesthesia, because the location of the patient's facial nerve prevented the safe creation of a cochleostomy. No airway problems that resulted in reduced S_{PO_2} were observed during or after the anesthesia. The patient's subsequent clinical course and recovery were uneventful. The patient was discharged on postoperative day 7.

CHARGE syndrome was originally reported by Hall [1] in 17 children with multiple congenital anomalies with choanal atresia, and was later independently reported by Hittner et al. [2] in 10 patients with coloboma. Pagon et al. [5] defined the term "CHARGE", an acronym summarizing six cardinal clinical features of the syndrome: ocular coloboma (C), heart defects of any type (H), atresia of the choanae (A), retardation (R), genital anomalies (G), and ear anomalies (E). CHARGE syndrome is an autosomal dominant disorder with a prevalence of 1 in 10 000 people. The gene *CDH7* in chromosome 8q12 was identified as a causative gene for CHARGE syndrome in approximately 60% of patients with a clinical diagnosis of CHARGE syndrome [6]. Most cases are sporadic; however, in rare cases, transmission from a mildly affected parent has been reported. In the present patient, no genetic diagnosis was made. Neither of the parents had clinical features suggestive of CHARGE syndrome. In addition to the canonical features, the following airway abnormalities have been reported in patients with this syndrome: retrognathia, glossoptosis, esophagotracheal fistula with or without esophageal atresia, stenosis or laryngeal paralysis, and cleft lip and palate [7]. A tracheostomy is indicated in 10% to 30% of the patients [8]. In addition, apart from choanal atresia and cleft lip and palate, 56% of patients have some form of other upper airway abnormalities [9]. Thus, the induction of anesthesia in patients with CHARGE syndrome is still challenging. The glottis and vocal cords of the present patient were not sufficiently demonstrated using a BFS, resulting in the failure of BFS-assisted ETT intubation. Following the planned strategy, an LMA was used to secure the airway. The LMA was placed in a good position; thus, the LMA could be replaced with an ETT using tube-exchange devices. According to the document provided by the manufacturer, a size 2.5 ProSeal LMA is made for children weighing 20 to 30 kg, a size 2.0 LMA is for children weighing 10 to 20 kg, and a size 1.5 is for children weighing 5 to 10 kg. At first, we tried in vain to insert a size 2.5 ProSeal LMA, which was larger than the one recommended for the present patient, with our intention being to later exchange the LMA for an ETT. Then, after the unsuccessful use of a size 2.0 ProSeal LMA, a size 1.5 ProSeal LMA was finally inserted smoothly. The patient had retrognathia and glossoptosis, both of which are characteristic features of the CHARGE syndrome. Therefore, the pharynx and larynx of CHARGE syndrome patients may be much smaller than normal for the patient's physique and so a smaller sized LMA should be used at first. Because the present patient was scheduled to undergo elective surgery, we had enough time to determine a strategy for airway management. Moreover, the patient's guardians also granted permission for a tracheostomy. However, some patients with CHARGE syndrome may require urgent surgical treatment for anomalies, especially when they have cyanotic heart and great vessel anomalies. As reported here, an LMA may be the device of first choice to secure the airway in patients with CHARGE syndrome. Naito et al. [7] reported the results of fiberoptic laryngoscopy performed in CHARGE syndrome patients. In addition to their anatomical anomalies and airway obstructions, patients with CHARGE syndrome had a significant risk of having an airway dysfunction [7]. Such patients frequently suffer from dysfunction of the upper airway, including hypotonic pharyngolarynx, facial nerve palsy, and saliva retention. In addition to the use of atropine, we used a ProSeal LMA to prevent saliva efflux to the airway. A ProSeal-type LMA has many advantages over classical LMAs in the anesthetic management of CHARGE syndrome patients. The above evidence indicates that, for achieving safe anesthetic management in patients with CHARGE syndrome, evaluation based not only on anatomical but also on functional aspects is very important.

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References

1. Hall BD. Choanal atresia and associated multiple anomalies. J Pediatr. 1979;95:395–8.
2. Hittner HM, Hirsch NJ, Kreh GM, Rudolph AJ. Colobomatous microphthalmia, heart disease, hearing loss, and mental retardation–a syndrome. J Pediatr Ophthalmol Strabismus. 1979;16: 122–8.
3. Keller C, Puhringer F, Brimacombe JR. Influence of cuff volume on oropharyngeal leak pressure and fibreoptic position with the laryngeal mask airway. Br J Anaesth. 1998;81:186–7.
4. Mizutamari E, Yano T, Ushijima K, Ito A, Anraku S, Tanimoto H, Terasaki H. A comparison of postoperative sore throat after use of laryngeal mask airway and tracheal tube. J Anesth. 2004; 18:151–7.
5. Pagon RA, Graham JM Jr, Zonana J, Yong SL. Coloboma, congenital heart disease, and choanal atresia with multiple anomalies: CHARGE association. J Pediatr. 1981;99:223–7.
6. Sanlaville D, Verloes A. CHARGE syndrome: an update. Eur J Hum Genet. 2007;15:389–99.
7. Naito Y, Higuchi M, Koinuma G, Aramaki M, Takahashi T, Kosaki K. Upper airway obstruction in neonates and infants with CHARGE syndrome. Am J Med Genet A. 2007;143:1815–20.
8. Roger G, Morisseau-Durand MP, Van Den Abbeele T, Nicollas R, Triglia JM, Narcy P, Abadie V, Manac'h Y, Garabedian EN. The CHARGE association: the role of tracheotomy. Arch Otolaryngol Head Neck Surg. 1999;125:33–8.
9. Stack CG, Wyse RK. Incidence and management of airway problems in the CHARGE Association. Anaesthesia. 1991;46:582–5.