Correspondence

Mandibular hypoplasia and narrow airway in goldenhar syndrome: Anticipation of difficult intubation with cone-beam computed tomography

Dear Editor,

Goldenhar syndrome (GS), also known as oculo-auriculo-vertebral dysplasia, is a rare congenital pathology of unknown origin with anomalous development of the first and second brachial arches and vertebrae. The disease comprises a wide range of clinical manifestations, but the association of unilateral craniofacial microsomia (characterized by incomplete development of the mandible, lip, soft palate, nose and ear), ocular dermoid cysts, and spinal defects tends to be constant [1]. Other organic anomalies may coexist, such as cardiac and vascular abnormalities.

Several surgical interventions may be necessary to restore these children’s craniofacial esthetics and function. For the diagnostic workup and virtual surgical planning, cone-beam computed tomography (CBCT) provides accurate 3-dimensional (3D) information of the facial bones and related anatomical structures, including the upper airway. In fact, characterization and quantitative assessment of the upper airway is accurate and reliable. Compared to conventional computed tomography, scanning time and radiation are substantially reduced, whilst maintaining high resolution and image sharpness for hard tissues. The resulting DICOM (digital imaging and communication in Medicine) images can be processed with third-party softwares to allow for 3D virtual treatment planning. Consequently, CBCT has become the gold standard image acquisition tool for children with GS.

On the other hand, anesthetic management of these patients is always a challenge. Facial asymmetry and airway anomalies may pose difficulties to face mask ventilation and tracheal intubation, respectively. Moreover, airway management difficulties tend to increase with age [2]. Together with the limited clinical experience due to the low incidence of this condition, the lack of evidence-based systematized protocols for airway management calls for a meticulous anesthetic evaluation and individualized planning [3]. In this context, the authors would like to share with your readers the value of CBCT—performed for surgical planning purposes—in the evaluation of the patient’s upper airway and subsequent anticipation of potential airway management complications. [4,5].

A case of a 6-year old child with GS is presented. His medical history was positive for right mandibular hypoplasia (Fig. 1a), moderate aortic stenosis, and unilateral right facial cleft number 7 according to Tessier’s classification (already corrected surgically at 3 years of age). He was scheduled for right mandibular hypoplasia correction with a costochondral graft.

A meticulous anesthetic evaluation was carried out by the prospective anesthesiological team. First, the preoperative CBCT study was used to analyze the patient’s upper airway. A 3D reconstruction of the airway was obtained with the appropriate software. It revealed a narrow airway, mainly on the right side (Fig. 1b). Second, clinical examination showed a restricted oral aperture and cross-bite malocclusion due to hemifacial microsomia. The combination of these radiological and clinical data suggested the risk of a difficult airway. Accordingly, the availability of the difficult-airway trolley was ensured in the operating room throughout the whole surgical procedure.

The patient was placed on a heating blanket and standard monitoring was applied, including noninvasive blood pressure control, pulse oximetry and electrocardiography. Inhalatory induction with sevoflurane was performed, followed by 2 mg kg−1 of fentanyl, 1 mg kg−1 of propofol and 0.6 mg kg−1 of rocuronium. The patient could be successfully intubated orally with a 4.5-mm internal diameter (ID) cuffed tube using GlideScope videolaryngoscope (Verathron, Bothell, WA, USA) and external laryngeal maneuvers. A 3-level intercostal nerve-block was performed for costochondral grafting. Dexamethasone 0.1 mg kg−1 was administered as postoperative nausea and vomiting prophylaxis, and amoxicillin-clavulamic 750 to 125 mg was administered as a single-shot antibiotic. Anesthesia was maintained with 2% sevoflurane. The surgery proceeded uneventfully and the patient was extubated awake without complications. He was transferred to the intensive care unit, and no further complications arouse.

Sometimes, classic formulas to determine the appropriate endotracheal tube size based on height, weight or age are poor. According to Matayama’s formula to chose a cuffed endotracheal tube [ID in mm = (0.25 × age in years) + 3.5], our patient would have required a 5-mm ID and 6.9-mm outer diameter (OD). However, we preferred to base our endotracheal...
tube size decision on CBCT information. In healthy patients, the narrowest diameter of the subglottic upper airway is the width of the air column at the level of the cricoid cartilage. This diameter governs the selection of the endotracheal tube size [6]. In our patient, the subglottic diameter was 6.5 mm measured at the most inferior aspect of the cricoid. Consequently, a cuffed endotracheal tube size 4.5-mm ID and 6.2-mm OD was chosen. Alternative imaging techniques for endotracheal tube size selection may include ultrasonography [7]. Correct tube size selection is essential in order to avoid complications. If too small, insufficient ventilation, poor reliability of end-tidal gas monitoring, leakage of anesthetic gases into the operating room environment, and an enhanced risk of aspiration may occur. If too large, upper airway damage (eg, local ischemia, ulceration, scar formation) and the potential for subsequent subglottic stenosis are potential risks.

In conclusion, the authors would like to highlight the relevance of CBCT imaging for the preoperative assessment of patients in whom an endotracheal intubation is foreseen. 3D reconstructions of the upper airway provide valuable morphometric information and, when available, should be part of the routine anesthesiological evaluation.

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