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Airway evaluation by CT imaging for cri-du-chat syndrome

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To the editor: We describe the effectiveness of preoperative airway evaluation by computed tomography (CT) (Fig. 1) imaging for a patient with cri-du-chat syndrome. Cri-du-chat syndrome results from a chromatin deletion on the short arm of chromosome 5(5p). In this syndrome, laryngeal abnormalities result in a characteristic cat-like cry during infancy. In general, patients with cri-du-chat syndrome exhibit a range of laryngeal abnormalities, including hypoplastic larynx, narrow laryngomalacia, and diamond-shaped larynx [1], as well as epiglottal abnormalities, including long, curved, floppy, hypoplastic epiglottis and hypotonic epiglottis [1], all of which present risks of difficulty in airway management during anesthesia.

Our initial experience involved the preoperative evaluation of a 15-year-old female with cri-du-chat syndrome scheduled to undergo a dental procedure. In this patient, preoperative chest X-ray revealed what appeared to be minor subglottic stenosis (Fig. 2). Based on this X-ray examination, we initially concluded that it would be possible to insert a 5.5-mm outer-diameter endotracheal tube. We found, however, that when we attempted to insert a 4.0-mm inner-diameter (ID) endotracheal tube, it could be passed between the vocal cords, but not through the subglottic region. The scheduled dental procedure under general anesthesia was postponed. Postoperative evaluation of the stenosis by CT imaging revealed that the diameter of the most stenotic part of the trachea was in fact 3 mm.

Based on this experience, we performed preoperative airway evaluation by CT imaging for a 33-year-old woman with cri-du-chat syndrome. This patient had been admitted for

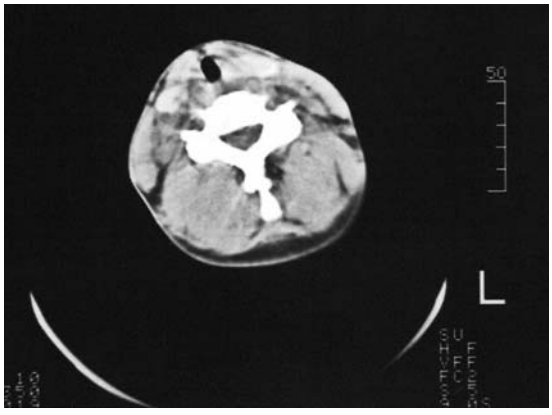

Fig. 1. Computed tomography (CT) image of the tracheal lumen below the glottis

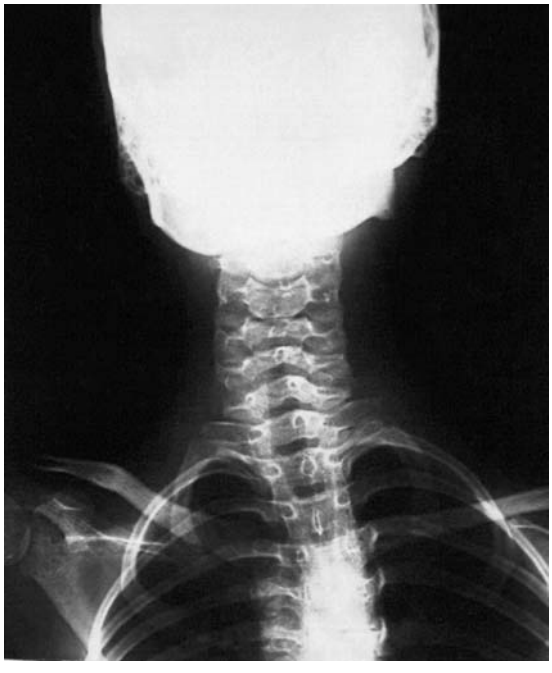

Fig. 2. Chest X-ray of a 15-year-old female with cri-du-chat syndrome, showing stenosis beneath the vocal cord

dental treatment to be conducted under general anesthesia, this being indicated because of the patient's mental retardation. Preoperative chest X-ray showed scoliosis but no stenosis of the tracheal lumen. We also performed preoperative airway evaluation using CT imaging (in slice sections at 5-mm intervals) under intravenous anesthesia with midazolam. The CT images showed no stenosis or abnormality from the epiglottis to the carina trachea. On the day of the dental procedure, a fiberoptic bronchoscope and a nasal flexible laryngeal mask airway [2,3] were prepared in the event of a difficult intubation. Anesthesia was induced with propofol $2\,mg \cdot kg^{-1}$. After ensuring controlled ventilation with a manual bag, a 6.5-mm ID nasal endotracheal tube was inserted into the trachea through a nostril after administration of vecuronium $0.1\,mg \cdot kg^{-1}$ intravenously. This procedure was performed without incident or difficulty. Anesthesia was maintained with oxygen, nitrous oxide, and sevoflurane. The dental procedures were also performed without complications and the patient's postoperative recovery was uneventful.

In patients with cri-du-chat syndrome, a diverse range of laryngeal and epiglottal abnormalities may be encountered [4,5], and, as described above, images from chest X ray are not always in complete agreement with those from CT imaging. We therefore strongly recommend that the preoperative evaluation of the airway by CT imaging is of great utility for patients with cri-du-chat syndrome and, in general, for any patients presenting the possibility of laryngeal, glottal, or tracheal abnormality.

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