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Apert syndrome, pediatric anesthesia, difficult airway, 3D-printed video laryngoscope, cleft palate repair, craniofacial anomalies

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## Through the Narrow Passage: Innovative Airway Securing in a Child with Apert Syndrome for Cleft Palate Repair

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### Abstract

Apert syndrome is a rare congenital disorder characterized by craniosynostosis, midface hypoplasia, and syndactyly, frequently associated with challenging airway management. Craniofacial malformations increase the risk of difficult mask ventilation, intubation, and postoperative airway obstruction. We report the anesthetic management of a 5-year-old child with Apert syndrome undergoing cleft palate repair. Anticipating a difficult airway due to craniofacial abnormalities, a comprehensive airway strategy was prepared, including pediatric-sized video laryngoscope blades, a fiberoptic bronchoscope, supraglottic devices, and surgical airway backup. Inhalational induction with spontaneous ventilation was followed by successful intubation using an endogenously developed 3D-printed pediatric video laryngoscope with integrated bougie port and a pre-shaped stylet. A 4.0 mm North Pole RAE tube was secured. Surgery proceeded uneventfully, and the patient was extubated fully awake with no complications. Midface hypoplasia, high-arched palate, and limited neck extension in Apert syndrome significantly increase airway difficulty. While fiberoptic bronchoscopy remains the gold standard, video laryngoscopy offers enhanced visualization and higher first-attempt success rates in pediatric difficult airways. In this case, the endogenously developed 3D-printed pediatric video laryngoscope with integrated bougie port provided a low-cost, customizable, and sterilizable solution suitable for resource-limited settings. Maintaining spontaneous ventilation until airway control, awake extubation, and a robust backup plan were critical to preventing complications. In pediatric syndromic patients, innovative, low-cost airway tools-such as endogenously developed 3D-printed pediatric video laryngoscopes with integrated bougie ports-can improve safety and efficiency. Careful preoperative planning, maintenance of spontaneous ventilation until airway control, and a robust backup strategy are key for optimal outcomes.

## INTRODUCTION

Apert syndrome, first described by Eugène Apert in 1906, is a rare genetic craniosynostosis syndrome with an estimated incidence of 1 in 65,000 live births<sup>[1]</sup>. It results from mutations in the fibroblast growth factor receptor 2 (FGFR2) gene, leading to premature fusion of cranial sutures and subsequent craniofacial dysmorphism<sup>[2]</sup>. Hallmark features include midface hypoplasia, high-arched palate, beaked nose, hypertelorism, and symmetric syndactyly of the hands and feet<sup>[3,4]</sup>.

Airway management in Apert syndrome is particularly challenging. Midface hypoplasia leads to reduced nasopharyngeal space, while limited mouth opening and abnormal cranial shape make direct laryngoscopy difficult<sup>[5,6]</sup>. Additionally, these patients may have associated comorbidities such as obstructive sleep apnea (OSA), congenital heart disease, raised intracranial pressure, and cervical spine anomalies<sup>[7,8]</sup>.

In children, difficult airway situations are more precarious due to smaller airway dimensions, reduced functional residual capacity, and higher oxygen consumption, which predispose to rapid desaturation during attempts at intubation<sup>[9]</sup>. Conventional direct laryngoscopy often fails, making fiberoptic bronchoscopy the gold standard for anticipated difficult airways<sup>[10]</sup>. However, video laryngoscopy has emerged as a highly effective alternative, offering improved visualization with less requirement for alignment of airway axes<sup>[11-13]</sup>.

Recent innovations such as 3D-printed airway devices are gaining interest in anesthetic practice. These devices are customizable, low-cost, and can be designed for pediatric anatomical dimensions<sup>[14,15]</sup>. This case report details the anesthetic management of a 5-year-old child with Apert syndrome undergoing cleft palate repair, where a sterilizable 3D-printed pediatric video laryngoscope enabled successful airway control.

## Case Presentation

**Patient Profile:** A 5-year-old male child, weighing 20 kg, with a known diagnosis of Apert syndrome, presented for elective cleft palate repair. Physical features included a tall, narrow skull, hypertelorism, beaked nose, midface hypoplasia, and syndactyly of both hands and feet. Airway examination revealed a reduced mouth opening (interincisor distance 2 cm), high-arched palate, limited neck extension, and micrognathia. Systemic examination was otherwise unremarkable.

**Preoperative Management:** Baseline laboratory investigations, including complete blood count, serum electrolytes, and renal function tests, were within normal limits. Echocardiography showed normal cardiac anatomy and function. There were no signs of

raised intracranial pressure. Given the craniofacial abnormalities, a difficult airway was anticipated. Preparation included availability of a pediatric fiberoptic bronchoscope, an endogenously developed 3D-printed pediatric video laryngoscope with blades, pre-shaped stylet, pediatric bougie, supraglottic airway devices (LMA sizes 1.5–2.5), and surgical airway instruments.

**Anesthesia Management:** The child was premedicated with intravenous midazolam 0.05 mg/kg and fentanyl 1 mcg/kg. Standard ASA monitors were applied, and preoxygenation was performed for 3 minutes. Intravenous induction was achieved with propofol 2 mg/kg, maintaining spontaneous ventilation. Using the custom 3D-printed pediatric video laryngoscope, a clear Cormack–Lehane grade II view was obtained, and the trachea was intubated with a 4.0 mm internal diameter North Pole RAE tube over a pre-shaped stylet. Correct placement was confirmed with continuous waveform capnography and bilateral chest auscultation. After securing the airway, atracurium 0.5 mg/kg was administered for neuromuscular relaxation. Anesthesia was maintained with sevoflurane in an oxygen–air mixture and intermittent atracurium boluses. The intraoperative course was stable, with no desaturation or hemodynamic instability.



Fig. 1: Post-extubation intraoperative view during cleft palate repair in a child with Apert syndrome, showing the repaired palate with midline sutures in place. The mouth is retracted using a Dingman mouth gag to provide optimal surgical exposure.

**Postoperative Care:** At the end of the procedure, neuromuscular blockade was reversed with neostigmine 0.05 mg/kg and glycopyrrolate 0.01 mg/kg. The patient was extubated fully awake after confirming adequate spontaneous ventilation and intact airway reflexes. He was transferred to the

pediatric ICU for overnight monitoring in room air. No airway obstruction or respiratory distress occurred in the immediate postoperative period.

**Follow-up and Outcome:** Recovery was uneventful. Oral feeding was initiated on the second postoperative day, and no airway-related complications reported.

## RESULTS AND DISCUSSIONS

Airway management in Apert syndrome requires careful planning due to multiple anatomical and physiological challenges. Midface hypoplasia narrows nasal passages, high-arched palate reduces oral space, and cervical spine anomalies can limit neck mobility<sup>[5,7,8]</sup>. Such features increase the likelihood of both difficult mask ventilation and intubation<sup>[9,10]</sup>.

Traditional fiberoptic bronchoscopy is often advocated for anticipated pediatric difficult airways<sup>[10,11]</sup>, but it requires expertise, availability, and patient cooperation. Video laryngoscopy offers an easier learning curve, enhanced visualization, and higher first-attempt success rates in difficult pediatric airways<sup>[12,13]</sup>.

In this case, the 3D-printed pediatric video laryngoscope offered unique advantages:

- Customization for pediatric dimensions
- Sterilizability for repeated use
- Low cost, especially in resource-limited settings
- Rapid prototyping for urgent cases<sup>[14,15]</sup>

Maintaining spontaneous ventilation during airway instrumentation is a widely recommended safety strategy in anticipated difficult airways<sup>[16]</sup>. This approach reduces the risk of “cannot ventilate, cannot intubate” situations. Additionally, a robust backup plan-including supraglottic airway devices and surgical airway readiness-was essential<sup>[17,18]</sup>.

Postoperatively, patients with craniofacial syndromes are at increased risk of airway obstruction due to edema or OSA<sup>[19]</sup>. Awake extubation, as performed here, minimizes reintubation risk<sup>[20]</sup>.

## CONCLUSION

When the airway is narrow, innovation must be wide. This case demonstrates that in pediatric patients with Apert syndrome, successful airway management hinges on meticulous preparation, maintenance of spontaneous ventilation until airway security, and the strategic use of innovative devices such as 3D-printed video laryngoscopes. Low-cost, customizable solutions can bridge technological gaps in resource-limited settings, turning high-risk scenarios into controlled successes.

**Credits:** We acknowledge Dr. Yachendra for the design and development of the endogenously developed 3D-printed pediatric video laryngoscope with integrated bougie port used in this case.

**Acknowledgment:** We acknowledge the contributions of all healthcare professionals involved in the diagnosis and management of the patient described in this case report.

**Conflict of Interest:** The authors declare no conflicts of interest related to this study.

**Consent Declaration:** Written informed consent was obtained from the patient's parent for the publication of this case report and any accompanying images.

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