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Case report: Anesthetic Management for an Adult Patient with Spondylothoracic Dysostosis and Tracheal Stenosis with Montgomery T-tube Insertion

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ABSTRACT

Background: Spondylothoracic dysostosis (STD) is a rare congenital disorder characterized by short-trunk dwarfism, vertebral anomalies, and severe thoracic insufficiency syndrome. Its management is challenging due to complex anatomical and respiratory features, often requiring multidisciplinary surgical interventions and advanced airway management techniques.

Case: We report a 19-year-old male with STD who underwent multiple thoracic and spine surgeries during childhood. He had a history of difficult intubation and subsequently required chronic tracheostomy. He developed tracheal stenosis following long-term tracheostomy use, which required laser tracheoplasty and insertion of a Montgomery T-tube (MTT) for tracheal stenting. The distorted airway posed significant airway challenges; a bronchoscopy-guided tracheostomy tube exchange was eventually performed, followed by an uneventful tracheoplasty and MTT insertion for stenting.

Conclusions: STD poses significant anesthetic and surgical challenges due to airway abnormalities and respiratory insufficiency. Multidisciplinary planning and individualized airway strategies are essential for safe perioperative care in such complex cases.

Keywords: Spondylothoracic Dysostosis; Crab-like thorax; Lavy-Moseley Syndrome; Jarcho-Levin Syndrome; Tracheal stenosis; Montgomery T-tube; Case Report

INTRODUCTION

Spondylothoracic dysostosis (STD) is an inherited disease characterized by short-trunk dwarfism with ribs and vertebral anomalies[1] that lead to thoracic insufficiency and restrictive lung disease. These patients often require

multiple surgeries during childhood, including thoracospinal corrective procedures and airway surgeries.[2] The combination of anatomical abnormalities, restrictive pulmonary physiology, and complex airway dynamics poses significant anesthetic and surgical challenges.

This report details a 19-year-old male with STD who developed tracheal stenosis after prolonged tracheostomy use. Successful management, including laser tracheoplasty and Montgomery T-tube (MTT) insertion, highlights the importance of multidisciplinary collaboration and tailored care in addressing the unique challenges of STD.

Written informed consent to publication had been obtained from the patient.

CASE PRESENTATION

Our patient was born with a short, immobile neck and a short trunk with prominent thoracolumbar lordosis. Imaging revealed multiple hemi-vertebrae and vertebral fusion; the thoracic cavity was diminished with symmetrical crowding of the ribs and costo-vertebral joint fusion, resulting in a crab-like appearance. The radiological features were compatible with Spondylothoracic Dysostosis.

He was intubated shortly after birth due to respiratory distress, and a tracheostomy was performed at 6.5 months of age. Staged expansion thoracoplasties with Vertical Expandable Prosthetic Titanium Rib (VEPTR) implantation were performed at 38 and 59 months of age, followed by multiple adjustments under general anesthesia. The implants were removed at 12 years old.

Despite the successful operations, anesthesia was challenging due to history of false-tracking caused by suboptimal manual tracheostomy tube exchanges. Subsequent attempts at intubation with fiberoptics, video laryngoscopes, and bag-mask ventilation were also unsuccessful. An uncuffed tracheostomy tube of size 4.5 was eventually inserted which became his long-term airway.

Chronic use of the tracheostomy tubes had unfortunately caused tracheal stenosis when he was 19. Flexible tracheoscopy showed a congested, elliptical, stenotic segment above the tracheostomy opening, measuring four millimeters at its narrowest point (Figure 1). CT imaging identified further stenotic segments along the trachea, with the tip of tracheostomy tube indenting the trachea near the aortic arch, raising concerns about potential erosion and an aorto-tracheal fistula.

Given the complex anatomy, a laser tracheoplasty and revision tracheostomy was arranged, followed by insertion of a MTT for stenting and as a back-up route of ventilation after the airway operation.

Pre-operative assessments and investigations

Our patient exhibited no signs of tachypnea or cyanosis. He weighed 31.9 kilograms and was 1.32 meters tall. Airway examination showed Mallampati score 2, with an inter-incisor distance of 2 centimeters and a short, immobile neck. The cardiovascular and respiratory examination was unremarkable.

Blood counts, liver and renal functions were unremarkable. Electrocardiogram showed a sinus rhythm of 105 beats per minute with right axis deviation. Cervical spine radiographs revealed vertebral fusion with minimal flexion-extension movement. Spirometry showed a mixed obstructive and restrictive pattern, with forced vital

capacity (FVC) at 0.69 liters (L) (18% of predicted), forced expiratory volume (FEV1) at 0.39L (11.5%) and FEV1/FVC ratio at 57%. Focused echocardiogram demonstrated a normal ventricular function and chamber size with no feature of pulmonary hypertension.

Given the underlying severe restrictive lung disease, postoperative ICU admission was arranged to monitor for potential respiratory failure as the patient transitioned to using his natural airway for the first time, which presented significantly higher resistance compared to the lifelong use of a tracheostomy tube.

Intraoperative anesthesia

Salvage veno-venous extracorporeal membrane oxygenation (VV-ECMO) was considered but not pre-emptively initiated due to favourable venous anatomy, as assessed by the intensivists.

Induction began with Otorhinolaryngologists scrubbed-in standing-by to assist exchange of tracheostomy tube. Patient was induced with intravenous propofol and remifentanil using target-controlled infusion. Examination of tracheostomy stoma confirmed a well-formed tract, the tracheostomy tube was then removed with replacement of a wire-reinforced, cuffed endotracheal tube of size 4.5 under fibreoptic bronchoscopy guidance (2.8-millimeter Ambu®) to confirm track and depth of insertion. The insertion of the wire-reinforced tube facilitated ventilation and provided airway protection during the excision of the tracheal stenosis proximal to the tracheostomy opening. Pressure-controlled ventilation was applied with inspiratory pressure of 15 cmH2O.

Following tracheoplasty and dilation of the stenotic segments, the wire-reinforced tube was removed to facilitate insertion of the MTT, while a size 5.5 microcuff endotracheal tube was inserted orally, using a CMAC D-blade (achieving Cormack-Lehane grade 2a visualization) for ventilation. The MTT, inserted via the tracheostomy stoma, is a T-shaped tube which provided a conduit for ventilation via the upper airway, while maintaining patency of the tracheostomy lumen in case of upper airway obstruction after oro-tracheal extubation. The patient was eventually reversed from anesthesia and extubated for spontaneous ventilation via the upper airway.

Postoperative course

The patient was monitored in the intensive care unit. Flexible endoscopy on postoperative day (POD) 11 confirmed a patent airway above and below the MTT. The MTT was removed on POD 13. Polysomnography conducted on POD 14 revealed mild obstruction with an apnea-hypopnea index (AHI) of 9.0. He tolerated nocturnal nasal continuous positive airway pressure (CPAP) and was discharged on postoperative day 24 with the device, while the tracheostomy stoma remained covered with dressings during the day.

DISCUSSION

First described in 1938,[3] Spondylothoracic dysostosis is a syndrome involving bilateral rib fusion at the costovertebral joints, resulting in a characteristic "crab-like" thorax. This thoracic rigidity causes significant restrictive lung disease and respiratory insufficiency.[4] Other features include vertebral segmentation or formation defects, cervical vertebra fusion and short stature; limbs are of normal length and intelligence is unaffected.[2] The prognosis is poor with only 25% of patients surviving to adulthood.[5] The inheritance is attributed to the Mesoderm Posterior 2 Homolog (MESP2) gene mutation disrupting somitogenesis.[6]

Anesthetic strategy for patients with STD

Anesthesia for patients with STD is challenging. Thoracic lordosis and fixed cervical spine render optimal position for intubation impossible, while reduced mouth opening and thyromental distance further worsens laryngoscopy views. Thorough planning with fibreoptic bronchoscopy, and with surgeons scrubbed-in standing-by for emergency front-of-neck airway, is advised. Emergency veno-venous extra-corporeal membrane oxygenation (ECMO) support should also be liaised in preparation of dire situations in which front-of-neck airway is deemed unattainable.

For patients with severe restrictive lung disease secondary to thoracic insufficiency, a lung-protective ventilation strategy, with a tidal volume of around 6-8 ml/kg, should be employed; Positive end-expiratory pressure (PEEP) should be carefully titrated to avoid overdistension while reducing atelectasis. Pressure-controlled ventilation is advised for tighter control of airway pressure. Excessively high respiratory rates should be avoided as they can lead to air trapping and dynamic hyperinflation.[7]

Should pulmonary hypertension be identified in preoperative workup, anesthetic management should aim at reducing pulmonary vascular resistance (PVR). This includes ensuring oxygenation, a low tidal volume (5-7ml/kg) and PEEP while avoiding medications that increase PVR.

STD patients should be monitored in intensive care for potential complications.

Tracheal stenosis and its anesthetic challenges

Tracheal stenosis is typically associated with prior history of endotracheal intubation; it may manifest as stridor or dyspnea, however symptoms may be inapparent unless the stenotic lumen is less than 5mm.[8] Spirometry shows a flattened inspiratory and expiratory curve. Computed tomography is helpful in delineating the exact location and size of the stenotic lesion, while bronchoscopy allows direct visualization of the stenosis.

Prior discussion between anesthetists and surgeons for airway management is mandatory. Laryngeal masks could facilitate flexible bronchoscopy performance, while insertion of endotracheal tubes, potentially through and bypassing the stenotic lesions, provides definite airway protection. Anesthesia is maintained using Total Intravenous Anesthesia (TIVA) to avoid awareness and volatile agent contamination during open airway surgery. The decision for spontaneous ventilation versus positive pressure ventilation (PPV) should be made on individual basis. While a lower airway and thoracic pressure is achieved with spontaneous ventilation, PPV allows a consistent and stable field for surgery with full control of ventilation parameters.

The decision of extubation is a shared decision with the surgeon post-operatively. It is a clinical dilemma while airway patency is ensured, the presence of endotracheal tube in a recently operated airway may impair wound healing.

Montgomery T-tube (MTT) and subsequent ventilation

The Montgomery T-tube is an effective solution to this dilemma. First described by Dr William Montgomery in the mid-1960s,[8] it is a device that provides stenting for the trachea while allowing ventilation through the

larynx and the tracheostomy stoma. It is an uncuffed silicon tube with a long limb placed within the trachea working as a stent and a conduit, whereas the short limb protruding through the tracheostomy stoma (Figure 4). To provide mechanical ventilation with the MTT in-situ, the extra-tracheal opening of the MTT is spigotted to prevent leakage of fresh gas. For ventilation via extra-tracheal lumen, it is connected to the breathing system via an adaptor, and the cranial portion of the intra-tracheal MTT should be occluded. Alternatively, a Y-piece connection, with one end to facemask and the other end to the short limb of the T-tube, had been described.[9,10]

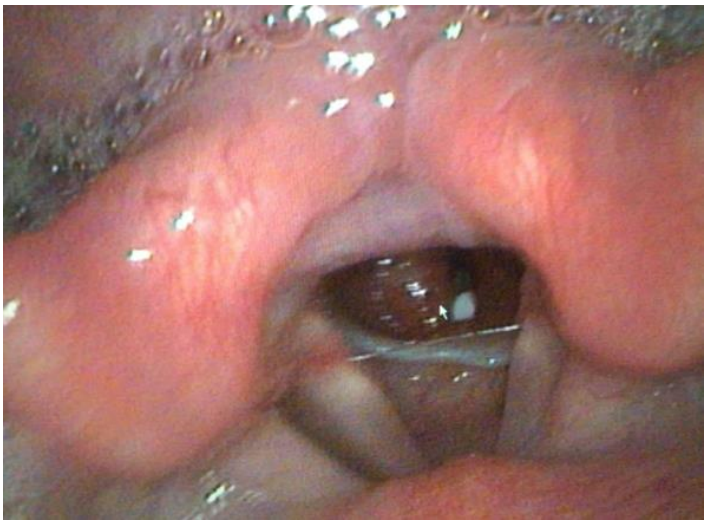

Figure 1: Tracheoscopy view of stenosis proximal to tracheostomy tube (stenosis marked with a white cursor).

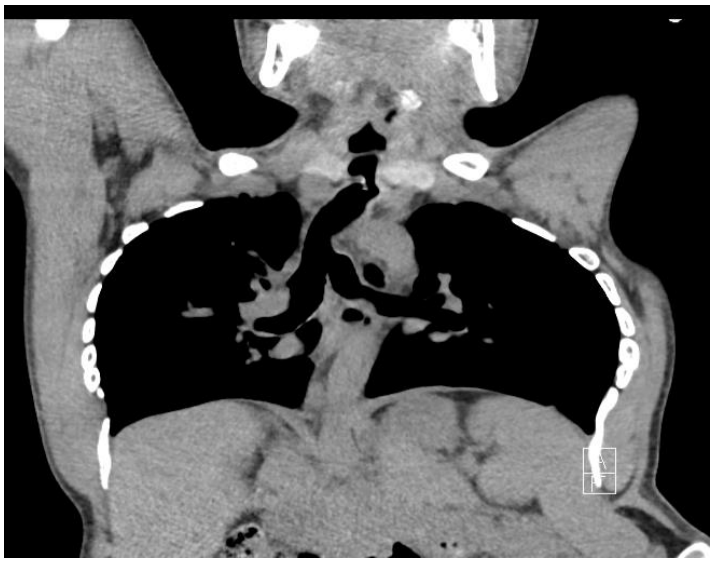

Figure 2: Coronal cut of the computed tomography scan of thorax showing multiple stenoses.

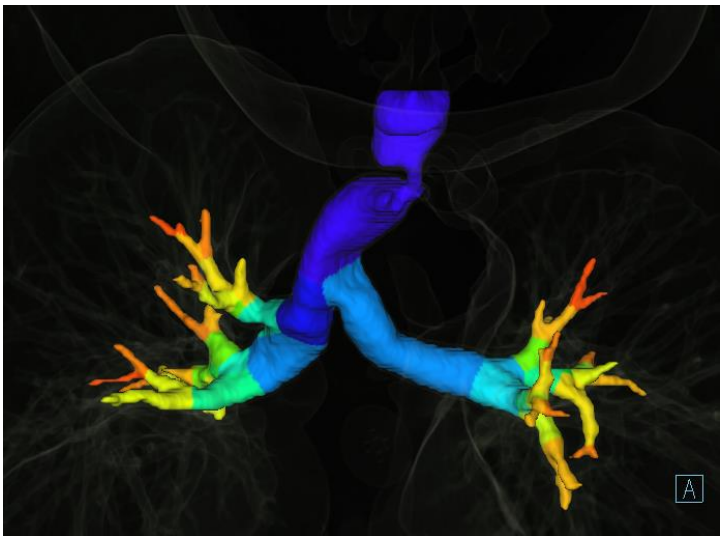

Figure 3: 3D reconstruction of the airway tree using images from the computed tomography scan.

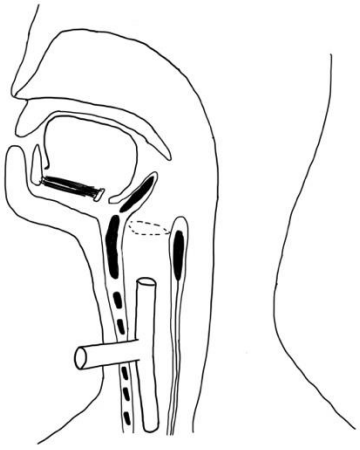

Figure 4: Illustration of the correct positioning of a Montgomery T-tube in trachea.

CONCLUSION

Spondylothoracic dysostosis poses significant challenges in surgical and anesthetic management due to severe airway abnormalities, restrictive lung disease and complex anatomical deformities. Comprehensive preoperative planning, including detailed airway assessment, readiness for extracorporeal oxygenation, multidisciplinary care and coordinated intraoperative management is vital to mitigate the risks associated with difficult airway management and compromised respiratory function.

Application of the Montgomery T-tube as a tracheal stent is effective for maintaining airway patency following tracheoplasty and serves as a backup ventilation conduit in the event of postoperative airway obstruction. This

case highlights the necessity of individualized airway-strategies to optimize outcomes and improve the quality of life in patients with STD.

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