INTRAOPERATIVE AIRWAY OBSTRUCTION RELATED TO TRACHEOSTOMY TUBE MALPOSITION IN A PATIENT WITH ACHONDROPLASIA AND JEUNE'S SYNDROME

RAMON E ABOLA[*], JONATHAN TAN[*], DAVID WALLACH[**],
CATHERINE KIER[***], PEGGY A. SEIDMAN[*,***]
AND JOSEPH D TOBIAS[****]

Abstract

A 14 year-old adolescent with achondroplasia and Jeune's syndrome (asphyxiating thoracic dystrophy) presented for cervical spine surgery in the prone position. Due to the need for home mechanical ventilation at night, the patient had a tracheostomy in place. With the first surgical procedure, the cuffed tracheostomy tube was left in place during prone positioning. Difficulties encountered with ventilation through the cuffed tracheostomy tube in the prone position necessitated aborting the case. During three subsequent surgeries, the tracheostomy tube was removed and an armored endotracheal tube was placed through the tracheostomy stoma prior to prone positioning. No further difficulties with ventilation were noted with the subsequent cases. There are currently no guidelines in the medical literature regarding perioperative management of patients with a tracheostomy requiring prone positioning for surgery. The management of such patients is reviewed and possible problems with tracheostomy positioning during prone positioning are explored. Given such issues, we would suggest removal of the tracheostomy tube and placement of an armored endotracheal tube through the stoma during surgical procedures in the prone position.

Introduction

Jeune's Syndrome is an autosomal-recessive disorder resulting in a constricted narrow rib cage with generalized chondrodystrophy. Given the restrictive respiratory defect, these patients frequently require tracheosomy and chronic mechanical ventilation. We present a 14 year-old adolescent with achondroplasia and Jeune's Syndrome with a tracheostomy in place who required prone positioning during cervical spine surgery. Intraoperatively, problems providing adequate ventilation resulted in the need to stop the surgical procedure and turn the patient supine. During subsequent surgical procedures, the tracheostomy tube was removed and an armored endotracheal tube (ETT) was placed through the trachestomy stoma. Despite the fact that patients with

---

*   Department of Anesthesiology, Stony Brook University Medical Center, New York, NY.
**   Department of Orthopaedics, Stony Brook University Medical Center, New York, NY.
***   Department of Pediatrics, Stony Brook University Medical Center, New York, NY.
****   Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, The Ohio State University, Columbus, Ohio.
     Corresponding Author: Joseph D. Tobias, MD, Chief, Department of Anesthesiology and Pain Medicine, Nationwide Children's Hospital, Professor of Anesthesiology and Pediatrics, The Ohio State University, 700 Children's Drive, Columbus, Ohio 43205. Tel: (614) 722-4200, E-mail: joseph.tobias@nationwidechildrens.org

tracheostomies frequently present to the operating room for surgical procedures, there are no definitive guidelines in the medical literature regarding the optimum means of airway management in such patients when the surgical procedure requires prone positioning. We describe the anesthetic management of a patient with a tracheostomy requiring prone positioning, explore the mechanisms whereby problems may occur with prone positioning of patients with tracheostomies, and present our recommendations for perioperative tracheostomy management for surgery in the prone position.

Case Report

Informed consent and written permission for review of this patient's medical record and presentation of this case report was obtained from the patient's guardian. A 14 year-old, 25 kg adolescent with achondroplasia and Jeune's syndrome presented to the emergency department with upper extremity weakness, complete paralysis of his lower extremities, and urinary incontinence. The patient's past history was also significant for the presence of a tracheostomy and chronic, night-time mechanical ventilation at home. Magnetic resonance imaging revealed spinal cord changes with increased vertebral mobility at $C2$-3. Orthopedic consultation recommended posterior cervical decompression and fusion from the occiput to C4.

The patient's past medical history was significant for nocturnal mechanical ventilatory support, restrictive lung disease, kyphosis and hypertension. His surgical history included a tracheostomy at 2 months of age and two thoracic expansion procedures using sternum division with a gortex patch at 1 and 2 years of age. The patient's pulmonary medications included budesonide, levalbuterol, montelukast and cetirizine. Pulmonary function test, prior to surgery, showed restrictive lung disease with an FEV_1 of 0.21 liters, FVC of 0.24 liters and an FEV_1/FVC ratio of 0.85. These values had not changed significantly over the past 5 years.

Preoperatively, corticosteroids were administered and there was return of full strength in the upper extremities and anti-gravity strength throughout the muscle groups of the lower extremities. Additional

preoperative evaluation revealed significant hypercarbia secondary to non-compliance with nocturnal ventilation Arterial blood gas (ABG) analysis revealed pH 7.31, $PaCO_2$ 114 mmHg, and PaO_2 68 mmHg and the underlying restrictive lung disease. The patient's pulmonary status was optimized with placement of a new cuffed tracheostomy tube and mechanical ventilation with a tidal volume of 10 mL/kg, rate of 18 breaths per minute and positive end expiratory pressure of 5 cm H_2O. Venous blood gas analysis prior to surgery revealed pH 7.36, $PaCO_2$ 69 mmHg and PaO_2 75 mmHg with an inspired oxygen concentration of 35%.

The patient was transported to the operating room and routine American Society of Anesthesiologists monitors were placed. Anesthesia was induced with propofol, fentanyl, and dexmeditomidine. The patient's tracheostomy was changed to a 5.5 mm cuffed Shiley tracheostomy tube (Tyco Healthcare, Pleasanton, CA) which was then sutured to the patient's neck. In the supine position, volume controlled ventilation with a tidal volume of 10 mL/kg was accomplished with a peak inflating pressure (PIP) of 26 mm H_2O maintaining an end-tidal CO_2 ($ETCO_2$) of 35 mmHg. Maintenance of anesthesia included total intravenous anesthesia using propofol, dexmeditomidine, and fentanyl. The patient was positioned prone for the surgical procedure. Ventilation became increasinly difficult in the prone position with lower tidal volumes (5-6 mL/kg) and increased PIP (32 mm H_2O). The $ETCO_2$ increased to 75 mmHg. ABG analysis revealed pH 7.11, $PaCO_2$ 105 mmHg, PaO_2 315 mmHg with an F_iO_2 of 1.0. Despite repositioning of and suctioning through the tracheostomy tube, ventilation remained inadequate with continued low tidal volumes and high peak airway pressures. The decision was made to return to the supine position and terminate the procedure. The patient was placed in a cervical halo for stabilization. The patient's pulmonary status was improved after returning to the supine position with a return of tidal volumes and PIP to baseline values. Postoperative ABG analysis revealed pH 7.30, $PaCO_2$ 65 mmHg, PaO_2 112 mmHg with an F_iO_2 = 0.4.

Thirteen days later, the patient returned to the operating room for completion of his cervical

INTRAOPERATIVE AIRWAY OBSTRUCTION RELATED TO TRACHEOSTOMY TUBE MALPOSITION IN A
PATIENTWITH ACHONDROPLASIA AND JEUNE'S SYNDROME

737

fusion. Induction and maintenance of anesthesia were accomplished in a similar fashion as described above. The patient's tracheostomy was exchanged with a 6.0 mm armored cuffed ETT (Mallinckrodt Inc., St. Louis, MO). $ETCO_2$ and bilateral breath sounds were confirmed. The ETT cuff was inflated to a leak at 20 cm H_2O and the ETT was sutured at the level of the patient's left clavicle. In the prone position, volume controlled ventilation with 8 mL/kg was accomplished with a PIP of 20 cmH_2O maintaining an ETCO2 of 40-45 mmHg. Posterior cervical decompression and fusion proceeded uneventfully. Two months later, the patient underwent a staged thoracic kyphosis repair. He was again placed in the prone position. His tracheostomy tube was exchanged with an armored cuffed endotracheal tube after induction of anesthesia for each surgery. Both procedures, in the prone position, proceeded without pulmonary complications.

Postoperative follow-up showed an improved neurological status with resolution of the weakness in his extremities. Currently, the patient is ambulating, his activity level has returned to baseline, and he is attending school. After his cervical fusion and kyphosis repair, there was improvement in his pulmonary function with an increased FVC to 0.51 liters, FEV_1 of 0.33 liters, and an FEV_1/FVC ratio of 0.64.

Discussion

Asphyxiating thoracic dystrophy, also known as Jeune's syndrome, is an autosomal-recessive disorder resulting in a constricted narrow rib cage with generalized chondrodystrophy.[1,2] Its incidence is estimated at approximately 1 per 100,000-130,000 live births. Patients with Jeune's Syndrome have variability in the severity of their clinical presentation and often present with dwarfism, short ribs, and short limbs.[2] The majority of patients die at a young age from respiratory failure secondary to impaired lung growth, recurrent infections, and atelectasis. Surgical treatment strategies involve procedures to expand the chest wall and allow for improved lung expansion. Three techniques have been described including sternal splitting with placement of a graft, lateral rib expansion, and vertical rib expansion.[3-6] None has been proven to be more efficacious for prolongation of life or improvement of the quality of life.

Our review of the medical literature revealed no preferred surgical technique for the management of Jeune's syndrome.[3-6] The perioperative management of these patients may be complicated by constriction of the narrow thorax and lung hypoplasia which can cause severe restrictive lung disease.[7] Anesthetic considerations issues include severe respiratory insufficiency, hypoxemia at rest, and rapid oxygen desaturation secondary to asynchronous rib and abdominal motion and associated small lung volumes. Suggested ventilatory management includes maintenance of low to normal peak airway pressures to avoid barotrauma.[7]

We postulate that the ventilation difficulties during the first surgery were secondary to tracheostomy tube occlusion. In the prone position, the weight of an anesthetic circuit can alter the position of a tracheostomy tube within the trachea so that the tracheostomy opening may become occluded by the membranous portion of the trachea adjacent to the esophagus (figure 1). This creates a "one-way valve" allowing for inspiration with limited exhalation during positive pressure ventilation. The prone ABG showing adequate PaO_2 but with inadequate CO_2 removal supports this "ball valve" concept. The

Fig. 1
Model representation of the airway. The clear tubing represents the trachea with the darker portion representing the membranous (posterior) portion of the trachea. A: Supine positioning with tracheostomy tube positioned appropriately in the tracheal lumen. B: Prone positioning with demonstration of how the weight of the circuit can alter tracheostomy position causing occlusion of the distal opening of the tracheostomy tube against the membranous portion of the trachea. C: Anterior view of the tracheostomy demonstrating occlusion during prone positioning. D: Armored endotracheal tube with distal opening positioned properly in the tracheal lumen.
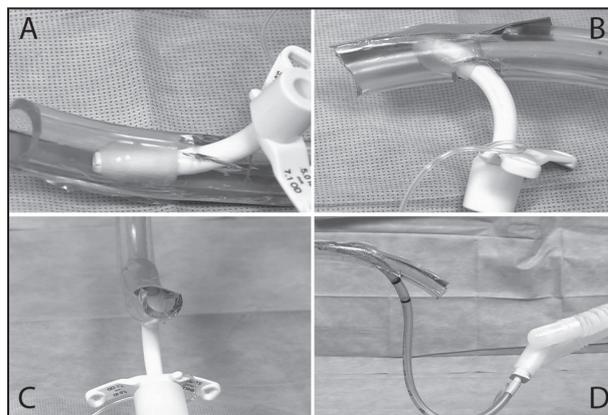
hypothesis is further supported by the elimination of ventilation difficulties with return to the supine position and in the prone position with an armored ETT. The armored endotracheal tube provides two advantages: (1) the Murphy's eye which may provide ventilation if the distal opening is occluded, and (2) the reinforced tubing which provides flexibility and patency in extremes of position thereby avoiding changes in position from the weight of the anesthesia circuit. Another possible etiology of the ventilation difficulties includes compression of the chest wall or trachea; however, all surgeries were done on a similar operating room table and frame (Jackson Table, Relton Hall Frame, Mizuho OSI, Union City, CA) and subsequently changing to an armored ETT provided effective ventilation without any change in patient positioning or use of a different operating room table. Although not done in this case, a fiberoptic bronchoscope could have provided additional information regarding the etiology of the ventilation difficulties with the tracheostomy in the prone position.

Perioperative management of this patient was complicated by the presence of a tracheostomy. Ross et al. proposed several strategies to manage a tracheostomy in the perioperative period including use of an existing tracheostomy, exchange to a cuffed tracheostomy, placement of an ETT through the trachestomy stoma,

or use of standard oro-tracheal intubation.[8] However, after our extensive search of the medical literature using several search engines including PubMed, Medline, and Google Scholar databases (From January 1965 to August 2008) as well as a review of several major Anesthesia and Otolaryngology textbooks, we did not find any guidelines regarding appropriate tracheostomy management for surgery in the prone position.

For patients who present for surgery in the prone position with a tracheostomy, we advocate exchanging the tracheostomy for an appropriately sized armored ETT. Use of the armored tube may avoid positional changes related to traction on the tube due to the weight of the anesthesia circuit (figure 1). We recommend securing the ETT with a suture to the anterior to the anterior chest wall. Exchanging a tracheostomy for an armored endotracheal tube should decrease the possibility of airway obstruction as an etiology of ventilation difficulties in a patient with a tracheostomy. However, given that the tracheostomy site enters the trachea closer to the carina that standard endotracheal intubation techniques, careful placement and auscultation of bilateral breath sounds is necessary to avoid endobronchial intubation. Further research is needed to establish airway management guidelines for tracheostomy patients undergoing surgery in the prone position.

References

1. Morgan NV, Bacchelli C, Gissen P, et al: A locus for asphyxiating thoracic dystrophy, ATD, maps to chromosome 15q13. *J Med Genet;* 2003, 40:431-435.

2. Oberklaid F, Danks DM, Mayne V, Campbell P: Asphyxiating thoracic dysplasia. Clinical, radiological, and pathological information on 10 patients. *Arch Dis Child;* 1977, 52:758-765.

3. Lewandrowski K, Campbell RM, Emans JB: Vertical rib expansion for thoracic insufficiency syndrome - Indications and technique. *Orthopaed J Harv Med Sch;* 2001, 3:65-73.

4. Phillips JD, van Aalst JA: Jeune's syndrome (asphyxiating thoracic dystrophy): congenital and acquired. *Semin Pediatr Surg;* 2008, 17:167-172.

5. Davis JT, Long FR, Adler BH, Castile RG, Weinstein S: Lateral thoracic expansion for Jeune syndrome: evidence of rib healing and new bone formation. *Ann Thorac Surg;* 2004, 77:445-448.

6. Sharoni E, Erez E, Chorev G, Dagan O, Vidne BA: Chest reconstruction in ashyxiating thoracic dystrophy. *J Pediatr Surg;* 1998, 33:1578-1581.

7. Borland LM: Anesthesia for children with Jeune's syndrome (asphyxiating thoracic dystrophy). *Anesthesiology;* 1987, 66:86-88.

8. Ross AK, Gooden CK, Golden S, et al: Society of Pediatric Anesthesia/American Academy of Pediatrics winter meeting review. *Anesth Analg;* 2007, 105:967-973.