Intraoperative airway obstruction in a Duchenne muscular dystrophy patient

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Abstract

Purpose To report a complication of airway obstruction during spinal deformity correction surgery in Duchenne muscular dystrophy (DMD) patient, due to lordoscoliosis, airway malacia, and prone surgical positioning, which was rectified by changing the position of the patient and surgery was successfully completed.

Case description A 15-year-old boy was diagnosed with DMD and admitted for surgical treatment of thoracolumbar scoliosis. The patient's preoperative Cobb's angle was 79° and the kyphotic angle was -19°. During the initial period of surgery, while in the prone position, peak inspiratory pressure (PIP) suddenly increased from 20–21 to 38–41 cmH₂O, wheezing sounds were heard on auscultation of both lungs, and his blood pressure began to fall. Under suspicion of airway problem, intraoperative fiberoptic bronchoscopy was performed which confirmed airway obstruction. Attributing patient's prone position as the

cause of airway obstruction, the surgical position of the patient was changed from prone to semi-lateral. After this change, the PIP stabilized to within normal limits (20–23 cmH₂O). The surgical correction was successfully completed with a posterior-only pedicle screw by the free-hand technique, with the patient in the semi-lateral position for the rest of surgery.

Conclusions Lordoscoliosis and airway malacia in a patient with DMD can lead to occlusion of the tracheobronchial lumen when the patient is in the prone position. Changing the patient's position from prone to semi-lateral can be of help to reverse this airway obstruction and complete the surgery. Pedicular screw insertion can be safely and effectively carried out in this position using free-hand technique.

Keywords Spine · Lordoscoliosis · Duchenne muscular dystrophy · Airway obstruction

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Introduction

Scoliosis is a frequently encountered deformity in non-ambulatory Duchenne muscular dystrophy (DMD) patients that often has negative effects on general health, sitting balance, and wheelchair transmission [16]. This disability ultimately leads to improper sitting posture, pressure sores, costo-pelvic impingement pain, and reduced pulmonary function [12]. For these reasons, surgical correction of the deformity is often necessary [13].

Usually, corrective spinal surgery in patients with DMD is performed through a posterior approach with the patient in prone position, because this allows for preservation of thoracic and pulmonary function, prevention of anesthetic complications caused by single-lung ventilation, and

a relatively short surgical time compared to an anterior approach with the patient in the lateral position [6, 22]. However, the surgical correction of spinal deformities of DMD patients in the prone position is sometimes complicated by deteriorating cardio-pulmonary function and upper airway problems [5, 9, 13]. In particular, thoracic lordosis concomitant with airway malacia can cause life-threatening intraoperative problems, such as airway obstruction, which may force the surgical team to abandon the procedure [1, 17, 19, 20].

We report a case of airway obstruction which developed while performing the surgical correction of scoliosis in prone position in a DMD patient. This problem was managed by changing the patient's position from prone to semi-lateral to alleviate the compression of the airway, and the surgery was concluded successfully.

Case report

A 15-year-old boy with a diagnosis of DMD who was wheelchair bound for 3 years (from 2008–2011) was admitted with progressive spinal deformity, back pain and positional imbalance (Fig. 1a). In the past, the patient used two types of customized wheel chairs: one without cushioning for short distances or brief movement, and one electric structured wheelchair with a seat cushion and back filler for long-term use. He did not have any problem with respiration, history of breathlessness and wheezing or any

medical history of pulmonary problems. He was more comfortable sleeping on the sides he was not used to sleeping prone.

On physical examination, the patient's height and weight were 158 cm and 32 kg, respectively. The patient was diagnosed with lordoscoliosis (Fig. 1b, c). His preoperative Cobb's angle measured from T8 to L5 was 79° and kyphotic angle measured from T3 to T12 was -19° . Computed tomography (CT) showed a distorted chest wall, deformed spine, and anterior-posteriorly flattened chest (Fig. 1d). Preoperative pulmonary function tests (PFTs) reported a forced vital capacity (FVC) of 44 %, forced expiratory volume for one second (FEV_1) of 37 %, FEV_1 /FVC ratio of 77 %, and a peak expiratory flow rate of 35 %. Comparing the patient's results to the predicted values, and based on his flow-volume curve, there were no signs of obstructive pulmonary pathology on his PFTs. Chest radiographs, electrocardiogram, and echocardiography were reported as normal. Based on this information, deformity correction surgery in the prone position with a posterior-only pedicle screw system was planned.

After the patient arrived in the operating room, general anesthesia was induced with an intravenous infusion of propofol and remifentanyl using a target-controlled infusion system (Orchestra Base Primea, Fresenius Vial, France) and endotracheal intubation was facilitated by rocuronium, 0.6 mg/kg. The patient's trachea was intubated using a 7.0-mm (I.D.) wire-reinforced tube (Mallinckrodt Medical, Athlone, Ireland). The insertion depth was 20 cm at the

Fig. 1 **a** The patient presented with scoliosis, pelvic tilting, and costopelvic impingement. **b** An anterior–posterior plain radiograph showed that the patient had scoliosis with a Cobb's angle from T8 to L5 of 79° . **c** A lateral plain radiograph showed that the patient had

lordosis on the thoracic spine, and the kyphotic angle from T3 to T12 was measured as -19° . The apex of the lordosis is located on T3 or 4 areas. **d** Computed tomography showed a decreased distance between the sternum and vertebrae (white arrow)

upper incisors, and both lung sounds were even and clear with manual bagging. Peak inspiratory pressure (PIP) was 17–18 cmH₂O in the supine position, with a tidal volume of 300 mL and respiration rate of 14 breaths per minute. After induction and confirming the patient's respiratory status, airway examination with flexible bronchoscopy was performed as per the protocol followed at our institute. On pre-positioning bronchoscopy, slight segmental airway malacia and narrowing of lower trachea was observed but other anatomical abnormalities were not detected.

Although PIP was slightly increased to 20–21 cmH₂O after moving the patient to the prone position; he showed no signs of respiratory distress and was maintaining his oxygen saturation adequately. Anesthesia was maintained with oxygen, medical air, and continuous infusion of propofol and remifentanyl. Additional muscle relaxants were not used to enable the monitoring of spinal cord and peripheral nerves, using motor-evoked potentials throughout the period of surgery.

After cleaning and draping free the surgical site, posterior midline incision was taken and spine was exposed using monopolar cautery for dissection and Cobb elevator for paraspinal muscle retraction. Thirty minutes after the start of surgery, the patient suddenly showed signs of airway distress including increased PIP to 38–41 cmH₂O and bilateral wheezing sounds. A flexible fiberoptic bronchoscope (Olympus LF-P, Olympus Optical Company, Tokyo, Japan) exam with manual ventilation was performed immediately, and revealed severely narrowed segment of the tracheo-bronchial lumen located approximately 22–25 cm away from the upper incisors (Fig. 2). The endotracheal tube was advanced along the fiberscope to 1 cm above the carina and was finally positioned at 25 cm at the upper incisors, but PIP remained high and ventilation did not improve.

Therefore, the position of the patient was immediately changed from prone to supine, and 10 min later, his PIP decreased to 20–22 cmH₂O. With close and vigilant monitoring of airway pressure, we tried to make him prone again but the PIP increased again making ventilation impossible.

From above, we inferred that prone positioning is causing airway obstruction and the position of the patient was changed from supine to lateral, and then to the left-up semi-lateral position (inclined about 45° to horizontal) with the hope of being able to continue the operation. In the semi-lateral position, PIP stabilized to 20–23 cmH₂O (Fig. 3) and the surgical procedure was resumed. Pedicle screws were inserted by the free-hand technique, being careful not to compress the chest [14, 15]. The patient's vital signs remained stable during the remainder of the operation. After surgery, patient was extubated under careful airway pressure and lung compliance monitoring. The extubation was performed without difficulty and the patient had no pulmonary complication.

Fig. 2 Lower trachea is distorted with almost total obstruction of the airway

Fig. 3 The patient position was changed from prone to semi-lateral during the operation

The postoperative Cobb's angle was 32° and kyphotic angle was 10° (Fig. 4a, b). On postoperative day 5, bronchoscopy showed airway malacia in the lower trachea and both proximal main bronchi (Fig. 4c, d).

Fig. 4 **a** An anterior–posterior plain radiograph showed that the scoliosis was corrected and that the postoperative Cobb's angle was 32°. **b** A lateral plain radiograph showed that lordosis on the thoracic spine was corrected and the kyphotic angle was 10°. **c** Narrowing of

the mid to lower trachea was confirmed on postoperative bronchoscope. **d** Airway malacia was diagnosed based on the reduced cartilage in the lower trachea and both proximal main bronchi. Except airway malacia, there were no other anatomical abnormalities

Discussion

Scoliosis is one of the most frequent complications of DMD, with an incidence rate of 69–90 %. Spinal deformity in DMD generally takes a long C shape in the coronal plane. In the sagittal plane, patients often have kyphoscoliosis, lordoscoliosis, or hyperlordosis with spinal rigidity [10, 18, 21]. Oda et al. [18] reported 39 % of patients having scoliosis in DMD have lordoscoliosis. The exact incidence of lordoscoliosis in DMD is poorly reported in the literature. Similarly percentage of DMD patients with airway malacia who do not require respiratory assistance is known to be uncommon although the exact incidence rate has not been reported through a well-structured study with larger numbers [2].

Based on this information, lordoscoliosis concomitant with airway malacia can be taken as rare. There is one case report in the literature describing these complex complications which happen when DMD patient is turned prone for surgery. The decrease of the sterno-vertebral distance by lordoscoliosis, and softening of the tracheal ring by airway malacia was taken as the possible cause for the airway obstruction [20].

If airway obstruction during surgery in the prone position occurs, continuation of surgery is practically not possible considering life-threatening status of the complication. In addition, correction of long-C-shape scoliosis is difficult through an anterior-only approach, and the insertion of pedicle screws in any other position except prone is also difficult due to the three-dimensional nature of the

deformity, which limits the use of a C-arm [20]. We overcame this problem by changing the position of patient to semi-prone and use of free-hand surgical technique to insert pedicular screws which eliminates the need of C-arm. To the best of our knowledge, there are no similar reported cases in the literature.

In our case, in spite of his benign, non-obstructive PFT pattern and lack of any history of pulmonary problems, the patient had three potential risk factors that could lead to airway obstruction. First, the patient's chest radiographs and CT showed a decrease in the upper thoracic anterior–posterior diameter due to thoracic lordoscoliosis (Fig. 1c, d). This anatomical change causes the trachea to become more vulnerable to compression by the spine posteriorly and to the manubrium sterni anteriorly [1, 19, 20]. The narrowed portion of the lower trachea (22–25 cm from incisors) corresponded to the apex of the lordoscoliosis in sagittal plane (T3 or 4) (Fig. 1). Furthermore, in prone position, the trachea could have been indented by the vertebrae posteriorly, or by the manubrium anteriorly, or both. Secondly, the patient also had airway malacia, in which the C-shaped cartilage of the trachea lose their curvature and cause the trachealis muscle, which covers the posterior side of the tracheal cartilages, to become loose and floppy as opposed to forming a tight muscle canopy [11, 19, 20]. Third, the scoliosis surgery in DMD patients is usually done in prone position and the pressure applied by the surgeon's hands and instruments while exposing the spine might have further aggravated the compression on the already vulnerable airways [19, 20]. Among the above-mentioned causes, the only

one that can be addressed and corrected during surgery is position. Therefore, we changed the surgical position to one that was acceptable for pedicle screw insertion without exacerbating the airway problem (i.e., the semi-lateral position). By changing the position from prone to semi-lateral, the trachea was likely decompressed, indicated by normalization of PIP, allowing us to proceed with the surgery.

When the surgical position of the patient is changed to semi-lateral from prone, additional rotation of the vertebrae is introduced by coronal and sagittal tilting of the body. The use of an image intensifier to insert pedicle screws may not be practical due to overlapping shadows of the bony elements in this position, and is further complicated by scoliosis deformity. Therefore, in this scenario the free hand technique is a better option as it does not depend on the use of an image intensifier and allows the surgeon to finish the operation without prolonging the average operation time [15].

In summary, surgeons should be prepared to handle intraoperative airway obstructions during surgery on DMD patients with lordoscoliosis and/or airway malacia. Bartlett et al. [1] suggested that all children with thoracic lordoscoliosis require regular monitoring of respiratory function in cases of scoliosis with neurofibromatosis or Turner syndrome. They also suggested that if patients have pulmonary symptoms or abnormal lung function tests, ventilation-perfusion scans or three-dimensional computed tomography scans should be performed during preoperative assessment for prevention of airway obstruction. This protocol is supported by the American Thoracic Society for the preoperative screening of patients to determine fitness for anesthesia and for extensive deformity correction surgery [3, 4, 7, 8]. Although intraoperative airway obstruction may not be expected, since the flow–volume curve of a DMD patient may be normal except for a decrease in volume due to muscular dystrophy, spine surgeons should be careful and have plans in place to control emergent airway obstruction. Changing patient position from prone to semi-lateral is one option to manage airway obstruction, and the free-hand technique can be used to successfully correct the curve and complete the surgery in such cases.

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Conflict of interest None.

Ethical statement This study was performed under approval of Institutional Border Review.

References

- Bartlett W, Garrido E, Wallis C, Tucker SK, Noordeen H (2009) Lordoscoliosis and large intrathoracic airway obstruction. *Spine* 34:E59–E65. doi:10.1097/BRS.0b013e318191f389 00007632-200901010-00029 [pii]
- Baydur A, Kanel G (2003) Tracheobronchomalacia and tracheal hemorrhage in patients with Duchenne muscular dystrophy receiving long-term ventilation with uncuffed tracheostomies. *Chest* 123:1307–1311
- Birnkrant DJ (2009) The American college of chest physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing anesthesia or sedation. *Pediatrics* 123(Suppl 4):S242–S244. doi:10.1542/peds.2008-2952J
- Birnkrant DJ, Panitch HB, Benditt JO, Boitano LJ, Carter ER, Cwik VA, Finder JD, Iannaccone ST, Jacobson LE, Kohn GL, Motoyama EK, Moxley RT, Schroth MK, Sharma GD, Sussman MD (2007) American college of chest physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing anesthesia or sedation. *Chest* 132:1977–1986. doi:10.1378/chest.07-0458
- Chong HS, Moon ES, Park JO, Kim DY, Kho PAB, Lee HM, Moon SH, Kim YS, Kim HS (2011) Value of Preoperative Pulmonary Function Test in Flaccid Neuromuscular Scoliosis Surgery. *Spine* 36:E1391–E1394. doi:10.1097/Brs.0b013e31820cd489
- Dobbs MB, Lenke LG, Kim YJ, Luhmann SJ, Bridwell KH (2006) Anterior/posterior spinal instrumentation versus posterior instrumentation alone for the treatment of adolescent idiopathic scoliotic curves more than 90 degrees. *Spine (Phila Pa 1976)* 31:2386–2391. doi:10.1097/01.brs.0000238965.81013.c5 00007632-200609150-00021 [pii]
- Finder JD (2009) A 2009 perspective on the 2004 American thoracic society statement, “respiratory care of the patient with Duchenne muscular dystrophy”. *Pediatrics* 123(Suppl 4):S239–S241. doi:10.1542/peds.2008-2952J
- Finder JD, Birnkrant D, Carl J, Farber HJ, Gozal D, Iannaccone ST, Kovesi T, Kravitz RM, Panitch H, Schramm C, Schroth M, Sharma G, Sievers L, Silvestri JM, Sterni L (2004) Respiratory care of the patient with Duchenne muscular dystrophy: ATS consensus statement. *Am J Respir Crit Care Med* 170:456–465. doi:10.1164/rccm.200307-885ST
- Gill I, Eagle M, Mehta JS, Gibson MJ, Bushby K, Bullock R (2006) Correction of neuromuscular scoliosis in patients with preexisting respiratory failure. *Spine* 31:2478–2483
- Granata C, Merlini L, Cervellati S, Ballestrazzi A, Giannini S, Corbascio M, Lari S (1996) Long-term results of spine surgery in Duchenne muscular dystrophy. *Neuromuscul Disord* 6:61–68
- Gray H, Standring S, Ellis H, Berkovitz BKB (2005) Gray’s anatomy : the anatomical basis of clinical practice. Elsevier Churchill Livingstone, Edinburgh, New York
- Majd ME, Muldowny DS, Holt RT (1997) Natural history of scoliosis in the institutionalized adult cerebral palsy population. *Spine* 22:1461–1466
- Mercado E, Alman B, Wright JG (2007) Does spinal fusion influence quality of life in neuromuscular scoliosis? *Spine* 32:S120–S125
- Modi H, Suh SW, Song HR, Yang JH (2009) Accuracy of thoracic pedicle screw placement in scoliosis using the ideal pedicle entry point during the freehand technique. *Int Orthop* 33:469–475. doi:10.1007/s00264-008-0535-x
- Modi HN, Suh SW, Fernandez H, Yang JH, Song HR (2008) Accuracy and safety of pedicle screw placement in

- neuromuscular scoliosis with free-hand technique. *Eur Spine J* 17:1686–1696. doi:[10.1007/s00586-008-0795-6](https://doi.org/10.1007/s00586-008-0795-6)
16. Modi HN, Suh SW, Yang JH, Cho JW, Hong JY, Singh SU, Jain S (2009) Surgical complications in neuromuscular scoliosis operated with posterior- only approach using pedicle screw fixation. *Scoliosis* 4:11. doi:[10.1186/1748-7161-4-11](https://doi.org/10.1186/1748-7161-4-11)
 17. Myer CM 3rd, Wiatrak BJ, Cotton RT, Bove KE, Bailey WW (1989) Innominate artery compression of the trachea: current concepts. *Laryngoscope* 99:1030–1034. doi:[10.1288/00005537-198210000-00011](https://doi.org/10.1288/00005537-198210000-00011)
 18. Oda T, Shimizu N, Yonenobu K, Ono K, Nabeshima T, Kyoh S (1993) Longitudinal study of spinal deformity in Duchenne muscular dystrophy. *J Pediatr Orthop* 13:478–488
 19. Oh AY, Kim YH, Kim BK, Kim HS, Kim CS (2002) Unexpected tracheomalacia in Marfan syndrome during general anesthesia for correction of scoliosis. *Anesth Analg* 95:331–332 table of contents
 20. Rittoo DB, Morris P (1995) Tracheal occlusion in the prone position in an intubated patient with Duchenne muscular dystrophy. *Anaesthesia* 50:719–721
 21. Smith AD, Koreska J, Moseley CF (1989) Progression of scoliosis in Duchenne muscular dystrophy. *J Bone Joint Surg Am* 71:1066–1074
 22. Sucato DJ, Elerson E (2003) A comparison between the prone and lateral position for performing a thoracoscopic anterior release and fusion for pediatric spinal deformity. *Spine (Phila Pa 1976)* 28:2176–2180. doi:[10.1097/01.BRS.0000084641.96288.8D](https://doi.org/10.1097/01.BRS.0000084641.96288.8D)