

Anesthetic Care During Lateral Thoracic Expansion for Asphyxiating Thoracic Dystrophy

Mineto Kamata^{a, d}, Roby Sebastian^a, Aymen Naguib^{a, b}, Joseph D. Tobias^{a, b, c}

Abstract

Jeune syndrome or asphyxiating thoracic dystrophy (JATD) is a rare autosomal recessive disorder with an estimated incidence of 1 in 100,000 to 130,000 live births. The typical phenotype of JATD is osteochondrodysplasia (narrow thorax and shorted ribs) in association with short limb dwarfism. The thoracic dysplasia results in respiratory insufficiency, usually requiring tracheostomy and chronic mechanical ventilation beginning in infancy and childhood. Lateral thoracic expansion is one surgical procedure that has been described in an attempt to enlarge the thoracic cage and alleviate the progressive respiratory failure. In addition to respiratory involvement, patients with JATD may have other co-morbid conditions involving the kidneys, pancreas, liver, and eyes. Given the underlying thoracic dysplasia and other associated comorbid conditions, the intraoperative care of such patients can be challenging. We present two patients with JATD who presented for lateral thoracic expansion. Previous reports of the intraoperative care of such patients are reviewed and options for anesthetic care are discussed.

Keywords: Pediatric anesthesia; Jeune syndrome; Asphyxiating thoracic dystrophy; Lateral thoracic expansion

Introduction

Jeune syndrome or asphyxiating thoracic dystrophy (JATD) was first described in 1954 by Jeune et al [1]. It is a rare, autosomal recessive disorder with an estimated incidence varying

from 1 in 100,000 to 1 in 130,000 live births [2]. The typical characteristic of JATD is an osteochondrodysplasia characterized by a narrow thorax and shortened ribs in association with short-limb dwarfism. The thoracic dysplasia results in progressive respiratory insufficiency with up to 60-70% of homozygous carriers dying from respiratory failure or chronic pulmonary infections. Despite the skeletal deformity affecting the thoracic cavity, pathologic studies suggest that alveolar and lung growth potential may be normal [2]. Although the clinical manifestations of respiratory insufficiency vary in magnitude from lethal to subclinical forms, comorbid conditions associated with JATD may lead to severe organ dysfunction affecting the kidney, pancreas, liver, and eyes in later years [3].

A number of surgical techniques have been reported in an attempt to limit the impact of the thoracic deformities on respiratory function. Lateral thoracic expansion (LTE) enlarges the thoracic cage in patients with JATD by separating, reconstructing, and lengthening the ribs to expand the thoracic cage. Although LTE remains a safe and effective procedure for this patient population, there are few reports discussing the intraoperative care with particular attention to acute, intraoperative changes in respiratory function. We present two patients with JATD presenting for LTE surgery. Previous reports of the intraoperative care of such patients are reviewed. Options for anesthetic care are discussed with a focus on acute changes in intraoperative ventilator settings and compliance changes.

Case Reports

Institutional Review Board approval for case reports involving two or fewer patients was not required by Nationwide Children's Hospital (Columbus, OH).

Case 1

A 19-month-old, 8.1 kg boy presented for general anesthesia for left LTE. He had a history of skeletal dysplasia, hypoplastic thoracic dystrophy, scoliosis, hip dysplasia, craniosynostosis, and pulmonary hypoplasia. Past surgical history included a right LTE at 7 months of age following a tracheostomy and craniosynostosis repair at 16 months of age. He was admitted to the hospital from home 1 day prior to surgery, requiring

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^aDepartment of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, Columbus, OH, USA

^bDepartment of Anesthesiology & Pain Medicine, The Ohio State University College of Medicine, Columbus, OH, USA

^cDepartment of Pediatrics, Nationwide Children's Hospital & The Ohio State University College of Medicine, Columbus, OH, USA

^dCorresponding Author: Mineto Kamata, Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, 700 Children's Drive, Columbus, OH 43205, USA. Email: Mineto.Kamada@Nationwidechildrens.org

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Table 1. Ventilation Parameters of Cases 1 and 2

Parameters	At incision	After division of ribs	After attachment of plates	At the end of surgery
Case 1				
PIP (cm H ₂ O)	25	31	31	31
Vt (mL/kg)	10	13	13	12
RR (bpm)	26	26	26	26
PEEP (cm H ₂ O)	6			
FiO ₂ (%)	39	39	40	40
SpO ₂ (%)	100			
EtCO ₂ (mm Hg)	43	50	50	47
I:E	1:2			
Case 2				
PIP (cm H ₂ O)	32	32	31	31
Vt (mL/kg)	13	13	14	13
RR (bpm)	23	25	22	22
PEEP (cm H ₂ O)	10			
FiO ₂ (%)	54	56	55	75
SpO ₂ (%)	100			
EtCO ₂ (mm Hg)	34	33	32	44
I:E	1:2			

Pressure-limited ventilation was used during the case. PIP: peak inspiratory pressure; Vt: expiratory tidal volume; RR: respiratory rate; bpm: breathes/minute; PEEP: positive end expiratory pressure; FiO₂: fraction of inspired oxygen; SpO₂: pulse oximeter oxygen saturation; EtCO₂: end tidal CO₂; I:E: inspiratory to expiratory time ratio.

chronic mechanical ventilation 24 h a day via tracheostomy using a Respironics Trilogy home ventilator. His preoperative oxygen saturation was 100% with synchronized intermittent mandatory ventilation (SIMV) in the pressure-controlled mode. Preoperative ventilator settings included: respiratory rate (RR) 26 breathes/min, peak inspiratory pressure (PIP) 30 cm H₂O, positive end expiratory pressure (PEEP) 12 cm H₂O, pressure support 16 cm H₂O, inspiratory time 0.7 s, and FiO₂ 30%. Mechanical ventilation was achieved through a 4.0 mm cuffed tracheostomy tube. Preoperative laboratory evaluation revealed a hemoglobin of 14.7 g/dL and a hematocrit of 43.2%. Prior to surgery, a 24-gauge intravenous cannula was placed and he was transported to the operating room. After American Society of Anesthesiologists (ASA) monitors were applied, anesthesia was induced with sevoflurane (end-tidal concentration 2%) and fentanyl (3 µg/kg). Neuromuscular blockade was provided by rocuronium (1.5 mg/kg). At the beginning of the surgery, intercostal blockade with 0.5% of bupivacaine was applied by the surgeon. There was no significant change in the vital signs during anesthetic induction. Following anesthetic induction, he was turned to the right lateral decubitus position and mechanical ventilation initiated with the anesthesia machine ventilator: PIP 24 cm H₂O, RR 26 breathes/min, PEEP 10 cm H₂O, inspiratory/expiratory time (I:E) ratio 1:2, and FiO₂ 40%. The expiratory volume varied from 12 to 14 mL/kg. Anesthesia was maintained with isoflurane (end-tidal concentration 1%), fentanyl (total intraoperative dose of 15.4 µg/kg), and rocuronium. Intraoperatively, the ventilator settings

were adjusted to maintain a tidal volume of approximately 10 mL/kg. The settings for mechanical ventilation are listed in Table 1 at various times including 1) prior to incision, 2) after division of the ribs, 3) after attachment of plates, and 4) at the completion of surgery. At each of these points, pressure-controlled ventilation was provided to achieve normocapnia. Following the surgical procedure, the patient was transported to the cardiothoracic intensive care unit (CTICU). Postoperative analgesia was provided by nurse-controlled analgesia (NCA) with fentanyl, rescue doses of enteral oxycodone, and scheduled intravenous acetaminophen followed by ketorolac. He was discharged from the CTICU and hospital on postoperative days 1 and 7, respectively. At discharge, his oxygen saturation was 100% with SIMV in the pressure-controlled mode. The ventilator settings included: RR 26 breathes/min, PIP 30 cm H₂O, PEEP 12 cm H₂O, pressure support 16 cm H₂O, inspiratory time 0.7 s, and FiO₂ 30%.

Case 2

A 16-month-old, 7.1 kg boy with JATD presented for general anesthesia for right LTE. He had a complex medical history including JATD requiring chronic mechanical ventilation via tracheostomy using a Respironics Trilogy home ventilator, pulmonary hypertension, and bilateral clubfeet. Past surgical history included a Nissen fundoplication and gastrostomy tube insertion at 6 months of age. He had been admitted for ap-

proximately 3 months prior to surgery due to pseudomonas aeruginosa bacteremia, tracheitis, and pneumonia. His preoperative oxygen saturation was 98% with SIMV pressure-controlled ventilation: RR 26 breathes/min, PIP 30 cm H₂O, PEEP 10 cm H₂O, PS 18 cm H₂O, pressure support 18 cm H₂O, inspiratory time 0.4 s, and FiO₂ 30%. Preoperative laboratory evaluation revealed a hemoglobin of 11.1 g/dL and a hematocrit of 34.5%. Prior to the surgery, a 24-gauge intravenous cannula was placed. Following premedication with midazolam, he was transported to the operating room. After ASA standard monitors were applied, anesthesia was induced with sevoflurane (end-tidal concentration 2-3%) and fentanyl (1.5 µg/kg). Neuromuscular blockade was achieved with rocuronium (1.5 mg/kg). There was no significant change in the vital signs during anesthetic induction. Following anesthetic induction, the 3.5 mm cuffed tracheostomy tube was changed to 3.5 mm cuffed endotracheal tube and he was turned to the left lateral decubitus position. Mechanical ventilation was initiated with the anesthesia machine ventilator using the following settings: PIP 24 cm H₂O, RR 25 breaths/min, PEEP 10 cm H₂O, I:E ratio 1:2, and FiO₂ 55%. The expiratory tidal volume was 109 mL (15 mL/kg). Anesthesia was maintained with isoflurane (end-tidal concentration 1%) and fentanyl (total intraoperative dose of 8.5 µg/kg). Intermittent doses of rocuronium were administered as needed to provide neuromuscular blockade. The settings for mechanical ventilation at the same points as case 1 are listed in Table 1. Following the surgical procedure, the patient was admitted to the CTICU. Postoperative analgesia was provided by nurse controlled analgesia with fentanyl and scheduled intravenous acetaminophen and ketorolac. The remainder of his postoperative course was unremarkable. At discharge, his oxygen saturation was 99% with SIMV in the pressure-controlled mode. The ventilator settings included: RR 26 breathes/min, PIP 30 cm H₂O, PEEP 10 cm H₂O, pressure support 18 cm H₂O, inspiratory time 0.6 s, and FiO₂ 30%.

Discussion

JATD is a disease process that may have effects in multiple organ systems. The respiratory symptoms vary widely from respiratory failure requiring tracheostomy placement and chronic mechanical ventilation from infancy to phenotypes that have mild respiratory involvement that may improve with age [4]. Given that the majority of JATD patients die from respiratory failure in early childhood, one of the keys in the perioperative management of these patients is to limit further respiratory deterioration. Involvement of the chest wall with limited growth and expansion significantly impairs lung growth, leading to pulmonary hypoplasia with restricted thoracic volumes thereby placing these patients at risk for perioperative barotrauma and pneumothorax during positive-pressure ventilation [5]. Although achievement of intraoperative ventilation by pressure-controlled ventilation with lower tidal volumes (6 - 8 mL/kg) has been reported, higher tidal volumes (10 mL/kg) were achieved in both of our patients with an acceptable PIP of 30 cm H₂O [6, 7]. In addition to the potential to result in

barotrauma, higher PIPs may result in increased intra-thoracic pressure and impaired venous return resulting in reduced cardiac output. Given these concerns, pressure-limited ventilation should be adjusted to keep the PIP ≤ 30 cm H₂O. Lengthening the inspiratory time and the use of high respiratory rates may allow for an effective tidal volume and minute ventilation while limiting PIP [8].

During the preoperative assessment, the home ventilation settings should be noted and attempts are made to mimic this pattern of ventilation intraoperatively. Given that the restriction to pulmonary expansion is related to the rigid restriction of the thoracic cavity, it is likely that minimal changes in resistance or compliance can be expected during general anesthesia and the use of neuromuscular blocking agents. This was the case in both of our patients as we noted no clinically significant changes intraoperatively in respiratory compliance with general anesthesia and neuromuscular blockade. Similarly, no immediate changes were noted in compliance immediately following LTE.

In any patient with respiratory insufficiency regardless of the cause, chronic hypoxemia and hypercarbia may result in chronic pulmonary hypertension, cor pulmonale, and right heart failure. Given these concerns, a preoperative echocardiogram is suggested to evaluate pulmonary artery pressures. In patients with pulmonary hypertension, mechanical ventilation and intraoperative therapies may need to be adjusted to minimize the risk of acute elevations of pulmonary artery pressure [9, 10].

In theory, alveolar growth may continue up until 8 years of age [11]. In patients with JATD, pathologic studies suggest that alveolar and lung growth are potentially normal, and that pulmonary impairment is secondary to the narrowing of the thoracic cage. Therefore, it is theorized that if the thorax is expanded, further lung growth can be expected. Various surgical techniques have been suggested for thoracic expansion in patients with JATD including sternal splitting, posterolateral rib cage expansion by placing plates, and vertical expandable prosthetic titanium rib (VEPTR) opening thoracotomy [12-14]. The primary goal of these procedures is to increase the thoracic volume and allow for lung expansion and growth.

LTE, the surgical procedure performed in our two patients, has been used to safely and effectively expand the thoracic cage in children with JTAD [15, 16]. The procedure involves separation of a segment of ribs from their underlying periosteum followed by staggered osteotomies with spreading and expansion maintained by bringing together the long segments of adjacent ribs and attaching them with plates. Although there have been reported improvements of respiratory function after surgery, there are limited or no data evaluating acute intraoperative changes in resistance and compliance [15, 16]. While anecdotal experience noted an intraoperative improvement in respiratory compliance during bilateral thoracoplasty and thoracotomy, ventilation returned to baseline after prosthesis placement [6]. In our case series, we noted no clinically significant improvement or deterioration in pulmonary resistance and compliance intraoperatively, thereby further suggesting that the true benefit of the procedure is to allow for subsequent increased lung growth.

Given the pre-existing impaired respiratory function,

postoperative management for JATD patients should focus on respiratory management with providing adequate pain control. Inadequate pain control can result in decreased respiratory excursion with rapid respiratory rates leading to loss of functional residual capacity (FRC) and atelectasis with its antecedent effects on pulmonary compliance. Atelectasis and a decrease in FRC may also elevate pulmonary artery pressure. Aggressive pulmonary toilet with suctioning and clearance of secretions is also important to limit the risks of nosocomial pneumonia. Postoperative analgesia for our patients was achieved by a multimodal analgesia technique including intercostal nerve blockade intraoperatively under direct vision by the surgeon, nurse-controlled analgesia with fentanyl, and adjunctive agents including acetaminophen or non-steroidal anti-inflammatory agents. Since both of our patients had a tracheostomy and were chronically dependent on home mechanical ventilation, they were transferred to the CTICU and mechanical ventilation continued postoperatively. With effective analgesia and aggressive pulmonary toilet, both transitioned quickly back to their home ventilation regimen.

In addition to involving the thoracic cavity, the skeletal dysplasia of JATD may also involve the spine. The thoracic and lumbar spines appear relatively normal in height and width, while cervical spine abnormalities can be life-threatening [17]. The posterior arch of C1 may curve anteriorly and compress the spinal cord. As such issues may impact airway management and intraoperative positioning, the cervical spine of patients with JATD should be evaluated preoperatively. Although instability is uncommon, if cervical stenosis is suspected on routine radiographic imaging, computed tomographic imaging or flexion/extension MRI studies of the cervical cord should be obtained. In addition to cervical cord compression, central apnea has been reported secondary to C1 compression [18].

The majority of patients with JATD have respiratory involvement either immediately following birth or develop respiratory insufficiency during the first few years of life, requiring tracheostomy and chronic mechanical ventilation. The severity of the thoracic deformity is a main factor of prognosis as the majority of patients will die from respiratory failure within the first few years. However, in specific phenotypes, the thoracic malformation and respiratory problems may be less severe, even decreasing with age [19]. Patients who survive the first year of life, may manifest extrathoracic comorbidities including renal, hepatic, gastrointestinal, and retinal dysfunction.

Renal involvement has been noted in approximately 40% of patients with JATD, beginning in early infancy [20-22]. Progressive renal dysfunction results from a ciliary defect in the nephrons resulting in tubulointerstitial nephropathy and cystic changes with tubular dysfunction, dilation, and fibrosis [23]. Given the primary tubular involvement, an inability to concentrate the urine is frequently noted. Additional clinical symptom includes polyuria, proteinuria, an increase in blood urea nitrogen (BUN) and creatinine, and hypertension [4, 21]. Renal failure remains the primary cause of death in patients with JATD between the ages of 3 and 10 years. In older patients, preoperative screening should include an evaluation of electrolytes, BUN, creatinine, and urinalysis. Perioperatively, urine output may be a poor indicator of intravascular volume

status, as a concentrating defect is frequently noted with polyuria and isosthenuria.

Hepatic involvement occurs in up to 30% of patients with JATD. Hepatic involvement includes prolonged neonatal jaundice, elevated liver enzymes, hepatomegaly, portal hypertension, hyperplasia of the bile ducts and hepatic fibrosis [24]. In most patients, hepatic pathological changes are noted only at autopsy and clinical signs of hepatic insufficiency are rare. However, neonates with cholestasis seem to have a poorer prognosis [25, 26]. As hepatic involvement with fibrosis may be noted in the patients who survive early infancy, an evaluation of hepatic function including hepatic enzymes, bilirubin, and coagulation function is suggested prior to surgical intervention in older patients.

Other comorbid conditions have been reported involving the eye and the gastrointestinal (GI) tract. Eye abnormalities including optic nerve hypoplasia, retinal dystrophy, abnormal retinal pigmentation, nystagmus, myopia, and progressive visual field loss are seen in 15% of the patients with JATD [21, 27, 28]. Age of occurrence has been variable. JATD can also occasionally be associated with GI malformations including cystic changes of the pancreas with fibrosis, Hirschsprung's disease, and malrotation as a manifestation of the ciliopathy [29, 30].

JATD is an autosomal recessive disorder primarily impairing respiratory function with thoracic restriction impairing lung growth. Given the underlying thoracic dysplasia and early mortality due to respiratory impairment, preoperative management for the patients with JATD should be focused on respiratory management and minimizing iatrogenic damage and perioperative deterioration of respiratory function. Comorbid conditions associated with JATD may affect the kidney, pancreas, liver, and eyes. Preoperative identification and evaluation of associated comorbid conditions which can impact the perioperative management is suggested with optimizing of end-organ function prior to surgical intervention.

Conflicts of Interest

None.

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