Total Knee Arthroplasty in a Patient With Diastrophic Dwarfism

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Diastrophic dwarfism is an autosomal recessive disease that predominantly occurs in the Finnish population (1 in 33,000) but has been known to occur worldwide. Affected patients present with multiple cartilaginous anomalies and early degeneration of weight-bearing joints. Once past infancy, life expectancy is favorable and patients may undergo multiple surgical procedures throughout their lifetime to repair or replace affected joints. The characteristic short trunk of these patients in addition to scoliosis, cervical kyphosis, and involvement of articular cartilages can create unique ventilation and airway challenges for anesthesia providers involved in their care.

Keywords: Anesthesia, diastrophic dwarfism, dysplasia, total knee arthroplasty.

A 50-year-old woman with known diastrophic dwarfism presented for right total knee arthroplasty. The patient's short limb length and stature of 132 cm was noteworthy, but the patient otherwise appeared alert, oriented, and younger than her stated age. The patient's weight was recorded as 54.4 kg, and her body mass index (BMI) was 31 kg/m². Previous surgical history for this patient included bilateral hip replacements, left shoulder replacement, left total knee arthroscopy, total abdominal hysterectomy with bilateral salpingo-oophorectomy, and gastric bypass. Her anesthetic history was clinically significant for nausea and vomiting. The patient also complained of an “extremely sore” throat that persisted for several days after a previous general anesthetic. The patient was assigned a Mallampati classification of 3 and an ASA score of 3. Previous anesthetic records indicated “difficult intubation,” use of a Macintosh No. 3 laryngoscope blade, and placement of a 7.5-mm endotracheal tube.

Several aspects of the patient's review of systems were of concern. The patient's pulmonary history was remarkable for obstructive sleep apnea which was said to have improved with weight reduction; however, she used a continuous positive airway pressure mask during sleep. The chest radiograph was clear, and no pulmonary function testing was performed. Past medical history was also notable for depression, occasional headaches of moderate intensity, and “feeling dizzy” with quick rotation of the head. Range of motion in the neck was found to be limited. The patient denied episodes of vertigo or blurred vision. Consistent with patient statements, further neurologic assessment found the presence of lower extremity weakness (left greater than right) but no existing paresthesias. The patient stated that she spent most of her time in an electric wheelchair. Ambulation was said to be infrequent and with walker assist. The patient admitted to gastric reflux and slept elevated on 2 pillows. All other systems were unremarkable, and laboratory testing was within normal limits. The patient's medication regimen included routine use of vitamin, mineral, and iron supplements and propoxyphene with acetaminophen, 1 tablet taken occasionally for arthritic joint pain.

The patient was brought to the operating suite and assisted to the operating table and then placed in the supine position. The use of preoperative sedation was withheld, and the emergency airway cart with fiberoptic scope was made available. Placement of standard monitors revealed a baseline blood pressure of 128/86 mm Hg, heart in normal sinus rhythm at 84/min, and respiratory rate at 12/min. The oxygen saturation was 100% on room air. Preoxygenation was achieved with administration of 100% oxygen via face mask for 5 minutes. The head and neck were stabilized. Induction and intubation was performed in rapid sequence fashion using fentanyl at a dose of 100 µg, propofol at 110 mg, and succinylcholine at 110 mg. Direct laryngoscopy was performed using a No. 3 articulating Macintosh blade, which revealed a grade 2 view with the blade in full articulation. The patient was successfully intubated with a 6.5-mm endotracheal tube on the initial laryngoscopy. Tidal volumes were maintained between 6 and 8 mL/kg, yielding peak airway pressures of 20 to 22 cmH₂O. Muscle relaxation was achieved with an initial bolus of 30 mg of rocuronium, followed by intermittent boluses of 10 mg approximately every 35 minutes to maintain a ratio of 2 of 4 twitches in a train-of-four at 40 mA. Despite an additional dose of 150 µg of fentanyl before surgical incision, the patient's blood pressure increased to 160/98 mm Hg approximately 40 minutes into the surgery. Additional fentanyl dosing was titrated to effect, with a total of 450 µg of fentanyl being administered before the end of this 140-minute case.

Upon conclusion of the surgery, 2 mg of neostigmine

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and 0.4 mg of glycopyrrolate were administered, resulting in 4 of 4 twitches in a train-of-four and 5 seconds of sustained tetanus at 50 mA. The patient began to breathe spontaneously with volumes ranging from 300 to 340 mL at a respiratory rate of 12/min to 14/min. Ondansetron, 4 mg, was given for nausea and vomiting prophylaxis. The patient was extubated when alert and following commands. Supplemental oxygen via a face mask at 8-L flow was delivered to the patient for transport to the postanesthesia care unit (PACU). On arrival at the PACU, the patient complained of pain at a level of 8 on a 0 to 10 pain scale, and hydromorphone, 1 mg intravenously (IV), was given in a divided dose to achieve patient comfort.

**Discussion**

Diatrophic dwarfism, or diastrophic dysplasia, was first described by Lamy and Maroteaux in 1960.1 A mutation of the gene known as the diastrophic dysplasia transporter gene results in impaired functioning of the sulfate-chloride exchange process in the cell membrane.2-5 Reduced uptake of sulfate by the cells and decreased proteoglycan sulfation leads to both functional and structural cartilage defects.2-5

Both general and regional anesthesia can be complicated by the manifestations of diastrophic dwarfism. Spinal deformities can include cervical kyphosis of varying degrees, spinal cord compression, spina bifida, scoliosis and narrowing between the lumbar pedicles, rendering regional anesthesia risky and/or impossible.2-8 In contrast, restrictive pulmonary disease, pulmonary hypertension, frequent respiratory tract infection, and reduced cardiac function can develop because of thoracic dystrophy and scoliosis complicating general anesthetic techniques.3,8 Furthermore, potential airway challenges such as cleft palate, micrognathia, and normal-sized dentition within a small oral cavity may complicate intubation. Patients who present with laryngotracheal stenosis and laryngomalacia may experience complete airway obstruction.3,8

Regional anesthesia was not chosen for this patient because of known spinal stenosis and thoracolumbar scoliosis. It should be noted, however, that other studies have described the successful use of regional anesthesia for total knee arthroplasty in patients with diastrophic dwarfism.5 It is clear that the debate between the utility of such a technique and its safety are still under discussion. Helenius and colleagues3 studied the success of 21 total knee arthroplasties in 14 diastrophic dysplasia patients between February 1992 and March 2000. Although the specifics of the anesthetic are not discussed, all patients were reported to have surgery using either spinal or epidural anesthesia techniques.5

Porter and Mendonca3 also reported the use of regional anesthesia in a 31-week gravid woman with diastrophic dwarfism undergoing cesarean delivery. A combined spinal and epidural anesthesia technique was attempted after magnetic resonance images of the spine were studied and the adequacy of a vertebral space could be determined.3 Despite the extensive preoperative study and anesthetic planning for this patient, the epidural catheter placement was unsuccessful.3 Unfortunately, the patient also suffered respiratory depression after midazolam dosing for anxiety related to an extended procedure, ultimately resulting in an adverse outcome.3

A plan for general endotracheal anesthesia was chosen for our patient after careful airway evaluation. Although the patient did appear slightly micrognathic, her thyromental distance was only slightly reduced and was estimated at 5 to 6 cm. The interincisal distance was also slightly reduced and estimated to be approximately 2.5 fingerbreadths. Range of motion in the cervical spine was found to be rather limited, however, and the patient complained of dizziness with rapid movements of the head. The latter findings were thought to be indicative of probable cervical stenosis and possible atlantooccipital joint instability. Despite these findings, the patient did have a history of difficult but successful endotracheal intubation in our institution. Therefore, careful in-line stabilization of the head and neck were maintained during a rapid-sequence induction. Sedation was withheld to extend optimal ventilatory efforts.

Past surgical records indicating prior intubations using a 7.5-mm endotracheal tube and patient statements of a prolonged postsurgical sore throat were believed to be indicative of probable laryngotracheal stenosis. Therefore, a smaller 6.5-mm endotracheal tube was chosen for intubation. Even with the patient’s previous history of successful intubation, fiberoptic equipment and a “difficult airway” cart were made available in the operating room for emergency backup. Our use of only short-acting induction agents for intubation further supported this plan. An articulating No. 3 Macintosh laryngoscope blade was used for direct laryngoscopy to lift the epiglottis and oropharyngeal tissues without manipulation of the head and neck. This technique yielded a grade 2 view, resulting in successful and atraumatic intubation.

Despite the smaller endotracheal tube, however, the patient complained again of a postsurgical “sore throat,” which she said was of “minor” intensity. An additional point of interest in this case was the patient’s total narcotic requirement of 450 µg of fentanyl. However, preoperative assessment of this patient did reveal chronic joint pain, which was said to be treated with propoxyphene and acetaminophen. Intermittent periods of intraoperative hypertension were therefore believed to be related to the need for pain management and the patient’s probable narcotic receptor upregulation. Consequently, the treatment of choice was a narcotic bolus, to which the patient responded favorably.

Anesthesia management of patients with diastrophic...
dwarfism can be particularly challenging. Anesthesia providers must perform a thorough preoperative evaluation in order to provide a safe anesthetic for this patient population. Computed tomography (CT) and magnetic resonance imaging may be necessary to evaluate atlantooccipital joint stability. Careful preoperative neurologic assessment must be performed and deficits noted. Care should be taken to stabilize the cervical spine throughout the intubation process and surgical procedure. Intubation with a fiberoptic bronchoscope, laryngeal mask airway, or Bullard or articulating laryngoscope may be helpful for reducing neck movement and extension. Although regional anesthesia has been used successfully in those with dwarfism, there should be a specific indication for use of this technique and a known positive benefit-to-risk ratio.

REFERENCES

AUTHORS
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