

# Epidural anesthesia for repeat cesarean delivery in a parturient with Klippel-Feil syndrome

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## Abstract

A patient with Klippel-Feil syndrome, morbid obesity, and scoliosis required cesarean delivery. Her previous cesarean deliveries were performed under general anesthesia. She desired a regional technique. Following aspiration prophylaxis and placement of standard monitors, ultrasound was used to identify midline and L<sub>2-3</sub> interspace. Unintentional dural puncture occurred at 10 cm, with an inability to advance the catheter. On second attempt, an epidural catheter was placed easily. After negative test dose, 18 ml of 2% lidocaine with epinephrine was administered to the patient. A T4 level was achieved. The patient tolerated surgery well. Complete block resolution occurred at 4 hours with no neurologic sequelae.

**Key words:** Cesarean, Klippel-Feil syndrome, neuraxial anesthesia, parturient

## Case Report

A 23-year-old female, G<sub>4</sub>P<sub>2012</sub> at 38-weeks gestation, and with Klippel-Feil syndrome (KFS) presented for repeat cesarean delivery and bilateral tubal ligation. Her pregnancy had been uncomplicated. The patient was morbidly obese, weighing 116 kg and 150 cm tall [body mass index (BMI) 51.5], with a history of scoliosis but no known cardiac or renal abnormalities. She reported a bony abnormality in her sacral area, which she could not further characterize. She had no neurologic deficits either subjectively or on examination. She exhibited the classic physical characteristics of a patient with KFS. She had a Mallampati class 4 airway, with extremely limited neck range of motion, and a thyromental distance of 4 cm. She had significant thoracic kyphoscoliosis as well. She had two prior cesarean deliveries under general anesthesia (GA) utilizing awake fiberoptic intubation (AFI) to secure her airway. She reported that this anesthetic was chosen secondary to the anesthesiologist's concern for potential neurologic damage during attempted neuraxial anesthesia.

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The patient strongly desired to avoid GA, if possible, and to be awake for the birth of her child. After lengthy discussion regarding the risks and benefits of regional anesthesia versus GA, the decision was made to attempt a continuous epidural catheter technique. The emergency airway cart was available in the operating room in the event the patient's airway needed to be secured. Preoperatively, the patient received famotidine, metoclopramide, and sodium citrate for aspiration prophylaxis. In the operating room, noninvasive blood pressure, pulse oximeter, and electrocardiogram were monitored. Cefazolin (2 g) was administered prior to skin incision. With the patient in the upright position, ultrasound was used to identify the midline and the L<sub>2-3</sub> interspace. The area was prepped and draped in sterile fashion. The skin was anesthetized with 1% lidocaine. A 17-G Tuohy needle was advanced using the loss of resistance (LOR) technique. Unintentional dural puncture occurred at 10 cm with brisk cerebral spinal fluid (CSF) flow. Attempts to thread a catheter were unsuccessful, and the Tuohy needle was then removed. No paresthesia was noted during attempted catheter placement. A second attempt was made at the same interspace with LOR to saline at 9 cm. The catheter threaded easily to 13 cm and was secured with adhesive, Tegaderm, and tape. The patient was placed supine with left uterine displacement. A ramp was used to obtain optimal sniffing position and supplemental oxygen was administered via nasal cannula. Catheter aspiration was negative for blood and CSF. Test dose was performed with 2 ml of 2% lidocaine with 1:200,000 epinephrine. No tachycardia or changes in blood pressure were noted. The patient had no subjective sensory or motor changes. Lidocaine

2% with 1:200,000 epinephrine was given through the epidural catheter in fractionated doses up to a total of 18 ml. The sensory level was assessed frequently to minimize the risk of a high block. After 25 minutes, a T4 level was achieved bilaterally to pinprick, and the patient was pain free upon incision. She experienced significant nausea and vomiting during the procedure, which was treated with ondansetron. She was hemodynamically stable throughout the surgery. Following delivery, 20 units of oxytocin in 1,000 ml of lactated Ringer's solution was infused intravenously and 2.5 mg of preservative-free morphine was administered via her epidural. The total surgery time was 45 minutes. The patient was transferred to the post anesthesia care unit for recovery. Her block resolved completely within 4 hours. She was followed up by telephone through postoperative day 11; she reported no evidence of post dural puncture headache or neurologic sequela as a result of her neuraxial anesthetic. The patient was pleased that she could avoid a general anesthetic.

## Discussion

Anesthesiologists are often faced with difficult decisions regarding what type of anesthetic is most appropriate. Patients with Klippel–Feil Syndrome (KFS) can have cardiac and/or renal anomalies as part of their syndrome. This patient had no known cardiac or renal anomalies, nor did she have signs or symptoms of cardiopulmonary disease. Her lung fields were clear and her respirations were unlabored. Her heart sounds were normal, and no murmur was appreciated on examination. Therefore, we did not perform any additional cardiopulmonary evaluation prior to her anesthesia. Patients with KFS are also known to have challenging airways. They typically have a very short neck as a result of missing or fused cervical and upper thoracic vertebrae.<sup>[1]</sup> The fusion may cause severely limited neck mobility. Many of these patients also have mandibular abnormalities and micrognathia.<sup>[2]</sup> Patients with KFS may experience instability in their cervical spine, making direct laryngoscopy and airway manipulation potentially dangerous. Severe neurologic injury in this patient population can occur spontaneously or following minor trauma.<sup>[3]</sup> As a result, these patients' airways are best secured using an awake technique to avoid both neurologic injury and failure to intubate.

In addition to having a high-risk airway, a large percentage of patients with KFS have associated Sprengel's deformity, kyphoscoliosis, lack of vertebral segmentation, and spina bifida, all of which make neuraxial anesthesia challenging.<sup>[4,5]</sup> Neuraxial anesthesia also carries the potential need for emergent tracheal intubation in the event of a high block, local anesthetic toxicity, block failure, or inadequate duration of surgical block. Only a small amount of literature exists

regarding the anesthetic management of parturients with Klippel–Feil syndrome as well as the safety of regional anesthesia in this patient population. In our search of the available literature, we found three cases describing the use of epidural for labor analgesia in patients with KFS, but none for cesarean delivery.<sup>[6,7]</sup> In 2001, O'Connor *et al.* described the successful use of a thoracic epidural for bilateral reduction mammoplasty in a non-pregnant patient with KFS.<sup>[8]</sup> In 1995, Dresner and Maclean described the use of a microspinal catheter in a parturient undergoing cesarean delivery.<sup>[9]</sup> To our knowledge, no one has reported the use of a continuous epidural catheter for anesthetic management of a parturient undergoing cesarean delivery.

When formulating our anesthetic plan, we had taken into account both patient safety and preference. The patient was thoroughly informed of the risks and benefits of both modes of anesthesia and we collectively chose to proceed with a regional technique. We were fully prepared to manage her airway in the case she needed to be urgently intubated. We do not routinely use ultrasound for spinal and epidural placement at our institution. However, the use of ultrasound has been shown to be beneficial when attempting to identify both midline and vertebral level.<sup>[10]</sup> Ultrasound imaging was particularly helpful in this patient, whose anatomy was not normal and whose bony landmarks were very difficult to palpate as a result of morbid obesity and significant thoracic scoliosis. Given the patient's history of an unknown sacral vertebral anomaly and no radiographic images available for review, identifying the correct level for needle insertion was vitally important. We chose to use a continuous epidural technique rather than a single-shot spinal for several reasons. First, the patient was of very short stature (150 cm), and it was unclear what dose of spinal bupivacaine would be appropriate. In addition, a high spinal in this parturient could potentially be devastating, if we were unable to secure her airway on time. This was the patient's third cesarean delivery, possibly resulting in significant scar tissue. This combined with her morbid obesity and addition of a bilateral tubal ligation may have increased the surgical time significantly. We did not want to risk the potential for surgical time to outlast our spinal anesthetic. A well functioning epidural would give us the ability to re-dose, should the surgery be lengthy. On our first attempt, an accidental dural puncture occurred. We attempted to thread a catheter, which would allow for a continuous spinal technique, but this was unsuccessful. This inability to thread the catheter may have been the result of an incomplete dural puncture. Given the patient's spinal abnormalities, we chose not to advance the Touhy further into the subarachnoid space. Another possibility is that our trajectory was slightly off midline and may have caused the catheter to come into contact with the dural sac as it exited the Tuohy needle, obstructing catheter advancement.

A continuous spinal would have been an excellent anesthetic choice, allowing for both slow incremental dosing and the ability to give additional local anesthetic in the event of prolonged surgical time. On the second attempt, we easily placed an epidural catheter. Because of the unintentional hole now present in the dura, we gave a small test dose to ensure that we did not have a spinal catheter. We chose to use the same interspace for the second attempt. We did not want to go above L<sub>2,3</sub>, and we also wished to stay far above the level of the sacral bony abnormality reported by the patient, as we were unclear what exactly this lesion represented.

The patient tolerated the epidural placement and the surgical procedure went well. She was able to remain awake for the birth of her third child, and experience this with her mother present. The patient never developed a dural puncture headache, possibly due to her morbid obesity.<sup>[11]</sup> Given that we were unable to thread a catheter into the spinal space, it is also possible that the puncture may have been incomplete, minimizing her risk of developing a post dural puncture headache.

We hope that this case report of successful anesthetic management of a parturient with KFS, using a continuous epidural technique, gives practitioners more clinical experiences on which to base their choice of anesthetic management in a patient with so many inherent challenges.

## References

1. Naguib M, Farag H, Ibrahim A el-W. Anaesthetic considerations in Klippel-Feil syndrome. *Can Anaesth Soc J* 1986;33:66-70.
2. Stoelting RK, Dierdorf SF. Anesthesia and Co-existing Disease, 4<sup>th</sup> ed. Philadelphia: Churchill Livingstone; 2002. p. 544.
3. Michie I, Clark M. Neurological syndromes associated with cervical and craniocervical anomalies. *Arch Neurol* 1968;18:241-7.
4. Salem MR, Klowden AJ. Anesthesia for Orthopedic Surgery. In: Gregory GA, editor. *Pediatric Anesthesia*. 4<sup>th</sup> ed. Philadelphia: Churchill Livingstone; 2002. p. 650-1.
5. Aebi M. Surgery of a sacral malformation in a Klippel-Feil syndrome. *Eur Spine J* 2009;18:1239-40.
6. Singh D, Mills GH, Caunt JA, Alderson JD. Anaesthetic management of labour in two patients with Klippel-Feil syndrome. *Int J Obstet Anesth* 1996;5:198-201.
7. Buist RJ, Mohandas PM, Wilson TJ. Management of a parturient with Klippel-Feil syndrome. *Anaesthesia* 1988;43:801.
8. O'Connor PJ, Moysa GL, Finucane BT. Thoracic epidural anesthesia for bilateral reduction mammoplasty in a patient with Klippel-Feil syndrome. *Anesth Analg* 2001;92:514-6.
9. Dresner MR, Maclean AR. Anaesthesia for caesarean section in a patient with Klippel-Feil syndrome. The use of a microspinal catheter. *Anaesthesia* 1995;50:807-9.
10. Arzola C, Davies S, Rofaeel A, Carvalho JC. Ultrasound using the transverse approach to the lumbar spine provides reliable landmarks for labor epidurals. *Anesth Analg* 2007;104:1188-92.
11. Faure E, Moreno R, Thisted R. Incidence of postdural puncture headache in morbidly obese parturients. *Reg Anesth* 1994;19:361-3.

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