



When There Seems to be no Options for Cesarean Section
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Objective

- Understand characteristics of Jarcho-Levin syndrome (JLS) and its unique challenges in Cesarean Section (CS).
- Learn about techniques to perform spinal/epidural anesthesia with patients with scoliosis.
- Acknowledge intrathecal catheter can fail despite return of cerebral spinal fluid (CSF) and the importance of having backup plans.

Case: Brief PMH

A 26-year-old female (height 132 cm, weight 32.2 kg, BMI 18.5) G1P000 at 34 weeks gestation, who presented for scheduled primary CS.

CS was indicated due to complex maternal history related to JLS, including spondylothoracic dysplasia status post implant insertion and rib expansion at age 8 with persistent severe restrictive lung disease (FVC 0.81 L; 31% predicted, FEV1 0.71L; 31% predicted), leading to no exercise tolerance at baseline.

Considering her height, distorted anatomy due to extensive back surgery and poor pulmonary function, decision was made to place intrathecal (IT) catheter and titrate up 0.75% bupivacaine until surgical level is achieved. At the end of the case, plan was to bolus 75 mcg of morphine IT for postoperative pain control.



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Evaluation

Initial attempt of placement of IT catheter was made by landmarks; however, CSF was not aspirated despite advancing the Tuohy needle after achieving loss of resistance.

Another attempt was made using ultrasound. CSF return noticed from the Tuohy needle and IT catheter was threaded through the needle.

After confirmation of CSF return through the catheter, 7.5mg (1ml) of hyperbaric bupivacaine was administered, and then 0.2 ml increment was given thereon. After completing administration of 2ml of bupivacaine, the patient only lost sensation to pinprick below right ankle. 12mg of isobaric bupivacaine 0.5% was also attempted, with no resulting block.

At this point, decision was made to induce general anesthesia with rapid sequence induction. She was intubated without difficulty and an arterial line was placed for serial blood gases. The fetus was delivered without issues and oxytocin infusion and tranexamic acid were given. The patient was extubated at the end of the case and sent to ICU for postoperative cares.



Pre-pregnancy CXR and CT abdomen and pelvis, showing vertical expandable prosthetic titanium rib implants and scoliosis, respectively



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Conclusion

- JLS has demonstrated autosomal recessive inheritance and is associated with chromosomal abnormalities, such as MESP2 and DLL3 genes. There are subtypes such as spondylocostal dysplasia (SCD) and spondylothoracic dysplasia (STD).
- JLS is characterized by short trunk of prenatal onset, craniofacial abnormalities, short rigid neck owing to cervical vertebral fusions, fused thoracic vertebrae, “crab-like” rib cage, owing ribs that flare in the fan-like pattern with extensive posterior fusion at the vertebrocostal junction. Significant thoracic restriction occurs in approximately 60% of the newborns, resulting in respiratory distress and around 45% mortality rate by the end of infancy for patients with STD. Scoliosis is not common but it can be severe in some cases.
- Due to her medical and surgical history, single shot spinal anesthesia, epidural anesthesia and general anesthesia were deemed not the best options for this case. Consider placement of IT catheter in patients with JLS but be cognizant of distorted anatomy of the spine, which can lead to failure of IT catheter and unexpected spread of medication. Recognizing that patients in this category have unique anesthetic challenges, preoperative planning is key and requires a multidisciplinary discussion.

References

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