

CASE REPORT

OBSTETRIC ANESTHESIA

Anesthetic management of a parturient with spondylocostal dysostosis (Jarcho-Levin syndrome) for cesarean delivery

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ABSTRACT

Jarcho Levin syndrome was first described by Saul Jarcho and Paul Levin in 1938. The syndrome is characterized by multiple skeletal deformities due to axial skeletal developmental defects during the embryologic stage. Generalized vertebral defects include fused, missing, or deformed vertebrae, and these are seen throughout the cervical, thoracic, and lumbar spine. The patients have short stature and kyphoscoliosis. In pregnancy, these skeletal defects may have pronounced effects on the parturient and the baby, so special considerations are needed by the obstetricians as well as the obstetric anesthesiologists. This case report highlights the challenges our anesthesiology team faces while providing patient care and points out the different anesthetic management approaches that can be used in cesarean delivery for parturients with spondylocostal dysostosis.

Key words: Jarcho-Levin Syndrome; Anesthesia; Congenital; Hereditary; Abnormalities; Growth Disorders; Physical Appearance

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1. INTRODUCTION

We present the anesthetic management of a 22-year-old Hispanic female patient Gravida 1 Para 0 for cesarean delivery at 34 weeks of gestation with a past medical history of Jarcho-Levin syndrome or spondylocostal dysostosis (SCD). The syndrome is characterized by defective embryological development of the axial skeleton early during pregnancy, leading to short trunk dwarfism, short thorax, severe shortening of the spine, and short rigid neck due to cervical vertebral fusion.¹ SCD has an incidence of 1:40,000, more commonly in the Puerto-Rican population.² A written HIPAA authorization has been obtained from the patient for the publication of this case report.

2. CASE REPORT

Our anesthesia team was first consulted to evaluate the patient at 22 weeks of gestation. On physical examination, the patient's height measured 133 cm, and their weight was 40 kg. Airway examination showed a Mallampati grade I view, high arched palate, and short neck with severely restricted extension. Physical examination showed significant kyphoscoliosis, abnormally increased anteroposterior chest diameter, and pectus carinatum (Figure 1). The patient's spine x-ray showed significant cervical spine fusion from the occiput to C-7 level, marked spine shortening, severe dysplastic, partially absent thoracic and lumbar vertebrae, and multiple rib fusion. Her pulmonary



Figure 1: Shows the skeletal features of the spondylocostal syndrome.

function tests showed a moderate restrictive ventilatory defect, and a

transthoracic echocardiogram showed normal cardiac size and function.

We used ultrasonography of the lumbar spine during the preoperative assessment, but we failed to visualize the ligamentum flavum, dura, or the epidural space; only the sacrococcygeal ligament and sacral cornu were visualized. We discussed with the patient that the placement of a lumbar epidural for labor analgesia will be difficult.

As a part of the preoperative evaluation, the otolaryngology team was consulted for difficult airway assessment, especially with significant limited neck mobility, which may intervene with a surgical airway if needed. The flexible laryngoscopy showed elongated epiglottis, a narrowed glottic opening, and mild subglottic stenosis.

At 34 weeks of gestation, the patient presented to the labor and delivery floor, complaining of dyspnea in the supine position. A multidisciplinary team, including obstetrics, otolaryngology, and obstetric anesthesiology, agreed to proceed with the cesarean section under general anesthesia using an awake fiberoptic intubation

approach with the otolaryngology team present in the operating room in case an emergency surgical airway was needed. Preoperatively, the patient received intravenous glycopyrrolate 0.2 mg and xylometazoline nasal spray. ASA standard monitoring was applied in the operating room, and sedation was started using TCI (Target Controlled Infusion) of dexmedetomidine (0.5 $\mu\text{g}/\text{kg}/\text{h}$) while the patient was in a semi-sitting position.

Awake nasal fiberoptic approach was decided to secure the airway after the topical spray of the patient's airway with 0.5% lidocaine. During the fiberoptic intubation, a tracheal web was observed, but it did not interfere with the insertion of the endotracheal tube (ID 5.5 mm) above the level of the carina. After securing the patient's airway and observing a sustained end-tidal capnography (EtCO_2) wave, general anesthesia was induced using propofol

80 mg, rocuronium 25 mg, and 2% sevoflurane for the maintenance of anesthesia. Pressure control with volume guarantee (PCV-VG) ventilation mode was used. An average tidal volume of 250 ml with a peak inspiratory airway pressure not exceeding 25 cmH_2O was achieved. After uneventful cesarean delivery, the neonatal APGAR score was 4 and 7 at 1 and 5 min, respectively. A multimodal analgesic regimen including intravenous fentanyl 100 μg , acetaminophen 600 mg, and ketorolac 15 mg was started. Dexamethasone 8 mg and ondansetron 4 mg IV were given as prophylactic antiemetics. At the conclusion of the surgery, sugammadex 80 mg was used for neuromuscular blockade reversal; spontaneous ventilation was restored, followed by fully awake extubation. The patient was transferred to the post-partum recovery, where she had an uneventful recovery and was later discharged to the ward.

3. DISCUSSION

We present successful anesthetic management of cesarean delivery for a 22-year-old female patient with a past medical history of Jarcho-Levin syndrome, also known as spondylocostal dysostosis (SCD).

This syndrome has a mortality rate of up to 44% in the first six months of life due to pulmonary comorbidities, recurrent aspiration, and restrictive pulmonary lung

disease.¹ SCD patients present with short trunk dwarfism, broad forehead, high arched palate, short thorax, severe shortening of the spine, generalized vertebral segmentation defects, and sickle cell shaped vertebrae. The ribs show extensive posterior fusion at the vertebo-costal junction, short rigid neck due to cervical vertebral fusions, pectus carinatum, and kyphoscoliosis. Schulman et al. described absent distal tracheal rings, wider carina, and narrowing of bronchial lumens, thus explaining the recurrent aspiration and pneumonia in pediatric patients with SCD.³

In addition to the missing vertebrae at the cervical, thoracic, and lumbar spine, spine radiography will show multiple formation defects varying between wedge vertebrae, hemivertebrae, and unfused anterior-posterior parts of the vertebrae, and sickle cell shaped vertebrae. Interestingly, though multiple skeletal malformations are seen throughout the spine, none are noticed in the sacrococcygeal region.¹ Patients' rigid chest wall due to the short thorax and vertebo-costal fusion explains the severe restrictive pulmonary pattern seen in patients' pulmonary function tests.

The planning for an appropriate anesthetic technique was challenging. The diffuse vertebral anomalies and the inability to identify anatomical landmarks using ultrasonography at the lumbar spine level made it difficult to offer lumbar spinal or epidural anesthesia. But as we were able to identify sacrococcygeal ligament, we discussed a possible neuraxial block using an ultrasound-guided caudal epidural catheter placement with careful titration of local anesthetics, considering the potential worsening of the patient's pulmonary status with a higher level of the block. MRI of the spine was scheduled at 34 weeks to confirm the continuity of the epidural space and rule out any interruptions due to the distorted anatomy. Unfortunately, this regional plan was aborted because of the deterioration of the patient's pulmonary status at 34 weeks of gestation and her inability to lie supine.

General anesthesia was our secondary plan, which was challenging due to her difficult airway. Additionally, we were concerned if we would be able to maintain adequate ventilation secondary to potential restrictive lung disease patterns and airway anomalies. In turn, this could have resulted in very high airway pressure and significantly low tidal volume, especially with high abdominal pressure during attempts of fetal extraction.

Up to our knowledge, few reports are available in the literature discussing the anesthetic management of SCD syndrome. Most of these cases were in the pediatric

patient population where supraglottic airway devices had been used to secure the airway.⁴

Dolak et al. described spinal anesthesia in a parturient with SCD with height-adjusted doses of local anesthetic rather than epidural placement due to the possibility of an interrupted epidural space.⁵

4. CONCLUSION

Each spondylocostal dysostosis patient should be assessed on an individual basis. Proper preoperative assessment is crucial. Studying the pathophysiology of the disease, detailed physical examination, investigations, and the review of radiological studies are of great importance in planning the appropriate anesthetic management. Although both regional and general anesthesia are challenging in such cases, the proper and safe anesthetic technique will depend upon the degree of anatomical distortions, patient's general condition, type of surgery, and availability of expertise and equipment facilities of the institute.

5. Conflict of interest

None declared by the authors.

6. Authors' contribution

Both authors took equal part in concept, conduction of study, manuscript editing and critical revision of the manuscript.

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