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Arthrogryposis and a Cesarean Delivery: A Case Presentation
Adam L. Weinstein M.D., Stephen Copeland M.D., Arpad Zolyomi M.D., John Wills M.D.

Introduction

Arthrogryposis multiplex congenita is characterized by progressive multiple joint contractures that are present at birth (dorsal joints are more affected than proximal joints). The origin is primarily neurogenic (>90% of cases) or myogenic with multiple and complex abnormalities. Arthrogryposis is a constellation of joint contractures of complex congenital neurologic and myopathic etiologies. It is often seen with a variety of other systemic defects: cardiac lesions, pulmonary lesions, CNS lesions, and craniofacial deformities. (1) Central core disease and myasthenia gravis is also common. 

Associated Conditions

Table 1

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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<tbody>
<tr>
<td>Central core disease</td>
<td>Pathologic abnormalities in skeletal muscle with spontaneous muscle fiber regeneration</td>
</tr>
<tr>
<td>Myasthenia gravis</td>
<td>Autoimmune disorder affecting the neuromuscular junction causing muscle weakness</td>
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<tr>
<td>Spinal muscular atrophy</td>
<td>Progressive muscle wasting and atrophy often beginning early in life</td>
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</tbody>
</table>

Case Presentation

We present the anesthetic management of a 31 year old female at 36 weeks and 5 days gestation with arthrogryposis complicating restrictive lung disease, vocal cord paralysis, mitral regurgitation, and gestational diabetes.

Figure 1

She was 5 feet 1 inch in height and weighed 55.4 kg. There was a noticeable hoarse voice, scoring a 47 on the Voice Handicap Index (VHI) - indicating moderate impairment. She had pre-pregnancy laryngoscopy and videoendoscopic examination which was consistent with Hypothyroid/Larynx Syndrome. The left vocal cord was paralyzed with fixation at the abducted position and the right vocal cord fold stopped in paramedian position (Figure 1). She had a vocal cord injection 10 years prior which was her most recent intubation. 

She had severe muscle wasting of her distal extremities, unable to abort her legs, and was predominantly wheelchair bound. Her hair was deemed a Mallampati class II with limited TMJ mobility, limited mouth opening, and a significantly decreased thyromental distance. Her past surgical history was significant for multiple ankle surgeries under general anesthesia without documentation of any complications.

Our anesthetic concerns stemmed from an increasing oxygen requirement to 3 liters and worsening feelings of dyspnea from 30 weeks gestation. Other anesthetic concerns can be seen in Table 1. One month prior to her delivery she had PTFS’ completed (Figure 2). PTFS demonstrate a restrictive pattern.

Figure

Her pulse oximetry readings were consistently between the mid-80s to low-90s throughout her pregnancy. We were also concerned about airway management due to her vocal cord changes and airway exam described above. There was some concern regarding a low plated level, but her RETOM was normal and she did not have a history of easy bruising or bleeding.

Our anesthetic plan was modified to accommodate our well informed patient’s preference. We initially performed an ultrasound scan of the lumbar spine to assess the possibility of neural axis anesthesia. With encouraging results from the ultrasound scan, the decision was made to proceed with neural axis anesthesia.

The patient was brought to the operating room with two large bore IV’s in place. The standard ASA monitors were applied and the patient was positioned sitting on the operating room table that was tilted 10 degrees to improve lumbar flex. Next the ultrasound scan of the lumbar spine was repeated and the L4-L5 interspace was identified and marked. An epidural catheter was placed on first attempt by a trained anesthesiologist. After which, the patient was an ENT surgeon on standby if an airway problem presented and became unmanageable. The patient delivered a normal appearing male with APGARs of 9 and 9 and weight of 2520 grams. One week status post caesarean section she endorsed pleasant mood and excellent appetite, normal breast feeding every 2-3 hours, and minimal vaginal bleeding. Her oxygen requirement had resolved prior to discharge from the recovery room.

Discussion

Arthrogryposis multiplex congenita has multiple anesthetic implications, especially in a parturient patient. The literature is sparse regarding the optimal anesthetic management for cesarean delivery in this population. Neuroanatomical abnormalities can be complicated by variations in the size of the dural sac affecting access to the epidural space. In addition, complications from a difficult airway can be exacerbated by pulmonary pathology limiting access to the epidural and intrathecal spaces. If the patient has pulmonary or musculoskeletal pathology, general anesthesia may be undesirable because of the risks of paradoxical airway response or uncomfortable or even unbearable as it increases work of breathing required to maintain oxygenation and ventilation.

Rozkowski and Spooner noted decreased sensitivity to depolarizing blockers in AMC patients due to a myopathic/inflammatory etiology. (2) Our patient had decreased sensitivity to succinylcholine and a history of myasthenia gravis.

Our anesthetic plan was modified to accommodate our well informed patient’s preference. We initially performed an ultrasound scan of the lumbar spine to assess the possibility of neural axis anesthesia. With encouraging results from the ultrasound scan, the decision was made to proceed with neural axis anesthesia.

In our patient we developed a multidisciplinary plan with the ENT surgeons and obstetric team in order to optimize patient management. Our first choice was an epidural so that we could titrate the level accordingly and avoid a high spinal due to potentially abnormal CSF fluid dynamics and decreased platelet count. If the epidural failed, we were prepared to place an intracranial catheter for slow titration to attain appropriate surgical level while minimizing the risk of a high spinal we were concerned about with a single shot technique. As an emergency fall back plan, general anesthesia and availability of advanced airway techniques would have been utilized.

Our plan was executed successfully and proves that in such a complicated patient a thorough knowledge of the individual patient presentation can prove fruitful when formulated a standardized anesthetic plan.

References