Clinical Anaesthesia File

Anaesthetic management of a parturient with severe muscular dystrophy, lumbar lordosis and a difficult airway

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Key words

ANAESTHESIA: obstetrical;
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Purpose: This case describes the management of a 19-yr-old wheelchair bound primigravida with severe muscular dystrophy who presented for Caesarean section after spontaneous rupture of membranes. Anaesthesia was influenced by several features of her systemic disease which were impediments to both neuraxial and general anaesthesia.

Clinical features: Other than for a prenatal record and the history obtainable from the patient, little additional medical information was available. Physical examination showed diffuse muscular weakness and an anatomically abnormal airway. Examination of the spine showed slight 10–15° thoracolumbar scoliosis and >45° lumbar lordosis. Fetal assessment was normal. Echocardiography revealed mildly decreased left ventricular function and was consistent with pulmonary hypertension. After discussion with the patient and her obstetrician, elective Caesarean delivery was deemed the best management. Neuroaxial anaesthesia was at an increased risk of failure due to the profound lumbar lordosis. A plan for awake intubation and general anaesthesia was described to the patient in case regional anaesthesia could not be initiated. A fiberoptic bronchoscope and difficult intubation kit were made available. General anaesthesia was expected to have increased risk of postoperative pulmonary complications, hence epidural anaesthesia was attempted. After difficult catheter insertion, a sensory block was titrated to a T₄ level. This was well tolerated by both mother and fetus. A healthy baby was delivered with Apgar scores of 9 and 9. Postoperatively the mother was transferred to the intensive care unit. After 72 hr, the patient's respiratory status allowed transfer to the ward.

Conclusion: This case illustrates the use of epidural anaesthesia in the successful management of a severely compromised patient with limb-girdle muscular dystrophy undergoing elective Caesarean section.

Objectif: Décrire la prise en charge d'une primipare de 19 ans clouée à la chaise roulante par une dystrophie musculaire grave, hospitalisée pour une césarienne après une rupture spontanée des membranes. L'anesthésie a été influencée par plusieurs propres à cette maladie systémique gênant à aussi bien l'anesthésie régionale centrale que l'anesthésie générale.

Caractéristiques cliniques: Peu de renseignements étaient disponibles à part le dossier prénatal et l'histoire obtenue de la patiente. L'examen physique révélait une faiblesse musculaire diffuse et voies aériennes anormales. L'examen du rachis montrait une légère scoliose thoracolombaire et une lordose dépassant 45°. L'évaluation foetale était normale. L'électrocardiographie indiquait une diminution légère de la fonction ventriculaire gauche et était compatible avec une hypertension pulmonaire. Après discussion avec le patiente et son obstétricien, un accouchement par césarienne programmée était décidé. Une anesthésie régionale centrale risquait d'échouer à cause de l'importance lordose lombaire. Pour pallier à un échec de l'anesthésie régionale, un plan comportant une intubation vigile et une anesthésie générale était expliqué à la patiente. Un bronchoscope à fibres optiques et une trousse pour intubation difficile étaient maintenus en disponibilité. Comme l'anesthésie générale augmentait le risque de complications pulmonaires, une anesthésie épidurale était entreprise. Après l'insertion laborieuse d'une cathéter épidural, le niveau de bloc sensitif était progressivement installé à T₄, ce que la mère comme le fœtus ont bien toléré. Un enfant en bonne santé avec des scores d'Apgar de 9 et 9 était mis au monde. Après l'intervention, la mère était transférée aux soins intensifs. Après 72 heures, la condition respiratoire de la patiente permettait le transport à sa chambre.

Conclusion: Cette observation illustre l'utilisation de l'anesthésie épidurale pour la prise en charge d'une patiente gravement atteinte de dystrophie musculaire des ceintures soumise à une césarienne programmée.

A 19-yr-old primigravida patient at 37 wk gestation arrived by air ambulance after spontaneous rupture of membranes 12 hr previously. Her prenatal care comprised four visits to her obstetrician. A history of "severe Duchenne Muscular Dystrophy" was recorded on the antepartum record. She described a history of muscle weakness beginning at the age of three years and progressing to confinement in a wheelchair by age 10 yr. The patient was now totally dependent on nursing care. A complete functional assessment was last done three years earlier in another institution, but the results of these tests were not available. Past surgical history included ventricular septal defect repair at 18 mo and tonsillectomy at 13 yr. The associated anaesthetics were described by the patient as uneventful. There had been no

admissions to hospital during the past three years and no regular follow-up by any medical specialists. She had had one or two bouts of pneumonia each year for the last few years which required only ambulatory treatment. She had stopped smoking at eight weeks gestation. She denied having fever, chills, cough, sputum, or any dyspnoea. The patient did not take any medications and had no allergies. Fetal movements were normal.

This patient presents two issues of concern for her anaesthetic management, muscular dystrophy and pregnancy. She gives a history of severe weakness and frequent respiratory infection, suggesting either impairment of normal clearance mechanisms and/or weakness involving the bulbar muscles with resulting recurrent pulmonary aspiration. Duchenne muscular dystrophy is most commonly an 'X' linked recessive disorder but has been reported in female patients.¹ In any event, the case will need to be managed without knowledge of the exact type of muscular dystrophy.

The anaesthetic considerations for the muscular dystrophies are similar in many respects. All patients with muscular dystrophy show sensitivity to medications including opioids and sedatives and the non-depolarizing muscle relaxants.¹ Patients have been described as existing in a state of "partial curarization."¹ The adverse events reported following administration of succinylcholine vary considerably. For example, some patients with Duchenne muscular dystrophy have suffered hyperkalaemic responses with rhabdomyolysis and cardiac arrest.² The risk of malignant hyperthermia with the use of succinylcholine and inhalational anaesthetics is probably increased both in patients with Duchenne muscular dystrophy and limb-girdle muscular dystrophy but this relationship is difficult to quantify. Avoidance of succinylcholine and the inhalational anaesthetics appears to be the most prudent approach.³

Pregnancy presents a number of considerations for anaesthetic management which are beyond the scope of this discussion. In this case, spontaneous rupture of membranes places the patient at an increased risk for sepsis which would then be a relative contraindication to regional anaesthesia.⁴

Physical examination revealed a gravid 59 kg woman near term in no acute distress. Her heart rate was 110 bpm, blood pressure 100/60 mmHg, respiratory rate 20 per minute. She was afebrile. Physical examination confirmed the presence of profound muscular weakness, which was more severe proximally. Air-

way examination revealed a small mouth, large tongue, reduced neck extension, and a thyromental distance of two fingers. The airway was assessed as Mallampatti Grade III. Air entry was decreased symmetrically, but no adventitious sounds were audible. Some use of the accessory muscles of respiration was apparent. Examination of the cardiovascular system revealed normal neck veins and minimal edema. A left parasternal lift was present with a loud, palpable P2. No murmurs or extra heart sounds were audible. The non-stress test showed a reactive fetal heart tracing with a baseline fetal heart rate of 130 bpm.

Physical examination revealed a high probability of difficult direct laryngoscopy and severe respiratory muscle involvement. Clinically this patient likely has pulmonary hypertension with adequate compensation since there is minimal evidence of right heart failure. To assess the severity of the pulmonary and cardiovascular disease further, arterial blood gases, spirometry, chest x-ray, and transthoracic echocardiography should be requested. In addition, the pulmonary, cardiology, intensive care, and neonatology services should be consulted to allow them to prepare for postoperative care.

The CBC was normal. Arterial blood gases taken with the patient breathing room air showed: pH 7.33, PCO₂ 45 mmHg, PO₂ 68 mmHg, and saturation 92%. Spirometry was performed with the patient supported in the sitting position and good effort was documented by the technician. The FEV_{1.0} was 0.25 L (7% predicted) and FVC 0.61 L (15% predicted). The ECG showed sinus tachycardia with a right bundle branch block and right axis deviation. Transthoracic echocardiography showed normal valvular function, a prominent pulmonary artery, a small ventricular septal defect with persistent left to right shunt, and moderate reduction in left ventricular contractility. Right and left ventricular wall thicknesses were at the upper limits of normal. The chest x-ray showed basilar atelectasis and borderline cardiomegaly but was otherwise unremarkable. The thoracic spine on chest x-ray was essentially normal.

The results of these investigations confirm severe respiratory involvement. A PCO₂ of 45 mmHg is a distinctly abnormal finding in pregnancy since the PCO₂ would be expected to be approximately 32 mmHg.⁴ This patient is in incipient respiratory failure.

This case has features compatible with several of the muscular dystrophies. For example, limb-girdle muscular dystrophy is an autosomal recessive disorder usually, but not exclusively, of intermediate severity with onset in the first to the third decades of life. Cardiac involve-

ment and congestive heart failure may occur. Typical ECG changes are sinus tachycardia and a right bundle branch block. Diaphragmatic muscle may also be involved.¹ Similarly, Duchennes muscular dystrophy is characterized by frequent cardiac and electrocardiographic abnormalities, respiratory weakness, and kyphoscoliosis. Although the exact type of muscular dystrophy presented here was not initially clear, the important consideration is recognition of the commonly shared features of the muscular dystrophies.

There is little or no information in the literature concerning the effects of the muscular dystrophies on uterine muscle function. Potentially, uterine smooth muscle function could be affected, as are cardiac and gastric smooth muscle. This might lead to an abnormal pattern of labour and possibly increasing the risk of postpartum haemorrhage secondary to uterine atony. These are only hypothetical concerns.

Given this patient's condition, a "stat" Caesarean section for fetal distress would be hazardous. At this point, the obstetrician should be informed of these issues and a decision made concerning operative delivery. There are two management paths to be considered. The first is general anaesthesia following awake fiberoptic intubation. If this is chosen, then neuromuscular blocking drugs and opioids should be used in the smallest amounts possible to facilitate the surgery and postoperative analgesia.¹ The second option is either spinal or epidural anaesthesia. Epidural anaesthesia has the advantage of allowing careful titration of block height, recognizing that identification of the epidural space may be technically challenging. In addition, fluid preloading should be done slowly because of the patient's pulmonary hypertension and decreased left ventricular function. Central venous or pulmonary artery pressure monitoring may be helpful in guiding fluid replacement. Finally, the epidural catheter could be used to administer narcotics/dilute local anaesthetic solutions for postoperative pain relief. Although epidural anaesthesia has certain advantages, it does not eliminate the possibility of intraoperative ventilatory failure due either to fluid shifts or additional weakening of the respiratory muscles.

Actual course

The patient was taken to the operating room 24 hr after the membranes ruptured. She was not in active labour and the fetal heart tracing had remained active. The attending anaesthetist decided to proceed with an epidural anaesthetic. If this had been unsuccessful, then an awake intubation was planned followed by induction of general anaesthesia. The

patient consented to this plan and was aware that she would be managed postoperatively in the intensive care unit. With the anaesthetic machine flushed and a fiberoptic bronchoscope, difficult intubation cart, and transtracheal jet in close proximity, the patient was brought to the operating room. Routine monitors were applied, oxygen by nasal prongs continued at $2\text{ L}\cdot\text{min}^{-1}$, and an arterial line was placed in the left radial artery. A large bore *iv* catheter was inserted for crystalloid infusion. Ampicillin and gentamicin were given for prophylaxis against bacterial endocarditis. Initially the epidural was attempted without success with the patient in the lateral position. She was then assisted to the sitting position which produced a minor decrease in the amount of lumbar lordosis. Cannulation of the epidural space was then successful. Using a total of 15 ml lidocaine 2% and 1:200,000 epinephrine in divided doses, the block was gradually titrated to the fourth thoracic dermatome. During this titration approximately 1.0 L of crystalloid was infused without perceived difficulty. The epidural block was well tolerated by both mother and fetus. No sedation was given. Blood loss was estimated at 500 ml. A vigorous 2850 g baby girl was delivered with Apgar scores of 9 and 9. Cord pH was normal at 7.29. At the end of the procedure 3 mg preservative free morphine was injected through the epidural catheter for postoperative pain management and the epidural catheter was left *in situ*. Intraoperative blood gases were omitted but should have been done to assess any worsening of this patient's respiratory status. The patient was then transferred to the intensive care unit for observation. The baby was treated for transient tachypnoea of the newborn which resolved spontaneously over the next few hours. For the next three days the patient's respiratory function was worse than it had been preoperatively. The PCO_2 , measured at 45 mmHg preoperatively, was consistently in the 50–60 mmHg range postoperatively. The patient had no complaint of dyspnoea and minimal postoperative pain. Respiratory function gradually improved over the next three days with oxygen by nasal prongs, vigorous chest physiotherapy, and incentive spirometry. Intubation and positive pressure ventilation were not required. She was transferred to a hospital ward on the third postoperative day and followed by the obstetrician and pulmonologist. The need for home oxygen therapy and home ventilation were to be assessed on an outpatient basis. Sleep studies were also to be arranged to rule out sleep apnea. The patient was subsequently lost to follow up.

Discussion

This case describes a patient with severe muscular dystrophy and an anatomically difficult airway who presented for Caesarean section.⁵ After careful assessment, the attending anaesthetist opted for lumbar epidural anaesthesia, a choice complicated by the patient's severe lumbar lordosis. Two concerns about the care provided were administration of a fluid load in the presence of pulmonary hypertension without central pressure monitoring and use of an untitrated dose of epidural morphine. Although the degree of pulmonary hypertension was not documented in this case, the echocardiograph showed a prominent pulmonary artery, a small VSD, and right and left ventricular wall thicknesses at the upper limits of normal. Nevertheless, the physical examination did suggest considerable pulmonary hypertension. Apparently, the pulmonary hypertension was adequately compensated and the fluid load was well tolerated, a fortunate outcome.

One can speculate that epidural morphine contributed to the ventilatory insufficiency seen postoperatively. This patient showed inadequate ventilation preoperatively on the basis of an elevated PCO_2 . Using patient controlled epidural analgesia to titrate a dilute local anaesthetic mixture, PCEA, rather than giving a fixed dose of opioid may have been a better option. This however is only speculation since the local anaesthetic itself can promote muscle weakness. It should also be remembered that even a successful regional anaesthetic is no guarantee that respiratory failure will not ensue intra- or postoperatively. The decision to intubate this patient's trachea would be a clinical one, based on a variety of measures such as respiratory rate, dyspnoea, blood gases, haemodynamics, level of consciousness, etc.

The muscular dystrophies are a heterogeneous group of diseases whose genetic basis is subject to active investigation. This patient was given the diagnosis of Duchenne muscular dystrophy because two male second cousins were diagnosed with that disorder. In order for Duchenne muscular dystrophy, an X-linked recessive disorder, to present in a female the "non affected" X chromosomes must be deactivated (the Lyon Hypothesis). It is theoretically possible for patients with Turner's syndrome who have only one functioning X chromosome to inherit muscular dystrophy, but these individuals would not be fertile. An alternative diagnosis is severe, early onset limb-girdle muscular dystrophy. Finally, spinal muscular atrophies, another diverse group of disorders, could present with this degree of muscle weakness but without the cardiac manifestations. Later in the course of this patient's hospital stay it was determined that the most likely diagnosis was in fact a severe form of limb-girdle muscular dystrophy. Muscle

biopsy should not be forgotten as a useful diagnostic tool. This patient underwent muscle biopsy at age 12 yr, but the results were reported as ambiguous and not helpful in delineating the diagnosis.

If a general anaesthetic had been required for this patient, then the basic approach would have been to perform an awake intubation, to provide a trigger free anaesthetic, and to use opioids and neuromuscular blocking drugs judiciously because of the patients incipient respiratory failure. Careful weaning from mechanical ventilation in an intensive care unit setting is a safe course of action postoperatively.

In summary, anaesthesia for patients with severe muscular dystrophy can be accomplished through a careful review of the patients respiratory status, anticipation of cardiac dysfunction, and careful selection of anaesthetic agents based on the potentially increased risk of malignant hyperthermia and their residual effects on respiratory function. Since perioperative care can be complex in these patients, a well organized multidisciplinary approach may be advantageous.

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