Case Report

Caesarean section in a patient with torsion dystonia

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We present a case of torsion dystonia in a 35-yr-old primigravida who presented for a Caesarean section under general anaesthesia. She had limb contractures and severe kyphoscoliosis associated with limited respiratory reserve and function. General anaesthesia was induced using thiopental and divided doses of mivacurium for rapid sequence induction. After the delivery of a healthy male baby, she received i.v. morphine and bilateral iliohypogastric, ilioinguinal blocks and had an uneventful recovery. Technical issues of supine positioning, intubation and respiratory support need to be considered during anaesthesia planning. Although regional anaesthesia is commonly offered for caesarean section, maternal compromise and technical factors may preclude this approach.

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Pregnant women with rare neurological disease often challenge our normal methods of anaesthetic delivery. We would like to report a patient who presented with torsion dystonia (Dystonia musculorum deformans; Oppenheimer disease) for a Caesarean section.

Case history

A 35-yr-old primigravida with a history of torsion dystonia presented for elective Caesarean section at 36 weeks gestation. She was diagnosed with torsion dystonia at 7 yr of age when she presented with sudden loss of arm and leg control. She gradually developed torticollis and permanent flexion of her right arm. Despite bilateral stereotactic thalamotomy, anticonvulsants and clonazepam, she developed progressive dysphasia, severe kyphoscoliosis and muscular imbalance. Progressive bilateral extensor spasm in the lower limb restricted her mobility. The patient was of normal intelligence and worked in a bank.

Past medical history was also significant for mild asthma and occasional anginal symptoms. She was prescribed glyceryl trinitrate as required. Limited antenatal cardiology investigations were inconclusive. She recalled several days of postoperative severe generalized muscular pain after previous general anaesthesia for dental surgery. She had received succinylcholine. Her intubation was recorded as Cormack and Lehane grade 1. She also experienced difficulty in breathing immediately after surgery. She required no intensive care admission.

The woman was of small stature, 1.47 m tall, weighing 54 kg and was extremely anxious. She had marked kyphoscoliosis and suffered from involuntary muscle spasms of her arms and legs with associated restricted neck movements (Fig. 1). She was orthopnoeic with a ventilatory frequency of 28 bpm but did not suffer from paroxysmal nocturnal dyspnoea. Air entry was reduced bilaterally in the lung bases. There was no evidence of crepitations in her lungs or peripheral oedema. Jugular venous pressure was not raised. Heart sounds were normal. Lung function tests revealed a severely restrictive disease state. Forced expired volume in 1 s (FEV1) and forced vital capacity (FVC) were 0.5 and 0.5 litre (predicted 2.6 and 3.1 litre) respectively and her peak expiratory flow rate 130 litre min−1 (normal 160–240 litre). FEV1 and FVC 5 yr previously were 1.0 and 1.1 litre, respectively. Arterial blood gas analysis on 35% oxygen revealed Pao2 20 kPa, Paco2 1.87 kPa, pH 7.47, HCO3 16 mmol litre−1, and BE −11 mmol litre−1.
The patient developed increasing dyspnoea and severe upper quadrant abdominal pain requiring i.v. opioid analgesia. A decision to proceed to Caesarean section at 36 weeks gestation was therefore determined. She was very anxious and refused regional anaesthesia. Anaesthetic concerns included potential difficult intubation, dystonic movements and postoperative respiratory compromise requiring intensive care admission. Assessment of her airway revealed a Mallampati score grade 2. Metoclopramide, ranitidine and sodium citrate were prescribed for antacid prophylaxis.

The left radial artery was cannulated under local anaesthesia and venous access established via a 16 G i.v. cannulae. The table was not placed in lateral tilt as the woman’s severe thoraco-lumbar kyphoscoliosis conferred an adequate natural tilt. Rapid sequence induction was achieved using thiopental and mivacurium. The mivacurium was given in two divided doses of 0.15 and 0.1 mg kg\(^{-1}\) 30 s apart to facilitate rapid intubation, which was performed 60 s after the last dose. Laryngoscopy revealed a Cormack and Lehane laryngoscopy grade 1. Ventilation revealed a Cormack and Lehane laryngoscopy grade 1. Ventilation was maintained with 50% oxygen, nitrous oxide and isoflurane and ventilated to an end-tidal CO\(_2\) of 4.5 kPa. After the delivery of a healthy 2.3 kg male infant (Apgar score 6 and 9 at 1 and 5 min, respectively), oxytocin 5 units and morphine 8 mg was given i.v. A further 5 units of oxytocin was given at the request of the obstetrician.

Bilateral iliohypogastric and ilioinguinal (IHIL) nerve blocks with bupivacaine 0.5%, 15 ml and diclofenac suppository 100 mg were given for postoperative pain control. Blood loss was less than 500 ml. She was placed in an upright position for extubation and neuromuscular block antagonized with neostigmine 1.25 mg and glycopyrrolate 0.25 mg. The tracheal tube was removed after adequate tidal volumes and appropriate response to verbal command. Postoperative arterial blood sampling revealed a \(P_{aO_2}\) 33 kPa (\(F_{I_0}\) 0.4), \(P_{aCO_2}\) 5.9 kPa, pH 7.32, HCO\(_3\) 22.9 mmol l\(^{-1}\), BE −2.2 mmol l\(^{-1}\), and Hb 10.7 g dl\(^{-1}\). Patient controlled analgesia with morphine was prescribed with regular diclofenac retard 75 mg twice daily. She was transferred to a labour ward high dependency room after a very stable 4 h stay in the recovery room. Her baby was discharged from the neonatal unit after 48 h observation. Both mother and baby were discharged home 6 days post partum with no postoperative sequelae.

**Discussion**

This is the first reported case of torsion dystonia in pregnancy requiring anaesthesia for Caesarean section. Torsion dystonia is a neurological disease with a spectrum of symptoms and presentation characterized by sustained excessive muscular contractions aggravated by voluntary movement. It can be primary or secondary and both forms present either during childhood or at a later stage. The primary type usually has a genetic cause. DYT 1, DYT 6, DYT 7 and DYT 13 genes have been identified as the defective focal area.\(^1\) Mutation of the gene coding for ATP-binding protein has been demonstrated in the DYT 1 gene anomaly.\(^2\) The mode of transmission is autosomal dominant with reduced penetrance. The childhood onset form is more common among Jews from Eastern Europe and those of Ashkenazi descent. It presents in the second decade of life and characteristically affects the lower limbs before becoming more generalized.

Adult onset torsion dystonia usually affects the upper body with torticollis, kyphoscoliosis, blepharospasm but is rarely generalized. Although the adult form presents later in life than the childhood form, it may also have a genetic cause. Tardive dystonia, secondary to drugs such as metoclopramide, dopamine agonists, antipsychotics, anticonvulsants and calcium channel blockers, is the commonest cause of the secondary adult dystonic form.

Severe kyphoscoliosis presents several technical challenges to both surgical and anaesthetic teams. Changes in cardiac and respiratory physiological function result in maternal morbidity and mortality.\(^3\) A thorough investigation that identifies reversible treatable factors such as bronchospasm, excess secretions, should be instituted. General anaesthesia allows ventilatory control while avoiding the technical difficulties of regional anaesthesia placement. Regional anaesthesia however, avoids airway manipulation and offers excellent postoperative analgesia. Severe kypho-

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**Fig 1** This is the A–P view of the patient showing severe kyphoscoliosis and truncal deformity. It was taken before her pregnancy.

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scoliotic curves may cause pooling of hyperbaric local anaesthetic solutions which can be avoided using isobaric local anaesthetic solutions.\(^4\)

General anaesthesia has been previously reported in two non-pregnant female patients with torsion dystonia.\(^5\)\(^6\) Both patients had identical clinical presentation of their conditions. Both had general anaesthesia and uneventful postoperative courses. In particular, tracheal intubation had been straightforward. Our patient had significant respiratory compromise and was unlikely to have tolerated the prolonged supine positioning after regional anaesthesia. She was also understandably, extremely anxious and insisted on general anaesthesia. Her lung function tests and arterial blood gas profile were caused by her disease process. The severity of her acidotic condition was somewhat puzzling. This seemed to have corrected itself with fluid replacement. She required a high dependency unit admission for observation but no ventilatory support.

There are no reported adverse effects of succinylcholine in previous case reports. She had experienced significant postoperative myalgia lasting several days after succinylcholine administration and was extremely anxious to avoid this. Mivacurium seemed a suitable alternative to succinylcholine for rapid sequence induction.\(^7\)\(^8\) It has a quick onset and offset action with minimal residual effects. It was administered in two divided doses as described by Ali and colleagues\(^9\) to facilitate rapid intubation. In their study of 200 patients, all but one were easily intubated with good to excellent intubation conditions. This dosing regime also demonstrated better recovery profile compared with rocuronium with minimal effects on cardiovascular parameters.\(^7\)

The benefits of regional anaesthesia are evident but the unwilling anxious parturient with difficult anatomy and potential respiratory difficulties may preclude its use. These complex parturients with challenging profiles need anaesthesia referral and workup early in their pregnancy. ITNL nerve blocks for postoperative pain are easy to administer and contribute to postoperative pain control. Although the majority of elective Caesarean sections are now performed under regional techniques, general anaesthesia still has a role for the very anxious woman with challenging skeletal deformity associated with significant cardiorespiratory compromise.

References

5. Davis NL, Davis R. Anesthetic management of a patient with dystonia musculorum deformans. Anesthesiology 1975; 42: 630–1