Caesarean section for twins in a patient with myotonic dystrophy

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The management of a patient with myotonic dystrophy undergoing Caesarean section for delivery of twins in breech position is reported. Anaesthetic management must reflect the multi-system nature of the disease in addition to the implications of pregnancy. Known triggers of myotonic crisis (succinylcholine, shivering) must be avoided. Attention to respiratory reserve is necessary in view of the restrictive defect this disease can impose. Cardiomyopathy and conduction system disturbances may be features. Upper gastrointestinal incoordination can superimpose the added risk of aspiration on the pregnant myotonic. Ineffective uterine contraction can cause vigorous post partum haemorrhage. Endocrine disturbances, especially diabetes mellitus, may be present. There may be increased sensitivity to sedative and narcotic agents. The newborn myotonic will need the expertise of neonatal care.

Key words

ANAESTHESIA: obstetric; SURGERY: Caesarean Section; GENETIC FACTORS: myotonic dystrophy.

Myotonic dystrophy is a degenerative disease involving smooth, skeletal and cardiac muscle and is of autosomal dominant inheritance with variable penetrance. The clinical syndrome manifests in early adulthood and is characterized by myotonia; facial, neck and distal extremity muscle wasting and weakness, cataracts, frontal balding, testicular and ovarian atrophy, and progressive decline of intellect.¹

Pregnancy is uncommon in this condition,² a point underscored by the paucity of management details in the obstetric and anaesthetic literature. The specific problems are: pulmonary aspiration, ventilatory failure, congestive heart failure, complete heart block and uncontrollable uterine haemorrhage.

Recently at our institution, we had a unique occurrence of a Caesarean delivery of monozygotic twin boys in breech presentation from a known myotonic patient. We felt our management of this unusual clinical problem warranted reporting as, to our knowledge, a similar situation has not been described in the literature.

The patient was a 29-year-old gravida two para one term female diagnosed by ultrasound as having twin breech fetuses. Six years earlier a normal pregnancy culminated in sixty hours of labour. Vaginal delivery was complicated by brisk post partum bleeding requiring transfusion.

The diagnosis of myotonic dystrophy was established by clinical demonstration of myotonia, i.e., the typical "myotonic handshake." Other symptoms were muscle spasms, lethargy, and chronic menstrual irregularities. Clinical evaluation of the family revealed a nephew with classic involvement.

Except for longstanding obesity the system review was non-contributory.

On preoperative examination, the patient was obese and of normal intelligence. Her weight was 103 kg, height 165 cms, blood pressure 140/90 torr, pulse 75/min, and respiration rate 18/min. Significant findings were mild facial weakness, minimal bilateral ptosis, moderate extremity weakness distally with mild weakness proximally. Percussion and effort-induced myotonia was present in both hands.

Preoperative haemoglobin was 128 g·L⁻¹, creatinine kinase 3078 U·L⁻¹ (normal 25-145

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TABLE I Arterial blood gases

	Preoperative	Intraoperative		Postoperative 100 min
		(time 1)	(time 2)	
pH	7.40	7.42	7.38	7.40
PCO ₂ (torr)	39	35	41	38
PO ₂ (torr)	67	216	194	67
HCO ₃ (meq·L ⁻¹)	24	23	25	23
O ₂ Sat (%)	93	100	99	93
FiO ₂	0.21	0.4	0.4	0.21

U·L⁻¹), with normal serum electrolytes. A poor inspiratory effort was noted on an otherwise normal chest x-ray. The electrocardiogram was also normal. The preoperative arterial blood gas results are shown in Table I and bedside pulmonary flow rates in Table II.

Cimetidine, 300 mg p.o., was administered eight hours and one hour before surgery. In the operating room a 16 gauge venous cannula was inserted in the left arm using one per cent lidocaine for skin anaesthesia. Ringer's lactate, 900 ml, was infused through a blood/fluid warming set. With the patient in the left lateral position an epidural catheter was inserted at the L4-5 interspace. She was then placed supine with a right hip wedge. An arterial line was inserted in the left radial artery; ECG electrodes, blood pressure cuff and axillary temperature probe were applied. An urinary catheter had previously been inserted. The room temperature was 25°C. Oxygen via face mask was administered (FiO₂ approx. 0.4).

A 2 ml epidural test dose of carbonated lidocaine was injected uneventfully, followed by a further 4 ml. Additionally, 12 ml of 0.5 per cent bupivacaine was given to render a T6 dermatomal level sensory block. Vital signs remained stable throughout the procedure. Relaxation was excellent and the patient was comfortable. Extraction of the babies

from the uterus was uneventful. Apgar scores of eight at one minute and nine at five minutes were observed. Muscle tone abnormalities were not evident as assessed by the neonatologist.

The uterus contracted adequately with intravenous infusion of five units of syntocinon followed by 1.5 units per hour. Fentanyl 50 µg was given in divided doses and additional local anaesthetic was given epidurally to relieve peritoneal closure discomfort. No respiratory difficulties ensued. Arterial blood gases were taken on two occasions (see Table I).

Immediately prior to transfer to the stretcher, two additional doses of fentanyl (25 µg) were administered. Two to three minutes following the last dose masseter spasm occurred, lasting about four minutes. A few moments later an episode of vigorous shivering arose lasting four minutes. A myotonic crisis did not follow.

Immediately after arrival in the recovery room an increased flow of blood was noted vaginally. Approximately 200 ml of blood was lost before massage and increased oxytocin became effective. A repeat creatinine kinase was 3156 U·L⁻¹ and an arterial blood gas taken two hours postoperatively is recorded in Table I. The remainder of the hospital course was uneventful.

TABLE II Spirometry results

	Preoperative	Postoperative (6 days)	Predicted	
			Pregnant	Non-pregnant
FEV ₁ (litres)	3.09	2.68	2.45	2.93
FVC (litres)	3.33	2.79	2.59	3.79
FEV ₁ /FVC	0.93	0.96		

Discussion

In managing any patient with myotonic dystrophy recognition of the multi-system nature of this disease is essential.

The major concern is induction of generalized sustained muscle contracture: the so-called "myotonic crisis." Ventilation, both spontaneous and controlled, can be severely compromised.³ As well, surgical exposure can be restricted especially during intra-abdominal procedures. The muscle contracture is not relieved by muscle relaxants or neural blockade. Direct injection of the muscle with local anaesthetic agents is reportedly effective.⁴ Systemic agents such as procainamide, quinine, and dantrolene may be effective. A role for calcium entry blockers has not been clarified.

All attempts should be made to avoid a myotonic crisis. Succinylcholine is the classic offender⁵ and is contraindicated. It is reputed to cause excessive potassium release in these individuals. Conditions that preclude shivering should be present; the operating suite should be warmed above normal room temperature and parenteral fluid heated to body temperature. Malignant hyperthermia has been reported in the presence of myotonic dystrophy with an unknown incidence,³ although this association has been questioned.⁵ Body temperature must be closely monitored.

Respiratory failure is a leading cause of morbidity perioperatively. Clinical assessment of respiratory reserve preoperatively is necessary. Pulmonary function testing and arterial blood gas results provide the data with which to compare thereafter. Obvious wasting of accessory muscles of respiration, especially the sterno mastoid,¹⁰ should warn of the potential for hypoventilation. Injudicious use of sedatives and narcotics perioperatively can seriously depress respiratory drive. The myotonic patient is extremely sensitive to these agents. Regional anaesthesia is preferable.⁷ If general anaesthesia is selected, readily excreted agents (e.g., volatile) would allow a rapid recovery post-operatively. Thiopentone may cause prolonged apnoea.⁴ Ventilatory adequacy should be evaluated prior to extubation. Arterial blood gas analysis facilitates assessment of on-going ventilatory changes.

Skeletal, smooth, and cardiac muscle may be involved in myotonic dystrophy.⁸ Cardiomyopathy

is a feature in a significant percentage of myotonics. If there is clinical evidence of myocardial involvement, then a pulmonary artery catheter is indicated and pharmacologic agents for haemodynamic manipulation should be available. A variety of conduction defects have been reported, most commonly first degree heart block.⁹

Decreased tone in the lower oesophageal sphincter and incoordinate action of the upper oesophageal sphincter¹⁰ increase the risk of aspiration in supine anaesthetized and awake patients. For general anaesthesia, methods to protect the patient's airway and raise gastric pH should be employed.

Diabetes mellitus commonly co-exists with myotonic dystrophy. Hypometabolism of non-thyroidal origin has also been observed.⁵

The myotonic parturient may be in particular jeopardy for several reasons. Hypoventilation is aggravated by the prenatal reduction in functional residual capacity. Many myotonics experience muscle weakness solely during pregnancy, only to abate post partum. The risk of aspiration is even greater because of upper and lower oesophageal sphincter weakness and incoordination superimposed on the progesterone and mechanical effects of pregnancy in the upper gastrointestinal tract. Blood volume changes associated with pregnancy and delivery may decompensate cardiomyopathy to the point of heart failure. Uterine contraction may be ineffective. A hypotonic uterus may bleed vigorously following apparently good initial contraction. A neonatologist should be involved with the delivery to assess and manage potential newborn hypotonia and hypoventilation.

Our patient presented with an uncomplicated myotonic dystrophy with findings referable to the musculoskeletal system. Potential complicating factors were: the possibility of hypoventilation during regional anaesthesia secondary to her obesity and the large uterine mass, the increased risk of postpartum haemorrhage from lack of uterine contraction and lastly the spectre of myotonic crisis. The one additional concern in this unusual case was the muscular status of the twin neonates.

Because of our awareness of potential difficulties, the vaginal bleeding was anticipated and managed early and known triggers were avoided. The fact that low doses of fentanyl were followed by

masseter spasm is interesting and we question if fentanyl should be included in the list of myotonic crisis triggers.

Myotonic dystrophy in pregnancy and certainly in multiple pregnancy is uncommon. Awareness of the many problems and readiness of therapeutic measures should lead to a successful outcome.

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Résumé

Les auteurs décrivent la conduite anesthésique chez une patiente souffrant de dystrophie musculaire, et qui, au terme d'une grossesse gémellaire avec fœtus en position de siège, doit subir une Césarienne. L'anesthésiste doit considérer la pluralité des systèmes impliqués en plus des problèmes associés à la grossesse. Les facteurs pouvant déclencher une crise myotonique, tels que les frissons ou la succinylcholine, doivent être évités. Une surveillance étroite de la réserve respiratoire est impérative. Une cardiomyopathie ou des troubles de conduction cardiaque peuvent être présents. Des troubles de motilité du tratus digestif supérieur augmentent le risque d'aspiration. L'atonie utérine est fréquente après la délivrance chez la patiente myotonique et peut être suivie d'une hémorragie importante. Des troubles endocriniens, principalement le diabète, peuvent s'ajouter au tableau clinique. Il peut aussi y avoir une sensibilité accrue aux narcotiques et aux sédatifs. Le nouveau-né, parfois myotonique, peut avoir besoin de l'expertise du néonatalogiste.