Anaesthesia for parturients with severe cystic fibrosis: a case series

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ABSTRACT
Cystic fibrosis affects 1 in 1600–2500 live births and is inherited in an autosomal recessive manner. It primarily involves the respiratory, gastrointestinal and reproductive tracts, with impaired clearance of, and obstruction by, increasingly viscous secretions. Severe respiratory disease, diabetes and gastro-oesophageal reflux may result. Improvements in medical management and survival of cystic fibrosis patients means more are committing to pregnancies. Although guidance for anaesthesia in this patient group is available, management and outcome data associated with more severe cases are sparse. Patients with severe cystic fibrosis require multidisciplinary input and should be managed in a tertiary referral centre. Close monitoring of respiratory function and preoperative optimisation during pregnancy are mandatory. The risk of preterm labour and delivery is increased. Pregnancy and delivery can be managed successfully, even in patients with FEV1 <40% predicted. Neuraxial anaesthesia and analgesia should be the technique of choice for delivery. Postoperative care should be carried out in a critical care setting with the provision of postoperative ventilation if necessary.

Keywords: Cystic fibrosis; Caesarean section; Neuraxial anaesthesia

Introduction

Cystic fibrosis (CF) affects 1:1600 to 1:2500 live births and results from mutations of the CF transmembrane conductance regulator protein on the long arm of chromosome 7. It is inherited in an autosomal recessive manner with a gene frequency of 1:22. Despite impaired longevity, the fertility of female CF sufferers is unaffected, and many have completed successful pregnancies. Although guidance for anaesthesia in this patient group is available, management and outcome data associated with more severe cases are sparse. We present a series of four CF patients with unplanned pregnancies who had very poor respiratory function, FEV1 <40% of predicted, and who all suffered regular infective exacerbations. Pre-pregnancy counselling, which should be given where possible to all CF females of childbearing age, was not possible. The patients were committed to their pregnancies, declined termination and were delivered by elective caesarean section. Multidisciplinary management included their CF team (CF physician, CF specialist nurse and physiotherapist), obstetric and anaesthetic review every four weeks initially. Three of the four patients required hospitalisation from 24, 28 and 30 weeks of gestation until their delivery. We present the cases and discuss their anaesthetic management during delivery and in the immediate postoperative period.

Accepted October 2013
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Case histories

In all cases preoperative optimisation included increasingly intense daily physiotherapy. Nebulised drugs were given immediately before caesarean delivery. Two patients received prophylactic doses of low-molecular-weight heparin (LMWH) which was stopped 12 h before surgery. Perioperative blood sugar concentrations were managed according to a standardised sliding-scale of insulin. After establishing peripheral intravenous access, oxygen saturation (SpO2), electrocardiogram (ECG) and direct arterial blood pressure monitoring, neuraxial anaesthesia was performed, before placing the patient in a semi-sitting position with left lateral tilt for surgery. All patients received a 500 mL crystalloid co-load while neuraxial anaesthesia was sited, which was continued at 100 mL/h. Intravenous co-amoxiclav was given before surgical incision. All four patients received intravenous oxytocin 5 U given over 5 min after delivery of baby, followed by an infusion of 40 U in 0.9% saline 500 mL at 62.5 mL/h. Estimated blood loss in all cases was < 400 mL.

Case 1

A 28-year-old nulliparous woman with a body mass index (BMI) of 26 kg/m^2 presented at 24 weeks of gestation after an incidental positive pregnancy test. Her FEV1 was 0.79 L (24% predicted) and co-morbidities included pancreatic insufficiency, impaired glucose tolerance, chronic colonisation with methicillin-resistant Staphylococcus aureus (MRSA) and *Pseudomonas aeruginosa*, and osteopenia. She required long term oxygen therapy,

nocturnal bi-level positive airway pressure (BiPAP) and had central venous access (Port-A-Cath®) and a percutaneous endoscopic gastrostomy (PEG tube) in situ. Due to poor glucose control at 30 weeks of gestation, she remained in hospital until caesarean delivery at 34 weeks, at which point her FEV1 had fallen to 18% of predicted.

Epidural anaesthesia was used for caesarean section. An 18-gauge epidural catheter was sited at L3-4, and 2% lidocaine with 1:200 000 adrenaline 10 mL was given to achieve a T6 level of block to cold. During surgery the patient complained of pain and a further 5-mL increment was given to achieve a T2 sensory level. Oxygen was administered via nasal prongs at 4 L/min. Respiratory compromise during the case required non-invasive assistance with BiPAP. Two 1 mg boluses of midazolam were given for anxiolysis after delivery of a healthy 2.2 kg baby boy who had Apgar scores of 9 at 1- and 5 min (See Table 1).

Case 2

A 36-year-old nulliparous woman with a BMI of 21 kg/m^2 and FEV1 of 0.85 L (32% predicted) presented in early pregnancy. Her co-morbidities included *Pseudomonas aeruginosa* and multiply resistant *Stenotrophomonas maltiphilia* colonisation, pancreatic insufficiency, insulin dependent diabetes mellitus (IDDM) and nocturnal PEG tube feeding. A left subclavian Port-A-Cath® was in situ. She required lifelong anticoagulation due to superior vena cava thrombosis with permanent stenosis, which also made her ineligible for lung transplantation. Continuous oxygen therapy was commenced at 17 weeks of gestation. She remained in hospital from 24 weeks due to infective exacerbations and became dyspnoeic at rest. She was delivered at 32 weeks due to low liquor volume.

Anaesthesia was performed with a needle-through-needle, combined spinal-epidural technique. A 25-gauge spinal needle was inserted via an 18-gauge Tuohy needle at L3-4. An intrathecal injection of 0.5% hyperbaric bupivacaine 1.8 mL was followed by a further 10 mL of 2% lidocaine with 1 in 200 000 adrenaline, in 5 mL increments given through the epidural catheter, to achieve a T6 block to cold. Oxygen was administered via nasal prongs at 4 L/min. Intravenous ketamine 10 and 5 mg and fentanyl 25 and 25 μg were given for inadequate analgesia after delivery of a healthy 1.49 kg baby girl. Apgar scores were 7 and 9 at 1- and 5 min, respectively. Intraoperative haemodynamic stability was achieved using a phenylephrine infusion (total dose 500 μg) (See Table 2).

Case 3

A 39-year-old nulliparous woman with a BMI of 20 kg/m^2 and an FEV1 of 1.05 L (36% predicted) presented in early pregnancy. She had chronic colonisation with *Pseudomonas aeruginosa*, multi-resistant *Staphylococcus aureus* and *Burkholderia cepacia*. Other co-morbidities included NIDDM and osteoporosis. She had peripherally-inserted central catheter (PICC)-related thromboses in both arms and required lifelong anticoagulation. She had one previous intensive care unit (ICU) admission for BiPAP ventilation. At 24 weeks of gestation, nasogastric nocturnal feeding was initiated due to her static weight. She progressed to 37 weeks of gestation and was admitted only two days before her planned delivery.

For caesarean section a spinal catheter was inserted at L3-4 and an epidural catheter at L2-3. After initially good flow from the spinal catheter, a sensory block to cold of T6 was achieved with 0.5% hyperbaric bupivacaine 3 mL, given in 0.5 mL increments. Oxygen was administered via nasal prongs at 4 L/min. Haemodynamic stability was maintained without the need for vasopressors. The patient complained of pain during surgery, and poor flow from the spinal catheter necessitated an epidural top-up using 2% lidocaine with 1:200 000 adrenaline 5 mL. A healthy 2.7 kg baby girl was delivered with Apgar scores of 9 at 1- and 5 min (See Table 3).

Case 4

A 20-year-old nulliparous woman with a BMI of 17 kg/m^2 and an FEV1 of 1.06 L (36% predicted) presented at eight weeks of gestation. She had chronic colonisation with *Pseudomonas aeruginosa*, pregnancy-induced IDDM and a right subclavian Port-A-Cath® had been inserted 11 years previously. The course of her CF, including during pregnancy, was typified by frequent infective exacerbations. She had no previous ICU admissions and had never required non-invasive ventilation. She was confined to hospital from 28 weeks of gestation due to an infective exacerbation with dete-

Table 1 Case 1: lung function tests and arterial blood gases

	Pre-pregnancy	24–26 weeks	27–29 weeks	30–34 weeks	Postop day 2
Weight (kg)	61	65	70	69	65
FEV1 (L) (% predicted)	0.94 (29%)	0.78 (24%)	0.71 (21%)	0.6 (18%)	0.62 (19%)
FVC (L) (% predicted)	NA	2.21 (59%)	1.67 (44%)	1.6 (42%)	1.55 (40%)
FiO_2 (mmHg)	NA	0.28	NA	0.28	0.34
PO_2 (mmHg)	NA	11.3	NA	9.47	8.51
PCO_2 (mmHg)	NA	5.01	NA	5.87	8.15

NA, data not recorded in clinical notes.