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CASE REPORT

Caesarean section in a patient with cystic fibrosis

M. Muammar, P. Marshall, H. Wyatt, V. Skelton

Department of Anaesthesia, King's College Hospital, Denmark Hill, London, UK

SUMMARY. Severely impaired pulmonary function due to cystic fibrosis is associated with a poor pregnancy outcome. A 19-year-old primigravida with cystic fibrosis had a forced expiratory volume in one second 34% of predicted and a low body mass index of 17.1 kg/m^2 before pregnancy. During pregnancy she required several admissions to hospital for deteriorating pulmonary function due to infection. She also developed gestational diabetes and required overnight feeding via a gastrostomy tube throughout pregnancy to improve her nutritional status. With close medical attention, pulmonary function tests remained stable. A joint decision was made to deliver the baby by caesarean section at 33 weeks' gestation as this was considered optimal time for mother and baby and to avoid potential catastrophic pulmonary deterioration later in pregnancy. A combined spinal-epidural technique provided adequate anaesthesia. Pulmonary function tests were performed in theatre before and after the spinal injection, as well as two and five hours postoperatively. Despite a spinal anaesthetic block to T5 bilaterally, lung function tests remained stable. An epidural infusion provided good postoperative analgesia. The outcome was successful for mother and fetus.

Keywords: *Anaesthesia*, obstetric; *Anaesthetic technique regional*, spinal,epidural,CSE; *Complications*, cystic fibrosis

INTRODUCTION

Cystic fibrosis (CF) is an autosomal recessive disorder that affects 1 in 2500 live births. Both sexes are equally affected and 1 in 25 Caucasians are carriers. The CF gene, located on the long arm of chromosome 7, codes for the CF transmembrane conductance regulator protein. The result is abnormal chloride and sodium ion transport across epithelial cell membranes. It is a multisystem disease mainly affecting the respiratory, gastrointestinal and reproductive tracts, due to impaired clearance and obstruction by viscous secretions. [1] Severe respiratory disease, diabetes and gastroesophageal reflux are the common features of concern to anaesthetists. Improved care of young patients has allowed many to survive into adulthood and therefore present for childbirth.

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Dr M. Muammar, Specialist Registrar in Anaesthesia, **Dr P. Marshall**, Specialist Registrar in Anaesthesia, **Dr H. Wyatt**, Consultant in Cystic Fibrosis, **Dr V. Skelton**, Consultant Anaesthetist; Department of Anaesthesia, King's College Hospital, Denmark Hill, London, UK.

Correspondence to: Dr. M. Muammar, Specialist Registrar in Anaesthesia, Department of Anaesthesia, King's College Hospital, Denmark Hill, London, UK, Tel.: +0044 207 346 3154; fax: +0044 207 346 4106; E-mail: muna@doctors.org.uk.

Premature labour and delivery remain a significant risk for pregnant women with CF. Maternal morbidity and mortality result from deteriorating pulmonary function. [2] We describe a patient with CF with poor respiratory function (less than 50% of predicted) who presented for caesarean section. Pulmonary function tests were performed repeatedly before and after regional anaesthesia.

CASE HISTORY

A 19-year-old primigravida with cystic fibrosis presented for elective caesarean delivery at 33 weeks' gestation. Before becoming pregnant, she suffered recurrent chest infections requiring two to four courses of intravenous antibiotics per year and her sputum was chronically colonised with *Pseudomonas aeruginosa*. Before conceiving she weighed 45.6 kg (body mass index 17.1 kg/m^2) and nocturnal gastrostomy feeding had been coincidentally started one month earlier. Before pregnancy, like many adolescents with chronic illness, she had failed to attend many outpatients' appointments, was reluctant to increase her calorie intake, did not use her inhalers regularly and received chest physiotherapy sporadically. She was counselled at 11 weeks' gestation regarding the risk of serious deterioration of her respiratory function during pregnancy, but decided to

continue with the pregnancy and made great efforts to improve her compliance with treatment. During pregnancy she required several admissions to hospital. At 12 weeks' gestation she required a two-week course of intravenous ceftazidime, which was given at home, for symptoms of respiratory tract infection.

At 17 weeks' gestation, she was admitted with deteriorating respiratory function (forced expiratory volume in one second [FEV_1] 1.1 L, 34% of predicted), shortness of breath and weight loss. She was treated with intravenous aztreonam and tobramycin, intensive physiotherapy, and overnight feeding. On discharge 15 days later, her lung function had improved (FEV_1: 1.5 L, 47% of predicted) and weight increased by 4 kg.

At 23 weeks' gestation, respiratory function deteriorated again and she was hyperglycaemic for the first time. She was admitted for three days to receive insulin and a two-week course of intravenous ceftazidime. Sputum cultures grew *Staphylococcus aureus* and *Pseudomonas aeruginosa*. One month later she was admitted again for two weeks with a chest infection and treated with intravenous tobramycin and aztreonam.

At 29 weeks' gestation, she was admitted for administration of dexamethasone for fetal lung maturation. She was started on an insulin sliding scale. A routine echocardiogram was performed at this stage which showed no evidence of pulmonary hypertension. Table 1 shows lung function tests performed before and during pregnancy.

Caesarean delivery was scheduled for 33 weeks' gestation, not only to avoid potential catastrophic deterioration later in the pregnancy but also because fetal growth began to slow down and the mother started to lose weight. She was admitted three days before surgery weighing 58 kg. Her gestational diabetes was well controlled but she had a productive cough. Intravenous ceftazidime was started, as well as colistin nebuliser and oral flucloxacillin. Before surgery she received a Pulmozyme nebuliser followed by chest physiotherapy. She agreed to regional anaesthesia for delivery.

Monitoring consisted of pulse oximetry, electrocardiogram (ECG) and direct intra-arterial blood pressure measurement. A 16-gauge peripheral cannula was sited intravenously and a 500-mL preload of Hartmann's solution was given. A combined spinal-epidural was performed in the sitting position. The epidural catheter was sited at L3/4 and a 25-gauge spinal needle was used to inject 0.5% hyperbaric bupivacaine 2.4 mL with diamorphine 200 μg. A bilateral T5 level of block to cold (tested with ethyl chloride spray) was achieved within 15 min. Oxygen was administered via a Hudson mask at 4 L/min and oxygen saturations were maintained above 95% throughout the procedure. Non-invasive positive pressure ventilation had been demonstrated to the patient pre-operatively and was available in theatre during the delivery but not required. Her systolic pressure was stable initially at 120-140 mmHg but, following delivery of the baby and i.v. administration of oxytocin 5 units given over 5 min, it dropped to 90 mmHg but responded to ephedrine. Pulmonary function tests were performed in theatre before and after the spinal injection (Table 2). Three readings were taken each time in a supine position and the best performance was recorded. Arterial blood gases taken before, during and after surgery are shown in Table 3. A healthy baby boy weighing 2.53 kg was delivered with Apgar scores of 9 at 1 and 5 min; he did not require respiratory support.

An epidural infusion of bupivacaine 0.25% with diamorphine 0.1 mg/mL at 3 mL/h was continued for three days postoperatively and provided good analgesia, allowing her to cough well and clear sputum effectively. She made an uneventful recovery and her diabetes completely resolved after delivery.

One year later she was coping well with her baby and her respiratory function had remained stable (FEV_1 = 1.3 L, FVC = 2 L, PEFR = 255 L/min).

DISCUSSION

Cystic fibrosis is a multi-system disease, but pulmonary complications play a major role in morbidity and mortality during pregnancy. In CF lower respiratory tract infection caused by *Staphylococcus aureus*, *Haemophilus influenzae* and *Pseudomonas aeruginosa* occurs early in life and is difficult to eradicate. The airway is predisposed to infection possibly due to a reduced volume of surface liquid, as a result of sodium and water

Table 1. Lung function tests during pregnancy

			Gestation (weeks)					
	Predicted	Pre-pregnancy	17	19	23	26	28	33
FEV_1 (L)	3.2	1.1	1.1	1.5	1.6	1.0	1.4	1.6
FVC (L)	3.7	1.6	1.6	2.2	2.2	1.4	2.0	2.0
PEFR (L/min)	426	240	255	270	240	245	250	280
FEV_1/FVC (%)	86	69	69	69	73	73	69	79

FEV_1 = forced expiratory volume in 1 s; FCV = forced vital capacity; PEFR = peak expiratory flow rate.