CASE REPORT

Anaesthetic management of vaginal delivery in a woman with Friedreich's ataxia complicated by cardiomyopathy and scoliosis

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SUMMARY. We report the anaesthetic management of vaginal delivery in a woman with Friedreich's ataxia, who had hypertrophic cardiomyopathy and had previously undergone thoracic spinal fusion with Harrington rod fixation. Combined spinal-epidural analgesia was used. Options for the anaesthetic management of labour and delivery are discussed.

INTRODUCTION

Friedreich's ataxia is an autosomal recessive inherited disorder occurring equally in males and females; sporadic cases occasionally occur. The disease is progressive with degeneration of the spinocerebellar and corticospinal tracts and concomitant dorsal root ganglion atrophy. It is, therefore, a mixed upper and lower motor-neurone disease with a predominance of cerebellar symptoms. Tendon jerks are lost at an early stage, muscle tone is decreased and the plantar reflexes are extensor. Scoliosis and pes cavus are almost invariable, and it is associated with hypertrophic cardiomyopathy. The disease is progressive and few patients survive more than 20 years after the onset of symptoms. Death is commonly due to heart disease either as a result of arrhythmia or from congestive cardiac failure.

We report the anaesthetic management of vaginal delivery in a woman with Friedreich's ataxia, who had previously had a fusion of her thoracic spine as treatment for scoliosis, and was known to have hypertrophic cardiomyopathy.

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CASE REPORT

A 23-year-old woman presented at 17 weeks gestation for antenatal care at the high risk clinic, which has multidisciplinary medical input. Friedreich's ataxia had been diagnosed at 10 years of age, when she presented with lethargy, poor exercise tolerance and clumsiness. Clinical examination at that time revealed a heart murmur and cerebellar signs. No family history of Friedreich's ataxia could be confirmed as she was adopted. Echocardiography and cardiac catheterization confirmed the presence of hypertrophic non-obstructive cardiomyopathy. She received general anaesthesia using thiopentone, pancuronium and halothane for her catheterization, which was without incident. It was also noted at the time of diagnosis that she had bilateral pes cavus and a thoracic scoliosis.

At the age of 18 she was free from cardiovascular symptoms, but her neurological condition had deteriorated so that she was practically wheelchair-bound. She was also suffering from increasing pain associated with thoracic scoliosis and, therefore, underwent thoracic spinal fusion with Harrington rod fixation. Surgery was conducted under general anaesthesia using thiopentone, vecuronium and isoflurane. She made an uneventful recovery from the procedure and within a year had resumed swimming and become Regional Disabled Champion.

At 21 years of age she developed angina. Symptom control was achieved using isosorbide mononitrate, aspirin, verapamil and glyceryl trinitrate spray. Her

echocardiogram showed generalized muscle hyper-trophy with no left ventricular outflow tract obstruc-tion and her left ventricular function remained good.

Two years later she presented at 17 weeks gestation in her first pregnancy. She had no cardiovascular or respiratory symptoms. Her echocardiogram remained unchanged and she was, therefore, advised to discon-tinue her medication due to uncertainty about the safety of verapamil in pregnancy. She received genetic advice concerning the possibility of her child inherit-ing Friedreich's ataxia and decided to continue with her pregnancy.

After careful consideration of the neurological and cardiovascular implications of both vaginal and opera-tive delivery, it was decided that she should be delivered by elective caesarean section using combined spinal-epidural anaesthesia. A 48-h admission for monitor-ing, assessment and review was planned between 32 and 34 weeks gestation. At this admission she was reviewed by the obstetricians, obstetric anaesthetist, cardiologists and neurologists. Her condition remained stable and delivery plans were confirmed.

At 35 weeks gestation the patient presented in early labour and wished to attempt vaginal delivery. After discussion between the patient, her partner and the obstetricians, it was decided to allow labour to continue and provide regional analgesia to reduce the associated cardiovascular stress. At 5 cm cervical dilatation com-bined spinal-epidural analgesia was planned and an intravenous infusion of normal saline was established. A needle-through-needle spinal-epidural was inserted using a 16 gauge Tuohy needle and a 25 gauge Whitacre needle. This proved technically difficult and was attempted at several lumbar spaces before successful placement at the L5/S1 interspace. Twenty-five micro-grams of fentanyl and 2.5 mg bupivacaine were injected intrathecally and complete pain relief was obtained within 5 min. The blood pressure was measured non-invasively every 5 min for 30 min and then at 15-min intervals. She remained haemodynamically stable.

Two hours later the patient required further anal-gesia. An epidural top up of 10 ml of 0.125% bupi-vacaine and 25 µg of fentanyl was given, but did not reduce her pain. Her blood pressure remained stable so a further epidural top up of 10 ml of 0.25% bupi-vacaine was administered. Unfortunately, pain relief was still incomplete, but as vaginal examination confirmed full dilatation of the cervix, the baby was delivered by ventouse extraction. Apgar scores were 8 at 1 min and 10 at 5 min.

After delivery the patient remained well with no cardiovascular or respiratory symptoms. Because breast-feeding had not been established successfully, on the 10th postpartum day she and her baby were transferred to her local district general hospital.

DISCUSSION

Friedreich's ataxia is a serious disease associated with significant cardiopulmonary and musculoskeletal complications. The physiological changes of preg-nancy may aggravate these problems, in particular:

- Increased ligamentous and joint laxity together with increasing uterine size may exacerbate the pain and weakness.
- Decreased functional residual capacity, upwards displacement of the diaphragm, splinting of the chest wall, and closing volume encroaching on tidal volume will all increase respiratory embarrassment.
- Generalized reduction in systemic vascular resistance and increases in cardiac output and blood volume will adversely affect myocardial function in a patient with a cardiomyopathy.

Despite these potential problems, previous reports suggest that, in general, pregnancy is well tolerated in patients with Friedreich's ataxia.[1]

Although there have been no reported cases of con-genital abnormalities resulting from the use of vera-pamil in the first trimester, fetal congenital hypertrophic cardiomyopathy is reported with the use of intravenous verapamil in the third trimester.[2] In view of this and the stable cardiovascular status of our patient, verapamil was discontinued.

Delivery by caesarean section may be planned either for obstetric or medical indications. The obstetricians considered that our patient's stature and musculo-skeletal abnormalities reduced her chances of successful vaginal delivery. The medical indications in our patient were controversial. Elective caesarean section avoids the potential cardiovascular stress associated with labour, but introduces the risks of surgery and anaes-thesia, in particular the risk of thromboembolic disease. Inadequately treated postoperative pain may also aggravate any pre-existing impairment of respira-tory function. Delivery by elective caesarean section minimizes the possibility of emergency caesarean sec-tion, which is associated with significantly increased morbidity and mortality. Emergency caesarean section also introduces the potential risks of either rapid onset spinal anaesthesia or rapid sequence induction of general anaesthesia.

Choice of anaesthetic technique for operative delivery is directed towards maintaining a stable haemodynamic profile, avoiding tachycardia and preserving normal preload and afterload.[3] General anaesthesia poses theoretical risks to patients with Friedreich's ataxia. Some of these risks are said to be the same as those for patients who have amyotrophic lateral sclerosis (ALS).[4,5] Patients with ALS are

vulnerable to hyperkalaemia following the administration of suxamethonium and therefore, suxamethonium should be avoided. Patients with ALS are also reported to have an increased sensitivity to non-depolarizing muscle relaxants,[4] but this has not been shown to be the case in Friedreich's ataxia.[5,6]

The cardiopulmonary complications of Friedreich's ataxia may also be adversely affected by the administration of general anaesthesia. Anaesthetic agents used routinely in obstetric anaesthesia result in decreased cardiac contractility, alterations in preload and afterload, and may predispose to dysrhythmias in a patient with cardiomyopathy. Intermittent positive pressure ventilation employed during anaesthesia is also likely to have deleterious effects on respiratory function. Provision of adequate analgesia after general anaesthesia is of paramount importance in minimizing cardiovascular and respiratory disturbance. Although the potential risks associated with the use of general anaesthesia may be theoretical, we decided that it would be wise to avoid it and therefore advised our patient to choose regional anaesthesia.

Regional anaesthesia using an incremental epidural technique is associated with less haemodynamic instability than subarachnoid anaesthesia. However, healed spinal fusion with Harrington rods poses technical problems, as the usual tactile landmarks are obliterated and the bone graft may impede entry to the epidural space. Fusion for scoliosis usually ends above L4 and the L5/S1 intervertebral space may provide a route of access to the epidural space. Nonetheless, epidural insertion may still be technically challenging, as was the case with our patient.[7] Even with successful placement of an epidural catheter, previous spinal surgery predisposes to poor spread within the epidural space and an inadequate block.[8] Daley et al report 21 attempts at continuous epidural anaesthesia for obstetrics after major spinal surgery in which satisfactory analgesia was achieved in 15 cases.[9] They found that a larger dose of local anaesthetic was necessary to provide analgesia in four of these cases, but there were no significant short- or long-term complications.

Kubal et al report the successful use of spinal anaesthesia for caesarean section in a patient with Friedreich's ataxia with no evidence of disease exacerbation.[10] In a patient with cardiomyopathy, a single-shot spinal may result in undesirable and dramatic changes in preload and afterload. Theoretically an incremental intrathecal technique using a spinal catheter provides excellent anaesthesia or analgesia with relative haemodynamic stability, and enables extension of the block if necessary. However, the insertion of microspinal catheters is not without

problems and is not a technique with which we are familiar. We, therefore, felt that a combined spinal-epidural technique would be most appropriate, as it provides both the haemodynamic advantage of low-dose spinal anaesthesia and the flexibility for extending the block with an epidural catheter in place.

A greater variety of regional techniques, providing a more flexible approach, has led to planned vaginal delivery becoming a reasonable alternative to delivery by elective caesarean section. The use of incremental epidural, combined spinal-epidural and spinal catheter techniques, together with invasive arterial and central venous pressure monitoring facilitates provision of good analgesia with haemodynamic stability. The use of epidural and spinal opioids has allowed smaller doses of local anaesthetic agents to be given, reducing the incidence and severity of side-effects without compromizing efficacy. Vaginal delivery still has the disadvantage that, if unsuccessful, it exposes the patient to the risks of emergency caesarean section. Patients who go into labour spontaneously may have more chance of successful vaginal delivery than those in whom induction and augmentation are necessary.

We decided to use a combined spinal-epidural technique to provide analgesia when our patient presented in labour. In view of good left ventricular function, invasive monitoring was considered unnecessary. The intrathecally administered drugs provided excellent analgesia as previously reported.[10] However, the epidural was not fully effective confirming the problems encountered by others.[11] Should our patient have required an emergency caesarean section, we could have repeated the spinal or alternatively administered a general anaesthetic using a cardiovascularly stable approach after instituting invasive monitoring.

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