Anaesthesia for Caesarean section in a patient with Jervell, Lange-Nielsen syndrome

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The anaesthetic management of a patient with Jervell, Lange-Nielsen syndrome (a form of congenitally prolonged QT interval) requiring emergency Caesarean section is presented. An epidural anaesthetic using 3-chloroprocaine produced a safe and satisfactory anaesthetic for the procedure in a patient prone to ventricular arrhythmias and cardiac arrest. High levels of circulating catecholamines have been postulated as one of the causes of the arrhythmias in these patients, making it advisable to select drugs and techniques known to minimize catecholamine levels. Although communication can be difficult in these profoundly deaf patients, it is important in order to reduce the emotional stress. Elective Caesarean sections under general anaesthesia have been reported; it appears that a well planned regional anaesthetic is equally safe.

There are several varieties of the prolonged QT syndrome (long QT syndrome, LQTS) which might either present as a congenital form of the disease, or be acquired in later life.¹⁻⁵ A case of Jervell, Lange-Nielsen (JLNS, cardioauditory or surdocardiac) syndrome is presented. This autosomal recessively inherited syndrome consists of a prolonged QT interval, neural deafness and syncopal attacks secondary to ventricular tachycardia, ventricular fibrillation or asystole, or sudden death.

The patient whose case is reported was known to have JLNS and required an anaesthetic to facilitate an emergency Caesarean section.

Key words

ANAESTHESIA: epidural, Caesarean section; SYNDROME: Jervell, Lange-Nielsen syndrome, prolonged QT syndrome.

Case report

A 19-year-old deaf Caucasian female presented at 38 weeks' gestation in early labour. She had a history of multiple syncopal attacks in childhood, which were fairly well controlled from age seven onwards by beta adrenergic blockers (initially propranolol, then atenolol, when depression and lethargy developed while the patient was receiving propranolol). At age 12 ventricular fibrillation and cardiac arrest occurred while she was en route to the dentist; after resuscitation she remained in the Intensive Care Unit for one week with aspiration pneumonia, mild hypoxic encephalopathy (from which she made a gradual recovery) and recurrent ventricular tachycardia that was controlled by lidocaine. During childhood the QT interval ranged from 0.66 to 0.49 seconds.

Family history revealed a cousin with syncopal attacks who died suddenly at age 22 and another cousin with prolonged QT syndrome, deafness and a seizure disorder.

On admission to hospital the patient's heart rate was 73 beats per minute; the electrocardiogram (ECG) revealed a QTc of 0.54 second. After initiation of ECG monitoring of the patient, syntocinon augmentation was begun. An initial fetal tracing showed poor variability and late decelerations that improved slightly after syntocinon was discontinued and oxygen was administered. As maternal beta-blockade can cause fetal hypoxia, and might attenuate the usual signs of fetal distress,⁷ the obstetrician planned a Caesarean section as soon as possible.

Epidural anaesthesia was selected because it was thought that a rapid-sequence induction and intubation might produce sufficient stress to induce dysrhythmias. Great care was taken to explain the procedure fully to the patient in order to reduce anxiety; since she was profoundly deaf and could not lip-read, this was accomplished with the assistance of a hospital social worker fluent in sign language. The epidural catheter was easily inserted in the L₃₋₄ interspace and a test dose of 3 ml of two per cent lidocaine without epinephrine was injected. In some patients with LQTS, lidocaine has caused further prolongation of the QTc.² This, coupled with the difficulty in

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communication of symptoms, should an inadvertent intravascular injection occur, lead us to choose an ester, 3-chloroprocaine, over lidocaine or bupivacaine. Incremental (3–5 ml) boluses were given until analgesia was obtained to the level of T₄, a total of 30 ml being given for the entire procedure. In the event that volume expansion and uterine displacement did not maintain the blood pressure, small (2.5 mg) doses of ephedrine would be given; however, both blood pressure and heart rate remained stable. Intravenous diazepam 2.5 mg was given after the delivery of a healthy boy (Apgar scores of eight at one minute and nine at five minutes), and epidural morphine 4 mg was given at the end of the case for postoperative pain relief. ECG monitoring was continued for three hours postoperatively; her condition remained stable, the QTc was 0.51 second (0.54 second preoperatively) and she was discharged to the ward. She and the baby did well, and were both discharged five days postoperatively. Five months later she had another cardiac arrest, from which she was resuscitated. She was unconscious and required mechanical ventilation for two days and had another arrest during that time. She was discharged in ten days with only a memory loss and unsteady gait as sequelae. A few weeks later after an argument with her husband she became pale and shaky. This had resolved and her ECG was as usual by the time she arrived in the Emergency Department.

Discussion

The Jervell-Lange Nielsen syndrome is rare; while the incidence is estimated to be 1 in 300,000 in the general population, the syndrome occurs in one per cent of patients with congenital deafness.⁸ The true incidence is difficult to determine as many with the condition die in infancy or childhood if not treated.

The abnormal repolarization of the cardiac muscle in these patients results from an imbalance in the cardiac sympathetic innervation. The imbalance is likely due to either decreased activity in the right cardiac sympathetic nerves (chronotropic activity) or perhaps to an increased activity of the left (inotropic activity). The altered sympathetic tone results in delayed ventricular repolarization, increasing the susceptibility to dysrhythmias. Adrenergic stimulation from physical or emotional stress or from exogenous adrenergic drugs might precipitate dysrhythmias.² Other proposed locations for the autonomic imbalance are the brain-stem, sympathetic chain or adrenergic nerve terminals.² Another possible mechanism is a deficient blood supply to the sinus node resulting in ischaemia or infarction causing sinus pacemaker failure which allows escape beats to develop. There may also be an enzymatic deficiency or metabolic abnormality causing the LQTS.² In 1975, Schwartz *et al.* reported that of

68 patients receiving no treatment 74 per cent died in infancy or childhood; of 79 patients receiving beta blockers six per cent died in infancy, childhood or early adulthood. Other medications that have been successfully used are phenytoin, phenobarbitone, primidone, calcium entry blockers, bretylium and lidocaine in acute attacks. Left stellate ganglion block or ganglionectomy has also been successful in blocking the sympathetic imbalance.⁹

In the anaesthetic management of these patients, reduction of both emotional and physical stress is of paramount importance. This is achieved by establishing good communication and rapport between the patient and medical staff, and by selecting anaesthetic agents and techniques that minimize either catecholamine release or cardiac sensitivity to catecholamines. Communication through an interpreter is time-consuming, but necessary to reduce anxiety. Emotional stress may have serious or even potentially fatal consequences, as is illustrated by the occurrence of a cardiac arrest in this patient seven years previously. This patient presented a challenge in that she was first made known to the anaesthesia department when she arrived in labour with signs of fetal distress.

There are few reports of anaesthesia for pregnant patients with LQTS. One report of labour in a patient with idiopathic LQTS mentions epidural anaesthesia being used for forceps extraction, but does not describe the anaesthetic management.¹⁰ Another report details two elective Caesarean sections in a patient with JLNS done utilizing general anaesthesia (diazepam was given as pre-medication, a rapid-sequence induction with thiopentone and succinylcholine was followed by maintenance with nitrous oxide, oxygen and halothane).⁵ In the present case a general anaesthetic was considered undesirable as the fetal distress would necessitate a light anaesthetic which, because of the maternal catecholamine release and subsequent risk of arrhythmias was to be avoided.

Regional (epidural) anaesthesia can be safely used for Caesarean section in patients with LQTS, with certain caveats acknowledged. The usual dose of beta-blockers should be continued, supplemented with intravenous propranolol as necessary. Stress, drugs, and techniques releasing adrenalin should be avoided, although pure alpha agonists or low doses of mixed vasoconstrictors are theoretically safe. Adequate hydration and proper positioning are the desirable methods of prevention of hypotension. With respect to drug selection, operations such as Caesarean section might require doses of local anaesthetics sufficient to cause systemic toxicity in the absence of epinephrine, yet the latter is to be avoided.² Alternative drugs (as chosen for this patient), or careful titration with avoidance of intravascular injection is required to prevent disaster. As in every case provision must be made for emergency defibrillation and resuscita-

tion should it be needed, and neonatal management including prevention of bradycardia and hypoglycaemia secondary to beta-blockade must be available.

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Résumé

La conduite anesthésique d'une patiente atteinte du syndrome de Jervell, Lange-Nielsen (une forme congénitale d'un intervalle QT prolongé) requérant une césarienne d'urgence est présentée. Une anesthésie épidurale utilisant le 3-chloroprocaine était sécuritaire et satisfaisante pour la procédure chez une patiente susceptible de développer des arythmies ventriculaires et un arrêt cardiaque. Des taux élevés de catécholamines circulantes ont été suggérés comme étant l'une des causes de ces arythmies chez ces patientes rendant plus sécuritaire le choix de drogues et de techniques connues comme pouvant diminuer le taux de ces catécholamines. Même si la communication est difficile avec ces patients atteints de surdit elle est importante afin de diminuer le stress motionnel. L'anesthsie gnrale pour une csarienne lective a t rapporte chez ces patientes et il apparat qu'une anesthsie rgionale bien planifie est aussi scuritaire.