ANAESTHESIA FOR CAESAREAN SECTION AND THE JERVELL, LANGE-NIELSON SYNDROME (PROLONGED Q-T INTERVAL SYNDROME)

J. V. FRESHWATER

SUMMARY

The anaesthetic management is described of a patient with Jervell, Lange-Nielson (JLN) syndrome presenting on two occasions for Caesarean section. The syndrome, its treatment and the choice of anaesthesia are discussed.

The syndrome of congenital deafness, syncopal attacks and an abnormal ECG showing prolongation of the Q-T interval corrected for heart rate (Q-Tc > 0.45 s) was described first by Jervell and Lange-Nielson (1957). It is a rare hereditary disease, inheritance being of the autosomal recessive type. The ECG abnormality resembles closely that found in the Romano-Ward syndrome which, however, is not associated with deafness, and has probably an autosomal dominant mode of inheritance. Both may present with a family history of syncope and of sudden death, often in infancy, occurring after exercise or emotional stress. Syncope may occur as a result of Stokes-Adams attacks, asystole or ventricular fibrillation occurring in the presence of atrio-ventricular block. Patients with prolonged Q-T interval syndromes may exhibit the “Torsades des pointes” phenomenon, polymorphic ventricular tachycardia, a rapid ventricular tachycardia characterized by changing QRS morphology. This is often self-terminating, but may degenerate into established ventricular fibrillation. Recent advances in therapy, notably the use of β-adrenoceptor blocking drugs, have improved survival, and a few patients presenting for surgery and anaesthesia are now recorded in the literature. The patient described is one of the few to have the classic Jervell, Lange-Nielson (JLN) syndrome and, possibly, the first reported to undergo Caesarean section.

CASE REPORT

A 20-year-old deaf Sikh female known to have the JLN syndrome presented for the management of her pregnancy. From age 5 yr she was subject to Stokes-Adams syncopal attacks most frequent during her early childhood. Two siblings had died in infancy. At 9 yr the JLN syndrome was diagnosed (Q-Tc 0.53 s) and treatment initiated with digoxin 0.0625 mg daily. This was later changed to practolol 200 mg twice a day with good control of the attacks. Propranolol 20 mg twice per day was substituted later.

On admission to hospital the patient had had no attacks for 3 years in spite of infrequent attendance for follow-up and, probably, poor compliance with therapy. She had an uneventful pregnancy, receiving propranolol 20 mg twice per day and was booked for elective Caesarean section under general anaesthesia. She was admitted for full preoperative assessment, including ECG. Propranolol 20 mg twice per day was continued until the morning of her operation. She was premedicated with diazepam 10 mg by mouth and after pre-oxygenation, cricoid pressure was applied. Anaesthesia was induced with thiopentone 250 mg, and suxamethonium 70 mg given to facilitate tracheal intubation. Ventilation was controlled, and anaesthesia maintained with nitrous oxide and 0.5% halothane in oxygen. She received intermittent injections of suxamethonium 50 mg during the 40-min procedure. A live, healthy male infant was delivered which was found to have no ECG abnormality. Immediately following operation the mother received papaveretum 5 mg, cyclizine 25 mg i.m. and diazepam 2.5 mg i.v. She had shown no additional ECG abnormalities throughout the procedure, and her course after operation was uneventful.

Two years later the patient presented for the management of her second pregnancy, and requested sterilization. She had remained well-controlled on propranolol 20 mg twice per day, with no further syncopal attacks. She was prepared for general anaesthesia as before, receiving her morning
dose of propranolol 20 mg and lorazepam 2 mg by mouth 1.5 h before operation. Her English-speaking sister accompanied her into the anaesthetic room to act as interpreter. Induction and maintenance followed the technique described already. During the somewhat longer procedure (some 70 min) she required two increments of suxamethonium 25 mg. After the second there was a decrease in heart rate from 60 beat min\(^{-1}\) to 49 beat min\(^{-1}\) which responded to atropine 0.2 mg i.v.—returning to 78 beat min\(^{-1}\). Two subsequent increments of suxamethonium 10 mg were given with no further alterations in heart rate. Papaveretum 10 mg i.v. was given after delivery. Her postoperative course was uneventful and she remains well, continuing to receive propranolol 20 mg twice per day. The second male infant is healthy and has no ECG abnormalities.

**DISCUSSION**

JLN syndrome is an autosomal recessive condition. Heterozygotes are clinically normal although their ECG may show prolongation of the Q–T interval. The frequency in the general population is 1 in 300,000 and in children with perceptive deafness, 1 in 100. Consanguinity of parents predisposes to the syndrome (Matthews, Blaint and Townsend, 1972): Sikhs usually inter-marry within their own community. The frequency of the syncopal attacks diminishes with age (Ratshin, Hunt and Russell, 1971). Syncope in patients with prolongation of the Q–T interval is usually related to exercise; in this patient emotional stress was a precipitating factor. Exercise tolerance was not tested in this patient as a child in view of her frequent and prolonged syncopal attacks at that time, and was probably not justifiable during pregnancy.

The pathogenesis of the condition is unclear, but is probably the result of an imbalance of tone in the cardiac sympathetic nerves, depressed on the right side or exaggerated on the left. This asymmetry in sympathetic tone delays repolarization of the ventricular myocardium, leading to an increased susceptibility to fibrillation. Recent work suggests that depression of the right side is more probable. Patients who have had percutaneous left stellate ganglion block (Moss and McDonald, 1971), or surgical left cervico-thoracic gangliectomy (Callaghan, Nichols and Sweet, 1977) showed clinical improvement with decreases in the frequency of syncopal attacks and sometimes a decrease in the duration of the Q–T interval.

Propranolol is the treatment of choice. Its beneficial effect is probably not a result of its influence on the Q–T interval, which it tends to prolong, but rather to direct anti-sympathetic activity, and an ability to increase the threshold for ventricular fibrillation (Milne, Camm and Ward, 1980). Unfortunately, not all patients tolerate therapy or are controlled adequately, and further anti-arrhythmic management may be difficult. Lignocaine and phenytoin have been used following surgery (Brown, et al., 1981). Percutaneous left stellate ganglion block has proved useful (Ponte and Lund, 1981), and surgical sympathectomy could be offered for long-term management.

The choice of anaesthetic technique for these patients is difficult. Anaesthesia must be hazardous where there is a risk of a life-threatening arrhythmia. The choice in this patient was further limited by her pregnancy since any technique chosen had to allay anxiety, provide cardiovascular stability, and satisfy the strict criteria for safety of mother and child inherent in any obstetric anaesthetic.

Communication between the patient and the medical team was restricted by her severe deafness and language problems. This difficulty and the risks of possible hypotension in the presence of β-blockade excluded extradural anaesthesia as a suitable technique. General anaesthesia, using a well-established technique and careful monitoring, offered the safest choice.

Preoperative visits, premedicants, the presence of a familiar relative in the induction room, and the excellent rapport between patient and cardiologist over some 15 years, undoubtedly decreased the perioperative anxiety. Lorazepam was substituted for diazepam on the second occasion, as it is less deleterious to the fetus (Whitelaw, Cummings and McPadyen, 1981).

Callaghan, Nichols and Sweet (1977) advised that factors which prolong the Q–T interval should be avoided—hypocalcaemia, hypomagnesaemia and hypokalaemia, as well as certain drugs which prolong the Q–T interval such as quinidine, lignocaine, procainamide, and some phenothiazines. They used a nitrous oxide, morphine technique with neournuscular blockade. General anaesthesia must be hazardous where there is a risk of a life-threatening arrhythmia. The choice in this patient was further limited by her pregnancy since any technique chosen had to allay anxiety, provide cardiovascular stability, and satisfy the strict criteria for safety of mother and child inherent in any obstetric anaesthetic.
tricular fibrillation. Intermittent suxamethonium is a well-tried, safe technique in obstetric anaesthesia. Any bradycardia occurring can be reversed by atropine. This was only required in this patient during the second, longer procedure, and a small dose (0.2 mg) under ECG control restored heart rate to close to its previous value.

This patient exhibited no problems in the period after operation, although others have experienced difficulty in managing arrhythmias in uncontrolled patients (Callaghan, Nichols and Sweet, 1977; Owitz et al., 1979; Wig et al., 1979). Our experience supports that of Medak and Benumof (1983) and suggests that the well-controlled patient with prolonged Q–T syndrome may be less at risk than expected. Most reported cases of the prolonged Q–T syndrome fall into the Romano–Ward type. Perhaps the JLN syndrome has a more benign natural history. Improvement with advancing age and a greater susceptibility to control of syncopal attacks by propranolol may render anaesthesia for these patients less hazardous.

ACKNOWLEDGMENTS

The author wishes to thank Dr Matthews, Consultant Cardiologist, at the Western General Hospital, Edinburgh for permission to report on his patient, Dr I. T. Davie, Consultant Anaesthetist at the same hospital for his encouragement and advice, and Mrs I. Blair for typing the manuscript.

REFERENCES


