PROLONGATION OF THE Q–T INTERVAL (ROMANO–WARD SYNDROME): ANALGESIC MANAGEMENT

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SUMMARY

The anesthetic management is described of a patient with prolonged Q–T interval which had been complicated by ventricular fibrillation at induction of general anaesthesia for a previous operation. This complication was prevented by effective premedication with i.v. propranolol and block of the left stellate ganglion.

Congenital delay of depolarization-recovery of ventricular myocardium is shown in the e.c.g. as prolongation of the Q–T interval. It is a rare inherited condition (Romano, Gemme and Pongiglione, 1963; Ward, 1964) which was first described in association with deafness (Jervell and Lange-Nielsen, 1957).

It can cause syncope or sudden death at any age, usually following physical exertion or strong emotions which may result in ventricular fibrillation. Surgery or anaesthesia may cause such an attack (Wig et al., 1979). We describe some preventive measures in the anaesthetic management.

CASE REPORT

A 27-yr-old female with a 5-year history of gynecological complaints was referred for laparotomy. She had been admitted for operation in another hospital earlier in the year, but the procedure was abandoned after induction of anaesthesia because the patient developed ventricular fibrillation when pancuronium 1 mg was given i.v. to prevent muscle fasciculations following suxamethonium. Cardiac massage was applied externally and the rhythm reverted to normal spontaneously.

The patient had suffered attacks of syncope from the age of 7 yr usually associated with physical exertion. The symptoms were worst at the age of 21 yr when they occurred several times per week. The patient was diagnosed as suffering from epilepsy for many years, but when she was referred to the Cardiology Department at Uppsala aged 22 yr, an increased Q–T interval was discovered. Propranolol was prescribed but she had failed to comply because of intolerance to exercise and bradycardia. However, the syncopal attacks diminished in frequency and the last before the one reported above had occurred 5 years previously.

Two pregnancies and two laparotomies under general anaesthesia had been without complications. The patient's sister had suffered from attacks of syncope as a child and her mother, and her two children, had prolonged Q–T intervals without symptoms.

Physical examination, heart size and all laboratory results were normal and e.c.g. showed a Q–T interval of 0.56 s in all leads with normal T waves at a heart rate of 80 beat min⁻¹ (normal value at this heart rate is 0.374 s). Metoprolol 200 mg orally was prescribed by the cardiologist for each of 2 days before operation. Premedication was with diazepam 15 mg orally 2 h before surgery and morphine 10 mg with hyoscine 0.4 mg i.m. 1 h before operation. Twenty minutes before induction of anaesthesia the left stellate ganglion was blocked percutaneously with 0.5% bupivacaine 8 ml, which produced obvious vasodilatation in the left arm, ipsilateral ptosis, but little difference in pupil size, probably because of the miotic effect of morphine. The patient was drowsy as a result of premedication and performance of the block had a minimal effect on heart rate. Propranolol 1 mg was given i.v. (parenteral metoprolol was unavailable) just before induction of anaesthesia, which was carried out with fentanyl 0.4 mg, thiopentone 200 mg and alcuronium 15 mg. Ventilation was controlled artificially via an endotracheal tube.
throughout the uneventful operation, e.c.g. was monitored at all times (precordial V5 lead) and a defibrillator was immediately available. After operation, two further injections of propranolol 1 mg i.v. were given at intervals of 8 h, and the e.c.g. monitored for 24 h.

Q-T intervals were normal shortly after induction of anaesthesia (fig. 1) but returned to pre-operative values in the course of the 2-h operation. The effect of the two injections of propranolol given during recovery on the Q-T interval was beneficial but small. However, the first measurement of the Q-T interval after induction of anaesthesia showed a remarkable relative shortening. Unfortunately, we could not obtain a paper record of the e.c.g. during the pre-induction period to measure Q-T intervals accurately and so we could not be certain its shortening had been caused by stellate ganglion block, beta blockade or anaesthetic agents used. To establish the therapeutic value of the stellate ganglion block in this patient, we decided to repeat it 4 days after operation, with the patient's co-operation, by percutaneous infiltration of the left ganglion with 5 ml of 1% lignocaine. Lead II of the e.c.g. was recorded at 5-min intervals and the Q-T intervals were briefly reduced to values within normal limits (figs 1, 2), in spite of minimal change in heart rate. A complete Horner's syndrome was apparent within 5 min of injection.

Atropine given during the operation also reduced the Q-T interval (fig. 1), with a corresponding increase in heart rate such that the change followed a line parallel to that defining the normal relationship between heart rate and Q-T interval (Bazett, 1920). Effects of arousal at extubation and of physical exercise were similar in nature to those of atropine. In figure 2, e.c.g. tracings are shown which correspond to some of the points plotted in figure 1, having superimposed the peaks of the first T waves for clarity. All the Q-T values plotted are averages of four consecutive measurements.

**DISCUSSION**

Schwartz, Periti and Malliani (1975) have reviewed more than 200 patients with this congenital syndrome; three-quarters of the untreated patients died, treatment with beta-blocking agents improved the survival rate considerably, and all those patients who had the left sympathetic supply to the heart surgically removed had survived.

The cause of Romano–Ward syndrome is unknown, but some evidence suggests that it is caused by imbalance of the sympathetic cardiac tone which is either exaggerated on the left or, more probably, depressed on the right (Schwartz, Periti and Malliani, 1975). This asymmetry in tone delays repolarization of the ventricular myocardium, leading to an increase in its susceptibility to fibrillation (Han and Goel, 1972).

Percutaneous blockade of the right stellate ganglion was shown in one patient to increase the Q-T interval even further, whereas a left block reduced the interval to normal values; surgical denervation following this test suppressed the symptoms (Mors and MacDonald, 1971). Additional reports have confirmed the clinical success of this manoeuvre, even if in some instances the Q-T interval was not reduced to normal values (Yanagida, Kemi and Suwa, 1976; Callaghan, Nichols and Sweet, 1977).
PROLONGATION OF THE Q-T INTERVAL

In the case presented here, the pre-operative left stellate ganglion block was successful in preventing complications, and was shown later to be effective in reducing the Q-T interval, indicating that this patient would possibly benefit from surgical sympathectomy, should her symptoms recur.

It was interesting to note the small effect of propranolol upon the Q-T interval in spite of its known therapeutic effectiveness (Milne, Camm and Ward, 1980).

Pancuronium is known to cause sympathomimetic effects on the heart and there is evidence in animal studies with clinical doses (0.2 mg kg⁻¹) that this is a pre-synaptic indirect action by increasing the local release of noradrenaline (Domenech et al., 1976); this may be more apparent in a patient with an increased sympathetic tone caused by apprehension. This effect of pancuronium or its known parasympatholytic actions, or both, might have been precipitating factors of the ventricular fibrillation at previous induction of anaesthesia in our patient. Therefore, the use of agents such as pancuronium or atropine is probably best avoided in this condition.


REFERENCES


PROLONGATION DE L'INTERVALLE Q-T
(SYNDROME DE ROMANO-WARD): CONTROLE DE L'ANESTHESIE

Nous decrivons ici le controle de l'anesthesie d'un patient ayant un intervalle Q-T qui avait ete complique par une fibrillation ventriculaire au moment de l'induction de l'anesthesie generale, lors d'une operation precedente. Cette complication a ete evitée par une medicament preoperatoire efficace, grace a du propranolol administre par voie intraveineuse et a un blocage du ganglion etoiyle gauche.
Es wird die Anästhesiebehandlung bei einem Patienten mit verlängertem Q-T Intervall beschrieben, das durch Kammerflimmern bei der Einleitung der Narkose bei einer früheren Operation erschwert wurde. Diese Komplikation wurde durch entsprechende Prämiedikation mit intravenös verabreichtem Propanolol und Blockade des linken Stellatumganglions verhindert.

Se describe la gestión de la anestesia de un paciente que presentaba un prolongado intervalo Q–T, el cual se había complicado por la fibrilación ventricular durante la inducción de anestesia general en una operación anterior. Esta complicación se previno mediante una efectiva premedicación con propanolol intravenoso y el bloqueo del ganglio estrellado izquierdo.