**Short reports**

**Asystole in the prolonged QT syndrome**

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**Summary** We report a 4½ year old boy with the Romano-Ward syndrome in whom an asystolic period of 18 seconds duration followed an episode of ventricular tachycardia. Treatment with propranolol and implantation of a demand pacemaker has prevented further attacks.

The Romano-Ward syndrome is a dominantly inherited condition in which hereditary prolongation of the QT interval is associated with syncopal attacks and sudden death in childhood. The hearing is normal unlike the variant described originally by Jervell and Lange-Nielsen in which the sufferers are profoundly deaf. Life threatening syncopal attacks are caused by paroxysms of arrhythmia, most commonly ventricular tachycardia. Asystole is a rarely recorded arrhythmia in this condition and only two reported patients have had pacemakers inserted. The patient of Olley and Fowler\(^2\) could not be paced because of resultant ventricular fibrillation and the authors considered implantation of a pacemaker to be contraindicated. Our patient has responded favourably to a combination of propranolol and a demand pacemaker.

**Case report**

The patient was a 4½ year old boy of unrelated parents. He had presented at another hospital at the age of 3 years with episodes of loss of consciousness lasting from a few seconds to several minutes; during these he became pale and rigid but there were no clonic movements and afterwards he was sometimes lethargic. The attacks were often precipitated by emotional stress or physical activity and in some there was premonitory abdominal pain.

The original diagnosis was one of epilepsy, although an electroencephalogram was normal. Carbemazepine seemed to produce an initial improvement but was stopped after the attacks recurred. At the age of 4½ years he was reviewed after a particularly prolonged attack and the possibility of a paroxysmal arrhythmia was considered. He became increasingly distressed, collapsed during an electrocardiographic recording, and after resuscitation was transferred to the Royal Liverpool Children's Hospital. The initial electrocardiogram showed sinus rhythm, T wave alternans, and prolongation of the corrected QT interval to 0·56 seconds (Fig. 1). The latter part of the recording showed torsade de pointes with a ventricular rate of 294 beats per minute. The ventricular tachycardia ended in asystole which lasted for 18 seconds (Fig. 2). Further investigation included echocardiography and determination of urea and electrolytes, calcium and magnesium, all of which were normal.

A temporary transvenous pacing wire was inserted under general anaesthesia and treatment with oral propranolol (15 mg, 8 hourly) was begun. Without the pacemaker episodes of nodal rhythm

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*Fig. 1  Lead II recorded during sinus rhythm before the onset of arrhythmia shown in Fig. 2. The QT interval is prolonged (corrected QT interval 0·56 seconds) with positive and negative T waves alternating.*
with a rate of 50/60 beats per minute occurred. A Cardiologic, programmable pacemaker was implanted into a left subcostal pouch and connected to a corkscrew electrode which was inserted into the diaphragmatic surface of the left ventricle. The pacemaker was set in demand mode at 70 beats per minute and several electrocardiographic recordings during the following 6 months showed a paced rhythm. Three episodes of faintness preceded by abdominal pain which occurred during the follow up period were thought to be brief periods of tachycardia and the dose of propranolol was increased to 25 mg, 8 hourly.

No family history of syncope or sudden death was elicited and the parents' electrocardiograms were normal. The patient's asymptomatic 8 year old sister, however, has a corrected QT interval of 0-57 seconds.

Discussion

The long QT interval when seen on an electrocardiogram is an important diagnostic pointer. Hereditary causes should be considered after excluding metabolic ones especially in patients with a history of syncopal attacks precipitated by stress. A family history of childhood attacks similar to fainting may be present. The diagnosis is made on the basis of a corrected QT interval in excess of 0-44 seconds. QT prolongation is not invariably present on the interval electrocardiogram and repeated tracings may be necessary. T wave alternans is a helpful diagnostic feature and warns of impending arrhythmia. Arrhythmias recorded during the syncopal episodes include torsade de pointes, ventricular tachycardia, and fibrillation. The mortality was extremely high in the series of Schwartz but has declined as treatment has improved.

The arrhythmia may end with a period of asystole as occurred in our patient and this may be an important cause of death in this syndrome. Electrocardiographic recordings showing this sequence have seldom been obtained but the introduction of miniature electrocardiographic tape records which may be applied to the chest during an attack will facilitate diagnosis and improve our knowledge of the electrophysiological mechanisms. Pathogenesis of this condition is incompletely understood but evidence from several sources indicates an imbalance of the autonomic control of the heart. Electrical stimulation or surgical ablation of the stellate ganglion alters the QT interval and left stellate ganglionectomy has been used with some success to control attacks in the long QT syndrome. Further support is given by the histological studies of James et al who showed severe, pathological changes in the cardiac nerves.

Treatment of patients with the prolonged QT syndrome is directed to the long term prevention of life threatening arrhythmias as the condition often remits after adolescence. Phenytoin and propranolol have been the most effective treatments, used either singly or in combination. Quinidine, which lengthens the QT interval, should be avoided. The episode of asystole in our patient indicated the need for a demand pacemaker in spite of the previous, unfavourable reports of pacing in this syndrome. Without long term pacing the use of beta blocking agents might, while reducing the incidence of tachyarrhythmia, increase the risk of bradycardia or asystole.

In conclusion, evidence of the prolonged QT syndrome should be sought in children of all ages with unexplained syncopal attacks. The combination of propranolol and long term pacing may give an improved prognosis in this syndrome.

References

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### Complete recovery after profound acidosis (pH 6·49)

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**SUMMARY** We describe an infant with profound acidosis caused by chronic therapeutic salicylate poisoning. The confirmed arterial blood pH of 6·49 must be close to the limit of tolerable acidity and is the lowest such value in our experience. Full recovery was made.

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**Case report**

A 2½ month old infant weighing 3·6 kg presented to the paediatric emergency ward with a three day history of a dry non-productive cough, diarrhoea, and vomiting. During the preceding three days she had reportedly received approximately 125 mg aspirin three times daily (administered by the mother) in an attempt to alleviate symptoms. The mother also reported that the infant had had generalised convulsions lasting 10 minutes on the day of admission.

Clinical examination showed an infant who was somnolent, dehydrated, hypothermic (temperature 35°C), hyperpnoeic (respiratory rate 72/minute), shocked (systolic blood pressure 50 mmHg), and anuric. There was clinical evidence of extensive, right sided, middle and lower lobe pneumonia. The infant's weight, head circumference, and length were well below normal. Arterial blood gas analysis and serum urea, creatinine, and electrolyte determinations confirmed a profound metabolic acidosis with respiratory compensation and hypertonic dehydration.

Initial baseline laboratory values were as follows: pH 6·82; PacO₂ 13·1 mmHg; Pao₂ 40·8 mmHg; bicarbonate 3·4 mmol/l; base deficit 32·0 mmol/l; oxygen saturation 36·7%; serum sodium 163 mmol/l; serum potassium 7·3 mmol/l; serum urea 38·7 mmol/l; serum creatinine 230 umol/l; serum osmolality 397; and whole blood glucose 18·2 mmol/l. A serum salicylate concentration of 1·36 mmol/l (reference value less than 0·36 mmol/l) and a high anion gap acidosis confirmed chronic, salicylate over-dosage. A blood specimen for prothrombin index failed to clot and a bleeding time (Ivy method) of 20 minutes confirmed a bleeding diathesis. A lumbar puncture on admission was normal.

Sodium bicarbonate 8·5% (2 mmol/kg) was given intermittently in addition to a continuous infusion at a rate of 0·03 mmol/kg/minute. While measures were taken to correct the severe metabolic acidosis and dehydration the patient developed generalised convulsions which were controlled with diazepam (1·5 mg, stat) and phenobarbitone (20 mg, IV stat) in single doses. Fifteen minutes later the patient developed a cardiorespiratory arrest but was successfully resuscitated and transferred to the intensive care unit. Prophylactic dexamethazone (2 mg IV, 6 hourly) was given for possible cerebral oedema. Fresh frozen plasma and vitamin K (2 mg IV, single dose) were given because of the deranged haemostatic values. Arterial blood gas determination immediately after cardiopulmonary resuscitation showed the following: pH 6·49; PacO₂ 43·1 mmHg; Pao₂ 30·1 mmHg; bicarbonate 3·4 mmol/l; and oxygen saturation 24%.

The above values were checked in triplicate. The patient was then rehydrated slowly, rewarmed, mechanically ventilated, and given inotropic support with isoprenaline (0·04 ug/kg/minute) and intravenous antibiotics (cefotaxime 100 mg IV, 6 hourly). Peritoneal dialysis (using 1·5% Dianeneal solution) was begun 6 hours after admission to the intensive care unit. Arterial blood gas analysis four hours later showed: pH 7·06; PacO₂ 33·9 mmHg; Pao₂ 108·4 mmHg, bicarbonate 7·3 mmol/l; and oxygen saturation 93·7%. At this point the ventilator settings were as follows: FiO₂ 0·8, respiratory rate 35/minute, expired minute volume 4·0 l/minute. Twelve hours later the patient was still acidic (pH 6·49) and a second lumbar puncture confirmed a normal pressure and absence of malignant cells. Chronic renal failure and failure to thrive led to death six weeks later.
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