TORSADE DE POINTES AND SYNCOPAL ATTACKS IN A 26-YEAR OLD WOMAN WITH CONGENITAL COMPLETE HEART BLOCK AND PROLONGED QT INTERVAL

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ABSTRACT

A 26-year old woman with congenital complete heart block and prolonged QT interval presented for the first time with syncopal attacks associated with torsade de pointes in adulthood. Cardioversion followed by overdrive pacing was needed to finally control the unstable rhythm. During episodes of non-capture, paroxysms of torsade de pointes leading to ventricular flutter were recorded by a 24-hour ambulatory electrocardiographic monitoring. Beta-blockade and permanent ventricular pacing finally abolished both the syncopal attacks and the torsade phenomena.

The prognosis of congenital complete heart block associated with QT prolongation resembles that of the Romano-Ward syndrome. Recognition of this variant would facilitate earlier treatment of this rare but potentially lethal disorder.

Keywords: Torsade de pointes, congenital complete heart block, prolonged QT interval, syncopal attacks, Romano-Ward syndrome.

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INTRODUCTION

Congenital complete atrio-ventricular block is known to be complicated by syncopal attacks and sudden death as a result of severe bradycardia or ventricular standstill from prolonged junctional exit blocks (1). However, congenital complete heart block with prolonged QT interval is uncommon being reported in no more than 15 or so cases, almost all being in the paediatric age group (2). We report here on an adult woman presenting with such a syndrome.

CASE REPORT

An asymptomatic 26 year old primigravid woman first presented to us at 34 weeks gestation, where she was found to be bradycardic at antenatal check-up. A resting electrocardiogram then showed complete atrio-ventricular block with a ventricular rate of 43 and an atrial rate of 90. The QT was 0.59 second (QTc 0.50 s). Exercise and intravenous atropine accelerated both the atrial and the ventricular rates to 110 and 62 beats per minute respectively. A 24-hour ambulatory ECG recording at that time was uneventful, no ventricular ectopy or prolonged ventricular standstill was detected. A full physical and an echocardiographic examination was normal. An audiographic examination was also normal. She was monitored during pregnancy and labour and successfully delivered a live normal baby daughter at 39 weeks. No syncopal or pre-syncopal symptoms or complex arrhythmia was noted. She was discharged well at one week post-partum.

A review of her past history suggested that she was first found by a childhood doctor to have had a 'slow heart' at 4 years of age. As a result her parents had always forewarned her against overly vigorous activities and sports. Her neonatal and childhood history was normal otherwise; immunisations were complete, and she had only a mild attack of measles at 5 years. Notably, she did not have recurrent sore throats, rheumatic fever, diphtheria or other illnesses which necessitated hospitalizations. Nevertheless, she was a fair sportsperson at school, being active in sprint events and netball. No fainting attacks or undue fatigue complicated her activities until this first pregnancy where her diagnosis was documented and confirmed. No other family history of sudden deaths or similar illness was noted among her six other siblings and parents.

In April 1987, 2 months after her delivery, she suffered her first 'grey-out' and fainted whilst bathing her baby. (The day before, she had complained of left-sided lower premolar toothache and had had a tooth extracted at a local dentist. She denied excessive residual pain, gum bleeding or fever following the procedure.) She recovered spontaneously after about 5 minutes but within the hour, she fainted again. Her husband brought her to the hospital where she was admitted to the coronary care unit.

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The admitting rest electrocardiogram (Fig 1) showed complete atrio-ventricular block with an atrial rate of 90 and a ventricular rate of 44, the QTc was 0.69 second (QT interval) = 0.80 second, using Bazzett's correction). The QRS interval was 0.08 second. Whilst on telemetric electrocardiographic monitoring, the patient again collapsed during micturition, but spontaneously recovered within minutes. The monitor showed short bursts of polymorphous ventricular tachycardia which later deteriorated into ventricular fibrillation (Fig 2), at which point the patient collapsed and developed a generalised seizure. Successful defibrillation followed by a lignocaine infusion was carried out until transvenous right ventricular overdrive pacing was performed at 120 per minute.

Other investigations were essentially normal: haemoglobin 12.4 g/dl, total white count 9 x 109, polymorphs 67%, lymphocytes 32%, eosinophils 1%; Blood urea 2.6 mmol/l, serum creatinine 70 mmol/l, potassium 3.9 mmol/l, sodium 142 mmol/l, calcium 2.42 and 2.34 mmol/l, albumin 43g/l, magnesium 0.91 mmol/l. Arterial blood gases were normal.

A 24-hour ambulatory electrocardiographic monitoring tape was commenced, alongside telemetric visual monitoring. When the pacing rate was decreased to 80 per minute, the patient developed several further episodes of torsade de pointes complicated by ventricular flutter or fibrillation (Fig 3). Defibrillation was delivered twice successfully, following the failure of overdrive pacing (at 120 to 180 per minute). During non-capture of the pacing mode, several short paroxysms of torsade were recorded but were otherwise asymptomatic. Metoprolol 50 mg twice daily was commenced.

Permanent right ventricular pacing was later performed with the ventricular rate set at 72 per minute. With beta-blockade, the torsade did not recur, and the QTc shortened to 0.49 second. She remained well at twelve months follow-up, with no recurrences of syncopal or near-syncopal attacks with maintenance metoprolol. A repeat electrocardiogram showed the rhythm to be pacer-dependent at 72 beats per minute.

DISCUSSION

Although syncopal, near-syncopal episodes and sudden death are known to occur with congenital complete heart block (3, 4), most of these episodes were thought to be caused by severe bradycardia either from prolonged ventricular standstill or inappropriate junctional exit blocks (5). From ambulatory electrocardiographic monitoring,

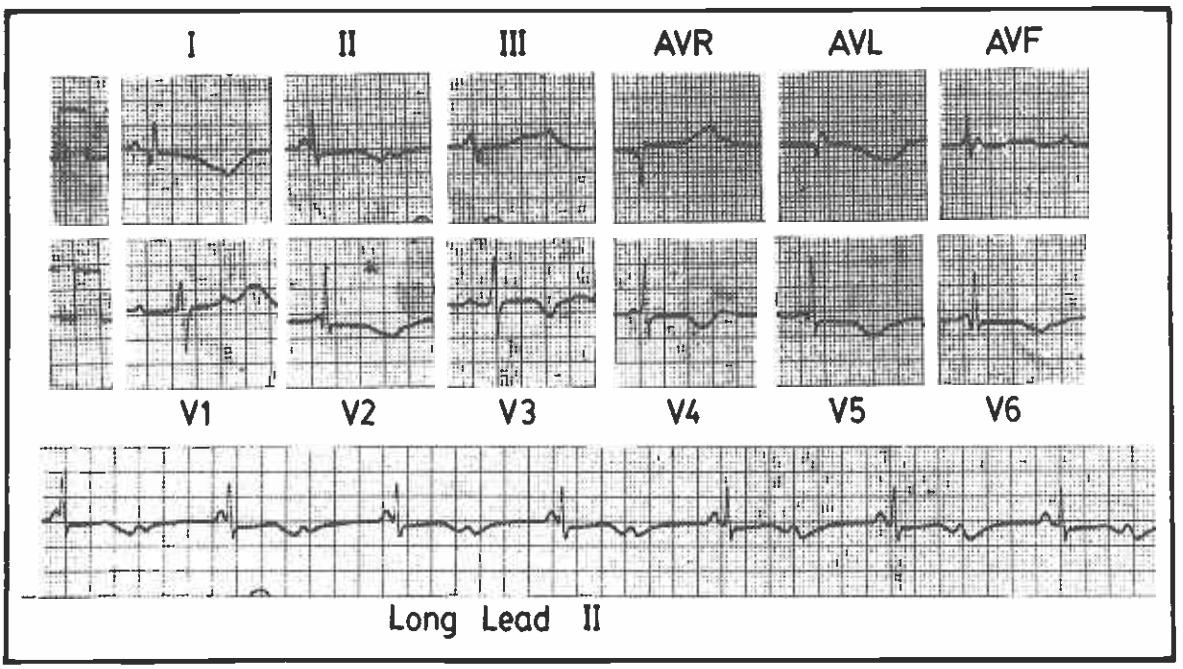

Fig 1
12-lead electrocardiogram showing complete heart block. The atrial rate was 90 and the junctional rate 44 per minute. The QT interval was markedly prolonged to 0.80 second (QTc 0.69 second, by Bazzett's formula). No abnormal notching or alternation of the T wave was seen.

Dewey and others (6), found that mean daytime junctional rate of less than 50 per minute carried a poorer prognosis with regards sudden death or eventual need for permanent pacing. They attributed this to a combination of junctional instability i.e. frequent episodes of junctional exit blocks, flat junctional response or associated tachyarrhythmias.

In 1962, Molthan and others (7) first described some three children with complete heart block who also had prolonged QT interval presenting with convulsive syncope, a typical ventricular tachycardia with and without asystole. These children died suddenly. Nikolic and others (2) have since reviewed some 14 cases with this syndrome including their own study of a 3-year old child who subsequently received permanent pacemaker.

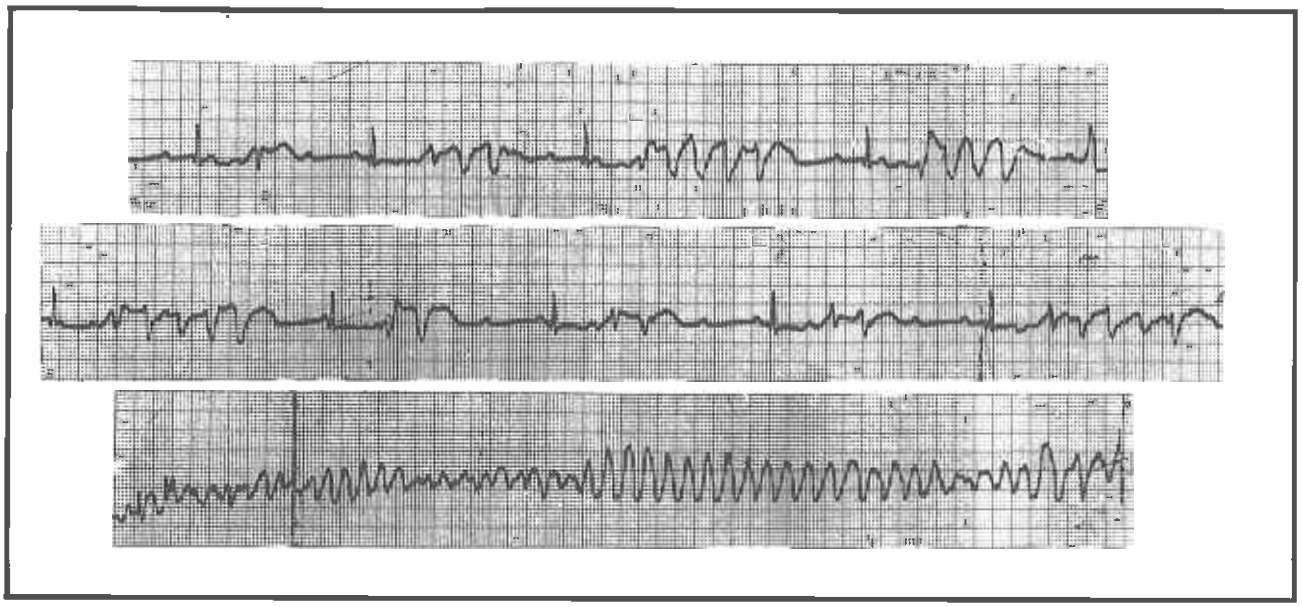

Fig 2
Electrocardiogram (continuous modified lead 2) showing ventricular premature beats falling on the abnormally prolonged QT segment with short polymorphic salvos and couplets. Finally a sustained run of torsade de pointes was seen during syncope.

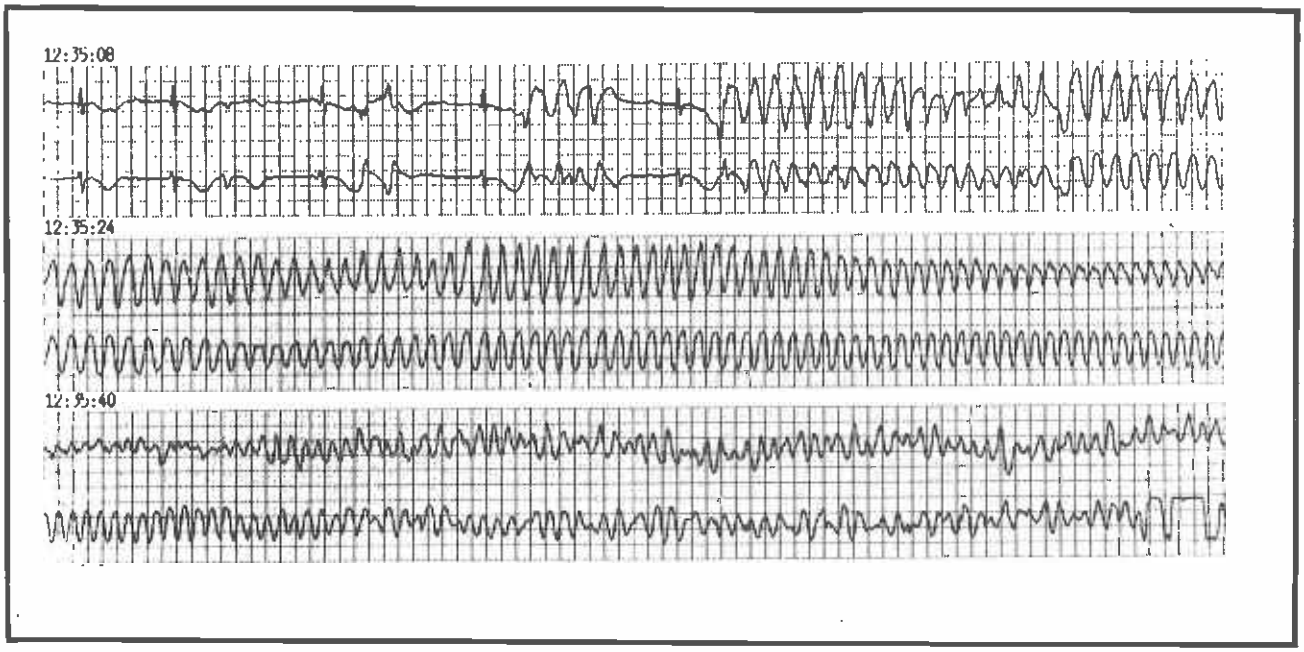

Fig 3
Holter recording showing ventricular premature complexes interrupting the abnormally delayed T wave, initiating short bursts of salvos and torsade de pointes terminating spontaneously in the first instance; then later, sustained torsade type ventricular tachycardia degenerating into ventricular flutter-fibrillation, requiring defibrillation.

Esscher and Michaelsson (8), in their review, showed that QT prolongation indeed carried a worse prognostic outcome with regards sudden deaths and Stokes-Adams attacks in congenital complete heart block. Because of the graver prognostic outcome of this subset of congenital complete heart block, Nikolic and others (2) have suggested that those with prolonged QT interval should be considered as sporadic variants of the Romano-Ward syndrome.

Our patient presented for the first time in adulthood having undergone an uneventful successful pregnancy. Curiously her syncopal episodes began only after what would ordinarily have been passed as routine dental extraction. This was the only temporally-related incident prior to her developing documented torsade de pointe type of ventricular tachycardia, the more prolonged episodes of which resulted in syncopal attacks and one witnessed convulsion. During this time her QT interval appeared to have been further prolonged to 0.08 second, thereby increasing her vulnerability to torsade de pointe ventricular arrhythmias.

Han and others (9) have shown that if activation time differences remain unchanged, the dispersion of repolarisation will be greater at slow than at fast heart rates. This increased dispersion might precipitate reexcitation or reentry circuits of disorganised ventricular tachyarrhythmias such as torsade and ventricular fibrillation. Although the mechanism of ventricular complexes initiating the arrhythmia is unknown, it is possible that increased sympathetic stimulation at the onset of arrhythmia increases diastolic depolarisation and precipitates a discharge of a latent pacemaker in the ventricular conduction system. Such multiple local reentry circuits may be caused by asynchronous recovery of the His bundle branch-Purkinje system, rather than the ventricular myocardium (10). Increased ventricular repolarisation (ie. longer QT interval) in this patient may have resulted from exaggerated imbalance between the left and right cardiac sympathetic innervation as proposed by some authors (11, 12). Could this usually prolonged QT interval be caused by excessive sympathetic stimulation of the left stellate ganglion from the pain or inflammation of the dental extraction? This would certainly be in keeping with the model proposed by Schwartz (13). It appears that a properly timed premature ventricular beat might be in the induction of ventricular arrhythmias in such patients, as might emotional and physical stress (13). Torsade de pointes appears to be an arrhythmia of maximal vulnerability.

We have documented here an adult 26 year-old woman with congenital complete heart block and prolonged QT interval. She presented with convulsive syncopal attacks associated with torsade de pointe type ventricular tachycardia and fibrillation. Several defibrillations, and overdrive pacing finally controlled her unstable rhythm. Beta-blockade appeared to have shortened the QT prolongation. It is noteworthy that no electrolyte imbalance such as low potassium, magnesium or calcium contributed to her sudden onset of symptoms. One possible contributing factor (at least temporally related) was that of a lower premolar tooth extraction the day before.

We believe that congenital complete heart block is not so benign especially when associated with markedly low ventricular or junctional rates or with QT prolongation. Like Nikolic and others (2), we believe that this patient suffered from a variant of the Romano-Ward syndrome with respect to prognosis. Treatment should include permanent pacing, with beta-blockade if necessary.

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