Unusual presentation of long QT syndrome

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We describe the case of a 9-yr-old child with undiagnosed long QT syndrome who experienced an intraoperative cardiac arrest after accidental intravascular injection of bupivacaine with epinephrine via a misplaced epidural catheter.

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Long QT syndrome (LQTS) is a genetically inherited condition that may not be recognized clinically unless the patient presents with syncope, seizures or sudden death.1 It is known to cause sudden death in young, apparently healthy people.2 The mechanism appears to be related to a polymorphic ventricular tachycardia, referred to as torsades...
long QT syndrome is characterized by prolongation of the QT interval on a resting electrocardiogram (ECG) or on exercise testing. The lack of anatomical abnormalities identifiable in life or at autopsy, a deceivingly normal ECG at rest and controversy about the interpretation and measurement of the QT interval duration often make the clinical identification of LQTS extremely difficult in asymptomatic patients. Patients with unrecognized LQTS undergoing procedures requiring general or regional anaesthesia will be at increased risk.

We describe an episode of intraoperative cardiac arrest after accidental intravascular injection of bupivacaine with epinephrine via a misplaced epidural catheter, in a child with previously undiagnosed LQTS.

Case report

A 9-yr-old boy, weighing 29 kg, with Pfeiffer syndrome (one of four acrocephalosyndactyly syndromes characterized by craniosynostosis, mild syndactyly of the hands and feet, and dysmorphic facial features) was born with anal atresia. He underwent surgery for correction of anal atresia and formation of a colostomy immediately after birth. He had several operations in the following years because of complications related to the colostomy. There were no reported problems caused by general anaesthesia during these operations. His latest admission was to take down the colostomy. The patient’s medical history was otherwise unremarkable. After being premedicated with oral midazolam 0.3 mg kg⁻¹, the patient was taken to the operating room. Standard monitors (three-lead ECG, pulse oximeter and arterial pressure cuff) were placed and the patient was induced with sevoflurane. One minute after the first dose of epidural bupivacaine 0.25% 3 ml with epinephrine 1:200 000 epidurally, after negative aspiration for blood. Approximately 3 min later and before surgical incision, the heart rhythm suddenly changed into ventricular tachycardia and a few seconds later into ventricular fibrillation and cardiac arrest. After a precordial thump, chest compressions were started. The desflurane was discontinued and the patient was ventilated with oxygen 100%. A single i.v. dose of epinephrine 1 mg was administered with immediate return to sinus rhythm, followed by sodium bicarbonate 30 mEq. The heart rate rose to 180 beats min⁻¹ and the arterial blood pressure to 150/100 mm Hg. A few seconds later, the patient had a second episode of ventricular fibrillation that resolved after brief chest compressions. The heart rate stabilized around 100 beats min⁻¹, and arterial pressure stabilized with systolic values around 90 mm Hg. Blood samples were taken for measurement of plasma bupivacaine concentration. Arterial blood gas was normal, with the exception of hypokalaemia (potassium 2.8 mmol litre⁻¹), probably because of the recent administration of bicarbonate. The patient was then extubated and taken to the cardiac intensive care unit. Before removal, we again aspirated the epidural catheter with a smaller syringe, and at this time we noticed a free return of blood. Repeated ECG in the first 24 h after the cardiac arrest revealed sinus rhythm and no evidence of Wolff–Parkinson–White syndrome. The QT interval appeared borderline normal, but the corrected QT was constantly increased (Table 1). An exercise test performed 48 h later showed a QT that became increasingly prolonged to 516 ms without ectopies (Table 1). The echocardiogram showed normal anatomy and a shortening fraction of 32%. The patient was started on the β-blocking agent nadolol, which decreased the resting heart rate from 88–110 to 58–70 beats min⁻¹.

The plasma concentration of bupivacaine 2 min after the cardiac arrest was 0.48 μg ml⁻¹. Cardiac and neurological signs of bupivacaine toxicity are reported in the adult population at total plasma bupivacaine concentrations of >4 μg ml⁻¹.¹³

Table 1 QT, corrected QT (QTc) and heart rate (HR) from ECGs recorded 30 min and 4, 20 and 48 h (on exercise testing) after the cardiac arrest

<table>
<thead>
<tr>
<th>Time after cardiac arrest</th>
<th>30 min</th>
<th>4 h</th>
<th>20 h</th>
<th>48 h</th>
</tr>
</thead>
<tbody>
<tr>
<td>QT (ms) (normally 340–430)</td>
<td>324</td>
<td>382</td>
<td>322</td>
<td>516</td>
</tr>
<tr>
<td>QTc (ms) (normally &lt;440)</td>
<td>483</td>
<td>480</td>
<td>457</td>
<td>560</td>
</tr>
<tr>
<td>HR (beats min⁻¹) (normally 60–100)</td>
<td>134</td>
<td>95</td>
<td>114</td>
<td>71</td>
</tr>
</tbody>
</table>

<https://academic.oup.com/bja/article-abstract/90/6/804/268326>
The direct family members (father, mother and one brother) were interviewed and asked to undergo exercise stress tests, which were all negative. There were no episodes of unexplained sudden death in the family, or events that could suggest a family history of LQTS.

**Discussion**

Intraoperative cardiac arrest is a rare event. The most recent survey on anaesthesia-related cardiac arrest in children (POCA Registry) reports an incidence of 1.4 per 10,000 cases. Thirty-two per cent of these episodes are secondary to cardiovascular problems and only 3% are attributable to arrhythmias. The reported incidence of arrhythmias after inadvertent systemic injection of local anaesthetic following intravascular migration of an epidural catheter is only 0.4%. It is well known that the methods commonly used to detect systemic injection of local anaesthetic, such as aspiration and return of blood, and evaluation of the heart rate and T-wave changes after a test dose, can fail, as documented in this case report. Fisher and colleagues showed that, even after combining detection of changes in T-wave amplitude (increase by 25%) with any alteration in the rhythm, it is possible to miss 3% of inadvertent systemic injections. We do not know why the initial test dose did not result in any significant haemodynamic or ECG changes in our patient. There is a possibility that the initial dose of epinephrine (10 μg), although within the recommended range in children (0.25–0.5 μg kg⁻¹), was too low to trigger any significant change. It is also possible that the epidural catheter migrated after the patient was repositioned on the operating table. Bupivacaine-induced arrhythmias include PR and QT interval prolongation, QRS widening, atriovenous block, ventricular tachycardia and ventricular fibrillation. We observed most of these electrocardiographic changes in our patient. Because of the short interval between the appearance of arrhythmias and the initial bolus of bupivacaine, we immediately suspected an intravascular injection of the local anaesthetic and did not administer further doses of bupivacaine. However, the bupivacaine concentration immediately after cardiac arrest was within normal limits. These plasma concentrations were significantly lower than those associated with neuro- and cardiotoxicity in adults. The patient received only bupivacaine 0.25 mg kg⁻¹, and several studies in healthy adults have shown that the i.v. administration of higher doses of bupivacaine (30–45 mg) over a short period (5–10 min) is well tolerated and followed by minimal cardiovascular effects. The subsequent discovery of a prolonged QT interval in this patient could explain the cardiac events. They could have been triggered by the small dose of epinephrine (15 μg given in conjunction with bupivacaine) after the epidural catheter had probably migrated intravascularly. Intraoperative torsades de pointes have been described in a child with LQTS after injection of a small dose (4 ml) of epinephrine (1:100,000) by infiltration for haemostasis. Arrhythmias in patients with LQTS are often triggered by physical exertion, emotional events or situations characterized by a high sympathetic discharge. Epinephrine is one of the drugs that should be avoided in patients with LQTS, and the treatment of this syndrome is mostly based on anti-adrenergic agents, such as β-blockers. We suspect that the accidental intravascular injection of epinephrine, which was combined with bupivacaine, after migration of the epidural catheter may have triggered the arrhythmia.

Long QT syndrome is a congenital or acquired disorder characterized by abnormal prolongation of ventricular repolarization, measured as lengthening of the QT interval on any of the 12 ECG leads. Other ECG abnormalities include bradycardia, increased QT dispersion and T-wave alterations. These abnormalities cause predisposition to ventricular tachyarrhythmias, such as polymorphic ventricular tachycardia and ventricular fibrillation. Acquired QT prolongation is most often attributable to the administration of drugs or electrolyte imbalance. There are two known congenital forms of LQTS, the Romano–Ward syndrome (autosomal dominant trait) and the Jervell and Lange-Nielsen syndrome (autosomal recessive inheritance, associated with congenital deafness). The pathophysiology of cardiac arrhythmias in these patients is related to impairment of outward potassium currents or to defective sodium channel activation. Both mechanisms result in reduced outward current during repolarization, with secondary prolongation of cardiac action potentials and lengthening of the QT interval. Our patient had been diagnosed with Pfeiffer syndrome in infancy because of craniosynostosis with minimal dysmorphic facial features. However, there are no reports of cardiac arrhythmias associated with Pfeiffer syndrome, and the patient did not have any of the other anomalies associated with this syndrome.

A screening ECG in every child undergoing general anaesthesia seems inappropriate, given the rarity of LQTS and the high incidence of a normal ECG at rest in patients with this congenital disorder. However, because of the potential for intravascular injection of the mixture of local anaesthetic and epinephrine during any regional anaesthesia technique, anaesthetists should consider the risks involved in performing central and peripheral nerve blocks in patients with known LQTS or a family history of LQTS. If it is decided to use a regional technique, anaesthetists should probably use a solution containing plain anaesthetic rather than a combination of local anaesthetic and epinephrine.

In conclusion, this case report confirms the difficulty of recognizing the intravascular migration of an epidural catheter. Other reasons besides local anaesthetic toxicity may be responsible for intraoperative cardiac arrest, and a full cardiac evaluation should be obtained in such cases.

**References**

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