by our wide range of interval odds ratios. A prospective multimodal evaluation would potentially further elucidate the determinants of this practice variation, an issue that our study was able to highlight.

**Declaration of interest**

None declared.

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Paroxysmal extreme pain disorder

Editor—We report the successful anaesthesia of a 15-yr-old girl (Miss A) with paroxysmal extreme pain disorder (PEPD) for laparoscopic ovarian cystectomy.

PEPD is a rare condition caused by inherited sodium channel abnormalities. There are a variety of clinical manifestations (Table 1). Sudden attacks of severe pain lasting seconds to hours occur, usually in characteristic distributions, namely rectal, ocular, and submaxillary. Flushing accompanies the pain, with harlequin skin changes in some patients (suddenly developing areas of sharply demarcated erythema). These attacks can escalate to seizures, hypertonia, respiratory arrest, and even asystolic cardiac arrest.1

Explanation of the constellation of symptoms is found in the identified sodium channel abnormalities. The majority of affected patients have mutations in the SCN9A gene. There is an autosomal-dominant abnormality in a voltage-gated sodium channel, Na+,1.7.2 This leads to hyperexcitability in the affected neurones. Na+,1.7 is found in the dorsal root and sympathetic ganglia. It is particularly highly expressed in nociceptors.3 The effects of hyperexcitability in these areas correlate to the clinical picture.

Miss A is unusual among PEPD patients in that she had had multiple episodes of cardiorespiratory arrest requiring resuscitation. The longest documented asystolic episode has been 45 s. Owing to these episodes, she has, from infancy, received 24 h care from an advanced life support provider. Bag-mask ventilation has occasionally been described as very difficult because of severe laryngospasm and hypertonia.

The anaesthetic plan was for conventional i.v. induction undertaken with particular attention to avoidance of provoking undue anxiety in Miss A. Emergency drugs were drawn up and all resuscitation equipment was checked and kept nearby but out of sight of the patient.

Routine monitoring was established before anaesthesia. A 22 G cannula was in situ and functioning. Preoxygenation was followed by induction of anaesthesia with fentanyl 100 mg, propofol 150 mg, rocuronium 40 mg, and glycopyrrolate 200 mg. Intubation was achieved easily with a Cormack and Lehane Grade 1 view. Anaesthesia was maintained with desflurane in an oxygen/air mix. An 18 G cannula was sited under anaesthesia. The siting of a larger cannula before induction of anaesthesia had been considered a significant risk of triggering an attack of PEPD.

Analgesia and antiemesis was provided by paracetamol 1 g, paracoxib 40 mg, dexamethasone 8 mg, and morphine 5 mg. Oxygen saturations were kept at 97% and above with an FiO2 of 0.5. Haemodynamic stability was maintained without support and 1 litre of Hartmann’s solution was administered over the course of the surgery. The procedure lasted ~45 min. A 6 cm haemorrhagic cyst was removed from the right ovary.

At the end of surgery, neostigmine 2.5 mg/glycopyrrolate 0.5 mg was given after confirmation of the presence of four twitches on train-of-four monitoring. Extubation was uneventful.

We feel cutaneous evidence of an attack at the end of a procedure and general anaesthetic should prompt consideration of continuing anaesthesia until the attack has resolved. The patient should be made as pain free as possible before emergence from anaesthesia is considered.

The patient remained stable and comfortable in the recovery room. Although considered before operation, a patient-controlled analgesia with morphine was not required for postoperative pain relief. The remainder of Miss A’s hospital stay was also uneventful; she has been seen in clinic after discharge and was fully recovered.

Although uneventful, we felt that this case was of interest due to the rarity of the condition and the potentially serious adverse outcomes including sudden cardiac arrest. There is no established guidance for anaesthesia in these patients reported in the anaesthetic literature. Conventional but meticulous anaesthesia with particular attention to minimizing distress and anxiety proved successful.

Table 1 PEPD signs and symptoms1

| Rectal, ocular, and jaw pain |
| Flushing |
| Limb swelling |
| Tonic non-epileptic seizures |
| Pupillary changes |
| Salivation |
| Apnoea |
| Asystole |
Continuous perineural infusion after lower extremity osteotomies in children: a feasibility and safety analysis

Editor—We wish to report our experience using continuous perineural infusions (CPNI) for postoperative analgesia in a paediatric population undergoing elective lower extremity osteotomies. While the use and safety of regional anaesthesia during postoperative care of paediatric patients undergoing orthopaedic procedures has been reported, placement of CPNI in children undergoing elective osteotomies of the lower extremity, which may have a higher postoperative risk for acute compartment syndrome (ACS), remains controversial.

After obtaining IRB approval, we reviewed our regional anaesthesia database and identified patients who had a CPNI placed as part of their care for lower extremity osteotomy. From March 2003 to February 2011, 222 patients had a total of 343 peripheral nerve blocks (PNBs) performed for postoperative pain management after lower extremity osteotomies. The mean (range) age was 12.9 (3–18) yr. Of the 343 blocks performed, 82 were single-shot nerve blocks (SS) and 261 were CPNI. Figure 1 provides a breakdown of the type and location of the blocks performed. Surgical characteristics of the patients revealed that 47% (105/222) had osteotomies of the foot, ankle, or both, while 53% of patients (117/222) underwent long-bone osteotomy (femoral, tibial, or fibular) for deformity correction. Block failure was recorded in 14 children (6%) with the absence of sensory block in eight patients and catheter dislodgement in six patients. Despite our use of dilute local anaesthetics (typically 0.1–0.2% ropivacaine or 0.125–0.25% bupivacaine), we had a 22% incidence of motor block (74/343 blocks). Sciatic nerve block (90%) represented the majority of motor blocks observed. All patients were followed acutely by the orthopaedic and pain service until catheter removal and block resolution, and then long term in the orthopaedic clinic. We did not identify any episodes of missed or delayed ACS, nerve injury, persistent numbness, or paraesthesia during follow-up.

Foot, ankle, and lower extremity long-bone osteotomies for deformity correction are common orthopaedic procedures in children and adolescents. While the use of regional anaesthesia is an attractive idea, this population may be at increased risk of postoperative compartment syndrome. There have been a number of case reports of compartment syndrome in the setting of neuraxial anaesthesia and CPNI, which may give paediatric anaesthesiologists and orthopaedic surgeons reason to pause when considering whether or not to utilize regional anaesthesia in these surgical populations. Review of our experience caring for patients undergoing lower extremity osteotomies with regional anaesthesia reveals a high success rate (94%) and a low complication rate. While actual invasive measurements of compartment pressures were not performed, none of the patients in our series revealed any postoperative symptoms of increased compartment pressures that warranted needle measurement of pressures.

In addition to the limitations inherent to retrospective studies, we provide no actual documentation of objective compartment pressures in any of the patients. However, given the absence of any clinical signs of increased compartment pressure after operation, the actual needle measurement of pressures in this cohort of children purely for research purposes was unacceptable. Additionally, even with 222 patients in this study, it is likely that the incidence of adverse events could have been underestimated due to their rarity. Despite these limitations, our data demonstrate

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Declaration of interest

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Fig 1 Location and types of PNB performed for management of foot, ankle, and long-bone osteotomies over an 8 yr period. Stim cath, stimulating catheter; Non stim cath, non-stimulating catheter.