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Case Report

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Between Seizures and Signals: Spine Surgery in Dravet Syndrome : A Case Report

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Abstract

Background
Dravet syndrome is a rare developmental and epileptic encephalopathy associated with a high susceptibility to seizures and specific perioperative challenges. Major spinal surgery requires effective analgesia and reliable neurophysiological monitoring, both of which may require adaptation in patients with Dravet syndrome.
Case presentation
An 18-year-old man with genetically confirmed *SCN1A*-related Dravet syndrome and pharmacoresistant epilepsy underwent posterior spinal fusion for progressive neuromuscular scoliosis. Because of concerns regarding sodium-channel blockade, surgical-site infiltration with local anaesthetics was omitted and intrathecal morphine was used as the main component of postoperative analgesia. Intraoperative spinal cord monitoring was also modified: somatosensory evoked potentials were maintained, whereas transcranial motor evoked potentials were omitted following multidisciplinary discussion because of concern that transcranial electrical stimulation could precipitate seizures. Total intravenous anaesthesia with propofol and remifentanil was used. Despite substantial blood loss and delayed emergence, no perioperative seizures or new neurological deficits occurred, and postoperative analgesia was satisfactory.
Conclusions
Major spinal surgery in patients with Dravet syndrome may require individualized adaptation of both analgesic and neurophysiological monitoring strategies. This case illustrates the importance of multidisciplinary assessment when the benefits of standard perioperative techniques must be balanced against seizure susceptibility.

Keywords

Dravet syndrome; *SCN1A*; scoliosis; spinal fusion; case report; intrathecal morphine; somatosensory evoked potentials; motor evoked potentials

Background

Dravet syndrome is a rare developmental epileptic encephalopathy, commonly associated with pathogenic variants in the *SCN1A* gene and characterized by pharmacoresistant seizures linked to cognitive and motor impairment[1]. Although anaesthetic management of patients has been previously described, to our knowledge, no previous report has addressed its perioperative management during major spinal deformity surgery. We report the anaesthetic management of a patient with Dravet syndrome undergoing posterior spinal fusion, highlighting the problems related to intraoperative neurophysiological monitoring and postoperative analgesia.

Case presentation

An 18-year-old man (158 cm, 44 kg) with genetically confirmed Dravet syndrome (pathogenic *SCN1A* mutation) was scheduled for elective posterior spinal fusion T3-L4 for progressive neuromuscular scoliosis. Dravet syndrome had been diagnosed at approximately 1 year of age following febrile seizures. His clinical course was

characterized by epileptic encephalopathy, generalized hypotonia, motor impairment, moderate intellectual disability and pharmacoresistant epilepsy despite administration of multiple anticonvulsant medications.

Chronic treatment included valproate (450 mg morning and 300 mg evening), stiripentol (250 mg twice daily), clobazam (10 mg twice daily), bilastine (20 mg once daily), methylphenidate (20 mg morning), trazodone (25 mg evening), and buccal midazolam (10 mg) as rescue medication.

Preoperative laboratory testing revealed mild isolated hypofibrinogenemia, considered by the haematology team to be related to valproate therapy and possibly potentiated by concomitant stiripentol. Repeated analysis before surgery showed blood test normalization.

Standard monitoring, frontal electroencephalographic monitoring (NeuroWave®), quantitative neuromuscular monitoring (TOF-Watch®), and peripheral intravenous access were established before anaesthesia induction. General anaesthesia was induced and maintained with target-controlled infusions of propofol (Eleveld model) and remifentanil (Minto model), with initial effect-site concentrations of 2 $\mu g \cdot mL^{-1}$ and 2 $ng \cdot mL^{-1}$, respectively, subsequently adjusted according to clinical requirements. Rocuronium (30 mg) was administered to facilitate tracheal intubation. Cefazolin and tranexamic acid were administered after induction.

Intrathecal morphine (160 µg) was administered at the L3–L4 interspace. A radial arterial catheter, urinary catheter, rectal temperature probe and cell salvage system were subsequently placed. Intraoperative neurophysiological monitoring consisted of electroencephalography and somatosensory evoked potentials (SSEPs). Following multidisciplinary discussion, motor evoked potentials were not used because the treating neurologist considered that transcranial electrical stimulation could precipitate seizures in this patient with pharmacoresistant Dravet syndrome. Surgery was initiated after complete recovery of neuromuscular transmission (train-of-four ratio 100%) and no additional neuromuscular blocking agent was administered.

The patient lay in prone position, and core temperature ranged from 35.1°C to 37.4°C and forced-air warming was used. Haemodynamic stability was supported with norepinephrine up to 0.08 $\mu g \cdot kg^{-1} \cdot min^{-1}$.

Surgery lasted for 4 hours, and total anaesthesia time was 9 hours and 36 minutes, including a delayed emergence. Haemoglobin decreased from 11.6 $g \cdot dL^{-1}$ preoperatively to 8.3 $g \cdot dL^{-1}$ at the end of surgery (estimated blood loss was 2.3 L). Intraoperative fluid therapy consisted of 4 L of crystalloid, 500 mL of 5% albumin, and 535 mL of salvaged autologous blood which was reinfused.

The patient was extubated in the operating room approximately 2 hours after discontinuation of anaesthetic agents and remained haemodynamically stable without clinical seizures. On postoperative day 1, he was easily arousable but somnolent. Electroencephalography showed no epileptiform activity. Neurological examination revealed no new deficits. Intravenous paracetamol and ketorolac provided satisfactory analgesia, allowing mobilization to a chair on postoperative day 1. Trazodone and methylphenidate were withheld because of persistent somnolence. No postoperative neurological complications occurred, and the patient was discharged home on postoperative day 7.

~1 year of age	Dravet syndrome diagnosed following febrile seizures; pathogenic *SCN1A* mutation identified
Preoperative assessment	Progressive neuromuscular scoliosis requiring T3–L4 posterior spinal fusion; mild isolated hypofibrinogenae
Shortly before surgery	Fibrinogen normalised without corrective treatment
Day of surgery	TIVA with propofol/remifentanil; intrathecal morphine 160 µg; SSEPs monitoring; TcMEPs omitted after multidisciplinary discussion
End of surgery	Estimated blood loss 2.3 L; delayed emergence; no clinical seizure
~2 h after anaesthetic discontinuation	Extubated in the operating room
Postoperative day 1	Somnolent but easily arousable; EEG without epileptiform activity; no new neurological deficit; satisfactory analgesia; mobilised to chair
Postoperative day 7	Discharged home without postoperative neurological complications

Discussion and Conclusions

Anaesthetic management of a patient with Dravet syndrome is challenging because perioperative factors including hyperthermia, physiological stress, interruption of antiseizure medications and exposure to potentially proconvulsant drugs may precipitate seizures[2]. In the present case, these considerations were particularly relevant because prolonged posterior spinal fusion requires general anaesthesia, management of substantial blood loss and the use of intraoperative neurophysiological monitoring.

Postoperative analgesia also required adaptation of our usual practice. In our institution, posterior spinal fusion is routinely accompanied by surgical-site infiltration with a local anaesthetic. In this patient, local anaesthetic infiltration was deliberately omitted because of theoretical concerns regarding sodium-channel blockade in the setting of *SCN1A*-related Dravet syndrome. Sodium-channel-blocking drugs are known to aggravate seizures in Dravet syndrome, and a generalized tonic-clonic seizure following intravenous lidocaine has previously been reported in a child with Dravet syndrome[3]. Although these observations do not establish a contraindication to local anaesthetics, we elected to avoid their use in this patient with pharmacoresistant epilepsy. Intrathecal morphine was therefore used as an alternative strategy to provide prolonged postoperative analgesia in combination with postoperative paracetamol and ketorolac. This approach provided satisfactory analgesia without the need for additional systemic opioids.

Total intravenous anaesthesia with propofol and remifentanil was selected to facilitate reliable SSEPs monitoring. Volatile anaesthetics can suppress SSEPs amplitude and increase latency, whereas propofol-based anaesthesia provides more favourable conditions for SSEPs recording during scoliosis surgery[4]. The question was particularly relevant in our patient because SSEPs constituted the primary modality for spinal cord monitoring after the decision to omit transcranial motor evoked potentials (TcMEPs).

The decision to omit TcMEPs monitoring represented an important trade-off in the present perioperative management. Combined SSEPs and TcMEPs monitoring is commonly used during scoliosis correction because the two modalities assess complementary spinal cord pathways[5]. TcMEPs require transcranial electrical stimulation which can induce seizures. Although this complication appears to be uncommon (reported 0.2% incidence during orthopaedic spine surgery), potential consequences warranted particular consideration in our patient suffering pharmacoresistant Dravet syndrome[6]. After multidisciplinary discussion, and on the recommendation of the patient's neurologist, we therefore elected to avoid TcMEPs stimulation and thereby to rely on SSEPs monitoring only. It is worth noting that epilepsy should not be considered an absolute contraindication to TcMEPs monitoring, hence our approach should not be generalized to all patients with epilepsy. Rather, this case illustrates the need for an individualized risk-benefit assessment when neurological benefit of multimodal spinal cord monitoring must be weighed against the patient's underlying susceptibility to develop seizures.

Core temperature was continuously monitored because hyperthermia may trigger seizures in Dravet syndrome[2]. Haemostatic management represented an additional concern. Valproate therapy has been associated with coagulation abnormalities, including thrombocytopenia, impaired platelet function, acquired von Willebrand disease, factor XIII deficiency and hypofibrinogenemia[7]. Although fibrinogen spontaneously normalized before surgery and no corrective treatment was required, identification of this abnormality was particularly relevant given the anticipated haemorrhagic risk of an extensive posterior spinal fusion (our patient’s blood loss of 2.3 L). The present case emphasizes the importance of preoperative haemostasis assessment in patients treated with valproate before major surgery associated with substantial blood loss.

Finally, emergence was delayed for approximately 2 hours after discontinuation of propofol and remifentanil. Postoperative EEG showed no epileptiform activity, making ongoing seizure activity an unlikely explanation for the delayed awakening. The patient's extensive anticonvulsant polytherapy may have contributed to delayed recovery. Particularly, stiripentol inhibits clobazam metabolism and increases exposure to clobazam and its active metabolite, N-desmethylclobazam[8]. Consequently, potential pharmacokinetic interactions should be kept in mind when delayed emergence occurs in patients with Dravet syndrome receiving multiple anticonvulsant medications.

The main limitation of this report is its single-patient nature, which precludes generalisation of the perioperative strategy described. In particular, the decision to omit TcMEPs monitoring and avoid local anaesthetics was based on an individualised risk-benefit assessment and should not be interpreted as evidence that these techniques are contraindicated in patients with Dravet syndrome.

In conclusion, the present case illustrates the competing considerations involved during major spine surgery in a patient with Dravet syndrome. Perioperative management requires minimization of potential seizure triggers while preserving reliable spinal cord monitoring, the anticipation of treatment-related coagulation abnormalities, and ensuring effective postoperative analgesia. In particular, the decision to omit TcMEPs highlights the importance of individualized multidisciplinary assessment when the benefits of standard neurophysiological monitoring must be balanced against seizure susceptibility.

List of abbreviations

EEG: Electroencephalography
SSEPs: Somatosensory evoked potential
TcMEPs: Transcranial motor evoked potential
TIVA: Total intravenous anaesthesia
TOF: Train-of-four

Declarations

Ethics approval and consent to participate
Not applicable. This manuscript describes a single clinical case and does not report research involving human participants requiring ethics committee approval.

Consent for publication
Written informed consent for publication of the clinical details contained in this case report was obtained from the patient's legal representative.

Availability of data and materials
All data relevant to this case are included in the article. Additional data are not publicly available in order to protect patient confidentiality.

Competing interests
The authors declare that they have no competing interests.

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Authors' contributions
BM conceived the case report, reviewed the literature, interpreted the clinical data, and drafted the manuscript. RZ contributed to the anaesthetic and perioperative management of the patient and critically reviewed the manuscript. MAD contributed to the conception of the case report, supervised its preparation, and critically revised the manuscript for important intellectual content. SFS was involved in the neurological management of the patient and contributed to the perioperative neurological and neurophysiological monitoring strategy. SV performed the surgical procedure and contributed to the surgical management of the patient. PL contributed to the anaesthetic and perioperative management of the patient and critically reviewed the manuscript. All authors critically reviewed the manuscript and read and approved the final version.

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