Schwartz–Jampel syndrome: Is risk of malignant hyperthermia the same as that of the general population?

Sir,
I read with interest the case report, “A successful anesthetic approach in a patient with Schwartz–Jampel syndrome (SJS)” by de Oliveira Camacho et al. However, I wish to point out that their conclusion that patients with SJS are at increased risk of malignant hyperthermia (MH) is not supported by the scientific evidence. Similar to the previous authors reporting anesthetic management of SJS cases, they based their conclusion on the case report by Seay and Zifer published in 1978. Seay and Zifer described a 23-month-old patient with SJS booked for cleft palate repair whose temperature rose by 1.5°C after induction of general anesthesia with ketamine (40 mg intramuscular [im]), nitrous oxide, atropine (0.2 mg im), and curare (1.5 mg intravenous). Her pulse rate also increased from 140 to 200 beats/min. Her temperature returned to preoperative values within 30 min without any treatment.

It is noteworthy that the patient was not exposed to suxamethonium and any volatile anesthetic agent, known MH-triggering agents. An in vitro muscle contracture test was not done afterward, although it is considered by some authorities not to be fully sensitive in children aged <10 years. It should also be noted that Parness et al., after carrying out a detailed review of the physiology of muscle excitability and excitation–contraction coupling, as well as the pathophysiology of MH and the myotonias, and the clinical literature on which the claims of MH susceptibility are based concluded that SJS patients have a risk of developing MH that is equivalent to that of the general population. Indeed, this is supported by the case report of a boy who had multiple anesthetics with sevoflurane with no evidence of MH. The authors however recommended that suxamethonium should be avoided in patients with SJS because of the risk of causing dangerous hyperkalemia.

In summary, the association of SJS, a rare chondrodystrophic myotonic syndrome, with MH in the literature is based on one case report in which no triggering anesthetic agents were administered to the patient. Recent scientific evidence suggests that no such association exists. Suxamethonium is however contraindicated in these patients.

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There are no conflicts of interest.

Michael Olusegun Ayeko
King Faisal Specialist Hospital and Research Centre,
Riyadh 12713, Saudi Arabia

Address for correspondence:
Dr. Michael Olusegun Ayeko,
King Faisal Specialist Hospital and Research Centre, Zahrawi Street,
Al Maaither, Riyadh 12713, Saudi Arabia.
E-mail: moayeks@doctors.org.uk

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Bilateral ultrasound-guided erector spinae plane block for postoperative analgesia in choledochal cyst resection surgery

Sir,

Choledochal cyst resection is not an uncommon surgery in pediatric age group. Opioids and epidural analgesia is mainstay for postoperative pain relief after the surgery. However, with opioids as these children are extubated and breathing spontaneously great care must be taken to avoid respiratory depression. Epidural analgesia is effective when properly placed and closely monitored. Nevertheless, a number of complications have been reported with epidurals. They include bradycardia, ventricular and atrial ectopics, transient apnea, and leakage around the epidural catheter.

[1]

The newly described technique ultrasound-guided erector spinae plane block (US-ESP) is a novel technique that anesthetizes the dorsal rami, ventral rami, and rami communicantes of the spinal nerves.

[2]

We report here the use of bilateral US-ESP for a pediatric case being operated for Choledochal cyst excision surgery. A 9-year-old 25 kg 140 cm male child posted for excision of choledochal cyst and hepaticoduodenostomy. Routine investigations were within normal limits. The ultrasound report showed there is fusiform dilation of extrahepatic bile duct with mild right and left hepatic duct. We planned bilateral US-ESP and general anesthesia for the patient. General anesthesia was induced with sevoflurane, fentanyl (2 μg/kg), and rocuronium (0.8 mg/kg). After the induction, we performed the bilateral US-ESP blocks. The ESP block was performed as follows the patient was placed in the left lateral position, and a high-frequency linear ultrasound transducer was placed in a longitudinal orientation 1 cm lateral to the thoracic sixth spinous process. The deep plane to the erector spinae muscle was identified. The ESP block was administered by injection of 0.25% bupivacaine 30 mL (15 mL injected into each side) into the fascial plane below erector spinae muscles [Figure 1]. Anesthesia was maintained with sevoflurane, fentanyl, and rocuronium. Intraoperative period was eventless, and no additional analgesic was given. The surgery lasted for 3 h and patient was extubated on operating table. Following extubation and transfer to the postanesthesia care unit, the patient was noted to have a Wong–Baker FACES pain rating scale score of 2 (consistent with mild pain). Postoperative analgesia was initiated with intravenous tramadol 2 mg/kg and intravenous acetaminophen 15 mg/kg every 8 h. The pain was assessed every 4 h using the Wong–Bakers FACES. The patient continued to report only mild pain at the surgical site up to 48 h after surgery. US-ESP achieves extensive multidermatomal sensory block of the posterior, lateral, and...