

Bilateral ultrasound transversus abdominis plane block in a patient affected from Menkes disease

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Abstract

This case report demonstrates the utility and safety of ultrasound transversus abdominis plane (TAP) block in a paediatric patient suffering from Menkes disease. Anaesthetists, and particularly paediatric and neonatal anaesthetists, have to make a careful assessment of these patients, who are subjected to frequent surgeries, despite their tender years. These operations are often necessary in order to improve the patient's quality of life as much as possible. The choice of anaesthetic technique must take the particular nature of the disease and the patient's medical history into consideration, while careful preparation and preoperative evaluations make it possible to administer the general or local anaesthetic safely. Official literature on the subject does not provide sufficient information about the use of local anaesthetic techniques. Moreover, the use of ultrasound TAP block in patients affected by Menkes disease has never been published in literature. In our experience, ultrasound TAP block has provided positive results, although the rarity of this childhood disease makes a detailed study difficult.

Introduction

Menkes disease is due to the mutation of a

gene located on the Xq1 3.3 chromosome, which encodes the ATP-7 A protein that transports copper within cells.¹ Menkes disease and the *occipital horn* disease are allelic. The copper deficiency alters the function of the copper-dependent enzymes and causes the symptoms of this illness, which include the following: retarded intrauterine growth and progressive neurological deterioration, with axial hypotony, spasticity, convulsions and hypothermia in the first months of life. The gliosis leads to the development of microcephaly. The hair has a characteristic phenotype: it is thin, hypo-pigmented, opaque, kinky and fragile. A microscopic examination reveals the presence of kinky hair. Osteoporosis is also present. Aneurysms develop in the long, winding arteries, with irregular lumen, and cause subdural, cerebral and intestinal haemorrhaging. Menkes disease can be diagnosed by measuring copper levels in the serum (low) and the fibroblasts (high). Death generally occurs in early childhood. Various genetic mutations linked to the X chromosome have been identified as being responsible for the illness. Female carriers (who may show signs of partial anomalies in their hair) and infected fetuses (prenatal diagnosis can be made by measuring copper concentrations in chorionic villi or cultured amniocytes) can be identified in families at risk by molecular analysis. The treatment involves parental copper injections. If treatment is started early enough, it can prevent the appearance of the neurological symptoms and life expectancy is increased. The illness affects around 1 in 300,000 live births.

Children affected by this illness often undergo surgery during their short lives in order to improve their quality of life as much as possible. Adequate anaesthetic preparation and the adoption of precautions in order to prevent undesirable complications are therefore necessary.²

Case Report

A 6-year-old male, 21 kg weight, suffering from Menkes disease, was presented for surgical treatment for bilateral inguinal hernia repair. The patient was carefully warmed with a blanket through a forced-air system before the start of anaesthetic procedures.

Analgesedation was induced by an initial intravenous administration of fentanyl 2 mcg/kg and propofol 3.0 mg/kg followed by the introduction of a proseal laryngeal mask airway (PLMA). No muscle relaxants were administered. Analgesedation was maintained by a continuous infusion of 80 mcg/kg/min propofol and 0.10 mcg/kg/min remifentanyl with Air/O₂ mixture (FiO₂=0.4) leaving the child spontaneously breathing through the PLMA.

Monitoring of vital parameters was carried

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out through electrocardiograph, pulse oximetry and non-invasive blood pressure measurement. After positioning the patient in a supine decubitus and skin disinfection, a 21 G 50 mm atraumatic needle, the same usually used as electro-neuro-stimulation, was placed for bilateral ultrasound transversus abdominis plane (TAP) block. A portable system with a linear array 10 MHz transducer was used. The probe was placed in a transverse plane to the lateral abdominal wall in the midaxillary line between the lower costal margin and iliac crest and an ultrasound TAP block was performed in real time observing the spread of the local anaesthetic in the plane between the internal oblique and transversus abdominis muscles targeting the spinal nerves in this plane. Levobupivacaine 0.25% at a dosage of 0.3 mL mg/kg was administered in each side.

The surgical procedure was performed without complications and all haemodynamic and respiratory parameters were always within the normal range. At the end of the surgical procedure the consciousness recovery was immediate and during the postoperative time no rescue analgesic drugs were necessary as the child had no pain.

Discussion

The problem of temperature regulation is particularly significant in this type of patient due to the cytochrome C oxidase deficiency

which leads to dangerous hypothermia. For these reasons we decided to warm the patient using blankets with a forced-air system. The neuromotor response appears to be another important anomaly in Menkes disease, scarcely coordinated and axial hypotonia and hypertonia of the extremities is often present, meaning that careful dosage of the curares and their antagonists is of fundamental importance. Neuromuscular monitoring with a neurostimulator and a reduction of the curare dosage is recommended.

During general anaesthesia critical increases in blood pressure due to the aforementioned vascular anomalies, which primarily involve the central nervous system and the intestines, should be avoided. Adequate cerebral perfusion can also be obtained by avoiding hypercapnia and hypoxemia, balancing the hydro-electrolytic equilibrium. The anaesthetic can be administered by inhalation or intravenously. In the first case sevoflurane appears useful and easy to handle, while in the second case the administration of pentothal seems to offer the advantage of causing cerebral vasoconstriction and reducing cerebral oxygen consumption, protecting the cerebral tissue from ischaemic episodes.

Hypoglycaemia, hypothermia and instability of the autonomous nervous system are the consequences of a reduction of the circulating levels of norepinephrine and the reduced adrenergic response. These alterations can be corrected through the administration of glucose solutions and sympathomimetic amines, especially in the case of marked falls in blood pressure.

The ultrasound TAP block performed in our patient allowed us to avoid general anaesthesia, orotracheal intubation and all the above described risks of complications that occur when general anaesthesia is performed.

Moreover, children suffering from Menkes disease often are affected by gastroesophageal reflux, which requires particular attention during intubation in order to prevent the risk of inhalation. Our patient maintained spontaneous ventilation through a PLMA so orotracheal intubation was unnecessary for reducing this risk.

Recently, the ultrasound TAP block has gained popularity for intraoperative and postoperative pain management in a variety of abdominal surgical procedures in adult, pediatric and neonatal patients.^{3,4} The abdominal wall consists of three muscle layers: the external oblique, the internal oblique and the transversus abdominis and their associated fascial sheath. This muscular wall contains the T7-T12 intercostal nerves, the ilioinguinal and iliohypogastric nerves and the lateral cutaneous branches of the dorsal rami of L1-L3. The above nerves run in a neurovascular plane between the internal oblique and transversus abdominis muscles and represent the *target* of local anesthetics. Anyway it has been demon-

strated that anatomical variants exist and this aspect is highly individual. For these reasons the traditional landmark-guided technique involving needle insertion at the triangle of Petit was often accompanied by a high rate of failure.^{5,6} The goal of achieving the excellent level of analgesia produced through ultrasound TAP block is due to a great precision during the anaesthetic administration and the progressive spread of the local anaesthetic over several hours. Another benefit is the absence of motor block that represents an advantage to patients affected from Menkes disease.

Concerning technique we used a portable system with a linear, 5-10 Mhz ultrasound probe, placed in a transverse plane to the lateral abdominal wall in the midaxillary line between the lower costal margin and iliac crest. The tip of the needle is positioned between the internal oblique and the transversus abdominis muscle in the fascial layer that separates the two muscle layers.

The administration of levobupivacaine 0.25% at a dosage of 0.3 mL mg/kg was found to be preferable over the older local anaesthetics such as bupivacaine and mepivacaine because it makes it possible to obtain good, longer lasting analgesia and low risks of toxicity of the levobupivacaine. Although the illness affects the central nervous system and vascular alterations are present, we did not come across any problems with the administration of local anaesthetic (although it should always be administered with the due precautions) nor were there any postoperative complications.

Postoperative analgesia was excellent and no rescue analgesic drugs were administered.

One of the most critical complication about TAP block is the possibility of visceral damage if the needle is advanced too far and the local anesthetic is administered out of the target, intraperitoneally for example. Other risks include bowel hematoma, liver laceration and femoral nerve palsy. In addition the spleen and kidney may be accidentally punctured. Ultrasound, together with the use of a fine gauge, blunt-tipped, short-bevel needle can reduce the risk of these complications.

TAP block may be performed with a continuous infusion of local anaesthetics through a catheter that may dislodge causing the administration of the drug to be out its target. Different access point approaches to TAP block can be used: subcostal, mid-axillary and via the lumbar triangle of Petit which depend on different nerves that must be involved. Contraindications to TAP block include tissue infection at the needle insertion site and coagulation disorders.

Official literature on the subject does not contain any comparative data for this anaesthetic technique, although the use of anaesthesia when treating this disease deserves greater study on a broader subject base, made

difficult by the rarity of this illness that affects just 1 in 300,000 live births.

Conclusions

The best technique for dealing with anaesthetic problems in children suffering from Menkes disease requires a knowledge of all its complications. Monitoring body temperature regulation and using new drugs, including the latest generation local anaesthetic with a low neurotoxicity together with echoguided locoregional anaesthesia, are the reference points for correct anaesthetic treatment in these patients. The ultrasound TAP block, with the due precautions, does not cause any problems worthy of note in our experience and his technique has given excellent results in postoperative pain management.^{7,8} Although a broader study group is necessary, it is hindered by the rarity of Menkes disease.

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