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Letter to the editor

Ultrasound-guided Supraclavicular Brachial Plexus Block Under General Anaesthesia for a Child with Conradi-Hünermann Syndrome*

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To the Editor,

We present the case of an 11-year-old girl with Conradi-Hünermann syndrome (CHS), a rare X-linked dominant genetic disorder occurring in approximately 1 in 200,000 births. CHS is caused by mutations in the ARSE gene which disrupt cholesterol biosynthesis, leading to toxic sterol accumulation and primarily characterized by skeletal abnormalities, cardiac and multi-organ involvement, as well as airway complications. It may also present with skin manifestations and cataracts[1,2]. Skeletal deformities like short stature, asymmetric shortening of the limbs, such as the shortened humerus, epiphyseal stippling, scoliosis, and craniofacial abnormalities like flattened face and saddle nose can compromise airway management[2]. A couple of cases reported management of general anaesthesia[3,4]. Yet to our knowledge, the presented case would be the first pediatric CHS scheduled for brachial plexus block under general anesthesia. Therefore, we aimed to present our management of ultrasound (USG) guided supraclavicular brachial plexus block under general anesthesia in a child with CHS scheduled to undergo radial head excision.

An 11-year-old girl weighing 45 kg with CHS was scheduled to undergo radial head excision surgery. The preoperative assessment revealed craniofacial deformities, including a flattened face and saddle nose, which raised concerns about a difficult airway, as well as a shortened right humerus and scoliosis-induced shoulder asymmetry (Fig. 1). Her chief complaint was right elbow pain with a preoperative visual analog scale (VAS) score of 4.

After obtaining signed written informed consent from the parents for publication, standard monitoring, and anesthesia induction were performed with intravenous (IV) 100 mg of propofol and 50 µg of fentanyl. Intubation was facilitated using IV 25 mg of rocuronium via video laryngoscopy to prevent possible anticipated difficult airway complications with an ID of 6 mm cuffed endotracheal tube. General anesthesia was maintained with sevoflurane (2%) in 50% air-oxygen and IV remifentanil infusion (0-0.2 µg.kg-1.min^{-1}). Afterwards, USG-guided (LOGIQ e, GE Healthcare, California, USA) supraclavicular brachial plexus block using 20 mL of 0.25% bupivacaine and infiltration of 5 mL 0.2% bupivacaine around the tourniquet area (Video 1). Additionally, 600 mg of IV paracetamol was administered intraoperatively. The patient's intraoperative course was uneventful, with stable hemodynamics and satisfactory anesthesia. Postoperative pain was evaluated using the Visual Analog Scale (VAS) in the recovery room, and the scores were recorded as 2, 3, and 5 upon arrival at the recovery unit, and at 6 and 12 hours postoperatively, respectively. The patient and her family reported high satisfaction, particularly with the painless first postoperative day. No complications were observed.

An 11-year-old girl having CHS scheduled for radial head excision surgery with brachial plexus block under general anesthesia underwent stable intraoperative surgical anesthesia and successful pain management, resulting in uneventful postoperative recovery. Although the choice of anesthesia and its management can be challenging in children with CHS, tailored approaches employing multimodal strategies might provide

Fig. 1 — (a) Upper images: Craniofacial appearance showing flattened facial features and saddle nose deformity. (b) Lower image: Shoulder region demonstrating scoliosis-induced shoulder height asymmetry and a shortened right humerus.

successful outcomes. General anesthesia with propofol, remifentanil, vecuronium, and sevoflurane in a nitrous oxide-oxygen mixture was performed for bilateral congenital cataract surgery in a 6-month-old boy without any difficulty during intubation[3]. Another congenital cataract surgery was managed under general anesthesia with sevoflurane in nitrous oxide-oxygen, fentanyl, and atracurium in a 4-month-old girl which required re-operation 4 years later[4]. In the present case, extremity surgery was needed instead of eye surgery. Craniofacial bone deformities, such as a flattened face and saddle nose, were examined (Fig.1. (a)). Fortunately, no difficulty with intubation was encountered. There were marked anatomical asymmetries, including scoliosis-induced shoulder height difference and a shortened right arm ([Fig.1. (b)). Although CHS is not directly associated with neuromuscular pathology, thoracic skeletal deformities such as scoliosis may reduce respiratory reserve, raising concerns about postoperative respiratory compromise. Therefore, to minimize opioid requirements, we planned a supraclavicular brachial plexus block under general anesthesia using propofol, remifentanil, rocuronium, and sevoflurane in an oxygen-air mixture. Supraclavicular brachial plexus was conducted under ultrasound guidance. During the block, potential brachial plexus variations were considered and ruled out under ultrasound guidance[5]. This highlights the usefulness of ultrasound in patients with skeletal anomalies, as it allows for more accurate block placement and increases the likelihood of success.

In conclusion, the effectiveness of combining general anesthesia with supraclavicular brachial plexus block via USG for an upper extremity procedure in this particular child with CHS was performed successfully. The current tailored multimodal anesthetic/analgesic approach to rare syndromic surgical cases would be valuable because the use of locoregional techniques can support enhanced analgesia and potentially reduce opioid-related risks. The USG-guided supraclavicular block provides multimodal anesthesia and/or analgesia, particularly in cases with thoracic skeletal deformities associated with reduced respiratory reserve while general anesthesia and airway instrumentation may still be required.

Video link: https://vimeo.com/1112688046?fl=pl&fe=sh

Real-time ultrasound imaging demonstrating the supraclavicular brachial plexus block procedure. The needle is visualized advancing toward the brachial plexus under direct ultrasound guidance, with 0.25% bupivacaine injected around the nerve structures to achieve an effective block.

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