Anesthetic management of CHILD syndrome: Not a child's play!

Dear Editor,

We would like to commend Pappu *et al*. for addressing the dearth of information regarding the perioperative management of a 13-year-old female with CHILD syndrome for syndactyly release and skin grafting.[1]

Disorders of keratinization are characterized by varying degree of scaling of the skin and are mostly inherited (due to genetic mutations encoding certain proteins) but may be acquired (due to malignancy, infections, or nutritional deficiency). Broadly the inherited ichthyosis can be further classified as non-syndromic (with only skin lesions) and syndromic (continuous gene syndromes like Kallmann syndrome, CHILD syndrome and Conradi-Hünermann-Happle syndrome).[2]

CHILD syndrome is a rare X-linked dominant disorder that is fatal in males, so mostly females may present for need of surgical intervention.[3] There is a defect in 3β-hydroxylsterol dehydrogenase enzyme which leads to defective synthesis of steroid hormones, bile acid, and central nervous system hypoplasia.[3] The patients typically present in neonatal period with unilateral skin lesions with ipsilateral abnormalities of the viscera and limb. Atrial and ventricular septum defects may also be associated, and a baseline echocardiogram should be done.

This patient had classical skin lesions but with no systemic involvement and had an uneventful anesthesia.[1] The erythroderma lesions may hamper the protective effect of skin,

make IV cannulation difficult, increase the risk of pressure sores, burns due to cautery and injury to superficial nerves. In addition, extensive cutaneous lesions may lead to increased loss of heat and water and make the children dehydrated and hypothermic.[4] So, adequate padding of pressure points and covering of skin lesions with emollient and dressing is desirable. Also, these children have increased caloric requirement due to increased turnover in the epidermal tissue. So, prolonged fasting should be avoided and early feeding in postoperative period should be considered.

Airway management may be difficult due to associated skeletal deformities like scoliosis, mandibular dysplasia, and abnormalities in ribs. Anesthesiologists must prepare accordingly and have a high index of suspicion for other unexpected findings like high arched palate (present case).

The nails are often keratotic, claw like, which may interfere with pulse oximeter probe placement and its readings.[4] So, advance monitors like Masimo SET® pulse oximetry is preferred (also used by the authors) and alternate sites like ear lobe should be considered in severe onychorrhexis cases.

Due to possibility of neurological defects and skin lesions, regional anesthesia should be carefully considered.[5] Since axillary area was involved, authors avoided brachial plexus block. However, considering the multitude of benefits, appropriate combination of ultrasound guided individual nerve blocks (radial/median/ulnar) could have been used. In the index case, authors have used extradural morphine but followed adequate precautions (ondansetron IV) to reduce its possibility. Opioids should be avoided in central neuraxial block as it may cause pruritus and aggravate skin lesions. If additives are desired, better alternatives like clonidine should be preferred.

In conclusion, CHILD syndrome is a rare disorder, which entails multiple challenges to the anesthetist. Ensuring uneventful perioperative course requires a thorough understanding of the disorder, meticulous preoperative assessment, administration of a safe anesthetic technique while protecting the skin lesions, and scrupulous perioperative monitoring.

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Conflicts of interest
There are no conflicts of interest.

Anju Gupta, Nishkarsh Gupta[1]

Anesthesiology, Pain Medicine and Critical Care, All India Institute of Medical Sciences, [1]Onco-Anesthesiology and Palliative Medicine, DRBRAIRCH, All India Institute of Medical Sciences, Delhi, India

Address for correspondence: Dr. Nishkarsh Gupta, Room No. 139, First Floor, DRBRAIRCH, AIIMS, Delhi, India.
E-mail: drnishkarsh@rediffmail.com

References

1. Pappu A, Kaur M, Gnanadhas JJ, Dawar R, Singh A. Anesthetic considerations of CHILD syndrome. J Anaesthesiol Clin Pharmacol. 2021;37:130-2.
2. Oji V, Tadini G, Akiyama M, Bardon CB, Bodemer C, Bourrat E, *et al*. Revised nomenclature and classification of inherited ichthyoses: Results of the First Ichthyosis Consensus Conference in Sorèze. J Am Acad Dermatol 2010;63:607-41.
3. Bergqvist C, Abdallah B, Hasbani D-J, Abbas O, Kibbi AG, Hamie L, *et al*. CHILD syndrome: A modified pathogenesis-targeted therapeutic approach. Am J Med Genet A 2018;176:733-8.
4. Dyer JA, Spraker M, Williams M. Care of the newborn with ichthyosis. Dermatol Ther 2013;26:1-15.
5. Ramphul K, Kota V, Mejias SG. Child syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 [cited 2020 Feb 25]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK507813.

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