REFERENCES:

Icthyosis means disorder of skin cornification. It is a group of skin conditions characterized clinically by scaling and histopathologically by hyperkeratosis.

Classification

Icthyosis can be classified on the basis of inheritance patterns, clinical features, associated defects, and histopathological findings.

Primary Icthyosis: the skin is predominantly or solely affected, these include:

- Icthyosis vulgaris, acquired Icthyosis, harlequin Icthyosis, recessive x-linked Icthyosis, lamellar Icthyosis and congenital ichthyosiform erythroderma (nonbullous), epidermolytic hyperkeratosis (bullous), ICE, Icthyosis hystrix.

Icthyosis with other systems involvement, these include:

- Netherton syndrome, KID, CHILD, Conradi–Hunermann syndrome, neutral lipid storage disease, Spilgren-Larsson syndrome, Rud syndrome, Refsum syndrome, Ophéme syndrome, ichthyosis follicularis

Palmoplantar keratodermas, hyperkeratosis is limited to palms and soles. They can be hereditary or acquired; diffuse, striate or punctuate; with or without other systems involvement, these may include:

- Unna–thost PPK, Vorner PPK, Mal de meleda, Vohwinkel PPK, Papillon – Lefèvre syndrome, Richner – Hanhart syndrome, Jadassohn – lewsaski syndrome

Definition

ABSTRACT

Moderate to severe Icthyosis can present challenges. The modifications made to monitoring instruments and surgical dressings can provide lessons on how to manage any patient with easily damaged skin such as burn patients.

Our medically challenging case involved a 5 year old 18 kg male with history of Icthyosis being seen for dental rehabilitation including multiple teeth extractions.

The anesthetic considerations for patients with Icthyosis include thermoregulation, elevated risk of dehydration and subsequent electrolyte disturbance, and patient’s elevated risk of infection due to the difficulty of line placement as a result of hyperkeratosis. As a practical matter, however, the most acute issue is the protection of skin and eyes. In our patient’s pre-operative consultation, his parents made many valuable suggestions including the use of preferred dressings such as Vigilon.

Preparation for the case one day prior to surgery involved the modification of monitors. We trimmed the neonatal EKG leads down to only the electronic probe and removal of adhesive bandage from the pulse-oximeter. All adhesives were removed by application of mineral oil on the EKG probes and the pulse-oximeter. The pulse-oximeter was wrapped with Tegaderm with adhesive EKG probes and the pulse-oximeter probe was wrapped with Tegaderm with adhesive pads.

Monitors: non adhesive pulse oximeters, ECG with needle electrodes or non adhesive pads

1. Thermoregulation: Hypo - or hyperthermia.
2. Dehydration.
3. Careful positioning and skin padding
4. Pharmacological considerations
5. Nutritional status due to protein loss
6. Cannulation, careful aseptic techniques, careful fixation, using petroleum jelly gauze, tie or suture down, no tapes
7. Monitors: non adhesive pulse oximeters, ECG with needle electrodes or non adhesive pads
8. Ophthalmology: ointments (Lacrilube), Eyepads for covering
9. Airway: perioral lesions, limited mouth opening, limited neck movement due to hyperkeratosis
10. Chronic steroid use, should be managed with appropriate stress dose
11. Higher risk of infection: due to skin disease and associated immunodeficiency
12. Restrictive lung movement, in severe forms
13. Electrolytes imbalance, esp. hyponatremia
14. Some types with neurological involvement (neuroicthyosis) may be associated with mental retardation, may limit patient’s cooperation
15. Associated prematurity, as in collodion baby

Pulse Oximeter and EKG leads after removal of adhesive and with wrapping

CONSIDERATIONS

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