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Introduction:

Harlequin ichthyosis (HI), the extremely rare, most dramatic form of congenital ichthyosis, is associated with a mutation in the gene for protein ABCA 12 in which thick plates of stratum corneum separated by deep fissures cover the patient at birth. The skin is hard and thick, restricting respiratory movements. Joints are contractured. Death is usually due to hypoventilation and pneumonia, or sepsis from cutaneous infection. The name is derived from deep purple fissures dividing the thickened skin into geometric plaques, giving the appearance of a harlequin's costume.

Case Report:

A 19-month-old male infant with a history of HI presented for circumcision. He had extremely red, thick, cracked and irritated skin which was lubricated with a hydrophilic cream by his mother prior to surgery. IV placement was difficult due to his thick skin, and once access was finally obtained, securing the IV was particularly challenging due to his slippery skin. Tape and tegaderm would not stick properly, so the IV was secured with a gauze dressing. Though the patient had some degree of limited mouth opening, intubation proved to be uneventful. The ETT was carefully secured with extra tape and Tegaderm given the greasy quality of the patient's skin. Though restrictive lung disease is often a concern in harlequin patients, we had no difficulty ventilating our patient. A caudal epidural block was deferred given his history of frequent cutaneous infections and red, irritated skin with multiple fissures present at the caudal site. He tolerated the procedure well. No complications were noted aside from difficult IV placement and the challenge of securing catheters and tubes.

Discussion:

Children with HI possess unique clinical features that introduce a number of anesthetic challenges. Because treatment is aimed at reducing or softening the scale by applying lubricants or keratinolytic agents, the oily base of these agents prevents adhesives from sticking to the skin. Perioperatively, tubes and catheters may need to be sewn or tied into place. Because of thick skin, peripheral IV access may be difficult. In severe HI, temperature regulation is impaired. Severe restrictive lung disease, difficulty intubating due to limited mouth opening and facial distortion, protein and electrolytes losses, dehydration, and risk for development of sepsis are all potential complications related to anesthesia in these patients. HI is an exceedingly rare condition; thus, it is important to raise awareness from others' experiences in order to avoid potential risks and complications specific to this disease process in the perioperative setting.



