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**Background:** Pediatric mastocytosis includes a rare spectrum of diseases characterized by abnormal mast cell infiltration in one or more organs. The cutaneous form (urticaria pigmentosa) is the most frequent pediatric clinical variant.<sup>1</sup> Various stimuli may cause mast cell degranulation leading to mediator release. However, there have only been few reports of serious anesthesia-related complications in pediatric populations.<sup>2</sup> Dexmedetomidine is a potential mast cell stabilizer that has been shown to block histamine-induced bronchoconstriction.<sup>3</sup>

**Case Description:** A 6-year-old female with history of mast cell mediator release due to the urticarial pigmentosa variant of cutaneous mastocytosis, factor VII deficiency, and urinary incontinence presented with increasing frequency of urinary tract infections and associated pyelonephritis. At baseline, she had macules with random distribution (Figure 1). She presented with the requirement of general anesthesia for a spine MRI and subsequent fatty filum release. **Spine MRI-** She underwent general anesthesia with an LMA and was maintained on sevoflurane without adverse events. Upon transport to the PACU, she experienced agitation/delirium with significant cutaneous flushing. Dexmedetomidine 0.5 µg/kg IV was administered with improvement, and the flushing subsided. **Lumbar Laminectomy-** Perioperative management included factor VII desensitization and early intraoperative infusion, avoidance of anaphylactic triggers, minimizing histamine-releasing medications, mast cell stabilization, and preparation for potential anaphylaxis. She had an IV induction with propofol and fentanyl with placement of an oral ETT. Anesthesia was maintained with sevoflurane, fentanyl boluses, and a dexmedetomidine infusion at 0.5 µg/kg/hr with a 1 µg/kg IV bolus prior to extubation. She tolerated the procedure well, was extubated in the OR, and monitored in the PICU post-operatively without any adverse events.

**Discussion:**

Pediatric mastocytosis can lead to provoked or unprovoked mast cell degranulation and mediator release. Although there are few reports of serious anesthesia-related adverse effects in the pediatric population, preparation and awareness for potential mediator release should be taken.

**References:**

1. *Anesth Analg* 2008; 107: 422-427.
2. *Pediatric Anesthesia* 2009; 19: 97-107.
3. *Anesthesiology* 2004; 100: 359-363.
4. *BJA* 1990; 65: 254-257.



Figure 1: Urticarial pigmentosa, a variant of cutaneous mastocytosis. These lesions are seen as unreactive (left) and reactive (right).

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