Avoiding Anaphylaxis:
Pediatric Cutaneous Mastocytosis and Anesthetic Challenges
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BACKGROUND
Pediatric mastocytosis includes a rare spectrum of diseases characterized by abnormal mast cell infiltration and proliferation into one or more organs, including skin, bone marrow, gastrointestinal tract, liver, spleen, and lymphoid tissue.1 The cutaneous form (urticaria pigmentosa) is the most frequent pediatric clinical variant, consisting of multiple reddish-brown hypopigmented macules, papules, or nodules at random distribution.2 Symptoms include GERD, flushing, pruritus, urticaria, hypotension, and provoked or unprovoked anaphylaxis. Anesthetic medications including muscle relaxants, opioids, and volatile anesthetics all raise concern for mast cell degranulation and mediator release. Despite this, there have only been few reports of serious anaphylaxis-related complications in pediatric populations.3

Dexmedetomidine is a potential mast cell stabilizer. In vitro studies have indicated that α2-adrenoceptor stimulation can lead to smooth muscle relaxation and bronchoconstriction prevention; in dogs, dexmedetomidine 0.5 µg/kg IV was shown to block histamine-induced bronchoconstriction.4

CASE DESCRIPTION
A 6-year-old female with mast cell mediator release due to the urticaria pigmentosa variant of cutaneous mastocytosis, factor VII deficiency, and urinary incontinence presented with increasing URI frequency and associated pyostomatitis (Figure 1). She had been diagnosed with cutaneous mastocytosis at 2 months-old by symptomology with a positive Darier’s sign and was on chronic medications including cromolyn, ranitidine, montelukast, and cetirizine. At baseline, she had macules with random distribution (Figure 2). She had a service dog that was trained to alert when detecting cutaneous cues of mast cell mediator release (Figure 1). The dog alerted to minor reactions by circling and more serious reactions by barking and tugging on the caretaker’s clothing. The patient presented under general anesthesia as a spine MRI, subsequent fatty filum release, and later a cystourethroscopy.

Spine MRI
The patient underwent an inhalational induction with sevoflurane in oxygen. An IV was secured and LMA placed without the need for IV medications, and she was maintained with sevoflurane in oxygen and air. She tolerated the MRI well, and the LMA was removed under deep anesthesia. During transport to the PACU, she exhibited emergence agitation/delirium and showed signs of significant cutaneous flushing. Dexmedetomidine 0.5 µg/kg IV was administered without vital signs were assessed. The patient quickly showed signs of improvement, the flushing subsided, and she awoke peacefully later in PACU. Her clothing worn during the MRI was later presented to the service dog, who alerted to a major reaction.

Cystourethroscopy and Pressure Induced Cystogram
Given the possibility of Deflux treatment during the procedure, the patient was admitted prophylactically for IV methylprednisolone. We were unable to attempt desensitization to the medication due to the nature of Deflux. Her service dog was allowed into the procedure suite as an additional monitor to detect evidence of mast cell degranulation. The patient was present under the chair of her handler near the anesthesia team (Figure 3). An IV induction with propofol was performed followed by LMA placement. Anesthesia was maintained with sevoflurane in oxygen and air, fentanyl, and dexmedetomidine infusion at 0.5 µg/kg/hr. The IV was given prior to incision without evident of mast cell mediator release. Her temperature was monitored closely for any abrupt changes, and she tolerated the procedure well with an EBL of 10 mL. Prior to extubation, dexmedetomidine 1 µg/kg was given. She was monitored in the PICU post-operatively where she remained hemodynamically stable. She was maintained on steroids, diphenhydramine, acetaminophen, and oxycodone and ultimately discharged home on POD #4 without any adverse events.

Lumbar Lamincotomy for Fatty Filum Release
Preoperative management consisted of a PICU admission for desensitization to recombinant Factor VIIa (rVIIa) with IV diphenhydramine and prednisone. An IV induction with propofol and fentanyl was performed with placement of an oral ETT. Pressure points and the hyperpigmented lesions were padded carefully. Anesthesia was maintained with sevoflurane in oxygen and air, fentanyl, and dexmedetomidine infusion at 0.5 µg/kg/hr. The rVIIa was given prior to incision without evidence of mast cell mediator release. Her temperature was monitored closely for any abrupt changes, and she tolerated the procedure well with an EBL of 10 mL. Prior to extubation, dexmedetomidine 1 µg/kg was given. She was monitored in the PICU post-operatively where she remained hemodynamically stable. She was maintained on steroids, diphenhydramine, acetaminophen, and oxycodone and ultimately discharged home on POD #4 without any adverse events.

DISCUSSION
Urticaria pigmentosa (UP) is the most common variant of mastocytosis in pediatric populations.2 Although many drugs used routinely in anesthesia reportedly cause mast cell degranulation, deviations from routine anesthesia techniques are not necessarily warranted.3 In contrast to adults, there are no reports of anesthesia-related deaths and few reports of serious anaphylaxis-related complications.4 However, heightened awareness of potential consequences of mast cell mediator release is warranted, and medications to treat should be immediately available. Special attention to position and protection of pressure points and UP lesions must be given. Perioperative management for this patient included IV desensitization, avoidance of mast cell degranulation triggers, minimization of histamine-releasing medications, efforts towards mast cell stabilization, and preparedness for potential mediator release leading to anaphylaxis. The role of a service dog has been shown to be beneficial in a family-centered care model.3 These service dogs also have the potential to be beneficial in functioning as an additional monitor in unique cases and circumstances.

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

REFERENCES