

# Non Triggering Anesthetic in a Patient with Mastocytosis and vonWillebrand's Disease

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## CASE HISTORY

5 year old male with history of mastocytosis, craniosynostosis, obstructive sleep apnea and von Willebrand disease (vWD) who presented for major cranial vault reconstruction.

Mastocytosis is a rare condition with elevated levels of cutaneous and systemic levels of mast cells<sup>1</sup>. Increased levels of mast cells place patients at elevated risk for anaphylaxis from drugs and physical stimuli compared to the general population. Lifetime prevalence of anaphylaxis is up to 50% in adults. Anesthesia, particularly general anesthesia for major surgery, has been associated with increased risk of anaphylaxis - up to 6% of patients, noting some anesthetic agents have increased risk of histamine release<sup>2,3</sup>. In addition mast cell secretory granules contain numerous factors, including heparin and tryptase, which can contribute towards coagulopathy and fibrinolysis respectively<sup>4</sup>.

## THE CHALLENGE

Delivering an anesthetic that would mitigate any systemic mast cell degranulation which could lead to anaphylaxis and coagulopathy due to histamine release.

## TREATMENT PLAN

An IV attempted preoperatively to administer Humate P<sup>®</sup> to treat coagulopathy due to chronic activation of mast cells and vWD. This failed due to patient being a difficult IV placement and agitation.

Proceeded to OR for inhalational induction. Patient induced with standard ASA monitors, radial arterial line and two PIVs inserted.

Baseline TEG and tryptase level drawn and then Humate P<sup>®</sup> given for vWD. Methylprednisolone, diphenhydramine given for anaphylaxis prophylaxis.

## TREATMENT PLAN

Anesthesia maintenance consisted of drugs that did not increase histamine release. Isoflurane, vecuronium, fentanyl and aminocaproic acid infusion.

Patient hemodynamically stable throughout, TEG and tryptase levels repeated to look for signs of systemic mast cell degranulation.

Patient extubated and went to PACU then ICU uneventfully.

## FIGURES/PICTURES

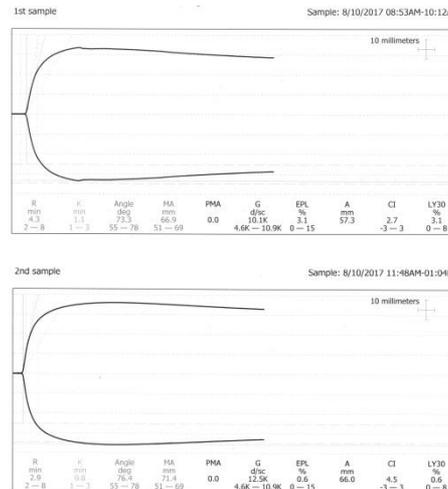


Figure 1: TEG at start and end of procedure

## FIGURES/PICTURES

	Before Incision	Incision Closure	Normal
Tryptase	192	109	<10.9

Table 1: Baseline tryptase and at closure

## CONCLUSION

We were able to safely administer general anesthesia to this complex patient without major bleeding or triggering anaphylaxis. This was done by administration of appropriate hemostatic factors, prophylactic medications and avoiding histamine inducing anesthetic agents. Pre and post surgical tryptase levels support this.

However, abnormally high tryptase levels at the start of the case, 192, raises concern that physical stimuli such as attempting PIV placement can lead to significant levels of histamine release with the potential development of anaphylaxis subsequently in this population.

While we avoided premedication in this case due to sleep apnea concerns, it should be given additional consideration in this population

## REFERENCES

- Muller, U.R., *et al.* The Problem of Anaphylaxis and Mastocytosis. *Current Allergy and Asthma Reports.* 2009, 9:64-70
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- Guilarte, M., *et al.* The Mast Cell, Contact, and Coagulation System Connection in Anaphylaxis. *Frontiers in Immunology.* Vol 18. July 2017. Pg 1-6.