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## SUMMARY

Life-threatening bleed in hemophilic inhibitor patients (pts) who are unresponsive to the bypassing agents alone, the addition of VIII conc should be considered, esp. when VIII inhibitor titer is <10BU/ml.

## INTRODUCTION

- Routine bleeding in hemophilic inhibitor is treated with the bypassing agents (BA, i.e., Novoseven®, FEIBA®).
- However, the use of BA alone in the life-threatening bleeding with massive amount of bleeding may be insufficient and inefficacious.
- Although pts who have been on BA exclusively may have their VIII inhibitor titers lowered, the use of VIII conc. is avoided in the usual bleeding in fear of anamnestic rise of inhibitor titer.
- A VIII inhibitor pt, who had a life-threatening intramural intestinal hemorrhage refractory to a large doses of BA (Novoseven®), which progressed to imminent surgery for decompression, promptly responded to the addition of large doses of VIII conc.

## CASE

• A 44 y.o. hemophilic with high titer inhibitor (152BU/ml) and high responder who had been on BA (Novoseven®) for at least >3 yr without exposure to VIII conc. was hospitalized with abdominal pain, and swelling.

• Initially the pt was treated with Novoseven® 250 KIU (90 ug/kg) q2~3 hrs for 6 days with progression of abdominal distension. Despite the increased dose of Novoseven to 500KIU (180ug/kg) q2 hrs the intramural hematoma worsened to the degree of imminent decompression procedure and the CT finding of progression of intramural hematoma and hemoperitoneum. Because of the lack of response to BA and the fact he had not been exposed to VIII conc., large doses of VIII conc. (100u/kg q 8 hr, 2 doses) was added to Novoseven®, which resulted in a prompt relief of pain within 6 hrs of VIII conc. and gradual relief of abdominal distension.



Fig 1. CT finding of intramural hematoma at the small bowel and hemoperitoneum. L. cross-section view, R. longitudinal view.

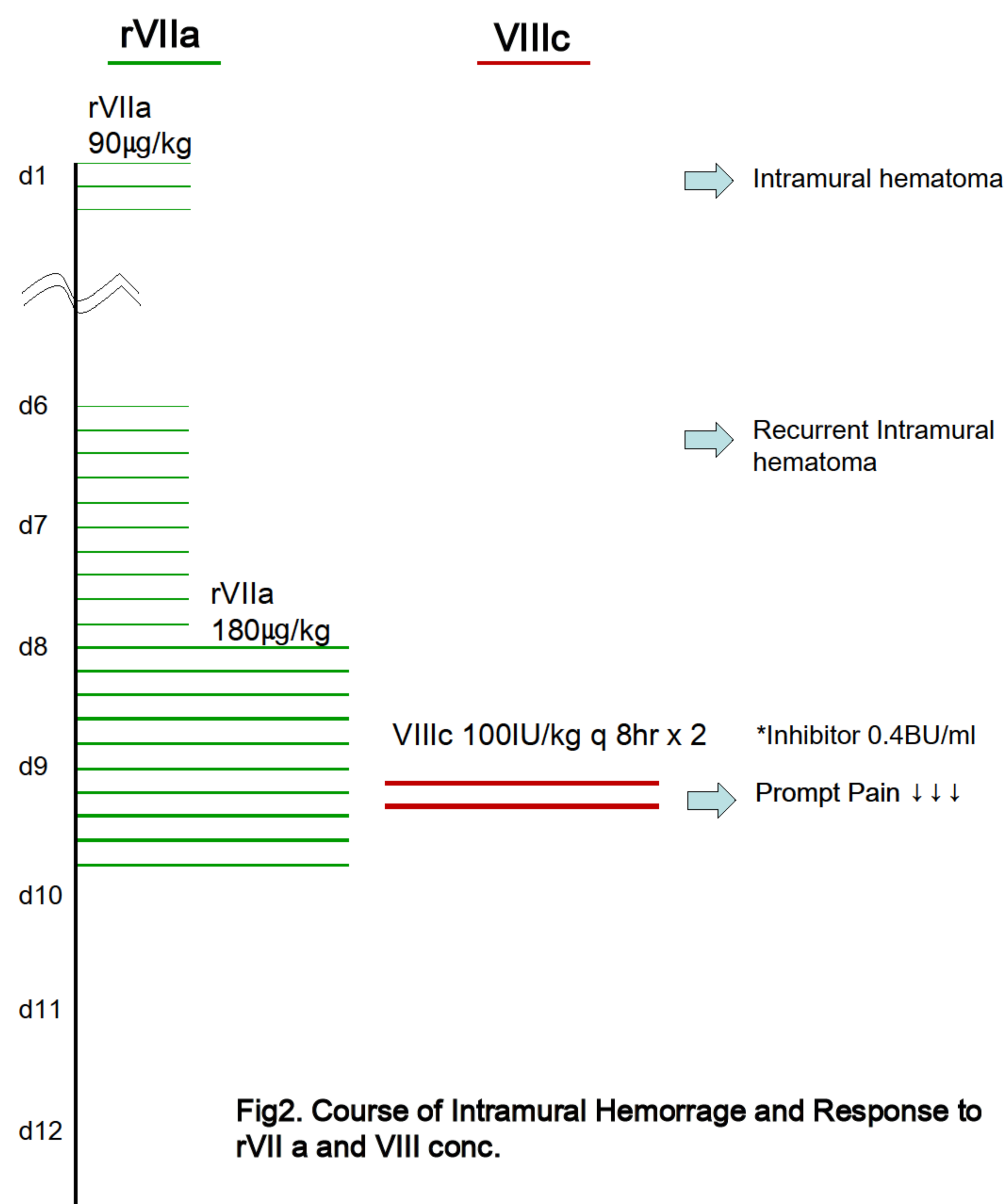


Fig2. Course of Intramural Hemorrhage and Response to rVII a and VIII conc.

## DISCUSSION and CONCLUSIONS

- Since high titered inhibitor pts are treated with BA alone for routine bleeding and are not exposed to VIII conc., their VIII inhibitor levels may be low enough to overcome with large doses of VIII conc in case of severe bleeding.
- Therefore, in hemophilic inhibitor pts with severe life-threatening bleedings, such as, intraabdominal bleed, CNS, etc, uncontrollable with BA alone, the use of large doses of VIII conc. (100u/kg) in conjunction with BA should be considered in order to save life.

## REFERENCE

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