

# Management of Hemophilia A in a Partial Hepatectomy

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## Case Presentation

Hemophilia A is an x-linked recessive hereditary disorder with factor VIII deficiency. This can lead to bleeding, clotting, and other intraoperative complications. Goals of care during the perioperative period involve optimizing both factor VIII activity and coagulation profile. If needed, adequate replacement of factor VIII levels is key to preoperative optimization.

A 68-year-old male with hypertension, hepatitis C, hemophilia A, and newly diagnosed cholangiocarcinoma in segment 6 of the liver presented for partial hepatectomy. Medications included amlodipine, hydrochlorothiazide, lisinopril, and clonidine. Review of symptoms was pertinent for fatigue and back pain. Physical exam and labs were within normal limits.

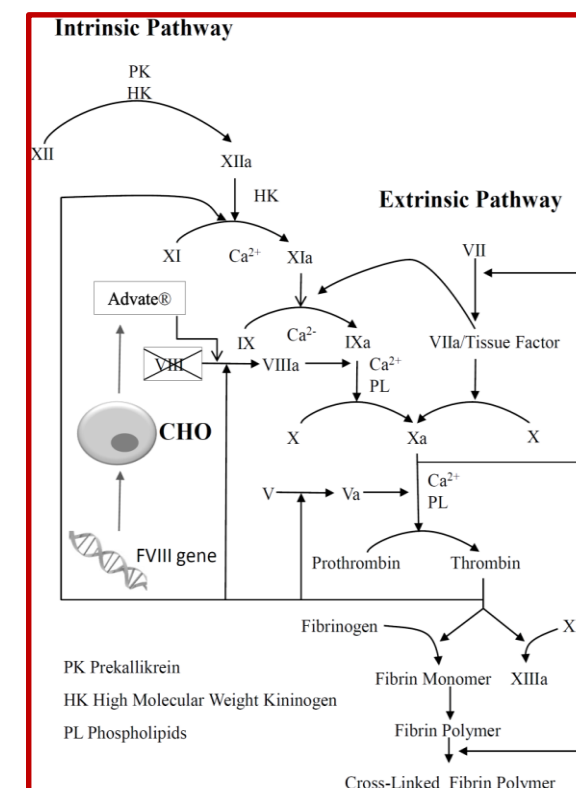
Severity of hemophilia	Blood clotting factor level
Normal	50% - 150%
Mild	>5% - <40%
Moderate	1% - 5%
Severe	<1%

## Preoperative Evaluation

The World Federation of Hemophilia recommends preoperative factor VIII levels of 50-80% for minor surgery and 80-100% for major surgery.

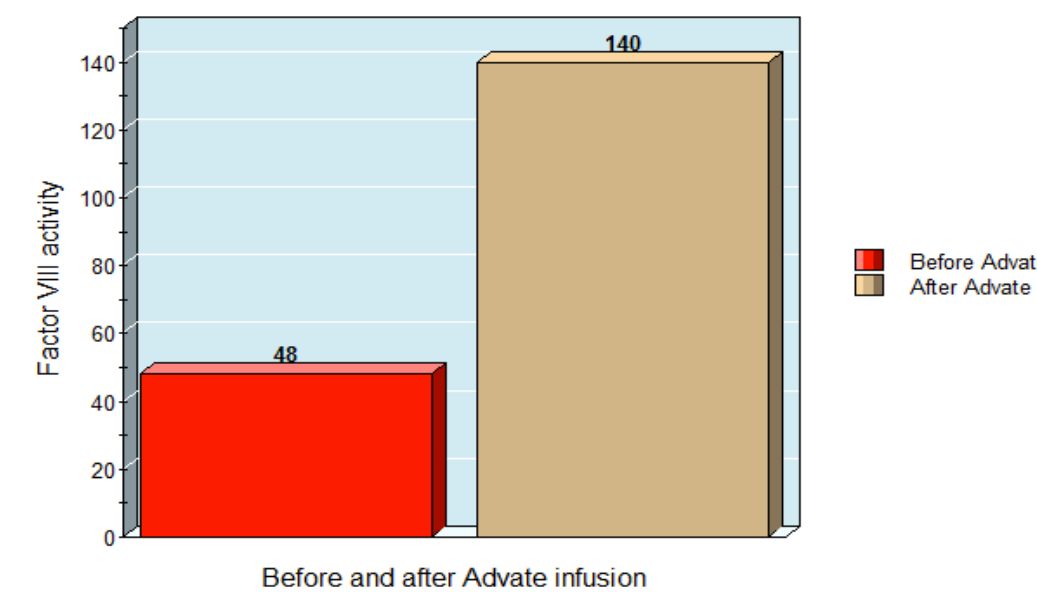
Patient was admitted prior to surgery with baseline factor VIII activity 45%. Hematology was consulted. Repeat Factor VIII activity was 48%, platelets 68,000.

Preoperatively, our patient received 50 IU/kg (4660 IU) infusion of recombinant factor VIII (Advate) on the day of surgery. Repeat factor VIII activity was 140%. Hemoglobin was 14.7. He had a 20G PIV in situ.



<https://www.semanticscholar.org/paper/USE-OF-RECOMBINANT-HUMAN-COAGULATION-FACTOR-VIII>

Factor VIII activity after Advate infusion



## Perioperative Course

Patient underwent general endotracheal anesthesia with addition of a 16G PIV and a radial arterial line. 4 units of PRBC were available on standby.

Vitals remained stable throughout the course with total phenylephrine boluses of 1000 mcg over approximately 4 hours. No blood products were administered.

Total IV fluids were 2300 mL of lactated ringers and 500 mL of 5% albumin. Estimated blood loss was 900 mL.

The patient was extubated and taken to the recovery room. Immediate post-operative hemoglobin was 11.7 with factor VIII activity 105%.

Hematology continued to follow the patient and checked factor VIII activity daily with the goal of infusing recombinant factor VIII 25 IU/kg for activity below 80%. No additional recombinant factor VIII was needed.

Patient was discharged on post-operative day 6 without complications and hemoglobin of 9.0.

## Discussion

Historically, elective surgery was not performed on patients with hemophilia, but in the setting of cholangiocarcinoma, surgery is the only hope for cure. Partial hepatectomy in patients with hemophilia A can lead to disastrous consequences if bleeding is not controlled. There are no standard guidelines for the anesthetic management of these patients.

One case study series described successful hepatectomy with median bolus injections of recombinant factor VIII of 3000 IU followed by continuous infusions of 4 IU/kg/h until 7-14 days after surgery. Another case with 887 mL blood loss gave an additional 2000 IU intraoperatively followed by 5 IU/kg/h until 7 days after surgery. Our patient only required one preoperative recombinant factor VIII bolus.

If preoperative factor VIII levels had been below 80%, hematology would have recommended an additional 25 IU/kg recombinant factor VIII infusion prior to surgery.

In the event of ongoing bleeding, additional blood products such as PRBC, FFP, platelets, and cryoprecipitate could have been administered. We discussed with pharmacy and had plans in place for additional recombinant factor VIII infusion intraoperatively if necessary. We did not discuss a continuous postoperative infusion. Thromboelastogram could also provide useful information to guide blood product administration.

## Conclusions

Early consultation with hematology, monitoring factor VIII activity level, and administration of recombinant factor VIII was critical to our patient's successful anesthetic outcome. It is important to have multi-disciplinary discussions to come up with a cohesive perioperative plan when caring for patients with hemophilia A.

## References

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