

# The experience of surgery in hemophilia patients with inhibitors

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## Introduction and Objectives

Inhibitors are the most serious complication of hemophilia therapy. Inhibitors significantly increase morbidity and also lower “quality of life” in hemophilia patients. Treatment of inhibitors and surgeries in hemophiliac patients with inhibitors are difficult and extremely expensive. We report that our surgical experience in 9 patients with hemophilia and inhibitors.

## Material and Methods

We retrospectively reviewed medical records of 16 patients with hemophilia patients with inhibitors who needed minor and major surgery. Activated prothombin complex concentrates (aPCC) was given a loading dose of 75 U/kg and followed by 150 U/kg/day in two divided doses every 12 hour. Recombinant factor VIIa (rFVIIa) was given 90 mcg/kg before and after surgery every 2 hour.

## Results

Characteristics of severe hemophilia A patients underwent surgery treated with rFVIIa

Age at Circumcision	Inhibitor Status (BU)	Weight (kg)	Complications of Circumcision and Time	Concurrent Operation/ Situation	DH (days)
13 years S.Ü.	23,6	33	2 <sup>nd</sup> day oozing and severe penile hematoma	None	20
13 years E.D.	25,6	35	1 <sup>st</sup> day, oozing	Cataract	4
16 years I. A.	30,7	52	7 <sup>th</sup> day oozing.	None	3
4 years O. I.	2,5	15	11 <sup>th</sup> day oozing	None	4
3 years N. T.	3,6	13	None	Laceration in tongue	3

Age at surgery	Surgery	Inhibitor Status (BU)	Weight (kg)	Complications and Time	Concurrent Operation	DH (days)	rFVIIa dose µg/Kg
3 years A.K.	Port a cath insertion (CIVAD replacement)	327,8	17	None	None	2	90/3 doses
5 months R.G	Subdural and intracerebral hematoma drainage	56,4	10	Exitus	None	2	100/12 doses
16 years E.D.	Toncil cauterizing and clot aspiration with bronchoscopy	25,6	60	None	None	3	90/12 doses during and 3 doses after intervention

Age at RAS	Surgery	Inhibitor Status (BU)	Weight (kg)	Complications and Time	Concurrent Operation	DH (days)	rFVIIa dose µg/Kg
16 years S.Ü.	RAS (right knee)	7,8	60	None	None	3	90/3 doses
16 years S. Ü.	RAS (right elbow)	7,8	60	Bleeding: after 3 days	None	3	270/2 doses
16 years S.Ü.	RAS (left knee)	7,8	60	None	None	3	270/2 dose
17 years B.S.	RAS (Bilateral knees)	6,4	57	None	None	2	90/3 doses

DH:Duration of hospitalization,

RAS:Radioisotope synovectomy

Characteristics of severe hemophilia A patients underwent surgery treated with aPCC

Age	Surgery	Inhibitor(BU)	Complications	Concurrent operation
16 y	Dental extraction	6,4	Severe haematoma	None
4y	Port a cath insertion	327,8	None	None
1 y	Circumcision	150	Haematoma	Inguinal hernia repair
17 y	Circumcision	6,4	None	Trafical accident
23y	Flexor tenotomy	10,25	None	None
16y	Root canal treatment	30,7	None	None
7 y	Nephrolithotomy	10,25	Hematuria	None
17 y	Full mouth prothetic rehabilitation	10,25	None	None

During this period, 24 surgical procedures were performed in 16 hemophilia patients with inhibitors, age ranging from 5months to 23 years with a average of 9,7 years. Only one patient with intracranial bleeding died postoperative periode. There were no tromboembolic complications. Blood transfusion was given in 3 patients.

## Discussion

With the introduction of by-passing agents, surgical interventions can be performed safely in hemophilia patients with inhibitors and we didn't see any thromboembolic complication other than mild and moderate bleeding depend on patient's surgery .

Surgical procedures in hemophilia patients with inhibitors can present a challenge because of the increased risk for bleeding complications and the potential difficulty in controlling bleeding during and after surgery. So, these patients need comprehensive hemophilia treatment centers. The centers approach ideally incorporates a number of specific pre-, intra-, and postoperative objectives for these patients undergoing surgery.

## References

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