Inherited Factor VII deficiency - perioperative replacement therapy is not necessary in most cases

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Background:
Inherited factor (F)VII deficiency is a rare bleeding disorder with a wide variation of bleeding symptoms less severe than in haemophilia A and B. Consistent evidence-based recommendations regarding perioperative replacement management do not exist for FVII-deficient patients. Recent publications (Benlakhal F et al for the French Study Group of Factor VII Deficiency. J Thromb Haemost. 2011:1149-56) suggest a replacement therapy only for patients with a factor VII activity below 10%. The aim of this study was to evaluate bleeding symptoms in daily life and pregnacy and perioperative bleeding in patients with factor VII deficiency.

Methods:
We collected retrospectively data of all patients with factor VII deficiency presented between 2001 and 2011 in two institutions (Vivantes Klinikum im Friedrichshain, Berlin and Zentrum für Blutgerinnungsstörungen, Leipzig).
We evaluated age, sex, factor VII activity, surgical interventions, perioperative replacement therapy, bleeding symptoms and pregnancies.

Results: We analysed 119 patients between 4 and 76 years old at presentation. 3 patients had a FVII activity below 1%, 7 patients below 10%, 24 patients below 30% and 85 patients below 60%.
Most patients showed up for a diagnostic work up with a reduced Quick value. Underlying molecular defects were mostly missense mutations but up to now only available for 43 patients.
16/119 patients had bleeding symptoms in daily life (1 patient with FVII < 1% had epistaxis and 15 patients with FVII > 20% hypermenorrhoe (8/15) and haematomas (7/15)).
37 surgeries were performed in these patients. 11/37 had bleeding complications during surgery before diagnosis of FVII deficiency. There was no relation between bleeding symptoms and FVII activity.
In 19 surgeries (10/19 major) after the diagnosis of FVII-deficiency no bleeds occurred. All patients received tranexamic acid. One patient (FVII < 1%) received one dose recombinant FVIIa before total hip replacement and one patient (FVII < 1%) received only tranexamic acid before hip replacement. 6 pregnancies (FVII: 3% to 55%) took place without bleeding complications and without any replacement therapy. The table shows additional details concerning pregnancies and surgeries in patients with FVII < 30%)

Conclusions:
Patients with FVII-deficiency have a low number of bleeding symptoms in daily life and during surgery. Bleeding symptoms were not proportional to the residual factor VII activity. Even patients with severe FVII deficiencies do not need routinely perioperative replacement therapy.

Factor VII-Activity (%)	Type of surgery	Replacement	Bleeding
<1	Larynx carcinoma	Tranexamic acid	No
<1	Total Hip replacement	1x 2mg NovoSeven, Tranexamic acid	No
<1	Total Hip replacement	Tranexamic acid	No
<1	Adenotomy	No	Yes
7	Tonsillectomy	No	No
10	Appenectomy	No	No
10	Hernia repair	Tranexamic acid	No
10	Ankle fracture	Tranexamic acid	No
21	Hernia repair	Tranexamic acid	No
25	Adenotomy	No	No
27	Hysterectomy	No	No
29	Tonsillectomy	No	No
30	Varicectomy	Tranexamic acid	No
3%	Pregnancy, spontanous Partus	No	No
6%	Pregnancy, spontanous Partus	No	No
14%	Pregnancy, spontanous Partus	No	No
33%	Pregnancy, spontanous Partus	No	No
35%	Pregnancy, sectio	No	No
44%	Pregnancy, sectio	No	No

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