

Effective management of surgical procedures of inherited FVII deficient patient – six-year experience at University Hospital in Ostrava

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Objectives:

Inherited factor VII (FVII) deficiency is the most common of the rare inherited bleeding disorders. The inheritance is autosomal recessive and prevalence of severe FVII deficiency is estimated 1:300 000 – 1:500 000 individuals. The clinical symptoms are highly variable, ranging from asymptomatic to severe life-threatening disorders. In laboratory investigations prothrombin time is prolonged.

The weak correlation between FVII coagulation activity (FVII:C) and bleeding symptoms is well-known. Efficient haemostasis can be achieved with the levels of FVII:C in the range of 10-15 %. Replacement therapy based on FVII:C levels and type of surgery is recommended.

Methods:

22 patients with FVII deficiency are registered in Hemophilia Treatment Centre in Ostrava (Czech Republic). Six of them have FVII:C < 1%, ten patients in the range 1-10 %, six patients 10,1 – 35 %. Ten of our patients underwent 14 surgical procedures between 2008 and 2013. Nine surgical operations were managed by substitution plasma-derived FVII (pdFVII, Factor VII Baxter, Baxter AG, Vienna, Austria). Five patients were substituted by activated recombinant factor VII (rFVIIa, NovoSeven®, Novo Nordisk, Bagsvaerd, Denmark).

Table 1: Characteristics of the patients who received substitution by plasma-derived FVII

n.	sex	age	baseline FVII:C %	type of surgery	IU	body weight (kg)	total IU/kg	period of substitution	bleeding outcomes
1	M	23	6,3	extraction of 3 third molars	1200	62	19	2x 600 IU á 4 h	excellent (suture)
2	F	44	6,5	extraction of 1 third molar	1200	65	18	2x 600 IU á 4 h	excellent (suture)
3	M	19	8,5	subperiosteal abscess, extraction of 1 third molar	1800	85	21	1200 IU+ after 5 h next 600 IU	excellent (suture)
4	F	39	4,0	mastopathy excise	6000	97	62	48 h	excellent
5	F	45	5,0	reduction mammoplasty	14400	64	225	6 days	excellent
6	M	50	3,9	repair of large incisional abdominal wall hernias	33600	101	333	7 days	excellent, 250 ml of serosanguinous fluid from drainage during first 24 hours
7	F	69	3,6	cataract	600	100	6	600 IU before surgery	excellent
8	F	47	0,5	hysteroscopy with a biopsy	3000	75	40	24 h	excellent
9	M	70	3,6	pacemaker reimplantation	3600	100	36,00	24 h	excellent

Table 2: Characteristics of the patients who received substitution by rFVIIa

n.	sex	age	baseline FVII:C %	type of surgery	mg	kg	total ug/kg	period of substitution	bleeding outcomes
1	M	63	1,4	radical nephrectomy (renal cell carcinoma)	82	120	683	7 days	excellent
2	F	47	0,5	cholecystectomy	64	74	865	7 days	excellent
3	F	49	4,6	repair of incisional abdominal wall hernias	70	109	642	7 days	excellent
4	F	70	0,1	gynaecological surgery for susp. ovary neoplasm (fibrom by histology)	67	80	838	7 days	excellent
5	M	52	3,9	TEP coxae l. sin.	81	97	835	10 days	excellent, < 300 ml during surgery + 640 ml from drainage next 48 h

Results:

rVIIa was used at a dose 15 - 30 ug/kg and pd FVII was used at a dose 8 – 20 IU/kg every 4h during the first two days and from the third day every 6h. Time of treatment was in the range 1 – 8 days depending on the type of surgery. Ten patients used tranexamid acid as a concomitant treatment, six patients received perioperative thrombosis profylaxis by low-molecular weight heparin (LMWH)

Conclusions:

During surgery in the substituted patients we did not register excessive bleeding loss in any group of patients. Also the postoperative course was without bleeding or different complications. Any thrombembolic disease in connection with replacement therapy was not observed.

References:

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