

Prophylactic treatment in factor XI deficiency patients undergoing invasive procedures.

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1 Aim of the study

The factor XI deficiency is a coagulation disorder leading to bleeding for a few patients.

There is not a good correlation between the level of the protein and the bleeding risk.

We studied in our cohort of patients the adopted strategy and the rate of complication for surgeries and deliveries stratified according to the factor XI level.

2 methodology

Population : patients known with a factor XI deficiency since 1995 and relatives with normal level but obligate heterozygotes in Angers university teaching Hospital

invasives procedures were analysed retrospectively and / or prospectively for each patient

Factor XI level was determined in a one-stage chromométric assay with deficient F XI (Diagnostica stago) and CK Prest (Diagnostica Stago)

3 results

175 patients were included. Age ranged from 2 to 87 years with a medium of 37 years

Patient distribution according to factor XI level :

UI/dl	<5	5 à 15	16 à 30	31 à 50	> 50
N=	8	5	44	96	22

46 (26%) had another abnormal blood coagulation test or disease with potential bleeding influence

Factor XIIc deficiency n=11, low PT without hepatic insufficiency =2, prolonged occlusion time on PFA 100® =1, vWD =4, acquired vWD =1, VIIIc deficiency =4, Haemophilia A carrier =3, thrombocytopenia =4, hepatic disease =3, factor VII deficiency =1, VII and IX deficiency =1, IX deficiency =1, factor V deficiency =2, hypofibrinemia =1, factor XIII deficiency =1, Carbohydrate Glycosylation Deficiency Syndrome =1, Noonan Syndrome =1, galactosemia =1, lysosomal acid alpha-glucosidase deficiency (Pompe disease) =1

APTT value according to FXI level :

Always abnormal	8	4	28	48	9
both	0	0	8 (20 %)	16 (17 %)	2 (9 %)
Always normal	0	0	4 (10 %)	24 (25 %)	11 (50 %)

Invasive procedures performed :

N procedure	26	8	87	138	27
Delivery	0	0	17	24	5
Caesarian	4	0	4	11	3
Tooth extraction	4	4	10	22	4
High risk surgeries	CIA correction under celioscopy, prostatectomy, fraumatic facial injury repair, cranial hematoma evacuation	Nissen fundoplication under coelioscopy with gastrostomy	Tumoral cerebral biopsy, hepatectomy, breast reduction, aortic valve replacement with coronary by-pass, scoliosis surgery, neuroblastoma, prostatectomy, cystectomy, pancreatectomy	Pancreatectomy, hepatic transplantation, renal angiomyolipoma, Hartmann's procedure for rectal carcinoma	cervical Chordoma
Written protocols for coagulation	7 patients 15 protocols	1 1	5 5	3 3	0 0
No FFP	3 2	4 0	75 0	126 0	24 0
Purified Factor XI	21 (81 %)	3 (37 %)	5 (6 %)	0	0
Tranexamic acid	6	2	11	10	0
dDAVP	0	0	4	0	2
others			Platelets transfusion 1	Vit K 1	APCC 1
Haemorrhages	3 (Day 5 to 7)	2	8	16	3
Red Blood Cell transfusion	1 (3,8 %)	0	4 (4,6 %)	6 (4,3 %)	0
DVT	2	0	0	0	0
Arterial thrombosis	0	0	0	0	0

Prophylactic strategy and complications :

4 Conclusion

26% of patient had another abnormal coagulation test or pathology with potentially bleeding influence.

Sensibility of APTT : many patients may have a surgical process with an unknown factor XI deficiency.

The factor XI level cut-off for systematic factor XI substitution in high risk surgery might be 15 %

anti fibrinolytic therapy should be used frequently

No thrombotic complication related to Factor XI substitution were observed

Twice in the same patient later diagnosed with JAK2 positive essential thrombocythemia

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