

RECOVERY AND HALF-LIFE OF FACTOR VIII

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OBJECTIVES

A most serious complication of haemophiliacs are development of inhibitors. It occurs approximately up to 30% of haemophilia A and only up to %5 of haemophilia B. Immune tolerance induction (ITI) is the best treatment option in these patients by using factor concentrates and/or immunosuppressed agents. Any bleeding of hemophilia with inhibitors can be treated with bypassing agents. In all of the treatment options when inhibitor is found negative, it must be confirmed by recovery and half life tests. We want to report our last two years results of recovery and half-life of FVIII in hemophiliacs with inhibitors.

METHODS

We retrospectively analysed the data from 1999-2013 at our center. The patients who have inhibitors and under treatment once the inhibitor had been found negative, it was confirmed by FVIII recovery test prospectively. At first FVIII and inhibitor measured and then FVIII was given 50U/kg dose; at 30-60 minutes and 4-6. hours FVIII and inhibitor was measured again.

RESULTS

Recovery test was performed 9 of the 21 haemophiliacs with inhibitors and median age was 19 (range;8-52). All of patients were treated by aPCC and rFVIIa. ITI was performed 5 of the 10 patients. But at the 2 adults of the 4 patients inhibitor levels were elevated after six months period so ITI was stopped. In the other 2 children; inhibitor became negative in 4,5 years and recovery test and half life was successful (>%66) in 1; in the other 1 inhibitor became negative about one year but at recovery test there is partial response (%39,6) and ITI is going on. The other patient who was treated with ITI several times and then continuing with rFVIIa, inhibitor was found negative after 14 years. On the other hand, five patients who are on demand treatment with aPCC or rFVII and on prophylaxis with aPCC, on follow up situation when inhibitor became negative recovery test was performed. Although inhibitors were negative, half life of plasma FVIII level was not adequate that we could say inhibitor was negative.

INHIBITOR NEGATIVITY DURING FOLLOW-UP

	0. minute		30-60. minutes		4-6. hours		Treatment	Result
	Inh (BU)	FVIII (%)	Inh (BU)	FVIII (%)	Inh (BU)	FVIII (%)		
#1 VE	6.4	<2	(-)	30.8	(-)	15.2	rFVIIa	No response
#2 MŞ	3.44	<%0.2	(-)	8.5	(-)	1.8	rFVIIa	No response
#3 ÇB	(-)	0.3	(-)	0.8	(-)	0.3	aPCC rFVIIa	No response
#4 YE	(-)	0.4	(-)	0.9	(-)	0.3	rFVIIa	No response
#5 MK	(-)	<0.5	(-)	49.7	(-)	31.2	rFVIIa	Partial response

PATIENTS -IMMUN TOLERANS INDUCTION

	0. minute		30-60. minute		4-6. hours		treatment	result
	Inh (BU)	FVIII (%)	Inh	FVIII (%)	Inh	FVIII (%)		
#1L.A.Y	(-)	0.4	(-)	58.7	(-)	49.8	ITI rFVIIa	Complete response
#2 A.T	37.5	1.1	7.5	2	37.5	2.3	ITI Feiba	No response
#3 B.B	42.5	0.5	50	0.8	62.5	0.5	ITI rFVIIa	No response
#4 E.Ş	(-)	<1	(-)	65.3	(-)	52.1	ITI FVIII	Complete response
#5 A.A	(-)	0.3	(-)	41.4	(-)	39.6	ITI Feiba	Partial response

CONCLUSIONS

In the follow up of the inhibitor positive patients with haemophilia, inhibitor can be negative at laboratory tests. Although inhibitor is negative, it can not be reached the enough FVIII or FIX levels. That's why it must be confirmed with recovery test and half life of FVIII/FIX.

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