Management of acquired hemophilia: a single center experience.

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Introduction and objective: Acquired hemophilia A is a bleeding disorder caused by autoantibodies to coagulation factor FVIII. The authors evaluate management of the first bleeding episode and the results of the immunosuppression.

Materials and methods: We performed retrospective observational analysis of our hemophilic registery. All patients with acquired hemophilia between years 1996-2015 were included in analysis.

Results:

1. Patient characteristics

PATIENTS, n	12
Age, y	73 (60-88)
Sex, n (%) – Female/Male	50/50
FVIII level (%)	3 (0-15)
Inhibitor titer BU/ml	19 (1,6-214)
Cause of bleeding, n(%) – spontaneous / traumatic	33/67
Severity of bleeding, n(%)	25/75

2. First-line hemostatic therapy for all first bleeding episodes (median)

Therapy	n	FVIII (%)	INH (BU/ml)	Initial dose (U/kg)	Total dose/patient	Bleeding control - %
aPCC	2	6 (5-7)	27(19-35)	43 (32-54)	33 500U (23 000-43 000)	100 %
aPCC+FVIII	5	8.5 (0-15)	8.5(1.6-32)	35(14-75) + 40(14-106)	13 500U (3 000-72 000) + 4 600U (3 000-18 000)	80%
rVIIa	1	4	4	90	No efect	0%

3. Response to the first-line immunosupression

			Days from the start of immunosupresion, median				
REGIMEN	n	CR n(%)	Inhibitor negative	FVIII>70%	Imunosupression stopped	Relaps n(%)	Complications
Steroid	4	75	25 (23-28)	25 (23-28)	135 (120-150)	0	Severe infections, diabetes,psychosis
Steroid + Cyclo-phosphamide	6	50	30 (28-32)	30 (28-32)	Steroid 60 +CFA 730	0	Severe infections, diabetes

Conclusions: Bleeding was controlled by lower doses of hemostatic agents than is recquired by the European Acquired Hemophilia Registry. Thromboembolic events were not reported. We suggest a prophylactic antiinfectious therapy for severe infectious complications during the immunosuppression.

References: 1. Managment of bleeding in acquired hemophilia A – EACH2 Registry, Blood, r2012, 2. Immunosuppression for acquired hemophilia EACH2, Blood, r2012

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