



Haemophilia A with inhibitor in children: Experience of a single center in the last 25 years

Zafer Şalcıoğlu¹, Hülya Sayılan Şen¹, Gönül Aydoğan¹, Ferhan Akıcı¹, Zafer Başlar²

¹Kanuni Sultan Süleyman Education and Research Hospital, Pediatric Hematology and Oncology Clinic, Istanbul, Turkey

²Istanbul University, Cerrahpasa Medical School, Internal Medicine Department, Istanbul, Turkey



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Intraduction and objectives

Development of factor VIII inhibitors remains the most serious and life-threatening complication, occurring in 25-30% of all patients with severe hemophilia. Inhibitors mainly occur in previously untreated patients (PUPs) with severe hemophilia A during the first 50 exposure days (EDs). Aim of this study is to report the incidence of inhibitor development in our haemophilia A population in the last 25 years and to describe characteristics and management of inhibitors patients.

Materials and Methods

All patients included in the analysis received a diagnosis of haemophilia A between 1990-2015. Information of patients with haemophilia A were retrieved from patient files and from the records contained in the electronic information processing environment created after 2005.

Results

During the above mentioned period, 134 patients received a diagnosis haemophilia A (77 severe, 37 moderate and 20 mild) and have been followed in our center. Seven patients develop inhibitors (5.2%). General characteristics of patients with inhibitor have been shown Table 1. The range of age at the diagnosis of (+) inhibitor was 2-15 years.

They are all severe patients (FVIII 1<%). All of patients had high responder (HR) inhibitor. Inhibitor titres range was 6-176 BU. As regard as FVIII gene mutation, we observed large deletion in three patient. All of patients with inhibitor have been exclusively treated with plasma-derived FVIII (pdFVIII) at least once in their life. Prior to inhibitor development three patients were treated for a number of EDs < 50, 4 patients for ≥50 EDs. Four patients underwent low dose Immun Tolerance Induction therapy (ITI). Two patients obtained a CR. One patient didn't obtain remission. One patient underwent ITI regimen.

Conclusions

Inhibitor incidence in our patients was lower than previously reported data from community of Turkey. Mild or moderate haemophilia A patients did not develop inhibitor.

Table 1. General characteristics of patients with inhibitor

Patients	Age	F:C	Genetic	Peak Inhibitör BU	Duration with inhibitor	Prophylaxis (aPCC)	İTT
1	10	0.1	-	60	4	+	-
2	12	0.1	-	80	6	+	+
3	11	0.1	-	32	3.5	+	+
4	22	0	Large deletion	60	15	+	+
5	4	0.3	-	98	1	+	-
6	27	0	Large deletion	176	11	+	-
7	14	0.5	-	6	3.5	+	+

