

# STATUS OF INHIBITOR HEMOPHILIA PATIENTS IN DIFFERENT AGE GROUPS

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## INTRODUCTION AND OBJECTIVES

Tendencies in inhibitors formation in patients with inherited hemophilia cause permanent attention of clinicians. The current analysis of continuous observational Russian registry which started in July 2014 aims at providing additional insights on clinical data distribution by age groups.

## MATERIALS AND METHODS

Clinicians from 53 centers participate in the investigator-initiated observational registry. By May 2016 totally 219 inhibitor hemophilia patients are included.

## RESULTS

By December, 2015 high-responding inhibitors (HRs) were identified in 88 patients; titer  $91.2 \pm 381.2$  BU (mean $\pm$ SD). The lowest numbers of patients were observed in three age groups: <2 years - 5 (3%); 12-15 years - 3 (2%), and 15-18 years - 6 (3%). All patients had severe hemophilia. The distribution HRs/ low-responding inhibitors (LRs) was: 60/40%, 0/100%, 50/50% in the groups <2, 12-15, and 15-18 years, correspondently. The majority of inhibitor patients were registered in three age groups: 5-7 years – 30 (15%); 18-25 years – 29 (15%), and 25-35 years - 37 (19%). In these groups more than 90% of patients had severe form of the disease. HRs constituted, correspondingly, 60, 41, and 38% in each age group. The total number of target joints was 200. The higher incidence of target joints: 68% in patients aged 35-45; 67% - 15-18; 57% - 18-25; 54% - 25-35, 50% - > 45 years. The lower incidence was registered in younger age groups: 20% - <2; 26% - 2-5; 30% - 5-7, 22% - 7-12; 33% - 12-15 years.

## CONCLUSIONS

Distribution of inhibitor patients by age does not demonstrate clear tendencies. Target joints numbers are clearly related to age groups: ranging from 20 to 33% in infants and children before 15 years and increasing to 50-67% in various age groups above 15 years. With consistent increase in patients on prophylaxis the annual bleedings rates dropped significantly, particularly in frequent bleeders (ABR > 6) – from 73% in 2014 to 38% in 2016.

Medication	N of prescriptions	Mean dose	N of infusions for bleed's termination	
			≤ 3	> 3
rFVIIa	121	90,8 mkg/kg	63%	30%
APCC	56	61,3 IU/kg	71%	19%

