

Joint outcomes in adults with hemophilia A on prophylaxis in Scandinavia: results from *the KAPPA register*

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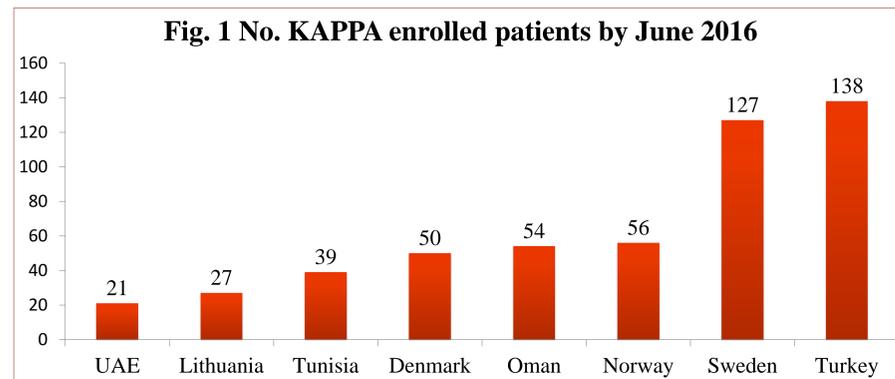


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Introduction and objectives

Prophylaxis for the first time was introduced in Sweden in late 1950s. Since then it has evolved over the time and the dose, injection intervals and products have changed. Denmark and Norway started prophylaxis later but nowadays treatment is very similar among the three countries. Lund university started KAPPA registry in 2013 to assess long term outcomes in moderate and severe haemophilia A. The aim of this study was to report on joint outcomes of KAPPA enrolled patients from Scandinavia.



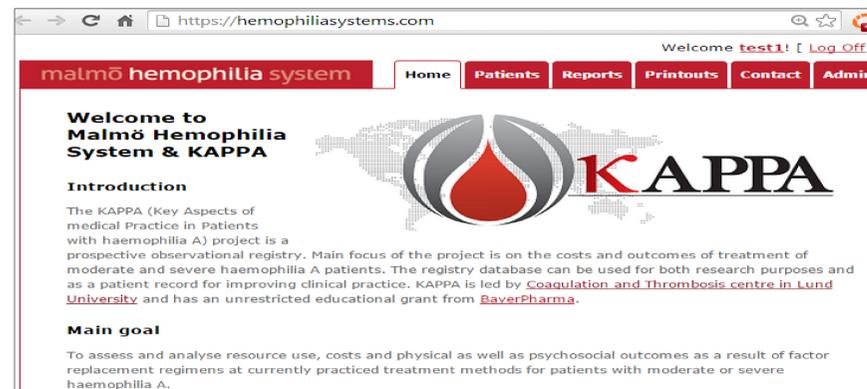
Methods

This cross sectional study implemented using data from KAPPA register (1). Eligible patients with moderate or severe hemophilia A were ≥ 15 years, on prophylaxis (at enrollment), negative for inhibitor and were residents of Denmark, Norway or Sweden. Data collection and clinical investigations were part of routine annual visits. For joint evaluations, hemophilia joint health score 2.1 (HJHS) was used. Total HJHS ranges 0-124.

Methods (cntd)

The scores of elbows, knees and ankles (both sides) each ranges 0-40. For the analysis we categorized patients into three age groups: g1(15-29), g2 (30-44) and g3 (≥ 45). Lund University regional ethics committee approved KAPPA register.

KAPPA online platform



Results and Conclusions

We recruited 106 participants including: 23 (25.5%), 27 (21.7%), and 56 (52.8%) patients respectively from Denmark, Norway and Sweden.

Table 1. Median and interquartile range (IQR) of HJHS scores

	15-29 y	30-44 y	$\geq 45y$
Sweden, median (IQR)	3 (2-8)	5 (3-23)	37 (23-43)
Denmark & Norway, median (IQR)	1 (3-5)	15 (5.5 -23)	39 (15-41)
Overall, median (IQR)	3 (2-6)	12 (3-23)	37.5 (22.5-42)

Results and Conclusions (cntd)

The overall median age was 37.3 years. Results of HJHS evaluations were presented in table 1. Overall HJHS were 3, 12 and 37.5 respectively in g1, g2 and g3 patients. In joint specific assessments among g1 patients, median (IQR) of elbows, knees and ankles were respectively 0 (0-0), 0.5 (0-2) and 2 (0-4). In comparison patients from g2 had median (IQR) score of 0 (0-8), 2 (1-7), and 4.5 (1-8) respectively for elbows, knees and ankles. Prophylaxis as dosed in Denmark, Norway and Sweden has given good long-term joint protection. Especially ankles were affected (2) in middle age groups, which emphasizes the need of better monitoring and treatment.

Keywords

Haemophilia A, Factor VIII, Prophylaxis, Joint outcomes, Registry

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