

ARTICULAR CONDITION IN CHILDREN AND ADOLESCENTS WITH SEVERE HEMOPHILIA A ON PROPHYLAXIS: CLINICAL ASSESSMENT AND ULTRASOUND EXAMINATION

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INTRODUCTION

Intra-articular damage in hemophilia patients may begin in childhood in response to clinical and subclinical bleedings even in those patients on prophylactic treatment. Identification of variables influencing the development of articular damage and tools for its early detection in childhood are needed.

OBJECTIVES

- 1- To determine the articular condition of patients < 18 year old (YO) on prophylaxis
- 2- To assess impact of type of prophylaxis and age of starting prophylaxis on the articular condition

METHODS

- Patients < 18 YO with severe Hemophilia A (SHA) on prophylaxis were invited to participated in the study.
- Last 12 months joint bleed rate (AJBR), age of first haemarthroses and start of prophylaxis were retrospectively collected. Current factor VIII (FVIII) trough levels and demographic data were also recorded.
- Articular condition was determined using the Hemophilia Joint Health Score (HJHS) and the HEAD-US score following the respective guidelines.
- Data are reported as median (min – max) or percentage as needed.
- P-value < 0.005 was assumed as statistical significant.

RESULTS

Thirteen SHA patients, age 8.3 (2.7-15.3) years old (YO) on prophylaxis were included. Ten patients (77%) and 3 patients (23%) were in primary prophylaxis (PP) and secondary prophylaxis (SP) respectively. Starting age of PP was 1.8 (0.3-7) YO and SP was 4.5 (4.3-8) YO. Median FVIII trough level was 0.3 (0.3-1.5) IU/dl. There was no correlation between trough levels and AJBR ($p= 0.111$). All patients on PP had AJBR=0 and HJHS=0. In this group, ultrasound determination showed only 1 patient (10% of total) (starting prophylaxis at 2.5 YO) with cartilage (HEAD-US score= 2) and bone (HEAD-US score= 1) deterioration at right ankle. In comparison with the PP group, patients on SP presented similar AJBR= 0 (0-6) ($p= 0.083$) but higher HJHS= 9 (0-17) ($p= 0.007$) and HEAD-US scores indicating synovial, cartilage and bone alterations for elbows and ankles in most of the patients.

CONCLUSIONS

- 1- HJHS and HEAD-US scores can be successfully measured in children even at early ages as 2.9 YO.
- 2- Both HJHS and HEAD-US score showed worse articular condition in children and adolescents on secondary prophylaxis compared with those in primary prophylaxis.
- 3- Elbows and ankles may be joints at risk of damage in pediatric patients even under prophylactic treatment.
- 4- HEAD-US score may be more sensitive than HJHS to identify joint cartilage and bone alterations.



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