2.05 Musculoskeletal issues

JOINT STATUS EVALUATION BY ULTRASOUND AND FUNCTIONAL ASSESSMENT IN PERSONS WITH NON SEVERE HEMOPHILIA ON PROPHYLAXIS WITH FOCUS ON THOSE WITH 5-YR FOLLOW UP

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Background and Aims. Chronic arthropathy causes morbidity in persons with hemophilia (PWH). and Joint damage can be detected also in non-severe (NS) PWH, even without overt bleedings. Identification of early signs of damage is crucial and point-of-care ultrasound (US) provides a rapid quantification of osteochondral and synovial changes. This study aimed to evaluate the joint status of a cohort of NSPWH on continuous prophylaxis (CP), focusing on US and functional assessments.

Materials and Methods. We reviewed clinical records of NSPWH currently on CP for severe phenotype or joint damage, collecting data about diagnosis, phenotype, treatment, Hemophilia Early Arthropathy Detection with Ultrasound (HEAD-US) and HJHS scores. In a subgroup of PWH with at least 5-yr follow-up the correlation between HEAD-US and HJHS scores at baseline and after a 5 years was evaluated, tracking the longitudinal progression of arthropathy.

Results. NSPWH on CP were 25/215: 5 with mild hemophilia A (HA), 14 with moderate HA (1 female) and 6 with moderate (hemophilia B) HB. Median age (range) was 32 years (4-67), at CP start 25 (0-63), being higher in mild HA than in moderate PWH (38, 11-63 vs 29, 3-62 years). Median baseline FVIII/FIX activity was 2% (1-10). Three moderate HA PWH were on CP with non replacement therapy, while the others with EHL factor concentrates. Baseline median total HEAD US score was 4, range 0-40. The score was ≥1 in 19/25 PWH (76%): 2 mild HA, 10 moderate HA, 6 moderate HB PWH, with highest rate for left ankle (LA, 17/25 ≥1, 68%). All but one PWH in this group, presented osteochondral change and again LA was the most affected joint by cartilage impairment (68%). 16 PWH has at least 5 years follow-up (2 mild HA, 4 moderate HB, 10 moderate HA; median age 34 years, 9-67). Median ABR in 5 years was 0.8 (0-3.8). Median HJHS was 9 (range 0-36) for this subgroup, being ≥1 in 12/16 PWH (75%). At baseline, a significant positive correlation was observed between HEAD-US and HJHS scores (Spearman r=0.869; p<0.001). After 5 years the correlation remained significant: r=0.759; p< 0.001. At longitudinal analysis, the mean HEAD-US score showed a negligible change (Δ=-0.50; Wilcoxon W=18.0, p=0.593). The mean HJHS score showed a trend to decrease (Δ=-2.00; Wilcoxon W= 36.0, p=0.298).

Conclusions. Our study showed joint damage in elevated rates of NSPWH, by both clinical and US assessments, highlighting the need for a regular follow up of joint status also in these patients, even in absence of overt bleedings. HEAD--US and HJHS scores exhibited a strong correlation over time, confirming that US mirrors clinical and functional joint impairment in hemophilic arthropathy and underlining the role of point-of-care US in the global care of hemophilia.