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ABSTRACT N. CS05

LE MALATTIE EMORRAGICHE CONGENITE

JOINT ULTRASOUND FOLLOW-UP IN PEDIATRIC PATIENTS WITH HAEMOPHILIA: A 12-YEAR PROSPECTIVE SINGLE-CENTER EXPERIENCE.

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Introduction. Periodic monitoring of joints status in patients with haemophilia (PwH) is recommended to identify and prevent the arthropathies. Ultrasound (US) is able to detect and quantify markers of disease activity and degenerative damage. The "Haemophilia Early Arthropathy Detection with Ultrasound" (HEAD-US) scoring scale is an accurate imaging method for identifying joint damage. Nevertheless, data on large cohorts of pediatric PwH, are still lacking.

Aim. To describe the joint status of 61 pediatric PwH assessed by US, using the HEAD-US protocol and analyse the efficacy of primary prophylaxis on the severe subset.

Materials and methods. We prospectively analyzed 61 pediatric PwH (severe hemophilia A=29, B=7; moderate A=8, B=2; mild A=7, B=8), referred to the "Haemophilia Centre, Regina Margherita Hospital, Turin, Italy" and collected data on joints status using the HEAD-US protocol, from November 2012 to November 2024. The Haemophilia Joint Health Score (HJHS) was performed in 44 patients and compared with the HEAD-US score.

Results. In total we performed 276 US exams, with a median of 4 exams per patient. Sorting our cohort into 3 age groups (early childhood: 1-5 years; school age: 6-14 years; adolescence and young adult age: 15-18 years) we found that in early childhood patients, the mean HEAD-US score was 0, in scholar age 0.56 (SD±1.17) and in adolescent and young adult patients was 1.76 (SD±2.22). Focusing the analysis on 31 patients with severe haemophilia treated with primary prophylaxis, we found that in early childhood patients mean HEAD-US score was 0, in scholar age 0.24 (SD±0.73) and in adolescent and young adult patients was 0.91 (SD ±1.38). In case of discordance between the HEAD-US and the HJHS scores (27%, HJHS=0 and HEAD-US>0), the HEAD-US score detected subclinical synovitis in 83% of cases.

Conclusions. Our data show that pediatric PwH treated with primary prophylaxis, show less joint damages compared to the same age patients, comprehending those not treated with primary prophylaxis. The HEAD-US score is a feasible and efficient clinical tool in pediatric PwH and, in our setting, it resulted even more sensitive in detecting subclinical joint damage, compared to the HJHS score.

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