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Effects of haemophilic arthropathy and age on subjective physical performance in people with severe haemophilia

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Introduction:

Adult people with severe haemophilia (PWH) suffer from intraarticular bleeding predominantly in large synovial joints, inducing haemophilic arthropathy (HA). HA leads to joint destruction which is accompanied by significant pain with consequences for physical performance [1]. A previous study has shown that HA increases with age [2]. However, less is known about the subjective physical performance (SPP) and its correlation with HA and age in PWH.

Subjects & Methods:

240 adults with severe haemophilia A (N = 212) or B (N = 28) had undergone an orthopaedic examination (knees, ankles, elbows) in order to evaluate their joint status. HA was classified applying the WFH physical joint examination instrument and pain scales (WFH score) [3]. Subjective physical performance was assessed by questionnaire HEP-Test-Q.

Tab. 1. *Anthropometric data; people with severe haemophilia (N = 240). The data are presented as mean ± standard deviation (min-max); n.a. (data not available)*

Age [years]	Height [m]	Weight [kg]	BMI [kg/m²]	HIV [N]	Hepatitis [N]
40 ± 12 (17 - 69)	1.78 ± 0.08 (1.55 - 1.96)	81.9 ± 14.6 (51.9 - 132.4)	25.3 ± 4.2 (17.7 - 42.9)	Yes: 48 No: 182 n.a.: 10	Yes: 166 No: 65 n.a.: 9

The WFH score is based on joint swelling, instability, axial deformity, crepitus on motion, muscle atrophy, range of motion and flexion contracture (Figure 1). A normal joint is scaled as zero and a higher score implies more distinct structural and functional joint deficits.

Fig. 1. *Items of orthopaedic examination for classification of ankle, knee and elbow status (WFH score)*

TShe Hep-Test-Q is a well-accepted and validated questionnaire assessing subjective physical performance of PWH, consisting of four domains 'mobility', 'strength & coordination', 'endurance` and 'body perception` [4]. The spearman rank test (Spearman`s rho [r_s]) was used to evaluate the correlation between the scores.

Results:

The total WFH score of PWH was 23 ± 12 (range: 3-63) and the total HEP-Test-Q score was 58 ± 22 (range: 0-100, higher values are better). Figure 2 shows the mean WFH scores for each joint. The values of the HEP-Test-Q domains were as follows: 'mobility' (63 ± 28, range: 0-100, N = 225), 'strength & coordination' (60 ± 27, range: 3-100, N = 225), 'endurance' (54 ± 22, range: 3-100, N = 217), 'body perception' (61 ± 23, range: 0-100, N = 225).

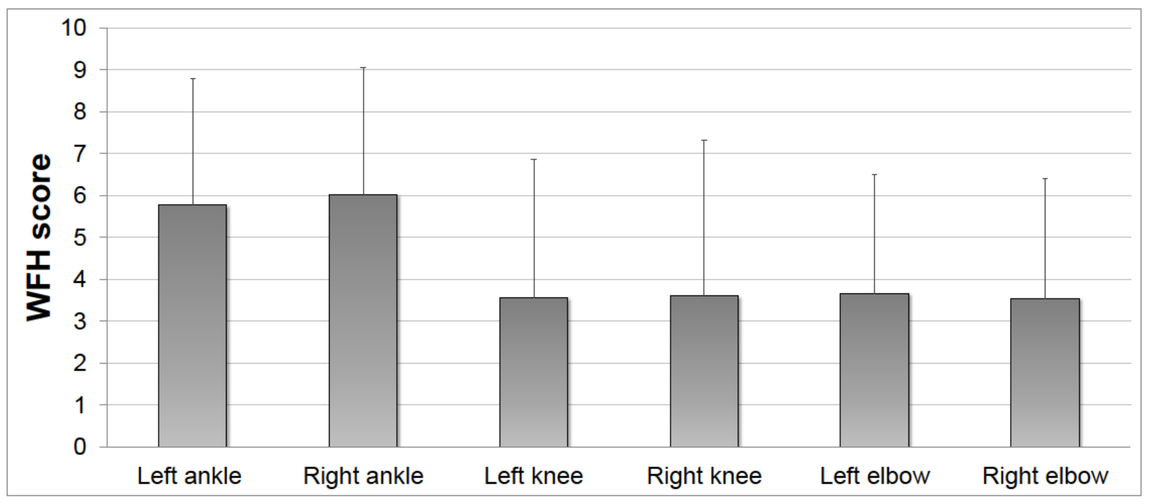

Fig. 2. *WFH score (mean ± standard deviation) for each joint in PWH (N = 240)*

A significant relationship was found between total WFH score and total HEP-Test-Q score (r_s = -0.63, p<0.001), whereby highest negative correlation existed for the HEP-Test-Q domain 'strength & coordination' (r_s = -0.72, p<0.001) (Figure 3). In contrast, the other domains showed only moderate negative correlations with the WFH score ('mobility': r_s = -0.57, 'endurance': r_s = -0.50; 'body perception': r_s = -0.37, p<0.001).

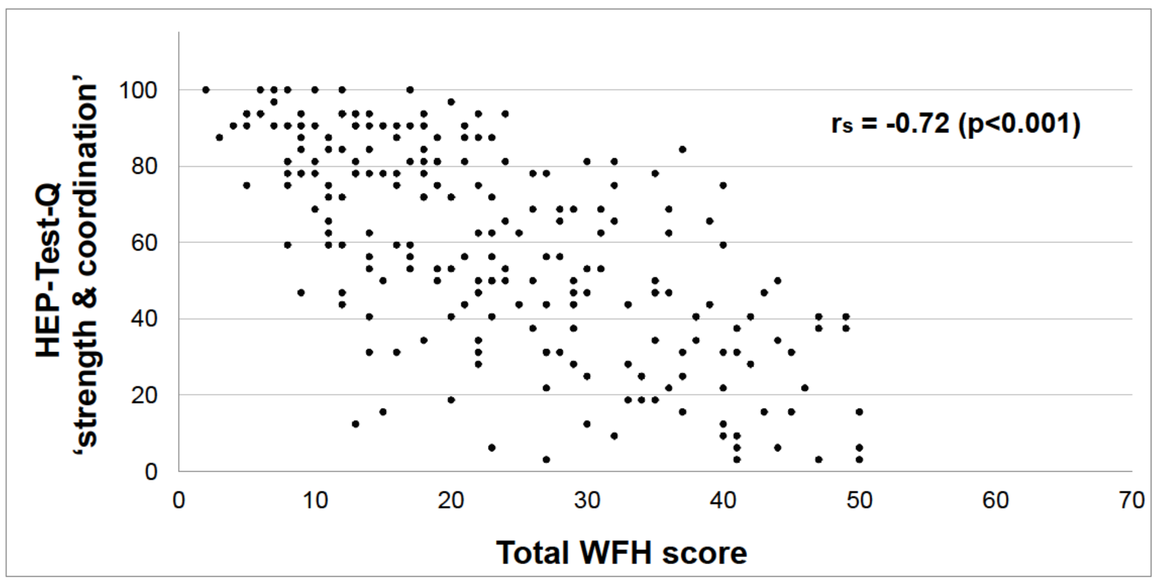

Fig. 3. *Scatterplot of the total WFH score and HEP-Test-Q domain ('strength & coordination') in PWH (N = 225)*

An inverse relationship was detected between age and 'strength & coordination' (r_s = -0.61, p<0.001), but also only moderate relationships were found between age and all other HEP-Test-Q domains ('mobility': r_s = -0.38, 'endurance': r_s = -0.42; 'body perception': r_s = -0.28, p<0.001).

Conclusion:

Our data indicate that HA obviously affects SPP more than age. Furthermore, this study provides evidence that worsening of joint status and increasing age were most clearly associated with impairment of strength and coordination. Therefore, the improvement of strength and coordination by sports therapy should be given a special attention in PWH.

References

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