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Age-related changes of joint status in adults with severe haemophilia in Germany

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Introduction:

People with haemophilia (PWH) are often affected by recurrent joint bleedings, commonly in ankles, knees and elbows. The consequence is the premature development of arthritis, termed haemophilic arthropathy (HA). Along with an increasing immobility for affected patients, HA leads to limited movements and malposition of joints accompanied by great pain. Due to recurrent bleedings during lifespan it can be assumed that the orthopaedic joint status worsens with age [1]. However, to date few data are available, which show a coherency of the orthopaedic joint status and age in PWH. The aim of this study was to analyse the age dependent orthopaedic joint status of PWH.

Subjects & Methods:

182 adults with severe haemophilia A (n=161) or B (n=21) and 81 non-haemophilic controls underwent an orthopaedic examination. The anthropometric data of the groups are shown in Table 1.

Tab. 1. *Anthropometric data; people with haemophilia (PWH) and controls. The data are presented as mean values ± standard deviation (min-max); age and Body Mass Index (BMI) with student`s t-test, height and weight with Mann-Whitney U-test.*

	PWH (n=182)	Controls (n=81)	p-value
Age [years]	40 ± 12 (18-67)	40 ± 14 (20-68)	0.845
Height [m]	1.78 ± 0.07 (1.62-1.96)	1.80 ± 0.68 (1.67-1.94)	0.005
Weight [kg]	81.9 ± 14.5 (51.9-132.4)	82.4 ± 11.4 (55.9-107.1)	0.277
BMI [kg/m²]	25.9 ± 4.2 (17.7-42.9)	25.3 ± 3.2 (19.4-36.1)	0.202

For assessment of the joint status of ankles, knees and elbows we used the joint physical examination instrument (WFH clinical score; Figure 1), which was introduced by Gilbert in 1993 [2]. The WFH score is recommended by the Orthopaedic Advisory Committee of the World Federation of Hemophilia and contains the following parameters: swelling, muscle atrophy, axial deformity, crepitus, range of motion, flexion contracture and instability. A higher score implies more distinct structural and functional joint deficits. The highest achievable score for the total WFH clinical score is 64. Correlations were determined using nonparametric spearman rank test (Spearman`s rho [r_S]). Statistical significance is defined as $p \leq 0.05$ using the SPSS 20.0 statistical package.

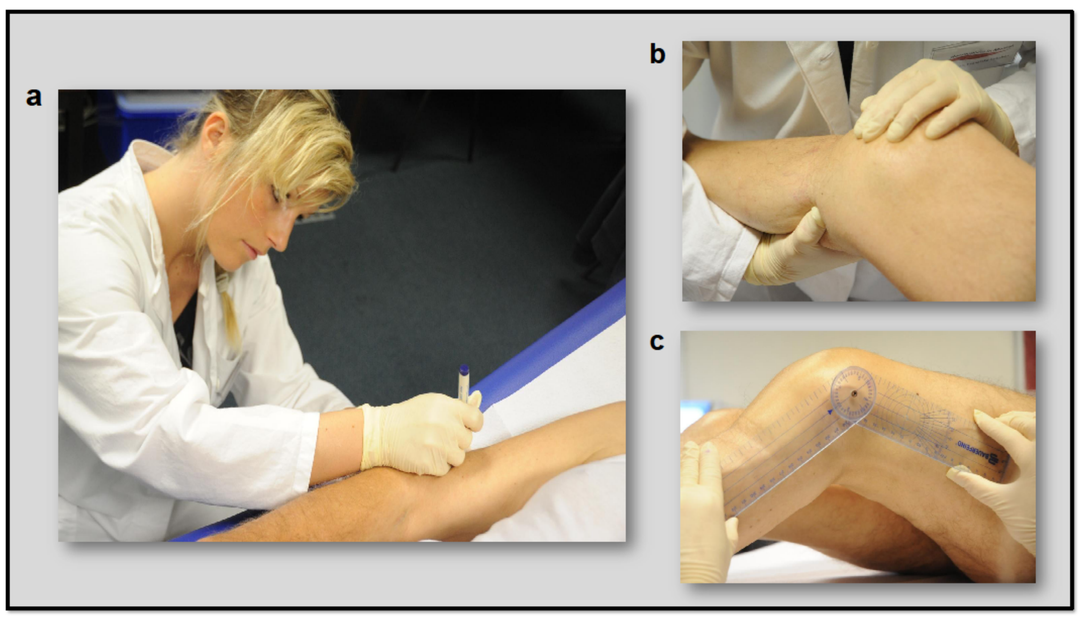

Fig. 1. *Orthopaedic examination for classification of ankle, knee and elbow status. (a: muscle atrophy, b: crepitus, c: range of motion)*

Results:

The median total WFH clinical score was 22 (IQR: 13-34), whereby ankles were the most affected joints in PWH. The WFH clinical score for each joint pair was highest for the ankles at 5 (IQR: 3-7), followed by knees at 2 (IQR: 1-7) and elbows with a median score of 2 (IQR: 1-6) (Figure 2).

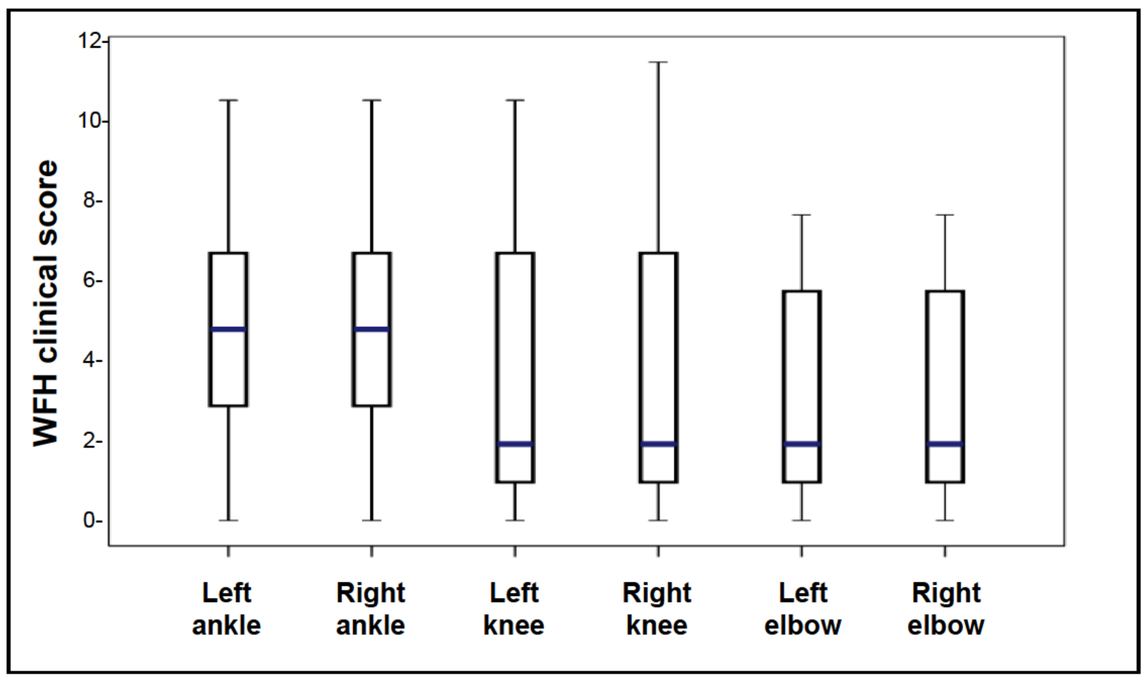

Fig. 2. *WFH clinical scores for each joint in PWH presented as box-plot with whiskers with maximum 1.5 interquartile range (IQR). The horizontal bar in the middle of boxes represents the median (n=182).*

A significant correlation was found between total WFH clinical score and age (r_S=0.77, p<0.001) (Figure 3). Moreover, the analysis of each joint also revealed a significant correlation with age (ankle left: r_S=0.54; ankle right: r_S=0.58; knee left: r_S=0.58; knee right: r_S=0.62; elbow left: r_S=0.55; elbow right: r_S=0.48; p<0.001). In contrast, no significant correlations were detected for the WFH clinical score and age in controls.

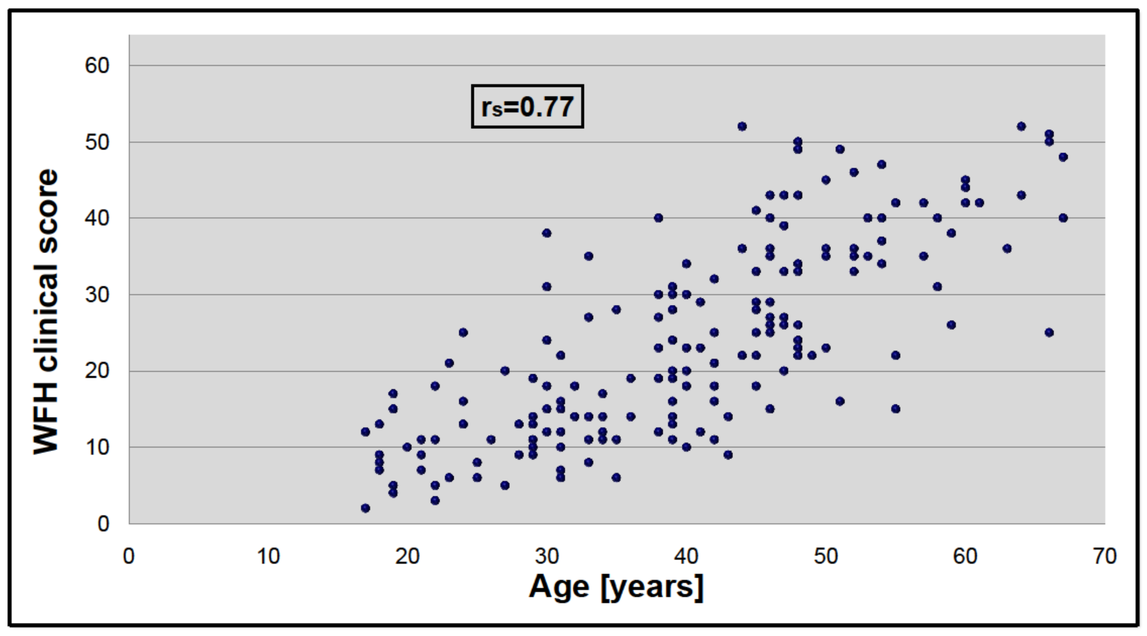

Fig. 3. *Scatterplot of the WFH clinical score and variables age in PWH (n=182).*

Conclusions:

Our data clearly show that the orthopaedic joint status in PWH highly correlates with age in contrast to people without haemophilia. Furthermore the most pronounced structural and functional joint deficit was found for the ankle, which becomes more and more important for today and in future.

References

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