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ORIGINAL ARTICLE *Clinical haemophilia*

The impact of sport on health status, psychological well-being and physical performance of adults with haemophilia

S. VON MACKENSEN,* C. HARRINGTON,† E. TUDDENHAM,† A. LITTLEY,‡ A. WILL,‡ M. FAREH,§ C. R. M. HAY§ and K. KHAIR¶

**Institute of Medical Psychology, University Medical Centre Hamburg-Eppendorf, Hamburg, Germany; †Katherine Dormandy Haemophilia Centre and Haemostasis Unit, Royal Free London NHS Foundation Trust, London; ‡Haemophilia Centre, Royal Manchester Children's Hospital, Central Manchester Foundation Trust, Manchester; §University Dept of Haematology Manchester Royal Infirmary, Manchester; and ¶Haemophilia Centre, Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK*

Background: There is increasing recognition that sport is important for individuals with haemophilia; however, there remains a paucity of data of the importance of this in adults, many of whom already have joint pathology related to childhood bleeds and treatment access. This multicentre, cross-sectional study presents the impact of sport on health-related quality of life (HRQoL), physical performance and clinical outcomes in adults with haemophilia. **Results:** Fifty adults aged 35.12±14.7 with mild ($n = 12$), moderate ($n = 10$), or severe ($n = 28$) haemophilia A (70%) or B (30%) from four haemophilia centres across the United Kingdom participated in the study. A total of 64% were overweight/obese according to their BMI; median orthopaedic joint scores using the WFH Orthopaedic Joint Score (OJS) were 6 (range 0–48). On a VAS pain scale (range of 0–10), patients reported mean score of 5.66 ± 2.4. 36% of participants reported not doing any sport, mainly due to their physical condition. However, 64% of participants reported undertaking sporting activity including contact sports, mostly twice per week in average 4 h week^{-1}. Participating in sport did not have a statistically significant impact on HRQoL; except in the domain 'sport and leisure' of the Haem-A-QoL. Patients doing more sport reported significantly better HRQoL than those doing less sport ($P < 0.005$). Those doing sport for more than 4 h week^{-1} had a significantly better physical performance than patients doing less sport (assessed with Hep-Test-Q). Encouraging physical activity and sport in older patients with haemophilia may have a direct impact on their HRQoL; thus, education about sport activity should be incorporated into routine haemophilia care.

Keywords: adults, haemophilia, health-related quality of life, physical performance, sport, sporting activity

Introduction

Haemophilia is an X-linked, usually inherited disorder of coagulation factors VIII (haemophilia A) or IX (haemophilia B) which occurs in approximately 1:5000 – 1:10 000 live births in the UK [1]. Haemophilia causes painful, spontaneous and trauma-related bleeding into soft tissues and weight-bearing joints [2]. In 1958, Biggs and Macfarlane categorized haemophilia as severe (with factor levels of <1 iu dL^{-1}), moderate (levels 1–5 iu dL^{-1}) and mild haemophilia (5–50 iu dL^{-1}) according to factor activity level (normal range 50–150 iu dL^{-1}) [3]. Bleeding severity correlates inversely with factor level. Thus, those with the lowest factor level are most clinically affected experiencing spontaneous or trauma-related bleeding from early childhood. Without factor replacement blood-induced joint damage occurs, resulting in haemophilic arthropathy in adults [4]. Due to sedentary life styles, people with haemophilia (PWH) often have an increased body mass index (BMI) [5], which may exacerbate their joint problems [6] as being overweight is recognized to impact on articular cartilage and ulti-

Correspondence: Sylvia von Mackensen, Institute of Medical Psychology, University Medical Centre Hamburg-Eppendorf, Martinistr. 52, 20246 Hamburg, Germany.
Tel.: +49 172 6822759; fax: +49 89 762316;
e-mail: s.mackensen@uke.uni-hamburg.de

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mately joint function [7]. Ullman *et al.* [8] report that overweight or obesity impacts on the ability to self-infuse due to difficult venepuncture related to adiposity, which impacts on time to treatment of bleeds and prophylaxis leading to worsening joint function.

Until recently, it was usual practice to discourage sporting activity (physical exertion which may be competitive in nature) for PWH because of the bleeding risk [9]. Many contemporary clinicians are more flexible about this believing that patients gain physical [10], social [11] and psychological [12] well-being from sporting activity. Tailored prophylactic therapy administered to prevent bleeds, minimize disability and improve quality of life (QoL) [13], which means that many older PWH are now able to participate in individualized sport. Despite this, adults with haemophilia consider their physical performance to be comparable to healthy age-matched controls [14]. There is, however, little data to support the non-physical benefits of sport in adults. Therefore, a multicentre study into the 'Evaluation of the Impact of Sport Activities on Health-Related Quality of Life of Haemophilia Patients' (EIS Study) was designed.

Study design and methods

The EIS study aimed to recruit up to 400 children (aged 6–17) and adults with haemophilia (aged 17–65), and parents of children. While data for children have been already published [15], this paper focuses only on the data from adults.

Seventy-five adults with haemophilia of any severity or type, with or without inhibitors aged 17 years or older from four haemophilia care centres (HTC) [two adult, two paediatric centres with patients in that age range] in the UK were invited to participate in the study. Participants were requested to complete questionnaires which studied the impact of sport on their lives. Validated questionnaires were selected specifically for the study to collect data on health-related quality of life (HRQoL) and physical performance. In addition, data were collected concerning sporting activities (e.g. frequency of sport per week, and number of hours spent participating in sport) and attitudes towards sport. Questionnaires were completed following patients' written consent at routine haemophilia appointments, information about the study was sent to patients two weeks before their clinic visits to enable them to have time to consider study participation. Ethical approval for the study was granted, by a local research ethics committee, before any study procedures were undertaken. Clinicians completed medical documentation including information about type and severity of haemophilia, bleeding, inhibitor history, concurrent illness, type and schedule of treatment and frequency of medical visits. The orthopaedic status was evaluated by the haemophilia specialist physiotherapist at each participating centre.

Instruments

Health-related quality of life was assessed using the disease-specific **Haem-A-QoL** [16,17], which consists of 46 items pertaining to 10 dimensions ('physical health', 'feelings', 'view of yourself', 'sports & leisure', 'work & school', 'dealing with haemophilia', 'treatment', 'future', 'family planning', 'relationships') and one total score with high values (range 0–100) indicating high impairments in HRQoL. The 'sport & leisure' domain of the Haem-A-QoL consists of five items: 'I had to refrain from sport because of haemophilia', 'I had to refrain from sport like soccer', 'I did just as much as others', 'I didn't have the freedom to travel', and 'it was necessary for me to plan everything in advance' which ask specifically about sport and leisure activities.

Physical performance was assessed by a patient-reported outcome using the HEP-Test-Q [18] and by an objective measure assessed by clinicians using the WFH Orthopaedic Joint Score [19]. The **HEP-Test-Q** assesses subjective physical performance and consists of 26 items pertaining to four dimensions (mobility, strength and coordination, endurance and body perception) with high values (range 0–100) indicating better physical performance. The **WFH Orthopaedic Joint Score** (**OJS**) measures joint health in the domain of body structure and function (i.e. impairment) of the joints most commonly affected in haemophilia (elbows, ankles and knees). Primarily designed for use in patients with established arthropathy, it is used as a physiotherapy outcome measure of joint function. The original OJS is composed of a clinical score (e.g. swelling, muscle atrophy, crepitus), a pain score and a bleeding score. All three scores are combined into a total score where high values imply worse impairment of the orthopaedic status. In the EIS study, we used a modified version and calculated only the clinical score. Three patients (17–18 years), from two paediatric centres underwent the Petrini haemophilia joint score [20] rather than the OJS in keeping with routine clinical care at that centre.

Statistical analysis

All statistical analyses were conducted using the SPSS program version 22 (SPSS Inc. Chicago, IL, USA). Descriptive data are shown as frequency distribution in percent or as mean ± standard deviations SD (range), median and interquartile ranges (IQR) and were tested for normal distribution using the Shaipiro–Wilk test. The comparison of differences between groups was examined by Student's test or Mann–Whitney *U*-test according to distribution; P values <0.05 were defined as significant.

In order to investigate the impact of sport on patients' well being (including clinical data in terms of

BMI, OJS, number of target joints, number of total bleeds, number of joint bleeds and number of days lost, HRQoL and physical performance), we considered the following variables: participating in sport (yes vs. no), sedentary life style (<1–2 h per day in front of television (TV) or computer vs. ≥3 h per day); for those doing sport, we distinguished those haemophiliacs who undertook sporting activities sport 1–2 times per week vs. ≥3 times per week, and those doing <4 h of sport per week vs. those participating in sporting activities ≥4 h per week. To determine the cut-off point for these variables, the median split was calculated.

Results

Socio-demographic data

The majority of PWH were single (59.2%). A total of 56% had at least a secondary level educational qualification, with 50% working part or full time. The vast majority (92%) lived within small or large cities; this probably reflects the geography of the centres participating in this study (see Table 1). There was no significant difference between severe and mild/moderate patients regarding socio-demographic data. By contrast, more severe patients (18.5%) spent >4 h in front of television/computer ($P < 0.014$) than mild/moderate patients (4.5%).

Table 1. Socio-demographic data ($n = 50$).

Socio-demographic data	*N*	Percentage
Marital status*		
Single	29	59.2%
Married	15	30.6%
Widowed	0	0%
Divorced	5	10.2%
Living with a partner*		
Yes	21	43.8%
No	27	56.2%
Number of children*		
0	27	60%
1	8	17.8%
2	7	15.6%
3	3	6.7%
Educational qualification†		
No qualification	6	12%
Lowest formal qualification	16	32%
Highest secondary qualification	18	36%
University degree	10	20%
Working		
Full-time	22	44%
Part-time	3	6%
Studying	9	18%
Looking after the home full-time	2	4%
Not employed/looking for work	11	22%
Retired	3	6%
Living in		
A big city	12	24%
The suburbs of a big city	13	26%
A town or small city	21	42%
A country village	4	8%

*Missing data.
†Lowest formal qualification = GCSE/O-Level; highest secondary qualification = NVQ or A-Level.

Clinical data

From the 75 adults invited to participate, 50 PWH were enrolled into the study (67%) with a mean age of 35.12 ± 14.7 years (range 17–66). A total of 70% had haemophilia A and half were severely affected (56%). Here, 54% had regular prophylaxis; 26% reported targeted prophylaxis before sport and three quarter had target joints. A total of 64% were overweight or obese according to their BMI [BMI = 25–29 or BMI = 30–100 respectively] (see Table 2). Significant differences between severe and mild/moderate patients were found (see Table 2).

Mean BMI values by age categories are shown in Table 3. No significant difference was found for BMI across age categories with the highest BMI (M = 29.24 ± 7.5) in the age group 55–64 years. Patients mildly affected by haemophilia had a trend to a higher BMI (M = 31.49 ± 10.3) compared to those with severe (M = 26.79 ± 4.1) or moderate haemophilia (M = 25.52 ± 6.5). Compared with the National Health Survey of the UK from 2010 [21], the prevalence of obese and overweight haemophilia patients in the age category 17–24 years was nearly two times the national average; in the age category 25–34 years, it was slightly higher, which might be due to the fact that in our cohort only a few patients were enrolled per age group. In the elder age groups (45–74 years), a different trend was seen; compared with data from the NHS survey, less PWH were overweight or obese (see Fig. 1). This could be explained as 11 of 17 PWH in these elder age groups had chronic hepatitis and three had HIV infection.

Overall bleeds were reported at a median of 2 (range 0–52, IQR = 5) in the 6 months preceding questionnaire completion; of these, a median of 0 (range 0–40, IQR = 3) was joint bleeds and 0 bleeds in median (range 0–4, IQR = 3) were attributed to sport. The WFH Gilbert/Petrini scores attained a median of 6.0 (range 0–48, IQR = 16.25). A visual analogue scale (VAS) was used to record chronic pain, (ranging from 0 to 10, where a score of 10 indicates maximal pain); 27 reported chronic pain in the preceding 6 months (54%) with a mean pain score of 5.66 ± 2.4 (range 0–10). A total of 56.3% of patients who reported chronic pain still participated in sport. Fourteen PWH missed days at work in the preceding 6 months due to haemophilia with a mean of 11.14 ± 11.7 (range 1–45) days lost; none due to sporting injury (see Table 4). A significant difference in clinical data was seen in the OJS ($P < 0.0001$) between severe patients (Med = 10, range 0–48, IQR = 22) and mild/moderate patients (Med = 1, range 0–46, IQR = 6).

Table 2. Clinical data of adult patients with haemophilia (n = 50); differentiated between severe and mild/moderate patients.

Clinical data	∑ (n = 50)	Severe (n = 28)	Mild/Moderate (n = 22)	χ^2
Severity				
Severe	28 (56%)			
Moderate	10 (20%)			
Mild	12 (24%)			
Type of haemophilia				
A	35 (70%)	21 (75%)	14 (63.6%)	*ns*
B	15 (30%)	7 (25%)	8 (36.4%)	
Inhibitor: past or actual	4 (8%)	3 (10.7%)	1 (4.5%)	*ns*
Tolerized	*3 (75%)*	*3 (100%)*	*0 (0%)*	
Current	*1 (25%)*	*0 (0%)*	*1 (100%)*	
Type of treatment				
On demand	23 (46%)	6 (21.4%)	17 (77.3%)	0.0001
Prophylaxis	27 (54%)	22 (78.6%)	5 (22.7%)	
Type of prophylaxis				
Primary	5 (18.5%)	2 (9.1%)	3 (60%)	0.030
Secondary	21 (77.8%)	19 (86.4%)	2 (40%)	
Intermittent	1 (3.7%)	1 (4.5%)	0 (0%)	
Prophylaxis prior to sports	13 (26%)	8 (28.6%)	5 (22.7%)	*ns*
Home treatment	40 (80%)	27 (96.4)%	13 (59.1%)	0.001
Presence of target joints:	37 (74%)	25 (89.3%)	12 (54.5%)	0.007
Chronic viral infections	22 (44%)	16 (57.1%)	6 (27.3%)	0.033
Type of Hepatitis				
HBV	*1 (4.5%)*	*0 (0%)*	*1 (16.7%)*	
HCV	*20 (91%)*	*16 (100%)*	*4 (66.6%)*	
HBV & HCV	*1 (4.5%)*	*0 (0%)*	*1 (16.7%)*	
HIV infection	6 (12%)	5 (17.9%)	1 (4.5%)	*ns*
Presence of chronic pain	27 (54%)	19 (67.9%)	8 (36.4%)	0.026
BMI				
Under weight	2 (4%)	0 (0%)	2 (9.1%)	*ns*
Normal weight	16 (32%)	9 (32.1%)	7 (31.8%)	
Overweight	19 (38%)	13 (46.4%)	6 (27.3%)	
Obesity	11 (22%	6 (21.4%)	5 (22.7%)	
Morbidly obese	2 (4%)	0 (0%)	2 (9.1%)	

ns, not significant.

Table 3. BMI across age categories (based on the NHS obesity classification).

Age groups	Number of patients N (%)	BMI Mean ± SD	BMI P
17–24	16 (32%)	28.95 ± 9.7	*ns*
25–34	13 (26%)	26.91 ± 3.6	
35–44	4 (8%)	24.78 ± 3.7	
45–54	11 (22%)	27.19 ± 5.8	
55–64	5 (10%)	29.24 ± 7.5	
65–74	1 (2%)	25.74 ± 0	
Total	50 (100%)	27.67 ± 6.8	

SD, standard deviation; *ns*, no significant difference across age categories.

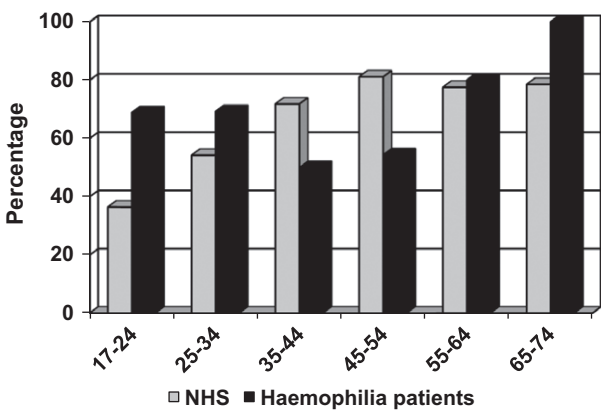

Fig. 1. Comparisons of frequency of overweight/obese patients across age groups compared with the UK National Health Survey.

No differences in clinical data in terms of BMI, number of days lost from work, number of total bleeds, number of joint bleeds and number of target joints were found for *sedentary lifestyle*, *doing sport*, *frequencies of sport* and *hours of sport*. Those doing sport 1–2 times a week had a worse orthopaedic status (M = 16.0 ± 18.2) compared with those doing sport three times a week or more (M = 9.38 ± 11.3), even though this difference was not significant. This trend was also found for hours of sport per week, as well as not significant; patients doing less than 4 h sport per week had a worse OJS (M = 15.33 ± 14.6) compared with those doing more than 4 h sport per week (M = 7.24 ± 11.3).

Sporting activity

Of the 50 participants, 18 reported not doing any sport (36%). The reasons for this were mainly their

Table 4. Clinical data of adult patients with haemophilia (n = 50).

Clinical data	M ± SD	Median [IQR]
BMI	27.67 ± 6.8	26.27 [7.18]
Orthopaedic status (WFH Score)	11.27 ± 13.4	6 [16.25]
No of total bleeds in the past 6 months	4.58 ± 9.2	2 [5]
No of joint bleeds in the past 6 months	3.56 ± 7.7	0 [3]
No of sports-related bleeds in the past 6 months	0.14 ± 0.6	0 [0]
Number of days lost in the past 6 months:		
Haemophilia-related	11.14 ± 11.7	9 [12.75]
Sport related	0	0

SD, standard deviation; IQR, Interquartile Range [Q3-Q1].

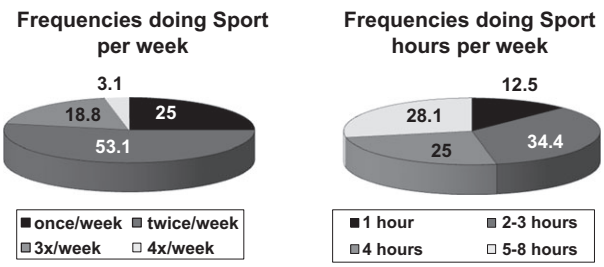

Fig. 2. Frequency and time doing sport.

physical condition (n = 14) and that they were afraid of hurting themselves (n = 8), nine reported that they were not allowed to do it by their parents (in the past) or their doctor and three mentioned that they did not like it, other reasons were time constraints or previous negative experience (more than one reason was given by some).

Thirty-two (64%) did sport; there was a statistically significant difference by age group ($P < 0.048$) with the age group 35–44 doing less sport than those aged 17–34 or 45–54 years; a significant difference was found as well by severity ($P < 0.017$), more severe patients (78.6%) did sport compared with only 45.5% of the mildly/moderately affected patients.

Patients reported participation in an average of two sports each; with the majority doing each sport twice weekly. They mainly performed sport with friends (81.3%); 37.5% did sports in a team or sports club and 50% at other places such as golf course or gym. Frequency and time doing sport are shown in Fig. 2. In total, men were doing a median of 4 h of sport per week (M = 3.71 ± 1.7). An extensive array of sporting activity was reported with the top sports being: swimming (50%), football (34.4%), jogging (25%), walking (18.8%), gym (18.8%), golf (15.6%) and cycling (15.6%). Compared with the children's study [13], adults reported less intensive impact or contact sports; with children reporting football and jogging as more frequently performed sports than swimming in adults. This may reflect joint function and pain in the adult cohort.

All PWH thought that doing sport was good, with arguments that it is: 'healthy and keeps you fit' (n = 19), 'social' (n = 7), 'good for self-esteem' (n = 6), 'keeping you competitive' (n = 4) and 'fun' (n = 4). Other arguments in favour of sport were that it 'helped maintain weight', was 'good for muscle strength' and 'keeps joints supple'. Twenty-eight said they would have liked to play sports [football (n = 13), rugby (n = 4), martial arts (n = 3), any/all sport (n = 3), basketball (n = 1), bungee jumping (n = 1), cricket (n = 1), ice hockey (n = 1) wrestling (n = 1)], but were advised against it. Despite this, 22 admitted having participated in football (n = 11), rugby (n = 2), martial arts (n = 2), basketball (n = 1), cricket (n = 1), wrestling (n = 1) or a variety of sports (n = 4). They reported adverse events related to sport, namely bleeds (n = 10), pain (n = 9), sports injury (n = 5) and decreased mobility (n = 1). Four patients thought that sport was bad or dangerous (8%): three considered specific sports such as football and rugby as dangerous; two were afraid of getting injured and one did not want to do it without professional help. Thirty-two reported that they would like to continue sports in the future (64%), 21 indicated they would also like to try a new sport (42%) including swimming (n = 7), football (n = 3), cricket (n = 2), running (n = 2), ice hockey (n = 1), gym (n = 1), fitness training (n = 1) and wheelchair sport (n = 1).

Health-related quality of life

The haemophilia-specific HRQoL was generally acceptable, apart from the physical domains. The highest impairment was reported in the domains 'sport and leisure' (M = 56.82 ± 5.1), 'view' (M = 46.9 ± 18.8) and 'physical health' (M = 43.98 ± 25.5) (see Fig. 3). Those with chronic pain reported significant higher impairments in the domains 'physical health' ($P < 0.0001$), 'sport & leisure' ($P < 0.002$), 'future' ($P < 0.002$) and the 'Total Score of the Haem-A-QoL' ($P < 0.008$) (see Table 5). Patients with a higher OJS reported a significant worse ($P < 0.031$) 'physical health' (M = 52.01 ± 24.0) compared with those with a lower OJS (M = 36.0 ± 25.7). Patients severely affected by haemophilia showed higher impairments in the domains 'sport & leisure' ($P < 0.045$) and 'treatment' ($P < 0.025$), while mildly/moderately affected patients reported higher impairments in 'dealing' ($P < 0.009$).

Sport did not prove to have a high impact on HRQoL. No differences were found between those doing sport vs. those not doing sport and for those with a sedentary lifestyle compared with those with a less sedentary lifestyle. No significant difference was found for those doing sport 1–2 times a week sport compared with those doing sport three times a week or more. Only one significant difference was found in

HRQoL in the domain 'sport and leisure' ($P <= 0.005$) for those doing less than 4 h sport per week (M = 68.25 ± 19.6) compared with those doing more than 4 h sport per week (M = 42.40 ± 27.2) (see Fig. 4).

Physical performance

Psychometric testing of the English version of the HEP-Test-Q revealed excellent values in terms of reliability and construct validity. Internal consistency in terms of Cronbach's alpha ranged from α = 0.859 for 'strength & coordination' to α = 0.949 for the 'HEP-Test-Q total score' (see Table 6); these values are similar to the German original HEP-Test-Q version [16]. The English version of HEP-Test-Q also showed good values for known group's validity demonstrating that PWH who suffered from chronic pain reported in all subscales a significant lower subjective physical performance than those without pain (see Fig. 5).

In general, PWH reported good physical performance (M = 60.11 ± 21.0) with highest impairments in the dimensions 'mobility' (M = 50.38 ± 28.9) and 'endurance' (M = 58.70 ± 21.5) (see Table 5). Those with a high OJS showed significant worse values in 'mobility' ($P < 0.001$) and the 'Total HEP-Test-Q Score' ($P < 0.016$) compared with those with a lower OJS. Patients severely affected reported worse values in 'mobility' ($P < 0.003$) compared with those with mild/moderate haemophilia (M = 40.18 ± 26.4 vs. M = 63.99 ± 26.9).

There was no significant difference between PWH doing vs. not doing sport, for sedentary lifestyles and for the frequency of doing sports in physical performance. Those doing less than 4 h sport per week reported significantly worse ($P < 0.007$) physical performance in all but in the dimension 'mobility' compared with those doing ≥4 h sport per week (see Fig. 6).

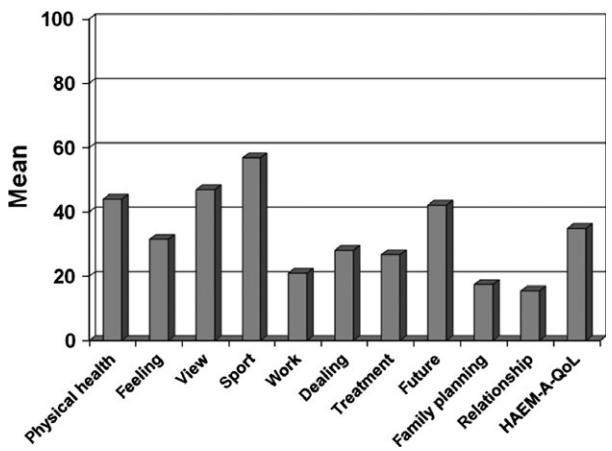

Fig. 3. Health-related quality of life profile (Haem-A-QoL).

Table 5. Health-related quality of life in adults with and without chronic pain.

	Chronic pain		
Haem-A-QoL	No Mean ± SD	Yes Mean ± SD	*P*-value
Physical health	28.26 ± 22.1	57.36 ± 20.2	0.0001
Feeling	26.63 ± 29.4	35.65 ± 25.2	*ns*
View	42.83 ± 19.0	50.37 ± 18.3	*ns*
Sport	45.30 ± 26.3	67.84 ± 18.3	0.002
Work	14.29 ± 22.1	27.68 ± 25.5	*ns*
Dealing	33.33 ± 26.2	23.40 ± 17.0	*ns*
Treatment	23.43 ± 19.9	29.57 ± 17.7	*ns*
Future	31.74 ± 21.4	50.93 ± 21.0	0.002
Family Planning	13.33 ± 19.0	21.35 ± 29.1	*ns*
Relationship	13.77 ± 21.7	17.00 ± 29.2	*ns*
Total	28.46 ± 15.4	40.59 ± 15.2	0.008

SD, standard deviation; *ns*, not significant.

Discussion

The majority of adults in our cohort reported good physical ability, this may be in part due to prophylaxis; with over 50% receiving primary, secondary or intermittent prophylaxis, or may reflect that 58% of participants were in the younger age range (17–34 years). The majority (80%) were on home treatment, which enhances early self-infusion in the event of bleeding, which helps to reduce further joint damage [8]. In this relatively small study of sporting activity in PWH, almost two-thirds of the participants reported participation in a wide range of sport.

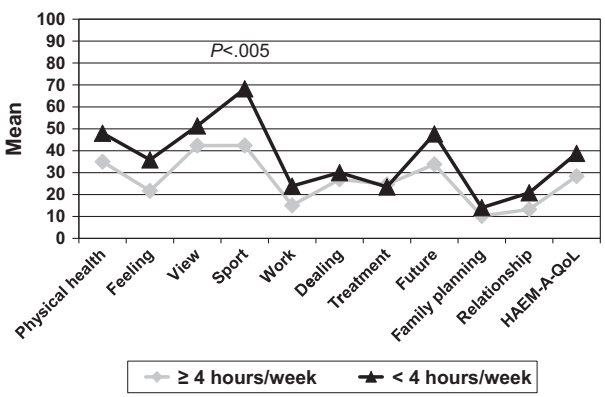

Fig. 4. Comparison of HRQoL (Haem-A-QoL) across 32 adult haemophilia patients doing sports more than 4 h week^{-1} compared with those doing sports less than 4 h week^{-1}.

Table 6. Psychometric characteristics of the HEP-Test-Q.

Domains	No of items	Mean	SD	Range	Cronbach's α
Mobility	4	50.38	28.9	0–100	0.920
Coordination	8	64.87	22.1	15.63–100	0.859
Endurance	8	58.70	21.5	15.63–100	0.866
Perception	5	62.55	25.9	5–100	0.870
HEP-Test-Q	25	60.11	21.0	13–100	0.949

SD, standard deviation.

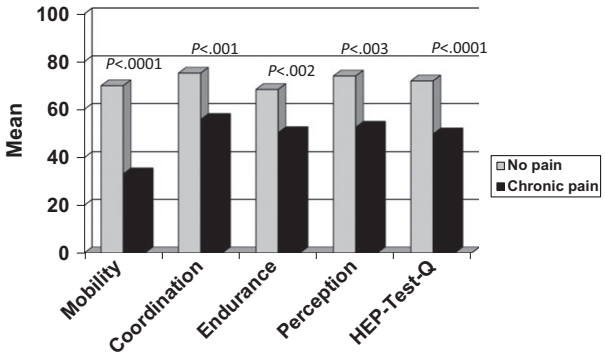

Fig. 5. Differences in physical performance between patients with and without chronic pain (HEP-Test-Q).

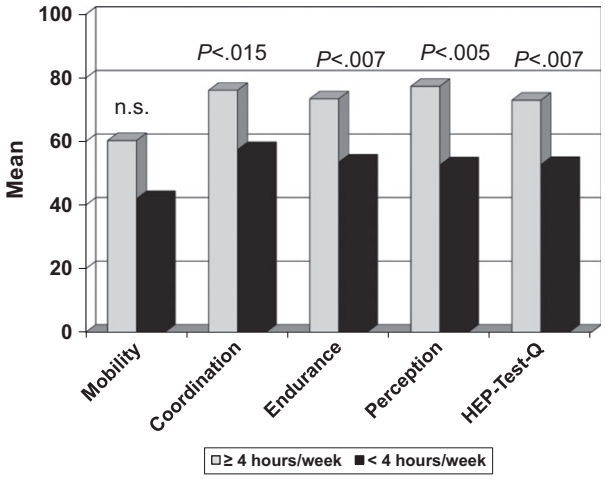

Fig. 6. Physical performance in 32 adults doing sport less than 4 h week^{-1} compared with those doing sports 4 or more hours per week (HEP-Test-Q).

Sport can not only improve physical health, but has also an impact on HRQoL. Highest impairments in HRQoL were reported in this study in the 'sport and leisure', 'view', 'physical health' and 'future' domains of the Haem-A-QoL. Sport did not have a high impact on these findings other than in the domain 'sport and leisure' where doing more sport led to a better reported HRQoL. Significant differences concerning subjective physical performance were found in all domains of the HEP-Test-Q with the exception of 'mobility' between those doing vs. those not doing sport. In men who reported chronic pain (54%), physical performance was significant worse in all domains of the HEP-Test-Q and for the HRQoL domains 'physical health', 'sport & leisure', 'future' and the 'Total Score' of the Haem-A-QoL.

Time of sporting participation was calculated by adding each sporting activity and time duration, resulting in an estimation of hours spent practicing sport per week. The majority (34.4%) reported four hours of sport participation per week. The level of sporting activity seen in this cohort of men with haemophilia was higher than might be expected, and is higher than the 150 min week^{-1} of moderate aerobic activity for adults aged 19–64, recommended by the UK Government [22]. This may be because of methodological issues with the questionnaire; we failed to ask open-ended questions about exact timings for sport participation. We have assumed that men doing sport for less than 1 h ticked the answer category '1 h', which may have resulted in a higher total amount of hours doing sport, this is a systematic error for all participants which did not lead to incorrect results. However, this result should be borne in mind when compared with other surveys.

Understanding the impact of sport on the health status and well-being of PWH is a relatively new concept; many of these men had limited access to treatment in the past when sport was not recommended. Recent attention has been given to the potential benefits of encouraging sport in this cohort of patients; Negrier *et al.* [23] recognized the physical and psychosocial benefits of sporting participation. Additional benefits of sporting activity such as improved personal physical health and being able to play with their own children/grandchildren have been cited as reasons to continue sport [24]. Sherlock *et al.* [25] reported an Irish cohort, where 55% of participants suffered sport-related injury and described this as an acceptable risk continuing to view sports participation in a positive light. Sporting injury can be prevented by appropriate training programmes [26,27] and by tailored prophylaxis [28].

McNamara *et al.* [29] reported the role of sport on maintaining a recommended BMI; reducing muscle atrophy and improving OJS are recognized as positive physical impacts of sporting participation. They also described how BMI in patients >41 years of age with severe haemophilia is reduced possibly due to muscle atrophy. In our study, we have also seen lower BMI in patients aged 35–64 when compared with the UK national cohort.

Ullman *et al.* [8] state that only 40% of American patients are on home therapy suggesting that 'more obese patients are less likely to self-infuse because of difficulties caused by adiposity', therefore raised BMI can impact on an individual's ability to self-infuse, which may in turn lead to loss of physical function and sporting activity. In our study, we found no difference in overweight/obese patients between those having home treatment and those not. In our cohort, 54% reported chronic pain compared with an EU survey where only 35% reported chronic pain [30], which was reported to restrict daily activities (89%) and mood (85%).

Elander [31] reported that HRQoL is worst in those with significant arthropathy; resulting in negative emotional thoughts, fear, anger and catastrophizing, reflecting emotional distress. However, he stated that

pain alone does not impact on an individual's ability to engage in valued activities (such as sport), which improve psychological well-being and QoL. There is also evidence among adults with osteoarthritis that sport has a positive impact on hip and knee pain as well as increasing mobility and well-being [32]. PWH in our cohort doing sports more than 4 h week^{-1} had better physical performance which concurs with the findings of a study where PWH underwent regular aqua gymnastics resulting in better endurance [33]. Moreover, sporting activity has been shown to enhance masculinity [34]. Previous studies have shown that not playing sport or being part of a team has a negative impact on masculinity in men with haemophilia [35]. Many men wish to participate in sport regardless of the known risks, especially in adulthood when they wish to play with their own children [22].

Initially, we had planned to enrol 200 patients in the adult arm of this study (100 per adult treatment centre); this proved to be impossible due to recruitment difficulties. Therefore, our study investigated only a small sample size of adults with mild (n = 12), moderate (n = 10) and severe (n = 28) haemophilia where milder disease is less likely to limit sporting activity, although in our cohort less mild/moderate patients were doing sport (45.5%) compared with severe patients (78.6%). This could reflect a desire to overcompensate on the part of patients with severe haemophilia. Predictably, patients with severe disease had significantly more target joints and more chronic pain. This, however, did not seem to limit sport participation, which can be seen as a tribute to human persistence in the face of adversity among our patients. Another limitation is that the data are only from four HTCs in the UK and cannot be attributed to all adults with haemophilia across the UK/Europe. The men in this study are treated at haemophilia centres where sporting activity is encouraged; following targeted prophylaxis and where specialist trained physiotherapy assessment is part of routine haemophilia follow-up. This may have biased the results that we report. However, it is encouraging to see a cohort of men across the adult age range who perceives sport to be a part of normal daily life, and not something to fear due to the risk of trauma-induced bleeding.

We have shown the health-related benefits of sport in adults with haemophilia in this article. Decisions about commencing sports programmes for others adults with haemophilia who may have target joints, limited range of movement and muscle strength should be made in conjunction with the patient and haemophilia healthcare providers [36,37]. This should include orthopaedic examination, fitness assessment and motion analysis [38,39]. Treatment regimens should be individualized to support sporting activity [9,23] while minimizing the risk of inducing further joint damage. The recommended level of physical activity for adults in the UK is defined as moderate intensity sport and active recreation averaging five or more times per week. People who have a physically active lifestyle are predicted to be at approximately half the risk of developing coronary heart disease compared with those who have a more sedentary lifestyle [40]. Physical activity is associated with increased functional ability and mental health as well as reduced incidence of diabetes, obesity, osteoporosis and colonic cancer [41]. Only 37% of men in the 2006 UK national survey were reported to achieve the recommended level of sporting activity [42]. Therefore, it is important to encourage PWH to be more physically active.

Increasing sporting activity may help to reduce BMI. Obesity is one of the major factors to the global burden of chronic disease and disability [4]. The WHO states that the obesity rates have increased at least threefold since 1980 in the United Kingdom [43]. In our study, we found a similar prevalence of overweight/obese men (64%) to that reported in a large American haemophilia cohort study (63%) [4,6]. Adams *et al.* [44] report that higher BMI is related to lower self-esteem and that patients with higher BMI are likely to watch more hours of TV. Furthermore, they suggest that health care professionals should address this through patient education encouraging less TV use and more active lifestyles for patients, which may impact also on HRQoL.

Haemophilia health care professionals recognize the physical and psychological benefits of sport for adults with haemophilia [45]. There are currently no internationally recognized sport recommendations for either children or adults with haemophilia. Indeed, encouraging participation in different types of physical activity and sport encourages boys and men to play sport that they are able to do well as well as those that they enjoy [46]. Negrier *et al.* [23] proposed that randomized clinical trials to promote evidence-based practice should be undertaken to assess the risks and benefits of sport in patients with all severities of haemophilia, we would also support this.

Conclusion

In this study of men treated at haemophilia centres where sporting activity is encouraged, with targeted prophylaxis to limit sporting injury related bleeding, we have described how sporting participation may improve patients' health status, overall HRQoL and physical performance. Sporting activity should be recorded and discussed as part of the haemophilia clinical review, including advice on joint protection/health, combined into HRQoL assessment. Sport participation did not result in bleeding in this cohort, further studies of the risks and benefits of sport for adult men with haemophilia are warranted.

Author contributions

SvM and KK designed the study and wrote the article. CH and ET gave input on the study design. KK coordinated the centres in the UK. KK, CH, AL and MF recruited patients and collated data at their HCTC. SvM analyzed the data. All authors contributed to the article and its revision.

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