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ORIGINAL ARTICLE

The impact of sport on children with haemophilia

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Summary. Sport is nowadays perceived as beneficial for children with haemophilia, as good muscle strength supports joints and may reduce bleed frequency; by contrast psychological benefits are less known. This study introduces the impact of sport on health-related quality of life (HRQoL) and physical performance in children with haemophilia. A cross-sectional, multi-site, study of boys aged 6–17 years with haemophilia A or B of any severity, current or past inhibitor, which assessed physical performance, sporting activity and HRQoL using age appropriate questionnaires including KINDL, Haemo-QoL and HEP-Test-Q. Eighty-four haemophilic boys (23 mild, 19 moderate, 42 severe) with a mean age of 11.52 years (SD = 3.4) were enrolled from two haemophilia centres in the United Kingdom. 28.4% were overweight/obese according to their BMI/age and had a good orthopaedic status (M = 1.55, SD = 3.3). Boys watching < 1–2 h of TV/PC/day had fewer days lost (M = 3, SD = 3.2) than those with a more sedentary lifestyle (M = 9.40, SD = 7.1) ($P < 0.032$). 90.5% participated in regular sporting activity; 79.9% at least twice a week. HRQoL in children was generally good, with highest impairments in boys aged 8–12 years. Boys aged 8–16 years reported good physical performance (M = 80.0, SD = 16.0) with highest impairments in the dimensions 'endurance' and 'mobility'. Boys doing sport had a significant better physical performance and HRQoL than boys not doing sport. Sedentary life styles had a negative impact on the subjective physical performance and number of days lost of children. Encouraging haemophilic boys to participate in sport will have a direct impact on their overall HRQoL.

Keywords: children and adolescents, haemophilia, health-related quality of life, physical performance, 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]. It affects boys, causing painful, spontaneous or trauma related bleeding predominantly in the weight bearing joints. Patients were sub-categorized in the 1950's by Biggs and Macfarlane [2] as severe (with factor levels of <1 iu dL^{-1}) moderate (levels 2–5 iu dL^{-1}) and mild haemophilia (5–50 iu dL^{-1}) according to factor level (normal range 50–150 iu dL^{-1}). Bleeding mostly correlates with factor level, thus those with severe haemophilia are most clinically affected. Children with haemophilia experience spontaneous or trauma related bleeding from an early age, without treatment early arthritic joint damage occurs [3].

Until the mid 1970's it was usual practice to discourage sporting activity in those with haemophilia because of the bleeding risk [4]. Today this attitude is more flexible, with many clinicians believing that sport is in fact beneficial for physical [5], social [6] and psychological [7] well-being. In part this is due to better treatment with prophylactic therapy being given to those patients with clinically severe haemophilia to prevent bleeds, minimize disability and improve quality of life (QoL) [8]. Many boys are now active sportsmen, participating at local, national and international levels, and have intensive prophylactic treatment tailored around their individualized sporting activity [9]. This enables them to participate fully in any activities they choose. The changes in treatment and resulting physical benefits have been shown to improve health-related quality of life (HRQoL) in these children [10]. There is, however, little data to support the non-physical benefits of sport in these

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boys. Therefore, a multi-centre 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 18–65), and parents of children. The data from children are presented in this article.

One hundred and twenty children with haemophilia of any severity or type, with or without inhibitors aged 6–17 years and their parents from two centres in the UK were invited to participate in the study. The children were divided into three age groups: [4–7 years (group I), 8–12 years (group II), 13–16 years (group III)] and were requested to complete age appropriate questionnaires which studied the impact of sport on their lives. The questionnaires were designed specifically for the study using validated questionnaires to collect data on HRQoL and physical performance. In addition, questions concerning sporting activities (e.g. frequency of sport per week and number of hours spent participating in sport) and attitudes towards sports were assessed using specially developed questionnaires, which were completed following parental consent and child assent at routine haemophilia appointments. 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 physiotherapist at each participating centre. Ethical approval for the study was granted by a local research ethics committee.

Instruments

The HRQoL was assessed using respective age-group versions [4–7 (group I), 8–12 (group II), 13–16 (group III) years] of the generic KINDL [11] and the haemophilia-specific Haemo-QoL instruments [12]. The KINDL questionnaire assesses self-reported HRQoL in six domains (physical function, psychological well-being, self-esteem, family, friends, school) and has an additional chronic-generic module with high values (range 0–100) indicating a good HRQoL. The disease-specific Haemo-QoL assesses self-report HRQoL of children with haemophilia. It consists of 8–12 dimensions dependent upon age (8, 10 and 12 dimensions, respectively) of HRQoL (physical health, feelings, attitudes, family, friends, perceived support, other persons, sports & school, dealing with the disease, treatment, future, relationships) with high values (range 0–100) indicating high impairments in HRQoL.

Physical performance was assessed by a patient-rated outcome using the HEP-Test-Q [13] and by an objective measure assessed by clinicians using the paediatric Petrini Haemophilia Joint Score [14]. The HEP-Test-Q was originally developed for adults with haemophilia and assesses four dimensions (mobility, strength & coordination, endurance and body perception) with high values (range 0–100) indicating better physical performance. In this study a child-adapted version of the HEP-Test-Q was included for children aged over 7 years, which varied only concerning the wording of some items compared with the adult version. The Petrini Joint Score ranks six joints (the elbow, ankle and knee, right and left) on swelling, muscle atrophy, axial alignment, crepitus on motion, flexion and extension loss, instability, joint pain, gait and strength. Scores of 0 indicate no joint problems; a maximum score of 156 would indicate severe joint damage and immobility.

Statistical analysis

All statistical analyses were conducted using the SPSS program version 17 (SPSS Inc. Chicago, IL, USA). Descriptive data are shown as frequency distribution in percent or as mean ± standard division SD (range), median and interquartile ranges (IQR) and were tested for normal distribution using the Kolmogorov-Smirnov 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.

To investigate the impact of sport on children's well-being we considered the following variables: doing sport (yes vs. no), sedentary life style [<12 h/day in front of television (TV) or computer vs. ≥ 2 h/day, frequency of doing sport (2 times/week vs. ≥ 3 times/week) and hours of sport (<5 h/week vs. <5 h/week)]. To determine the cut-off point for these variables the median split was calculated.

Results

Clinical data

From the 120 children invited to participate 84, with a mean age of 11.52 years (SD = 3.4, range 5.83–17.86), were enrolled into the study (70%). 92.3% had haemophilia A, half were severely affected, 22.6% had moderate and 27.4% had mild haemophilia. Two-thirds of the boys had regular prophylaxis; 16.9% reported targeted prophylaxis before sport. Nine boys had target joints and 28.8% were overweight or obese according to their BMI and age (see Table 1).

Overall bleeds were reported at a median of 0 (range 0–18, IQR = 2) in the 6 months preceding questionnaire completion, of these a median of 1 (range 0–10, IQR = 3) where joint bleeds and 0 bleeds in median (range 0–6, IQR = 0) were attributed to

Table 1. Clinical data of children with haemophilia (n = 84).

Clinical data	N	Percentage (%)
Type of haemophilia: A	77	91.7
B	7	8.3
Severity: severe	42	50.0
moderate	19	22.6
mild	23	27.4
Inhibitor: past or current	18	21.4
still present	5	27.8
Type of treatment: on demand	28	33.3
prophylaxis	56	66.7
Prophylaxis prior to sports:	14	16.9
Home treatment:	64	76.2
Presence of target joints:	9	10.8
Blood-borne infections:	0	0
Presence of chronic pain (M = 6.2, range 1–10)	22	27.5
BMI: under weight	27	33.8
normal weight	30	37.5
overweight	12	15.0
obesity	11	13.8

sport. A visual analogue scale was used to record chronic pain, (ranging from 0–10, where a score of 10 indicates maximal pain), 22 boys (26.2%) reported chronic pain in the preceding 6 months with a mean pain score of 6.19 (SD = 2.2 range 1–10). The Petrini score attained a median of 0 (range 0–15, IQR = 1), thus the boys in this study had evidence of good joint function. Nineteen boys missed days at school, 18 due to haemophilia: with a mean of 6.56 (SD = 6.5, range 1–26) days lost in the preceding 6 months. Three missed a mean of 3 days at school due to sporting injury (SD = 2.6, range 1–6). Clinical data across all three age groups are described in Table 2.

No differences in clinical data in terms of orthopaedic status, BMI, number of days lost from school and number of bleeds were found for sedentary lifestyle, doing sport or frequency/hours of sport. The only significant difference was found for number of days lost ($P < 0.032$) for boys watching <1–2 h of TV/computer games per day who had fewer days lost (M = 3, SD = 3.2) than those with a more sedentary lifestyle (M = 9.40, SD = 7.1) and for those boys doing sports more than 5 h week^{-1} (M = 4.3, SD = 3.9) compared with those doing sports less than 5 h/week (M = 11.5, SD = 8.2) ($P < 0.032$).

Sporting activity

Of the 84 participating boys only eight reported not doing any sport. The reasons for this were: that they weren't allowed to do it (n = 5), that they did not like it (n = 4) or that they were afraid of hurting themselves (n = 4) (more than one reason was given by some boys).

Seventy-six boys (90.5%) did sport. They reported participation in an average of four sports each, with the majority of boys doing sport twice weekly (see Fig. 1) They mainly performed sport with friends (80%) and at school (80%), although 40% of children reported participation in team sports at a sports club. In total, boys were doing an average of 4.9 hours sporting activities per week (range 1–13 h). Just over half of the boys (59.2%) participated in sport for 2–5 h/week with 35.5% stating they did 6–9 h, with 2.6% doing as much as 10–13 h/week (see Table 3) An extensive array of sporting activity was reported with the top five sports being: football (77.4%), jogging (76.2%), swimming (59.5%), gymnastics (36.9%) and cycling (25%).

Seventy-seven boys (95.17%) thought that doing sport was good, with arguments that it: 'is healthy and keeps you fit' (n = 36), 'is fun' (n = 21), 'makes you active' (n = 6) and 'is social' (n = 2). Other arguments in favour of sport were that it 'got you outside', 'gives you confidence' and 'is good when eating junk food'. Seventy-one boys (93.4%) reported that they would like to continue sports when they are older. Of these boys, 57 would like to try new sports such as rugby (n = 4), boxing (n = 3), cricket (n = 3), golf (n = 3) and hockey (n = 2).

Eight boys (9.5%) thought that sport was bad or dangerous: four boys considered specific sports such

Table 2. Clinical data according to age groups.

Clinical data	6–7 years (n = 15) M ± SD Median [IQR]	8–12 years (n = 41) M ± SD Median [IQR]	13–17 years (n = 28) M ± SD Median [IQR]	∑ (n = 84) M ± SD Median [IQR]
BMI	17.74 ± 2.4 17.72 [4.1322]	19.04 ± 4.7 17.36 [3.8075]	22.95 ± 4.4 21.72 [5.3375]	20.11 ± 4.7 18.92 [5.7869]
Orthopaedic status (Petrini Score)	0 ± 0 0 [0]	1.32 ± 2.8 0 [1.5]	2.7 ± 4.3 0 [5]	1.53 ± 3.2 0 [1]
No of total bleeds in the past 6 months	2.13 ± 4.5 1 [2]	1.88 ± 4.0 0 [2.5]	1.57 ± 2.5 0.5 [2]	1.82 ± 3.6 0 [2]
No of joint bleeds in the past 6 months	1.60 ± 3.1 0 [2.25]	2.26 ± 3.2 1 [3]	1.84 ± 2.8 1 [3]	1.96 ± 3.0 1 [3]
No of sports-related bleeds in the past 6 months	0 0 [0]	0.72 ± 1.6 0 [0.25]	0.41 ± 0.8 0 [0.5]	0.45 ± 1.2 0 [0]
Number of days lost in the past 6 months:				
haemophilia-related	11.67 ± 12.4 5 [22]	6.22 ± 5.2 6 [9]	4.50 ± 3.9 3.5 [5]	6.56 ± 6.5 4.5 [9.25]
sport related	0 – [–]	3.5 ± 3.5 3.5 [5]	2.00 ± 0 2 [0]	3.0 ± 2.6 2 [5]

IQR, interquartile range.

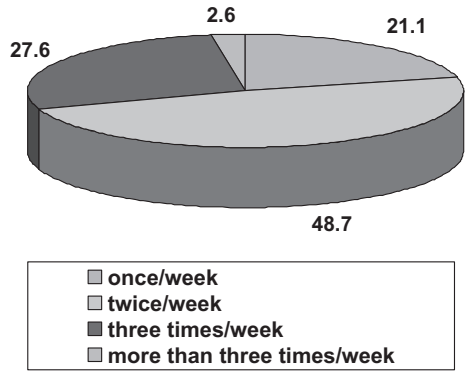

Fig. 1. Frequency of reported sporting activity.

Table 3. Hours of sporting activity across age groups.

Hours	Age 6–7 *N* (%)	Age 8–12 *N* (%)	Age 13–17 *N* (%)	∑ *N* (%)
1 hour	0	2 (5.3%)	0	2 (2.6%)
2–5 h	12 (85.7%)	21 (55.3%)	12 (50%)	45 (59.2%)
6–9 h	2 (14.3%)	13 (34.2%)	12 (50%)	27 (35.5%)
10–13 h	0	2 (5.3%)	0	2 (2.6)
∑	14 (100%)	38 (100%)	24 (100%)	76 (100%)

as boxing, rugby and hockey as dangerous and stated they considered getting hurt ($n = 4$) or falling ($n = 2$) too risky.

Health-related quality of life (HRQoL)

In the generic KINDL questionnaire children reported a quite good overall HRQoL in the total score, being highest in the youngest age group (I: M = 77.61, SD = 14.2; II: M = 70.40, SD = 8.9; III: M = 70.38, SD = 12.3), while for the chronic-generic module best values were reported by the oldest age group (I: M = 82.67, SD = 17.6; II: M = 82.98, SD = 20.9; III: M = 86.88, SD = 11.8). Haemophilic children reported the highest impairments in the dimension 'school' (see Table 4).

The haemophilia-specific HRQoL in children was generally good. Children in age group II (8–12 years) reported the highest overall impairment in the Haemo-QoL (M = 41.43, SD = 10.5). The youngest boys (6–7 years) reported highest impairment in the domains 'sport and school' (M = 44.44, SD = 25.7) followed by the dimensions 'treatment' (M = 32.14, SD 37.2) and 'other' (M = 32.14, SD = 30.1). In contrast, boys in age group II reported highest impairments in the domains 'dealing' (M = 75.74, SD = 16.3) followed by 'treatment' (M = 68.47, SD = 8.0) and 'friends' (M = 64.3, SD = 27.0). Adolescents reported highest impairment in the domains 'perceived support' (M = 50.8, SD = 27.5) 'friends' (M = 44.75, SD = 24.3) and 'future' (M = 33.11, SD = 20.5) (see Table 4).

Sport did not prove to have an impact on the HRQoL of children in age group I. By contrast there was a significant difference in HRQoL between children in age groups II and III grouped together doing sport and those not doing sport. Boys who did not do sport were more impaired in the dimension 'feeling' ($P < 0.014$) and 'family' ($P < 0.13$) than those doing sport (see Fig. 2). Children practising sport ≥ 3 times per week (M = 22.34, SD = 15.5) reported a better 'view of themselves' ($P < 0.017$) than children doing sport twice a week or less (M = 33.83, SD = 18.5). Children doing <5 hours sport per week (M = 44.53,

Table 4 HRQoL scores across age groups (KINDL, Haemo-QoL).

HRQoL questionnaire	Domains	Age 6–7 Mean (SD)	Age 8–12 Mean (SD)	Age 13–17 Mean (SD)
KINDL	Physical	75.00 (21.1)	74.41 (19.0)	74.31 (16.8)
	Emotional	76.67 (24.0)	79.49 (12.8)	77.31 (15.7)
	Self-Esteem	75.00 (23.1)	59.46 (17.1)	63.43 (21.9)
	Family	81.67 (24.0)	73.96 (18.6)	72.99 (18.1)
	Friend	81.67 (24.0)	79.55 (16.9)	75.45 (18.7)
	School	75.00 (31.0)	55.01 (17.2)	57.93 (20.4)
	KINDL TOTAL	77.61(14.2)	70.40 (8.9)	70.38 (12.3)
	Chronic-Generic	82.67 (17.6)	82.98 (20.9)	86.88 (11.8)
Haemo-QoL	Physical Health	22.12 (26.1)	28.39 (26.6)	22.69 (20.0)
	Feeling	14.44 (17.7)	18.38 (24.7)	13.17 (15.1)
	View	14.29 (25.4)	36.1 (15.1)	23.93 (20.1)
	Family	31.39 (32.9)	21.08 (19.4)	25.65 (26.2)
	Friends	23.33 (32.0)	64.3 (27)	44.75 (24.3)
	Support	–	57.94 (27.7)	50.80 (27.5)
	Other	32.14 (30.1)	12.39 (18.8)	17.11 (20.5)
	Sport	44.44 (25.7)	46.48 (10.3)	25.72 (20.8)
	Dealing	–	75.74 (16.3)	19.36 (15.2)
	Treatment	32.14 (37.2)	68.47 (8.0)	21.30 (14.5)
	Future	–	–	33.11 (20.5)
	Relationships	–	–	9.26 (16.5)
	Haemo-QoL TOTAL	28.09 (14.8)	41.43 (10.5)	24.44 (12.7)

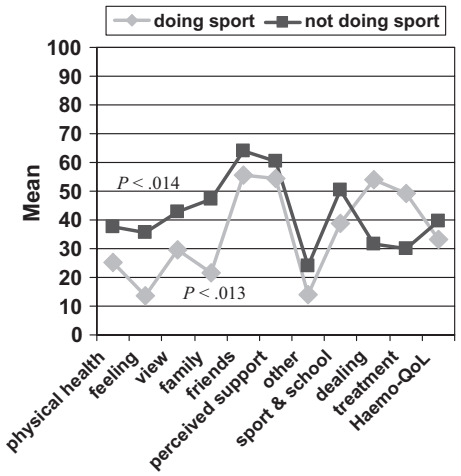

Fig. 2. Comparison of HRQoL (Haemo-QoL) across boys doing sport and those not doing sport.

SD = 23.8) felt less impaired ($P < 0.021$) in their 'perceived support' than children doing ≥ 5 h sport per week (M = 61.58, SD = 28.8).

Physical performance

Psychometric testing of the child-adapted version revealed excellent values in terms of reliability ranging from Cronbach's $\alpha = 0.855$ for 'endurance' to $\alpha = 0.936$ for 'the HEP-Test-Q total score'. The child-adapted HEP-Test-Q version showed as well good values for discriminant validity demonstrating that children who suffered from chronic pain reported in almost all subscales a significant lower subjective physical performance than those without pain.

The 67 boys in groups II and III (aged 8–17 years) completed the HEP-Test-Q. In general they reported good physical performance (M = 80.0, SD = 16.0) with highest impairments in the dimensions 'endurance' (M = 73.70, SD = 19.1) and 'mobility' (M = 75.29, SD = 24.0). There were no differences between age groups II (8–12 years) and III (13–17 years).

There was significant difference between children doing and not doing sport in all domains of physical performance (see Table 5). This demonstrates that children who do not do sport perceive their physical performance worse than those who do. A difference was also seen for sedentary lifestyles of children who watched television or played video games for more than 1–2 h on a weekday, who reported more impairment in 'endurance' ($P < 0.001$), 'body perception' ($P < 0.024$) and 'total physical performance ($P < 0.004$) than children who spent less time in front of the TV/computer (see Fig. 3).

Boys doing sport ≥ 3 times per week (M = 93.43, SD = 7.9) had significantly better 'co-ordination' ($P < 0.009$) than children doing less frequent sport

Table 5. Physical performance in 67 children doing sport and those not doing sport (HEP-Test-Q).

HEP-TEST-Q	Children not doing sport ($n = 5$) Mean (SD)	Children doing sport ($n = 62$) Mean (SD)	*P*-value
Mobility	50.00 (22.1)	77.40 (23.1)	0.013
Endurance	41.07 (23.7)	76.37 (16.2)	0.000
Co-ordination	52.63 (9.3)	88.34 (13.4)	0.000
Body perception	56.00 (16.4)	86.80 (18.6)	0.001
Total	48.58 (7.1)	82.52 (13.7)	0.000

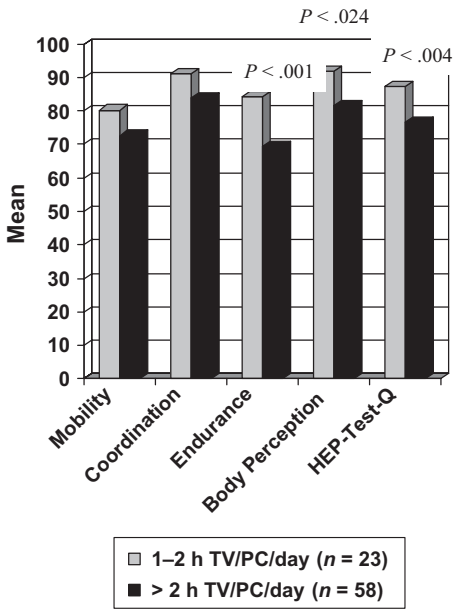

Fig. 3. Physical performance (HEP-Test-Q) based on hours of TV/computer games.

(M = 85.55, SD = 15.0). Those boys who reported doing ≥ 5 h sport per week (M = 81.52, SD = 13.0) reported significantly better 'endurance' ($P < 0.008$) than children practicing less sport (M = 70.69, SD = 17.6).

Discussion

Almost all boys in this study did sport, of which 80% played sport at school and 40% were part of a sports club. It is recognized that this level of sport in young boys will encourage sporting activity in adulthood where additional benefits such as improved personal physical health and individuals being able to play with their own children/grandchildren are cited as reasons to continue sport [15].

The impact of sport on children's health status and well-being was based on; doing sport, sedentary life style, frequency of doing sport and hours of sport. The only impact of sport on the health status was found for the number of days lost from school, where

boys with a more sedentary lifestyle had significantly more days lost from school. Differences in HRQoL were found in some domains of the Haemo-QoL ('feeling', 'family', 'view') for doing sport and frequency of doing sport per week. By contrast, significant differences concerning the subjective physical performance were found in all domains of the HEP-Test-Q between boys doing vs. boys not doing sport; significant differences in their physical performance were found for other aspects such as sedentary life style, frequency of doing sport and hours of sport.

The boys in this study are treated at haemophilia centres where sporting activity is encouraged, where prophylaxis is targeted around sport and where physiotherapy reviews are part of routine haemophilia care. This may have biased the results that we report. Nevertheless, it is encouraging to see a cohort of children who perceive sport to be a part of normal daily life, similar to their non-haemophilic peers, and not something to be scared of because of the risk of trauma induced bleeding.

Initially, we had planned to enrol 400 patients in this study (100 per centre), but this was not possible due to the lack of interest of patients in participating in this observational study and completing the very complex questionnaire. Therefore, our study investigated only a small sample size of children with mild, moderate and severe haemophilia where milder disease is less likely to limit sporting activity. Another limitation is that the data are only from two children's comprehensive care centres and cannot be attributed to all children in the UK/Europe.

According to international recommendations children should participate in moderate-to-vigorous physical activity (MVPA) at least 60 min daily [16,17]. In our study children were asked for how long they were doing sport every time they practised a respective sport; answer categories were '1 h', '2 h' or 'more than 2 h'. To calculate the average time they were doing sport in total we added the hours for each sport they were practicing resulting in 4.9 h/week in average (range 1–13 h). This is higher than might be expected; in the international HBSC REPORT [18] in the UK only 18% of boys aged 15 years (23% of boys aged 13 years, 27% of boys aged 11 years) were doing at least 1 h of MVPA daily [19]. This is probably due to the fact that we did not ask in an open-ended question the exact time for each sport. Even though this is a methodological error related to the answer categories provided in the questionnaire (we assume that children doing a specific sport less than 1 h ticked the answer category '1 h', which results in a higher total amount of hours doing sport), this is a systematic error for all participants which did not lead to incorrect results, but this result should not be compared with other surveys, where an exact time of MVPA is assessed.

Health-related quality of life in children with haemophilia is not only influenced by disease severity and treatment but also by the impact that these have on the ability to participate in routine childhood activities in day-to-day life. For boys, perhaps more than girls, there is a social pressure to conform to peers by participating in sporting activity. This has been shown to enhance masculine identity and is increasingly important in adolescence [20]. Past studies have demonstrated how not being able to play sport or be part of a team, has had a negative impact on masculinity in men with haemophilia [21]. These men described the wish to participate in sport vs. the risk of joint damage as a 'mental battle that you have to overcome as a child' and that this continues into adulthood when they can not do sport with their own children [22].

Young British children have been shown to find exercise enjoyable when it is not only competitive but something that encourages experimentation with different activities that are beneficial to them [15]. Parents play an important role in enabling their children to play sport safely [23] this may be part of an overall health and fitness campaign. Recent British Government recommendations around weight and activity state that 70% of the population should do 30 minutes moderate exercise, five times per week [24]. Only 37% of men in the UK were achieving this benchmark in 2006; one reason for this is negative participation experience at school [20]. We have shown that many of the boys in our study already exceed this target.

In this study 95.17% of boys thought that doing sport was good and 90.5% participated in leisure sports, mainly in football, jogging, swimming, gymnastics and cycling; this is comparable to a German study, where 44 children with any form of haemophilia (aged 4–16 years) were investigated. In the German cohort 88.6% of the adolescents actually performed one or more leisure sports, mainly swimming, tennis and football and 79% considered exercise in daily routine to be important or very important [25].

In the current study no differences were found in clinical data for children doing sport vs. those not doing sport. This is in line with other studies conducted in the US with boys with severe haemophilia, where level of athletic participation was not a significant prognostic factor for joint haemorrhage [26], or in the Netherlands, where no relationship between exposure to risk during sports participation and physical outcome measures was found [27], or in Israel, where level of physical activity in 44 young patients (aged 12–25 years) with severe haemophilia, assessed by the Godin and Shephard questionnaire, did not show a difference in bleeding frequency and pain [28].

Only few data on the impact of sport on HRQoL in children with haemophilia are available [29,30]. In the Australian cohort of haemophilic boys, no correlation was found between quality of life and fitness. In

general their HRQoL was high (measured via the Haemo-QoL) and comparable to that of haemophilic boys from Europe [29]. In a Dutch study [27] no differences for risk exposure factor and HRQoL measured with the Haemo-QoL Index [31] were found.

In our study we found a higher prevalence of heavy and overweight boys (28.4%) compared with a Dutch study of 158 haemophilic boys, where only 16% were classified overweight [32]. In the Dutch cohort a negative association of being overweight and HRQoL evaluated via the Haemo-QoL Index was found. This was not corroborated in the present UK study nor in another Dutch study involving 13 severe haemophilic boys, which demonstrated that there was no significant relationship between the child report of HRQoL, as assessed via the long-form of the Haemo-QoL and physical fitness, as evaluated via both absolute and relative peak oxygen uptake [30].

Haemophilia treaters now recognize that sport is beneficial both physically and psychologically for children with haemophilia [33]. There are currently no internationally recognized sport recommendations for boys with haemophilia, only on national levels [34] and individual children participate in many sports depending on their own interests and abilities. Indeed encouraging participation in different types of physical activity and sport encourages boys to play sport that they are able to do well, as well as those that they enjoy [35].

Better HRQoL is demonstrated in those doing sport more than three times per week than those doing twice a week or less. Therefore, encouraging boys with haemophilia to participate in sport will have a direct impact on their overall HRQoL. Sporting activity should be recorded as part of the haemophilia clinical review and combined into HRQoL assessment. Decisions about the most suitable sport for individual children should be made by haemophilia clinicians, physiotherapists, patients and parents [36,37], including orthopaedic examination, fitness check and motion analysis [38,39]. Treatment regimens should be individualized to support sporting activity [9] whilst minimizing the risk of long-term joint damage [5].

Even though it is widely recognized that physical activity is important for boys with haemophilia as it promotes healthier joints and reduces the risk of bleeding, the selection of an appropriate sport that minimizes the risk of injury and matches the patient's skill and needs is important [38].

Conclusion

Boys participating in sport had a significant better physical performance and HRQoL than boys not doing sport. Sedentary life styles had a negative impact on the subjective physical performance and number of school days lost by children. Therefore encouraging boys with haemophilia to participate in sport will have a direct impact on their overall HRQoL. Sporting activity should be recorded as part of the haemophilia clinical review and combined into HRQoL assessment. Participation in sport did not increase the risk of bleeding or developing target joints. Interventional studies are needed assessing the impact of sports on boys with haemophilia.

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Author contributions

KK and SvM designed the study and wrote the paper. KK coordinated the centres in the UK. KK and AL recruited patients at their HCTC. SvM analysed the data. All authors contributed to the paper and its revision.

Disclosures

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