









ORIGINAL ARTICLE

Social participation and hemophilia: Self-perception, social support, and their influence on boys in Canada

Aubrey S. Chiu MSc¹   | Victor S. Blanchette MB BChir (Cantab), FRCP(C), FRCP^{1,2}  |
Maru Barrera PhD^{1,2}  | Pamela Hilliard BSc, PT¹  | Nancy L. Young MSc, PhD³  |
Audrey Abad HSc¹  | Brian M. Feldman MD, MSc, FRCPC^{1,2} 

¹The Hospital for Sick Children, Toronto, Ontario, Canada

²The University of Toronto, Toronto, Ontario, Canada

³CHEO Research Institute, Ottawa, Ontario, Canada

Correspondence

Brian M. Feldman, 555 University Avenue, Toronto, Ontario, M5G 1X8, Canada.
Email: brian.feldman@sickkids.ca

Funding information

No funds were received for the conduct of this study.

Handling Editor: Pantep Angchaisuksiri

Abstract

Background: Patients with hemophilia may experience joint damage, which can impair participation, yet few studies have examined the impact hemophilia may have on social participation and quality of life.

Objectives: The aims of this study are to assess the relationship between patient social participation and self-perception, social support, and impact on the family.

Patients/Methods: A random representative sample of 50 boys with hemophilia from The Hospital for Sick Children, Toronto, Canada, completed measures of social participation (Participation Scale for kids), self-perception (Self-Perception Profile for children and adolescents), and social support (Social Support Scale for children). Participants' parents completed Family Impact Module of the Pediatric Quality of Life Inventory. Data were analyzed using Pearson product-moment correlations.

Results: Social participation was strongly correlated with self-perception subscales Social Acceptance ($r = -0.5, p = <0.001$) and Global Self-Worth ($r = -0.6, p = <0.001$) for all participants. The Athletic Competence subscale was strongly correlated for adolescents only ($r = -0.6, p = <0.01$). There were strong correlations between social participation and social support from parents ($r = -0.6, p = <0.001$), teachers ($r = -0.5, p = <0.001$), and classmates ($r = -0.6, p = <0.001$) and moderate correlations for support from close friends ($r = -0.4, p = <0.01$). There were no significant correlations with family impact.

Conclusion: In the context of a country with unlimited access to safe clotting factor concentrates, boys with hemophilia have few social participation restrictions. Although correlational findings do not represent causality, they suggest that encouragement of social participation may be beneficial in boys with hemophilia to increase self-perception as well as strengthen their social support network.

KEYWORDS

child, Hemophilia, quality of life, self concept, social participation, social support

This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2021 The Authors. *Research and Practice in Thrombosis and Haemostasis* published by Wiley Periodicals LLC on behalf of International Society on Thrombosis and Haemostasis (ISTH).

Essentials

- Boys with hemophilia may experience joint damage, which may impact participation in activities.
- We explored how social participation, self-concept, social support and family impact are related.
- Boys reporting high social participation also reported high self-perception and social supports.
- In countries with unlimited access to treatment, few boys report participation restrictions.

1 | INTRODUCTION

Hemophilia is an inherited bleeding disorder caused by abnormalities in the genes for factor VIII (FVIII) or factor IX (FIX).¹ These gene defects result in deficiencies of FVIII (hemophilia A) or FIX (hemophilia B) leading to impaired clotting. Boys with severe hemophilia, defined by FVIII or FIX levels <1%, typically experience recurrent bleeding into muscles and joints from an early age, leading in some cases to clinically significant arthropathy. The effects of chronic pain and reduced mobility in patients with hemophilia may impact social participation, ranging from occasionally missing a social activity to complete isolation from an activity and/or the individual's peers.

The International Classification of Functioning, Disability, and Health (ICF) developed by the World Health Organization (WHO) provides a framework embracing the multiple components of health.² This framework states that a person's functioning and disability is the result of the dynamic interaction between the person's health conditions and contextual factors, both personal and environmental (Figure 1). The model is bidirectional in nature such that each component impacts and is impacted by other components. The WHO defines participation as "involvement in a life situation"²; however, there has been debate over the WHO definition and its measurement.³ For the purposes of this study, we have chosen to redefine participation as "social participation" to emphasize the social aspects of participation. It has been suggested that for children with chronic illnesses, social participation restrictions may be more important to them than their disease and that the inability to participate like one's peers could both lead to and result in stigma, which may further restrict social participation.³ The aforementioned effects of chronic pain and reduced mobility in patients with hemophilia may restrict participation in the activities that one's peers engage in.

Personal factors include "the particular background of an individual's life and living, and comprise features of the individual that are not part of a health condition or health states."² These may include self-esteem, self-perception, and perceived social support. The contribution of each of these factors and their dynamic interaction may determine the level and extent of an individual's functioning, including social participation.² For example, the ability of a boy with hemophilia to participate in a 1-km run for physical education class is determined by his health condition (hemophilia), body functions and structures (does he suffer from arthropathy?), activities (is he able to run?), environmental factors (is the running surface appropriate?), and personal factors (does he have confidence that he can run? Do his parents or teachers allow him to run?). The interplay between all

these factors reflects the multifaceted nature of participation and the importance of examining multiple components.

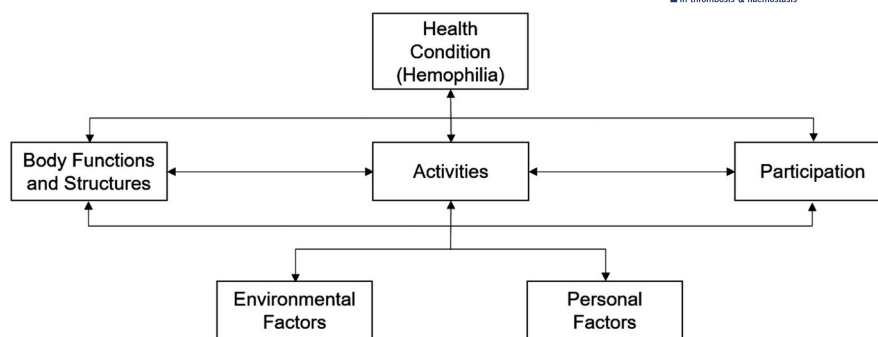
Self-perception may impact social participation and refers to one's perception of self, including feelings of competence and adequacy in specific developmental domains, overall self-esteem, and self-worth.⁴ For children with chronic diseases, social participation restrictions may have a significant impact on their well-being as the inability to participate could both lead to, and result in, stigma that could further restrict their social participation.³ Difficulties with mood changes and general psychological well-being are often reported for patients with hemophilia, including depressive symptoms, poor self-perception, and low self-esteem.^{5,6}

To optimally engage in life's activities, an individual must feel accepted by, and have the social support, of others. Social support can be broadly defined as the approval or positive regard of others and may come from family, classmates, health care professionals and others, which may help reduce social isolation.^{7,8} If opportunities to engage in social activities are restricted, or if the individual feels socially isolated, his or her self-perception and social participation may be negatively impacted. Previous research has shown that a lack of social support for adult hemophilia patients was significantly associated with patients reporting higher depressive symptoms.⁹

It is important for health care providers to recognize that chronic illness impacts the entire family, not just the patient.¹⁰ Chronic health conditions can affect multiple domains for parents and the family, including physical, emotional, social, and cognitive functioning, communication, and worry.¹¹ If family members perceive the disease to have a high impact on the family, they may be less inclined to encourage social participation. In a previous study, parents reported having difficulty when their children wished to play sports as they had to choose which sports were suitable and not suitable for the child to participate in.¹⁰ The social participation of the individual may consequently be affected by the family's attitudes, support, and encouragement to engage in certain activities.

To our knowledge, no studies to date have measured the association between self-perception, social support, impact on the family, and social participation in boys with hemophilia. The rationale for this study is based on the premise that understanding how patients interact socially with their peers may inform efforts to improve social participation (and, perhaps, vice versa). Our primary hypothesis proposes an association between self-perception and social participation in boys with hemophilia, which is bidirectional in nature, in keeping with the ICF model. Additional hypotheses explore if there are associations between social participation and social support, as well as impact on the family.

FIGURE 1 This framework states that a person's functioning and disability is the result of the dynamic interaction between the person's health conditions and the contextual factors in their life



2 | METHODS

2.1 | Design

A cross-sectional observational study was conducted at The Hospital for Sick Children ("SickKids") in Toronto, Canada.

2.2 | Sample

Eligible subjects were boys 7–18 years of age registered in the pediatric comprehensive care hemophilia program with a diagnosis of moderate (baseline factor level 1%–5%) or severe (baseline factor level <1%) hemophilia A or B (including patients with inhibitors). A random sample from the eligible pool of subjects was generated using an electronic randomization program, proportionately stratified for hemophilia type and severity based on baseline clinic proportions. Subjects had to have no history of an acute bleed within the last 2 weeks before assessment.

2.3 | Procedure

Research Ethics Board approval was obtained and the study was carried out in accordance with the Declaration of Helsinki. All participants and parents/guardians were required to give written informed consent or assent as applicable. Participants were recruited and participated in the study between June 2011 and October 2013.

2.4 | Assessment measures

Except for the Pediatric Quality of Life (PedsQL) Family Impact Module and demographic questions, all other measures were completed by the patient or administered by a study staff member, if needed. Questionnaires were completed in the outpatient clinic setting and patients were encouraged to complete them on their own whenever possible. If questionnaires were administered by a study staff member, questions were posed to the patient and answers were recorded accordingly. Study staff provided clarification on questions, if needed. Parents/guardians may have remained

present during the questionnaires. Participants took approximately 30–40 minutes to complete questionnaires.

2.5 | Social participation

Social participation was measured using a modified version of the Participation Scale, which measures the impact of nonparticipation in adults (Cronbach α 0.9; intertester reliability = 0.8).³ The Participation Scale accounts for nonparticipation in activities that are irrelevant or of no interest to the subject. Twenty-three scale items prompted subjects to self-report if they participated as much as their peers did in an activity. Items were scored 0 to 5, where higher scores indicated greater problems with social participation.

With permission from the developers of the adult Participation Scale, questions were modified to be relevant to children. For the purpose of this study, the scale is referred to as the Participation Scale for kids. The comparison to one's "peer" was changed to "classmate" for ease of understanding. Two questions relevant to North America were added (to explore if participants play games and use social networking sites as compared with their classmates). Grades of participation restriction were proportionately redefined as: no significant restriction (0–15), mild restriction (16–27), moderate restriction (28–40), severe restriction (41–66), and extreme restriction (67–115).

To test the reliability of the modifications made to the participation scale, internal consistency was tested using Cronbach coefficient alpha. Both the overall raw alpha ($\alpha = 0.7$) and the standardized alpha ($\alpha = 0.7$) met the minimum alpha recommended for research purposes.¹²

2.6 | Self-Perception

Self-perception was measured using the Self-Perception Profile for children (ages 7–12) or the Self-Perception Profile for adolescents (ages 13–18).^{4,13} According to the scales' author, as people age they evaluate themselves across an increasing number of domains and the content of those domains will change.¹³ These scales have been used extensively, including in subjects with hemophilia,^{6,14} and have demonstrated strong internal consistency (Cronbach $\alpha \geq 0.8$) when

administered to North American children as well as face, factorial, convergent, and construct validity.^{4,13}

Three domains were selected for study inclusion to reduce participant burden and to align with subscales available in the Self-Perception Profile for Children: Social Acceptance (extent he felt accepted by peers or felt popular), Athletic Competence (competence in sports and outdoor games), and Global Self-Worth (degree to which he liked himself as a person).⁷ These domains were selected as they had the most face validity suggesting a relationship with social participation.

2.7 | Social support

The Social Support Scale for Children is a self-report tool that measures perceived support and positive regard from significant others. The scale has strong internal consistency (Cronbach $\alpha = 0.7$ – 0.9) and demonstrated validity for each of the subscales.^{7,15}

Four sources are measured: parents (parents understood or cared about him), teachers (there was a teacher who helped him achieve his best and cared about him), classmates (his classmates liked him the way he was or asked him to join in play), and close friends (there were close friends who understood him or who listened to him).¹⁵

The items on the Self Perception Profiles for children/adolescents and the Social Support Scale for children are scored 1–4, where higher scores indicate higher self-perception, more perceived social support, or positive regard.

2.8 | Impact on the family

The impact on family members was assessed using the 36-item PedsQL Family Impact Module. The eight subscales measure Physical Functioning, Emotional Functioning, Social Functioning, Cognitive Functioning, Problems with Communication, Problems with Worry, Problems with Daily Activities, and Problems with Family Relationships.

Respondents answered items on a scale from 0 to 4. Items were subsequently reverse-scored and linearly transformed to a 0–100 scale, where higher scores indicated less negative impact on the family and better family functioning.¹¹

2.9 | Analysis

Double data entry of 100% of the data was performed to ensure accuracy. Single imputation was used to account for missing data and sensitivity analysis of worst- and best-case scenarios (10th and 90th percentiles, respectively) were performed for missing data.

Pearson product-moment correlation coefficients were calculated to assess the linear relationships between variables. The

bidirectional nature of the ICF model influenced our decision to use correlations for the analysis to not imply causality between any of the variables. Hypotheses were specified in advance; therefore, no post hoc corrections were made. The following guidelines for interpreting relationship strength were used (in absolute values): ≤ 0.1 represented small correlations, 0.3 were moderate correlations, and 0.5–1.0 were large correlations.¹⁶ Differences between children and adolescents were assessed using independent samples *t*-tests.¹⁷ SAS version 9.2 was used for statistical analysis.

3 | RESULTS

3.1 | Sample characteristics

Participant characteristics are displayed in Table 1. Of 61 patients assessed for eligibility, 57 (93.4%) were deemed eligible. Exclusions from the study were due to patients unable to independently answer questions ($n = 2$), psychosocial issues ($n = 1$), and family issues ($n = 1$). Fifty-one (89.5%) gave informed consent/assent. Fifty of 51 consented participants completed all assessments, which included 30 children (aged 7–12) and 20 adolescents (aged 13–18) from 7.7 to 17.0 years old (mean = 12.1, SD = 2.8).

Ethnicity was self-reported by Canadian national standard categories (Caucasian/White, Black, Asian, Native) and subcategories. Most participants indicated ethnicity as North American (Canada and United States), followed by South Asian, European, then Middle Eastern. One-fifth of the sample identified their ethnicity as Other, a category generally used when reporting two or more ethnicities (e.g., North American and European). Forty-nine (98.0%) participants attended school (one was home-schooled, 2.0%); school grades ranged from 2 to 12.

Descriptive statistics of the family situation were collected from participants' primary guardian. The majority (82%) reported married/common-law marital status (82%) and 74% reported both mother and father were primary guardian. More than one-half (53%) reported high household income (greater than CAD\$80,000), 35% reported medium (\$30,000–\$80,000), and 12% reported low (<\$30,000).

The sample was composed predominantly of participants with hemophilia A (82%) and the majority had severe hemophilia (70%). Standard of care for most boys with severe hemophilia at The Hospital for Sick Children is tailored primary prophylaxis; as such, more than two-thirds of the participants were on prophylaxis, defined as regular clotting factor infusions given at least once per week to prevent bleeding. Of the severe hemophilia patients, 29 (82.9%) patients were on prophylaxis. The majority (86.7%) of patients with moderate hemophilia receive treatment on demand. Six cases had a history of an inhibitor to FVIII; however, all subjects were responsive to infusions of FVIII at the time of study. Two subjects had three or more bleeds into an index joint (both knees) within the 6-month period before enrollment. Forty-nine subjects were receiving a

recombinant factor VIII concentrate at the time of study; one subject was receiving a plasma-derived FVIII concentrate.

Less than 5% of the data was missing for any one variable; as such, there were minimal changes witnessed in the analyses of worst- and best-case scenarios. There were no significant differences for severity of hemophilia; therefore, results are presented jointly. There were differences in the correlations for children and adolescents between social participation and both self-perception

and social support. However, when comparing the means of the children and adolescent groups, allowing for unequal variances, no significant differences were found for the subscales; therefore, results are presented jointly.

TABLE 1 Participant characteristics (N = 50)

Variable	n (%)
Type of hemophilia	
A	41 (82.0)
B	9 (18.0)
Phenotype	
Moderate	15 (30.0)
Severe	35 (70.0)
Ethnicity	
North American	16 (32.0)
Other	10 (20.0)
South Asian	6 (12.0)
European	5 (10.0)
Middle Eastern	3 (6.0)
African	2 (4.0)
Caribbean	2 (4.0)
Chinese	2 (4.0)
Korean	1 (2.0)
Mediterranean	1 (2.0)
South American	1 (2.0)
Vietnamese	1 (2.0)

3.2 | Social participation

When assessing children and adolescents, the mean score for restricted social participation was 6.5 (SD = 6.2) of a possible 115 (see Figure 2). Forty-six (92.0%) participants had no significant restriction, 3 (6.0%) had mild restriction, and one (2.0%) had moderate restriction. No participants indicated severe or extreme restriction.

When assessed separately, the mean score for children on social participation was 7.6 (SD = 7.2), whereas adolescents scored slightly lower at 4.9 (SD = 3.7). When comparing the means of the children and the adolescents, the differences were not statistically significant ($t(45.4) = 1.8, p = 0.1$).

3.3 | Self-perception

Overall, participants scored highly on self-perception subscales (Table 2). Although children reported slightly lower mean self-perception scores than adolescents, when comparing the means of the children and adolescents, no statistically significant differences were found; therefore, results for combined data are presented (Table 3). Given that the maximum subscale score was 4.0, the mean scores of 3.2 and above suggest that self-perception was not significantly impaired in this sample. When comparing scores from this study to comparable reference groups assessed by the

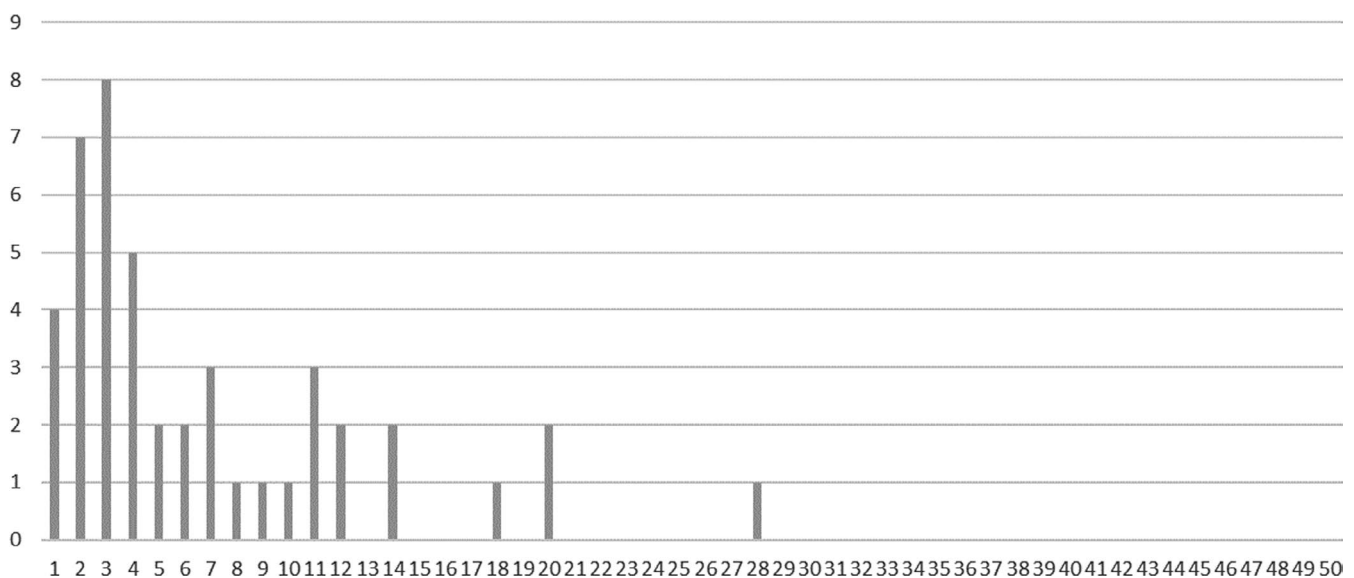


FIGURE 2 The distribution frequency of social participation as measured by the Participation Scale for kids, where higher scores indicated more restrictions with participation (N = 45)

Variable	M	SD	Min-Max	Pearson <i>r</i>
Social participation				
Overall	6.5	6.2	0–28	–
Children	7.6	7.2	0–28	–
Adolescents	4.9	3.7	0–12	–
Self-perception				
Social acceptance	3.3	0.7	1.2–4.0	–0.5*
Athletic competence	3.2	0.7	1.7–4.0	–0.2
Global self-worth	3.5	0.6	1.7–4.0	–0.6*
Social support				
Parental support/regard	3.6	0.5	1.8–4.0	–0.6*
Teacher support/regard	3.4	0.7	1.2–4.0	–0.5*
Classmate support/regard	3.5	0.6	1.3–4.0	–0.6*
Close friend support/regard	3.5	0.6	1.0–4.0	–0.4*
Impact on the family				
Total score	3074.0	571.1	1275.0–3600.0	–0.2
Physical functioning	91.4	14.0	37.5–100.0	–0.2
Emotional functioning	80.8	19.7	25.0–100.0	–0.2
Social functioning	89.8	18.9	25.0–100.0	–0.3*
Cognitive functioning	90.5	19.9	20.0–100.0	–0.2
Communication	80.8	20.2	25.0–100.0	–0.2
Worry	73.2	22.5	15.0–100.0	0.0
Daily activities	88.8	19.1	33.3–100.0	–0.3
Family relationships	87.9	18.1	20.0–100.0	–0.2

Note: Participation Scale for kids: higher scores indicated greater problems with social participation. Self Perception Profiles for children/adolescents: higher scores indicated higher self-perception. Social Support Scale for children: higher scores indicated more perceived social support or positive regard. Pediatric Quality of Life (PedsQL) Family Impact Module: higher scores indicated less negative impact on the family and better family functioning.

Abbreviations: M, mean; Max, maximum; Min, minimum; SD, standard deviation.

*Correlation is significant at $p < 0.05$.

authors of the scale (mean scores of 3.1 and above),⁴ the means of this sample were higher, suggesting no impaired self-perception for the sample.

3.3.1 | Correlations between self-perception and social participation

When looking at the relationships between social participation and the self-perception subscale scores, a strong correlation was found between social participation and Social Acceptance ($r = -0.5$, $p = 0.0003$) as well as Global Self-Worth ($r = -0.6$, $p = <0.0001$; see Figure 3). The negative correlation suggests that when participants had better self-perception regarding Social Acceptance and Global Self-Worth, they tended to report fewer social participation restrictions. The correlation between social participation and Athletic Competence was not statistically significant ($r = -0.2$, $p = 0.1$).

TABLE 2 Descriptive statistics and correlations with social participation (N = 50)

3.4 | Social support

Participants scored highly on social support subscales with all subscale mean scores above 3.3 (maximum, 4.0). When comparing the means of children and adolescents for social support, there were no significant differences between the two groups (Table 3); thus, combined data are presented (Table 2). Much like self-perception, when this sample's scores were compared with reference groups (mean scores 2.9 and above), the means of the sample were higher.¹⁵

3.4.1 | Correlations between social support and social participation

Strong correlations were found between social participation and parent ($r = -0.6$, $p = <0.0001$), teacher ($r = -0.5$, $p = <0.0001$), and

TABLE 3 Comparing the means of the self-perception and social support subscale scores between children and adolescents (N = 50)

Subscale	Children M (SD)	Adolescents M (SD)	df	t	p
Self-perception					
Social acceptance	3.2 (0.7)	3.5 (0.7)	40.7	-1.4	0.2
Athletic competence	3.2 (0.7)	3.2 (0.7)	38.3	-0.2	0.8
Global self-worth	3.5 (0.6)	3.6 (0.6)	42.0	-0.8	0.4
Social support					
Parental support/regard	3.6 (0.6)	3.7 (0.4)	47.7	-0.3	0.8
Teacher support/regard	3.4 (0.7)	3.3 (0.8)	38.9	0.7	0.5
Classmate support/regard	3.4 (0.6)	3.5 (0.6)	41.2	-0.6	0.6
Close friend support/regard	3.5 (0.5)	3.5 (0.7)	33.2	-0.1	1.0

Note: p is significant at <0.05.

Abbreviations: M, mean; SD, standard deviation; df, degrees of freedom; t, t value.

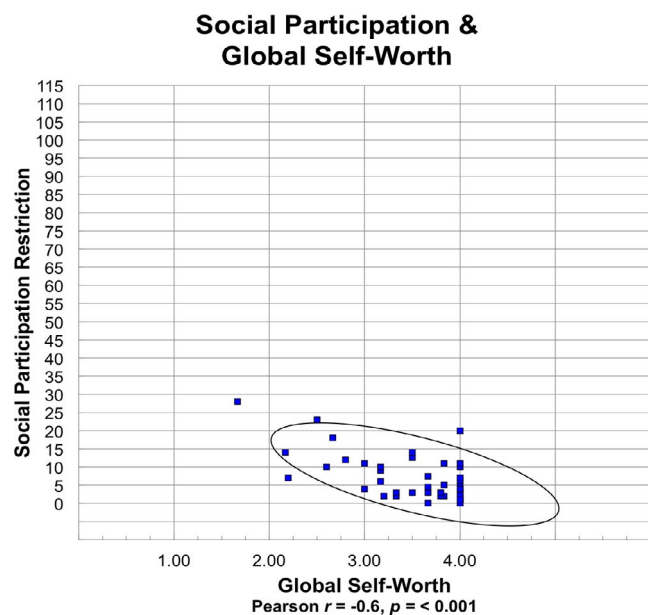


FIGURE 3 Social Participation and Global Self-Worth is illustrated using the correlation between social participation as measured by the Participation Scale for kids, where higher scores indicated more restrictions with participation, and Global Self-Worth as measured by the Self-Perception Profile for children or adolescents, where higher scores indicated more adequate self-perception. The confidence elliptical curve shows 95% prediction

classmate support/regard ($r = -0.6, p = <0.0001$). A moderate correlation was found between social participation and close friend support/regard ($r = -0.4, p = 0.002$).

3.5 | Impact on the Family

Scores on the PedsQL Family Impact Module were high (mean = 3074, SD = 571; maximum score of 3600) suggesting that a diagnosis of hemophilia had minimal negative impact on the family (see Table 2).

3.5.1 | Correlations between family impact and social participation

There was a moderate correlation between social participation and social functioning ($r = -0.3, p = <0.05$), which suggests that when parents/guardians reported better social functioning, their children tended to report better social participation.

All other subscales did not have statistically significant correlations.

4 | DISCUSSION

Boys with hemophilia with access to comprehensive care and unrestricted access to safe, effective clotting factor concentrates report having few social participation restrictions, high levels of perceived competence, and high levels of perceived social support. Positive relationships between social participation and self-perception as well as social participation and social support were observed. No significant relationship was found between social participation and impact on the family.

The ICF model suggests a dynamic interaction between hemophilia and the personal factors in one's life. Using this model, the results observed support the conclusion that self-perception and perceived social support are associated with social participation. Given the bidirectional nature of the model, social participation may also exert influence on these factors. The interplay observed between these variables determines the individual's functioning and disability, including social participation.

An important finding in this study was that most participants experienced few social participation restrictions and no participants reported severe participation restrictions. This may be explained in part because the majority of participants were on treatment programs of tailored primary prophylaxis from an early age in life that resulted in very low spontaneous rates of bleeding into joints and muscles.¹⁸ This finding is comparable to previous reports of patients on prophylaxis being less impaired than those using on-demand

treatment.¹⁹ Future studies on the influence of personal and environmental factors such as access to treatment, severity of illness, inhibitor presence, number of target joints, arthropathy, and socioeconomic status (including ethnicity) may glean additional insights.

Previous research is inconclusive about self-perception in boys with hemophilia. Some suggest that boys with hemophilia are more likely to report lower perceived competence than healthy peers,^{20,21} whereas others argue that self-esteem and self-perception are comparable to healthy peers.^{14,22} Consistent with research in the Netherlands,¹⁴ our findings support the notion that boys with hemophilia whose disease is well managed and are well-supported socially are no more susceptible to lower levels of self-perception than their healthy peers without hemophilia. Future studies may explore self-perception in patients with hemophilia with unrestricted access to long-term prophylaxis compared with children without chronic illness.

The results indicate moderate to strong correlations between social participation and self-perception, supporting our primary hypothesis. Accordingly, participants who reported better self-perception also reported fewer social participation restrictions. Social participation and athletic competence were not correlated when data from children and adolescents were analyzed together. There was a significant correlation when examining adolescents only, which may indicate that participation in sports and outdoor games are more significant for adolescents than younger boys with hemophilia.

Regarding the exploratory hypothesis, results from this study suggest that social support and social participation are associated. There was a moderate correlation between social participation and support from close friends, but a strong correlation between social participation and support from classmates. These results indicate that boys with hemophilia who endorsed better support/regard from their social network also had better social participation scores. Perhaps participants who perceive classmates to be supportive are more inclined to engage with them socially, and conversely, individuals who participate more may be more popular with their classmates and invited to engage in social activities more often.

Overall, the participants in this study felt they were well supported by their social network. There is no universal conclusion when it comes to self-perception of chronically ill children when compared with healthy peers. Possible reasons for the differences observed in our study include effective disease management (the majority of participants were receiving long-term prophylaxis introduced from an early age in life and enrolled subjects had to have no acute bleeds 2 weeks before assessment) and the age of participants (the study cohort included more young boys with hemophilia than adolescents who have perhaps not experienced negative social consequences from not participating in activities).²³ Some studies conducted in countries where patients have access to safe clotting factor concentrates have found that patients with hemophilia may report depressive symptoms and lower self-perception, but not necessarily at a clinically significant level.⁵ Other research has found

adolescents and adults with hemophilia to have difficulties with perceived support.^{9,19}

The importance of a social support network has far-reaching consequences. If a patient perceives that he has good social support, he may feel less depressed, have better compliance, and higher health-related quality of life.^{9,19,24} Furthermore, although many parents may worry about their child with hemophilia having activity-related bleeding, forbidding boys to engage in specific activities may lead to them concealing their involvement in at-risk activities.²⁵

The data mostly did not support the hypothesis that impact on the family and social participation were associated. In this sample, hemophilia had limited impact on the family with few parents/guardians reporting any difficulties. Possible reasons may be that this sample included older boys with well-established prophylaxis regimens who may have adapted to living with hemophilia. Finally, findings support previous research that found that parents of children with hemophilia report fewer psychosocial strains when compared with parents of children with other chronic illnesses.²⁶ Other studies found that parents reported that activities or relations were almost never disturbed because of extra care required for the patient; hemophilia was perceived as a minor burden on daily practical problems.²⁷

There are potential limitations that must be considered when interpreting this research. Elements of social desirability in the participants' responses are possible, as participants may not have wanted to report severe limitations to their health care team, some of whom were involved in the research study, or in the presence of family members. It is possible that the emphasis on participant confidentiality and encouragement to complete self-report questionnaires independently addressed this concern. The broad age range included in this study allowed for a representative sample; however, there may have been some heterogeneity in how participants responded to the questionnaires. To mitigate this, we used age-specific versions (e.g., children vs. adolescent) whenever possible. This study was limited to English-speaking patients, who were physically well at enrollment (having had no major bleeds recently), which may prevent the generalizability of the results to other patients with hemophilia. However, only four patients were excluded from the study, which ensures the sample is representative of the entire hemophilia population in a pediatric comprehensive care hemophilia center located in a large, multicultural city (Toronto, Canada).

Although the participation scale modified for this study was untested before our research, our tests for internal consistency met minimum recommended cutoffs. The modified scale addresses the personal and subjective nature of quality of life²⁸ and this feature was the primary reason why this scale was selected. Future work may include determining the psychometric properties of the scale for children.

Finally, the questionnaires alone may not have adequately assessed the impact of hemophilia. Patients with hemophilia today in Canada can live a relatively normal life, but intermittently the disease may cause pain associated with bleeds and the burden of

illness becomes apparent even in countries with access to good medical care and safe clotting factor concentrates as in the center participants were treated. Although hemophilia may be more of an inconvenience than a disability on a daily basis in countries where there is unlimited access to safe, clotting factor concentrates free of cost to individuals with hemophilia (as is the case in Canada), there are countries where a diagnosis of hemophilia is severely limiting because of a lack of access to adequate medical care and safe replacement clotting factor concentrates. These challenges were not applicable for our study cohort, but they do exist globally and may not always be fully captured by questionnaires. Future studies may wish to explore qualitative methodologies to add richness to the data investigating the impact of childhood hemophilia in Canada and explore social participation in countries where access to treatment is more limited.

In keeping with the ICF model, the data illustrate that the interaction between personal factors as well as the health condition of hemophilia are associated with the social participation of a person with hemophilia. Although the majority of participants reported few participation restrictions, there were some who reported very low self-perception scores in each of the domains. In spite of all that has been achieved regarding hemophilia care, it is important to bear in mind that chronic illness is a part of these boys' lives and that sometimes hemophilia can be a very difficult illness to cope with.²¹ By expanding knowledge about social participation in boys with hemophilia, this study may facilitate and inform program planning, monitoring, and evaluation, including the assessment of interventions regarding social participation,³ and may guide psychosocial interventions when needed.

ACKNOWLEDGMENTS

The authors thank the participants for their time and effort to complete this study. They also thank Marjorie McLimont and the rest of the research staff in the Pediatric Comprehensive Care Hemophilia Program at The Hospital for Sick Children for their support and valuable contributions to this study. Preliminary results were presented at poster sessions at the World Federation of Hemophilia World Congresses in 2012 and 2014.

RELATIONSHIP DISCLOSURE

Dr. Blanchette reports that he is Chair of the International Prophylaxis Study Group (IPSG), a cooperative study group funded by education grants from Bayer Healthcare, Bioverativ/Sanofi, Novo Nordisk, Pfizer, Shire/Takeda, and Spark Therapeutics to the Hospital for Sick Children ("SickKids") Foundation. He has received fees for participation in Advisory Boards/Education events supported by Novo Nordisk and Takeda. Additionally, he has participated in Data Safety Monitoring Boards (DSMB) for Octapharma and Takeda. He has received investigator-initiated, industry-sponsored funding from Bayer Inc., Bioverativ/Sanofi, and Takeda. Dr. Feldman has served as a consultant for, and received research funding from, Novo Nordisk. The remaining authors declare that there is no conflict of interest regarding the publication of this article.

AUTHOR CONTRIBUTIONS

Aubrey S. Chiu, Victor S. Blanchette, Maru Barrera, and Brian M. Feldman conceived the study; Aubrey S. Chiu was the main contributing author. Aubrey S. Chiu, Brian M. Feldman, and Nancy L. Young contributed to statistical analyses. Victor S. Blanchette, Pamela Hilliard, and Audrey Abad collected data from participants. Victor S. Blanchette, Maru Barrera, Nancy L. Young, Pamela Hilliard, Audrey Abad, and Brian M. Feldman contributed to revision and rewriting of the manuscript. All authors read and approved the final manuscript.

CONSENT TO PARTICIPATE

All participants and parents/guardians were required to give written informed consent or assent as applicable.

CODE AVAILABILITY

SAS version 9.2 was used for statistical analysis; code available upon request.

ORCID

Aubrey S. Chiu  <https://orcid.org/0000-0001-7056-9700>

Victor S. Blanchette  <https://orcid.org/0000-0003-3341-5010>

Maru Barrera  <https://orcid.org/0000-0002-4079-6686>

Pamela Hilliard  <https://orcid.org/0000-0002-3123-3400>

Nancy L. Young  <https://orcid.org/0000-0002-1739-3299>

Audrey Abad  <https://orcid.org/0000-0001-8396-3168>

Brian M. Feldman  <https://orcid.org/0000-0002-7813-9665>

TWITTER

Aubrey S. Chiu  @AubreySChiu

REFERENCES

1. Carcao MD. The diagnosis and management of congenital haemophilia. *Semin Thromb Hemost*. 2012;38(7):727-734.
2. World Health Organization. *International Classification of Functioning, Disability and Health: ICF*. World Health Organization; 2001.
3. van Brakel WH, Anderson AM, Mutatkar RK, et al. The participation scale: measuring a key concept in public health. *Disabil Rehabil*. 2006;28(4):193-203. doi:10.1080/09638280500192785
4. Harter S. *Manual for the Self-Perception Profile for Children: Revision of the Perceived Competence Scale for Children*. University of Denver; 1985.
5. Coppola A, Cerbone AM, Mancuso G, Mansueto MF, Mazzini C, Zanon E. Confronting the psychological burden of haemophilia. *Haemophilia*. 2011;17(1):21-27. doi:10.1111/j.1365-2516.2010.02280.x
6. Trzepacz AM, Vannatta K, Davies WH, Stehbins JA, Noll RB. Social, emotional, and behavioral functioning of children with hemophilia. *J Dev Behav Pediatr*. 2003;24(4):225-232.
7. Harter S. *Social Support Scale for Children: Manual and Questionnaires*. University of Denver; 2012.
8. Khair K, Holland M, Carrington S. Social networking for adolescents with severe haemophilia. *Haemophilia*. 2012;18(3):e290-e296. doi:10.1111/j.1365-2516.2011.02689.x
9. Iannone M, Pennick L, Tom A, et al. Prevalence of depression in adults with haemophilia. *Haemophilia*. 2012;18(6):868-874. doi:10.1111/j.1365-2516.2012.02863.x

10. Beeton K, Neal D, Watson T, Lee CA. Parents of children with haemophilia – a transforming experience. *Haemophilia*. 2007;13(5):570-579. doi:10.1111/j.1365-2516.2007.01494.x
11. Varni JW, Sherman SA, Burwinkle TM, Dickinson PE, Dixon P. The PedsQL™ family impact module: preliminary reliability and validity. *Health Qual Life Outcomes*. 2004;2(1):55. doi:10.1186/1477-7525-2-55
12. Nunnally JC. *Psychometric Theory* (2nd edn). McGraw-Hill; 1978.
13. Harter S. *Manual for the Self-Perception Profile for Adolescents*. University of Denver; 1988.
14. Hegeman AK, Van Genderen FR, Meijer S, Van Den Briel MM, Tamminga RYJ, Van Weert E. Perceived competence in children and adolescents with haemophilia: an explorative study. *Haemophilia*. 2011;17(1):81-89. doi:10.1111/j.1365-2516.2010.02357.x
15. Harter S. *Manual for the Social Support Scale for Children*. University of Denver; 1985.
16. Hemphill JF. Interpreting the magnitudes of correlation coefficients. *Am Psychol*. 2003;58(1):78-79. doi:10.1037/0003-066X.58.1.78
17. Norman GR, Streiner DL. *PrettyDarnedQuick (PDQ): Statistics* (3rd edn). People's Medical Publishing House; 2003.
18. Feldman BM, Rivard GE, Babyn P, et al. Tailored frequency-escalated primary prophylaxis for severe haemophilia A: results of the 16-year Canadian Hemophilia Prophylaxis Study longitudinal cohort. *Lancet Haematol*. 2018;5(6):e252-e260.
19. Gringeri A, Von Mackensen S, Auerswald G, et al. Health status and health-related quality of life of children with haemophilia from six West European countries. *Haemophilia*. 2004;10(suppl 1):26-33.
20. Agle DP. Hemophilia - psychological factors and comprehensive management. *Scand J Haematol*. 1984;33(S40):55-63. doi:10.1111/j.1600-0609.1984.tb02545.x
21. Thomas D, Gaslin TC. "Camping Up" self-esteem in children with hemophilia. *Issues Compr Pediatr Nurs*. 2001;24(4):253-263. doi:10.1080/014608601753260344
22. Kellerman J, Zeltzer L, Ellenberg L, Dash J, Rigler D. Psychological effects of illness in adolescence. I. Anxiety, self-esteem, and perception of control. *J Pediatr*. 1980;97(1):126-131. doi:10.1016/S0022-3476(80)80152-1
23. Williams KA, Chapman MV. Social challenges for children with hemophilia: child and parent perspectives. *Soc Work Health Care*. 2011;50(3):199-214.
24. Petrini P, Seuser A. Haemophilia care in adolescents - compliance and lifestyle issues. *Haemophilia*. 2009;15:15-19. doi:10.1111/j.1365-2516.2008.01948.x
25. Buzzard BM. Sports and hemophilia: antagonist or protagonist. *Clin Orthop Relat Res*. 1996;328:25-30.
26. Wiedebusch S, Pollmann H, Siegmund B, Muthny FA. Quality of life, psychosocial strains and coping in parents of children with haemophilia. *Haemophilia*. 2008;14(5):1014-1022. doi:10.1111/j.1365-2516.2008.01803.x
27. Varekamp I, Suurmeijer P, Rosendaal FR, van Duck H, Bröcker-Vriends A, Briët E. Family burden in families with a hemophilic child. *Fam Syst Med*. 1990;8(3):291.
28. Feldman BM. Issues in the measurement of quality of life in hemophilia. *Rev Bras Hematol Hemoter*. 2013;35(5). doi:10.5581/1516-8484.20130118

How to cite this article: Chiu AS, Blanchette VS, Barrera M, et al. Social participation and hemophilia: Self-perception, social support, and their influence on boys in Canada. *Res Pract Thromb Haemost*. 2021;5:e12627. doi:[10.1002/rth2.12627](https://doi.org/10.1002/rth2.12627)