Description of a Canadian Cohort of Youth and Young Men with Hemophilia Based on Health-Related Quality of Life Measures

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BACKGROUND

Health-related quality of life (HRQoL) is an important treatment outcome in chronic conditions such as hemophilia, and is becoming increasingly relevant as treatment options evolve. Little is reported on the factors that impact HRQoL in hemophilia A (HA). This poster is the first in a series of publications that will describe the longitudinal patterns of HRQoL among youth and young men with HA over a 3-year period.

OBJECTIVES

- To describe the baseline characteristics of a Canadian cohort of youth and young adults who have haemophilia
- To provide a detailed description of the HRQoL scores from this sample
- To lay foundation for an ongoing longitudinal study of this same cohort

METHODS

Participants: Males, 13 to 29 years of age, with moderate or severe HA ,who were treated with Helixate FS, were recruited from 6 Canadian treatment centres. The presence of an inhibitor, HIV infection and symptomatic HCV infection were exclusion criteria.

Data Collection: Demographic and clinical information was collected at baseline, including: severity of hemophilia, treatment program and history of bleeds and target joints.

HRQoL was measured by self-report using an age and disease-specific measure of HRQoL: the Canadian Haemophilia Outcomes – Kids Life Assessment Tool (CHO-KLAT)[1,2] version 2.0 for youth; and the Haemo-QoL-A[3] for adults. In addition, all participants completed the generic SF-36[4] to enable comparison between the youth and adult groups and to other clinical populations.

A physical examination was performed by a haematologist and joint status was measured by a trained physiotherapist using the Hemophilia Joint Health Score (HJHS)[5] version 2.0.

Analysis: HRQoL scores were summarized by age group. Relationships between the various measures were assessed using Pearson's correlations. The impacts of age and joint status were explored using linear regression.

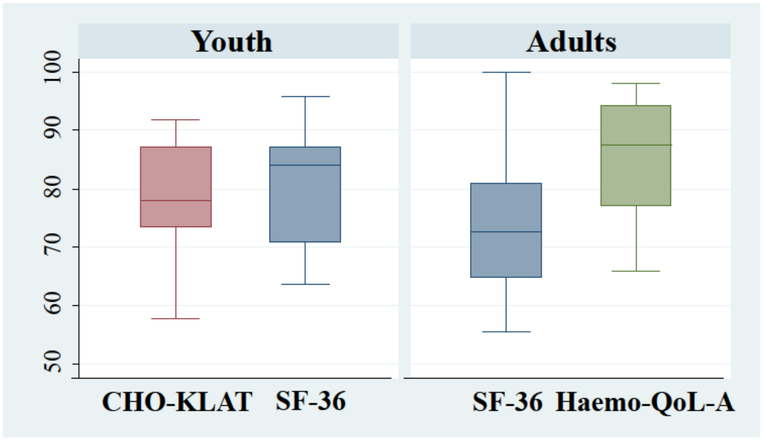
RESULTS

Sample:
13 youth (mean age=15.7; range=12.9-17.9 years)
35 adults (mean age=23.5; range=18.4-28.7 years)
6% moderate and 94% severe hemophilia A
47 were receiving prophylaxis & 1 on-demand infusions

HRQoL Scores from this Study Sample:

Age Group	HRQoL Measure	Mean	Std. Deviation
Youth	CHO-KLAT	77.7	11.1
	SF-36	80.9	9.8
Adults	Haemo-QoL-A	85.3	9.4
	SF-36	73.7	10.6

Quality of Life Score Distributions:



Mean HRQoL scores from previous studies:
CHO-KLAT: 74.6 (SD=14.2)[1] Haemo-QoL-A: 79.1 (SD: 12.3)[3]

Relationship Between Disease-Specific and Generic Measures:

The CHO-KLAT$_{2.0}$ was weakly correlated with the SF-36 (r=0.23, p=0.47) in the youth sample. This is due, in part, to the narrow distribution of scores and small sample of youth (n=13). The Haemo-QoL-A was moderately correlated with the SF-36 (r=0.56, p=0.001) in the adult sample.

Factors associated with HRQoL:

Age: Participants ranged from 12.9 to 28.7 years of age.

We explored the relationship of other factors to HRQoL and found:

- a weak trend of declining CHO-KLAT$_{2.0}$ scores with increasing age (0.73 points per year of age) among the 12 youth with complete data, but this was not statistically significant (p=0.78); and
- a stronger trend of declining Haemo-QoL-A scores with increasing age (0.80 points per year of age) among the 35 adults, but this did not reach statistical significance (p=0.15).
- The impact of age across the combined group of 48 participants was assessed using the SF-36, and demonstrated a reduction of 0.90 points in HRQoL for each year of age, that was statistically significant (p= 0.01).

Joint Status: HJHS scores ranged from 0 to 17 in the youth (median=3) and 0 to 34 in the adults (median=13). CHO-KLAT$_{2.0}$ scores were not correlated with joint status in youth (r= -0.08, p=0.80) and Haemo-QoL-A scores were weakly correlated with joint status in adults (r=-0.27, p=0.03). This is likely due to the broader distribution of HJHS scores in the adults. The SF-36 was weakly correlated with joint status (r=0.27, p=0.07) in the pooled sample.

Note: It was not possible to examine the relationship with severity or treatment program, due to a small number of participants with moderate severity (n=3) and receiving on-demand therapy (n=1).

CONCLUSIONS

- The youth in this cohort had good HRQoL scores and minimal joint disease, while the adults demonstrated slightly worse HRQoL scores and more joint disease.
- In the youth: the disease-specific CHO-KLAT$_{2.0}$ scores were 3.1 points higher than what has been reported in the literature,[1] and 3.2 points lower than generic SF-36 scores. A very different picture was observed in the adults, who had Haemo-QoL-A scores that were 6.2 points higher than what has been reported in the literature,[3] and 11.6 points higher than generic SF-36 scores. However, our adult cohort is younger and had less comorbidity than that reported by Rentz et al,[3] which partially explains the high Haemo-QoL-A scores. Further exploration of the Haemo-QoL-A is warranted.
- The SF-36 appears to provide a good estimate of HRQoL across the age range 13-29 years.

Contribution: These results form the foundation for a longitudinal study that will examine the impact of biological factors and life events on the HRQoL of youth and young adults with haemophilia.

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