Establishing a Registry and Improving Diagnostic Capabilities and Treatment Outcomes for Persons with Haemophilia in Jamaica

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INTRODUCTION

- Jamaica has a population of approximately 2.9 million persons. It is estimated that there are approximately 200 persons with moderate/severe haemophilia (PWH) living in the country. These individuals do not have regular access to clotting factor concentrates and thus rely on blood products such as fresh frozen plasma (FFP)/Cryoprecipitate for prevention/treatment of bleeding .
- It is predicted that affected individuals in Jamaica will have significant complications related to inadequate treatment including musculoskeletal complications, transfusion transmitted infections as well as quality of life issues.
- Funding was obtained from the Novo Nordisk Haemophilia Foundation (NNHF) to characterize PWH in Jamaica with the ultimate goal of developing a Comprehensive Care Haemophilia Program. The Project is conducted in collaboration with the Comprehensive Care Haemophilia Programs in Toronto and Kingston, Ontario, Canada .

METHODS

- In 2014 training of multi-disciplinary team members in Kingston, Jamaica by health care professionals from the Comprehensive Care Haemophilia Programs in Toronto (adult and pediatric) and Kingston , Ontario , Canada was conducted:
  - 5-day program; 44 participants : medical, physiotherapy and laboratory modules.
- In 2015, additional training was conducted in Jamaica with an emphasis on musculoskeletal complications of hemophilia : assessment and management.
- A REDCap-based Haemophilia Registry was developed for the University of the West Indies (UWI) with patient demographics, laboratory results, and selected outcome measures.
- List of outcome measures:

The Hemophilia Joint Health Score 2.1 (HJHS), Pediatric Haemophilia Activities List (PedHAL), Activities Scale for Kids (ASK), Canadian Hemophilia Outcomes-Kids Life Assessment Tool (CHO-KLAT), Pediatric Quality of Life Inventory (PedsQL), Haemo-QoL-A, Functional Independence Score in Hemophilia (FISH)

RESULTS

Characteristics of Patient Cohort

Total of 13 pediatric (<18 years of age) and 33 adult (≥ 18 years of age) patients were registered in the REDCap system. The patient characteristics are listed below (Tables 1 and 2).

Table 1: Patient Characteristics & Laboratory Results

	Haemophilia A	Haemophilia B	Total
Sample	42	4	46
Age: median (range)	26.3 (0.7-69.3)	39.1 (18.7-48.4)	27.1 (0.7-69.3)
Severity			
Mild (6-40%)	4	0	4
Moderate (1-5%)	22	1	23
Severe (<1%)	15	3	18
Laboratory Results			
VWF: Ag: median (IQR: 25%-75%)	134% (99-150) (n=25)	145% (65-153) (n=3)	135.5% (97.5-151.5) (n=28)
VWF activity level: median (IQR: 25%-75%)	100 (79-125) (n=25)	125 (68-171) (n=3)	103.5 (78-125)

Table 2: Annualized Bleeding Rate (ABR) and Target Joint by Severity

	Severe (n=18)*	Moderate (n=23)**	Total (n=41)
ABR median (range)	5.5 (3-12)	10 (3-20)	6 (3-20)
Target Joints Present	9 (50%)	14 (61%)	23
# of Target Joints			
1	3	4	7
2	5	6	11
3	0	4	4
4	0	0	0
5	1	0	1

Target Joint : At least 3 -4 bleeds within a 3 – 6 month period

Outcome Measures

Overall, scores from outcome measures were low indicating a significant burden of disease (Table 3-5). Results from the CHO-KLAT, Haemo-QoL-A and HJHS were lower than those reported from Canada or Other Developed Countries (Figure 1).

Table 3: Pediatric Outcome Measures

	n	Mean (SD)	Min	Max	Best possible score	Worst possible score
CHO-KLAT Child	9	58.8 (12.8)	39.3	78.7	100	0
CHO-KLAT Parent	10	56.8 (13.8)	42.6	83.9	100	0
PedsQL	9	65.0 (15.3)	47.8	93.5	100	0
PedHAL Child	8	84.5 (11.9)	70.5	100.0	100	0
PedHAL Parent	12	79.3 (16.6)	48.3	99.5	100	0
ASK	8	82.0 (8.0)	70.5	95.0	100	0

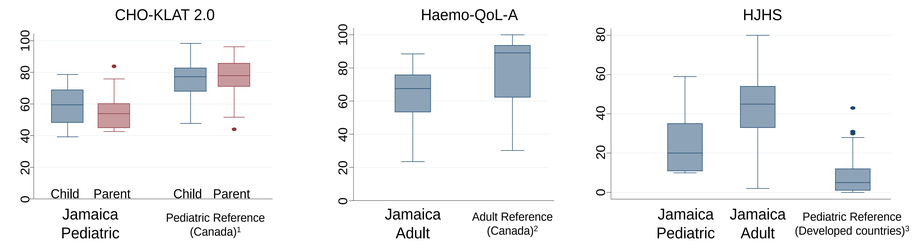
Table 4: Adults Outcome Measures

	n	Mean (SD)	Min	Max	Best Possible Score	Worst Possible Score
Haemo-QoL-A	33	62.0 (18.5)	23.6	88.6	100	0

Table 5: Physical & Function Measures

	n	Median (IQR:25%-75%)	min	max	Best Possible Score	Worst Possible Score
HJHS	39	42 (25-53)	2	80	0	128
FISH	37	25 (19-28)	13	32	32	0

Figure 1: Score Distributions of the CHO-KLAT, Haemo-QoL-A, and HJHS



SUMMARY & CONCLUSIONS

- The specialized training has enabled cataloging of PWH in Jamaica using standardized clinical and laboratory outcome measures, and patient-reported outcomes. Preliminary data indicate a very high burden of disease in the quality of life and physical functioning of persons with moderate and severe haemophilia in Jamaica.
- These detailed observations are an essential first step in advocating for the development of a Reference Comprehensive Care Hemophilia and Inherited Bleeding Disorders Program in Jamaica.

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ACKNOWLEDGEMENTS

- The capacity-building haemophilia project in Jamaica is funded by the Novo Nordisk Haemophilia Foundation (NNHF). The additional advocacy study was funded by the World Federation of Hemophilia advocacy grant, complementing the NNHF project.