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Short Communication

Has the Quality of Life Improved for Children with Haemophilia A?

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Abstract

Intuitively it would be assumed that the quality of life of current paediatric haemophilia patients has improved due to treatment modalities and integrated haemophilia centre support programs. This study was performed to determine if this is indeed the case. A small number of 8-12 year old haemophilia patients were asked to fill out the Haem-o qol questionnaire. Adult haemophilia patients were also presented with the 8-12 year questionnaire and asked to fill it out as they remembered their childhood experiences. Examining the median overall ratings, the adults reported higher values. However, of the eleven subscales only four showed a statistically significant difference ($p<0.05$) between the children and adults (view of yourself, friends, dealing with haemophilia and global health). Quality of life has improved for paediatric haemophilia patients, with adults having a higher median value for all of the eleven subscales. Apart from sport and school where improved treatment has contributed to a better physical outcome a major area of improvement is the psychosocial interaction of the paediatric population. There are still areas where improvement can be made to increase the QoL of persons with haemophilia.

INTRODUCTION

Haemophilia A is an X-linked bleeding disorder, predominantly affecting males, which results from reduced activity of factor VIII [1]. Bleeding into joints can result in significant morbidity with restricted movement in the joint impairing daily activity [2]. There is also a strong psycho-social component to the disease with persons with haemophilia (PWH) viewing themselves as different and being concerned about their interactions with their peers [3-5].

Thus in the case of haemophilia, quality of life (QoL) assessment necessarily involves both physical and social issues. With the marked improvement in treatment and support through haemophilia treatment centres, the quality of life for young people with haemophilia would be expected to be higher than that experienced by their older counterparts.

However, in comparing themselves to their non-haemophilia peers, the fact that they still have physical restrictions along with the inconvenience and pain of treatment, may result in them assessing their QoL as no better than did older PWH at the same age.

This study, on a small number of paediatric and adult PWH, was performed to determine if the premise that the quality of life of paediatric persons with haemophilia has improved and also to establish any areas where improvement in QoL have not been met [6].

MATERIAL AND METHODS

The study was conducted through the haemophilia centres at the Women's and Children's Hospital, North Adelaide and the Royal Adelaide Hospital, Adelaide, South Australia. Ethics approval for the study was obtained from both institutions.

The Haemo-qol 8-12 year questionnaire [7] (see http://www.haemoqol.org for details) was distributed to eight paediatric (8-12 year old) haemophilia patients, (6 severe, 2 mild) and also to 20adult PWH (aged 44-73, median 57) (4 severe, 10 moderate, 6 mild). Due to the small number of subjects available all degrees of severity were included. The adults were asked to complete the Haemo-qol 8-12 year questionnaire by remembering their feelings and perceptions as an 8-12 year old. While this may be perceived as a bias many of the respondents reported it had been easy to recall their childhood, with most recalling bleeding episodes as a base point.

The subscales investigated in this age group through the questionnaire are as follows -:

Physical health, feeling, self -view, family, friends, perceived support, other persons, sports and school, dealing with haemophilia, treatment and global health.

High scores obtained for the subscales are an indication of reduced quality of life. The mean raw scores obtained for each patient for each subscale were transformed to a percentage using the following formula -:

Raw score – minimal possible raw score (of that subscale)

TSS = 100 x possible range of raw scores (of that subscale)

The transformed subscale scores were then used in the statistical comparisons between the children and adults. Due to non-normality in some of the data or a significant Bartlett's test,

for consistency, results from the non-parametric Kruskal Wallis rank sum test are reported for the comparison of the two groups across all the subscales.

RESULTS

Examining the median overall ratings, the adults reported higher values, which were not unexpected (Figure 1). However of the eleven subscales only four showed a statistically significant difference between the children and adults (view of yourself, friends, dealing with haemophilia and global health) (Table 1). For the subscale "view of yourself" a significant difference was observed between the children (median score = 11.1) and the adults (median score = 33.3) (p=0.04). The median score for the children on the "friends" subscale was 31.3 compared to 71.9 for the adults (p=0.003). For the "dealing with haemophilia" subscale the median score for the children was 21.4 compared to 32.1 for the adults (p=0.049). Finally, a significant difference was observed for the "global health" subscale (p=0.0006) – the median score for the children was 0 and for the adults it was 50.

DISCUSSION

There were several limitations to this study. Firstly the ability of the adults to recall their childhood to respond to the questionnaire. It has been reported that events that were extremely salient to a person are easily recalled. Beckett et al., in a longitudinal study indicated that health conditions having an impact on daily activities were repeatedly reported [8]. Krall et al [9], found that childhood communicable diseases were accurately recalled at age 50. These were one off episodes and therefore chronic illnesses should allow for easy recall.

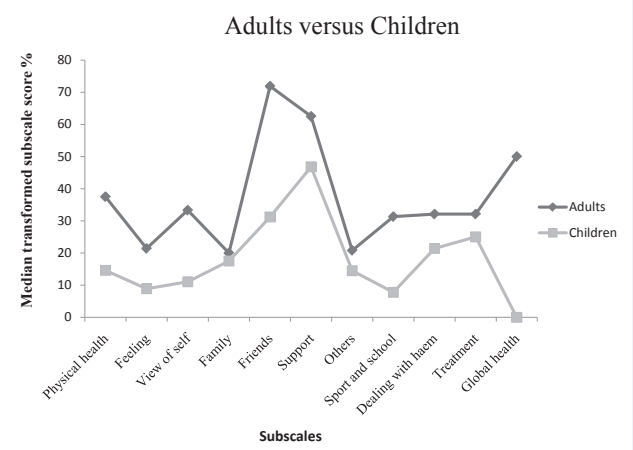

Figure 1 Median transformed subscale scores (TSS) % for each of the subscales.
TSS% = 100 x (Raw score – minimal possible for that subscale)/max possible score for that subscale)

Table 1: Statistical analysis of the children versus adults for each of the subscales investigated.

Subscale	*P* value (Kruskal- Wallis Rank Sum Test)
Physical health	0.56
Feeling	0.23
View of self	**0.04**
Family	0.61
Friends	**0.003**
Perceived support	0.61
Other persons	0.20
Sports and school	**0.06**
Dealing with haemophilia	**0.049**
Treatment	0.78
Global health	**0.0006**

Secondly the high percentage of severe cases in the paediatric group could bias the QoL in a negative fashion. Despite this the paediatric group still reported a much better QoL.

Overall the adults indicated they felt more self-conscious about having haemophilia and that it made their life more difficult. They reported feeling isolated with their haemophilia. The current paediatric group was more likely to have a friend that was supportive.

The difference in the sports and schools subscale tended towards significance (p=0.06).Presumably current treatment regimens mean there is less restriction on sporting activity for the young boys even with severe haemophilia [10]. Education of teachers has allayed their fears for the student, allowing them to participate in the majority of school activities.

Interestingly the older group felt that they were not necessarily in control of their situation. They indicated that although having accepted they had haemophilia, they did not feel well informed about the condition. They also indicated they were less likely to recognize a bleed than the current paediatric group.

Global health produced the most significant difference between the adults and children. However this was due to the fact that the adults generally reported their global health as good, while the children reported their health as excellent.

The effect haemophilia has on the family does not appear to have changed with time. Parents have not changed in their protection of their children nor does it appear more or less convenient for current parents caring for their children. Perhaps a surprising outcome was that both groups had a similar rating for the subscale treatment. This was one subscale where one would have anticipated a distinct difference. This subscale is less about the effectiveness of treatment and more about the processes surrounding treatment - the haemophilia centre and inconvenience of injections. Until the use of longer acting factor replacement or alternatives become routine [11,12] this area of the quality of life for PWH will not show improvement.

Considerable progress appears to have been made in improving the children's comparison of themselves and their peers. The ability of the boys to relate to a special friend has shown a dramatic improvement.

The disappointing outcome is that perceived support has not shown any great improvement and was a high scoring subscale for both the adults and children (Figure 1) [13]. The multi-disciplined approach employed by modern day haemophilia centres will hopefully overcome this perception. The centres need to provide both substantial physical and psychological support. Thus, while the quality of life has shown improvement for paediatric patients with haemophilia, there are still areas where this can be advanced.

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