

## **An exploration of health related quality of life in adults with haemophilia – A qualitative perspective**

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## **Summary**

Musculoskeletal dysfunction is a common feature of haemophilia and along with other manifestations of this condition, there is a general perception that health related quality of life (QoL) will be affected. Previous research using standardised questionnaires has demonstrated that QoL is lower in haemophilia groups compared to normal populations. However interviews with disabled people and disability studies suggest that many disabled people experience positive life changes as a result of their illness and an affirmative model of disability has been proposed. A qualitative study involving focus groups and interviews was undertaken to explore these issues in a group of 19 severely affected adults with haemophilia. The focus groups and interviews were tape recorded and fully transcribed and the results subjected to thematic analysis. This paper focuses specifically on key issues that impacted on perceptions of QoL. The findings suggest that the participants' perceptions of QoL were very positive. Possible reasons are proposed including the benefits of factor replacement, participants recollections of their lifestyle before factor replacement, that having haemophilia was integral to the 'self' and finding a niche where they could be successful. Suggestions for a more positive affirmation of haemophilia could be considered when developing QoL measures.

## **Introduction**

Musculoskeletal dysfunction is a common manifestation of haemophilia particularly in severely affected individuals as bleeding can occur spontaneously into joints and muscles. This can lead to arthropathy and contractures, which may be associated with pain, deformity and loss of function. Current medical management focuses on prompt and adequate factor replacement to treat bleeding episodes and prophylactic treatment programmes to minimise bleeding into joints and muscles are now utilised [1-3]. However, in spite of the availability of factor replacement, adults with haemophilia still experience bleeds and those adults who did not have access to treatment in their early life may have developed arthropathy. They may also have to deal with complex psychological and social issues as a result of haemophilia [4] and may have been infected with HIV and hepatitis as a consequence of using contaminated blood products in the past. It is not surprising therefore that there is a general perception in the health care community that these issues will impact on quality of life (QoL).

### *Quality of life studies in haemophilia*

There has been an emerging interest in recent years in evaluating the effect of haemophilia on QoL with an increasing number of published studies on this topic [5-14]. Most studies have used generic QoL questionnaires, in particular the Medical Outcomes Study SF36 (SF36). The SF36 is a well-known example of a standardised QoL questionnaire that evaluates eight dimensions of life to assess the quality [15]. The results of the SF36 are often compared to normal population figures in order to evaluate the impact of a specific health condition. In the haemophilia population the majority of the studies undertaken have been cross section studies. The test retest reliability and sensitivity of the SF36 has not been specifically evaluated for individuals with haemophilia therefore its value as an assessment tool has not been fully explored. More recently, haemophilia specific questionnaires have been developed [16-18]. Disease specific questionnaires tend to ask more relevant questions and therefore be more sensitive to change but results cannot be compared to normal population data and the questionnaires may be less likely to detect unanticipated effects of the disease [19].

Overall the results of published studies have demonstrated a lower QoL for adults with haemophilia in comparison to those people with no specific health problems [13,20]. Several factors have been perceived to reduce QoL when correlating the

scores from standardised questionnaires with various personal attributes. These include having impairments due to arthropathy, repeated bleeds, a history of orthopaedic surgery and being HIV positive, however findings between studies have been conflicting. Possible reasons for this may include assessment tools not being sensitive enough to detect changes, findings being dependent on when the questionnaires are administered or that these issues may not impact on QoL [12,21].

#### *Rationale for the study*

One of the potential limitations with standardised QoL questionnaires is that they do not consider individual patient values and therefore some components of the questionnaire may not be relevant to specific participants or important aspects of a patient's QoL may be omitted [22]. Cella [23] cautioned against the use of standardised questionnaires alone to evaluate QoL as patients may misunderstand the questions or issues important to the patient may not be asked. The authors recommended that any standardised questionnaire should be supplemented with an interview. Qualitative methods such as interviews promote the collection of data which can be analysed for interpretation and meaning and may provide greater insights into how QoL is affected [24].

An interesting concept has emerged when authors have interviewed people who have chronic illnesses about their QoL. Participants with apparently poor health often reported good QoL and this has been called 'the disability paradox' [25]. Interestingly, several authors [5,6,26] who have investigated the impact of haemophilia on QoL expected QoL scores to be lower than the scores actually achieved. They concluded that coping strategies and other mechanisms may reduce the impact of haemophilia.

Many disabled people express positive life changes as a result of becoming disabled and do not perceive disability as 'a tragedy', a common perception of health care professionals [27]. These positive perceptions have been described within the affirmative model of disability [28]. This model espouses the view that disability can 'enhance life or provide a lifestyle of equal satisfaction or worth' [28 p570]. The focus is on a non-tragedy view of disability with disability being acknowledged and celebrated as an integral part of the self [28]. Reynolds [29] highlights the practical, social and emotional difficulties that people face when experiencing a chronic illness. Peoples' response to illness will be associated with 'their emotional state, belief system, coping strategies and support mechanisms' but will also be related to the seriousness of the illness and the societal response to that illness [29 p162].

Although people may report negative perceptions related to chronic illness including difficulty coping with unpleasant symptoms, depression, loss of self esteem, uncertainty about the future, loss of job and social isolation, individuals often regard their illness positively. Indeed people with long term or congenital disorders have been reported to accept their condition as a part of normal life [29]. A number of explanations why people might describe chronic illness as a positive experience have been proposed [29]. These include improving relationships, benefits from developing social networks by assisting others with similar problems and re-evaluating priorities and personal values [29-33]. This will have implications for how individuals evaluate their QoL and for any instrument that purports to measure that QoL. Certainly, studies have demonstrated that patients often rate their QoL better than health care professionals, carers or partners suggesting that QoL is a personal issue [34]. These concepts may have relevance to people with haemophilia that have yet to be explored.

Extensive database searching did not identify any published literature that specifically interviewed adults regarding if, and how haemophilia impacts on QoL. The aim of this research was therefore an exploratory qualitative study to investigate this question.

### **Methodological considerations**

A qualitative research methodology was employed including focus groups and semi-structured individual interviews. Focus groups can be a useful method when researching an unexplored field as they provide an opportunity for participants to discuss issues that they consider important which will therefore inform the researcher [18,35]. Focus groups usually involve 6-8 participants who meet for 1-2 hours to discuss a particular topic [36]. However, smaller groups can still be successful if the participants have interest and experience in the topic of discussion [37]. The proposed advantage of a focus group versus an individual interview is that it is the interaction between the participants that enables them to explore attitudes and opinions and clarify their views [35,38]. Focus groups have been successfully employed in order to ensure that a QoL questionnaire for children reflected the experiences of the participants [18].

### *Method*

Ethical approval was granted for the study. Only severely affected adults were included as it was perceived that their QoL was most likely to be affected by haemophilia. All adults (n=105) over the age of 16 years with Haemophilia A or B

registered at the Royal Free Hospital were contacted by letter to determine whether they would be prepared to attend a focus group at the Haemophilia Centre to discuss QoL issues. Individual interviews were also offered to explore the views of participants if they did not wish to attend a group discussion.

Twenty adults (19%) returned reply forms indicating their interest in being involved in focus groups and/or interviews. Ten adults opted to attend a focus group but two adults could not attend on the dates arranged and had individual interviews instead. One participant who returned a reply form could not be contacted.

There were two adult focus groups. One focus group involved three participants and the other group had five participants. The ages of the participants in the focus groups ranged from 27-73 years with a median age of 34 years. The median age of all severely affected patients registered in the centre was 35.55 years. All the participants in the focus groups had Haemophilia A. One participant in each group was HIV positive and one participant used a wheelchair due to severe arthropathy of his lower limbs as a result of insufficient treatment with factor concentrate in his youth. There was also a set of twins in the larger focus group. Six of the eight participants were working. One of the HIV positive participants was not working and one participant was retired.

Eleven interviews with adults with haemophilia were conducted. Nine adults chose an individual interview. Reasons for not attending a focus group included not wishing to discuss issues with other participants, practical issues associated with difficulties attending the haemophilia centre and one participant who had had previous experience of organising focus groups as part of his work and had not found them beneficial. The ages of the participants who were interviewed ranged from 38-72 years with a median age of 50 years. Six participants had haemophilia A, two of whom had inhibitors and five had haemophilia B. One of the participants with a high titre inhibitor used an electric wheelchair due to marked arthropathy. Four participants with haemophilia A were HIV positive, three of whom were working. Of the other seven participants, three were working, two were not working due to haemophilia related problems and two were retired.

### *Procedure*

The respondents were contacted via email or telephone to arrange a suitable time and place for a meeting. All the focus groups and the majority of the interviews took

place in the haemophilia centre. For practical reasons two interviews took place in the author's (KB) main place of work. One other interview took place in the interviewee's own home. All of the interviews and focus groups lasted between one to two hours.

At the beginning of the focus groups participants were informed that information discussed within the group was confidential and should not be disclosed outside that setting. After a general introduction, the opening question was 'so how does haemophilia impact on QoL'. Participants in both focus groups and interviews were free to explore issues relevant to them. Prior to the discussions, the interviewer had developed a series of prompts relating to different aspects of QoL. These were based on clinical experience of haemophilia as well as from the literature on QoL. These prompts were addressed as relevant to the situation and the focus of the discussion. Issues raised in previous sessions were explored with subsequent participants to ascertain their relevance. All the focus groups and interviews were tape recorded and then fully transcribed. Once the tapes had been transcribed, they were checked and then returned to the individuals concerned for verification and any additional comments. Sixteen transcripts were returned. One participant made extensive additional notes on his personal reflections of haemophilia and several participants made minor editing changes to their transcripts. The tapes were destroyed on completion of the study.

#### *Analysis of data*

This was an exploratory qualitative study that seeks to develop understanding and meaning from the data [39]. The transcripts were all read several times and a preliminary analysis was undertaken looking for common themes between the transcripts. Key phrases and topics were highlighted from each focus group and interview. These were summarised and then integrated with responses from other participants and collated under various categories or themes. [40] argues that reliability between coders is only applicable for semi-structured interviews not unstructured interviews. The interview style in this study was more related to unstructured interviewing as described by Morse [40] than semi-structured interviews. Participants were not all asked the same questions as the aim was to explore issues of relevance to the participants rather than follow a set format of questions. Therefore the themes were not 'verified' by another researcher. Such manipulation of the data can lead to a superficial overview rather than a deeper analysis that is possible for a researcher more intimately involved with the data [40].

However it is important that the researcher does not miss important topics or themes by becoming too narrow in perspective which could threaten the credibility of the study. Therefore two independent peers read three transcripts (one focus group and two interviews) that had been randomly chosen by the researcher and identified the issues that they felt emerged from the data. This process of peer collaboration was undertaken to avoid 'tunnel vision' and enabled the data to be considered from different perspectives so enhancing credibility [41].

The Nvivo computer program was subsequently used to assist in the coding of the data. Six broad themes emerged from the data. These were

- functional activities
- factor replacement
- social impact
- coping with additional illnesses
- psychological impact
- the impact of haemophilia on health related QoL.

From within these broad themes a number of sub categories were developed. The data in each category was then reviewed and categories with a large number of entries and a variety of issues within the category were further analysed into smaller sub categories. This paper will focus specifically on key issues that impacted on perceptions of QoL.

## **Results**

The findings have been considered under the following headings. These include benefits of factor replacement, recollections of lifestyle pre factor replacement, haemophilia is integral to the self, finding success in life, altruism and the impact of these issues on QoL.

### *Benefits of factor replacement*

Availability of factor replacement was a crucial aspect of participants' QoL as it reduces the incidence and severity of bleeds. It gave the participants' freedom to live a 'normal' life, and importantly enabled them to plan their life and be able to look to the future. They could also undertake social activities with less concern about whether they would have a bleed and have to cancel an engagement.

*Int3: Impossible to describe how living with severe haemophilia with no treatment affects you. That does make a difference. The uncertainty. The total disruption to your life. Increasing*



*disability. With treatment that goes. It revolutionises your life. Like patients on dialysis having a kidney transplant.*

Having factor replacement available enabled them to put haemophilia to the back of their mind. The participants wanted to be independent and have control and be able to make their own decisions based on their perceived assessment of risk. Participants in one of the focus groups recognised that between them, they used different strategies in managing their haemophilia. For example, some participants were on prophylaxis and considered that beneficial to their lifestyle, others preferred to be 'on demand' treatment. The reasons for their choice included the number of bleeds they experienced, their perception of the inconvenience of undertaking the prophylaxis, perceived difficulties accessing their veins and concerns regarding safety of blood products. The ability to be able to choose what was right for them at a particular time in their lives was important to the participants. This enabled them to perceive that they had haemophilia under control and regard it as 'an inconvenience'.

#### *Recollections of lifestyle pre factor replacement*

The median age of the participants in the study was 34 years and 50 years respectively and so many of the participants could recall their lifestyle prior to availability to factor replacement. This appeared to be an important issue as it impacted on their views of how they managed their haemophilia and how they perceived their QoL now in comparison to their early life. Several participants recalled how they had missed a lot of time from school and had experienced severe pain, immobility and restricted activities when they were younger.

*Int10: The war was on so I was away from home and it wasn't very funny to have a bleed because .... the black-out would go up on the windows and no light would come in, your door would be closed at nine or ten o'clock at night and you were just left alone in terrible pain until 7 or 8 the next morning.*

These experiences made them appreciate the advances in medical care and the benefits of the availability of factor replacement. Several participants reported that they had not expected to live as long as they had, having been given negative predictions by health care professionals regarding expectations of mobility or life span. This made them feel quite proud that they had defied medical opinion by still being alive and mobile decades after it had been predicted.

*Int10: I suppose I am a bit smug, a bit pleased with myself. My parents were told I stood very little chance of living beyond 7. .... Later on when I was at school it was reported back to me that a teacher said 'Aren't you going to do something about x?' and another teacher said 'No there is no point, he will be dead by the time he is 19!'*

Many of the participants also considered that having to manage severe pain and long periods of immobility and isolation from activities with their peers in early life made them stronger and a more resilient person in later life. They were also aware that many of their friends and colleagues were no longer alive due to haemophilia related illnesses such as HIV.

*Int5: I think anybody that has been through haemophilia for quite a long time has to have a fairly hard shell to have survived. That has had an impact .... as well.*

Several of the participants were coping with other health problems such as HIV, hepatitis or inhibitors. One participant illustrated the challenge of living with HIV for twenty years and the uncertainty that they had to live with.

*Int5: It's absolutely the uncertainty, it's the fact that 10 years ago, it was almost easier because 10 years ago one had a reasonable expectation that one was going to be probably dead in about 4/5 years time, 10 years on one isn't which is rather irritating, ironically! I think the problem now is almost that although I am very well I've no idea how long that's going to last.*

#### *Haemophilia is integral to the self*

Participants could not envisage what life would be like without haemophilia as they had lived with the condition all their life.

*Int5: The problem is that when you have got some kind of chronic condition, you don't know what not having it is like! So when people say how are you or are you feeling tired? You've no idea what kind of benchmark anybody else is working to.*

Haemophilia appeared to be an integral part of the 'self'. Their awareness of having haemophilia had emerged gradually as a child, often as they began to realise that not everyone had haemophilia. As a result of this, they found it difficult to regard haemophilia as a problem.

*Int8: I can't look on haemophilia as a particular problem really because I am so used to it, I have had it all my life. If it is a problem who is it a problem for?*

Participants did not report being angry, hostile or depressed as a result of having haemophilia. They expressed their acceptance of the condition and they lived within the limitations of the condition.

*Int4: My attitude to haemophilia has been: 'I didn't ask for it, but I have got it. So let us fight it. It is no good getting upset and blaming the world'.*

One participant felt that his life would have been worse without haemophilia, as it gave him strategies and a way of dealing with things that he would not have had otherwise.

*Int8: So I can't say it is a bad thing to have actually! How would my life have turned out without it? It could have been a lot worse because of being without it.*

### *Finding success in life*

The availability of regular factor replacement ensured that participants generally lost little time from work due to haemophilia related problems. Participants valued their work and the contribution they could make in the workplace.

*Int3: because you have a disability it is important to be able to contribute. I think that I do my job reasonably well. It's important to me, to give something back that I have taken out of the NHS. It is important to be involved, and pay taxes, have a sense of identity.*

Having a job where they were able to compete on an equal basis with their peers was a tremendous boost to self-esteem. This ability to focus on their personal qualities was something that developed as an adult rather than as a child. As a child they were often more aware of being different to their peers and were often unable to participate in activities and sports. This was partly due to less effective treatment being available at that time. The participants had used the time that they had not been able to be involved in sporting activities to their advantage and as they got older their involvement in sporting activities became less important. The necessity to focus on other areas of life was then seen as an advantage of having haemophilia.

*Int5: I found that it (haemophilia) probably was beneficial in some ways, because given that I couldn't participate in sport, I developed my interest in skills like argument and debate, and so I found well I can't beat someone on the cricket field, but I can certainly beat them in the debating chamber.*

For some participants not being able to do certain activities because of haemophilia was seen as an advantage rather than a disadvantage especially if it was something they did not enjoy. For example one participant was unable to undertake a more physical part of his job due to his arthropathy. He perceived this actually improved his QoL. Even those participants who were not working took advantage of opportunities to make the most of their lives. They had the opportunity to travel abroad and several of the participants had traveled extensively. They pursued hobbies and interests, and met people with similar interests.

*Int11: I have been quite lucky and there is no point in feeling bitter about what you could have had, you just have to make the best of what you can and use your abilities in other ways....I can't do the physical (aspects) so you just find some other way where you can contribute to it, to be still within it but without having to rely on physical strength.*

### *Altruism*

Their gratitude to the health care profession caused them to behave in an altruistic manner. As a result they were often willing to participate in health professionals' research projects, they contributed to charity committee work and were involved in fundraising to support others. This was regarded as an important part of 'giving back' to society and contributing to the NHS. This benevolence may in part be because the participants were very aware of the costs of factor replacement and the scarce

resources of the NHS. Their first hand experience of haemophilia also enabled them to be in effect 'the experts' and by sharing information and insights into their condition, they could improve the experience of others.

*Int9: My main skill area is being a patient and seeing things from both sides of the fence and a certain amount of fund-raising, ideas and working on it with people, policy things.*

This was particularly relevant if they regarded these activities as something that they were skilled at doing. This contributed to them feeling valued and able to contribute to society.

*Int5: I will always be involved in something, that's my nature. I wouldn't be here if it wasn't for the NHS or the welfare state. If I can give something back in some way then I should. I would have been dead years ago..... It's something I can do, I have a skill there, I'm good at it, if I can use it to some advantage or help somewhere then I will. It also keeps the intellect going, and that's the most important thing as far as I am concerned. An awful lot of me doesn't work but fortunately my brain does. My arms and legs might be useless, but my brain works so I will make the most of it.*

As a result of their past experiences of their lifestyle as a child, the availability of factor replacement now, their ability as adults to contribute to society through work or other activities, the participants were able to maintain their self esteem and feel valued. They reported that they were 'fortunate', 'lucky', and were able to live life to the full. They demonstrated a positive attitude to life and this impacted on how they perceived their QoL.

*FG2(5): I don't think haemophilia has ever got me down. Does not stop me doing what I want to do, it doesn't stop me having a good life. Obviously there are things that I don't do but I live life to the full. I think I am very fortunate.*

## **Discussion**

Haemophilia is a condition that can affect many aspects of an individual's life. Previous research investigating QoL in haemophilia has demonstrated that the participants generally have a lower QoL compared to people who do not have haemophilia. However most of the research has used standardised questionnaires. The participants in this study, despite being HIV positive and /or having a variety of musculoskeletal dysfunctions were still able to report a good QoL. A number of explanations can be proposed to explain these findings.

People who develop chronic illness in later life often experience biographical disruption where their whole projected life history is affected by the onset of an illness [42]. People with congenital illness may perceive their illness differently and are less likely to experience feelings of loss or change as a result of their illness [29,43]. The discussions with these participants suggest that because haemophilia is a congenital condition it is difficult to separate from the 'self' and therefore the impact is less likely

to be perceived as a problem or a loss compared to an illness that may occur later in life.

The participants recognised the enormous advances and improvements in haemophilia care over the last 20 years. They therefore considered that they were 'lucky' in many aspects of their lives. Access to factor replacement reduced the pain and immobility associated with uncontrolled bleeds and enabled them to plan for the future. Indeed, the memories of their early experiences of haemophilia appeared to have a positive impact on their perceptions of their life now. They seemed to make the most from their lives, seizing opportunities, travelling, being involved in social activities and living life to the full. In many cases they felt that they did more and achieved more than many people with no health problems. Participants sometimes evaluated their life in comparison with others who they perceived as less fortunate than they were, either people with other health conditions or people with haemophilia in countries who did not have ready access to regular supplies of factor replacement. This downward social comparison may be a strategy for maintaining or boosting self esteem [44].

The literature provides evidence on how a chronic illness can lead to a loss of self [45]. The findings of this study demonstrated that these participants rather than experiencing a loss of self were able to maintain, and in some cases enhance, their sense of 'self' and self esteem. The participants focused on finding a niche in life where they could be successful. They chose friends who had similar interests and aspirations. They focused on their own personal qualities and aptitudes that they were good at, so that they were still able to make a contribution to others and to society and this enabled them to feel valued. Participants often found success in the workplace. This is important in a society that values individuals who contribute to society. Several participants who appeared to have an inner drive and ambition in the workplace did not feel that haemophilia had restricted their ambitions. In fact having haemophilia seemed to open up new avenues and opportunities that might not have been explored otherwise and enabled them to focus on aspects of life that they would not otherwise have done.

Studies involving people with chronic ill health have identified that they often place less value on their health and work and more on relationships and helping others [29]. The participants in this study still valued their opportunity to work, possibly because they had not had a sudden change in health status in later life. Work

provided them with a sense of identity and enabled them to maintain their self esteem.

Having a good QoL was regarded as a bonus rather than an expectation that may characterise those without health problems. However strategies needed to be employed to take advantage of different experiences and opportunities. Planning around various illnesses was regarded as an inconvenience rather than a major compromise and did not prevent enjoyment of life. There was also a general feeling of optimism about the management of haemophilia through the possibility of more effective treatment in the future or even a cure through gene therapy. Psychological traits such as mood, coping style and self-evaluation of the health condition will impact on how individuals assess their QoL [46]. The positive outlook expressed by the participants may begin to explain why they reported a good QoL. Standardised QoL questionnaires tend to focus on negative aspects of health rather than any positive aspects of life as a result of the health condition [46].

One participant noted that it was difficult to evaluate QoL because they were not sure if their expectations were the same as other people. Possibly some people with haemophilia may have lower expectations but because they have no benchmarks to compare they may not recognise that their expectations are lower. Stensman [30] described a model to explain the relationship between fulfillment and expectations. People with high fulfillment in life and low expectations may be predicted to be more likely to report a good QoL and this may be an issue for some people with haemophilia which may warrant further study.

Despite practical difficulties and limitations of their mobility, these adults reported a good QoL by being in control of their haemophilia and focusing on their strengths. These participants conceptualised their values and priorities with reference to haemophilia and valued their life and their achievements more in light of the challenges they faced. Their focus on strengths and the positive aspects of their life enabled them to live life within their limitations but they were still able to enjoy a good QoL.

Although qualitative research methods are viewed by many researchers as 'unscientific', there are a number of strategies that can be employed in order to ensure that the analysis of the data is meticulous and thorough [39]. Terms such as internal validity, external validity and reliability may not be so appropriate for

qualitative data and the terms credibility, transferability, dependability and confirmability are preferred [47]. The credibility of the study was enhanced by ensuring accuracy of the transcripts, verification of transcripts by participants and submitting transcripts to peers for their interpretations. Transferability relates to external validity or generalisability of the data. The nature of qualitative data is such that it is not usually appropriate to generalise the findings beyond the group studied but the aim is to gain an understanding of the participants and their experiences through the analysis and interpretation of rich data. The sample was self selected and the characteristics that many demonstrated, including being highly motivated and altruistic, may encourage this type of individual to volunteer for research activities. It is recognised that their perceptions may not be the same as other individuals with haemophilia. It is also important to acknowledge the median age of participants in this study. Their experience of the less optimal management of haemophilia when they were younger differs from younger adults who have always had access to factor replacement. These experiences may impact on their perceptions of QoL. Further study of the perceptions of QoL in younger adults is warranted.

These issues may be of relevance for those health care professionals who are interested in evaluating QoL for adults with haemophilia. The findings of this study suggest that health care professionals may wish to consider additional methods of assessment as well as standardised measures, when assessing QoL. The negative format of standardised questions could also be reviewed to provide a more affirmative perspective of having haemophilia. Interestingly, negative emotional factors such as anger and sadness were rejected by children as not being relevant to having haemophilia [18]. In addition, a reduced level of functioning on a scale may not equate to a poor QoL.

## **Conclusion**

This study supports the findings of other studies involving qualitative methods that individuals with a chronic health condition can report a good QoL [30,44]. The findings also supports the affirmative model of disability suggesting that disability can lead to positive effects on life [28]. Issues that may impact on the views of participants may include being unable to separate haemophilia from the 'self', as haemophilia is integral to who they are as a person. Due to the ages of the participants, many of them could recall their lifestyle pre factor replacement that appeared to influence how they judged their QoL now. Their gratitude for the improvements in their health was expressed by altruistic behaviour, a common

behaviour of those with chronic health conditions [29]. The good QoL reported by these individuals was also influenced by their ability to be successful in various aspects of their lives and their ability to offer a positive contribution to society. These issues may have relevance for clinicians interested in evaluating QoL of their patients.



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