Haemophilia
The Official Journal of the World Federation of Hemophilia,
European Association for Haemophilia and Allied Disorders and the Hemostasis & Thrombosis Research Society

Physiotherapy outcome measures of haemophilia acute bleed episodes: what matters to patients?

Journal:	*Haemophilia*
Manuscript ID	HAE-00127-2019.R2
Manuscript Type:	Original Article
Date Submitted by the Author:	22-Jul-2019
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Keywords:	acute bleeds, outcome measures, patient opinion, core outcomes

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2nd April 2019

Dear Sirs,

Re: Physiotherapy outcome measures of haemophilia acute bleed episodes: what matters to patients?

Please see included the above article for your review. If you require any further information I am more than happy to be contacted for this.

This work has not been already published nor is it not being currently considered for publication elsewhere by myself or any of the other authors.

Kind regards,

Lizzie

Elizabeth Bradshaw
Specialist Musculoskeletal Physiotherapist
University Hospitals Bristol NHS Foundation Trust.

Prepared Domains with Measures Used. **Bold** measures are haemophilia specific. *Italics* are routinely used at the participating CCC

Domain	Comments	Measures
General function	Questionnaires that can be self-completed or respond to verbal questioning Questionnaires given to patients (e.g. FISH) to gauge current function.	**HEP-Test-Q[15],** McMaster Toronto preference questionnaire[27], **FISH (functional independence score in haemophilia)[14], HAL (haemophilia activities list)[17],** MAQ (modifiable activity questionnaire)[28], COPM (Canadian Occupational Performance Questionnaire)[24], HAQ (health assessment questionnaire)[18], Colorado score (for adults) [29]. *EQ5D (usual activities section)[16]*
Joint function	Scores completed by therapists. Scores give overall indication of impact of disease on joints.	***HJHS (Haemophilia joint health score[7]),*** **WFH Score (Gilbert score)[30],** Colorado score (for adults)
Mood	Can be self-completed or respond to verbal questioning regarding current mood status.	BDI-SF[19], SF36[20], *EQ5D (anxiety and depression section)[16]*
Pain	Patients are asked to rate pain using visual, verbal or numeric descriptors of type and severity.	Likert scale[22], Visual analogue scale[21], *Pain medication use, Verbal pain intensity[21],* Wong Baker Faces scale[23] *EQ5D;* not a pain specific scale but has a pain section [16].
Quality of Life	Can be self-completed or respond to verbal questions regarding the impact of their condition on quality of life.	**Haem-A-QOL[13],** SF36, *EQ5D[16]*
Range of movement	A measure of the amount of movement at the affected joint.	*Goniometry: a large protractor like device that gives a reading of degrees of movement [31].*
Strength	A measure of the current strength of the patient in the affected area.	Manual measurement: Resisted strength either through range or a static (isometic) measure on a machine such as the biodex. *Therapist assessed: resisted by therapist either through range or statically [32].*
Swelling	A measure of the size and width of bleed or affected limb.	*Bleed size: can be palpated by therapist measured with tape measure. Can demonstrate resolution of bleed. Global swelling (e.g. around calf circumference) could also be taken.*
Work	Self-reported or verbally taken questionnaires discussing impact of general activities or specific activity restrictions.	COPM (Canadian Occupational Performance Questionnaire)[24] Reported number of days off work (by patient) *EQ5D; This asks about usual activities which would incorporate work if a patient normally works. [16]*

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		Haem-A-QOL; this is a quality of life measure but has a specific section about the impact of haemophilia on work or normal activities [13].

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Table 1

1. Title page

1. Full title:

Physiotherapy outcome measures of haemophilia acute bleed episodes: what matters to patients?

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5. Running title

Physiotherapy acute bleeds: outcome measures

Key words: acute bleeds, outcome measures, core outcomes

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2. Abstract

Introduction

The research was conducted at a UK teaching hospital and Haemophilia Comprehensive Care Centre (CCC) as part of a research program investigating physiotherapy for acute bleed management.

Aim

The aim of the study was to understand the perspectives of people with haemophilia (PWH) on validated outcome measures (OM) and whether these measures capture changes relevant to them whilst recovering from an acute bleed episode.

Methods

Any person with haemophilia registered to the CCC who reported an acute bleed within the past 2 years was invited to participate. Semi-structured interviews or workshops (activity-focused discussions with small groups) were conducted with PWH who had received physiotherapy treatment in the previous two years. These were used to explore opinions of PWH of commonly used outcome measures.

Results

Eight male PWH participated, mean age 61 years, ranging between 39 and 71. Seven participants had severe Haemophilia A and 1 had Von Willebrands. Participants described numerical rating scales of pain as abstract and expressed a preference for verbal or visual descriptors. In relation to function, the men generally found haemophilia specific OM to be more relevant. The EuroQol 5-Dimension 5-Level (EQ5D-5L) and Haemophilia and Exercise

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Project Test Questionnaire (HEP-Test-Q) were considered as good measures due to brevity and ability to capture relevant changes promptly.

Conclusion

Participants in this study reported a preference for short OMs that allow them to reference their ability during the acute bleed episode in comparison to their normal function.

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3. Main body of text - Word count 2995

Introduction

Haemophilia is a congenital bleeding disorder caused by a deficiency of coagulation factor. People with haemophilia have a tendency for prolonged bleeding, often into joints or muscles[1]. Haemophilia management has changed significantly over recent years due to increased availability of effective and safe clotting factor agents preventing ongoing bleeding (including recombinant), as well as prophylaxis in countries where it can be afforded. Despite use of prophylaxis reducing incidences, acute bleeds remain common[1] and can result in joint arthropathy, chronic pain, restricted movement and poor physical function[2]. Effective management of PWH is crucial for minimising detrimental impact of bleeds. Physiotherapy plays a key role by helping to restore movement and function and prevent arthropathy[3,4].

Considerable variation currently exists in physiotherapeutic management of acute bleeds in the United Kingdom(UK). A lack of consensus exists between physiotherapists regarding outcome measurement (OM)[5], suggesting the need for clearer guidelines on both managing PWH following an acute bleed, and standardising OMs[6]. For longer-term joint health, service-provider guidelines mandate use of the Haemophilia Joint Health Score[7]. There is however no consensus on core OM for recovery following an acute bleed[8-10]. In addition, little work has been carried out to investigate priorities of PWH when attending physiotherapy generally. Research into use of OM in other long-term conditions found clinicians recognise their value in allowing care to be tailored to individuals. Patients also recognised this value but expressed concern regarding OM reducing time available to build an effective therapeutic relationship between clinician and patient[11].

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Previous work around haemophilia OM has focussed on long-term management and joint function. Despite use of prophylactic factor, bleeds regularly occur and can have a considerable short-term impact[2]. To ensure services best meet need, it is important to gauge opinions of PWH regarding service design and ensure OM used are acceptable and meaningful to them. The study aim was to understand perspectives of PWH on validated outcome measures (OM) and whether these measures capture changes relevant to them whilst recovering from an acute bleed episode.

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Material and Methods

This research was part of a program investigating physiotherapy for acute bleed management at a UK teaching hospital and Haemophilia Comprehensive Care Centre (CCC)[3,5]. Data were collected between January and March 2017.

Any PWH registered to the CCC who had attended physiotherapy for management of a bleed within the past 2 years was invited to participate in a workshop. Usual practice in the CCC is for physiotherapy review to occur as soon as possible after report of a bleed, as per UK Guidelines[2]; with most PWH seen by a physiotherapist within 48 hours. Exclusion criteria were; aged under 16 years or no reported acute bleed. Letters of invitation were sent to those eligible. A reminder letter was sent after 2 weeks. Those who indicated interest were contacted by phone to explain the research further and arrange an appointment. Written consent was requested from individuals on attendance.

The workshops were activity-based groups used to promote discussion around haemophilia OMs and were facilitated by EB and CMcC. Participants who were unable to attend a workshop were invited to a one-to-one interview, which were conducted by EB. Interview schedules (identical for workshops and interviews) were developed by the research team with input from a patient research partner and the hospital's Quality and Information Service (QIS). Interviews and workshops were audio-recorded and transcribed (by EB), with exception of one interview for which notes were taken immediately afterwards due to recording failure. The purpose of the qualitative study was to explore patients' experiences and understanding of OMs and common haemophilia symptoms. This was done by facilitating discussion around key symptom domains.

Prior to data collection, haemophilia OMs from the literature and a national survey of current physiotherapist practice[5] were grouped into key domains: for example function, pain, quality of life (table 1). To prompt discussion, participants were asked which of these key domains most impacted their lives during an acute bleed. They were also prompted to comment on which outcomes were important to them in seeking physiotherapy treatment during these episodes. Participants were encouraged to consider and discuss the different OM sets. Only OMs used in routine practice at the participating centre were familiar to patients (table 1). No domains were identified that had not been prepared in advance and some prepared domains were not discussed. Deductive thematic analysis was used as an appropriately pragmatic approach to synthesise data [12].

Ethical approval was gained from the Faculty of Health and Applied Sciences Research Ethics Committee at University of the West of England, Bristol and approval for the study was also provided by the Trust QIS.

Results

Thirty-seven PWH met the inclusion criteria. Eight male PWH participated, mean age 61 years, ranging between 39 and 71 (standard deviation 11.2). Seven participants had severe Haemophilia A and 1 had moderate type 2M Von Willebrands. Two workshops of two

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participants were run. A further four participants were interviewed on a one to one basis to fit with planned attendance to physiotherapy appointments. No response was received from 29 participants. In line with the approved protocol, no attempts were made to seek reasons for non-response.

OM specific results

The most common domains identified by participants were pain (raised by all 8 participants), range of movement (7 participants), general function (7 participants) and strength (5 participants). The domains are considered below with key quotes to represent the range of opinion.

Domains: General Function and Quality of life

In general, participants recognised the impact of an acute bleed on general function.

Once it goes and pain comes in, everything goes bad….you can't do anything, you are down. (participant 4: interview).

With longer measures particularly, participants were concerned with time being taken away from treatment and practicalities within a painful bleed episode.

If I come in with a bad bleed…the last thing I want to do is fill in…questionnaires….whilst I can understand needing to do it, it's a case of "just get on and treat it". (participant 3: interview)

- Haem-A-QOL (Haemophilia Quality of Life Questionnaire for Adults)[13]

Some participants recognised the wider impact of a bleed on overall well-being. Whilst participants felt it was important for a physiotherapist to acknowledge this, the Haem-A-QOL was felt to be too long for use in the acute bleed phase.

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… I…think…someone….treating…does need to think…what kind of impact is it having? But….it's easier to…talk…to someone….than fill…in a form. A…..physio should be saying, "are you a bit depressed"….because…..if you are feeling depressed, the last thing you want to do is fill in a form. I am feeling fed up and then you want me to fill in seven pages about how fed up I feel. (participant 8: workshop)

- FISH (functional independence score in haemophilia)[14]

The FISH was also viewed as too long for within a bleed episode.

if you are having a bleed you wouldn't want to do all of this. (Participant 4: interview)

- HEP-Test-Q (Haemophilia and Exercise Project Questionnaire)[15]

The format and brevity of the HEP-Test-Q was favourable. However, even within the haemophilia specific OMs, some participants felt measures asking about specific activities, may not capture changes relevant to them following a bleed.

Trouble is, if we fill in those questions, a lot of them are aren't relevant, it's like the [question about] shopping. So basically, I drive, my wife shops; I don't do any shopping normally. So it's not relevant. (participant 7: workshop)

- EQ5D-5L (Euroqol 5-Dimensions 5-Levels)[16]

The EQ5D was generally viewed positively as it enabled all participants to reference their normal, rather than asking about specific activities.

That is..useful as you are giving….definitions..you know "I have no problems doing my usual activities…you've got….reference points…. (participant 3: interview)

- HAL (haemophilia activities list)[17]

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Participants all generally liked shorter measures. However, despite being a longer measure, participants all felt the HAL would capture changes relevant to them.

I like this one. It shows you all the areas that might be affected by a bleed. It is more relevant to me because it gives you more ways to express how you feel…This shows you all the activities you can do during a bleed and after a bleed. (participant 4: interview)

- HAQ (Health Assessment Questionnaire)[18]

As a shorter measure the HAQ was liked, although some found the descriptors hard to relate to their activities.

The HAQ. You can just quickly answer this one and it is easy to understand, with a tick box, this one is easier to do. (participant 4: interview)

'Much difficulty' what does that mean? Participant 3: interview

Domain: Mood

Only one participant raised the issue of mood being impacted during a bleed. The mood section of the EQ5D was not deemed specific enough to capture changes.

This hasn't really got enough in it, although it is about self-care. (participant 6: interview)

The Beck Depression Inventory[19] while being *"interesting to try"* (participant 6:interview) was felt to be too in depth to capture changes occurring around an acute bleed episode.

If you are measuring mood…this might be the thing…just to see how I was. (participant 6: interview)

The mood section of the Short Form Survey (SF36)[20] was felt to be the best of the three to capture relevant changes in an acute bleed.

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This one (SF36) would capture it (meaningful change) more than the BDI. (participant 6: interview)

Domain: Pain

Acute bleed episodes were uniformly recognised as very painful. Physiotherapists often aim to record pain levels as a sign changes are occurring from their treatment. Participants however struggled with many common pain measures used in both research and practice.

It's very difficult if you are used to pain. … when we suffer bleeds we get pain all the time, it's something…we get used to well, I say we get used to it, it hurts like blazes and we want it to go away. (participant 7: workshop)

All participants struggled with the concept of measuring or rating pain, particularly as a number on the visual analogue scale for pain[21]:

(Sighs). These are impossible… pain is un-measurable. If anyone came up with a pain measure….they could plug you in and say, yes you really are in that amount of pain. (participant 6: interview)

They generally preferred Likert scale[22] measures.

I would find it easier to say it is moderate or severe pain than 4 or 6. (participant 8) .

However others found the Likert descriptions on the EQ5D-5L too limiting due to the condition variability.

I don't like the descriptions as I couldn't just say one. It varies .I would have to tick all of them even on one day. (participant 5: interview)

Participants generally avoided taking pain medication, regardless of pain levels hence this was deemed an inappropriate OM.

That would not have much benefit because I rarely take any form of analgesics. (participant 3: interview)

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Despite being a paediatric measure, the Wong Baker faces scale[23] was felt to be a good measure as it gave a visual reference point.

I'm a…visual person, so the smiley faces thing, whilst it appears quite childish, it actually works wonders for me, because…it's…providing a reference point to provide some…criteria. (participant 3: interview)

Domain: Strength and Range of Movement

Objective measurement of strength and range were viewed as valuable to give feedback on progress, if confidence was held in the physiotherapist measuring them. Participants valued the consistency of having the same physiotherapist.

[Physio] knows how to use them, and the fact…you know how to use them means…you've got a guide every time we come, so it's accurate. So, if you can see….we're doing better, obviously we are working and doing what you have asked us to […] then it works for me". (Participant 7: workshop)

As soon as (physio) measures it and says you're doing better, you feel ok. (participant 4: interview)

Domain: Swelling

Participants all felt, other than in severe cases, a tape measure would suffice for swelling measurement rather than a scan. As with range of movement and strength, this measure could provide helpful feedback about progress.

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"You are seeing face to face about how…swelling changes" (participant 4:interview).

Another participant recognised that swelling may not be a good measure of bleed resolution as it can be impacted by other issues.

"My legs swell up…at work. You should see my calf at the end of the day" (participant 5:interview)

Domain: Work

Only one participant was working at the point of data collection. They suggested time off work was not always reflective of recovery from an acute bleed due to wider issues regarding impacts of haemophilia.

Days off would not be a measure for me as my work is guided by not wanting to lose pay and also….if I am off I have to have a meeting when I go back and get told off. (participant 5:interview)

Participants views about the Canadian Occupational Performance Questionnaire [24] within an acute bleed were varied.

> *I prefer less open questions. I found this hard*. (participant 2: workshop)
>
> *[I prefer]….COPM; it is shorter* (participant 4: interview)

Discussion

Whilst acute bleeds are a self-limiting condition, episodes are recognised to contribute to development of joint damage, impacting on longer term function[10] . Limited research has

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focussed on OM for recovery from acute bleeds or more generally for haemophilia. Currently, no condition specific OMs exist for use within an acute bleed episode, despite UK guidelines recommending physiotherapy input within this period[2]. It is recognised that several measures discussed were not designed specifically for acute bleeds. In this study, PWH were asked to consider whether OMs currently used in practice and research reflect changes important to them during recovery from an acute bleed episode.

Generally, participants preferred condition specific measures of function to generic measures. Participants also expressed concern around how OMs might contribute to a changing culture in health care around data collection and reduce time with therapists. Literature on use of OMs in other long-term conditions reports similar findings, for example whilst people with diabetes recognised the value of OMs in clinical encounters, concerns were expressed about the potential negative impact on the clinician-patient relationship[11].

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Participants within this study valued measures that could reflect positive progress, for example joint range of movement. A common concern was however, raised regarding the time and effort required to complete measures. This was particularly a concern as acute bleeds can be extremely painful. Perhaps for this reason, participants indicated a preference for shorter measures. Potential burdens created by OMs are recognised in literature, although this research within haemophilia is specific to children[9]. Perspectives of PWH reported recommend simple, rapid measures are used routinely, and more detailed measures used in defined periods such as annual reviews[25].

All participants felt rating pain as a number was an abstract concept. There was concern regarding the perceived lack of standardisation a Visual Analogue Scale (a continuous single scale measure of pain intensity[21]) rating held between patients. Other recent research within haemophilia also found PWH preferred measures of pain with a functional component, for example relating pain levels to an impact on usual activities, rather than purely listing pain as a number[26]. Despite acute bleeds being recognised as significant episodes with wide-reaching effects, participants remained stoic, preferring to avoid taking medication despite potentially high pain levels. This finding may have differed with a younger cohort.

Limitations of this study include a lack of younger participants giving a narrower representation of functional ability and differing historical experiences of acute bleed treatment which could have impacted results. Lower recruitment of younger participants was most likely due to the limited times available for data collection as a result of clinical commitments and funding restrictions. Participants were encouraged to talk openly about their opinions but the location (physiotherapy department) and professional background of the researcher (physiotherapist) may have negatively impacted the participants' willingness to speak freely.

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Despite limited participant numbers and age ranges, this work adds to the existing evidence-base regarding PWH views on OM specifically within acute bleed episodes. No current OMs fully reflect what matters to PWH or the prognosis of recovery from acute bleeds. Further work is needed to review optimum uses of OM in acute bleed episodes, as well as exploring opinions of people with bleeding disorders on relevance of longer-term measures to their daily life. Future work would benefit from engaging a wider breadth of age ranges, functional abilities and experiences of acute bleed episodes, in addition to exploring use of person-centric measures and views of health professionals involved in acute bleed management.

Conclusion

Participants in this study found numerical scale ratings of pain abstract and prefer verbal or visual descriptors. Generally, participants preferred measures that were quick and easy to complete. Haemophilia specific OMs were identified as more relevant to capture function. The EQ5D-5L and HEP-Test-Q were favoured for their shorter length and ability to capture overall function promptly. Use of wording comparing ‘usual activities’ or ‘usual function’ were deemed more relevant than longer scales listing several specific activities that may not be relevant. All participants raised issues about use of OMs generally and felt that a skilled communicator can gain far more valuable information verbally. Participants valued the role of a knowledgeable and trusted physiotherapist when experiencing acute bleeds. The use of OMs can provide objective evidence of treatment effect for service providers. However, within acute bleed episodes therapists should consider carefully which measures are used to avoid additional burden at a challenging time.

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Implications

This work is part of a wider research programme investigating physiotherapy management of acute bleeds and ongoing joint pain. Findings will inform OMs used in future trials as well as guiding routine clinical practice for patients with an acute bleed. Work is ongoing to establish a core set of OMs to help standardise care across specialist haemophilia units throughout the UK. This work contributes a view from PWH on the ability of measures to capture changes relevant to them within acute bleed episodes. Future research with large samples representing a broader range of PWH is warranted. These results underscore the pivotal role played by physiotherapists not only in the management of bleeds but also in communicating their implications to PWH as part of ongoing management.

4. Acknowledgements

This work was funded by a Health Education England Integrated Clinical Academic Bridging Award. The authors have no competing interests.

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Appendix: Authorship

E. Bradshaw was the principal researcher on this paper. She ran the workshops with CMcC, conducted the interviews and was primary author of the manuscript.

C. McClellan was involved in the protocol development, review of the manuscript and ran the workshops with EB.

P. Whybrow was involved in the analysis of the results and development and review of the manuscript.

F. Cramp had academic oversight for the research work. She was involved in protocol development and development and review of the manuscript.

For Peer Review

Dear Prof Hermans and reviewing team,

Thank you again to you all for taking your time to read through and review the revisions to our paper. we appreciate the time taken to do this and for the comments.

We have addressed the comments as follows (*in italics)* :

Comments	*Response*
Reviewer: 1	
Page 6 line 25 - '...detrimental bleed impacts' ... this reads a bit strangely - consider rewording/reformatting sentence.	*Re-worded to 'detrimental impact of bleeds'*
Page 6 line 41 - 'commissioning guidelines' - this is a very particualr NHS term... consider other terminology for clarity ... eg mandated provider service specifications, payers, etc	*Thank you for raising this. Re-worded to 'service-provider guidelines mandate the use of'.*
Page 7 line 6/7 - 'despite use of ..... short term impact' ... I feel you know a ref here to qualify this statement.	*Added a reference to Hanley et al 2017 who discuss the impact of acute bleeds.*
Page 7 line 31 - 'reported to physiotherapy' ... could sound patriarchal .. consider 'attended' maybe?	*Changed to 'attended'.*
Page 7 line 33 - '... past 2 years was invited to participate' ... add in 'to a workshop' at the end here for clarity.	*'in a workshop' has been added.*
Page 7 line 42 - (n=37) is a result - move to results section	*Moved to the start of the results section 'Thirty-seven PWH met the inclusion criteria'.*
Page 7 line 56 - using the term focus group here now... stick to one description	*Thank you for highlighting, this has now been changed to 'workshop'.*
page 15 line 20/21 - 'All participants raised issues.... valuable information verbally' I think this sentence would be better suited in the conclusion section - with added informtioan relatig to the high importance PWH placed on the Physio guiding and advising in the acute phase.	*Sentence moved into conclusion with brief discussion point as suggested.*
I feel you could emphasies more the role of the physio and its value in communcation/care over that of the OM's here	*We feel that this has now been addressed through the above change.*

Thank you for your comments which have been very helpful to guiding our revisions.

Kind regards,

Elizabeth Bradshaw

For Peer Review