Treatment of hemophilia

Citation for published version (APA):

von der Lippe, C., Frich, J., Harris, A., & Solbrække, K. (2017). Treatment of hemophilia: A qualitative study of mothers' perspectives. *Pediatric Blood & Cancer*, 64(1), 121 – 127.
<https://doi.org/10.1002/pbc.26167>

Document status and date:

Published: 01/01/2017

DOI:

[10.1002/pbc.26167](https://doi.org/10.1002/pbc.26167)

Document Version:

Publisher's PDF, also known as Version of record

Document license:

Taverne

Please check the document version of this publication:

- A submitted manuscript is the version of the article upon submission and before peer-review. There can be important differences between the submitted version and the official published version of record. People interested in the research are advised to contact the author for the final version of the publication, or visit the DOI to the publisher's website.
- The final author version and the galley proof are versions of the publication after peer review.
- The final published version features the final layout of the paper including the volume, issue and page numbers.

[Link to publication](#)

General rights

Copyright and moral rights for the publications made accessible in the public portal are retained by the authors and/or other copyright owners and it is a condition of accessing publications that users recognise and abide by the legal requirements associated with these rights.

- Users may download and print one copy of any publication from the public portal for the purpose of private study or research.
- You may not further distribute the material or use it for any profit-making activity or commercial gain
- You may freely distribute the URL identifying the publication in the public portal.

If the publication is distributed under the terms of Article 25fa of the Dutch Copyright Act, indicated by the "Taverne" license above, please follow below link for the End User Agreement:

www.umlib.nl/taverne-license

Take down policy

If you believe that this document breaches copyright please contact us at:

repository@maastrichtuniversity.nl

providing details and we will investigate your claim.

RESEARCH ARTICLE

Treatment of hemophilia: A qualitative study of mothers' perspectives

Charlotte von der Lippe¹ | Jan C. Frich² | Anna Harris³ | Kari Nyheim Solbrække²

¹Centre for Rare Disorders, Oslo University Hospital HF, Rikshospitalet, Oslo, Norway

²Institute of Health and Society, University of Oslo, Oslo, Norway

³Department of Technology and Society Studies, Maastricht University, Maastricht, The Netherlands

Correspondence

Charlotte von der Lippe, MD, Centre for Rare Disorders, Oslo University Hospital HF, Rikshospitalet, P.O. Box 4950 Nydalen, 0424 Oslo, Norway.
 Email: uxhelc@ous-hf.no

Abstract

Background: In Norway, boys with hemophilia usually begin treatment after their first bleeding episode. Boys with severe hemophilia usually start prophylactic treatment around 18–24 months. Health professionals administer factor concentrate initially, but when boys are around 4 years old most parents start treating their children at home. There is a lack of research on how parents, and especially how carrier mothers, experience the medical treatment for their sons' hemophilia. Our aim was to investigate how carrier mothers experience this treatment in the hospital setting and at home.

Methods: In this qualitative study, we interviewed 16 mothers of boys or men with hemophilia A or B. Data were collected via semistructured interviews and analyzed using an inductive thematic analytical approach.

Results: Mothers experienced both practical and emotional challenges in relation to their sons' treatment, and repeated venipuncture was especially difficult emotionally. Parents preferred home treatment to hospital treatment because it was less time-consuming, less disruptive to family life, and provided a greater sense of control. Encountering healthcare professionals who were unfamiliar with hemophilia was a second major stress factor, especially when parents felt that health professionals lacked competence and were unwilling to seek advice.

Conclusion: While home treatment for hemophilia enables freedom, flexibility, and autonomy for the boys as well as for the family, mothers may experience treatment of hemophilia as a burden. Health professionals should provide tailored practical and emotional support to parents by probing into their experiences with treating their sons' hemophilia.

KEYWORDS

experiences, hemophilia A, hemophilia B, mothers, qualitative, treatment

ABBREVIATIONS: BWH, boys with hemophilia; IVAD, intravenous access device

1 | INTRODUCTION

Hemophilia A and hemophilia B are rare X-linked disorders caused by mutations in the factor VIII (*FVIII*) and factor IX (*FIX*) genes, respectively. The deficiency in factor VIII or IX clotting activity results in abnormally prolonged bleeding. Treatment consists of intravenous administration of an external coagulation factor prophylactically and/or episodically when bleeding occurs. A healthcare professional at a hospital or a child's guardian at home administers treatment by peripheral venipuncture or by using an intravenous access device (IVAD)/central venous catheter.

Prophylaxis for children with severe hemophilia consists of infusion of a factor concentrate two or three times a week in order to keep blood levels of factor VIII or IX high enough to prevent bleeding. On-demand therapy involves infusion of a factor concentrate immediately after bleeding starts in an effort to prevent joint or muscle damage. Treatment regimens vary between countries.^{1–3} Prophylactic treatment is generally recommended because it reduces the number of bleeding episodes and hospital visits and results in less joint damage and need of orthopedic surgery, increased participation in sports and activities, and less time lost from work or school.^{4–8} In Norway, medical personnel usually administer factor concentrate in a hospital

setting in children younger than 4 years of age, which we refer to as hospital treatment in this paper.

Parents learn to infuse factor concentrate when the child is about 4 years old. In this paper, we refer to treatment administered by parents or guardians as home treatment. After successfully completing a 5-day course at the national competence center for hemophilia (Centre for Rare Disorders, Oslo University Hospital) parents/guardians are certified to administer treatment at home. During this course, they learn how to identify and treat a bleed, order and store factor concentrate, and perform venipuncture. At home, parents may administer prophylaxis and on-demand treatment for uncomplicated bleeds. For complicated bleeds, requiring more than two doses of factor concentrate, or serious injuries such as head trauma, parents must contact a hospital. Parents and health professionals can contact the competence center for advice from 8 a.m. to 3:45 p.m., 5 days a week. For "out-of-hours" advice, parents contact their local hospital and health professionals can contact the national hospital, Rikshospitalet, Oslo, Norway.

Although parentally administered prophylaxis gives the family more flexibility, the treatment is not without worries or challenges. Even when recommended treatment regimens are in place, hemophilia affects children's education and leisure activities.⁹ Parents find it stressful to have a child with hemophilia,¹⁰ and experience uncertainty relating to home treatment and parenting.¹¹ Prophylactic home treatment may be time consuming or challenging,¹² and for the nonmedically trained, learning how to administer peripheral injections usually takes time.¹³

Caregivers of children with hemophilia report reduced health-related quality of life compared with caregivers of healthy children.¹⁴ Women are often the primary caretakers,¹⁵ and mothers of boys with hemophilia (BWH) are usually more involved in daily childcare than fathers and are believed to have higher risk for psychosocial problems.¹⁶ The mother is often a carrier of the mutation that causes her son's hemophilia¹⁷ and may experience shock and sadness when a son has been diagnosed with hemophilia.¹⁸ One study suggests that mothers are more depressed and anxious because of their children's disease than fathers.¹⁹ A more recent study did not detect increased anxiety and depression in mothers of BWH compared with a reference group.²⁰

Research on how parents, and especially mothers, experience the medical treatment of hemophilia is limited. This study investigates how carrier mothers experience the medical treatment of their sons' hemophilia in the hospital setting and at home. Our aim is to offer qualitative insights that can facilitate better support and guidance from healthcare professionals who work with BWH and their mothers.

2 | METHODS

2.1 | Design

We used an explorative qualitative research approach with in-depth interviews to gather data. This present study is part of a larger study on the experiences of being a carrier of hemophilia with an affected son.

2.2 | Recruitment

A nurse at the national competence center for hemophilia, Centre for Rare Disorders, Oslo University Hospital, Oslo, Norway, invited women known to be carriers of hemophilia A or B to participate. In order to be included in the study, women had to be carriers of the disorder, have an affected child, and understand and speak Norwegian or English. We aimed to recruit 10 women. In an attempt to achieve diversity in the sample, the nurse selected 11 women of different ages from different parts of Norway. She contacted seven women by phone and approached four women about the study in conjunction with their sons' annual follow-up. All women received written information about the study and all accepted the invitation to participate. Five additional women contacted the first author directly after hearing about the project from the Norwegian Haemophilia Society. All 16 women, ethnic Norwegian aged 27–72 years (mean = 42), were included in the study. Table 1 summarizes clinical and demographic characteristics of the participants.

The first author contacted the participants by phone after they agreed to participate in the study. All participants gave written consent and chose when and where they wanted their interview to take place. We obtained ethical approval for the study from the Regional Committee for Medical Research Ethics (Health Region South-East, reference number: 2012/1617).

2.3 | Data collection

The first author conducted all 16 semistructured interviews at a university hospital or at participants' homes between February 2013 and March 2014. The interviewer, a clinical geneticist, was explicit about her role in the project as a researcher. Average duration of the digitally recorded interviews was 70 min (range 40–100 min). All interviews were transcribed in verbatim. The interview guide included open-ended questions, some of which are listed as follows: Tell me about how, and when, you learned you were a carrier, and what you feel about being a carrier? How do you feel about having an affected child? How do you communicate about having a genetic disorder in your family? How does your son's disorder affect your and your family's daily life? What are your experiences with the treatment and the healthcare system?

The semistructured interview with open-ended questions allowed the participants to expand upon aspects of their experiences that were important to them.

2.4 | Data analysis

An inductive thematic analytical approach was used to analyze the data.²¹ The first author listened to the interviews, read the transcripts several times, and summarized key aspects of each mother's experiences in writing. The second and fourth authors also read the transcripts. The third author participated in the subsequent analysis and discussions. The first author coded the entire material line-by-line in tandem with the last author using the software program HyperRESEARCH.²² All authors discussed the coding and

TABLE 1 Participants' clinical and demographic characteristics

Participant (Pseudonym)	Age	Carrier of hemophilia A or B	Number of affected sons	Hemophilia known in the family prior to having a son with the diagnosis
Kari	35	Hemophilia A, severe	1	Yes
Lisa	32	Hemophilia A, severe	1	Yes
Anna	41	Hemophilia A, severe	1	Yes
Heidi	27	Hemophilia A, severe	1	No
Lisbeth	39	Hemophilia A, severe	2	No
Marianne	35	Hemophilia A, severe	1	Yes
Ingrid	27	Hemophilia B, moderate	1	Yes
Thea	30	Hemophilia A, severe	(1, early termination)	Yes
Lena	40	Hemophilia B, moderate	2	No
Tanja	65	Hemophilia A, severe	3	Yes (distant)
Wilma	55	Hemophilia A, severe	1	Yes
Mette	72	Hemophilia A, severe	2	No
Victoria	34	Hemophilia B, moderate	2	No
Jane	56	Hemophilia A, severe	1	Yes
Therese	28	Hemophilia A, severe	1	Yes
Astrid	49	Hemophilia A, severe	1	Yes

grouped codes by main themes. Data analysis in this study focused on the mothers' experiences with the healthcare system and treatment. The Norwegian authors translated the quotes from the interviews and allocated pseudonyms.

3 | RESULTS

We present the results from this study exploring the three main themes of the mothers' experiences related to medical treatment: challenges with treatment; advantages with home treatment; and trust and mistrust in doctors.

3.1 | Challenges with treatment

The mothers described several situations when treatment was challenging: When the doctor or parent had trouble accessing a vein, when the procedure was painful for the child, when the child was uncooperative or when he complained that it was unfair that he needed to be treated, and when siblings interrupted during treatment. Going on vacation could also be a challenge and the mothers preferred to travel to places and countries where they knew access to proper treatment in a hospital was available, as Victoria stated:

For us it means we do not go on a jungle safari, we do not travel to remote places but to places close to a good hospital.

Although the mothers discussed practical challenges, it was the emotional challenge of caring for their sons that was emphasized. Participants described the emotional toll of venipuncture, either performed in a hospital setting or at home as an especially significant burden. Tanja remembered vividly when her oldest son was younger and

received treatment at a hospital. Hearing him scream was very hard for her, and it made her consider future reproductive choices:

He screamed so much [when he had treatment], I thought I will never do this to another child. ... You have to make yourself tougher than you are.

Venipuncture and failure to insert the cannula was also challenging for parents in the home setting, as Therese explained:

It was inconceivably difficult. It was a nightmare. It was a battle. It was screaming and yelling. I got angry and he got angry, and I got upset and he got upset. It was extremely tough. It has been so hard!! So hard!! To hold him tight to give him the medication, then you miss [inserting the cannula], then he cries, then I cry!

Most mothers had a partner who could administer treatment, and almost all couples shared the responsibility—or "that burden," as Lisa said. Therese, a single mother, described the difficulties of being a single parent with sole responsibility for managing her son's condition:

I can never really shut the door and close the curtains and turn off my phone, because I have no one to share the responsibility with. ... I don't know how it feels to shut off my phone, it is always on. Even if I am in the shower, the phone is close by, in case someone calls about my son.

One group of mothers believed they treated their son with hemophilia and their other child/children the same, while others felt they were overprotective of the son with hemophilia.

Practically speaking, more time has been spent on him [the child with hemophilia]. It really has, and that hurts

sometimes. I have felt bad for my other son, for not spending more time on him. (Astrid)

Similar challenges were described by both older mothers and younger mothers.

3.2 | Advantages with home treatment

All mothers considered administration of the factor concentrate at home to be more convenient than receiving treatment in a hospital setting. They described the latter as very time consuming due to travel and waiting. Meeting new health professionals who were unfamiliar with hemophilia was frustrating and could give rise to concerns about whether the child would receive appropriate treatment. Administering treatment at home made life easier, as reflected by Lisbeth:

After we learned how to do it [administer treatment] ourselves, we have noticed life has become easier for all of us. We don't have to go to a hospital, where they may not be familiar with the boys, and I have to explain things. It takes more time [at the hospital].

Although treatment is free at point of delivery in Norway, participants recounted financial issues such as travel expenses and loss of income. Several families found living close to a university hospital an advantage in case of emergencies and because they were more likely to encounter doctors and nurses familiar with hemophilia in this setting. Home treatment was less time consuming, allowed the child to be perceived as "normal" because he was able to avoid trips to the hospital, and provided valuable freedom and flexibility for the family. Anna, one of the mothers, said

First of all, it [treatment] takes a lot of time, and secondly one is more ill if one has to be absent from kindergarten every time one gets treatment [at the hospital], than if he can have the treatment at home and be like everyone else. So, it is absolutely an advantage to have home treatment.

3.3 | Trust and mistrust in doctors

Participants spoke about encounters with doctors who had little knowledge of hemophilia. Lacking competence became an issue especially in acute situations requiring hospital visits. The mothers accepted that not all doctors had extensive knowledge of a rare condition such as hemophilia, as Heidi expressed: "They [doctors] are not computers, they are humans, right." It was not the lack of competence or knowledge per se that mothers found difficult, but what they perceived as the physicians' attitude; their unwillingness to admit their shortcomings and to seek information and advice. Many mothers said that it was most difficult when the doctors disguised their lack of knowledge. Astrid told how her father, who had hemophilia himself, had advised her to never trust doctors or the healthcare system and to always have 100% control. Mothers tried hard to not "step on anybody's [doctors'] toes," but at the same time felt they knew more about hemophilia and its treatment than many of the physicians they

met. Knowing how to interact with doctors wisely and in a respectful way, while still being critical, was perceived as an important competence. Several mothers felt they were a burden to the healthcare system and that doctors viewed them as "hysterical mothers." Negative experiences had a damaging effect on the mothers' relationships with doctors. As Lena described:

We try to keep calm and take care of things by ourselves, because we do not want any contact with them [doctors at the local hospital], because it is never a positive experience.

As an example, Lena recounted an episode when her son had a stomach infection and rectal bleeding. The parents were worried and brought the boy, not yet on prophylactic treatment, to the hospital. They were expecting treatment with factor concentrate to reduce the bleeding. The junior doctor treating their son refused to administer factor on the grounds that it could camouflage the infection; reasoning the parents did not understand. The father consulted the national hemophilia competence center that confirmed that it was safe to give factor concentrate. The doctor did not change his mind, leaving the parents feeling that they had to deal with the situation on their own.

So, we asked permission to go home for the night, and then we went home and gave him medication [factor concentrate]. Of course the bleeding was less the next day when we came back [to the hospital], and they were happy [laughs] because the bleeding had stopped by itself. They thought so, and we did not tell [about the medication]. And this is what is wrong, because we can't tell. We feel like criminals going behind their backs doing things we should not do. It makes me very unsure, I feel they [her boys] do not get the right treatment there [at the local hospital], and that makes us feel unsafe.

Some participants explained how having a child with hemophilia had changed their views of doctors and the healthcare system. For example, Heidi said:

It is a big change [to be the expert on your child's disease], because I have been brought up to believe that the doctor always knows everything, and then maybe now I know more than the doctor about certain things. It has been a change to be critical of the healthcare system because they don't know everything. It is not always the doctor who knows [what is right].

As a way of coping with her lack of trust, Heidi asked new doctors questions she already knew the answers to. She became worried when the doctor gave her a wrong answer and she would only trust a doctor if she/he knew the right answer—or if she/he admitted to not knowing but would try to find the answer.

It was important for all the mothers to be able to engage with healthcare professionals who were familiar with hemophilia and could provide advice and support.

4 | DISCUSSION

Our findings revealed that carrier mothers experience challenges with treatment both in the home environment and in contact with the healthcare system. In the home setting, parents must learn to identify bleeds and administer factor treatment intravenously^{23,24} and gain confidence in their expertise and skills.¹¹ Inserting a cannula in your child can, no matter how good ones skills are, be a stressful experience. Treating your child with advanced medical procedures at home may give rise to a conflict between the role as a parent and as a medical caregiver. Parents of sick children experience both acute and prolonged medical traumatic stress,^{25,26} which may evoke difficult feelings.

Taking care of a child who needs specialized medical treatment alters the nature of parenting.²⁷ A parent usually has the role of comforting the child and protecting him or her from anything hurtful. Performing venipuncture produces pain, and the parent enters the role of a medical profession by undertaking the task. The mothers may feel hypervigilance, as expressed by Therese, who could never turn off her phone. Parents of children with life-threatening conditions report hypervigilance,²⁸ and in some cases, mothers may be overprotective for their child with hemophilia.²⁹

A process of normalization³⁰ has been described previously in families with children with hemophilia.³¹ A normalization process means reconstructing family life and accepting the new situation as normal,³² as the mothers in this study who accepted they could not travel “all over the world.” Mothers viewed the ability to administer treatment “secretly” at home as allowing the child to be “normal.” Hemophilia is an invisible disorder in many respects, although being absent from daily activities like kindergarten and school, and going to the hospital, is visible and might be stigmatizing.³³ Absence from kindergarten and school for visits to the hospital make people around the boy with hemophilia aware that he has a disorder. BWH may also feel socially stigmatized because of limitations in sports and other activities.³⁴ An experience of being different from peers may increase feelings of being stigmatized.³⁵ Barlow et al.³⁶ showed public misconceptions regarding bleeding disorders, perhaps linked to the history of patients with hemophilia being infected with HIV and hepatitis C. For the carrier mothers, who may feel guilty for having passed on the mutated gene,³⁷ stigmatization and misconceptions, perceived or real, may be an extra burden. This may influence how mothers experience the emotional challenges with treatment, as described by Tanja, who had thoughts about reproductive choices hearing her son scream.

Another important finding in this study is the mothers’ descriptions of doctors pretending to know, or not admitting to their lack of knowledge, about hemophilia. Both younger and older mothers and carriers of severe and moderate hemophilia reported feelings of mistrust. Such experiences had a negative effect on mother–doctor relationships and led mothers to avoid contact with hospitals. This finding resonates with other studies showing that health professionals have a lack of knowledge about rare bleeding disorders^{36,38} and patients with bleeding disorders experience difficulties in communicating with healthcare workers, resulting in delayed treatment.³⁸ Conversely, parents of children with rare disorders often have expert knowledge of the disorder,³⁹

and when parents are certified to do hemophilia treatment at home, they are expert caregivers.⁴⁰ The parent’s role as an expert caregiver is not easy for either the parent or the health professional in a hospital setting.⁴¹

This type of tension also points toward the phenomenon of shared decision-making between clinicians and patients, which is a dynamic process wherein both parties must try to understand each other’s needs and wishes in a context of a relationship built on trust and respect.⁴² In an acute situation at a hospital, it is difficult to develop a trustful relationship. Doctors may recognize lay expertise⁴³ and may not acknowledge the certification parents of BWH have. When parents try to communicate their expert knowledge to physicians but feel mistrusted, the parent–physician relationship deteriorates further⁴⁴ and leads to avoidance of hospitals. The tragic history from the 1980s where viral infected blood products were administered to persons with hemophilia⁴⁵ may for some mothers influence their trust in doctors. Mistrust can result in delayed treatment and extensive bleeding with severe consequences for the boy.

5 | LIMITATIONS

Currently, there is no registry of carriers of hemophilia in Norway. We invited women of different ages and from different parts of the country to participate, and we tried to do this randomly. We also included five women who contacted the first author directly. Our recruitment procedure resulted in a selection of participants who may not be representative for all carrier mothers. Another limitation is that we did not interview doctors, or other health professionals, and do not know how they experience the treatment of BWH. The intent of this qualitative study was however to understand more about mothers’ perceptions of the treatment of their sons. For further research, it would be interesting to explore how doctors experience the relationship with the carrier mothers regarding the treatment of the BWH.

6 | CONCLUSION

While home treatment for hemophilia promotes freedom, flexibility, and autonomy for the boys and the family, mothers may experience treatment of hemophilia as a burden. Health professionals can provide tailored practical and emotional support to parents through probing about mothers’ experiences treating their sons’ hemophilia.

ACKNOWLEDGMENTS

We thank the participants for sharing their insight and experiences with us. We thank nurse Siri Grønhaug from Centre for Rare Disorders, Oslo University Hospital, for recruiting the participants and for information about the certification program for home treatment in Norway, pediatrician Heidi Glosli for information about treatment of BWH in Norway and medical geneticist Trine Prescott for valuable discussions and comments on the manuscript.

CONFLICT OF INTEREST

All authors declare that there is no conflict of interest.

REFERENCES

- Berntorp E, Shapiro AD. Modern haemophilia care. *Lancet*. 2012;379(9824):1447–1456.
- Ljung R. Paediatric care of the child with haemophilia. *Haemophilia*. 2002;8(3):178–182.
- Bolton-Maggs PH, Pasi KJ. Haemophilias A and B. *Lancet*. 2003;361(9371):1801–1809.
- Hoots WK, Nugent DJ. Evidence for the benefits of prophylaxis in the management of hemophilia A. *Thromb Haemost*. 2006;96(4):433–440.
- Steen Carlsson K, Hojgard S, Glomstein A, et al. On-demand vs. prophylactic treatment for severe haemophilia in Norway and Sweden: Differences in treatment characteristics and outcome. *Haemophilia*. 2003;9(5):555–566.
- Teitel J, Barnard D, Israels S, Lillicrap D, Poon MC, Sek J. Home management of haemophilia. *Haemophilia*. 2004;10(2):118–133.
- Yee TT, Beeton K, Griffioen A, et al. Experience of prophylaxis treatment in children with severe haemophilia. *Haemophilia*. 2002;8(2):76–82.
- Ljung R. Hemophilia and prophylaxis. *Pediatr Blood Cancer*. 2013;60(Suppl 1):S23–S26.
- Holstein K, von Mackensen S, Bokemeyer C, Langer F. The impact of social factors on outcomes in patients with bleeding disorders. *Haemophilia*. 2016;22:46–53.
- Torres-Ortuno A, Cuesta-Barriuso R, Nieto-Munuera J. Parents of children with haemophilia at an early age: Assessment of perceived stress and family functioning. *Haemophilia*. 2014;20(6):756–762.
- Beeton K, Neal D, Watson T, Lee CA. Parents of children with haemophilia—A transforming experience. *Haemophilia*. 2007;13(5):570–579.
- Hacker MR, Geraghty S, Manco-Johnson M. Barriers to compliance with prophylaxis therapy in haemophilia. *Haemophilia*. 2001;7(4):392–396.
- Petrini P. Identifying and overcoming barriers to prophylaxis in the management of haemophilia. *Haemophilia*. 2007;13(Suppl 2):16–22.
- Lindvall K, von Mackensen S, Elmstahl S, et al. Increased burden on caregivers of having a child with haemophilia complicated by inhibitors. *Pediatr Blood Cancer*. 2014;61(4):706–711.
- Navaie-Waliser M, Spriggs A, Feldman PH. Informal caregiving: Differential experiences by gender. *Med Care*. 2002;40(12):1249–1259.
- Wiedebusch S, Pollmann H, Siegmund B, Muthny FA. Quality of life, psychosocial strains and coping in parents of children with haemophilia. *Haemophilia*. 2008;14(5):1014–1022.
- Konkle BA, Josephson NC, Nakaya Fletcher S. Hemophilia A. In: Pagon RA, Adam MP, Ardinger HH, et al., eds. *GeneReviews*® [Internet]. Seattle, WA: University of Washington, Seattle; 1993–2016. Sept. 21, 2000 [Updated June 5, 2014]. <http://www.ncbi.nlm.nih.gov/books/NBK1404/>
- Myrin-Westesson L, Baghaei F, Friberg F. The experience of being a female carrier of haemophilia and the mother of a haemophilic child. *Haemophilia*. 2013;19(2):219–224.
- Saviolo-Negrin N, Cristante F, Zanon E, Canclini M, Stocco D, Girolami A. Psychological aspects and coping of parents with a haemophilic child: A quantitative approach. *Haemophilia*. 1999;5(1):63–68.
- Limperg PF, Haverman L, Peters M, Grootenhuis MA. Psychosocial functioning of mothers of boys with haemophilia. *Haemophilia*. 2016;22:e57–e60. doi:10.1111/hae.1283157-60
- Braun V, Clarke V. Using thematic analysis in psychology. *Qual Res Psychol*. 2006;3(2):77–101.
- Hesse-Biber S, Dupuis P, Kinder TS. HyperRESEARCH: A computer program for the analysis of qualitative data with an emphasis on hypothesis testing and multimedia analysis. *Qual Sociol*. 1991;14(4):289–306.
- Furmedge J, Lima S, Monagle P, Barnes C, Newall F. 'I don't want to hurt him.' Parents' experiences of learning to administer clotting factor to their child. *Haemophilia*. 2013;19(2):206–11.
- Shaw D, Riley GA. The impact on parents of developments in the care of children with bleeding disorders. *Haemophilia*. 2008;14(1):65–67.
- Woolf C, Muscara F, Anderson VA, McCarthy MC. Early traumatic stress responses in parents following a serious illness in their child: A systematic review. *J Clin Psychol Med Sett*. 2016;23(1):53–66.
- Landolt MA, Ystrom E, Sennhauser FH, Gnehm HE, Vollrath ME. The mutual prospective influence of child and parental post-traumatic stress symptoms in pediatric patients. *J Child Psychol Psychiatry*. 2012;53(7):767–774.
- Kirk S, Glendinning C, Callery P. Parent or nurse? The experience of being the parent of a technology-dependent child. *J Adv Nurs*. 2005;51(5):456–464.
- Heller KS, Solomon MZ. Continuity of care and caring: What matters to parents of children with life-threatening conditions. *J Pediatr Nurs*. 2005;20(5):335–346.
- Abali O, Zulfikar OB, Karakoc Demirkaya S, Ayaydin H, Kircelli F, Duman M. An examination of the symptoms of anxiety and parental attitude in children with hemophilia. *Turk J Med Sci*. 2014;44(6):1087–1090.
- May C, Finch T. Implementing, embedding, and integrating practices: An outline of normalization process theory. *Sociology*. 2009;43(3):535–554.
- Emiliani F, Bertocchi S, Poti S, Palareti L. Process of normalization in families with children affected by hemophilia. *Qual Health Res*. 2011;21(12):1667–1678.
- Deatrick JA, Knafel KA, Murphy-Moore C. Clarifying the concept of normalization. *Image J Nurs Sch*. 1999;31(3):209–214.
- Goffman E. *Stigma: Notes on the Management of Spoiled Identity*. 1963 ed. New York: Simon & Schuster, Inc.;1986.
- Williams KA, Chapman MV. Social challenges for children with hemophilia: Child and parent perspectives. *Soc Work Health Care*. 2011;50(3):199–214.
- Engebretson J. Understanding stigma in chronic health conditions: Implications for nursing. *J Am Assoc Nurse Pract*. 2013;25(10):545–550.
- Barlow JH, Stapley J, Ellard DR, Gilchrist M. Information and self-management needs of people living with bleeding disorders: a survey. *Haemophilia*. 2007;13(3):264–270.
- Featherstone K, Atkinson PA, Bharadwaj A, Clarke AJ. *Risky relations: family, kinship and the new genetics*. Oxford: Berg 2006.
- Barlow JH, Stapley J, Ellard DR. Living with haemophilia and von Willebrand's: A descriptive qualitative study. *Patient Educ Counsel*. 2007;68(3):235–242.
- Skinner D, Schaffer R. Families and genetic diagnoses in the genomic and internet age. *Infants Young Child*. 2006;19(1):16–24.

40. Sadler E, McKeivitt C. 'Expert carers': An emergent normative model of the caregiver. *Soc Theory Health*. 2013;11(1):40–58.
41. Allen D. Negotiating the role of expert carers on an adult hospital ward. *Sociol Health Illn*. 2000;22(2):149–171.
42. Lown BA, Clark WD, Hanson JL. Mutual influence in shared decision making: A collaborative study of patients and physicians. *Health Expect*. 2009;12(2):160–174.
43. Prior L. Belief, knowledge and expertise: The emergence of the lay expert in medical sociology. *Sociol Health Illn*. 2003;25:41–57.
44. Nuutila L, Salanterä S. Children with a long-term illness: Parents' experiences of care. *J Pediatr Nurs*. 2006;21(2):153–160.
45. Evatt BL. The tragic history of AIDS in the hemophilia population, 1982–1984. *J Thromb Haemost*. 2006;4(11):2295–2301.