

**Becoming and being mother to a boy with  
severe haemophilia**

**by**

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## **STUDENT DECLARATION**

### **Concurrent registration for two or more academic awards**

I declare that while registered as a candidate for the research degree I have not been a registered candidate or enrolled student for another award of the University, or other academic or professional institution.

## **ABSTRACT**

### **Background**

Haemophilia is a recessive genetic disorder which manifests symptoms in males but is carried by females. The disordered coagulation associated with severe haemophilia causes atraumatic joint and muscle bleeds and prolonged mucosal bleeding. These acute bleeding episodes frequently occur without any obvious injury and cause severe pain which is only relieved by administering intravenous treatment accompanied by rest, ice, compression and elevation. The management and treatment of haemophilia has evolved over the past fifty years from a dependence on lengthy hospital admissions to community based care with parents, usually mothers, administering bolus intravenous infusions to their boys at home. A review of the literature revealed that, despite being essential contributors to their sons' care, there was a paucity of literature relating to mothers experiences of their sons disorder.

### **Research Aim**

To gain a richer understanding of what it is to become and be mother to a boy with severe haemophilia.

### **Methodology and Methods**

This qualitative study was informed by hermeneutic phenomenology. Data were collected from mothers of boys with severe haemophilia (n=5) using unstructured interviews. Data were interpreted and given meaning through immersion in the interview transcripts, coding and the development of themes.

### **Findings and Discussion**

The mothers in this study discussed how, at times, they felt being responsible for giving their boys treatment burdensome and isolating, and that this was an immense added responsibility for them as mothers. This sense of burden, isolation and responsibility could be lessened by Haemophilia Teams addressing the difficulties of out of hours' services; developing a mother to mother support network, and by an expectation that more than one family member should be able to administer treatment in the home. The mothers also talked of how their lives were in some ways enriched by their sons' haemophilia as they accessed previously undiscovered strengths and skills and discovered a new sense of normal. All of the mothers had moved from a position of vulnerability to independence, although they still experienced ongoing concerns around what limitations to place on their sons' activities, disclosure of the diagnosis to outsiders and imagining their boys' futures. Core findings from this study resonate with those from other studies about mothers' experiences.

### **Conclusion**

In conclusion this study shows that mothers develop skills and knowledge that enable them to manage their boys' haemophilia independently. Home based services are clearly beneficial in promoting and facilitating independence but Haemophilia Teams need to address the perceived negative aspects of haemophilia care and support mothers as their reality changes to encompass that of becoming and being mother to a boy with severe haemophilia.

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## **CHAPTER 1**

### **INTRODUCTION AND LITERATURE REVIEW**

#### **Introduction**

The origins of this thesis lie in my observations accrued through my work as a nurse specialist in a Haemophilia Comprehensive Care Centre (CCC) with families affected by haemophilia. In particular it arose from observing how mothers move from a period of vulnerability and dependence on hospital services at the time their sons first present with unexplained symptoms through to an independence that would have been hard for them to imagine on the day of their sons' diagnosis with severe haemophilia. I have observed how this journey to independence is convoluted and for some mothers littered with obstacles. This research is driven by my curiosity and desire to gain a deeper understanding of these mothers' felt emotions and experiences and the meanings that lie therein, including what sustains and thwarts them as they become mother to a boy with severe haemophilia.

This thesis comprises five chapters. The first chapter introduces the origins of the thesis, provides an introduction to the clinical condition of haemophilia, a historical context to its management and treatment followed by a review of the literature relating to mothers' experiences of haemophilia. Chapter two discusses the methodological influences on this qualitative study and goes on to describe the method used to gather the data and its analysis. Chapter three, the findings chapter, uses the mothers' rich data to illustrate my interpretations of the participants' experiences. The fourth chapter discusses my interpretation of the mothers' meaning of what it is to become and be mother to a boy with haemophilia. The concluding chapter provides a summary of my findings; provides



recommendations and recognises the limitations of the study before giving an explanation of how the findings of this research project will be disseminated.

## **Overview of Haemophilia and the History of its Treatment**

Haemophilia is a sex linked recessive genetic disorder which manifests its symptoms in males but is carried by females (Appendix 1: Diagram of inheritance pattern). Disordered coagulation does not cause more rapid bleeding but haemostasis takes longer to achieve. Haemophilia A, the absence or reduction of clotting factor VIII (FVIII), is the most common form of the disorder affecting 1:5000 live births; haemophilia B, factor IX (FIX) deficiency, 1:25,000, is the second most frequently occurring (Haemophilia Alliance, 2006). In two thirds of cases there is a family history of an affected male relative, in the others there is no known history of the disorder. Presentation, similar in both forms of the condition, may be mild, moderate or severe; the phenotype runs true within families. In the UK, 1859 people (665 under 18 years of age) have severe haemophilia A or B (UKHCDO, 2011). Along with overt symptoms of disordered coagulation such as atraumatic bruising and prolonged mucosal bleeding, the more significant problems in severe haemophilia are caused by bleeding into muscles and joints. These acute bleeding episodes frequently occur without any obvious injury having been sustained; they cause severe pain, relieved only with the administration of bolus intravenous FVIII/FIX accompanied by rest, ice, compression and elevation (RICE) of the affected area. Recurrent muscle and joint bleeds lead to muscle wasting, synovial hypertrophy and haem-arthrosis which eventually cause irreversible haemophilic arthropathy with associated chronic pain and disability (Melchiorre et al., 2011).

Treatment for haemophilia evolved during the second half of the twentieth century, from cryoprecipitate in 1964, through human derived FVIII/FIX in 1968 to genetically engineered (recombinant) product in 1992 (Manucci, 2008). In the early 1980s it was established that plasma derived FVIII/FIX was contaminated with non-A non-B hepatitis (later known as hepatitis C) and HIV. In the UK, most people with haemophilia who had received plasma derived FVIII/FIX were tested for the presence of HIV-1 antibody between 1985 and 1989. The gathered data indicates that 1246 people with haemophilia tested HIV-1 antibody positive (Darby et al., 2004) and, when in 1992, testing for hepatitis C became widely available 4670 people were confirmed to be either co-infected with HIV or infected with hepatitis C alone (Haemophilia Society, 2011).

Until the 1980s hospital visits and admissions for haemophilia were frequent; treatment and analgesics were administered for acute bleeding episodes and bed rest was enforced. Lengthy hospital admissions equated with long absences from school and, although some boys attended residential schools for the physically handicapped (McAfee, 1966, Britten et al., 1966), educational opportunities were limited (Oldenburg et al., 2009). With the increase in home therapy and the advent of recombinant FVIII/FIX twenty first century, care for haemophilia changed. Currently, optimum haemophilia management focuses on bleed prevention, which is effected by administering prophylactic, intravenous bolus injections of FVIII/FIX therapy (Berntorp et al., 2010). The aim of prophylaxis is to transiently change the haemophilia from severe to mild-moderate; the injected recombinant FVIII/FIX elevates the circulating FVIII/FIX to near normal levels, its effect diminishes over the following 48 (FVIII) to 72 hours (FIX). Unexplained bleeding events may still occur but once a full prophylaxis protocol is established, usually between 4.9 and 6.9 years of age (Dodd and Watts, 2012), a-

traumatic bleeds are reduced from a frequency of four or more a month (World Federation of Haemophilia, 2012) to approximately three a year (Fischer et al., 2011).

Prophylaxis is initially administered by a member of the Haemophilia Team, based in a Haemophilia Comprehensive Care Centre (CCC). However, as parents become more familiar with haemophilia and its effects, they are given the opportunity to develop the skills necessary to take on this role so that they can administer treatment at home until the time when their son can administer their factor by self-infusion. The ability to administer treatment at home facilitates independence from the CCCs and there is an expectation that there should be few restrictions in activities of daily living (Manucci, 2008). In the UK a child is registered with one of 23 CCCs through which they have access to a consultant haematologist, specialist nurses, physiotherapists and other specialist health care professionals (Haemophilia Alliance, 2006).

There is much to suggest that life has improved for the family affected by severe haemophilia (Manucci, 2008; Oldenburg et al., 2009). Factor VIII/IX supplies are kept in the home allowing prophylaxis to be straight forward and treatment for acute bleeding episodes to be quick and easy; visits to the CCC are sometimes reduced to routine clinic appointments. However, this move towards independent home management shifts the burden of treatment responsibility and decision making from health care professionals to the primary carer, most frequently the mother (Shaw and Riley, 2008).

## **Literature Review**

A search of the English language based literature published between 1980 and 2013 using the CINAHL, Medline, PsycINFO and SocINDEX databases found a paucity of literature

directly relating to women's history, memories, and knowledge about haemophilia and their past experience. The search terms h(a)emophilia; mother\*; parent\*; family; prophylaxis; cop\* (cope/coping) and experience\* were used because these terms had been identified from an initial scoping of the literature as core search terms. The time frame was chosen as it encompasses the period of change from hospital to home based care and the move towards prophylaxis as the preferred mode of haemophilia management. However, it was during the 1980s that the impact of HIV and non-A non-B hepatitis became apparent; consequently much of the haemophilia related literature of the 1980s and 1990s that discussed experiences, coping and treatment of haemophilia was influenced by the effects of these blood borne viruses. This literature was not useful for the focus of this study and so has not been included in the review.

The results of the literature search resonated with my existing knowledge of the literature on haemophilia; although literature that I was not familiar with was also identified; such as Emiliani et al.'s (2011) study on the process of normalisation in families with children affected by haemophilia and Myrin-Westesson et al.'s (2013) hermeneutic study of carrier mothers' experiences of being mother to boys with haemophilia. The literature reviewed reflected studies undertaken in the United Kingdom (UK), Europe, Australia, North America and Canada.

The literature that had been selected was saved in preparation for its usefulness to this study. An initial reading was made to check for relevance. Some papers were discarded after reading the abstracts, including Dunn's (2007) study which focused on the needs of women and girls with a diagnosed bleeding disorder and Potnis-Lele and Kar's (2003) study looking at family modification strategies in families affected by haemophilia in India. The

information obtained from these abstracts was sufficient to show that these papers would not enrich the study. Other papers were discarded after a more thorough analysis such as Young et al.'s (2006) review of the Canadian haemophilia-outcomes Kids Life Assessment tool (Chok-Lat) which initially appeared to be relevant to the study but which focused on the design of the assessment tool and the child-parent concordance found in the results. A paper by Barlow (2007) was also excluded as the thematic analysis was of unstructured interviews with adult participants. Twenty seven relevant papers were saved, critically re-read and scrutinised; analytical notes were taken and points of particular interest highlighted.

Literature on coping styles in haemophilia, family communication about haemophilia and home therapy and prophylaxis was drawn on to inform the development of the proposal and has guided the study. In the critical review of the literature that follows, three core areas are addressed:

- Coping and normalisation;
- Home therapy and prophylaxis; and
- Carrier status and diagnosis

### **Coping and normalisation**

As an inherited disorder prone to unpredictable exacerbations, severe haemophilia is a burden on family processes (Steele et al., 1997). Families, with the support of CCC staff and their social network, evolve their own strategies to accommodate haemophilia's 'fickle' nature. These strategies and experiences may in turn be used to inform the sisters and daughters of men with haemophilia when becoming and being mother to their own affected

son (Varekamp et al., 1990; Steele et al., 1997 Miller et al., 2000). Carrier mothers' past experiences of family life with haemophilia frame its current management, treatment options and life style limitations within an historical context. This is a perspective denied to those mothers without a family history of haemophilia (Beeton et al., 2007; Gregory et al., 2007).

Developing a new knowledge of haemophilia, learning how to navigate their way round the health care system and how to give treatment to their boys were key turning points in achieving reconciliation with a changing life for the thirteen haemophilia carrier mothers in Myrin-Westesson et al.'s (2013) hermeneutic study. This study found that in the time after diagnosis the mothers experienced a deep sense of sorrow at burdening their boys with haemophilia. The mothers also felt guilt that their faulty gene had been responsible for their boys' haemophilia and for the disruption to family life. At this early stage the mothers, at times, felt overwhelmed with the worry that they might miss the signs of a bleed and when looking ahead felt an overwhelming sense of fear for their boys' future. However, once they had achieved competence in administering treatment the mothers became more confident, with some describing their personal growth, improved decision making and practical skills as giving their lives a new, enhanced sense of meaning (Myrin-Westesson et al., 2013).

The ability to administer prophylaxis was liberating for some parents. Emiliani et al. (2011) found that prophylaxis was only a contributory factor to the process of normalisation in families with one or more sons affected by severe haemophilia. In their two phase study Emiliani et al. (2011) used semi-structured interviews with thirteen parents from 10 families whose sons only received FVIII/IX replacement therapy for bleeding episodes (on demand therapy). The second phase of the study was conducted five years later. Two couples from

the original cohort were re-interviewed about their experiences in the four years since their boys had switched from on demand therapy to prophylaxis. Both sets of parents were in full employment and all four were described as middle class. The first family had four children, only one boy had severe haemophilia; his mother was not a carrier for the disorder. This family had many friends who they trusted and with whom they felt able to discuss any difficulties they were facing. Both parents and son had a positive relationship with clinical staff, and felt able to contact them if they had any concerns about prophylaxis or treatment. They also had a valuable acquaintance with a young man with haemophilia who was optimistic about his own future. They found that their supportive network had helped them to achieve a new and acceptable sense of normal. In contrast, the second family were isolated from social support, their son was their only child and his mother was discovered to be a carrier after his birth. These parents experienced their son's haemophilia as an immense burden which placed restrictions on them and their child alike. They felt unable to spend time together without their son and much of their life became a ritual with haemophilia and prophylaxis at its core. They constantly sought new information from sources outside their CCC and found themselves at odds with the staff who worked within it. They had difficulties in adapting to a new normal and seemed unable to benefit from support available from outside their small family group (Emiliani et al., 2011). This study found that although prophylaxis assisted families in gaining a sense of routine, other elements such as support, knowledge and understanding of haemophilia are required for normalization to be achieved (Emiliani et al., 2011).

The inheritance pattern of haemophilia means that the mothers of approximately two thirds of boys with newly diagnosed haemophilia are carriers for the disorder. The genetic

responsibility for their sons' haemophilia caused some mothers in Emiliani et al.'s (2011) study to experience a deep sense of guilt which frequently manifested its symptoms as depression and anxiety. Coppola et al.'s (2011) questionnaire based study found that although some non-carrier mothers experienced anxiety and depression; a sense of guilt was felt to a lesser degree. In this study, mothers' feelings of anxiety and depression coincided with paternal estrangement as fathers tended to withdraw from their boys' clinical care and experience a sense of their own exclusion from contributing to that care (Coppola et al., 2011). Coppola et al.'s findings reflect those of Herrick et al.'s (2004) survey based quantitative study of fathers' psycho-social adaptation to their boys' haemophilia. In this study there was a correlation between fathers who had been left out of decision making about their sons' clinical care and poor marital adjustment. And for those fathers who had experienced frustrating interactions with clinicians there was a demonstrable reduction in parenting satisfaction. (Herrick et al., 2004)

The ability to administer factor replacement as a small bolus injectable infusion has many benefits such as the child's ability to participate in most peer group activities and the ability for parents to administer treatment at home (Shaw and Riley, 2008). However, it can cause the interface between clinician and family to be reduced to a brief, irregular encounter which can fail to take into account the physically and emotionally disruptive effect of this chronic disorder on all family members (Bottos et al., 2007). Concern that these needs had not been addressed led to a programme of counselling and psychological support being developed for parents who attended a CCC in Padua, Italy. Bottos et al., (2007) examined depression, anxiety and coping strategies in thirty parents of boys with haemophilia before and after attending this programme of support. Three questionnaires; COPE (Coping,



Orientation to Problems Experienced) (Sica, et al., 1997), BDI (Beck Depression Inventory) (Beck, 1978) and STAI-Y (State-Trait Anxiety Inventory) (Spielberger, 1989) were used and the collected data analysed. The group support increased the parents' use of problem focused coping strategies and reduced their dependence on emotion focused coping. Levels of anxiety and depression had also reduced to a level that was felt by the parents to be manageable (Bottos et al., 2007).

Coping as an adaptation to the impact of stressful events is divided into two distinct domains. The first is an effort to be in control of the stressor; the second to manage the negative emotions associated with the stressful event (Miller et al., 2000). These strategies are broadly divided into 'active' where the focus is on developing a method of dealing with the problem and 'avoidant' where the stressor is internalised and blunted (Steele et al., 1997; Miller et al., 2000). Active coping with an adverse situation is usually more effective than internalisation alone which can lead to maladaptive coping and associated health issues (Miller et al., 2000).

Coping strategies used in childhood may be reused in adult life. This is significant for families with past experiences of haemophilia where the daughters of men or sisters of boys with haemophilia become mothers to sons with haemophilia (Steele et al., 1997). Steele et al. (1997) used predictive modelling to look at the impact that having a father with haemophilia had on family processes. Sixty nine families consisting of both parents and one child over seven years of age - participated in the study. The severity of the father's illness symptoms and symptoms of depression in either parent affected the child's coping strategies. The more intrusive the father's illness symptoms and the greater the severity of either parent's depression, the greater the internalisation of emotion and use of avoidant coping strategies

by the child were found. Where difficulties existed in the relationship between mother and child, the child's use of avoidant coping strategies increased further (Steele et al., 1997).

Miller et al.'s (2000) quantitative study used "The Revised Ways of Coping Checklist" (Vitaliano et al., 1985) for parents and "Kidcope" (Spireto et al., 1988) for children as a base from which to study "adaptive functioning in the face of adverse experience" (Miller et al., 2000 p4) in families where one or more members were effected by severe haemophilia. Eighteen families (a total of 59 participants) were asked to complete a coping check list in relation to a problem they were currently experiencing. Differences in coping styles were found between parents, boys with haemophilia and their siblings. Parents most frequently used active coping strategies such as social support, problem focus and cognitive restructuring. All the children (age range 6 years to 15 years 9 months) favoured more passive strategies with 'wishful thinking' being the most frequently used. The boys with haemophilia used distraction from adverse events as an alternative method of coping whereas siblings became more active using social support, problem solving and emotional regulation as their next best strategies.

### **Home therapy and prophylaxis**

The development of home treatment programmes for acute bleeding episodes in the late 1980s and prophylaxis in the 1990s radically changed the ways families live with haemophilia. Regular prophylaxis reduces the incidence of acute bleeding episodes to a median of 3.2 bleeds per year (Berntorp et al., 2010, Fischer et al., 2011); with joint bleeds and the associated acute and chronic pain occurring significantly less frequently in those who receive their prophylaxis in the mornings (Fischer et al., 2011). A longitudinal

comparison of primary prophylaxis (in which a schedule is started before the first joint bleed occurs; this is almost always before two years of age) and secondary prophylaxis (where the schedule commences after the first identified joint bleed) showed that breakthrough bleeding events occur more frequently in the latter group (Berntorp et al., 2010, Dodd and Watts, 2012).

Frequent visits to hospital for treatment, and the long periods of admission experienced before home treatment regimens were established (Teitel et al., 2004, Oldenburg, et al., 2009) have been replaced by educational visits to the CCC for parents and later the affected boys to learn the home management of haemophilia, the administration of intravenous treatment and to discuss life style issues (Oldenburg et al., 2009). A positive relationship with the haemophilia team at the CCC, in particular the nurse specialist, which is based on trust is integral to improving the quality of life of parents, siblings and boys with haemophilia (Teitel et al., 2004, Emiliani et al., 2011, Dodd and Watts, 2012).

Home based treatment gives independence from CCCs but can leave families with a sense of isolation when making crucial decisions; walking the tightrope between allowing their son to join in 'normal' activities and enforcing restrictions in physical activities which can limit independence (Teitel et al., 2004). Administering treatment at home helps older boys to conceal their difference and aids efforts to connect with peers (Williams and Chapman, 2011). For parents of younger boys, home treatment is not always so straight-forward. Shaw and Riley's (2008) study used semi-structured interviews and their findings show that parents recognised prophylactic home therapy as the rational way to manage their child's disorder but some expressed anxiety at administering a treatment which, in the wake of past contamination of FVIII/IX with blood borne viruses, no clinician would guarantee as

completely safe. This burden was made worse when trying to administer the intravenous treatment as prophylaxis to a reluctant child (Beeton et al., 2007, Furmedge et al., 2013). Parents in a focus group, when asked to recall their first experiences of administering their boys' treatment via a central venous port described sweating, shaking and feeling intensely emotional as they anticipated inserting the needle through their boys' skin so that they could access the port (Furmedge et al., 2013). Although they were concerned that the procedure might be painful for their boys their greater fear was that their aseptic technique might be flawed and that they could be responsible for introducing infection into the blood stream. Despite the weighty responsibilities of home treatment, the freedom from frequent visits to the CCC, the reduction in uncertainty and increase in control has made this responsibility, for most families an acceptable price to pay (Verekamp et al., 1990, Shaw and Riley, 2008, Beeton et al., 2007).

In their review of home management of haemophilia, Teitel et al., (2004) record that although parents were aware that regular prophylaxis could improve school attendance, participation in physical activities and integration with peers, adherence to a programme of prophylaxis was sometimes problematic. One hundred and thirty eight patients (aged over 13 years) and 42 parents of children (aged between 2 and 12 years) from six European countries participated in a highly structured quantitative study looking at influences on adherence to haemophilia treatment regimens administered in the CCC or in the home (De Moerloose et al., 2007). The data gathered were analysed by an independent market research company. De Moerloose et al. (2007) found adherence to treatment of between 80-87%; much higher than the reported 50% adherence expected for life long disorders in developed countries (WHO, 2003). The active involvement of CCC staff was considered by

many participants to be an important influence on following treatment regimens.

Respondents in England and Spain cited their parents before haematologists as the strongest authority on haemophilia; those in Sweden, Italy, Germany and France cited their haematologist first. Like Shaw and Riley's (2008) findings, this study identified parents' need for greater support and more information on the safety of treatment from members of the Haemophilia Team (De Moerloose et al., 2007).

### **Carrier status and diagnosis**

The genetic inheritance of haemophilia does not seem to be openly discussed in all families.

Gregory et al. (2007) in their study using semi-structured interviews with seven affected men, 19 obligate carriers<sup>1</sup> and 13 non-obligate carriers<sup>2</sup> reported that fathers found it particularly difficult to tell their daughters that they were obligate carriers for haemophilia.

Other studies found between 49- 57% of carriers unaware of their potential carrier or obligate carrier status before becoming pregnant (Maclean et al., 2004, Dunn et al., 2004).

Women were more likely to be aware of their potential to be a carrier for haemophilia if their brother was affected and the administration of FVIII treatment was not concealed.

Where fathers were affected by haemophilia and had chosen to give themselves their treatment in private, their daughters were at best uncertain, or some, entirely unaware that they were obligate carriers for the condition (Gregory et al., 2007). Those who did know their carrier status were unaware of the true implications and statistical risk for their own

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<sup>1</sup> Obligate carrier: In autosomal recessive disorders such as haemophilia this refers to a female who may be clinically unaffected but who must carry the gene mutation as her father is affected by the disorder.

<sup>2</sup> Non-obligate carrier: In autosomal recessive disorders such as haemophilia this refers to a female whose mother is a carrier for haemophilia and so has a 1:2 risk of being affected by the disorder.

daughters being carriers or their sons being affected by haemophilia (Maclean et al., 2004, Gregory et al., 2007, Dunn, et al., 2008, Myrin-Westesson et al., 2013). Awareness of 'carriership' in advance of pregnancy emerged in several different ways (Gregory et al., 2007). Where fathers were willing to be open about their disorder or a brother had haemophilia the women developed an informal understanding of their obligate or possible carrier status but factual discussions rarely took place (Gregory et al., 2007). One father described a feeling of intense guilt and responsibility when he considered telling his teenage daughters of their obligate status (Gregory et al., 2007). Some were told of their carrier status during their early teens, by a member of the CCC team. Some participants in Gregory et al.'s (2007) study recalled that during a scheduled appointment they had received a detailed explanation of haemophilia accompanied by diagrams of inheritance patterns; they described this as being similar to a science lesson and had not realised that this information directly related to their own lives.

In a small qualitative study, parents expressed experiencing a "huge emotional shock" on learning of their child's diagnosis of haemophilia (Beeton et al., 2007 p572). This shock appeared to be no less for those who knew their carrier status, apparently indicating that past experience does not necessarily prepare parents for this event. Although it is not clearly evidenced, it may be that the emotional shock experienced by obligate carriers is in part influenced by past family history of the disorder (Beeton et al., 2007). Parents found learning more about the nature and management of the condition helped to improve their quality of life as they reached a level of acceptance and eventual mastery of the condition (Beeton et al., 2007) and reconciliation with their changed lives (Myrin-Westesson et al., 2013). Such mastery is easily threatened by the unanticipated trauma of spontaneous

bleeding, urgent treatment and the disruption to family life, but the sense of control was increased by the ability to administer treatment as required at home. Myrin-Westesson et al.'s (2013), Beeton et al.'s (2007) and Gregory et al.'s (2007) studies are informative for the proposed area of research as they identify the widely varying preparedness and knowledge base from which women are learning to be mothers of boys with haemophilia.

## **Conclusion**

Although there is some literature relevant to the topic of this study, the literature review demonstrates a lack of evidence relating to mothers' experiences of raising a boy with severe haemophilia. There is lack of United Kingdom based and contemporary literature that investigates the essence of becoming and the early years of being mother to a boy or boys with severe haemophilia. This study aimed to address this deficit.

## **CHAPTER 2**

### **METHODOLOGY AND METHOD**

#### **Introduction**

This study is informed by both my workplace experiences as a practitioner providing care for children and families with haemophilia and the apparent lack of previously published literature specific to this topic. The study used a qualitative approach influenced by hermeneutic phenomenology to interpret the phenomena experienced by mothers who have one or more sons affected by severe haemophilia.

#### **Aim of the study**

This study aimed to explore what it is to become and be mother to a boy with severe haemophilia.

#### **Methodological Influence on the Study**

I chose a hermeneutic phenomenological approach as it was important to me to explore each mother's individual reality. I recognised that each mother's reality was likely to evolve through a process of constant renewal, influenced by her interpretation of and responses to her own past and present experiences. I acknowledged that such interpretations are dynamic, constantly reworked and reframed through temporal distance from the experience and informed by experiencing new phenomena.

Hermeneutic phenomenology recognises that pre-understanding and prejudice serve to inform, and are inextricable from, present understanding and that it is impossible to



separate one's understanding from one's self (Binding and Tapp, 2008). In conducting this study, I acknowledge that I have not bracketed (Binding and Tapp, 2008) my experiences of haemophilia gained as a practitioner providing haemophilia care. Instead when interpreting the phenomena and experiences as told by the mothers, I was aware that I brought to the investigation my own preconceived notions and pre-experiences (Salsberry, 1989). Whilst I did not bracket, I did adopt a highly reflexive way of working which I describe in more detail later in the thesis.

In hermeneutics, individuals are considered responsible for giving their own life meaning; making sense of the world through their existence (Annells, 1996). Experience and interpretation of phenomena provides each individual with their own reality in which "individuality is a fundamental value of human life" (Wartenberg 2008 p5). Sartre (1956) describes the essence of the individual as developing with living, and that only in the process of experiencing events being in the world (Heidegger, 1962) does that individuality develop. My pre-experiences, as Koch (1994) proposes, have inevitably influenced my interpretation of the data collected in this study. However I hoped that this inter-relationship between myself as researcher and my research participants would allow what Debesay et al. (2008) describe as new knowledge and new understanding to emerge. I wanted to generate knowledge and understanding of what it is to 'become and be mother to a boy' with severe haemophilia.

## **Method**

The purpose of this study was to hear the mothers' voices in each story; to hear their narrative as their own and their interpretation of the experiences which, when drawn

together, would create a unique reality of what it is to become and be mother to a boy with severe haemophilia. My intention was to minimise constraint and conformity and to give each mother the space, time and opportunity to recall her experiences with minimum direction from me, as the researcher. I wanted to encourage each mother to access her authentic self (Wartenburg, 2011) as she interpreted her memories and experiences veiled by the effect of time.

It was with this purpose that I chose unstructured interviews as the method of data collection. I chose this approach so that I could converse with each mother in the original, Latin sense of 'wandering together' (Kvale and Brinkmann, 2009 p 48), allowing the ebb and flow of the narrative to be led by each mother as she recalled her past experiences. This notion of wandering through a conversation intended to facilitate the interpretation of these experiences as they became reinforced or revised by 'inter-subjective' understanding (Standing, 2009) between myself and each of the mothers. I hoped that adopting this approach to the interviews would result in the emergence of what Gadamer (1981) proposed as being a new, co-constituted meaning of the mothers' lived experiences. The choice of a conversational approach to data collection fitted both with the chosen methodological approach and with my existing practice-based skill set. I utilise a conversational approach to haemophilia care in my clinical practice as this openness facilitates a sharing of experience between me, as practitioner, and the child and family; creating a deeper understanding and greater knowledge of haemophilia.

Care was taken when planning the interviews to put in place measures to ensure that the mothers felt safe and comfortable and were aware that I would not press them to reveal

aspects of their lives they did not want to. How these measures were operationalised are now discussed in more detail.

Before beginning the interview with each mother time was taken to engage in generic conversation aiming to create a comfortable dialogue. As the research was conducted during the winter months much of each conversation was spent talking of the effects of the weather or even simple day- to-day tasks. As the conversation evolved and a relationship developed, the discussion drew towards haemophilia, the research and its rationale. Once I felt sure that the mother was informed about the study, written consent was obtained (for a detailed presentation on consent, see later section 'Ethical Considerations and Research and Ethics Approvals'). In preparation I tested the digital recorder and positioned it to ensure a clear recording. I began the interview by asking each mother an open question about her own experience of haemophilia and from there I followed her narrative; I did not use a topic guide. This style of interviewing gave each mother the opportunity to tell her story as her own; stopping and starting as she chose, safely in control of what she disclosed. I was mindful that discussing past experiences of haemophilia might cause a mother distress and so I tried to use reflection and open questioning sensitively as we conversed. I used my knowledge of haemophilia, its history and its management, gained through my work as a practitioner, to help me to understand and contextualise each mother's story. Once the interview was complete, further conversation took place, the mothers talked a little more generally about their families. We discussed the purpose of the research again, in the context of the interview now being complete. I checked that each mother had my contact details and that I had theirs. In time, as we disengaged from our meeting together, the conversation moved towards the rest of the day's activities and the practicalities of my

departure. Time was spent on the process of disengagement so that each mother would feel able to contact me if they had further questions about the research or wished to withdraw from the study. It also gave me the opportunity to ascertain if the mother felt any sustained sense of distress after the interview process had been completed; and to provide support as needed.

### **Sampling and Inclusion and Exclusion Criteria**

The focus of this study was on mothers with or without a past history of haemophilia and who had a son or sons aged between 1 and 9 years, with severe haemophilia A or B (Factor VIII/IX <1-1.9%) and who had developed independence from the CCC. The target CCC had a caseload of approximately 30 families with 14 mothers who fulfilled the core study criteria: all were invited to participate in the study.

This study focussed on mothers whose sons were in their early years of living with severe haemophilia. This early period was chosen as the first years of being mother to a boy with haemophilia involves an intense period of learning and frequent visits to the CCC for advice and treatment. Part of the difficulty of managing haemophilia, experienced by parents and practitioners alike, is its unpredictability and the sporadic nature of bleeding episodes.

Parents find detecting the early signs of an atraumatic joint or muscle bleed exceptionally hard. This is particularly problematic for parents in the early years when their son is least able to communicate his needs. It is during these first years that many treatment and lifestyle decisions have to be made by parents; for example, when to present to the Centre for treatment and when to embark on prophylactic therapy. Mothers who have a close family relative with haemophilia may use their memories or experiences to help with their

decision making. The ability to administer intravenous clotting factor at home brings with it independence, greater opportunity to choose the activities in which their sons can participate and the responsibility of making clinical decisions. All of these factors supported the decision that mothers of boys aged between 1-9 years were likely to be able to share particularly pertinent insights into becoming mother of a boy with severe haemophilia.

The host CCC is situated in a city with an ethnically diverse population but 83% describe themselves as White British. The second most frequently occurring ethnic group is British Asian, who constitute 7% of the city's population (Office for National Statistics, 2011). The host CCC provides a haemophilia service for the city and the population of the surrounding towns and rural areas. The decision to exclude mothers who were unable to use or understand conversational English had a minimal effect on the numbers of mothers invited to participate in this study as few of the caseload were unable to converse in English. The decision to exclude this group was made as in depth interviews encourage a participant to provide a narrative of her experience. The gathered thoughts, pauses and feelings are an expression of the life lived. The use of a translator, would have given this personal story an interpretation outside the control of both the participant and the researcher, different to that which had been meant to be conveyed.

### **Participant Recruitment**

Following full ethics and research governance approval (see later section 'Ethical Considerations and Research and Ethics Approvals') I met with the nurse specialist at the CCC host site to discuss the research proposal and its design.

The mothers who fulfilled the research criteria were identified by the clinical team. The nurse specialist at the host site sent these mothers a letter of invitation to participate (Appendix 2: Letter of Invitation) accompanied by an information sheet (Appendix 3: Information Sheet) and a return stamped addressed envelope. A reminder letter (Appendix 4: Reminder Letter) was sent two weeks after the letter of invitation. This letter was posted to all of the mothers who had been sent the letter of invitation. Where it was thought possible that families were in crisis the nurse specialist at the study CCC used her understanding and knowledge of their situation as she considered the appropriateness of sending a letter of invitation. The letter invited the recipient to read the enclosed information sheet that explained my involvement with haemophilia, the purpose of the study and method of data collection, along with recognition of the need for confidentiality. My work phone number (with availability times), email and work address were on the letter and the mothers were encouraged to contact me by telephone, letter or email according to their preference, for clarification about the study. A cut-off date for reply was clearly stated in the letter; this assisted me in developing an interview appointment framework. Responses from mothers who were interested in taking part were returned to me at my work address. I then made an initial telephone call to all those who wished to participate and made provisional plans to interview them. Once the last date for replies had passed, I contacted the potential participants again by telephone to confirm the venue, date and time for the interview.

### **Ethical Considerations and Research and Ethics Approvals**

Approval for the study was given by the Greater Manchester-West; Research and Ethics Committee; reference number 12/NW/0727 with local approval from the Research and

Development Committee at the recruiting site, Sheffield Children's NHS Foundation Trust. Following this approval the study received ethical approval through the BuSH (Built, Sport and Health) Ethics Committee at the University of Central Lancashire (UCLan).

Concerns about clinical care provision (Karnelli-Miller et al., 2009) including confidentiality and anonymity can influence potential participants' decision to engage in health related research. My decision to recruit my sample from a CCC where I was not involved with any aspect of a child's care aimed to alleviate the tensions inherent in a practitioner-researcher working with families on her own caseload (NMC, 2008). Further to this, although the letters of invitation were sent to the mothers by the CCC's nurse specialist, they were returned to me at a separate address; the staff at the host CCC are unaware who chose to participate in the research. The study used an opt-in rather than an opt-out approach to recruitment. All eligible mothers were provided with detailed but accessible information packs containing key information about the study to allow them to consider their decision about whether to participate.

Consent was confirmed through signature before the interview took place but after there had been the opportunity for dialogue. I made it clear that participation in the study did not bring any direct benefit to them, as participants, or to their child and that their child's care would not be affected in any way by their decision to either participate or decline to participate in the study. When both participant and I felt confident that there was an understanding of the research purpose, that issues of confidentiality and anonymity had been addressed and a position of informed consent had been achieved, the consent form (Appendix 5: Consent Form) was signed (RCN, 1995). I did not divulge participant names to anyone including other participants or CCC staff. Total confidentiality could not be assured

as conversation between participants, potential participants and others could have occurred inside or outside the CCC, which I could not control. Assurance was given that at any time before, during or up to eight weeks after the date of interview a participant could choose to withdraw her consent and be given the option of having any electronic data deleted and hard copy data (e.g. transcripts) destroyed. No participant chose to withdraw from the research.

Reports to date and any future reports arising from this study will not contain any quotes directly attributable to participants. The risk of disclosing identity has been and will be minimised by omitting or altering very specific contextual details as needed. However where consent has been obtained, the digital recordings will be kept for a period of five years so that audio extracts may be used within presentations; allowing the participants' own voices to be heard. Where staff from the participants' own CCC attend these presentations there is a possibility that the participant's voice might be recognised and the written and audio quotes linked together; this was made clear to the participants through the information sheet and reiterated prior to gaining consent.

During the interview process, issues relating to bereavement, loss and guilt surfaced; I dealt with these matters as sensitively as possible and, where appropriate, discussed with the participant the possibility of referral to the psychological services linked to the CCC. I also made available to the participants the contact details of local support and counselling agencies to which either a self-referral or General Practitioner referral could be made. I was provided with support by the supervisory team at the Children's Nursing Research Unit (CNRU) Alder Hey Children's NHS Foundation Trust and UCLan.



I made participants aware that I work as a nurse specialist in the Alder Hey CCC. This information could have triggered them to ask for opinions or advice directly relating to their son's condition or treatment. However, before the interviews began I explained that it would not be possible to give advice, but that if requested I would pass on any points of concern to the staff at their CCC. No such requests were made. Similarly had a participant disclosed methods of managing their son's disorder which could have caused him harm, have an adverse effect on his condition or in other ways cause concern; these issues and any concerns around safeguarding or illegal activities would have to be reported. The need for such a report would first have been highlighted with the participant and the sharing of information would have been in accordance with the host Trust's guidelines. I did not have to deal with any instances of concern.

In preparation for meeting the participants in their own homes I familiarised myself with the Sheffield Children's NHS Foundation Trust lone worker guidance. I left information about where the visit was taking place and the timing of the visit with a core contact. Prior to entering the property and again after exiting, I sent a telephone message, confirming my safety. On entering the property I identified potential exit routes and I planned to use my judgement to ensure that I did not enter a home if I felt unsure or unsafe. No instances occurred where I felt unsafe.

## **Data Analysis**

All of the digital data (audio recordings and transcripts) were encrypted and transcription of the interviews was undertaken by an experienced transcriber who followed the NHS protocol for confidentiality. The transcriber and I agreed a system of participant identifier

codes; M1 (Mother 1) to M5 (Mother 5) before transcription began. I made a separate decoder key which contained the participants' identifying information and demographics such as each mothers' name and those of family members and details of the schools and hospitals attended. The coded, anonymised transcripts are kept separately from the decoder key and from the encrypted digital files. The participants had agreed to be contacted after the interview by telephone if parts of the digital recording were unclear, or if I was uncertain about what they had meant by something they had said. I did not need to contact any of the participants after the interviews had been completed. Although participant numbers were small, I initially separated the transcripts into those with past experience or knowledge of haemophilia (two participants) and those without (three participants). I felt that it was possible that past knowledge or experience of haemophilia might have an effect on the mothers' experiences with their own son.

Before interpreting the data I familiarised myself with some of the accepted methods of data analysis in qualitative research. Kvale (1996) advises that there are no 'magical tools' (Kvale, 1996 p 187) that will unlock the meaning hidden in interview transcripts; and reading other authors' perspectives on qualitative research (Braun and Clarke, 2006; Miller, 2003; Standing, 2009) I could see that this opinion was shared. I recognised the need to immerse myself in the words of the mothers but like Miller (2003) initially felt a reluctance to categorise or break the transcripts of the mothers' conversation words into units of meaning. I was unwilling to take the reductionist approach described by Hycner (1985) who also urged the use of bracketing, which I was certain I would find impossible. Colaizzi's (1978) 'seven procedural steps to analysis' as described by Miller were a logical framework and Braun and Clarke's (2006) six step approach seemed manageable. Together these

helped me to realise that to make it possible to interpret the mothers' meaning it was necessary to identify a way of breaking down the transcripts into units so that I could identify emerging themes. But in keeping with many qualitative researchers (Kvale, 1996) although my interpretative process was informed by such logical and step-wise techniques and methods I did not follow one specified framework.

Throughout this research project I have used field notes to record my thoughts, reflections and processes. My first notes were made in the period immediately after interview, when I wanted to capture my initial responses to the participants; my first thoughts and feelings. My more detailed notes were written as I familiarised myself with the mothers' stories, explored their meanings and pondered on emerging themes. These later notes were more analytical in nature and were of my reflections on the data as I interpreted the mothers' individual and collective meaning and experiences.

I began to familiarise myself with the mothers' stories by listening and re-listening to the audio recordings of the interviews. As a practitioner I use interpretation of intonation, pauses and vocal expression, as a means of understanding what is being said by mothers and their boys during my clinical interactions. I felt that my interpretation of the interview transcripts in this study would be helped if I could recall the participants' voices as I was reading the typed documents.

I found it difficult to take the step from familiarising myself with the transcripts to beginning to code and 'break' the mothers' words into fragmented units. I felt concerned that I would ascribe meaning to the mothers' stories that was not meant, or place emphasis on parts of the narrative that the mothers had not felt important. I felt it was essential to treat these

stories with integrity and respect as in conversing with me the mothers had shared part of their inner being. During this period, where I was feeling unable to move forwards, finding it difficult to 'risk' making any interpretive decisions, I was supported by my academic supervisors as I navigated a period of intense reflection and I used supervision as an opportunity to discuss my thoughts and tentative interpretations.

Coding at last began. Initially as I read through the transcripts I highlighted phrases and words which I felt were important and had meaning and resonance. I gathered these words and phrases and identified ideas and concepts which were individually experienced or shared by the other mothers in this study. As I read and re-read the transcripts and developed my thoughts; broad themes began to emerge. I created a multipage excel database to record codes and themes with supporting quotes from the transcripts. Each page of the database was labelled, and the detail from each of the mothers' transcripts was assigned a row (M1-M5). Each column had a heading and a hyperlink to the relevant part of their interview (See Appendix 6: Table 1 as an example).

Although the contents of the database contained quotes directly attributable to each mother it remained a fairly superficial repository for data and my struggle to allow myself to ascribe deeper meaning to the mothers' words continued. To help me over-come my hesitancy I broadened the scope of my background reading. I grappled with an introduction to existentialism (Wartenburg, 2011) and attempted to draw a deeper understanding of emotion by reading a translation of "Sketch for a theory of the emotions" (Sartre, 1962); while these were deeply informative, my limited grasp of their meaning meant that they were also in some ways intimidating. My attention was diverted and I began to look at concepts and theories as presented in boundary ambiguity (Mu et al., 2005; Berge and

Holm, 2007); the impact of the uncertainty associated with chronic illness (Holm et al., 2008) and biographical disruption (Bury, 1982; Delbene, 2011). I reflected on stigma and parenting (Francis, 2007) and the social construction of motherhood (Collett, 2005).

I also wondered if the mothers' experiences could be interpreted through the construct of Posttraumatic Stress Disorder (Landolt et al., 2005; Cabizuca et al., 2009) but felt that this model was too constrained. I was concerned that recognising elements of these concepts in my interpretations of the mothers' experiences represented a conflict with hermeneutics. Later I found that they were valuable aids to my interpretations. I then focussed my reading on papers including those that looked at phenomenology of practice (van Manen, 2007) hermeneutics as embodied existence (Schuster, 2013) and the influences of Husserl and Heidegger (Koch, 1995). I read articles that used a hermeneutic approach to interpreting experiences of chronic health difficulties such as cerebral palsy (Glasscock, 2009) and feeding difficulties (Hewetson and Singh, 2009). All these contributed to my still embryonic knowledge and understanding. But two papers on hermeneutic phenomenology, the first by Annells (1996) on hermeneutic phenomenology as a philosophical perspective and the second which focused on closing the hermeneutic circle (Debesay and Slettebo, 2007) along with Binding and Tapp's (2008) paper on developing an understanding through three levels of conversation led me to feel that my developing understanding of hermeneutics as a philosophical view was sufficient that I should move onwards.

This emerging understanding enabled me to trust in my belief that interpretations of experiences are not rigid and unbending but are flexible; yielding to the effects of time and newer experiences. Interpretation of our own and others' experiences are not locked into a position of right or wrong but are dynamic. I was now able to delve deeper into the data and

became more confident in my interpretations and the gathering together of themes.

However before I was able to commit myself completely to these themes I had to check and reflect on whether my framing of a concept was supported by the mothers' words. I needed to be certain that my experiences as a practitioner did not impose on or dominate the mothers' stories. At last I felt that the themes which had emerged through my interpretations captured the depth of the mothers' experiences; echoing the essence of what it is to become and be mother to a boy with severe haemophilia.

## **Conclusion**

Having undertaken a careful and considered approach to the planning, design, and implementation of my study and been conscientious with the way in which I engaged with my data, I felt confident that I could portray the mothers' meaning through my final themes. It is these, five distinct but interwoven themes which form the basis of the following chapter.

## **CHAPTER 3**

### **FINDINGS**

#### **Introduction**

Five mothers responded to the letter of invitation declaring an interest in participating in the study. They were offered the option of their interview taking place at their home, avoiding the need to travel, or in a mutually agreed venue where the interview could take place without interruption. Travel (if necessary), after school club and baby-sitting costs could have been reimbursed through a grant from the Roald Dahl Foundation; no mother required these services. All of the mothers chose to be interviewed at home. The interviews were digitally recorded and lasted between fifty five and 102 minutes (mean 61 minutes).

All of the mothers were Caucasian and married; all of their husbands were in paid employment as were three of the mothers. None of the mothers had daughters and three were first time mothers. Two of the mothers had three sons; one of these mothers had one, the middle, affected son; the other mother's oldest two boys had haemophilia the youngest did not (Table 1). Two of the mothers had one of their children with them for all or part of the interview; one husband was present briefly for part of the interview.

Five distinct but interwoven themes emerged from the mothers' interviews: the diagnosis of haemophilia; managing haemophilia and experiencing hospitals; giving treatment at home and regaining control; daily anticipation and imagining their boys' futures; and separateness, isolation and support. These themes although shared by the mothers were individually experienced and interpreted by the mothers' in the context of their own history.

	Family history of haemophilia	Number of children	Number of sons with haemophilia	Age group of affected son(s)	Results of genetic assessment	Treatment route	Relationship Status	Employment status	Partner's employment Status
<b>M1</b>	None	1 boy	1	Pre-school	M1 confirmed carrier her mother unaffected, haemophilia started in M1	Portacath	Married	Part Time	Employed
<b>M2</b>	None	3 boys	Middle son	Infant school	M2 and her sister confirmed as carriers	Peripheral veins	Married	Full time at home	Employed
<b>M3</b>	Yes, affected father	1 boy	1	Infant school	M3 obligate carrier	Peripheral veins	Married	Full time at home	Employed
<b>M4</b>	Yes affected uncle	1 boy	1	Pre-school	M4 and her mother confirmed as carriers	Portacath	Married	Full Time	Employed
<b>M5</b>	Discovered after eldest son's diagnosis	3 boys	First and second son	Junior and Primary school	M5 carrier as is her mother, geneticists extrapolate that M5's deceased uncle and brother succumbed to catastrophic haemophilic bleeding episode	Portacath	Married	Part Time	Employed

**Table 1 Characteristics of interviewed mothers**



## The Diagnosis of Haemophilia

The diagnosis of severe haemophilia in their sons brought with it a sense of shock and maternal guilt for the loss of a healthy child and the responsibility for their boy being different.

All of the mothers in this study were carriers for severe haemophilia. One mother described how she was dismayed to discover that the genetic error had started with her, *“so ... they tested my mum but she wasn’t [a carrier], so it’s something that has just started with me.”* (M1). A second mother found out that haemophilia was ‘in the family’, after her son was diagnosed at 16 months of age, that she and one of her sisters *“who... has got the same condition”* were carriers, although she explained *“my parents haven’t shown an interest to have bloods taken to find out where it’s from”* (M2).

The other three mothers had a family history of haemophilia but only one knew of her carrier status before she became pregnant. These mothers had a range of different memories of the effects that haemophilia had on their family as they had been told or had overheard complete or incomplete stories of its impact. M4 was aware of her carrier status and talked of how she could not remember her father ever complaining about his haemophilia although she recalled how he talked in a fairly matter of fact way about its impact:

*“He just used to say his elbow is hurting...or something... He could ride a bike up until he was in his teens and then [with] the arthritis in his joints because of previous bleeds he couldn’t do it anymore and he had to stop it.”* (M4).

She described how her father's independence and her knowledge that treatment had improved since her father was a child had helped her to plan for her baby's delivery once she knew that it was a boy:

*"He's [Dad] always remained really independent and done everything for himself, he's never let anything stop him. So he's just gone out and done whatever he wanted to do....[so] as soon as I found out it was a boy I was right he's got it, that's it.... let's have an elective section; then all the teams are there who need to be there, It's not an emergency, not rushed. They were on standby to take a cord blood sample. It was the day after that I found out....I just thought 'Right. Crack on; crack on!'" (M4)*

M4's day-to-day familiarity with haemophilia and her father's pragmatic approach to its effect on his life appeared significant in her planning and preparation for her son's arrival.

M3's awareness was a step removed and her familiarity less secure. She was aware of her uncle's severe haemophilia and his increasing immobility caused by recurrent bleeding into knees and ankles, but she was not aware of its implications for her as a future mother until she gave her family history to the midwife when she was 10 weeks pregnant. After discussion the midwife ensured that she was referred to the (adult) haemophilia team where specially arranged blood tests confirmed that she was a carrier for the disorder. She recalled:

*"The family had always been told that it [uncle's haemophilia] was a genetic mutation because there had been no previous history in the family.... I had a lot of contact with my uncle over the years....you can't conceal your haemophilia, I knew*

*that he developed mobility problems but I didn't really know the impact that it was actually having on his life" (M3).*

During the first consultation and with her carrier status confirmed, the haemophilia team were keen to contextualise haemophilia for her in terms of its contemporary treatment, control of symptoms and potential impact. She recalls with irony *"this big positive spin"* as they reassured her about her son's future should he be diagnosed with haemophilia and they explained that:

*"It's not the same as the previous generation, I know your uncle has haemophilia and that it was the blight of his life but it's not like that now and they can have a normal life and everything's fine." (M3).*

Although M3 described this positive representation of life with haemophilia in the 21<sup>st</sup> century as *"a good thing... when you are pregnant you don't want to know everything"* (M3); this mother's initial experience was far from positive and despite her ante-natal consultation with the haemophilia team and the reading about haemophilia that she had done in the latter days of her pregnancy, she felt ill-prepared for the horror of her child's first bleed. This episode occurred within hours of his haemophilia being confirmed, after his traumatic delivery by emergency Caesarean Section. Mother and infant had just transferred to the post-natal ward from the high dependency unit when she found herself shouting for help:

*"'Can somebody get a doctor? My baby's head's started getting bigger.' And they were like, yes there's one coming.' And then it all went a bit sort of crazy because his head was getting bigger.....They had difficulty finding veins, it took them a while,*

*and he had factor. But we [parents] were still quite calm, you think he'll have factor and it will be ok" (M3).*

Then, almost unable to control her distress, she whispered as she recalled:

*"[A doctor said] 'He could be bleeding on the brain'. They took him to the children's hospital [for] an MRI but we couldn't go with him. So he got taken away [the doctor explained]: 'He'll have the MRI, we've got neurosurgeons on standby if he needs to have surgery it will happen straight away'" (M3).*

After his MRI, her son was brought back to the post natal ward; she and her husband were shocked and relieved when they were told that there had been no bleeding on the brain. *"He's actually fine!" (M3).* The scan had revealed an extra-dural bleed which the haemophilia team explained would have no long term developmental consequences. However, this trauma had long lasting repercussions for this mother. She explained how she spent the early months of her son's life wrestling with feelings of guilt and self-blame. Initially this focused on what she felt had been her lack of assertiveness when she was in the early stages of labour, she revealed how:

*"I started to blame myself because I had read all about the statistics of delivery and C section versus normal delivery and so started to question whether I should have been more assertive...I had asked for a C section early in my delivery and they said – 'No, no, no, you don't need one, we can keep going'" (M3).*

These feelings of guilt persisted and defied logic, as she blamed herself for ruining her boy's life by inflicting him with her genetic flaw. She explained:

*“Logically, yes, I carry the gene. But that doesn’t stop the mother side of you kind of feeling guilty...and beginning to think this is all my fault...oh my god it’s your baby ... I thought he was going to have a normal life, I was feeling relieved but then [thought] I’m a terrible person ..... What is his life going to be?”(M3).*

Gradually she adjusted to her new reality and recognised that the outcome of her son’s first bleed, although traumatic had had no lasting effect on him. Further helped by several months without another emergency she was aware that she could not allow herself to continue to feel so negative and was able to comply with her husband’s request to:

*““Pull yourself together’....” and reflect that “It’s happened now, he has haemophilia; he didn’t get brain damage, let’s get on and deal with it” (M3).*

The steps leading towards diagnosis for the other mothers were characterised by individual series of events. Each mother had her own story to tell; each was differently affected. M5’s first day back at work was interrupted when her seven month old son toppled over and cut his chin at his maternal grandmother’s house. This minor injury led to a series of unexpected explanations and family revelations. Initially grandma had felt that the injury would be easy to manage, but the bleeding persisted and once her daughter had arrived home they embarked on a number of increasingly anxious trips to and from their local Accident and Emergency Department. Here staff attempted to Steri-strip™ the wound; but the bleeding continued, dislodging the Steri-strips™. At last a referral was made to the Children’s Hospital and her son was admitted. M5 talked about how her anxiety increased over night as the nurses’ attempts to stop the bleeding were ineffective and their attempts to get a

doctor to review the situation failed. She laughed nervously as she recalled her fear as she looked at her little boy smeared in blood and naked except for ineffective bandages:

*“All through the night the nurses kept phoning for the doctor to come and by the end of it, I mean I’m laughing now but, at the time he’d got this wad on his chin a bandage all the way round his head to hold it on! He ended up completely naked because he was just covering vests and baby-grows and everything with blood.”*  
(M5).

Although this was her first child, it was obvious to her that the bleeding was not normal and, having spent a night worrying about the lack of improvement, her anxiety changed to indignation when the anaesthetist arrived to assess her son’s fitness for anaesthetic. She recalled:

*“The anaesthetist came round because obviously they were going to stitch him up in theatre and went – ‘Where are these bruises from? There is something not right’. Obviously initially that sort of thing put my hackles up.....when they ask ‘Where’s this bruise from?’ you get defensive.”*(M5).

Blood samples were taken and after 22 hours of bleeding, without the results of the tests, the surgery took place. Despite the implied concerns in the anaesthetist’s remarks, no allegation of non-accidental injury was made although another doctor asked M5 if she knew of any family history of heavy bleeding. She recalled being shocked, when, later that day she spoke to her father on the telephone, recounting the day’s events:

*“I remember being on the phone to my dad saying – ‘They’re talking about blood disorders in the family’ [I told them] ‘there’s nothing’; [Dad] went ‘Oh there is, your mum’s brother died’, and I was like ‘Aaagh!’”*(M5).

Despite being stunned by this *“family history....hidden; regarded as some dark secret”* (M5) she felt relief when the haemophilia team came to talk to her about her son’s diagnosis:

*“I saw [the specialist haemophilia team] at the side of his cot ‘Right, this is what we’ve found’. I kind of took the news quite open-minded I’m not saying that I didn’t go away and have a little cry ‘what have I done?!’”* (M5).

In the context of the previous night’s traumatic experience, M5 was glad to have an explanation for her son’s dramatic bleeding episode. She soon realised that the diagnosis of haemophilia also helped to explain her boy’s previously unaccounted for swellings and bruising. These outward signs of injury had caused her to avoid attending baby clinic for fear of judgement by health care professionals:

*“[I] was actually quite relieved; it explained a lot of things; he used to bang his hand all the time on his cot and it used to be so swollen. I wanted to take him to be weighed at baby clinic I looked at his hand, and I thought ‘I’m not taking you anywhere!’ Thank god we’ve found this out now rather than months later when something serious happens, or somebody calls social services on me because my son’s black and blue”.* (M5)

These first months were a period of adjustment for M5. She spent time learning more from her mother and uncles about their brother who had died from a bleed at nine years of age. Shortly after her son’s diagnosis M5 and her mother were seen by the ‘adult’ haemophilia team. Blood tests confirmed that mother and daughter were carriers for haemophilia and it appeared that both were mothers to boys with severe haemophilia. M5 explained:

*“My mum, when she had my oldest sister mentioned haemophilia to her midwife. She just told her not to be so silly. So mum never talked about it again. Later [the year] before she had me she had a little boy [my brother] who was a forceps delivery; he*

*died a week later from a brain haemorrhage. The doctor said, from the family history, it must have been haemophilia. For mum and dad it was almost a weight lifted, after all these years, got almost like a justification for it, as to why it had happened.”(M5).*

M2 had a similar Accident and Emergency experience to M5 when her second son at 16 months of age fell and pierced his tongue with his two teeth. The bleeding would not abate; she recalled the reassuring words that accompanied her visits to the Accident and Emergency department *“There are a lot of veins in the tongue”* and explained that it was usual for there to appear to be blood everywhere when a tongue had been bitten; *“Go home everything will be fine”* (M2). However, there was no reduction in bleeding and she found little comfort in their advice. Her dilemma was complicated by an invitation to her sister’s wedding which was to be held at a venue over a hundred miles away. She described her reluctance to attend and her sense of relief at being summoned to return home by her mother in law who said: *“Look he’s still bleeding!’ What looked like a bubble had formed, he’d move his tongue and he’d knock it all off again”* and the bleeding would start again with what seemed to be renewed vigour. M2 talked of how terrified she was, saying *“I’ve never seen so much blood; it was horrendous, really scary. It made me physically sick. It was like someone had stabbed him”* (M2). Her terror turned to panic, incredulity and anger at the decisions made when she returned again to Accident and Emergency:

*“The very last time we went he was really pale, really lethargic and he was really flopping. [They did] his obs [observations] [and said] ‘you are not going home’ They took loads of blood, they were saying ‘Give him an ice pop, [it will] stop his bleeding’. I thought to myself: ‘Are you stupid? That’s what I’ve been doing.’ After two hours they said ‘Well he has to stay in’ [moments later] ‘Look it’s an emergency you need*



*to take him up to [the Children's Hospital] can you make your own way?' I was absolutely disgusted. I was so annoyed" (M2).*

Enlisting support from her in-laws to look after her older son, she and her husband drove the long journey with their sick child to the Children's Hospital fearful of what news might await them there. She recalled the far from reassuring parting words from a member of the Accident and Emergency staff that played over in her mind as they travelled into the night:

*"I don't want you to panic,' which obviously straight away you do. 'The ward that you are taking him on there, you will see patients that have got leukaemia and cancer and no hair.' It really did put the fear of God into me!"(M2)*

She recounted, through tears, the first few minutes after they arrived on the ward, still unaware of the diagnosis:

*"We got there and he was violently sick so all the blood came up. As soon as I saw that I was crying, screaming, he was screaming" (M2).*

The diagnosis came within an hour of their arrival on the ward. But these traumatic few days led life to *"completely, completely change!"(M2)*. She went on to describe how her older son was left on the periphery of their new life, finding it difficult to include him while they adjusted to managing the haemophilia. Talking about this situation she explained:

*"He [son] has severe type A so it was a bit of a nightmare, I wanted to wrap him up in cotton wool...., [but] I felt sorry for [other son]; he felt left out in a way. It wasn't intentional but [our youngest] was the focus just because of his haemophilia and the treatment. It was all new. We ended up having to go to [the children's] hospital quite a few times and he was being left with grandparents. I don't think he liked that at all" (M2).*

The fifth mother in the study, M1, described utter devastation on receiving her son's diagnosis. Unlike the two mothers who had sought help when their boys were bleeding overtly, M1 had noticed a series of more subtle signs. She seemed almost to regret the day that these symptoms finally led her to pluck up the courage she needed to visit the GP with her son; the visit that led her to discover that her son "...had been a haemophiliac for a while....we had not known."(M1). That day her sense of normality was overthrown, despite her efforts to prepare herself, as she recalled:

*"I'd done my research on the internet when I thought something was wrong, and I thought 'Oh it might be a mild case of Von Willebrand's Disease or something'. I didn't think it was going to be anything [as] serious as severe haemophilia..... So I went to the GP and the GP sent us to see a [local] paediatrician. He had a blood test done and it came back that something was wrong.... you always think it's going to be normal, you know. You always think they are just over-cautious" (M1).*

They were given an appointment in the region's only CCC based in the children's hospital, an hour's drive from home so that they could learn more about the diagnosis. M1 described how the shock was so great that initially all she could remember was being told about the treatment, and that she would one day administer it herself; at the time this had seemed an impossible task:

*"It was a big shock I was really, really upset and I suppose devastated in a way because you don't know what it's going to mean....I was so shocked [that afterwards] I didn't even [recognise the doctor] who first explained the diagnosis. The uncertainty, the treatment was such a big thing, it seemed like, you know, that it's*

*going to be too devastating to do.....I had to go outside, I was crying. I was very focused on sort of 'What if he has a car accident or something will he just bleed to death?'"(M1).*

The diagnosis of their boys' haemophilia led these mothers along a pathway of unexpected challenges; different from those experienced by mothers of unaffected boys. These challenges seemed insurmountable for some as they reeled from the shock and began to learn what severe haemophilia meant to them and their boys.

### **Managing Haemophilia and Experiencing Hospitals**

In the months after their sons' diagnosis the mothers experienced many visits to hospital for advice, treatment and appointments. This dependence continued up until the ability to administer treatment at home created a sense of independence from the CCC.

'Managing haemophilia and experiencing hospitals' was a core and evocative theme identified from the data. The mothers talked of the, at times, shocking experiences of the planned and unplanned visits to hospital with their boys. They described the anxiety they experienced when their sons were assessed and treated by health professionals who did not work in the CCC and so were unfamiliar with managing haemophilia. This was at times made more distressing when there were difficulties with venous access.

Ever since her son's diagnosis with haemophilia, M2 had tried to protect him and be there for him; this had meant that, almost without fail, she had been one step away from him. She explained: *"Every time he ran anywhere I was there behind him. It was an absolute nightmare."* (M2). Despite this constant attention and her close proximity to her son she could not always stop things happening, she re-lived a terrifying incident, in a children's play

area, in which everything seemed to happen in slow motion when he was just out of arms reach:

*"We went to a park and he was running on the rubber floor, I could see it happening and I could just not get there quick enough; and oh my goodness! He'd [just] been diagnosed with haemophilia and as he fell.... there was this steel step of a ladder and his head just smacked it, and bounced off and he came running round and he'd split his head open. Oh! I was absolutely screaming in this park. I'm shouting, screaming at the top of my voice 'Somebody help! Help me, get me an ambulance!' I literally thought he was just going to bleed to death there. I've never seen so much blood. I bet they thought 'that stupid woman!'"(M2).*

Although "eventually" (M2) an ambulance car and 'first responder' arrived M2 described how her sense of panic increased as the responder made it clear that he did not know much about haemophilia. There was no relief when they arrived at the local Accident and Emergency department to discover *"they [A&E staff] have never, ever dealt with a haemophiliac. Never, ever mixed a treatment up for a haemophiliac!"* (M2) By this time she was so fearful that her son would die that she phoned the specialist nurse at the CCC for help. A strong echo of the emotion she experienced at the time was evident as she explained:

*"I've got the phone to my ear to speak; I was crying, sobbing my heart out. 'I want them to mix this Factor up quick.' I could feel my heart going, I was getting really, really angry because he was still bleeding and bleeding I'm thinking – he's going to*

*die .... 'This is absolutely ridiculous, just give me the medication and I will go, I will go up to the Haemophilia Centre and they will do it there'" (M2).*

Beside herself with distress, she did not have to wait long to discover that the journey to the CCC would not be necessary as a phone call between nurse specialist and triage was made and the treatment given. Scarred by this experience, M2 stated she has since felt unable to use the local Accident and Emergency services for any of her three boys:

*"I still don't trust them [local A&E] now; to be honest I would rather travel and feel safer. I know that sounds silly! But even if it was [one of the other two boys] if they were ill, that would not be my first choice" (M2).*

Emergency trips to the haemophilia out of hours' service, based on the oncology ward, could be similarly stressful. In the months after her son was diagnosed, M5 ironically recalled *"the joys of out of hours"* (M5) that seemed to become part of the weekend routine as bumps and swellings needed to be assessed as possible joint or muscle bleeds. She blamed herself for these visits as she remembered how she had tried to adhere to the reassuring words of advice given by the nurse specialist on the day of her boy's diagnosis:

*"In all of it just remember that he is a boy first and he has haemophilia second. Don't wrap him up in cotton wool; he needs to learn his own limitations'. And that's always stuck with me.... much as grandparents freak out about the boys doing stuff, I'm like – 'yeah, yeah, let them go and let them get on with it'... But I don't think at that point I realised how much time [my eldest] would be backwards and forwards to hospital in the first couple of years, because we were...quite a lot. [So] everybody else wrapped my son up in cotton wool and I didn't'; when I was at work [his care] was*

*split between my mother and the child minder; Then I'd come home and I'd kind of let him be [himself]."*

Although she was not always sure about the need for treatment, delaying treatment was not an option. She recalled:

*"There were quite a few Friday nights you just go 'I'll phone the hospital' and then you'll speak to doctor and – 'Oh I think we need to see him', 'come on' [to my husband] and you'd get in the car. And you'd just be there [out of hours on the oncology ward] for hours and hours....There was the odd joint bleed, but mainly lots of soft tissue, lots of heavy bruising" (M5).*

She went on to describe how, despite trying to remain *"calm [and] relaxed"* (M5) the visits to the ward were frequently harrowing with the doctors repeatedly sticking needles into her son's skin searching for veins in order to successfully administer the necessary treatment.

She explained:

*"We'd [appear to] be very calm, when they were getting on with things [trying to find veins], but by the time we had got to the lift you could guarantee I had got tears running down my face. I remember one time that I'd stopped counting at the fourteenth attempt [to insert a cannula]" (M5).*

She described how she was so desperate on one occasion that her calm façade cracked:

*"One of the doctors said that she was going to try and do it in his neck and I did lose my temper at that point and shouted – 'Get me somebody who knows what they are*

*doing!’ They brought an anaesthetist down to get his cannula in and they did first time” (M5).*

These experiences gave her confidence when her second son was confirmed to have severe haemophilia, hours after he was born. *“I said to [my husband] – ‘well they are not going to mess with him like they did with [his brother]’. I know now, I know!” (M5)* She felt that the balance of control had returned to her and although she had been unable to fully protect her first boy she was able to use her acquired expertise in protecting her second boy, stating that:

*“The first time it’s a whole new environment, you just trust everybody and what [they] say because they are the doctors and nurses they know best. But once you have been round the block.... On the second time, I can stand up and say what I thought; whereas the first time I’d just sit there and let them do what they want” (M5).*

Haemophilia and Oncology services are co-located within the study setting. During office hours, the haemophilia team managed all care and appointments. During the weekends and evenings, the oncology inpatient ward provided urgent assessments and treatment. This co-location created a sense of close proximity between the children with haemophilia and those with cancer. The mothers talked of leukaemia or cancer as a form of emotional benchmark against which to measure their own experiences; a way of controlling how they felt. They were fearful that their anxieties around their boys’ haemophilia could be perceived as trivial by oncology staff and the mothers of children of cancer who, they felt,

were facing the greater terror of malignancy. Anxiety permeated many of the mothers' memories.

One mother described an experience on the oncology ward which left her with feelings of terrible inadequacy. M3 became increasingly upset by her son *"being pinned down and treated as a pin cushion"* when several clinicians attempted unsuccessfully, over a 12 hour period, to give treatment for a cut lip which had been bleeding intermittently for a week. Her words suggest that she feels it acceptable for a mother to feel stressed about their child's cancer but that her own increasing levels of anxiety for her son were a sign of her weakness:

*"You know it doesn't look good when the parents of the kids that have cancer are looking at you with pity!....It looks horrendous, he's just sitting in the main room covered [in blood] it's pouring everywhere; we're covered in it and he's covered in it, and he's screaming. [I was] quite stressed really, really quite stressed.... and it's difficult because you are in a ward with people whose kids have cancer and I really shouldn't be like this because their problems are so much worse than mine, but I still find this quite stressful."*(M3)

One mother admonished herself as she recalled becoming increasingly upset as her son was being prepared for theatre. She had tried to convince herself that although her experience was upsetting it could have been worse:

*"They tried both his feet and both his hands. I was just in hysterics. Then you feel embarrassed because it's the same ward and the same place as children with cancer*



*go and leukaemia and things and it's difficult because you realise that compared to that it's not serious" (M1).*

Later in the interview she talked about how she had begun to process her response to her son's diagnosis and treatment and how her anticipation of her perfect new born had suddenly been replaced by shock and the distress associated with her son having been born with a life-long condition: *"You have a child and the worst thing is if something's wrong with him. You don't really expect it" (M1).* She went on to explain how she had wanted to justify to herself her hysterical response to her son's distress as she restrained him for pre-operative treatment. She described how her attempt to minimise the seriousness of his haemophilia in comparison to cancer had only temporarily controlled her distress; but how someone else's words *"This is for the rest of his life; whereas [leukaemia] maybe not" (M1)* had helped to explain her feelings of being overwhelmed by the permanence of haemophilia.

The service provided by the oncology ward was praised by M4, whose father had haemophilia. Her previous intimate experience of haemophilia as well as being a health care practitioner may have had a protective effect which the other mothers did not experience. She would phone the ward to speak to a doctor and then, if advised to, would attend. She described her experience:

*"You speak to the doctor on call and describe the symptoms and then he will tell you whether the [child] needs to go in to check it out or not. It's mainly if he's had a big haematoma they just want to look at it and decide what course of action they should*

*take ..... "It's been a really good service because if they've told us to go in we've gone in and they've been expecting us, so they make you feel welcome there"(M4).*

She described the impact on her of seeing children with cancer on the ward and in clinic. She was conscious that the children with cancer were different in appearance to her son and, so as to reassure and prepare him she tried to explain this difference. However, what were obvious differences to her seemed to be immaterial to her five year old:

*"...When I started going onto the ward and seeing the kids with cancer and that; that upset me more than [my son's] condition. I just think there are people worse off than you anyway..... I thought [my son] might ask because a couple of times he's said 'Why have they got a tube in their nose?' And I've tried to explain it's an NG tube and things. But last time we went he didn't say anything and he was playing with another child who had obviously got [cancer] because they've obviously got no hair, and I thought he might ask me questions why they've not got any hair but he didn't" (M4).*

The mothers' experienced a strong sense of support from the Haemophilia Team and the opportunity for learning about their sons' haemophilia when they visited the CCC. This was a positive counter-balance to the uneasy relationship they had with the out of hours' service which they attended in an emergency and where their sense of place and acceptance at time felt uncertain.

## **Giving Treatment at Home and Regaining Control**

The need for frequent visits to hospital waned once the mothers were routinely administering their sons' treatment. This was achieved after they had been shown how to inject their sons and gained the required skills to administer treatment at home. Where treatment could not be given successfully through peripheral veins, central venous ports (ports) were used. Five of the six boys had a port that had been surgically implanted under the skin of their chest wall enabling reliable venous access. Only one boy had never had a port, his mother having been taught to give factor replacement through peripheral veins. Another of the boys had initially had a port but it had been removed and his mother had since become proficient in giving treatment using a butterfly giving set. Two of the mothers' partners had not been trained to give treatment; the other three partners were able to give it but rarely did so. All of the mothers discussed the freedom of home treatment; how it provided the opportunity to incorporate haemophilia management into family routine and to tailor prophylaxis to their boys' activities. However, some mothers elaborated on how home treatment brought with it a conflict of emotion as they welcomed the independence achieved, but they found the responsibility, at times, burdensome.

Some mothers approached the responsibility of managing their sons' treatment with trepidation, especially when treatment in the hospital setting had been troublesome. M1 recalled the trauma of hospital visits where administering factor was often only successful after several, painful attempts to access her son's veins. Eventually it became clear that, until his veins became more visible and easier to access, an alternative method of giving treatment was needed. She remembered discussing the need for a port with the Haemophilia Team.

*"They kept saying 'It is your decision' but I don't know if it was our decision really. He couldn't go on with them trying to poke him in veins that they couldn't find really. And I would never have been able to do [treatment]. [The nurse specialist] said 'I will eventually train you to do it'. I don't think I ever thought I would!" (M1).*

Her son's port was implanted and, over time, M1 learnt the practical skills required to give the treatment, *"Every time we went she [nurse] showed me what to do and then eventually let me start doing it" (M1).* Although acquiring the practical skills was a challenge, throughout the interview M1 recounted events which indicated that the emotional process was far more complex than the physical process of administering the treatment. She described how even now, from the moment of applying his local anaesthetic cream, she anxiously anticipates administering her son's treatment and how she imagines postponing the event. Despite the twice-weekly treatment regime and her son's relative indifference to the procedure, her feelings of guilt did not dissipate as she explained:

*"We tend to do it [treatment] in the evenings at about seven o'clock on a Wednesday and a Sunday [My husband says] 'Put his cream on' because that has to be put on an hour before and part of me thinks 'I'll just put it off.' The first time I injected him into his port it was awful, you feel guilty for doing it. I still feel like that. I'm still glad when it's over with....I mean [my son] is great, he sits, on Dad's knee, and watches telly, he even watches the needle, he just sits there.... I know he can't necessarily feel it, it's not painful, but I don't think it's nice for him. .... Maybe I have just got to get it into my head that I've got to do it and get it over and done with" (M1).*

Later in the interview she talked of how being able to administer treatment meant that the family was able to go away on holiday although this required the right preparation as they had to “...make sure we have enough stuff, tons of factor, a fridge full.”(M1). Although this was a break from home she did not feel able to relax as she had sole responsibility for giving treatment as her husband had “not been trained up to do it; he didn’t really want to!” (M1) She went on to explain the worry that accompanied this sense of responsibility:

*“I take the responsibility for the treatments. If I’m ill or anything, or if anything ever happened to me you know, I worry about who is going to do his treatments and things. It’s the responsibility, it’s a responsibility!” (M1).*

This burden almost seemed too great for her to bear but later in the interview she talked of how protective she had become of the port and it seemed that her worries could have been greater still had her husband shared giving the treatment, she confided that:

*“I think now I’m a little bit sort of [well] I don’t like anybody touching his port kind of thing! I do get really worried about infections and stuff. That’s my main concern” (M1).*

M2’s son no longer had a port but she vividly recalled how her decision to take sole responsibility for giving treatment and preventing port infections had made her the focus of her son’s wrath when he exploded with fury at each treatment time:

*“I was taught how to use the port-a-cath; how clean everything had to be frightened me. I kept thinking what if something’s mucky and I make him ill? [At treatment time] I felt as if I was a bad mummy [he saw] me coming with this [treatment] and would honestly shout ‘I hate you! He’d go running to daddy; [he] didn’t want me. It*

*was absolutely horrendous and that's why I said to him [husband], 'It's heart breaking, you need to take some of this responsibility'" (M2).*

She later went onto explain that although in some ways she had wanted her husband to share the focus of their boy's anger and the responsibility for treatment, this feeling was overridden by her concern that her husband might not give the treatment properly:

*"In a way I was kind of glad that he didn't do it [treatment] because it was my baby. I felt that I was the only one who knew how to. I would watch him do it and I would go 'you are not doing it right, don't do that'" (M2).*

With her son's port no longer in situ, M2 had become skilled in administering treatment into his peripheral veins. She explained how the ability to give treatment at home enabled her, and school, to be quite flexible:

*"We have had to jiggle about with his medication because getting up in a morning with three kids to get ready for school is just well, chaotic in a morning. So we've had to jiggle about and he has it [treatment] every Tuesday morning, Thursday morning, [on] Saturday we can be a little bit more laid back as I am not rushing around, and they are his days that he actually does PE at school..... School have been really good, if I can't get his medication done [before school] I can go back up; they've got a little room there that I can give it [treatment] in, so he is covered" (M2).*

Severe haemophilia and its treatment was part of M4's childhood because her father was affected and so from her son's early days she felt that it was important for him, now five years old, to understand his haemophilia and be involved with his treatment. She described her approach to this:

*"I explain to him that he has a condition, same as his granddad called haemophilia, which means his blood is a bit poorly and that's why he gets the bruises and has to have medicine to help. He used to think when he was younger that if somebody had a bruise that they needed treatment. 'Right you've got to have your medicine now, you've got a bruise!'" (M4)*

Her knowledge of haemophilia, as well as her health related experiences, contributed to her quickly learning how to give treatment through peripheral veins to her toddler son. *"I think [the nurse specialist] did it once or twice and I just watched, [then] I just got on with it."* (M4). Her early independence was supported by the knowledge that the haemophilia team was available at the end of the line to give advice as she explained:

*"She [the nurse specialist] is really good; she's on the end of the phone. I mean sometimes when I've had a few queries I've rang her up just to double check things. Sometimes I think 'oh he could do with an extra dose he's having a bleed.' and so I'll discuss it with her first" (M4).*

Compared to M1 and M2, when M4 was administering treatment it was a relaxed affair for mother and son:

*"We tend to do it [treatment] in here [sitting room] and he's sat watching the telly or looking at a book or something or telling me how to mix it up. I try to get him to participate because I know that he will be taught to do it when he is older. So he [is able to] connect the syringe up to the vial and push the water in and draw it back out. Then we'll put the needle in and he can even inject it himself" (M4).*

However, this relaxed atmosphere and sense of confidence was lacking when M4's husband was giving treatment. She described how although she recognised his need for encouragement their son was less tolerant:

*"It.... took him [husband] a bit of practice and sometimes he's still not quite confident. If he misses the vein, he doesn't want to do it again. Because [my husband] panics it goes onto [my son]; and he'll say 'I want mummy to do it because SHE can!'" (M4).*

M5 lives with her husband and three boys an hour's journey away from the CCC. Two of her boys have haemophilia and receive their treatment through their ports at home. With their different treatment regimens, normal schooling would have been almost impossible to sustain without M5's determination to weave treatment into their daily home routine, avoiding the disruption of almost daily treks to the CCC for treatment. She explained the timetable they had developed:

*"[One has it] on a Monday, Wednesday and Friday; [the other] is on Monday, Wednesday, Friday, Sunday but then we jump to Tuesday, Thursday so that week is an everyday job virtually.... We get them sat on the sofa and I end up with [the unaffected son] sat at the side holding his T-shirt up as well! It's a case of 'One, two, three, a deep breath in and in we go and that's it'" (M5).*

She went on to describe how she had accepted that both her husband and boys were reluctant for him to give treatment and how she attempted to accommodate their wishes:

*"Daddy can do it. But.... he will kind of dilly and dally in front of it for a bit rather than just like stick it in, 'just stick it in!' He did miss quite a few times on [one son] so he*



*now says 'Aagh, you're not doing it anymore, Mummy is.' The other week I had to be at work for half-past six so it was 'Right I haven't got time to do everything but I'll put the needle in' then dad could actually give him his factor and take it back out."*(M5)

Routine and predictability are an essential part of life for M3, her husband and son. Having a port in situ has made it possible for her to give prophylaxis at home, giving her a sense of control that she had previously not experienced:

*"Prophylaxis does make life easier, it takes a lot of the stress out you don't have the unpredictability of..., 'is he suddenly going to fall?'[Or rather] the implications aren't there if you are on prophylaxis. Everything is done on sort of the ebb and flow of his levels. We have high and low activity days we are very much a family of routine so it's treatment on a Monday and then it's playgroup because he can run around and do whatever he wants. Treatment on a Friday and playgroup Tuesdays and Saturdays we'll still do some high activity but Wednesdays, Thursdays and Sundays are the calm days so we go swimming and we play in the house and that works. Since he's been on prophylaxis there has been no need for any 'on demand' treatment" (M3).*

The ability to give treatment gave the mothers some freedom from the CCC. Their sense of independence increased further as they became confident enough to alter prophylaxis schedules to incorporate individual family activities; and to give extra doses of treatment when an injury occurred or a bleed was suspected.

## Daily Anticipation and Imagining their Boys' Futures

All of the mothers discussed the ways in which haemophilia could impact on the choices they made for their boys and the restrictions that they placed on them to keep them safe. They described how they were passionate in their desire for their boys to be treated normally by others although, as mothers, they recognised their sons' differences. Parts of M2's interview illustrated the emotional see-saw experienced by the mothers as they balanced their boys' needs against their own. They wanted the boys to be included in activities along with their peers whilst wanting to protect them from the emotional distress of being an outsider. Parties could be particularly difficult as *"There'd be a bouncy castle and I'd think 'Oh no, he can't go on that', or if he did, I had to [too]!"* (M2). Similarly they wanted to protect the boys from the physical harm that could be caused by ill-advised activity *"it's absolutely heart-breaking telling him that, with all that contact, actually it's [rugby] not really the best sport for you!"* (M2). It seemed that at times anticipation of their boys' emotional or physical pain was as disabling to the mothers as the haemophilia itself.

M4 talked about her father's positive attitude to his haemophilia *"He doesn't let anything stop him....he just carries on"* (M4) and how this had influenced her determination to ensure that her five year old son would experience haemophilia as an incidental part of him, which he could control.

*"I've seen what my dad's grown up with. I don't always think 'he [son] can't do this and can't do that.' I just think 'Let him get on with it let him be a child, so it doesn't affect him'..... If something happens we'll deal with it and carry on. I don't want him to be like, segregated, and treated different...I'm happy for him to try anything."*

*Obviously not full physical contact sports, but if he finds something he's interested in then I'm all for wanting him to have a go" (M4).*

She went on to describe how engaging her son in his treatment was also an important part of helping him to accept and control his haemophilia and had given him confidence

*"I try to get him to participate with it [treatment], have as much to do with it as possible because I know that he will be taught to do it when he is older.... So he can connect the syringe up to the vial push the water in and draw it back out. Then we'll put the needle in and he can even inject it himself. It has given him confidence" (M4).*

However she felt that her son's complete assurance that haemophilia was only a small part of his life was contingent on the staff at school being confident and not restricting his activities. She described how, when he started at the school, she had explained the condition and its treatment to the staff. They were accepting of his haemophilia but initially showed a high level of caution with this new responsibility.

*"I said to the teachers 'just let him carry on'... When he first started school they used to ring me up every day! It did settle down more as they've got used to him. I mean, we still get quite a few bump notes even if it's just a little graze. But [for emergency treatments] there is a little room, one of the teachers will sit in, they'll bring a book and they'll do a bit of reading between them and I'll give him his treatment and he just carries on!" (M4).*

M2 wished that haemophilia could be only one fine thread running through her family's life as she described how she was consumed by an anxiety for her boy which stretched from day time *"He wants to go out and play in the street, I won't let him" (M2)*, into the night when

*"He cries out a lot in the night and I think he's in pain" (M2) and on in to his future; "[I] hope that when he gets older he is going to have really sensible friends who know about his condition and who can help"(M2). She wished that his childish suggestion that she should always give him his treatment could be true. 'You're doing it mummy, you are just going to do it all the time'... I'd love it, just keeping him!"(M2). During her interview she went on to explain how her "totally, totally different relationship" (M2) with her six year old son was interpreted through his haemophilia. The middle of three boys he is the only one affected and the prospect of him having an injury-induced bleed caused her to "feel like I need to protect [him] a lot more than I do the other two; and I really, really worry about [him] all the time" (M2). However, the need to protect him, at times, led to disputes between the older two boys and between M2 and her husband.*

*"[The boys] have got this love-hate relationship! No word of a lie the oldest will say to me 'Well how come it's alright for him to hit me and I'm not allowed to hit him?'... I had tears off him yesterday because they were fighting and I told him off and he said 'You are always on [my brother's] side! He's allowed to punch me but I'm not allowed to hit him,' which he's not really. ....my husband will say to the oldest 'If he hits you, hit him back!' And I say 'How can he hit him back, you know it's really dangerous. You can't hit him back!' So that's really difficult you don't want to seem like you are just on one side" (M2).*

Despite knowing that it exposed her son to potential risk, M2 was keen for others to treat her son normally. She realised that this contradicted her own management, as she looked after him quite differently to her other two children. When he went to play with friends she found herself anticipating what could go wrong if his activities were not restricted. However

she preferred to accept that he could possibly be injured rather than him feeling that he was on the outside of daily rough and tumble with his friends:

*“He has been round to friends for tea and I have let the parents know about it [haemophilia]....I try to wrap him up in cotton wool because I don’t want him to get hurt, but I wouldn’t want them to.... I don’t want him to be treated any differently. Why should he be? .... But boys can get rough and [my boy] is so fiery, he’s got a right kick and I just think ‘Oh he’s going to end up having a bleed; it’s going to be his foot!’”(M2).*

Like M2, M5 has three boys; the older two have haemophilia. She described how her relationship with her eldest boy was slightly different than with her other two boys. She explained how her first experiences of haemophilia had caused her to “....kind of wrap [the eldest] in this ball... [we] almost suppressed him” (M5) and how she often wondered if this protection had led him to become “quite a lazy child, not at all sporty” (M5). She explained how her experiences with him had meant that she did not restrict her second son as much as she was better able anticipate problems for him, reducing unnecessary encounters with the hospital “They are not going to mess [him] around like they did with [the first] ....We were more relaxed”(M5).

She explained how being in tune with her eldest son’s behaviour meant that she was able to recognise the changes that heralded the early signs of an unprovoked bleed:

*“I could tell you, before you can see the bleed, [his] temperament, it’s just completely different. I can go ‘He’s not right, there’s something wrong,’ because it’s almost like a dark shadow on him he will just get upset and cry. If it’s taken him in such a bad way*

*he will retire to his bedroom and just lay there because he's so glum.....I can't tell [yet] with the younger one, it's not the same"(M5).*

She recounted how, despite being in tune with him she had not anticipated a recent, close encounter with rugby, considered by all those involved with haemophilia to be the forbidden team sport. Her description highlights the tensions inherent in her decision making. Having acknowledged that her son was not interested in sports his clear excitement and desire to play rugby caught her off guard; eliciting an instinctive and immediate 'no' response. M5 wanted her son to be sporty, not lazy, but there were limits to the risks she would allow him to take:

*"[He] came home from school with a letter saying that the rugby club were going to do after school rugby practice with the kids. [He] said 'Can I do it, can I do it?' [He] doesn't get excited about things but he got excited about this, then I just went 'No'. And it's the first time I've actually turned round and said 'No, you can't do that' and that did upset me.... It's the ultimate. I'm just like "'No, no, no, no, you can have a go at anything else but you are not doing rugby! Rugby is completely out, there is no way you are doing that'..... If we have a rugby player it will have to be [the youngest]! Oh yes we need a rugby player definitely!" (M5).*

M3, like M2, was keen for her two-year-old boy to be involved in normal activities, but had some reservations. She recalled how, in the months after her son learnt to walk, she was constantly anticipating injury and the need for treatment:

*"He walked early... and then it's really difficult kind of waiting 'When is he going to have a joint bleed?' And so you are kind of on tenter- hooks all the time because any*

*time there is unexplained distress you are whipping all his clothes off and examining him” (M3).*

She went on to explain that being vigilant contributed to her feeling able to give her son the opportunity for a range of experiences. She felt her vigilance reduced the risk of either significant injury or the more subtle twists and strains which might store up joint damage detectable only in the future. She explained:

*“You are trying to strike the right balance, thinking ‘What is the best thing for [my son]?’ I don’t want to deprive him of doing things that the other children are doing, but at the same time don’t want him to hurt himself. I’m not talking about the bruises that they get, they are unsightly, I’m talking about a bleed or, you know, a nasty injury to an eye or a cut lip or something that toddlers get....You have to watch everything he does because it won’t necessarily cause a problem then but it could cause a problem much later” (M3).*

M1 found herself anticipating her son growing up, when she would not always be involved in his day-to-day activities. *“With everything that he’s been through, we are that little bit closer, which maybe will make it harder when he goes [to school]” (M1).*

In the immediate period after his diagnosis she had been frightened for him and could only imagine catastrophes *“What if he has a car accident or something major happened, will he just bleed to death?”(M1);* and rejection *“At first I was worried that we wouldn’t get him into the school we wanted because they wouldn’t want him” (M1).* But with time she developed an understanding of haemophilia and its effects on her son:

*"I think if it was an accident we would probably ring them [CCC] but if it's a bleed then we know we've got to give him his treatment; ... it's strange but I suppose you are used to your child and you could tell [a bleed] straight away" (M1).*

This helped her not to fear the worst, but led her to a different concern. She talked of being worried about other people's perception of the disorder and their ability to know what to do in an emergency; this meant she was reluctant to leave others with this responsibility:

*"It has to be somebody who is aware of what's the matter with him. They would know that they need to ring us immediately if anything happened. It was difficult, I guess, the first time I had to let him stay overnight at his grandma's. I suppose I get uneasy" (M1).*

She tried not to dwell on thoughts of handing over her son's care to school; as a mother she had come to know her boy's requirements but anticipated problems with persuading the school of the significance of his haemophilia:

*"I'm starting to kind of work it through my head a little bit, how it's going to work. I know that you can explain it [haemophilia] to them and I am hoping that they will listen and be good about it. They [staff at the hospital] said 'You just tell them to call you if anything happens'....Yes, it will be difficult to leave him with people that that don't realise" (M1).*

She found herself looking far into a future in which her son carried the responsibility for treating himself; anticipating him resenting his diagnosis:



*"I'm really worried, for some reason, about when he has to start doing it himself and things and what if he forgets to do it. I feel that I've kind of burdened him for life, maybe he will feel 'Why me?' I just think maybe he will feel the way we did [at the beginning]" (M1).*

The mothers straddled the divide between feeling normal and feeling different with a varying sense of dis-ease. Sometimes they feared being apart from their boys and sometimes they worried about their futures. The mothers recognised that haemophilia had made their relationship with their boys different; simultaneously wanting them to be free to participate in activities with their friends and stay close by, out of harm's way.

### **Separateness, Isolation and Support**

The mothers all expressed an awareness of how, since having a boy with haemophilia, their relationship with others had changed *"it's very difficult to talk about it with friends because they don't understand"* (M3). Some of the mothers expressed a sense of estrangement from their established networks and of standing on the periphery of groups in which they had previously had a place. M1's sense of emotional separateness was painfully tangible as she described attending a playgroup which she no longer felt a part of:

*"I went to a regular children's play thing. I remember [after diagnosis] going a few times and looking at the other kids thinking 'I just can't believe it', that it's happened to us, and to be honest a bit angry and jealous, you know" (M1).*

M3 illustrates how her son's haemophilia had taken away her confidence, left her feeling socially uncertain and at times without a voice. She wanted to be able to include haemophilia in conversations as it was part of her life and yet did not want to draw

attention to herself. She wanted to remain within her social groups and yet was left feeling apart; she explained:

*"It's not that you lose friends, we haven't lost any friendships, it's just that you see people less and I don't think they necessarily understand why... When you try and explain it you want to explain it in a way that doesn't sound like you are wanting sympathy, because I don't, I don't want sympathy, that's not what I'm after" (M3).*

Later in the interview, she described how, during a particularly difficult time in the summer, she had felt that she owed friends an explanation for her lack of availability; but was uncertain of the required etiquette:

*"They [friends] would say 'Well how are you?', or 'How have you been?'.... We are [nearby] and we'd like to stop by.' 'It would be great but..... [he] fell over and then they couldn't get a vein and then he ended up having emergency surgery.....' And they go 'Aaagh, that sounds awful - god that's horrendous!' And you are kind of like 'Ah, I've given too much information haven't I?' But then you don't want to sound rude and just say when they want to visit 'No I'm sorry we can't fit you in'" (M3)*

Her search for the correct etiquette continued as she tried to work out how best to explain what haemophilia was. She described her pervading sense of discomfort on one social occasion when a group of parents were discussing their children and began to discuss her son's haemophilia. She was left with the impression that the other parents thought her unnecessarily bleak, confirming her position outside the group, unable to find support through these friends:

*"One [colleague] has three kids, every [one] has been easy [to look after] [He said] 'It must be terrible if he falls over and cuts himself' [I say] 'No, I'm not really bothered if he cuts himself, it's if he jars his elbow at an angle.' ....I [know] they think we are being over dramatic 'Really do you need to worry about going to the park? You are painting a very negative picture!'" (M3)*

These experiences left her feeling quite alone with no one other than her husband to share experiences with:

*"I really did get isolated because there was nobody, other than my husband, to share the experiences with and then all you are doing is just kind of sharing your own experience you are not getting any feelings that there are other people out there like us" (M3).*

M1 shared M3's sense of uncertainty in approaching conversations with friends about her son's haemophilia. Initially she had talked a lot about the diagnosis, but with time she had become more veiled and self-conscious in her approach, heavy with the knowledge that the haemophilia came from her, she felt that she should deal with it alone, she explained:

*"At first I think we talked a lot about it to other people but as we've gone along we just....Ummm..... I guess being his mother you feel like you've brought him into the world and he's a haemophiliac and therefore you should be the one that deals with it" (M1).*

She felt her sense of difference and separateness was compounded by the knowledge that she could only leave her son with someone who understood what needed to be done in an emergency:

*“It has to be somebody who is aware of what’s the matter with him.... [so] he’s only really been to my mum just overnight and my dad’s looked after him for sort of an hour. They would know that they need to ring us immediately if anything happened. I suppose I get uneasy, I do worry about what’s going to happen” (M1).*

M2 found that her concerns that an emergency could occur at any time meant that she “never got any support from [my family] but that her parents in law had “been really good, [and] offered support” (M2). Her parents in law felt able to look after her unaffected sons but it seemed that witnessing M2’s ability to administer treatment, contributed to their reluctance in looking after her boy with haemophilia. M2 felt that he would always remain a little apart:

*“For them to see him having treatment,[it] really hit home. They would [say] ‘I just can’t believe he can sit and you can do that! You are really good.’ it’s not the fact that I’m really good it’s what we have to do, this is our life now, he has to have it Our [eldest] stays [with my parents’ in law] but they will not have [my son with haemophilia] because they are absolutely petrified; it is too much responsibility. I completely understand; I wouldn’t want him to [stay with them] either” (M2).*

The sense of isolation from family and friends experienced by these mothers was countered to some degree by the support found through regular contact with the nurse specialist at the CCC. The mothers missed the closeness of their contact with her when the independence of home treatment made their hospital visits less frequent:

*“When we did see her [nurse specialist] more, she was one of the few people that I could really speak to about it, .....The only person really that seems able to say anything about what might happen in the future with him” (M1).*

But they knew that her support was readily available by telephone *“If I’m a bit concerned [an injury] might develop into a bleed then I would just speak to her [nurse specialist] for some advice.” (M4).*

As the mothers began to develop skills in recognising bleeds and became increasingly accomplished in administering the boys’ treatment, visits to the CCC dwindled and the mothers found themselves looking for other sources of support. Some of the mothers found a social media site that was specifically for mothers of boys with haemophilia a refreshing change. The fact that all the site users, most of whom were based in the UK, had haemophilia in common meant there was a shared understanding and that no complicated explanations were required:

*“The point is everybody else is having a similar sort of experience. Obviously some people have a more difficult time than others; inhibitors, target joints all sorts of things. But that’s good because it gives you some perspective” (M3).*

Virtual access to the implicit understanding of each other’s experiences and concerns engendered a richness of support that allowed for celebration and commiseration in equal measure. M5 described the importance to her of being able to share in other mothers’ joy and fears.

*“One of the mums’ son [had] been fighting his inhibitor for years, he had [had] six months in a wheelchair because he couldn’t walk. They moved him onto plasma*

*[based treatment] and just before Christmas [she] got the news that the inhibitor had disappeared. And for her to put that on there it was like 'Wow!' You just felt the absolute joy and relief for her. [She] had hundreds of postings of people going 'Yeah!' We were so happy for her.....Another one [of the mums], her boy's in hospital with a suspected port infection and she's actually put 'Oh god I hope it's a viral infection!' We all understand that. 'Fingers crossed!' If anybody else were to read that they'd think 'What?'"(M5).*

Despite the importance of support through the social media site they later discussed how they would welcome the opportunity to meet other mothers face-to-face outside the confines of the out-patient department. M1 explained how she wanted to meet others to help her as she tried to gain insight into the unknown:

*"It would be beneficial I think to speak to other mums with kids with haemophilia. Just to know what to expect with bleeds and things; so much of it is unknown" (M1).*

M2 felt that being able to share experiences of haemophilia with other parents would help reduce her feelings of being different and to feel more understood:

*"I think it would be nice for us mums and dads [to] have a chit chat and talk about experiences. Just to meet other people, to know if they've been through it, feeling the same as what I've felt. Because you kind of feel nobody can understand if they haven't got a son with haemophilia they can't understand what you have been through"(M2).*

M4 favoured a buddying system of support which would take into account both the large geographical area served by the CCC and the relative rarity of haemophilia. This method would allow mothers to support each other practically and socially:

*“That extra person who you can make contact with and say: ‘How are you doing?’  
Give each other support and stuff; have a coffee or a more formal get-together with  
people in your area. Or, phone each other and say: ‘I’m going through this problem  
have you had it before?’ ....If you are living [near] you could go round and help give  
them extra support. Help them build confidence up” (M4).*

All of the participants identified establishing strong mother-to-mother support as an opportunity they would relish. Much of their own and their sons’ lives were geared towards being normal but there was an incontrovertible difference between these mothers and their contemporary social groups. Having haemophilia as a shared norm would provide these mothers with the opportunity to develop a network of shared knowledge and experiences.

## **Conclusion**

These five themes form my interpretation of the mothers’ narratives of what it is to become and be mother to a boy with haemophilia. A more detailed examination of these themes forms the framework for the three areas of discussion in the next chapter.

## **CHAPTER 4**

### **DISCUSSION**

#### **Introduction**

The findings in this study illustrate these mothers' lived experiences of what it is to become and be mother to a boy with severe haemophilia. They talk of how being mother requires them to balance on a tightrope as they travel the divide between: Feeling and being separate and supported; feeling and being in and out of control and restricting and enabling their boys. These three interlinked areas form the content of the discussion that follows.

#### **Feelings of Being Separate and Supported**

The mothers in this study recalled the sense of anticipation and excitement they experienced during their pregnancy and up until the point they suspected that there was something wrong with their boy. Their imagined future as a mother was uncomplicated but their sons' diagnosis changed this. Like other mothers, their imaginings of motherhood were perhaps romantic and unattainable, elevated to the unachievable through representations marketed by the media and in parenting literature, reflecting little of the reality of becoming and being mother. Wolf (2005) and Douglas and Michaels (2004) discuss how the mythology surrounding perfect motherhood can leave mothers with feelings of inadequacy; a sense of being parted from the ideal. This romanticised link between the imagined and the real is stretched still further when mothers' hopes are shattered with the diagnosis of their child's life-long condition. Wright (2008) describes the point of diagnosis as being the first departure from other mothers as the diagnostic label separates mother and child from other mothers and their children.



The sense of separation described by Wright (2008) was evident in this study. Having a son newly diagnosed with haemophilia initiated a sense of being different for all of the mothers and some of them expressed feelings of devastation as M1 eloquently explained: *"I was distraught....You have a child, and the worst thing is something's wrong with him. You don't really expect it"* (M1). The mothers in this current study found that their sons' diagnosis of haemophilia gave them a solitary sense of burden, setting them apart from their families due to their carrier status for the faulty gene. Similar themes have been identified in other studies exploring the quality of life of parents of boys with haemophilia, finding that the parents' feeling of difference was intensified for the mothers; aware that they had passed on the disorder to their son (Emiliani et al., 2011, Wiedebusch et al., 2008, Madden et al., 1982). Like the mothers in Myrin-Westesson et al.'s (2013) study, the mothers in this study experienced a sense of distress with feelings of injustice that it had happened to them and it was their family affected.

The unpredictability of haemophilia, particularly before the mothers were able to administer prophylaxis at home, made it difficult for several of the mothers to access support either from friends or from the reassurance of participating in routine activities such as toddler groups. This was influenced, in part, by the practical concerns of having to respond to an unexpected injury or bleed but also by other factors such as the emotional toil of explaining to others about their sons' haemophilia and having to manage their own feelings and fears (Cohen, 1995) while searching for an appropriate reaction to their questions. The mothers' hesitance to participate in routine social activities has been noted in previous work, which found some mothers unable to discuss their sons' condition with others whilst they were feeling vulnerable; these feelings did not ease until they had

reached a position of acceptance of their boys' haemophilia (Emiliani et al., 2011, Beeton et al., 2007; et al., Myrin-Westesson et al., 2013). Described as living on the margins by Hewetson and Singh (2009), these feelings of isolation are particularly strong until a sense of a new normality can be constructed; this was also evident within this study.

The Haemophilia Team at the CCC became the first, vital source of support for the mothers in this study as, in keeping with the national standards (Haemophilia Alliance, 2006), the staff used their experience and expertise to provide the required information and treatment. However, despite the support from the Haemophilia Team, the mothers described finding a much deeper sense of support through their empathic relationship with the nurse specialist whom they described as being their primary source of reassurance and advice. Her role in the families' lives was succinctly described by M5 who explained *"She said she would always be there for us....and she is; like one of the family!" (M5)*. The reliability of the relationship with the nurse specialist was integral to the mothers' confidence in providing care for their boys and confirms the findings of other studies of parents' relationship with their nurse (Madden et al. 1982; Beeton et al. 2007). The benefit of nurse specialist support has been identified in studies of parents of other chronic conditions including atopic eczema (Cork et al. 2003), cystic fibrosis (Hodgkinson and Lester, 2002) and juvenile type 1 diabetes (Moyer, 1989).

The support of the nurse specialist did not prevent the mothers from experiencing episodes of crushing sadness, these feelings were not constant but would appear, unbidden, when their boys faced haemophilia-related challenges. These challenges included painful joint bleeds, enduring others' unsuccessful attempts to administer intravenous treatment or, when their wish to participate in an activity had to be rejected thus magnifying the loss of

‘what might have been.’ These feelings, although deeply felt, were somehow intangible and often transient and therefore hard to share, creating a further sense of being apart. Similar feelings, first described as ‘chronic sorrow’ by Olshansky (1962), have been recognised in other studies, in which parents have begun a process of adaptation to their child’s illness, only to have their sense of containment usurped by the unexpected impact of an event or interaction. As attention is drawn to their child’s difference from the norm, the parents – as in other studies - were emotionally returned to the feelings of sorrow first felt on receiving their child’s diagnosis (Webb, 2005, George et al., 2007; Bowes et.al, 2009; Kepreotes, et al., 2010).

Despite being a potential source of support only two of the mothers sought and found regular support from their own parents; the others doubted that their family really understood the impact haemophilia was having on them, engendering a reluctance to leave the boys in their care, and contributing to their sense of isolation. Some of the mothers had shared little of their daily experiences with family members as, together with their partner, they underwent a process of, what has been described in previous work as ‘internal regrouping’ (Mastroyannopoulou et al., 1997). This process required the mothers, supported by their partners, to progress away from feelings of inadequacy which were linked to a sense of being responsible for their sons’ difference (Williams and Chapman, 2011), whilst adjusting their expectations and incorporating haemophilia into family life. Although not evident from the interviews conducted with these mothers, studies have shown that in families with poor cohesion, the diagnosis of a childhood chronic illness may deepen the fissure in already fractured relationships (Mastroyannopoulou et al., 1997; Varekamp et al., 1990).

Many mothers experience periods in their mothering when they feel exposed to their own and others' criticism (Collett, 2005). At these times they feel solely responsible for their children's behaviour, health and wellbeing, a position reinforced by societal norms and expectations (Power, et al., 2011). The deeply felt sense of being responsible for their own sons' haemophilia left the mothers in this study feeling vulnerable. This vulnerability made it hard for them to access support from family or friends as they felt exposed to the scrutiny and judgement of others. These dilemmas of uncertainty are seen in other studies involving parents of boys with haemophilia (Beeton et al., 2007; Madden et al., 1982) and have also been found amongst parents of acutely, ill children, as they navigate the maze of unwritten social mores that surround seeking help and advice (Neill et al., 2013).

The concerns that the mothers experienced when needing to obtain advice or treatment for their boys were compounded when they knew they had to turn to the out of hours' service which was based on the oncology ward, where the staff were unfamiliar with haemophilia. Lack of haemophilia knowledge amongst staff has been found, in other studies, to be a key driver for parents to learn how to administer replacement factor (Furmedge et al., Yee et al., 2002). But sometimes, even where mothers were able to administer treatment they were uncertain of the significance of their boys' symptoms or how to manage them; the decision to seek out of hours' review was reluctantly made, and was done only after the mothers had assessed and reassessed their boy's symptoms. Shaw and Riley's (2008) study found the level of uncertainty and need for support so great amongst parents in their study argued for an increase in the onsite availability of haemophilia specialist staff (Shaw and Riley, 2008). In this study the boys' unscheduled need for attention from the staff on the oncology ward seemed to disrupt the rhythm and orderliness of inpatient routine, emphasising the

mothers' feeling of being an outsider; their son being different. The mothers recognised that the children with cancer and their parents faced different challenges to their own and imagined how it would be to have a child with cancer or leukaemia. They used illnesses as an emotional benchmark against which to measure their own situation and coping strategies. This benchmarking triggered them to 'count their blessings.' This cognitive restructuring (Miller et al., 2000) was not always easy, when imagining their own sons' futures. M1 summarised the feelings shared by some of the other mothers *"compared to [leukaemia] it's not serious [although haemophilia] is for the rest of his life whereas that maybe not."* (M1).

The mothers' difficulty in deciding whether a fall was hard enough or the twist of a limb awkward enough for them to seek urgent, out of hours advice was a significant challenge which has also been found in other studies of parents' experiences of managing and treating their boys' haemophilia (Shaw and Riley 2008; Beeton et al., 2007; Bottos et al. 2007). This challenge is mirrored by the dilemma faced by parents of children without long term health problems in deciding when to seek medical help for their acutely unwell child (Neill 2010). Like the parents in other studies, the mothers feared being dismissed for being over-anxious although this anxiety was matched by the fear of the consequence of not taking action in haemophilia a bleed which, if left, would cause a significant amount of pain (Shaw and Riley, 2008, Beeton et al, 2007, Wiedebusch et al., 2008, Yee et al., 2002).

Once the decision to attend the ward had been made the mothers were sensitive to the reception they received and the care their sons were given. When staff were unresponsive, unwelcoming or experienced difficulties administering treatment the mothers' confidence in their decision to attend the ward were shaken; reinforcing their sense of isolation. Similar

anxiety and uncertainty was found in a study of parents of children with sickle cell disease and cystic fibrosis (Kepreotes et al., 2007) and a large study of parents whose children were affected by one or more chronic illnesses when they experienced negative interactions or responses from clinical teams (Nabors et al., 2013). Over time, with the support of the Haemophilia Team, the mothers in this study gradually reframed their perspective on their boys' condition and developed the skills necessary to assess their sons' clinical needs and to administer their treatment; with these developments came an expanded sense of being mother; enveloping clinical expert and care co-ordinator; reducing their dependence on the out of hours service and the CCC. Other studies of parents' experiences of childhood chronic illness have described how adjustment to their child's needs and acquisition of new skills has had an enabling effect, reducing feelings of isolation and separateness (Nabors et al., 2013, Kapreotes et al., 2010, Kratz et al., 2009).

### **Feeling and Being In and Out of Control**

Some of the mothers recalled with vivid clarity the moment at which their sons' diagnosis was confirmed. The shock of diagnosis and their perception of haemophilia left them feeling disembodied; they talked of being unable to recall the other information they were given at the time. This meant they had little to mitigate the shock of the diagnosis and the lack of contextual information added to their sense of disembodiment, confusion and feeling out of control. This period of confusion, also experienced by other haemophilia carrier mothers is described as 'existential chaos' by Myrin-Westesson et al. (2013) and is evident in other studies of parental response to the diagnosis of chronic illness in childhood such as Duchenne muscular dystrophy (Webb; 2005), chronic feeding difficulties (Hewetson and Singh, 2009) and juvenile type 1 diabetes (Lindolt, et al., 2005).

The trauma of diagnosis brought intense feelings of being out of control as the mothers grappled with the unfamiliar label of haemophilia and the uncertainty of its impact on their sons and their families. Uncertainty is a feature identified in other studies focusing on parents of children with haemophilia (Furmedge et al, 2013; Wiedebusch et al., 2008, Beeton et al 2007, Varekamp et al., 1990). In this study, uncertainty also impacted on how the mothers viewed and had to review the new and unexpected responsibilities that the diagnosis brought to becoming mother. Their altered responsibilities led to a sense of role ambiguity (Berge and Holm, 2007) which had a ripple effect on other family members as all were required to shift their roles to accommodate a boy with haemophilia and a mother with clinical duties. Whilst haemophilia creates particular challenges for parents, similar shifts in responsibility and role ambiguity are also apparent in parents having to accommodate chronic illness of a different nature such as cystic fibrosis (Berge and Patterson, 2004) and epilepsy (Mu et al., 2005).

Diagnosis of a child's chronic illness is often described as being shocking for parents (Myrin-Westesson et al., Marshall et al., Webb 2005). However, although their boys' diagnosis had a shocking effect on four of the mothers in the study, the one mother with close family experience of haemophilia did not share this sense of shock; forewarned that she was pregnant with a boy she chose to control his arrival with an elective section. Personal experience of her first son's haemophilia had a similarly protective effect on M5, so that when her second son was diagnosed she felt unsurprised. These findings are consonant with those of Wiedebusch et al., (2008) who found that parents who had a son with haemophilia and a family history of the disorder had a better quality of life than parents of children with juvenile type 1 diabetes or juvenile idiopathic arthritis.

Feelings of uncertainty were felt most intensely by the mothers at their boys' diagnosis, but were re-experienced with each un-interpretable sign of their boys' distress, each injury which might initiate a bleed and these feelings were re-enforced by each unwanted visit to the out of hours' service for treatment. The intensity of the mothers' emotional response to their boys' symptoms is concordant with uncertainty in illness theory (Mishel, 1981) (as applied to parents). This theory maintains that parents find it difficult to assign meaning to an 'illness event,' such as a possible bleeding episode as they are unclear how the event, or bleed, will be managed and are uncertain of the likely immediate restrictions or long term effect on the child's quality of life (Mishel 1981, Santacroce 2003, Holm et al. 2008). Such intensity of feeling was not constant but came in waves, so that during periods when the boys' symptoms of haemophilia were in abeyance feelings of uncertainty were less intrusive with the 'humdrum' of family life providing respite from feelings of being out of control. The findings of other studies of parental response to childhood illness (Cohen 1995, Sällfors and Hallberg, 2003) show similar patterns of the heightening and respite of such feelings.

Gaining a sense of control over their boys' haemophilia was essential to reducing the risk of the mothers succumbing to the emotional and physical exhaustion known to be associated with uncertainty (Mishel 1981, Mastroiannopoulou et al., 1997, Holm et al., 2008).

Achieving control was an evolving process that required a combination of knowledge and practical skills as well as emotional and psychological adjustment.

Gathering information about a child's diagnosis, prognosis and treatment are keystones in parental adjustment to their child's illness. These measures alone do not eradicate uncertainty, but they do help to reduce heightened anxiety (Holm et al 2008, Berg and Holm, 2007). Developing the, previously unimagined, skills required to administer treatment



at home was essential to the mothers gaining control over their boys' haemophilia (Furmedge et al., 2013, Beeton et al., 2007, Yee et al. 2002). These skills gave the mothers the opportunity to weave haemophilia into the fabric of family life: the threat of urgent trips to hospital for treatment receding into the background (Furmedge et al., 2013).

Responsibility for preparing and giving the boys their treatment was the mothers' domain; as in other studies the fathers rarely, if ever, gave treatment (Coppola et.al, 2011, Herrick et al. 2004; Yee et al., 2002). Some of the mothers described this responsibility as onerous but seemed reluctant to relinquish the burden as they had concerns that their husbands were not up to the task.

Maintaining personal control of the boys' haemophilia and excluding the fathers is a powerful theme that has been recognised in other studies of haemophilia and of other chronic childhood illness (Coppola et al., 2011, Herrick et al., 2004, Swallow et al. 2011). All of the mothers in this study provided rational, functional reasons for their partners' lack of involvement: *"He doesn't want to...and I am not keen!"*(M1) *"I would watch and go 'you are not doing that right!'"* (M2) *"He is hopeless at multi-tasking!"* (M3); *"He is not confident!"* (M4); *"He dillies and dallies!"* (M5). Fathers giving treatment would mean ceding control of their boys' haemophilia to them and returning the mothers to a position of uncertainty.

The mothers recognised that there was an inherent risk to home treatment being dependent on one person, yet they remained unwilling or unable to forego their expanded role as clinical mother. This suggests that their desire to control home treatment had origins beyond the functional. Some of the mothers described feeling 'genetically responsible' for their sons' haemophilia and experiencing episodes of self-blame, guilt and sadness (Myrin-Westesson et al. 2013; Dunn et al., 2008). It may be that this genetic responsibility

contributed to their need to control their boys' haemophilia by taking on primary responsibility for treatment; in some way assuaging an associated feeling of guilt (Emiliani et al., 2011, Gregory et al., 2007, Varekamp et al., 1990; Madden et al., 1982).

Maternal gatekeeping (Allen and Hawkins, 1999) is a theory that provides an explanation for women's resistance to men's involvement in family work. In this theory the men are unable to achieve the standards expected by their partners and are required to stay within a pre-determined role which confirms the mother's position as carer and home maker (Allen and Hawkins, 1999). This theory does not provide an historical or cultural context for this behaviour nor does it explore the influence of partner expectation. However, if administering treatment is considered to be an extension of the function of maternal caring then this theory goes some way to explain the differentiated roles found in this study. The role division may have been further reinforced because the mothers received home treatment training in the absence of their husbands.

Keeping fathers out of treatment procedures and decisions has been found to have a negative effect on marital adjustment and parenting satisfaction in families with haemophilia (Herrick et al. 2004). This is contrary to the findings in this study where, although the mothers found that giving treatment could be burdensome, they considered this to be a lesser burden than coping with the concern that their husbands would be unable to give treatment correctly.

### **Restricting and Enabling their Boys**

A conundrum faced by many mothers as their children grow, is how to balance keeping their boys safe whilst allowing them to engage in activities and experiences that may expose

them to risk. It can be difficult for mothers of boys with haemophilia to decide which activities are allowable and which are out of bounds. Although the decisions that mothers make about their boys' engagement in activities seem in some ways arbitrary, with some mothers enabling their participation while others restrict them, it is likely that these decisions are influenced by societal contexts such as 'boys needing to be boys' (Francis 2012, Hays 1996). The mothers in this and other studies, such as those by Beeton et al. (2007) and Varekamp et al. (1990) found that being mother to a boy with haemophilia brought a further layer of complexity to making decisions about bringing up a boy and facilitating their engagement in activities. The potential risk of an activity-induced bleed needed to be set against the risks, particularly as the boys grew older, of inactivity or social exclusion (Williams and Chapman, 2011, Madden et al., 1982). Over time, informed by their own experiences and advice from the Haemophilia Team, the mothers appeared to develop a kind of internal algorithm against which to measure the risk associated with their boys' activities. As mothers, they navigated the course between the nurse specialist's exhortations to "remember he is a boy first... his haemophilia comes second" (M5) and the opposing mantra repeated by many haemophilia specialists: "He has haemophilia; he will never play rugby!" (Park, 2000).

In this study the mothers talked of the stifling effect their boys' diagnosis had on 'being mother' to them, particularly in the early months after diagnosis and before they were able to administer treatment themselves. Like other mothers (Myrin-Westesson et al., 2013, Emiliani et al., 2011, Beeton et al., 2007) they were aware that early signs of a bleeding event were difficult to recognise and did not always have an obvious pre-cursor. Despite some bleeds occurring without an obvious trauma the mothers found themselves restricting

their boys' activities. The mothers discussed a variety of strategies they had used to cocoon their boys from harm; from "checking and rechecking [his] body for signs of swelling" (M1) to "two layers of underlay beneath a new thick carpet" (M3) and "flattening out [bumps] in the back garden." (M4). This vigilance was supported by the mothers' avoidance of events or activities that they feared could put their boys at risk; such as toddler birthday parties and bouncy castles. These episodes of hyper-vigilance and avoidance have been described in other studies of parents of boys with haemophilia (Bottos et al., 2007, Madden et al., 1982) as well as in parents of children with juvenile type 1 diabetes (Marshall et al., 2009, Lowes et al., 2004). Keeping their boys safe through their vigilance, the mothers navigated a tricky route between being mother and clinical assistant (Berge and Holm, 2007).

The mothers went to great lengths to maintain the impression of their boys' normality; they ensured that they timed the prophylaxis administration day, which transiently shifted the boys' severe haemophilia to mild, to the boys' advantage. In the pre-school days, high activity was arranged for when factor levels were at their peak and, once the boys had started at school, prophylaxis was fine-tuned to be at its most effective on days when the boys were doing physical education. The mothers adopted tight control of 'who did and who did not know' about their sons' haemophilia as they wanted their sons to appear normal. This fits with Goffman's (1963) notion of 'passing' as normal. The mothers of the school aged boys described a process of 'selective disclosure' (West, 1986) of their boys' haemophilia. Mothers balanced disclosing the diagnosis to another child's mother before allowing her boy to 'go and play' against the alternative of restricting his social activities and keeping the haemophilia concealed.

Like other mothers of boys with haemophilia (Furmedge et al., 2013; Shaw and Riley, 2008, Yee et al., 2002) the mothers valued the flexibility and liberating effect that prophylaxis afforded them in helping to reduce the sense of stigma surrounding themselves and their boys; it helped to create a new sense of normal.

For the three mothers, whose boys were of school age, the issues of restriction and enablement were a particular cause for concern. They struggled to ensure that their boys engaged in activities with their peers but also to ensure that their boys recognised their limits and the need to withdraw from an activity when it had become too risky. Conferring responsibility to a child for managing aspects of their own health occurs in other studies of haemophilia (Park, 2000; Madden et al 1982); and in juvenile type 1 diabetes (Marshall et al., 2009). The mothers described the quandary they faced; a quandary also experienced by mothers in other studies (Park 2000, Mulder et al., 2004) when they balanced the short term benefit of their boys withdrawing from risky activities against the potential for this withdrawal to bring with it a label of 'wimpy boy' (Williams and Chapman, 2011). They were concerned that this accentuated difference (Madden et al., 1982) could cause their boys to always be on the outside of groups (Park, 2000, Evans, et al., 2000). Such exclusion from group activities would result in the boys being unable to continue passing as normal (Gray 2002), possibly their sons' first experience of enacted stigma (Quinn and Earnshaw, 2012).

The mother of the oldest boy in this study suspected that it was wrapping her son in cotton wool during his early years rather than his genetics that had led him to be a cautious 'rather lazy' boy. He professed disinterest in sport - preferring to stay at home or hang around with friends - similar problems resulting from over-protection have also been identified by Mulder et al., (2004). Although M5 was keen for her son to one day embrace some sort of

sporting activity there were limits to her ambitions for him. She was horrified subsequently when he came home from school one day begging to be allowed to accept an invitation to join the after-school rugby training sessions *"No, no, no! ...anything else...but not that!"* (M5). This dilemma typifies the internal debates these mothers experienced as they grow to understand how the effect of haemophilia reaches beyond the physical (Myrin-Westesson et al., 2013, Beeton et al., 2007). The mothers' recognised the need for their boys to obtain a group identity, sense of camaraderie, self-worth and co-operation that is particularly associated with team sport (Mulder et al., 2004) but also, despite the protective effect of prophylaxis worried when their boys went off to play that they might cause a bleed (Shaw and Riley, 2008).

All of the mothers talked of how they spent time imagining their boys' futures. Looking towards a time when the boys were able to take on greater responsibility for their treatment and choosing their own activities. One mother, who had close family experience of haemophilia, was already involving her five-year-old son in his treatment. In contrast another mother, while recognising the impracticality, considered that she would feel comforted if her son remained dependent on her, stayed at home so that she could continue to manage his activities and treatment; this view was shared by mothers in other studies (Beeton et al., 2007; Bottos et al., 2007). Another imagined her toddler son as a young adult; being angry at her for her 'genetic failing' and for being the cause of his haemophilia, the reason for him needing treatment and the reason for him not being 'normal'. These feelings of guilt and maternal responsibility have been described in other studies of haemophilia (Emiliani et al., 2011; Myrin-Westesson et al., 2013) and X linked recessive disorders (Webb 2005).

## **Conclusion**

This discussion shows how the mothers in this study have moved forward from a position of vulnerability and dependence on the staff in the CCC at the time of their sons' diagnosis to one of independence. In many ways they have been changed by their experiences as they accessed previously undiscovered strengths and skills and developed a new sense of normal.

## CHAPTER 5

### CONCLUSIONS AND RECOMMENDATIONS

#### Conclusion

The diagnosis of severe haemophilia leaves mothers in a state of shock about their sons. They are compelled to depart from their imagined way of mothering so that they can accommodate the effect this life-long disorder has on their boys and their families. This study set out to explore what it is to become and be mother to a boy with severe haemophilia. Using a qualitative approach, influenced by hermeneutic phenomenology, unstructured interviews were used to gain a deeper understanding of the experiences that face mothers of boys with severe haemophilia.

This study shows how the mothers moved forward from a position of vulnerability and dependence on the staff in the CCC at the time of their sons' diagnosis to one of greater independence. The journey to independence took time, was neither smooth nor linear but was strewn with episodes of fear and assurance; anxiety and joy; distress and relief as each mother developed their knowledge and understanding of haemophilia, its unpredictability and its effects on their family. This journey was mirrored by the mothers developing recognition that not all health care professionals were aware of the clinical and emotional needs of boys with haemophilia, their mothers and their families.

The availability of recombinant FVIII/IX, prophylaxis and the opportunity to administer factor replacement therapy in the home have brought positive changes in haemophilia care. For the mothers in this study these improvements relate to the ability to treat acute bleeding episodes without recourse to hospital visits, and greater flexibility in planning



preventative treatment schedules, allowing the mothers to tailor prophylaxis to suit the requirements of their sons and their activities.

However the reduction in regular visits to CCCs led some mothers to feel a sense of isolation, with little opportunity to share their experiences. Haemophilia related admissions have become unusual and it would seem from what the parents have said that the rarity of admissions has left ward based staff unfamiliar and ill prepared to make an assessment of or administer treatment for acute bleeds.

In many ways these mothers have been changed by their experiences as they have accessed previously undiscovered strengths and skills and developed a new sense of normal. However it is incumbent on Haemophilia Teams to address the perceived negative aspects of haemophilia care and find ways of providing support as mothers' reality change to encompass that of becoming and being mother to a boy with severe haemophilia.

### **Limitations of This Study**

When I embarked on this project I had more than twenty years' experience as a nurse specialist working with families affected by haemophilia but little experience as a nurse researcher. In my clinical practice the focus of my work is on gaining a deeper understanding of how families experience haemophilia and on providing support as they adjust to create a new sense of normal. Arguably my approach to providing haemophilia care has threads that link to philosophical hermeneutics, but this is somewhat distant to using hermeneutic phenomenology as an approach to research. Thus this research is limited by my lack of experience as a researcher and as a hermeneutic phenomenologist; my

understanding of both these tasks has developed throughout the project, supported unstintingly by Professor Carter and Dr Bray.

This study is further limited by the small number of mothers who agreed to participate in this study. The five mothers represent a third of the invited participants. All of the mothers were Caucasian and had lived for all or most of their adult life in the area; all were married to men who were in paid employment; this shared demography reduced the breadth of societal influences and social experiences.

### **Dissemination**

A short report entitled “Haemophilia Through Mothers’ Voices” was submitted in September 2013 to the Roald Dahl Different Disciplines Project, who provided funding for aspects of this project. A lay summary of the work will be given to all mothers who have contributed to the research. The results will also be shared with the CCC from where the participants were recruited. It is hoped that the project will be of interest to those who work with both adults and children with haemophilia as it offers a window to the world of being mother to a boy with severe haemophilia. The biennial Northern Haemophilia Nurses meeting would provide a forum for discussion of issues raised. The Haemophilia Nurses Association/UK Haemophilia Centre Doctors Organisation is an annual meeting attended by those who specialise in the field of haemophilia but which actively encourages the participation of those new to the speciality and those who are involved with haemophilia through their work in District General Hospitals; the results of the study could make a contribution to the greater understanding of the effect of living with this chronic disease. Finally a paper will be prepared and submitted to the journal ‘Haemophilia.’

## Recommendations for Clinical Practice

1. Out of hours' service: Out of hours' services are used most intensely by boys with haemophilia in their early years, this coincides with the time when their mothers are at their most vulnerable, lack experience, have limited knowledge and when the boys' venous access is at its worst. As in other CCCs the out of hours' service was run from an oncology unit. This means that the mothers' vulnerability was met with uncertainty by staff whose specialism is working with children who may be incredibly sick but whose needs are more predictable than those of boys with haemophilia. ***The out of hours' requirements of boys with haemophilia would be better addressed on a ward familiar with accommodating children with chronic conditions prone to acute exacerbations such as juvenile arthritis or diabetes.***
2. Nurse education: Education of medical staff is an important part of their training but often junior doctors association with a particular ward or speciality is transient. The more consistent work force over a period of time is likely to be the ward nurses. ***Ward nurses should be encouraged to engage in haemophilia education; facilitating a better understanding of the clinical and emotional needs of boys affected by haemophilia.***
3. Parent-to-parent support: The mothers move towards independence often begins in the pre-school years; this can mean that their visits to the CCC are only for routine three to six monthly clinic appointments. Despite the support of the Haemophilia Team mothers reported feeling alone with no-one to identify with or share their experiences as mother of a boy with haemophilia. The mothers talked of how face-to-face, parent-to-parent, one-to-one or group support might provide an opportunity to share experiences, offer and receive advice and tips, give

encouragement in times of difficulties and celebration when hurdles have been overcome. ***Mothers should be supported and assisted in their ambition to develop a system of mother-to-mother support.***

4. Training more than mother: Administration of treatment was largely the mothers' domain. Often because they are the most readily available the mothers were the first to be trained to administer their boys' treatment. In some families the mother was the only one to have been trained. Although the mothers appeared mostly to prefer to be in control of their boys' treatment there were potential difficulties if mothers were to become unwell or in other ways unavailable. ***The Haemophilia Team should expect and show enthusiasm for more than one family member being trained and provided with opportunities to develop the skills required to administer treatment.***

### **Recommendations for Research**

1. Expanding this study to other CCCs: A similar study to this one, which involves a larger number of mothers, could contribute to adding depth to the findings of this study. This would allow the exploration of different situations and settings such as alternative ways of delivering out of hours care and would provide an opportunity to include mothers from minority ethnic groups.
2. Becoming and being father: The experiences of becoming and being father to a boy with haemophilia are poorly researched. Qualitative research using a hermeneutic approach would serve to develop Haemophilia Teams' understanding of what it is to be a father affected by haemophilia.

3. The stories and experiences of primary school aged children with severe haemophilia. These children's stories are rarely told. An exploration of these experiences would provide an added dimension to our understanding of how it is to be a boy with severe haemophilia; this would serve to facilitate enhanced child centred care.

### **Concluding remarks**

This study has its roots in my clinical practice; from my observations of mothers and their experiences of haemophilia and its effects. The five mothers who participated in the interviews have given narrative to 'mothers' experiences and feelings; a narrative which in the past I had only imagined and guessed at. I continue beyond this research as a practitioner with a richer and deeper understanding than before.

Contemporary haemophilia care means that boys who access timely treatment with FVIII/IX therapy are unlikely to experience lengthy hospital stays, irreversible joint and muscle damage or experience severe limitations in their activities. However, this should not lead Haemophilia Teams to be complacent. The individual and collective experiences of these mothers illustrate how they continue to need our support and empathy as well as our skills in administering their boys' treatment and over time teaching the mothers to do the same.

This study has achieved its aim of deepening understanding of what it is to become and be mother to a boy with severe haemophilia.

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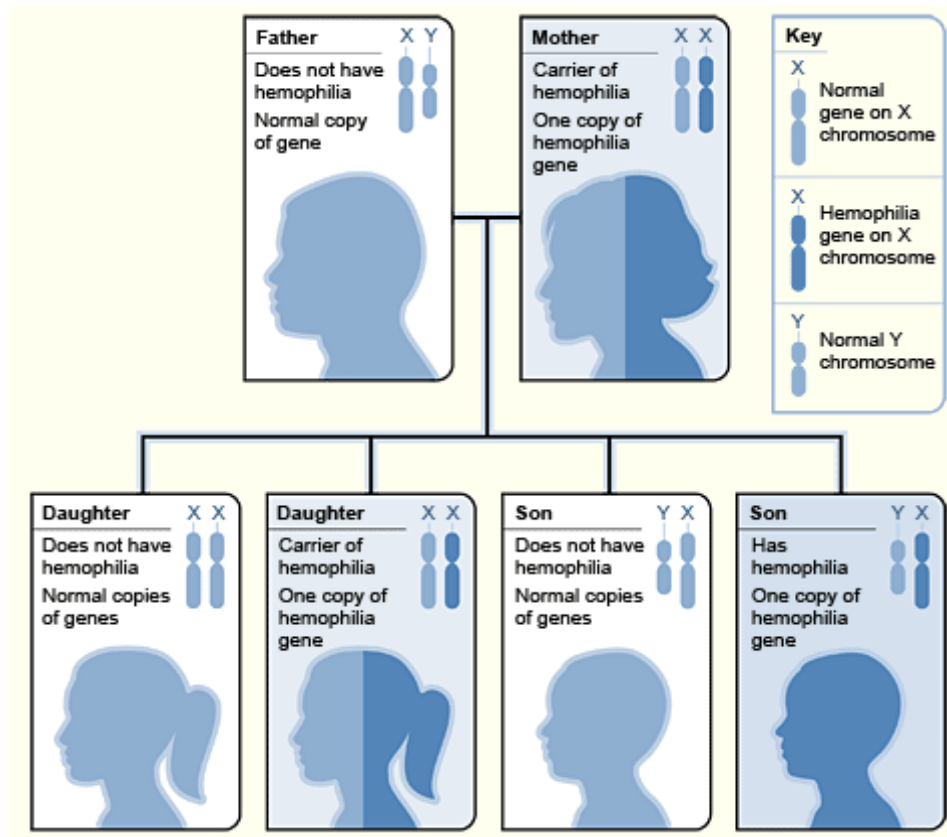
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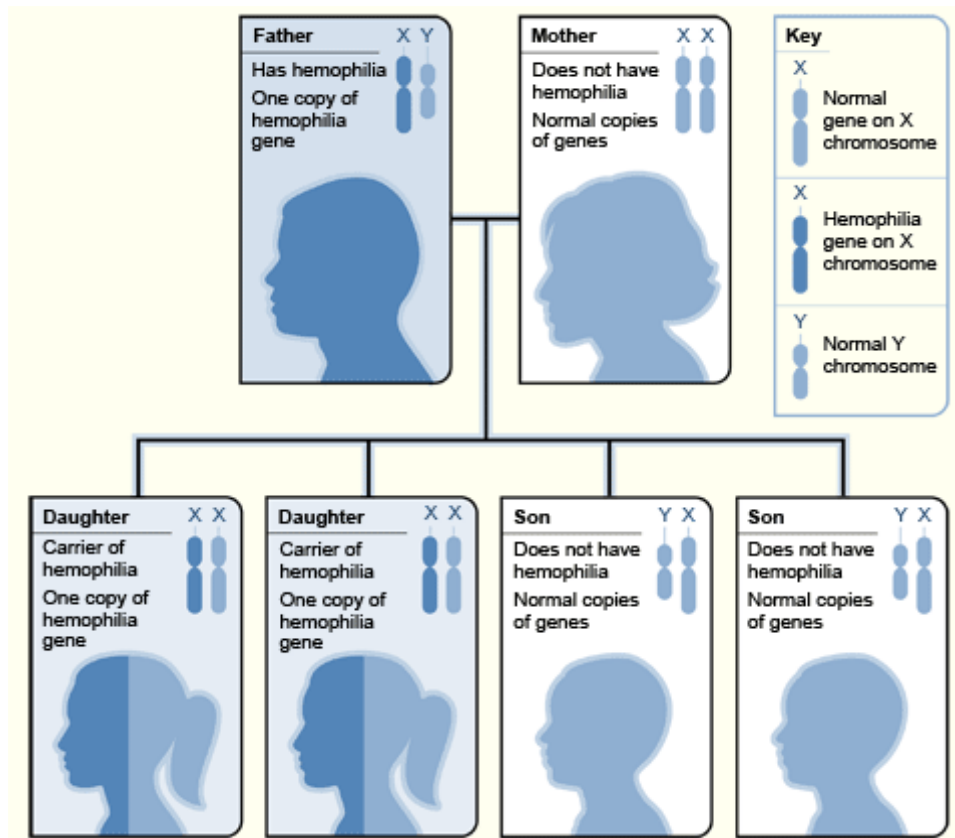
# Diagrams of inheritance pattern

## Haemophilia: Carrier Mother



NHLBI (2012)

## Haemophilia: Affected Father



NHLBI (2012)



Mss Nicki Mackett Haemophilia Nurse Specialist  
Haematology Treatment Centre  
Alder Hey Children's NHS Foundation Trust  
Eaton Road  
Liverpool L12 2AP  
Direct Dial 0151 252 5079 (9 AM – 5 PM)  
Email [nicki.mackett@nhs.net](mailto:nicki.mackett@nhs.net)

Dear

We would like to invite you to take part in our research project

**“Being Mother to a boy with severe haemophilia”**

We have asked Vicky Vidler (Haemophilia Nurse Consultant) to send the enclosed information sheet to all mothers of boys who are aged between 1 and 9 years, have severe Haemophilia A or B and who attend The Roald Dahl Haemophilia Centre at Sheffield Children's Hospital.

After you have read the information leaflet we hope that you will consider participating in this research. For further information about the study please contact Nicki Mackett using the above details.

If you would like to take part in this project please complete the reply slip below and return it in the stamp addressed envelope provided within 6 weeks of receiving this letter.

Yours faithfully

Nicki Mackett

On behalf of the research team: Professor Bernie Carter, Nicki Mackett and Dr Lucy Bray

-----  
I would like to participate in the research project:

**“Being Mother to a boy with severe haemophilia”**

**My Contact Details:**

**Name:**

**Email:**

**Telephone Number:**

**Best times to 'phone:**

**Address:**



### **Information Sheet: Being a mother to a boy with severe haemophilia**

**Name of Project Team: Nicki Mackett, Dr Lucy Bray and Professor Bernie Carter**

#### **Introduction**

Thank you for reading this information. You are being invited to take part in a project which is exploring what it is like to be a mother to a boy with severe haemophilia. Before you make your decision it is important for you to understand why the study is being done and what it will involve. Please take time to read the following information and discuss it with others if you wish. If anything is not clear to you, or if you would like further information, please contact Nicki Mackett, whose details are at the end of this sheet. Nicki works as a haemophilia nurse specialist at Alder Hey Children's NHS Foundation Trust, Liverpool. Nicki is also a student who is undertaking this study as part of her MSc by Research at the University of Central Lancashire.

Please take time to decide whether or not you wish to take part.

#### **What is the purpose of this study?**

This study wants to find out about the experiences of being mother to a boy with severe haemophilia between 1 and 9 years of age. By doing this project we hope to better understand what it is like to be a mother to a boy with severe haemophilia. This in turn may lead to professionals being better able to support mothers and boys.

#### **Why have I been chosen?**

The nurse specialist at your haemophilia centre has identified you as having at least one son who has severe haemophilia.

#### **Do I have to take part?**

No, it is entirely up to you to decide whether or not you take part in this project. If you do decide to take part you can change your mind, and withdraw, at any time up to 8 weeks after you have contributed to the research. Deciding not to take part or changing your mind about being involved will have no impact on the care you or your child receives.

#### **What will happen to me if I decide to take part in the project?**

If you would like to take part in this project please return the reply slip in the enclosed stamp addressed envelope or contact Nicki using the details at the end of this information sheet. She will then contact you to talk through the project and arrange a time and place to meet.

Nicki, the main researcher, will meet with you to hear your story about what it is like for you, to be a mother to a boy with haemophilia. The interview can take place in your home or, if you would prefer it, at a venue close to home.

We would like to audio record the interview to help us to remember in detail what you have said. You can stop the interview at any time and withdraw from the study without it affecting your child's standard of care.

**How much of a time commitment will this be for me?**

We realise that you are busy, so will arrange to meet at a time which best suits you and your family. The interview is expected to last approximately 60 minutes and no longer than two hours.

**How do I give my consent to take part?**

If you decide to take part, then before you are interviewed you will be asked to sign a consent form in which you agree to participate in the project. Before you sign the form Nicki will discuss the project with you and answer any questions you may have. You will be given the option of the audio recorded interview being kept for ten years so that short anonymous extracts of the interviews can be used at professional meetings, (such as conferences). If you agree to this we would ask you to initial a specific section of the consent form.

You do not have to agree to us using extracts of your interview at meetings. We would still like to hear your story.

**What will the research cost me?**

We will pay for any travel costs if you choose an alternative interview venue to your home. We can also reimburse any reasonable and pre-agreed extra child care costs, such as After School Club or extra hours with a child minder, which are incurred because you are taking part in the research project.

**What are the possible disadvantages and risks of taking part?**

We do not think there are any disadvantages to you taking part in this study but sometimes talking about or recalling a difficult day with your son's haemophilia can be distressing. If this happens and you would like us to, we can contact the haemophilia centre or your GP to request that you receive some extra support.

**What are the possible benefits of taking part?**

We hope that you enjoy taking part in this project. The information that you share with us could help us find out how to better support and care for other mothers who have boys with severe haemophilia.

**What happens when the research stops?**

Once you have taken part in the interview you will not need to do anything else. At some time in the weeks after the interview has taken place Nicki may need to contact you by phone to check she has understood some of the things you said during the interview. We plan to have finished the study and have written the reports within fifteen months of your interview. If you would like, we will send you a short summary of the research when it is finished.

**Will my taking part in this study be kept confidential?**

The Children's Haemophilia Care Team in Sheffield have sent out the letters of invitation and information sheets on behalf of the research team, but we will not tell them if you decide to take part in the project. All the information collected during this project will be kept confidential. Your name and other details will not be shared and will be removed in the final report so that you and your family cannot be recognised. The exception to this confidentiality would be if you were to disclose information suggesting that someone is coming to harm; this information would be reported with your knowledge.

Where you have consented to us being able to play extracts of your interview at meetings anyone who knows you may be able to identify you from your voice.



**What will happen to the results of the project?**

The results of the study will be written in a report which will be shared with Haemophilia Care Teams and The Roald Dahl Marvellous Children's Charity. The research team plans to write articles and present some of the research findings at meetings and conferences, this will help us to share our ideas with other professionals.

**Who is funding the study?**

The funding for this study is coming from the Roald Dahl Marvellous Children's Charity and Nicki's fees for her MSc have been paid for by the Children's Nursing Research Unit at Alder Hey Children's NHS Foundation Trust.

**Who has reviewed this study?**

This study has been reviewed by an ethics committee (name of committee to be inserted here) and the research committee at Sheffield Children's Hospital and the referee for the SWESH Research Degree Sub Committee at the University of Central Lancashire (UCLAN).

**Who can I contact for further information?**

If you would like further information on this study please contact Nicki Mackett  
Telephone (direct dial) 0151 252 5079; email [nicki.mackett@nhs.net](mailto:nicki.mackett@nhs.net);  
HTC Alder Hey Children's NHS Foundation Trust Eaton Road Liverpool L12 2AP

**What if something goes wrong?**

In the event that something does go wrong and you are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Alder Hey Children's NHS Foundation Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you.

**What if I have any concerns?**

If you have any concerns about the research and wish to speak to someone independently, please contact

Wendy Swann Research Manager at Sheffield Children's Hospital

Telephone 0114 271 7417 Email [wendy.swann@sch.nhs.uk](mailto:wendy.swann@sch.nhs.uk)

You may also contact Professor Bernie Carter who is Director of Studies for Nicki's MSc by Research based at the University of Central Lancashire. Email [bcarter@uclan.ac.uk](mailto:bcarter@uclan.ac.uk)

**Thank you for reading this information sheet.**

## Appendix 4: Reminder letter of invitation



**Ms Nicki Mackett Haemophilia Nurse Specialist**  
**Haematology Treatment Centre**  
**Alder Hey Children's NHS Foundation Trust**  
**Eaton Road**  
**Liverpool L12 2AP**  
**Direct Dial 0151 252 5079 (9 AM – 5 PM)**  
**Email [nicki.mackett@nhs.net](mailto:nicki.mackett@nhs.net)**

Dear

We have asked Vicky to send this reminder letter to everyone who we first invited, two weeks ago, to take part in our research project.

### **“Being mother to a boy with severe haemophilia”**

We have asked Vicky Vidler (Haemophilia Nurse Consultant) to send the enclosed information sheet again to all mothers of boys who are aged between 1 and 9 years, have severe Haemophilia A or B and who attend The Roald Dahl Haemophilia Centre at Sheffield Children's Hospital.

After you have read the information leaflet we hope that you will consider participating in this research. For further information about the study please contact Nicki Mackett using the above details.

If you would like to take part in this project please complete the reply slip below within the next four weeks and return it in the stamp addressed envelope provided.

If you have already returned your reply slip or if you have chosen not to take part in this study please ignore this letter.

Yours faithfully Nicki Mackett

On behalf of the research team: Professor Bernie Carter, Nicki Mackett and Dr Lucy Bray

I would like to participate in the research project:  
**“Being Mother to a boy with severe haemophilia”**

#### **My Contact Details:**

**Name:**

**Email:**

**Telephone Number:**

**Best times to 'phone:**

**Address:**

## Appendix 5: Consent to participate



### Consent to participate in interview

**Project Title: Being a mother to a boy with severe haemophilia**

**Name of Project Team: Nicki Mackett, Dr Lucy Bray & Professor Bernie Carter**

Please  
initial  
box

I confirm that I have read and understand the Information Sheet (version. 1) for the above study and have had the opportunity to ask questions

☐

I understand that my participation is voluntary and that I am free to withdraw at any time up to eight weeks after the interview without giving any reason and without my legal rights being affected

☐

I understand the interview I take part in will form part of the data collection for this study

☐

I understand that the interview will be audio recorded, with my permission, and that some of the things I say in the interview may be quoted in the final report or publications and I understand that these quotes will be anonymised

☐

I understand that data collected during the study may be looked at by individuals from regulatory authorities or from the NHS Trust, where it is relevant to my taking part in this research. I give permission for these individuals to have access to the records.

☐

**In addition to the above, I understand that I can also agree to the following additional element.**

**I understand that, with my permission, voice extracts of the audio recorded interview will be stored for ten years and may be used at professional meetings**

☐

I agree to take part in the above study

Name of Participant

Date

Signature

Name of Researcher

Date

Signature

## Appendix 6:

**Table 1: Example of database showing code phrase adjacent to hyperlinked interview text for Page Label: Management of Haemophilia, Hospitals**

	Out of hours service	Hyperlink: Out of hours	Difficulties with treatment	Hyperlink: Difficulties with treatment	DGHs	Hyperlink: DGHs
M1			He used to scream and scream. It just felt awful putting him through it	It was really bad, especially when they try to take blood tests from him. He used to scream and scream, he had to have it done and it just felt awful, awful putting him through it.		
M2					They had never ever dealt with a haemophiliac	I've got the phone to my ear crying, sobbing my heart out. I wanted them to mix this medicine up as quick as (possible). They had never, ever dealt with haemophiliac. Never ever mixed a treatment up for a haemophiliac!... he was still bleeding and bleeding and I'm thinking – he's going to die
M3						
M4						
M5	Oh the joy of out of hours	if something was going to happen it would be at weekend and I'd be on out-of-hours and – oh god, the joys of out-of-hours!... They struggled quite badly with his veins				