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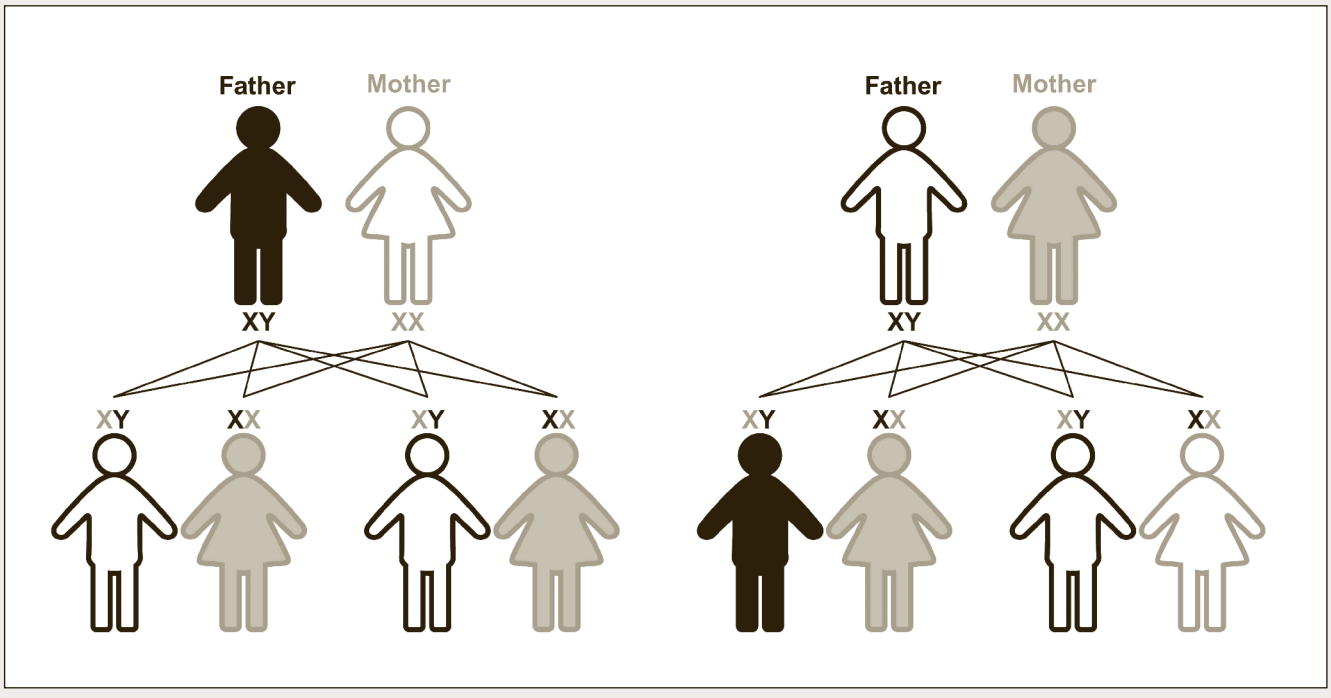
The Social Ecology of New Technologies and Haemophilia in New Zealand

A Bleeding Nuisance revisited



Julie Park and Deon York

The Social Ecology of New Technologies and Haemophilia in New Zealand

A Bleeding Nuisance revisited

The "Living with Haemophilia, alias the 'Bleeding Nuisance' research" studied the everyday life of people with haemophilia in New Zealand. The report "A Bleeding Nuisance" (1995) was the first publication of its kind. This volume focuses on the many significant changes since that time in the lives of people with haemophilia and for the community as a whole.

What is it like to have a child who is diagnosed with haemophilia and subsequently to live with this genetically inherited bleeding condition?

How are people with haemophilia and their families responding to newly available technologies, including reproductive technologies, and treatments?

This volume presents the voices of people in the haemophilia community and analyses their experiences in the context of rapid changes in technology, treatment, support organisations and the health services, and in the aftermath of the blood-product-related aspect of New Zealand's Hepatitis C epidemic. An accessible read, written for people in the haemophilia community and everyone interested in social studies of health.

The Authors

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To people with haemophilia, their families and all those who care for them.

The Social Ecology of New Technologies and Haemophilia in New Zealand

— *A Bleeding Nuisance* revisited

Julie Park and Deon York

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Abstract

This research builds on previous studies conducted by the "Living with haemophilia" researchers over the past decade in New Zealand. The current study investigates the implications of new treatments, new technologies, and changes in health care for people and families with haemophilia and those who care for them, in the context of everyday living with haemophilia. The research design used semi-structured face-to-face interviews and/or telephone interviews with 37 people, and participant-observation at a range of haemophilia gatherings. We asked people to share with us their perceptions and/or experience of prenatal genetic diagnosis, preimplantation genetic diagnosis, gene therapy, and new treatments for hepatitis C, as well as their everyday experiences with haemophilia.

The study design and the reasons we undertook it are described in the introduction. The first substantive section highlights the everyday issues of living with haemophilia as a bleeding disorder. The second discusses the organisational ecology of haemophilia. The third traverses issues concerning haemophilia as a genetic disorder, passed down the generations, and the final section explores the presence of hepatitis C in the haemophilia community.

In the conclusions we note that there are still some difficulties around the timely diagnosis of haemophilia. However, treatment for many people has changed from on-demand to prophylaxis and from the provision of blood products to recombinant products. These technologies have had significant effects on perceptions of the seriousness of haemophilia, on the safety of products, on daily living, and on relationships with the treatment sources: from products made from donations, to those manufactured by multinational pharmaceutical companies. There was a high level of awareness of the costs of treatment, compared to the earlier studies.

The formation of a National Haemophilia Management Group, which was a result of years of work between the Haemophilia Foundation of New Zealand (HFNZ), medical experts, and Ministry of Health officials, was a very welcome development in 2006. The HFNZ continues play an important part in many people's lives.

Despite a continuing emphasis on women as carriers, there is a greater realisation that men, too, pass on haemophilia, and that women can suffer from bleeding problems. Parents were exercised by the timing of when to tell their daughters about their carrier status, but carrier testing very seldom incurs the long delays of earlier years. Issues around carrying haemophilia on and reproductive choice are handled with great care in this community. A wide range of views were encountered, tempered by respect for the positions of others.

Discussion of gene therapy was a little passé in this community, as it had been on an ever-moving horizon for many years, and because new alternative treatments were seemingly offering considerable benefits. However, gene therapy was not dismissed as a future possibility.

Hepatitis C has had important effects on this community and on the individuals within it: effectively there is a hep C generation and a post-hep C generation. It was heartening that those undergoing the most recent form of treatment appeared to be experiencing better outcomes, although the treatment itself was gruelling. At the end of this research period, a Government announcement of acknowledgement, compensation and treatment was made, fulfilling a decade and a half of struggle for recognition of harm.

Chapter One: Introducing Haemophilia and the Study

Haemophilia is a rare hereditary bleeding disorder in which the ability to produce clotting factor is either impaired or not present. There are two main types of haemophilia, Haemophilia A, in which clotting factor VIII (FVIII) is missing, and Haemophilia B, which involves a reduction in clotting Factor IX (FIX). As a consequence, without factor replacement therapy, that is, infusion of FVIII or FIX, a person with haemophilia will bleed longer than usual. Cuts and external bruising are not usually a major problem; it is internal bleeding that causes pain, swelling and damage in joints and muscles. Haemophilia can be mild, moderate or severe. While in New Zealand there is adequate treatment overall, there is currently no cure for this disorder.

As an X-chromosome-linked condition, the affected gene is inherited by both men and women (see Figure 1). Because men have only one X chromosome, they express the condition, i.e., have bleeding problems. Women, with two X chromosomes, have some protection from the condition, but carry it into the next generation, as do men. About one-third of women carriers also have bleeding problems, and these are usually, but not always, mild or very mild.

A small research group based in the Department of Anthropology, The University of Auckland, has been working with people with haemophilia since 1993, researching the social aspects of living with this condition. This is part of a programme in medical anthropology. In this current study we wished to delineate what had changed and what had stayed the same for people with haemophilia, compared with the baseline study of 1994–95 (Park *et al*. 1995, Park *et al.* 1999) and with a small update study of 1999 that focused particularly on hepatitis C. (See Appendix A for a summary of the 1995 study.) Our participants included families and individuals who had been diagnosed with haemophilia since 1994, as well those who had been invited to participate in the original study. The focus of the study was on how people with haemophilia and their families understand and deal with key changes (actual or potential) in technologies for the diagnosis and treatment of haemophilia and related conditions, especially hepatitis C. This research was planned in the context of debate about the high costs of current haemophilia treatment (Carnahan 2003, Faed 2003, Harper *et al.* 2003).

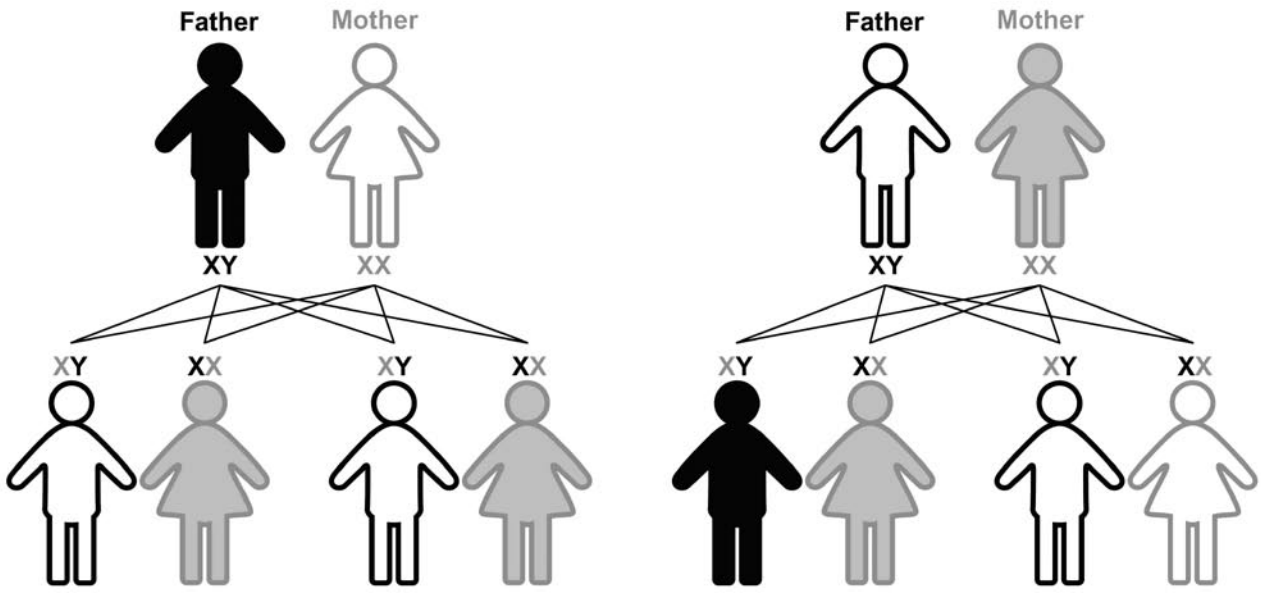

Figure 1. The inheritance of haemophilia, an X-chromosome-linked condition (Re-drawn by Peter Quin, after an image from the Australian Haemophilia Society).

The initial aims of this current research, as expressed in our research proposal, were:

- to provide trustworthy, cogently analysed information from the New Zealand haemophilia community to assist general community debate and social policy formation on the important health, social, personal, clinical and bio-ethical considerations that arise from new treatment and diagnostic technologies for haemophilia;
- to contribute to the international literature and theory development on the basis of New Zealand data; and
- to provide an overview analysis of the views, experiences and behaviours of people with haemophilia in New Zealand with regard to these new technologies, in order to assist this community, their clinicians and health planners as they grapple with the issues arising.

These aims were translated into the research questions based on our academic training in anthropology and specifically on a review of our previous publications and recommendations, our readings of relevant academic debates about haemophilia, new technologies, health-based social movements, and blood-borne viruses, and especially our observations of issues for people with haemophilia and conversations with them. The research was discussed with the Council of the Haemophilia Foundation of New Zealand (HFNZ) and received its support.

Our plan was to focus on four domains: new reproductive technologies, carrier issues, gene therapy, and hepatitis C, which are all further described below. But as befits an ethnographic approach that emphasises experience, these domains were approached in the context of a general research conversation with each participating individual or couple about living with haemophilia, and were amplified by participant-observation. Therefore the contents of this report ranges much more widely than these four specific domains.

Pre-natal Technologies and Testing for Haemophilia

Amniocentesis and chorionic villus sampling for prenatal genetic diagnosis (PND) was available at the time of our initial study in 1994. Although PND was much discussed, relatively few women who might have been carrying a foetus with haemophilia had made use of the service, and within the haemophilia community there were a wide range of views on the acceptability of this practice (Park and Strookappe 1996). The possibility of choice was enabling for some, but created severe personal distress for others. This diversity is paralleled by international studies with people with genetic disabilities (Rapp 1994, 1999). The past decade has been a time of rapid developments in new birth technologies, and these have provided several different options for people with haemophilia. For example, the technology of preimplantation genetic diagnosis (PGD) was available in Australia and was under discussion in New Zealand when we planned the current study. Use of PGD could enable people with haemophilia to avoid having daughters who carried haemophilia or sons who had the disorder. However, its use raises ethical, religious and social questions that require considerable public discussion (Finkler *et al*. 2003). Research in Sweden by Tedgård (1998) indicates that some women, especially those with genetic disorders, experience considerable psychological distress as a consequence of PND. It is likely that the newer technologies also are interpreted ambivalently and have considerable personal costs.

Testing for haemophilia is not an individual matter, as samples for analysis are also required from family members. Positive or negative individual results therefore have implications for family members, as they do with any genetic disorder (Hall *et al.* 1998). Indeed, it is common for women to find out that they are likely carriers through tests conducted on their male children. The question of when to test girls or women who are likely to be carriers (i.e., those whose mothers are carriers; the daughters of men with haemophilia being "obligate carriers") exercises people in the haemophilia community, with many parents preferring to have daughters tested at an early age so that they will "always know", as they believe that this will lead to healthier psycho-social development. However, concepts of informed consent and privacy codes formulated on an individual rights basis introduce conflicting demands and constraints.

New Treatments for Hepatitis C

The majority of people with haemophilia treated before 1986 were exposed to hepatitis C via human plasma used to treat haemophilia. When hepatitis C was first identified, there were no treatment options available. Until very recently treatment was relatively ineffective, and initially was not widely available. Consequently, a substantial proportion of those infected were expected to progress to serious liver disease (e.g., Lee 1999). There have been significant improvements in treatment options recently — e.g., pegylated interferon and ribavirin combination therapy had just become available in New Zealand as we began the study. Although this therapy has different success rates, depending on the viral strain, between 55% (strain 1) and 80% (strains 2 and 3) of people with hepatitis C can expect to clear the virus. Unfortunately, the hard-to-treat strain 1 is common among people with haemophilia (HFNZ 2004). Consequently, despite this welcome improvement, there are still issues associated with treatment for hepatitis C, and, for those for whom this has proved to be a chronic disorder, issues related to living with this infection acquired through their haemophilia treatment.

In addition to these issues concerning daily life, people with haemophilia and hepatitis C were hoping for recognition and an apology from the Crown for those who had died and for the survivors. This is a moral issue, but there is anecdotal evidence from several countries that the unassuaged grief, anger, and personal devaluation created through the bad blood experiences of HIV and hepatitis C, and the ways they have been handled by governments, are inimical to quality of life and contribute to mortality (Keshavjee *et al.* 2001).

Gene Therapy

While gene therapy for the treatment of haemophilia is not a 'cure', it offers the prospect of significantly improving the lives of people with haemophilia (Bolton-Maggs and Pasi 2003). If gene therapy were to succeed, a person with haemophilia would have a more convenient, longer-lasting treatment that could potentially maintain factor levels for a longer period of time. It would reduce the chronic disease burden and could be available to those who would not normally have access to treatment. It may be less expensive and it would minimise the risk of having blood products that potentially contain viruses. The possibility of gene therapy for the treatment of haemophilia has been widely discussed among the international and New Zealand haemophilia communities and has had varied responses over the past decade. Perceptions and responses from this community affect how gene therapy is understood and the willingness to support it, including taking part in clinical trials.

Research Design

This research was carried out by two social anthropologists: Julie Park, who has been researching social issues relating to haemophilia since 1993; and Deon York, who as a person with haemophilia has been involved with the haemophilia community for over two decades. Our positioning in relation to the haemophilia community cannot be described in a word. Both of us are insider-outsider researchers, depending on what aspects of our identity are relevant to a particular research encounter. As a man with haemophilia, Deon is 'one of us' in certain contexts; as a parent Julie is 'one of us' in other contexts. Both of us are outsiders in the sense that we analyse our own experiences and those of others in the light of our training in anthropology, and in that we convey these understandings to the non-haemophilia world.

We found that our participants were not bothered by which one of us interviewed them, and this extended to Māori participants for whom we were Pākehā researchers. In all cases, the people with whom we spoke were interested in the topic, seemed keen to talk to us, and were eager to receive feedback from the research.

Theoretically, our study was informed by interpretative approaches within anthropology. We recognise that research findings are constructed in partnership with research participants, but that the researchers have the responsibility to control and direct the research process. Ethnographic accounts are never complete and are always delivered from a position: 'partial' in both senses of the word. Objectivity, in this paradigm, consists of struggling to be aware of and to describe our positionings, being scrupulous about crafting our account on the basis of the data that we have produced, and not over-extrapolating our conclusions. Ethnographic accounts are constructed, but within the limits of the available data.

The study received a small grant from the University of Auckland Faculty of Arts Research Fund, and was approved as a multi-centre study, with Auckland Ethics Committee X acting as the lead committee. This oversight was later transferred to the national multi-centre committee, based in Wellington (see Appendix B for "Letter to participants"). Interviewing began in June 2005 and finished in March 2006, with most interviews being completed between November 2005 and February 2006. Analysis began in March and a draft report was completed in June of 2006.

Community consultation began in September 2006. As a part of this process of consultation, all of the longer quotations or paraphrases, and the surrounding text, was emailed or mailed to the individual participants concerned, who were invited to comment and to let us know of any errors or differences of interpretation. We took this opportunity to make a final check that participants consented to our use of the information that they had contributed, and we attempted to contact everyone by mail, email or telephone, who did not reply. The draft report was also sent as a confidential document to the Council of the HFNZ for similar reasons. Small factual errors were corrected, but no other changes were requested. The report was finalised in December 2006. A summary report was sent to all participants at this time. The full report was submitted for publication review in January 2007, and was accepted, subject to revisions, in July. It was revised and resubmitted in September 2007. The publications from the earlier studies are listed in Appendix C.

Information about this update study and invitations to take part were issued through the magazine of the HFNZ, *Bloodline*, the HFNZ website, and via personal invitations issued by outreach workers based in Christchurch and Auckland, nursing staff at the Haemophilia Centre in Auckland, people attending the Young Families camp in Rotorua, and members of the HFNZ National Council.

Interviews. The main research method we used was that of the semi-structured interview (Bernard 2002). Interviews were tailored to the circumstances of the participants, and covered prenatal testing, including prenatal genetic diagnosis, gene therapy, carrier testing and bleeding issues for carriers, and hepatitis C, as these topics were relevant. In most cases, the interview also included a 'catch-up' or 'getting to know you section',

depending on whether the person had taken part in earlier studies and was known to the interviewers. Deon and Julie also attended a number of meetings, camps and workshops, and had many informal discussions with a wide range of people in the haemophilia community, which provide further context for this study.

The interview outline can be seen in Appendix D. The outline was constructed by the researchers to lead to satisfying discussions around the topics of interest, as well as to provide context about the individual, couple or family. Some questions were taken from earlier interview outlines, but most were developed for this study. The outline was trialled several times before it reached its final form. However, we should stress that each interview was different and was adapted to the circumstances of the individual, couple or family, in line with the theoretical and methodological underpinnings of this research.

All of the interviews were digitally recorded, with the permission of the participants. The telephone interviews were recorded using a speaker-phone and digital recorder. The recordings were transcribed verbatim by the interviewers. There was little difference between the telephone and face-to-face interviews, apart from the rather obvious fact that verbal responses (mmm, yeah, uh ha) were used much more often on the telephone, where a smile or a nod sufficed in the face-to-face setting. The same outline was used for both.

Interview participants. A total of 35 individuals or couples indicated that they would like to be part of the study, and 33 interviews were completed. The two that did not eventuate were because of personal or scheduling reasons. Julie conducted 21 interviews, and Deon the remainder. Because four couples were interviewed, a total of 37 people participated. The relatively small number of couple interviews was a result of many of the interviews being conducted by telephone.

Thirteen participants were men: nine had haemophilia and four were partners and/or fathers of people with haemophilia. Of the 24 women, 23 were carriers; five had not had children, and the other 18 were women who were both carriers and mothers of children with haemophilia. The remaining woman was not a carrier, but was the mother of a boy with severe haemophilia. Several women had bleeding issues themselves.

Participants came from various parts of New Zealand, from Auckland to Southland, from both rural and urban areas. Most, but not all, participants had severe haemophilia in their families.

The age of participants ranged from 17 to 60-plus years, with the majority in the 20–50 age group. However, because parents were asked about their children, information about children from a few months to 16 years of age was also included. Several members of the same families participated: aunts, cousins, siblings, parents and children, sometimes living in different locations.

Participants were not specifically asked their ethnicity, but conversation revealed that four people either identified as Māori or had Māori connections. The remainder were largely Pākehā, although there were a small number of people who were born overseas.

Other sources of information. In addition to interviews, both Deon and Julie attended a number of haemophilia organised social occasions, meetings, workshops, camps and conferences. These interactions added considerable depth to our interview material, and, because we had met many participants through these occasions, they added an extra dimension to the interviews. Some people in the haemophilia community have been known to each of us for well over a decade; and to Deon for his lifetime. The many informal conversations and other occasions for sharing information that we have had with people who were not interview participants give us confidence that our findings are relevant beyond the 37 participants. Reports and newsletters relating to haemophilia (and hepatitis C), along with media attention, provided further information for our study.

Analysis. Analysis began during each interview and continued during transcription. Once most interviews were completed, both researchers discussed the themes emerging from them, which included those relating to the research questions as well as less expected themes. When transcribed, the interviews were systematically thematically analysed following standard ethnographic procedures (Anderson and Jack 1991, Emerson et al. 1995). This involved repeated reading (and listening) to the interviews, and coding them in terms of the main topics of conversation (e.g., gene therapy and prenatal testing) and the characteristics of the participants (e.g., male/female, relationship to haemophilia). Further coding was done as analysis proceeded and topics emerged. For example, quite a number of people mentioned that they had been adopted, so this was coded and explored. Concerns about costs and what we have called 'the business of haemophilia' also emerged. The themes of risk and disclosure were also discernable and coded. Information and support, the role of the Foundation, health services and treatment, as well as the ordinary matters of daily living were further coded.

The qualitative analysis program N6 (QSR International, 2002) was used to assist the analysis. This allowed considerable flexibility in exploring various hunches about the data. By using N6, we could be sure that each item of data relevant to the theme being considered was consulted while that theme was being written up. In addition, we were easily able to make enquiries such as: "Do men with haemophilia talk about prenatal testing in ways that are different from others in the haemophlia community?" We were also able to check for artefacts, such as differences between telephone and face-to-face interviews. Julie carried out

the analysis, and the results were discussed by both authors before the draft report was finalised. In a few cases, background information from earlier studies has been used to provide a little more context. Every person who contributed an interview appears in this report. Extended direct quotations from field notes are indented and appear in a different font; those that appear within the body of the text are in quotation marks.

Writing the report. In writing the report, we are very aware of the in-depth knowledge that people with haemophilia have about one another, and therefore the grave responsibility of preserving anonymity. While shared personal knowledge exists in the community, it does not necessarily encompass all of the things that we discussed with participants, and nor is this personal knowledge evenly distributed within the community. We have taken considerable pains with this aspect, using pseudonyms and also using more generic labels, such as 'a mother' or 'the husband', so that some parts of people's stories are less easily connected with other parts. We have not mentioned place names if we thought that these might help identify individual participants. This is frustrating, as it does not allow us to show the details of the excellent services provided by Auckland and Christchurch haemophilia centres, for example, or the experiences of ill-informed service that some participants have suffered in a few hospitals, or the good systems that other smaller hospitals have set up.

Although most participants were appreciative of the quality of the services they received from their specialist haemophilia centres and the HFNZ, there were some criticisms too. We have included these where possible, again trying to preserve anonymity, because it is through understanding all of these different perspectives and accommodating them that an organisation becomes stronger. Criticisms of the Foundation were made especially hesitantly, because those making them also benefited from the Foundation and appreciated its work, and often were involved as volunteers.

We have written this monograph primarily as a community report. For that reason, and to keep the volume to a reasonable size, we have limited our academic references, discussions and debates.

Historical context. The study took place under the District Health Board (DHB) health service structure. The 21 DHBs are budget holders for primary and secondary health services. If they do not provide the services themselves, they purchase them (Ashton et al. 2005:255) to service the health needs of a New Zealand population of just over 4 million. Each phase of this long-running research project has been undertaken under a different health structure, providing a basis for comparison in terms of haemophilia services.

Several significant developments were fermenting at the time of fieldwork. Negotiations were well advanced on a national funding plan for haemophilia that would overcome some of the adverse consequences of a highly provincialised health structure. However, this was not yet in place. Similarly, HFNZ was anticipating a satisfactory settlement of the long outstanding hepatitis C grievances through direct negotiations with Government. A number of people with hepatitis C were undergoing a new treatment regimen, which, although gruelling, was proving more effective than previous regimens. The Human Assisted Reproductive Technology Ethics Committee approved preimplantation genetic diagnosis (PGD) mid-2005 and funding of two cycles per couple and 40 procedures per year for serious genetic conditions, including haemophilia, was announced in December 2005. Thus this study took place at a particular moment and an exciting one, indicating once again that the experience of this genetic condition, haemophilia, is thoroughly shaped by the historical and social contexts.

Chapter Two: Living with Haemophilia: A Bleeding Disorder

In this chapter we present and discuss themes related to everyday life with the bleeding disorder of haemophilia. We start with the process that begins when parents or medical staff realise that something is wrong with a baby or young child, through to diagnosis and the beginning of treatment. The chapter follows a life course model, first with the child in the home, then moving out into the world of pre-school, school, tertiary study and/or employment, sports and other leisure pursuits. At this point we consider a number of topics that we found were important to the participants: dealing with bleeds and accidents, including the use of complementary therapies; groups with special needs; and the indirect costs of haemophilia to individuals and families.

Finding Out About Haemophilia

In every generation, approximately 30% of people with haemophilia do not inherit it from a long family line; either they themselves, or more likely their mother, has had a spontaneous mutation in that part of the gene that controls the clotting proteins. Thus some instances of haemophilia are always going to be a surprise. For three of the participants in this study, a diagnosis of haemophilia for themselves or their children was a complete surprise because they were adopted and this medical information had not been recorded or perhaps was not known. The other adopted person uncovered incomplete information about haemophilia when she was 18. For seven other people of the 16 with whom we talked about this topic, the diagnosis was also completely unexpected as there was no known family history. We did not speak to everyone about initial diagnoses, as several people had participated in earlier studies where this information had been discussed.

Known carrier mothers also have to find out if their sons have haemophilia to establish how the '50% chance' falls for them. Often this was done via cord blood testing, but sometimes it occurred as a result of an accident. For example, the baby boy (now at primary school) of one known carrier was diagnosed a few days after birth, when he became very ill due to a brain bleed. His birth had involved a breech presentation and forceps had been used.

We have selected some stories that demonstrate several of the dimensions of haemophilia diagnoses in 'unknown' families. These are all diagnoses that have occurred since our first study, but the issues are consistent with those identified 10 years earlier. The rarity of haemophilia means that it is not uppermost in most medical professionals' minds, which can lead to delays in diagnoses. On the other hand, an observant person who has seen haemophilia before can suspect it in a flash.

The steps leading up to diagnosis in those families in which haemophilia was unexpected were often traumatic and sometimes long and drawn-out. For example, Ursula's son was eight months old when a very small accident led to serious bleeding and eventually to a diagnosis of severe haemophilia. Before this, his bruises had been assessed as normal. Although they lived near a city with a large hospital, they were referred to their closest hospital first.

> … but he cut his finger and it bled and bled and bled, and then I took him to a doctor who said he must have cut an artery. I didn't think [so] — it was just a little nick. So the next time, he again had an injury and I took him to the GP and the GP said, "This time we'd better test him", so, ah, they went and did a blood test, and the person doing the blood test was quite incompetent and couldn't find a vein and poked around for quite some time, and then got somebody else to do it but in the meantime had done quite a bit of damage. By the next morning, Uri had a bleed from his arm down to his fingers, a large bleed from all the (needle) holes. So that was our first one. He was immediately admitted to hospital because he was really unwell, and unfortunately the paediatrician there didn't send him immediately to [City haemophilia centre] where he should have been, and gave him a blood transfusion, which shouldn't have happened. And that was our first introduction, it was pretty horrific! But once we were hooked into the [haemophilia centre] carers it was, it's been brilliant ever since. They knew what they were doing. They had a really good haemophilia nurse at that time, and the care we received from then on was wonderful.

Ursula's story has some similarities to that of Barbara, who lived in a small provincial centre. She, too, had taken her son (now a pre-schooler) along to her doctor with various small bleeding issues, without any tests being done until a significant bleed developed.

> It wasn't 'til he was 13 months when he had a bruise on his bottom and we were changing his nappy about 4 o'clock in the afternoon on a Saturday, and I said to my husband, "Something is not quite right here", the bruise

was tracking round underneath his scrotum, and I said to him, "I don't think that is right, something is not right." So we took him up to the Accident Clinic and from there they diagnosed, it took about three days, 'cause it wasn't showing like they thought it was going to be for haemophilia. But I just remember, too, probably about a month before that, I was cutting his finger nails, and I cut just a tiny bit of skin and he had a white stretch-and-grow on, took him up to the hospital, it looked like he'd been in a car accident, blood just everywhere ... Like on his skull, he had a lump on his head, which I hadn't got checked out because I didn't think it was that bad … he hadn't shown signs of concussion, about a week before. And they were more worried about his head and how I'd cut his finger, whether I had done something, and then when I turned up with the one on his bottom, later the doctors informed me that the nurse had actually gone, "I think we've got child abuse" … 'cause they kept asking us with his finger nail and his head, "How did it happen?", and I was more worried about his finger.

Julie: That was pretty [intimates unpleasant].

Barbara: I mean they were doing their jobs, and that's okay.

Tui had a particularly difficult time with her son, who is now of primary school age. She lived in the countryside, a long way from any hospital and much further from a haemophilia centre. The nature of her son's injury — a bleed in his brain detected at three months — as well as the delay in diagnosis, had very serious effects on him. Her son was also suspected to be a child abuse case, and Tui assumed that this was because she and her family are Māori. As it happened, it was the police investigation that led to his diagnosis and then, at last, to treatment.

Tui: With my son, we were in [a provincial] hospital for about a month and we didn't know what's happening … and when we finally went to Starship [the tertiary-level children's hospital in Auckland], they told us he probably wouldn't make it through the night. … So that night was "help"! … But to wake up and walk over there and see his little face, that was a bonus, but he was stressed and in pain, and everything. … It was all so hard. Since it wasn't in my family, they didn't really know what they were looking for. So they kind of thought that we had beaten him, done something to his head.

Julie: I see.

Tui: 'cause we were Māori. So we had the police, and social welfare were coming in.

Julie: Police and social welfare?

Tui: Mm, 'cause of his injury, and they were coming in, and they brought an eye specialist in, and it was the eye specialist that actually found out that he was a haemophiliac.

Julie: Oh?

Tui: 'Cause they say that after a blow to his head the veins behind the eye have red rings when they look at them. They are burst. But when he looked at my son's eyes, they were all good, there was none burst. And they did another blood test on him and that's when they found out he had haemophilia and they just immediately put in his portacath, his port and that.

Julie: So they wouldn't have treated him with blood products then until that time?

Tui: By then he was just bleeding, an internal bleed and it really damaged his brain while they actually found out what he had. Yes, half his brain was gone. That is the main thing. But at his age, now, the side that he has lost, his right side, the left side has compensated for it … . But he has come on a very long way.

[Although this is a phone interview, Julie had previously met Tui's son at a Foundation event.]

Julie: Oh yes, he is doing so well. It's a joy to see him, after all he has been through.

In two cases, both of which had occurred five to seven years before the interview, diagnoses of other diseases — meningitis and viral-activated arthritis — were being pursued before haemophilia was diagnosed. A lumbar puncture to test for meningitis caused a damaging bleed into one baby's back, which alerted the doctor to possible haemophilia and the baby was diagnosed without delay. The diagnosis for the other baby was more delayed. First, the local doctors saw the bruises as just normal for kids, but a recently trained locum spotted something amiss on a repeat visit for a swollen knee. Getting the diagnosis involved considerable trekking by the parents from their rural home to various hospitals, but eventually fluid was aspirated from the baby's swollen knee at a hospital where there was a haemophilia specialist, blood was detected in the fluid, and haemophilia quickly diagnosed. This diagnosis, when it came, brought relief — "Oh, that's good", the mother reported herself saying when she heard it — because she had been worried it would be leukaemia.

Being in the right place at the right time led to several speedy diagnoses. For example, a little boy was listless after a caesarean section where forceps had been used. He was born in a large city hospital with a haemophilia centre nearby, and was diagnosed within two days. The diagnosis of Carolyn's little boy was not quite so quick,

but he was diagnosed within a week, mainly because of a sharp-eyed nurse:

> He was born on a Wednesday, and by Sunday he was really, really ill. He got taken to the hospital, from one hospital back to the neonatal unit in the ambulance, and then it was Sunday and I think it was the Monday or the Tuesday they started talking about haemophilia. I think it was about the Tuesday that I think they actually diagnosed him, because one of the nurses had put a line in on the Sunday and she noticed it was still weeping. ... And when he was born he had quite severe bruising on his head, which was just, obviously continued to bleed, and sort of four days later he became quite ill.

One participant still did not have a complete diagnosis for her son, who initially was found to have von Willebrand Disease (vWB) inherited from his father when he was about a year old. Some months later he was found to have Factor VIII haemophilia as well, and possibly something else. This was clearly a very difficult diagnosis to make, and our participant explained that so far the geneticist had been working with DNA samples from about 15 family members and it was taking months for a complete diagnosis to be made. She had found the diagnostic uncertainty upsetting and difficult to deal with, especially because it had not been possible to establish a routine treatment process, and she did not know what the future held for her son.

Nita: He's looked after fairly well when he is in hospital, although with the diagnosis and tests and that, they just don't seem to have any idea of why he is like he is, which is really frustrating, because I can't sort of [sighs] … It's hard, that side of it is really hard, because I need to know, if you like I need to be able to put him in a [diagnostic] box so I can understand exactly why and what to expect from him, but I can't do that yet.

Julie: Yeah, so that is really a lot of uncertainty there for you.

Nita: Very much so. I feel like my life has been put on hold until I really get him sorted and we work out exactly what is going on with him.

Discussion

It would have been very pleasing if this update study had shown that the problems of diagnosing haemophilia were a thing of the past, and especially that birth-related head injuries no longer occurred. One of these injuries was avoidable, in the sense that the mother's carrier status was known and the breech presentation was known. The protocol is for all carrier women to be treated at birth as if they have bleeding problems themselves and as if their baby has haemophilia. For known haemophilia carriers, a head scan of baby boys rather than precautionary treatment has been accepted as standard practice. In the other cases of spontaneous or unknown haemophilia, it is difficult to see how these incidents can be avoided, when haemophilia is so rare at the population level. Prompt detection via head scans and haemophilia testing, and timely treatment, plus the actions of alert staff or parents are obviously crucial.

Most of the stories of delayed diagnosis involved an initial phase of the parents repeatedly taking their child to a general practitioner (GP), private accident clinic or a hospital Accident & Emergency (A&E), or having discussions with the Plunket nurse about unusual bruising, but this did not trigger a referral to a paediatrician or haematologist. Clearly, there is some educative work to be done in this area around best practice. But, with the exception of Tui's son, once the child was referred on, even with the wrong provisional diagnosis, it did not take long for the correct diagnosis to be made, although, as noted above, invasive diagnostic procedures can also be very damaging.

Tui was by no means the only person whose child was identified as a child abuse victim, but she may be the only one where that 'diagnosis' may have significantly delayed her son's haemophilia diagnosis during the one-month period when he was in a smaller hospital. Paradoxically, this allegation probably ultimately saved her son's life once it was investigated in Starship. Tui assumed in a very matter-of-fact way that the suspected child abuse, and hence the delay in testing for haemophilia, was related to her family being Māori. There is a disturbing implication here about the role of ethnic stereotyping in health care.

Dealing with Haemophilia

In our initial study, *A Bleeding Nuisance,* our main aim was to describe living with haemophilia as it affected the different dimensions of life: family living, education, employment, leisure, community activities, and health and health care. In this update, we did not systematically explore these dimensions as our focus was elsewhere. However, in talking with people who had participated in earlier phases of research, and in meeting new participants, we inevitably talked about how these issues were for them and their families and talked over significant events in the previous few years. The same challenges were there, but with some new twists. Newly diagnosed families had a great deal to learn; other families usually had updating to do, and partners had to acquaint themselves with information about haemophilia.

Beginning Treatment. Typically, boys with severe haemophilia were not put on prophylactic treatment until about age two — in part, so that their patterns of bleeding could be observed, because some people with severe haemophilia have few spontaneous bleeds,

whereas others quite frequently have spontaneous heavy bleeding. In addition, delay in starting treatment was thought to protect against the development of inhibitors (antibodies to treatment). All of the younger people with haemophilia were currently treated with recombinant clotting products, which are genetically engineered, and the Haemophilia Foundation has worked with haematologists, drug companies and Pharmac (the national drug-purchasing agent) to make this possible. Recombinant products have almost completely obviated safety issues around viral contamination of clotting factor products.

Many parents experienced this period of the first two years as being difficult, particularly once their baby had started moving around and was bumping into things or falling, as they had to make the judgements about whether treatment was necessary and they were apprehensive about trauma with no prophylactic cover. Those few participants who had mild familial haemophilia, or girls with bleeding problems that were equivalent to mild haemophilia, were always in this situation of having to decide on the seriousness of the injury because they used on-demand treatment. With more severe haemophilia, once prophylaxis was instituted, this new routine had to be normalised into family life, but it was usually experienced as much less stressful as it provided the child with a degree of cover which greatly reduced the number of spontaneous bleeds and provided some protection from accidents.

Not too long after prophylaxis had started, and sometimes simultaneously, home treatment usually began. This generally involved one or both parents becoming competent in sterile procedures and intravenous access, or in accessing the portacath, if one had been implanted. There were regional differences in portacath versus peripheral venous access. Sometimes other family members were enlisted to help with treatment, such as a grandmother who had treated her own son. Occasionally, until the parents were able to take on the task themselves, a nurse did home visits, or a shared home-hospital or doctor's surgery arrangement was made. One mother, whose own mother was already good at giving treatments, did not want to do them herself. She wanted to be the one who held her son's hand while his grandmother gave the treatment. Her son had a number of health problems and she "has to do all the other bad things". Another family with a pre-schooler opted to take their son to a clinic once a week and have a nurse visit the other times. We learned that the father of one of our women participants, a man with mild haemophilia, had recently learned to do home treatment himself, in his 50s, because he wanted to travel overseas to take part in a sporting event. As a person with mild haemophilia and good access to treatment at his local health centre, he had not felt the need to do this before.

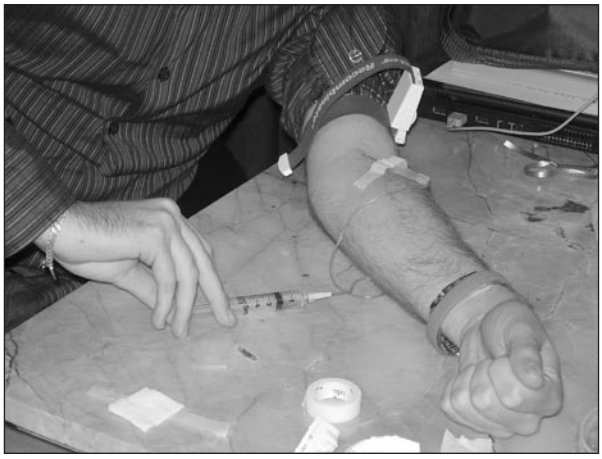

Figure 2. Home treatment of haemophilia with replacement clotting factor.

About 10% to 20 % of all those on treatment products develop inhibitors or antibodies to treatment. This generally runs in families. Sometimes they can be successfully treated with Factor VII, but a process called "tolerisation" is carried out if at all possible. This is preferably done when the child is young, while their body weight is low and before they have joints which have had repeated bleeds (target joints). But sometimes older children who have missed out on tolerisation when young are also treated. The idea is to overcome the inhibitors that prevent effective treatment. Tolerisation involves the infusion of large quantities of clotting factor over a concentrated period. It requires large amounts of treatment products. One of the young boys in this study had had a serious reaction to one brand of synthetic Factor VIII, went on to another brand of Factor VIII, developed inhibitors, and subsequently went on tolerisation. Unfortunately, it was not successful and so he began on yet another recombinant product. This is an unusually complicated situation. In earlier studies, we met several children for whom tolerisation had been successful and they were having effective prophylaxis with normal recombinant clotting factor as a result. But even without inhibitors, dealing with haemophilia is no picnic.

Ursula was a sole parent of a pre-schooler and did not have other family members to call on. She lived rurally and learned how to administer treatment herself.

Ursula: Now he has prophylaxis treatment every third day, with quite a high dose so he doesn't have it every second day. That seems to work best for him. He doesn't like it too often and I do it at home. So it's really working well.

Julie: And does he have a portacath?

Ursula: No, I do it into the vein.

Julie: How about you learning how to do it?

Ursula: Well, I was a needle phobic, so it was horrendous for me. Now it's fine. Initially I used to have heart palpitations and it was

just a nightmare. I hated it really, and I hated it because I initially wasn't very good, and sometime we didn't hit the vein and it would be unpleasant. Now it's pretty well one go, and so it's not [too unpleasant].

Julie: And you learned to do that at the haemophilia centre?

Ursula: The haemophilia nurse showed me how to do it. So I think that you really just have to. The horrible thing is you have to practise on a live person really, to get competent. But he's been unbelievably good and has been tolerant. Very, very early on he figured out that to sit still and nicely was the way.

Ursula prepared a treasure hunt after each treatment to help keep the experience positive. Even though treatment went so well, it took about two hours on treatment days to get through the whole process in an unhurried and unstressed way, from waking up, putting on the local anaesthetic cream, waiting, doing the treatment, waiting, then getting on with the rest of the morning routine.

When Tui was in Starship with her son, she had one day of instruction in home treatment, via a portacath, and after that, with a little help from her local hospital, she and her mother were on their own, although the outreach worker was only a telephone call away.

Tui: We had a crash course [laughs] in hospital. As soon as he got it [the port], and they found out what it was, the next — the day before we were coming home — some nurses from the Haemophilia Foundation, they used to be nurses, and we spent the day with them and they actually showed us how to do it on that puppet thing. Oh no, I've forgotten his [the puppet's] name, and after that, we — yeah! one day! — and we more or less taught ourselves after that.

Julie: Goodness me, that's a big deal.

Tui: Yeah, at the [local] hospital, the nurses a couple of times they'd do him, and they helped us, the sisters and that, so we had a couple of lessons in there, and then home James!

Julie: And then you were flying solo! Wow! That's quite a learning thing … And who else is there at home, in terms of adults, to help you with all this?

Tui: I actually do it myself now he is older, but my mum and myself used to do it. My mum used to hold him down when he was a baby and I used to do it. But now he helps himself get the stuff ready, and he's got a portacath, and I saw all the kids at the camp, it was so neat, doing it themselves, and, and I thought, "Oh, I hope one day you learn how to do this."

The little boy mentioned above who was not sucessfully tolerised needed treatment every day for a long period. In addition, his veins were poor and he did not tolerate peripheral venous access well, because of many trips to hospital. Both of his parents, his mother's brother and her father had learned to give treatment through the port. This provided back-up and allowed the parents the occasional time-out. They said that when they go on to three-days-a-week treatment it will seem like a holiday.

Even when parents became competent with delivering intravenous treatment and there were no other difficulties, it was not always plain sailing. Both Ivy and her husband were trained to give treatment through a port, but then a haematoma developed and the port was eventually removed, and Ivy had to learn to access her boy's veins. She practised on family members, but, compared with their veins, her pre-school son's veins were very tiny.

Ivy: It was awful. For a start-off, it started off quite good. I don't know, he was really good for a start-off, 'cause we started off doing it twice a week in his port and once in his arm, sort of thing, and then we went twice in his arm and once in his port, and then we weaned him right off and were just doing the port once a month, because we had to flush it. He was quite good, but once his port was actually gone, he sort of changed. And, I don't know. Maybe, they said, maybe he thinks his safety net has gone, and he was really naughty, I don't know if you would say naughty — screaming, yelling, kicking. It was awful.

Julie: It must have been pretty tough.

Ivy: We started going to the hospital again, which isn't very far to go [in their own town], it didn't really matter. Then they started coming out at home, then they started me doing it at home again and slowly got him back, and me building up my confidence again, you know.

Other parents, too, reported their children becoming upset when they changed from port to veins.

By the time children are in primary school, most parents had the whole process down to about half an hour. But even competent parents could lose their confidence after one or two bad days of being unable to get access and, as one couple pointed out, the children do not spare them criticism if they make a mistake and do not get into a vein on the first try, demanding that dad do the treatment if mum has not done well the last time, or vice versa.

There is no set age when children start to do their own treatment. Often the process is quite gradual, with them taking over more of the preparation and tidy-up as they get older, and, when they and the parents are ready, giving themselves the whole treatment, initially under

parental supervision. By intermediate school age, many children are able to treat themselves. The treatment room at camps where haemophilia nurses, outreach workers and often a haematologist or two are present, as well as other families doing treatments on their youngsters and people treating themselves, is often the place where a child learns the finer points of accessing peripheral veins or portacaths, and indeed where the relative merits of the two kinds of access are discussed or silently observed.

Towards Independence. Introducing the concept of haemophilia to the day-care or pre-school, and later to school, was a challenge that most parents seemed to take in their stride. They were assisted in this by printed and video materials supplied by the Foundation. In many cases, the outreach worker or Haemophilia Centre staff were available to provide back-up information via a telephone call. Many parents commented on how helpful the school or pre-school staff had been. If parents did not feel confident in the institution's ability to keep their child safe and respond appropriately to any accident, typically they moved their child to somewhere where they felt he or she was better protected. Parents took safety and the nature of relationships promoted in the school into account when they chose where to send their child.

At the Young Families Camp and in several interviews, several parents joked about how their children seemed to have inherited 'the monkey gene' along with haemophilia. By this they meant that kids with haemophilia seemed to be particularly energetic, always climbing, jumping, running and generally tearing around at breakneck speeds. It was rare to find parents who described their child as "more of an indoor boy", although such children did exist.

Despite being careful about the choice of school, parents still had concerns which arose from their child's inevitable realisation that things were a bit different for him or her than for other children. Julie had asked Ursula to look ahead and talk about her hopes and fears for Uri and her.

> He is not going to be able to do things that other children do in terms of contact sports and trampolining, so I am expecting, because he is a very strong-willed, confident child, that adolescence is going to be a nightmare. That's what I'm expecting for the future. But at the same time, it will get — it's got — easier and easier in terms of his capabilities. He doesn't hurt himself so much, he's so much more competent and that makes it a lot easier. He isn't aware that he is different from other children at the moment, because he isn't different. It will only be when he goes to school and can't do the contact sports, I think, that he will really know, so I'm not looking forward to it.

It was at this stage of life, when the child was outside the home for some or all of the day, that the now ubiquitous cell phone came into its own. All the school had to do was to dial the number or send a text message and a parent would be within earshot. This, of course, was a new development since our initial study, and it has provided parents with a greater sense of security when they are away from their landlines, and has allowed mothers, especially, to be more mobile.

Getting a good education is highly valued in the haemophilia community. This time, apart from tertiary students undergoing hepatitis C treatment, there were no stories of long periods of missed schooling as a result of haemophilia. Students were encouraged to get tertiary qualifications and many did so. In fact one of the very rewarding aspects of this update study was to hear about and meet so many well qualified young people working in a wide range of professions, including some in the general area of sports and fitness.

By the time the boys are in their teens, most have become very competent at self-treatment. Over time, with new preparations, the treatment time has reduced significantly. Trish described how she no longer knows which mornings are treatment mornings for her son, Ben, as he is so quick with it.

> We certainly noticed it, going from plasma-derived to recombinant products, because the recombinant, you're only taking a small amount, 10–15 mls, whereas prior to that we were doing 40–60 mls. You actually had to draw it up in several syringes and the whole giving, it took half an hour as opposed to the five minutes it takes now. Occasionally I'll say to Ben, "When's treatment day this week?" Or "When was the last time you had treatment?", and often he'll have had it that day and I just haven't noticed, it was sort of so quick. But he is very independent with it now.

Leaving home can be a significant time for young people with haemophilia, because, in addition to all the other things that they are now responsible for, they have their treatment, getting their supplies, having regular reviews, and so on, to think about. Some young people would have taken over these responsibilities while they were still at home, but there was always the backup of mum and dad. Albe, who had been living independently for several years, talked about his active approach to staying healthy. He was on prophylaxis for severe haemophilia.

Albe: Yes, that's the one. … I do about 1000 units every second day, and that's a high dose now. I'm on a much higher dose now than when I was young. I recently, about two years ago, looked at the CSL — Commonwealth Serum Laboratories — recommendations for my weight, and I was on far too low for a severe haemophilia,

so I upped the dose as they suggested and it's been good. See then I was doing more less [frequently], now I'm doing less more. So really, what I am doing is trying to keep that sine curve [of the clotting factor levels in his blood] from going too low, too high, so there's going to be smooth.

Julie: Every two days is quite often, so how do you find it fits in with everything else you have to do?

Albe: See the whole thing with it is it's the beginning of a normal life: treatment. Without it, my life isn't so normal. It's ah … I think I put up with it more when I was younger, the bleeding and the pain. I don't have as much time for it now, I think that is where, as you get older, you don't have as much time for a lot of nuisances. [Laughs.] … Mum's not there, Dad's not there. I have to be more able — to put it simply — so that means, look after yourself and prevent any possibilities you have to knock yourself back.

Sports

Although there is definitely a much wider range of sports on offer now for children, the popularity of contact sports has not waned. Not all children with haemophilia want to play them, but when they do it does create a difficulty for the parents, and the child. These days, the Foundation can provide parents with a helpful book written for New Zealand conditions on sport and exercise for people with haemophilia (Joll 2005) and there, in the list of sports to be avoided, are some of those closest to the hearts of kiwi kids, including the various forms of rugby.

Art and Anna are very pleased that their son has really taken to swimming, helped along by the Foundation's swimming programme. They have a covered heated pool in their small town, so it is a year-round activity. Their son has well muscled joints and has been able to play cricket in summer without any problems. In winter, occasionally rugby becomes an issue:

Art: Well, it is to the extent that being in a small place. Probably more so last year, because last year was the first year the boys were registering in his class for rugby, six-year-olds.

Anna: Not all of them.

Art: Son was saying he is the only one who didn't.

Anna: No, no, no. [Another couple of boys did not.]

Art: I knew it was a big deal when that triathalon happened. The Friday before that was when they registered for it [rugby], and he was chivvying me all weekend and I was saying you won't be able to go and do that, you won't be able to play rugby. And he made the school triathalon the Tuesday and did the swimming length for his team and got them well up in the places from where they were, and the others were saying "Good swim" to him, and he suddenly went from being on this lower plane of not being able to do that stuff to being [gestures a hero], so he was back in with all the boys. "I don't have to play rugby 'cause I'm back in" sort of thing.

Art and Anna are aware that a bit of rugby goes on at playtimes and that their son has taken part, and got a bruised eye as a result. Playtimes are a source of anxiety for many parents, including for another mother with a "rugby mad" little boy.

Ivy: And we've never ever — My husband, he used to play rugby when he was a boy and he watches all the rugby on the tele, but we've never pushed him towards it, but then we have never steered him away either. But for him to pick one sport that he really wants to do, for it to be rugby! It could have been anything, you know.

Julie: So how are you handling that?

Ivy: Oh, all right. He knows that he won't be able to play rugby and we bought him a whistle and said, you know, "You can be the referee", but I do know that in the playground at school, that a bit of rugby goes on, but you can't, there's nothing really you can do about it, you know what I mean.

Julie: No, there isn't much.

Ivy: 'cause you don't want to single him out and say, "You, sit", you know. You want him to sort of be normal, but he can't be normal, you know what I mean?

It is no surprise that this child chose rugby. In addition to her husband's rugby-watching, Ivy explained that from age eight in the winter at his school all the girls play netball and all the boys play rugby and they are hooked into a district-wide competition. Their community, too, is fixated on rugby, Ivy reported: "We've had some people say to us, 'I don't know how I could handle it if my son couldn't play rugby.' Well, we both think 'Well, so — it's not the end of the world!' " Hearing this, it seems that nothing much has changed since our initial study (Park 2000), at least outside of the main centres.

Sporting activities can also cause some angst for older boys and men, and for women with bleeding problems. These activities are very important to them in creating a meaningful life, a point which some feel is not sufficiently appreciated by their clinicians. One young man had started with a semi-contact team sport as a little boy and was still playing at a high level in his later teens. His mother explained:

We don't make a big deal of the fact that our son is in sport, is playing in a tournament, things like that. We just kind of keep it to ourselves,

> and obviously, if there was a big trauma and he needed to be hospitalised, they'd soon find out, but we prefer to just keep that under our hat. I mean he says that he plays [the sport]. I don't think they are aware of the level that he is playing at. So we've had to be a little cautious about what info we share with the haemophilia centre …
>
> Generally I think it's actually kept him very fit and he loves it with a passion, Julie, he just loves it and to deprive him of it would … Again it's the benefit thing, and it's just what he loves doing. I would much prefer him to do something safer, but that's his passion and everyone needs to have their passion, but he's coming to the stage, and I've actually said it for the last couple of years, "I think this will be the last year you'll play because of getting a bit older." I've said to him, "I wonder if you are going to thank me at 30 that I let you play [this sport] in your teens."

A man who was about 30 was equally passionate about his water sport. He had never been interested in rugby, he ironically confided in a near-whisper. He wore a helmet, but he felt that his haematologists disapproved because they were concerned about trauma. But trauma was not a major worry for him; rather it was the health of his joints which his sport helped. He just loved it, it made his life enjoyable and had enabled him to give up both drinking and smoking, generally live a more healthy life, and be in a better mental and spiritual state. He said that he wished his doctors condoned it, but he would continue anyway.

Bleeds and Accidents

Despite good treatment and prophylaxis, accidents do occur and bleeds happen that need more than prophylaxis to treat. Because so many people are on home treatment, nearly everyone has products at home, even people with moderate or mild haemophilia who will need only occasional treatments, so that a telephone call to the haemophilia centre and/or haemophilia caregiver and extra home treatment is all that is required along with RICE (rest, ice, compression, elevation). This means that emergencies involving rushing off to hospital generally now occur only when something quite major has happened — for example, when one little chap was hit over the head by a kindergartener with a hammer, or a couple of other children broke their legs, and some older ones had serious sports-related injuries.

Listening to the women with significant bleeding problems talk about their experiences, however, was quite another story. An older woman who was also a keen sportsperson seemed to have had rather hit-and-miss treatment. Not surprisingly, she was reluctant to come forward for treatment, after getting hepatitis B after one blood transfusion and hepatitis C after another treatment. She, too, had sustained joint damage. A young woman, with Factor IX haemophilia, also a sporty type, had sustained a whole raft of accidents, causing bleeds which she mainly treated with arnica. She was more particularly troubled by extraordinarily heavy menstrual periods, which left her very anaemic, and she had had enough of being on the Pill with its risks and side-effects. Eventually, with the help of a doctor at Family Planning, she found a satisfactory solution which involved an IUD device which delivers a very small dose of hormone directly to the uterus and so is much less risky than the birth control pill that most carrier women with menstrual problems use.

One young mother of a boy with Factor VIII moderate or mild haemophilia and an expressed carrier daughter had been and still was active and sporty. She had not known that haemophilia affected women until very recently. She had difficulties with many of her joints, had arthritis, and menstrual problems that earlier had been mainly controlled by the Pill. She had just learned about DDAVP and tranexamic acid by attending the Young Families Camp and was going to discuss their use with her doctor. Although it was too late for her to protect herself from any damage, she could prevent future damage and had learned about it in time to help her young daughter avoid serious joint damage. The following conversation took place shortly after she had realised, through a haematologist's presentation, that girls and women with low clotting factor levels have the equivalent of mild haemophilia.

Noeline: My daughter has a clotting factor level of 20%.

Julie: Oh, okay, she's got more or less mild haemophilia, then.

Noeline: Yes, she basically has. That's what they consider 'symptomatic carrier'. Basically, she is a haemophiliac, but because it is not so well known in females then they call her that.

Julie: Yeah, what do you think about that?

Noeline: I think it sucks! And she does too. Just because it's kind of understanding, people don't think that females can be haemophiliacs. We didn't know until my daughter was diagnosed. … That kind of makes things a lot more difficult, because we don't have the support that we need, and even people within the Haemophilia Foundation themselves don't really recognise us, so I found that a lot of the camp was great, but I didn't find out a lot of the stuff that was more important to us.

Julie: Mmhmm. And what sort of stuff would you have … do you have an idea of the sort of stuff you would have liked to have found out?

Noeline: Yep. I would have liked to have found out information that I can pass on to my daughter

> when she is getting older, like basically avoiding some of the mistakes that I made growing up, with some of the sports and things, like what ones to avoid, and to give her the education of what might happen when she starts being old enough to get her periods, and I mean she is always covered in bruises. Yes, how do I know when to give them [her daughter and her son] treatment? And do I do them here at home, or do I rush into the hospital? I can never tell. They are always constantly covered in bruises and I don't know whether I am meant to take them to hospital, or anything. So that kind of stuff would have really helped me a lot.

Complementary Therapies

Several people had found the use of complementary therapies very helpful. Arnica was commonly used for bruising. Several people also mentioned various forms of massage and touch therapies as helpful in relieving pain and assisting muscle development. Many people with haemophilia require physiotherapy at various times. The group of children who had had head and central nervous system bleeds while babies had particularly rigorous physiotherapy programmes in which the parents were, of course, heavily involved. Some parents found that they needed to remind the physiotherapists that their child had haemophilia, or otherwise a bleed could be started.

One mother of a child — now adolescent — with mild/moderate haemophilia had weighed up the 'two evils' of treatment with its risks of blood-born infections versus damage and pain from bleeds. She had decided to treat him with the conservative RICE formula and herbal remedies, about which she was knowledgeable. She herself had needed treatments on only a few occasions but had contracted both hepatitis B and C, so she had good reason to be wary. When her son had knocks, and even when he had teeth out, his bleeding was able to be controlled with the herbs. He has entered adolescence with no joint damage and is active in various forms of water sports. Interestingly, one of the haemophilia nurses mentioned that she felt that rest, elevation and playing quietly for a few days after a bleed, which used to be standard practice, was not used as much as it could be to assist the control of bleeding even with the use of blood products.

Groups with Special Needs

In this update, there were four girls and women with haemophilia — the so-called symptomatic or expressed carriers — whose symptoms were experienced as significant, and several others who had some problems but did not rate them as very important We also found that a couple of women who had had bleeding problems initially attributed to haemophilia alone had since been diagnosed as having vWB, as well as lower factor levels. One of the four with significant problems had been involved in earlier phases of this study, but the other three were interviewed for the first time and included a mother and daughter who both had low factor levels, even though the familial haemophilia was at a mild level.

The inaugural 2005 Young Women's Workshop Weekend was for all young carrier women, and this included some with significant bleeding problems for whom there was a special session. This was definitely seen as helpful, but not everyone could attend and none of those whom we interviewed with significant problems had attended. There was an observable degree of isolation among the women with bleeding problems, especially the younger ones, during this study as there had been in earlier phases. An electronic discussion group managed through the Foundation may be useful in addressing this, as more and more people have email access, or could be assisted with this, and it does not require everyone to be in one place at one time.

Something similar might also be helpful for Māori members of the Foundation. Tui, for example, had no idea that there were other Māori people affected by haemophilia. In fact, she had thought that it was an exclusively Pākehā problem, and her son was the only Māori boy with it, until a few years after her son's diagnosis she saw an article about Māori with haemophilia in *Bloodline*. Her son was at primary school when she attended her first camp. There she was delighted to meet several other Māori families, including some longstanding and staunch Foundation supporters with young adult sons with haemophilia, and some other newcomers. At the camp she heard about various Māori families with haemophilia who denied it "because it was not a Māori thing", and she was keen to talk with them about her experiences to see if that would make them more accepting of the various forms of help available. A Māori study, led by Māori members of the haemophilia community with the support of the HFNZ, was in its initial planning stages in 2007.

Indirect Financial Costs of Haemophilia

Parents of children with haemophilia incur considerable indirect financial costs and sometimes it has cost them their employment. One sole parent, for example, had expected to return to her managerial job after taking maternity leave. Instead, she found herself being a full-time mother of a child with severe haemophilia, living on the domestic purposes benefit. She also had to make a decision about where to live, leaving her own house in an area far from a haemophilia centre, and relocating in the rural outskirts — because she could afford the rent and enjoyed the beauty — of a large city. Although her son was nearly five, she had only just received her full entitlement of supplementary payments from WINZ for a child with a disability and this had taken a struggle to receive. She found the benefit just enough for day-to-

day living, but she was unable to replace any household equipment and was concerned about her car, which is essential safety equipment for anyone with haemophilia living rurally. She was planning to retrain once her son was at school, so that she would be able to find a job nearer to where they lived.

Another mother, also a sole parent at the time, had twice tried to return to work after having her baby, but each time she had had to resign when her son had a bleed and needed time in hospital and recovery time at home. Although she found WINZ helpful, it was very difficult for her to manage financially.

Many of the partnered women with children had part- or full-time work, which, as one said, allowed her to keep her sanity, as well as contributing to the family income. In one household where the mother had part-time work, they also experienced hardship because the father was a contractor and was paid only for the time he could work. They lived in a small town several hours' drive from their nearest hospital, and their son's condition required several trips in the first few years of his life, some involving several days away. There needed to be two adults in the car. Eventually, they got into a situation where the husband could not afford to take any more time off work, so the help of the husband's mother was enlisted. Despite being grateful to her mother-in-law, it was very difficult for the mother not to have her husband's support during these emotionally gruelling hospital stays, and there came a point when she needed professional counselling to help her deal with the whole situation.

In another situation where the wife worked part-time and the husband full-time on a salary, he used up well over his annual allocation of compassionate leave, because not only did they have their boy with haemophilia, but their little daughter became ill and subsequently died. The large public organisation in which he was employed allowed employees to donate their compassionate leave to people who needed it, so he did not have to take unpaid leave.

People with haemophilia, too, sometimes have to adapt their employment to be manageable with their condition — for example, take a desk job which might not be so satisfying or as well paid, rather than a job which involves lugging heavy pieces of equipment around but is more commensurate with the person's training. The desk job could be done with or without a bleed, whereas with the other job more time off would be necessary. As noted earlier, the choice of occupation in the first place is also often made with haemophilia in mind, and education is stressed to give young people a choice of jobs.

Chapter Three: The Social Ecology of Haemophilia

In Chapter Three we focus on the interactions between people with haemophilia and the organisations that assist them. We begin with a discussion of the health service and the experiences that people with haemophilia have had with it. This is followed by a consideration of the direct costs of haemophilia to the individual and the nation. Haemophilia is big business. How this impacts on individuals and the community is discussed next. A major mediator between individuals and families with haemophilia in New Zealand, and between them and the wider world and services providers, is the HFNZ. The Foundation's many roles are outlined. One of its major activities has been the effort towards ensuring a national service for haemophilia. This was achieved at the end of 2006, after our study was completed. Another major focus of effort has been the hepatitis C claim, which is discussed in the final chapter. Finally with an eye on the future, we look at how people with haemophilia understood the new science of gene therapy and what they thought about it is presented.

Interactions with the Health Service

Regular reviews are part of the treatment plan for people with severe haemophilia and other people with haemophilia who have bleeding problems. These reviews usually took place every six months for children, and annually or every two years for adults. Young children tend to have more frequent visits with the haematologist and/or paediatrician. Several people in the South Island were grateful that the Christchurch haematologists took clinics in regional hospitals from Nelson to Invercargill, which greatly reduced the difficulties they would otherwise experience in getting to their appointments. These arrangements involve sharing care between different DHBs and the specialists located within them. There is obviously potential for communication difficulties and so the nature of the teamwork has to be carefully established. While some people described teething difficulties while the organisation of care got underway, the people we spoke to were generally happy with their care. Some people who were closer to Dunedin Hospital by-passed it to obtain service from Christchurch, whereas others attended Dunedin.

In the North Island, the main centres for service were the Wellington and Auckland city hospitals, including Starship Children's Hospital. Waikato Hospital, located in Hamilton, provided a limited service, because the haematologist there was in private practice and did limited clinics (despite the very large central North Island area that Waikato was responsible for). A service was also provided from Palmerston North that covered Wanganui and Taranaki, but we did not have any participants who regularly used that service in this study. As well as the haematologists, paediatricans, and the rare paediatric haematologist, haemophilia nurses, where they existed, provided a crucial service. They were the main contact people, and were especially crucial to new families. There was considerable disquiet expressed about the Wellington service when the experienced nurse left, and alarm at any plans to reorganise the service and remove or reduce that speciality in any hospital where there were haemophilia nurses. In this update study, most participants were cared for by either Christchurch or Auckland, with just a few from Wellington, Dunedin and Hamilton.

Below we introduce some examples of the experiences and arrangements of people living outside the main centres for haemophilia treatment. Anna lived over two hours' drive from her son's treatment centre, in a small town with a district hospital nearby. Apart from when he was diagnosed, her son had only been hospitalised to have his portacath inserted. When Julie arrived at their house for the interview, the son (aged seven) greeted her at the door, and asked what she was going to talk about. She said "About haemophilia", and he pulled up his tee-shirt to show his port, remarking in a very everyday sort of a voice that he had haemophilia. Anna was very positive about the medical support available in this rural area. "We've got great support up here. Fantastic GP, great support staff at our medical centre, all medical centres here — we can go to any one of them, basically." When her son started day-care, their haematologist rang the day-care centre to discuss any issues they had, but other than that, and reviews, they have needed very little extra help. They can telephone their haematologist at any time.

Not everyone living rurally is so fortunate. At a haemophilia camp, Julie spoke with an older woman whose husband had haemophilia. They also live about two to three hours away from their haemophilia centre. On a recent visit to his local hospital with a mouth bleed, her husband was asked by the doctor attending him how long he had had haemophilia. This question is a typical alarm bell as it indicates the doctor has no idea, so she rang her outreach worker for advice, and was advised to ring her haematologist who asked them to come in immediately. Her husband was given clotting factor treatment. In another area, a woman took her son in to A&E with a joint bleed and was told to come back when

the bruise appeared — another reason for a telephone call to the haemophilia centre. A further example was of a mother who had to attend her local hospital several times with her son before she was permitted to see the paediatrician and start treatment for him. A final example was of a boy who had a leg injury that caused acute pain. He was certainly treated for haemophilia at the nearby hospital, but the mother knew that a bleed would not cause serious pain so quickly. However, the person treating her son would not consider an X-ray, so she drove quickly to her own hospital 100 kilometres away where an X-ray revealed a fracture.

At the camp for new families, there was a wide-ranging discussion of treatment issues, during which people who lived near smaller hospitals described their experiences and also the systems that they had put in place, usually with the help of a staff member from a haemophilia centre or an outreach worker. In some hospitals the agreement was that the parents would telephone ahead and go straight to the ward. In others, because of protocols about infection control or perhaps security, they would telephone ahead and someone would come from the children's or haematology ward to meet them in A&E. In others, there was a letter of agreement between the hospital and a haemophilia specialist. When a hospital was identified as a trouble-spot, the haematology clinical staff present worked out with the family how to deal with it, such as a haematologist's authorisation to have the child identified as a Code 2 on the triage system, meaning he required immediate, specific, treatment. As a haemophilia nurse said, parents have to be assertive, explain that they know what they are talking about, and "be prepared to bring in the big guns". This means that as well as all the work on national protocols and particular arrangements for individuals with haemophilia, the cell phone number of a haematologist or haemophilia nurse is a necessary tool in the work of getting effective service for haemophilia.

One youngish man who was quite happy with his treatment regimen had difficulty expressing what he thought was a limitation with the current haemophilia service in New Zealand. He did not appear to have an entirely trusting relationship with his clinicians. He had seen men younger than himself with poor joints and felt that the treatment was not aggressive enough to preserve good joints. He recognised that there were haematologists working on a whole lot of projects, but no one, he felt, who was wholeheartedly interested in the "guys with haemophilia": how haemophilia affects their whole lives. This contrasted with his experience in Australia where he had lived for several years from his late teens: "and I was a kiwi as well, you got to remember that! [laughs] — but it was 'There's a haemophiliac, get him', really aggressive treatment. It was good." This treatment was focused on maintaining healthy joints. He was the only person to raise as an issue what he perceived as the absence of a "champion for the guys" with haemophilia from within the haemophilia service, someone who took a holistic view of living with this condition.

However, similar sentiments about the need for a holistic view surfaced more obliquely in other discussions, especially in relation to haematologists' disapproval, or anticipated disapproval, of young guys undertaking some of the moderately risky outdoor activities. The clinicians' focus on the risk of possible trauma was seen as too narrow. On the other hand, sports-people themselves were aware of the costs that extra treatments could incur, and this introduced a certain wariness in their approach to their treatment centres.

Direct Costs of Haemophilia

The cost of clotting factor replacement products — and especially the constant reminders of how expensive they are — was a bugbear mentioned by many participants, who pointed out that it was not as if they had any option but to take the product. While it is informative for people in the haemophilia community to know the costs of the different treatment options available — currently plasma-derived and recombinant products cost much the same — the mention of cost every time a prescription is required has a negative effect on the caring relationship for many people with haemophilia. Cost of lifelong treatment is certainly one of the factors that several people noted that they think about when contemplating having children — a major change from our earlier study (Park and Strookappe 1996).

Treatment costs, and how to organise existing treatment plans to provide maximum protection for sporting activities, are also at the forefront of parents' minds and those of adults with haemophilia. The costs are so high that once people hear of them they do not need further reminders. In fact one person indicated that he had extended his review period to reduce the number of times he was told how much his treatment — or, as he put it, "I" cost: not a helpful outcome. He had gone from annual reviews to "about every two years".

Albe: Now I am often reminded of how much I cost.

Julie: Are you?

Albe Yes.

Julie: In what contexts?

Albe: At a review. Yes, "I'll give you this much and we know that that will cost about this figure." "Well, I know that, doctor! [irony and laughing] What about the knee?" I'm not proud of it, I hate it and being reminded of it, I really resent that. I used to think "He's doing his job", but why bother? He knows that I know, maybe he doesn't know that I know, but what are you going to do? Silence.

The parents of a young boy with haemophilia revealed that they felt "quite embarrassed" about how much their son's treatment cost — "that we are such a drain on the health system because of the costs, I struggle with that a lot". The mother who was a nurse often thought about "how many knee ops" you could do for the price of a year's haemophilia treatment. However, they could see the enormous difference that the treatment made in their son's life and in the lives of the other children. Even so, they often sat on a bleed 'to see' before treating immediately, in order to conserve the product. They did not feel pressure from their haemophilia service to under-treat, however. It was something that they had internalised.

Children under two years were generally not put on prophylaxis, and parents understood this to be partly because their patterns of spontaneous bleeds could then be observed and also to prevent the development of inhibitors. However, some thought that it was also to save money.

At an earlier stage of his life, Albe did under-treat because of his awareness of costs. But now:

> So I got to the point a few years ago: so, if you do treat less and you are putting up with it in terms of pain, and there's disability — stop it, just stop it. It's just ridiculous. You treat as much as you have to, to get you a normal life.

Nadine was concerned that her brother was not getting the amount of factor he needed as often as he needed it, because of the cost.

> I think it is available a lot more now, but I still don't think … it's still not enough. Like recently, my brother was put on daily treatment and he was getting a lot better, feeling a lot better, but they're cutting him down again and you can definitely tell the difference. I don't know the full story, but I think it is a problem with the amount that he is using that will … they can't give that much to one person, but I don't know, I mean it costs a lot.

Participants who had been involved with haemophilia for a decade or more noted that the emphasis on costs of treatment in the clinical context was a new feature. Five or six years before, we were told, clinicians did not talk about the costs of treatment. These comments about the new emphasis on cost were made in several different areas of New Zealand, and were not confined to one haemophilia centre or clinical. Albe noted, on a return visit to Australia, that a similar change was apparent there.

These observations coincide with our own experience when in 1994–95 clinicians were worried about the budget blow-outs due to haemophilia treatment, given the emphasis then on the business model and bottom lines in the health sector as a result of the recent health 'reforms'. However, with limited exceptions, they did not share these concerns with their patients, except in the context of prenatal testing. Our suggestion at that stage was that it would be helpful if people in the haemophilia community in general were knowledgeable about the costs of treatment, rather than finding out that it was shockingly expensive just at the time they were making decisions about starting or extending their families. If it were common knowledge, then it could be more easily integrated into decisions about family planning, as well as occupational choice and choices about leisure activities. The pendulum seems to have swung to the other extreme, so that now people who are dependent on a lifetime of treatment are reminded seemingly at every turn of how much 'they' cost. Yet the benefits of good treatment are everywhere apparent as young people with good joints enter adulthood.

People who worked in the health sector noted that this emphasis on cost was pervasive, and in some areas of health it meant that patients did not have access to the most effective drugs or treatments in the interests of saving costs. They pointed out that the emphasis on cost in haemophilia treatment was not unique, but what was different was the very high cost of the standard haemophilia treatment that everybody needed, and needed over their lifetimes. Many people understood that there was a 'top-down effect', with budgetary constraints and anxieties about overspending being passed down the whole health sector hierarchy until it impinged on managers of haematology services and clinicians.

Compared to haemophilia treatment, costs for other health problems tended to fade into insignificance. This was very apparent when people were talking about the costs of PGD. Making several cycles available to couples carrying haemophilia who found the procedure acceptable was rated as much more cost-effective than limiting the number of funded cycles to just two, because if the IVF were not successful, the couple might subsequently have a boy with haemophilia who would need a lifetime's treatment.

A DHB employee had jokingly told her sister who had a boy with haemophilia that it would be cheaper for a DHB to buy a family with haemophilia a house in another DHB area than to treat the family. This last scenario is, of course, not just about the perceived expense of haemophilia, but also about a health service that divides up a country of only 4 million people into a complicated mosaic of funding territories which then have to cope with the random but clustered nature of an expensive rare condition like haemophilia. Haemophilia has the capacity to bankrupt a medical service in one DHB, whereas the neighbouring DHB may have no people with haemophilia within its boundaries. This is the situation that the national treatment plan, described below in the section on the Foundation, addresses.

Despite the well-recognised cost of haemophilia borne by the health budget, there are still some areas where costs of treatment that have to be born by the patients cause hardship. The first example bears out Noeline's perception that women with bleeding problems are still something of an anomaly. In the case described by Frances, below, this was not through any deliberate policy but because she was probably the first person with haemophilia to use a particular, relatively new treatment which had been designed for women with different health problems. Consequently, haemophilia was not listed by Pharmac as one of the conditions for which they would fund this device, which Frances, who has Factor IX bleeding problems, has now used successfully for over five years.

Frances: Mirena, which is an IUD device. Back then it wasn't sponsored by Pharmac. And yeah, they gave that to me, and [prolonged, heavy periods] completely disappeared. I have one or two [bleeding] days in the month. It was a godsend! And that was five years ago and it expired in September of this year, so I went back and my outreach worker from the Haemophilia Society [Foundation] helped me to have a little argument with Pharmac and get them to approve the funding line, 'cause it wasn't in the Pharmac rules to help women with haemophilia, it was only to help women with haemoglobin and feratin. And so my outreach worker finally got the Pharmac people to approve it, and I was told I had to go down to [a hospital 150 kilometres away] to have it done and I objected to that because I would have to take a day off work and my partner would have to go with me to drive me home, 'cause there is generally a little bit of discomfort after it. And so they finally … there is a specialist from [that hospital] that comes up here every month and he did it about two weeks ago.

Julie: And how is it going?

Frances: It's fine, back to normal. But the two months I had between September and November, when it expired, they went back to being how they were, yeah. But it would be nice if there were an easier way to do it. 'Cause that costs $330, if it is not funded by Pharmac, plus I've got a bill of $190 for the gynaecologist's consultation which I am going to claim on my health insurance or try and get Pharmac to pay for.

A quite different example relates to a family whose son required frequent visits to the main city hospital about 30 to 45 minutes' drive away. Their son was quite young and was much less stressed if one parent could sit with him in the back of the car. It was just not possible for both parents to take so much time off work, so several days a week the mother found herself fighting her way through heavy traffic, trying to find a car park at the hospital, and simultaneously trying to calm and reassure her son. Their social worker eventually suggested they take a taxi, saying that all they needed to do was to collect the receipts, which they did. However, when it came time for reimbursement, once they had spent $500 on taxi fares, none was forthcoming because "Your consultant has not approved a taxi."

Although there were some other examples of this nature relating to costs of accessing treatment services, and confusion about entitlements for refunds or other assistance with associated costs, this theme was by no means as marked in this update as it had been in our first study.

A third example related to PGD. One couple had determined to fund PGD themselves after several years of letter-writing and lobbying officials to try to get PGD approved by the ethics committee and funded for people with haemophilia. They had found the umbrella organisation for people with rare disorders, the New Zealand Organisation for Rare Disorders (NZORD), extremely helpful and supportive. Eventually, they ran out of patience and, after ethical approval had been granted, they opted to go ahead. However, while they were still undergoing preliminary tests, the funding of two cycles per couple approved was announced, so it seemed likely that they would not need to pay for the whole process themselves. However, as much of their initial care would take place in a clinic about an hour's flight from their home, they would likely incur travel and related costs, even if their PGD were publicly funded.

The Business of Haemophilia

Products for the treatment of haemophilia and all of the devices that allow that treatment to proceed safely and as easily as possible are big business. This is evident at the World Congresses, where the major multinational drug companies have elaborate displays and offer hospitality to many of those attending, and even pay conference attendance costs for some delegates. It is clear at the national level, where, for example, the HFNZ has a bevy of these companies as sustaining patrons and is careful not to be beholden to just one. The companies provide education days for people in the health sector, and Pharmac negotiates deals for supply with them, seeking the best deal. And it is also evident at the individual level, where parents often have direct dealings with the company representative and might refer to their children as 'Bayer kids', if they are treated with the factor made by that company. Children become aware at quite a young age that they are sought-after consumers.

Win: A couple of years ago, 18 months ago, our haematologist looked at Wyeth rFVIII for us. I forget what it was called now, and I said, well you know, I'd like to have all the facts and figures

> before we sign on to anything. And son, being 13 at the time, said, "Oh, what's in it for me?" Other boys he's spoken to, you know, 'cause we're Baxter, and Bayer provided kids with cell phones and bags of all sorts of goodies for being a Bayer boy, and of course Baxter don't supply those sorts of niceties for the Baxter kids. So my son said to the haematologist, "What's in it for me?" And the haematologist said, "What do you mean?", so son explained all the cell phone and all the dramas, and he said, "Well [name], I'm sure I could run to a can of Coke and a bag of chips." [We laugh.] And my son said, "That'll do." And you do have these boys … There was this 13-year-old thinking, "Oh if I change products, oh there could be something in it for me."

Julie: Interesting, the market thing is there.

Win: Yes, let's do market research: what's in it for me? But, as it turned out, I was more concerned that if he developed an inhibitor with a new product or had a reaction against it — for whatever reason it didn't work — we couldn't go back on to Baxter and I was too afraid to lose what we already had that worked.

Julie: So did you stay with Baxter?

Win: Yeah, my son's probably one of the original Baxter kids … I mean he's been on Baxter products since he was about four years old, you know when they first introduced the six months free to get you hooked on it.

In contrast to Win's son, Pam and Peter's son has had products from every pharmaceutical company providing recombinant clotting products in New Zealand, because it has been very difficult to find a suitable treatment for him. Pam and Peter have consented to his photo being used by one of the companies because these products have made such a difference to their son's life. Pam singled out for praise a representative from a company whose products they found that they could not use. This did not make any difference to her relationship with them, and she still popped in to see them and kept in contact. Pam summed up:

> I think our dealings with the pharmaceutical companies have been fine. I think that they have to make a business and that on the whole they are fine. And when it comes to the genetics and trying gene therapy, they are scientists just like the one that years ago developed recombinant Factor VIII and people probably had the sense that those people were spending money and are money-hungry, which they are, but they've made a huge difference.

During the fieldwork period, the work being done by one company on a five-day product which would halve the number of treatments for many people was mentioned with considerable interest. Non-intravenous delivery of product was another area where people sincerely hoped work would be successful. Another aspect of work being done by one company was mentioned: the release of a Palm Pilot that is able to communicate about bleeding episodes with the hospital to users of a recombinant product. Users described how they are able to enter into

Figure 3. Multinational pharmaceutical companies display their products at the Haemophilia World Congresses, held every two years.

the Palm Pilot the bleed, site of bleed, product batch used, and other information about pain and the site of injection. This information then gets sent to the hospital via a modem. Thus the innovations being worked on by the drug companies are being eagerly awaited.

The few times that there has been a shortage of product or a scare — such as the CJD scare with a company's product in 1999 — the leaders of the Foundation and people who were aware of the issues believed that these situations were handled well. Nonetheless, some people have expressed feelings of vulnerability with regard to product supply, because New Zealand is a small and distant market for these companies. The cost of products and of anticipated new products also raises anxiety. The fact that their first duty is to make profits for their shareholders is not forgotten, as one of the older men who had taken leading roles in the Foundation pointed out:

> All of this cutting-edge research is done in the private sector and not in the state sector. Obviously there is going to be a price tag on this. One thing I've learnt is that price has nothing to do with cost. Price has something to do — well, a lot to do — with value of the commodity to the community. And because there is going to be that type of what, by this stage, will be treatment [gene therapy] that would replace the treatment that we are conventionally used to today — with an infusion every second day, of a couple of thousand dollars worth of pharmaceuticals — it seems to me that the price of gene therapy is going to be extraordinarily expensive to actually have that three-month injection.

Another man discussed the cost of products and the different aspects that can affect price. He made the point that the revenue from current products funds research into new ones.

> For products like FVII [which is used to treat people with inhibitors] they are using it in a lot of other [ways], like surgery. American military probably uses it as well, in the field, so there's other uses for the factor to stop bleeding for anyone, and so there're probably more demands for the product. They can get the price they want. Yet … the patent thing limits the suppliers, so then the price is higher. Yes, it's difficult. I suppose that researchers doing that research could invent a product and patent it and give it to the world for free, but then probably the research wouldn't take place in the first place because there wouldn't be funding for it. So it's a whole dilemma. I suppose an ethical dilemma for researchers to decide how they will fund their own research.

One family had researched in great detail the procedure of sperm sorting. They had gained ethical approval and funding to have the procedure done, but finally did not go ahead with it, for the reasons they explain in the following quotation. Sperm sorting can increase the chances of having a child of the desired sex. In this instance, the sperm-sort company was American and the couple would have had to travel overseas. This couple were unwilling to "discard" a fertilised embryo, as in PGD, or have an abortion following chrionic villus sampling (CVS), but initially believed that sorting sperm was acceptable to them.

> Wife: I think we sort of went in and looked at the company a lot more and actually found that while they were doing this for medical reasons, actually they were covering the cost of medical reasons by those people who were just choosing a boy or girl.
>
> Husband: While it was free for haemophilia and genetic disorders, they would do free treatment, but then the rest of their company was based on 'family balancing' as they called it — choosing one sex — and in some ways we didn't feel it was right to be used in that manner, in terms of they are using a good thing to justify something that is a bit more debatable in terms of, yeah, I can just see more danger in society choosing on gender and then wonder how far that will go.
>
> Wife: And especially just when … they prefer to have a boy or a girl in their family. And I think also … we felt they were solely using us to get a collaborator to use it here in New Zealand, and I didn't agree with that.

This situation demonstrates the degree of thought and ethical debate behind some of the decision-making that people with haemophilia undertake. These examples also show the complex relationships that people with haemophilia have with commercial firms and the market. On the one hand, they rely on the firms' products and processes; on the other hand, they are wary of being used in ways that they cannot approve, and of the unintended consequences that might flow from their quests to find the best products or the most acceptable procedures.

The Haemophilia Foundation of New Zealand: Support and Information

When one young couple with a boy with mild haemophilia decided to move from a large city to a small city, one of the things that gave them pause was the lack of haemophilia support. A visiting specialist would come perhaps only every two months and there was no HFNZ branch activity in the area. However, they still would be able to ring the outreach worker. The mother in this young family was an obligate carrier and was one of those who had always known about haemophilia. Despite being quite knowl-

edgeable already, the couple found the branch network very effective and felt that they had both learned a tremendous amount just by being involved. In addition, they had attended conferences in New Zealand and Australia, which the husband, in particular, had found informative. Their planned move enabled them to be closer to grandparents and siblings, so they were not so concerned about personal support. More, they worried about no longer being in the thick of things where they seemed to absorb new information as they did their voluntary work. Only half-jokingly, they said that, with their extended family and the other families there whom they had yet to meet, maybe they could start up a sub-branch.

The role of the HFNZ as an education and support organisation was significant for many of the people we interviewed, as it was for this young couple. The informal interpersonal ties created at local branch level and through the national council, as well as via the various camps, workshops and conferences that the Foundation ran, were also very important, especially for new families, families with young children, and anyone having a crisis. The political role of the Foundation was also frequently acknowledged.

Over the past 10 years, the HFNZ (formerly the New Zealand Haemophilia Society) has grown and changed. During this update study, the national office was in Christchurch where it had been for several years, staffed by a chief executive officer, an office manager, and an information officer. A professional staff of three outreach workers was also employed by the Foundation. One worked from the Christchurch office, and the other two had small offices in Wellington and Auckland. The elected National Council members were voluntary, as were the branch committee members and the others who served on the special committees, such as the education trust. A Medical Advisory Council sat alongside the Foundation and contributed its expertise and advice on a wide range of medical matters. In addition, a National Haemophilia Management Group, comprising health professionals and including a representative from HFNZ, was in the process of being formed while this update was in progress (see below). The National Council was also represented on a number of government-sponsored groups relevant to haemophilia and hepatitis C policy. The Foundation and its many activities were funded from a variety of sources: contributions from pharmaceutical companies who were among its sustaining sponsors, the Kiwifirst philanthropic organisation and other charities, government grants for specific purposes, local funding sources for specific regional activities, bequests and donations, and the contributions of its members.

The services of the outreach workers, the newsletter, camps and workshops are the most public aspects of Foundation activities. However, through the Foundation many individuals are personally assisted in a variety of ways. These extend from getting shoes — people with haemophilia require well-made support shoes to protect ankles — to assistance with swimming programmes, educational grants, payment of carer support in specific circumstances, and other specifically tailored help.

Everyone we interviewed was a member of the Foundation and received its newsletter, which was perceived as very helpful, but not everyone was active in the sense of attending meetings and camps. A few people had only recently become involved, after an absence. This was particularly the case for women who had attended camps with their brothers or father when they were young, had had a period away, and then had got involved again as they themselves started to think about having children. Around a third of those we interviewed were, or had been, local or national committee members.

Although the results of all the Foundation's work and lobbying were noted appreciatively by many members, one or two of the longer-term members expressed some regret that it was now such a political group whereas in the past it had been much more focused on grass roots support. Despite this, several new families reported that talking with other Foundation members was their most helpful source of advice in the early days of coming to grips with haemophilia.

The HFNZ outreach workers were cited by many participants, especially those with young children, as being a wonderful resource. People described telephone

Figure 4. Haemophilia Foundation of New Zealand Young Families Camp, 2006.

calls and visits to and from these women, and the timely help that they received. At the time of the study, an outreach worker had recently been appointed for the Wellington region. For a considerable time before that, there were only two for the whole country (based in Auckland and Christchurch), which had made covering the whole population a real challenge.

One family had had a particularly difficult time over a period of several years. They were well-informed, confident people and appeared to 'cope', but they felt that their continuing needs for support and assistance were not adequately recognised, and help was forthcoming only at times of crisis. As well as the competing demands on the outreach worker's time limiting the amount of help available, it is possible that the professional background and competent demeanour of this couple may have masked their need for support at times, a suggestion that is consistent with other research on coping and support in middle-class families in New Zealand (e.g., Park 1983). Especially with so many people having interferon combination treatment in 2004 and 2005 for hepatitis C (see Chapter Five) and needing considerable extra support to cope with the treatment, the outreach workers' capacities were indeed stretched. They provided one-on-one information and support via telephone and visits, helped their clients deal with bureaucratic processes, organised national camps and workshops, supported the branch activities, kept close liaison with clinical workers, and kept up to date with new developments or old issues as they resurfaced. In recent years, some women with personal experience of haemophilia as well as professional training have been employed in these positions.

In our interviews we found that there were people who had no one with haemophilia in roughly the same age group or situation living nearby, who would have appreciated parent-to-parent or woman-to-woman support. Sometimes an outreach worker could 'buddy up' two families for mutual support. In other cases, people who had met at camps or in Starship (the children's hospital in Auckland City), when both their children were hospitalised at the same time, kept up telephone contact, which was helpful, but not a complete substitute for in-person support. Those involved in the Foundation and their local committee knew the other people with haemophilia in their area and could offer support. For example, as described in the discussion of treatment, sometimes people on home treatment lose their confidence after missing a vein, or the child decides suddenly that he does not have haemophilia any more and does not need treatment, or becomes fearful. When a young boy and his mother had a crisis over treatment, a more experienced home-treater and her teenage boy who treated himself went over to their home a few times and the teenager did his treatment alongside the little boy while one mother encouraged the other until the situation was back to normal.

Travelling around the country and meeting people with haemophilia, and telephoning people in their homes, we could see the difference that it makes having a support network of people who know about haemophilia around you. One young mother could not speak highly enough of the help that she had received from the outreach workers, but what she really wanted was to know other women who were symptomatic carriers or who had symptomatic carrier daughters to help guide her in bringing up her girl. Another woman had had tremendous help from her (distant) outreach worker and staff at her haemophilia centre, and good family support, but there was no one living around her with experience of haemophilia on whom she could call. She herself said that after about two years of dealing with quite difficult situations, it had all got "just too hard". Fortunately, she did seek professional counselling. This is the kind of situation in which a somewhat more experienced family can provide that day-to-day support for the continuing struggles with bleeds, setting realistic boundaries for the child, dealing with medical authorities who do not know much about haemophilia, setting up good access systems at the local hospital, and all the myriad other things that a new mother or couple with unexpected haemophilia have to deal with, in addition to coming to an acceptance that this disorder is now in the family.

The camps and workshops were a time of often intense sharing of information and experiences among those attending. As an example, for Tui not only was it the first time she had met other Māori people with haemophilia, and other parents who "really understood" about haemophilia, but she learned about factor levels, so that at her next review she would be asking about the tests for her son's levels. This camp was the first time that her son, now at primary school, had met any other boys with haemophilia. Because he was the only one with it in his school and among his family, for a long time he did not realise there were any other boys with it at all. Other families whom we interviewed shortly before or just after the January 2006 camp spoke about how much they were looking forward to it, or how much they and the children had enjoyed it and had got out of it. Many people went away with new friends as well as with new information that they would be discussing with their specialists at their next appointment.

But camps were not for everyone and nor could everyone attend. One father at the camp, for example, said that he found just being in such a large group — there were around 120 people — quite taxing, despite the benefits of the education sessions and the activities for the children. A woman expressed disappointment that she could not persuade her nephew with haemophilia, who had been infected with hepatitis C, to go to the Hepatitis C Conference. She thought that he would go to the proposed camp, later in 2006, for men with haemophilia where there could be an opportunity to talk about hepatitis C, but going to a conference identified as

being for 'hep C' was another matter. The young woman and her daughter who were symptomatic carriers were the only ones at the Young Families camp. Although they learned a great deal about haemophilia there, they did not meet other women with similar problems and learn specifically about symptomatic carriers. Interestingly, another young symptomatic carrier whom we interviewed had been to many camps when she was younger, accompanying her father, but she had always thought of the camps as "being for the boys", which of course they were, until recently.

Haemophilia conferences were rated very highly by attendees as places where they could access up-to-date information, generally in a fairly easily digestible form. These were the places where the latest research on gene therapy or new treatments or physiotherapy and so on was presented, and where there was also a good deal of sharing between individuals and small groups. One person pointed out that a lot of the science behind haemophilia is very complex, and for the lay person repeated exposure to the concepts is necessary to build up an understanding. A contingent of New Zealanders, which can number up to 30, usually goes to the Australian haemophilia conference, and a much smaller group, perhaps about six, to the World Congress. One Australasian conference has been held in New Zealand and it was very well attended. One person wondered if it might be a better use of Foundation money to have more conferences in New Zealand to allow more New Zealanders to attend. She thought it would not be difficult to attract good speakers.

As well as the sources of information provided by the HFNZ and the haemophilia centres, participants learned about haemophilia from a wide range of sources: television, movies, GPs (rarely) and the internet. A few people expressed some frustration that, rather than their doctors providing them with information, the process often happened in reverse. The internet was seen as a slightly mixed blessing: it could overwhelm you with information, and it was not always easy to assess the information's quality, but it was a powerful source of basic information as well as a way to keep abreast of the latest developments. For example, those couples interested in new technologies, such as sperm sorting or PGD, had used the net to do their research, and so had people interested in naturopathy and related complementary approaches to healing.

Towards a National Service for Haemophilia

Over the past ten years, the HFNZ had been working towards a national service for haemophilia. Our earlier research showed clear inequalities between regions which were purely an artefact of the way that the health and blood services were regionalised (Park *et al.* 1999). Consequently, we are of the research-based opinion that a national service is required for equitable and good quality treatment, and we agreed entirely with the HFNZ goal. At particular times this was almost achieved or was achieved for a brief moment. For example, after a few years under the regional health authority (RHA) system, one RHA was designated as the contracting agent for haemophilia services: an 'as-if' national service when there were four RHAs. No sooner was this achieved than the RHA system was dismantled and replaced by the Transitional Health Funding Authority and the National Health Funding Authority — both excellent news for haemophilia. But with every re-structuring, officials changed and their replacements had to be re-educated. Before the national system had time to draw breath, the district health board system (DHB), resulting in 21 DHBs , was instituted — a very retrograde step for the haemophilia community, in our opinion.

However, in the meantime a national New Zealand Blood Service, which formerly had been regional, was created. Thus purchase and supply of both the plasma-derived products from the Blood Service and the increasingly-used recombinant clotting products purchased by Pharmac, became nationally-based. This had considerable advantage: for example, at the Young Families Camp, a speaker announced that Pharmac had recently concluded a deal with one drug company for the supply of their product for the next three years at half the cost of the product in the United States. People who were on other products did not have to change, and new patients could be prescribed alternate products if there were a clinical reason. The arrangement would be reviewed in three years. This was reckoned a good deal, although there was some anxiety about the single supplier and possible shortages if something went wrong with production.

During the 1990s, the haematologists who advised the Foundation (the Medical Advisory Council) and Foundation members worked with other interested parties to produce National Guidelines for the Treatment of Haemophilia, which have been influential in creating a more consistent service within New Zealand. Thus, many of the building blocks of a national service were in place by the first decade of the 21st century. The main stumbling block was the way that the funding of haemophilia services was delivered through the 21 DHBs.

As described earlier, some families with haemophilia have shared care between two DHBs. Other families who live in one DHB area have their care from another. Because of the cut-back in service from Waikato Hospital, for example, we were told that some Waikato people seek some or all of their care from Auckland DHB, and some Otago and Southland people had care from Christchurch. Complicated cases and emergencies typically end up in the Christchurch or Auckland city hospitals. Two people in this study had actually moved their place of residence to be in a DHB where they believed care was superior, and this had

also occurred with participants in earlier studies. One of the haemophilia nurses told us that she sometimes tells her patients that they cannot expect to have a full haemophilia service in their local DHB area and they should consider moving to a larger centre with a better service. This is part of acknowledging that they have haemophilia and that certain adjustments have to be made to be able to live safely and fully. If they cannot move, then they have to expect to travel to treatment.

While it is obvious that in a small country like New Zealand it is not possible to have haemophilia specialists and a complete service in each DHB area, shared care between a specialist haemophilia centre and the local hospital or health centre can deal with this aspect. If the funding of treatment were not funnelled through the different DHBs, it would be possible to have the same satisfactory level of treatment irrespective of which DHB a person lived in. It is this that the National Haemophilia Service is aimed at. In the words of one of the people who had been involved in the negotiations with the Ministry of Health, this is what the plan was in December 2005.

> And the Ministry, I don't know if [our representative has] worn them down with correspondence or what, but the Ministry seems to be very keen to have a national service and the majority of DHBs have [agreed].

> My understanding [is] there was only actually two [DHBs] that weren't that keen. One of them didn't have anyone with haemophilia in their area. Funnily enough they had someone move there and another one go on holiday and have an accident while there, and they suddenly changed their mind! …The Ministry, to get the DHBs on board, has thrown some sweeteners in there to the extent that, can't remember, but 3 to 4 million [dollars] over three years allowed in their specialist service budget for tolerisation and that sort of thing, and if that is not used it can go back into the national pot. The national group that's being set up, it goes to them and they get another lot each year. And if it's not used, it's fully paid out to them under contract, if it hasn't been used for tolerisation, etc., and a couple of months of the year are left, they've got choice. They can say, we're going to do these six elective surgeries [for people with haemophilia], or whatever. But coming back to the DHB level for those sorts of things [i.e., the system at present], you will find that someone in Auckland will be able to get that sort of thing and someone in Southland might not 'cause the money has run out.

He then went on to explain that even the differences between individuals can be great and this can have an impact on a DHB budget. He compared his son who has almost never used extra treatments, apart from his regular prophylaxis, with a couple of friends, one a few months older, the other a few months younger. Both of them have older siblings and are always getting bumped around and need extra treatments.

Elective surgery for people with haemophilia typically would be something like a knee replacement for an arthritic or fused knee which has resulted from repeated bleeds into the same target joint: something that prophylaxis now prevents in most cases of younger people. Elective surgery requires a stockpile of treatment products to keep the factor levels up during and after surgery.

It was excellent news to hear, after our draft report was completed, that this national service had eventuated.

Gene Therapy

As a possible treatment in the future, gene therapy provides an excellent example of the ways in which people with haemophilia deal with the complicated and sometimes futuristic science that impinges on their everyday lives. We were interested to know how gene therapy was thought about and participants' attitudes towards it.

The ability of gene therapy to provide treatment of haemophilia for the present generation was greeted with considerable scepticism. Virtually everyone had heard about the possibility of gene therapy. Those people who had followed the fortunes of gene therapy trials for a decade or two described this therapy as an always-distant horizon or as a ship that never comes closer. It appeared to be as far away in 2005–06 as it had been 10 or 20 years before. Some of the younger people and those who had only recently come in contact with haemophilia were more enthusiastic about its application; others were highly sceptical. One mother surmised that this was a protection against disappointment.

A few understood it as a once-and-for-all procedure carried out on an embryo to "get rid of the bad gene instead of aborting the child", or perhaps on a grown person, in which case it would provide a lifelong cure. The once-and-for-all scenario was noted by an older man who had followed gene therapy closely for decades as the initial way it had been described to him. But in recent years the 'top-up' approach was what was mostly talked about. Most understood it to be not a cure but a form of prophylactic treatment that would at best allow someone with severe haemophilia, for example, to have sufficient clotting factor to be classified as moderate or even mild. How this might happen might be through 'a jab' or a pill or perhaps some kind of implant of genetic material that would, for a while at least, manufacture clotting protein in one's body. A few people mentioned that trials had been done on dogs and some on humans, but that there were unresolved problems including some major health issues. A number talked about the virus

vector that was used to introduce the 'clotting gene', two mentioning that a modified HIV virus was one of those experimented with — "an unsettling thought, given the history of haemophilia".

The 'ray of sunshine on the horizon' was that gene therapy would be necessary only infrequently, compared with current prophylactic treatment, and would not be intravenous. 'Infrequently' might be once a week, once a month, once every three months, or even once a year. When discussing frequency, several people noted that the drug companies are at present trying to extend the half-life of their recombinant products, so that people who are now treating themselves three times a week or more often would be treating only once every five days if this work is successful. This was seen as a great boon, and considerably more likely to eventuate in the near future than gene therapy.

Conversations about gene therapy indicated that, although nearly everyone had a degree of confidence in scientific medical trials and would not consider making a major change in treatment that had not been trialled, trials alone were not quite enough. People live with haemophilia their whole lifetime, families live with it for generations. Compared with this time-span, trials are short-term. Although not the only concern, the long-term consequences of gene therapy are the major issue for people with haemophilia.

So long as gene therapy were properly tested and trialled and the trial subjects monitored long-term, most of the people interviewed would consider using it for themselves, or would be prepared to discuss its use with or for their children or grandchildren. Once it was available, they would weigh up the pros and cons against other available treatments. However, they pointed out that it would not be their decision, but that of the younger generation.

No one interviewed opposed gene therapy outright on the grounds that it involved genetic modification (GM). However, several people revealed that had they not become involved with haemophilia they would have opposed it because of their prior stance on genetic engineering (GE).

Ursula: I used to be totally against GM, but of course I have totally changed that view because of our situation, and if a cure could have eventually…

Julie: So you have changed your view around this area, but what about other areas of GM?

Ursula: Not in terms of crops, I think that is still dangerous, I don't like that idea at all. For medical [purposes] only.

This routine use of a GE product by most members of the community, especially the younger ones, was highlighted by quite a few people as significant in causing them to rethink their unreflective anti-GE stance, and to make them more open to gene therapy and other new technologies, such as PGD. As Nita, the mother of a nine-month-old, said, so long as gene therapy has been properly tested:

> … if it makes my baby healthy, happy, and well 'protected' is sort of the wrong word, but more like everybody else, so he can play or run or do whatever he wants, without mummy having to fear that he is going to be hurt and end up in hospital, why would you not use it?

Unintended long-term consequences were noted by several people as their main reservation about gene therapy, even when trials had been properly completed. This was often expressed in terms of what was natural, or going against what the body was designed to do. Don used the internet to read up on gene therapy, and he and his wife, Deb, had discussed it in some detail. Although they would consider gene therapy for a child, to them it was "scary because it changes the molecular structure". Tui had heard about gene therapy from movies and television, and although she thought that:

> …it doesn't sound very natural, if there were older adults who have used it and go through it and then to hear first hand from somebody that has been through all that, and then I might consider it.

New Zealand "not being at the cutting edge" was seen as an advantage when it came to gene therapy. There would be time to observe its implementation elsewhere. Several people expressed admiration for men who subjected themselves to trials of new treatments, saying that they would not be first in line for it. Two men, one in his 30s and another around 60, said that they would be prepared to be part of a trial. For genetic therapy of any kind, and this included PGD also, participants noted that there was always the possibility of what Anna described as "science gone mad" — technologies which had the potential for good being used for evil. On the other hand, Ashley cautioned that anything to do with genetics has to be approached very carefully, as it may have unintended consequences, and people have to think about it and avoid the hysteria evoked in slogans such as 'designer babies'.

A number of practical obstacles were mentioned. Funding in New Zealand's stretched health services would be an issue, for example. A leader in the haemophilia community warned that gene therapy would come with a price tag that was not necessarily the cost of production and distribution, but the price based on its high value to the community. Then there would be the effects of this treatment possibly replacing what are at present conventional treatment products (plasma-derived or recombinant) with a consequent reduction of options and changes in cost structures. He reflected on the contrast that he had observed in the haemophilia community between worries about the safety of current blood products and the relatively

relaxed approach to gene therapy, despite its experimental nature. Several people noted that gene therapy would probably involve taking immuno-suppressants to prevent rejection, and this could cause problems. One person thought that if the exact genetic mutation causing each person's haemophilia were pinpointed — which it is for most people through DNA studies — then the amount of foreign genetic material that would need to be introduced to stimulate clotting would be reduced, and so, therefore, would the possibility of rejection of that material.

Chapter Four: Haemophilia: A Genetic Condition

In this chapter we discuss how individuals and couples respond to the possibility that they and/or their children may have the genetic condition: haemophilia. As an X-chromosome-linked condition, haemophilia is passed by both men and women to the next generation. Men pass it to all of their daughters, who become carriers, and to none of their sons. Women with a gene mutation for haemophilia have a 50% chance of passing it on to their daughters and to their sons. This pattern means that the testing of girls to see if they are carriers is only necessary if their mothers are carriers or if there is a suggestion of a spontaneous mutation. About 30% of haemophilia in any generation is reckoned to be new, due to new mutations. These are subsequently inherited in the usual way.

Carrier Testing

All of the women participants, the men in couples, and two of the men with haemophilia discussed carrier testing. Of course, all men with haemophilia are also carriers, but they do not need further tests to establish this. Four of the women were obligate carriers, but only three had always known their status. The other woman had been adopted and had discovered her probable status from her birth parents when she was 18 years old. Even then, her biological father's status was not definite, but it has since been confirmed through the pattern of haemophilia in her birth family. Of the non-obligate carriers, four were from families with known histories of haemophilia, but one was adopted and had discovered she was a carrier only when her son was diagnosed. The birth grandmother, contacted for the first time after our participant's son was born with a head bleed, recalled that her brother was a 'bleeder'. In the case of the second adopted probable non-obligate carrier, her birth mother was unaware of any family history of haemophilia so our participant was probably a spontaneous carrier, although the possibility of obligate carrier status was not ruled out. Two women were still waiting for results, but were the first known people in their families to have a child with haemophilia. The other women were spontaneous carriers so far as DNA testing could establish. Two of the women over 40 years had not had a DNA test: one because it had not been available when she was having children, the other because their familial mutation had proved very difficult to discover. Both were established as carriers by having more than one child with haemophilia. However, one other older woman who had given birth to her child with haemophilia before DNA tests were available had a test later to confirm her status.

DNA testing of the daughters of women carriers, or the mothers of single children with haemophilia, is necessary to establish their haemophilia status. Clotting factor level tests are indicators, but while a low level indicates haemophilia, a normal level does not rule it out. Women who are alerted to haemophilia because of the unexpected diagnosis of a son are tested to establish if the mutation occurred with the son, or with the mother, or perhaps earlier in the family history. The results of this test can have implications for the woman herself with subsequent pregnancies, and for the woman's sisters, daughters and nieces. Such tests usually require blood samples from several family members. Sometimes the carrier women's bleeding problems are so marked that it is this that leads to the decision to test for haemophilia. This was the case for one of the participants, and for a daughter of a participant.

The interview conversations about carrier testing included descriptions of the testing process and its consequences, as undertaken by the women participants as well as for any daughters. Where there were no daughters, participants were asked about what they would do, or thought should be done, for potential carrier girls, particularly what time they thought was best to test, and when and how to tell their daughters, especially if the test results were positive.

There was absolutely no debate about the desirability of carrier testing; however, there was considerable discussion about the timing of testing and whose decision it was to have a test. The exception to this was the testing of symptomatic carriers, for whom testing was seen as a necessary safety measure. Our interviews showed a range of practices around the country in terms of the testing of asymptomatic carrier girls. In fact, at a workshop on the social implications of prenatal testing that Julie conducted during the Young Families Camp, two women who attended the same hospital, but who were treated by different clinicians, found that there were different practices within the same hospital. The status of some daughters of mothers with haemophilia was known from the time they were a 12-week foetus, as a result of prenatal testing, several others were tested using cord blood at birth (although it was not always clear if these were clotting factor level tests to see whether bleeding problems were likely, or DNA tests), others as quite young children, several around the ages of 11–14, others as older teenagers, and another we heard of, at the age of 30, when she was starting to think about having children.

Two of the non-obligate carrier women who were in their 20s also raised the issue of carrier testing in relation to PGD, a topic that they had discussed at the Young Women's Workshop. They were interviewed separately, but both expressed the same opinion, even though they were both quite cautious about the use of PGD.

Hannah: There was the screening process itself [as part of PGD]. We had a few sessions with how they won't test [for] carrier girls and how that decision is left up to the next generation whether or not they want to go through the process again. We thought that that was a bit bizarre. Um, you know, that you can screen boys to see if they have the haemophilia gene but you don't screen girls. Which was quite a bizarre thing.

Deon: So, if there is a known history in the family, you would screen the boys ...

Hannah: Yep, yep, but you won't ...

Deon: You won't screen the girls; see what is happening with them?

Hannah: Yep, which we thought, you know, if you can have a carrier girl or a non-carrier girl, why would you not have a non-carrier girl, you know. It's the same as getting the haemophilia gene out [i.e., eliminating the mutation from the next generation], which is what you are doing by, you know, by having a boy who doesn't have haemophilia. And because it doesn't directly impact on their lives you say ... you know, you can't do that, which was a bit bizarre, but, hey, I'm sure they'll wrinkle it out and realise, see sense, hopefully.

Hannah was fully aware of the eugenic effects of screening for male embryos with haemophilia and thought that this should be available for females too.

People who knew their daughter's haemophilia status from prenatal testing or cord blood, or from a young age, were in a similar position to those with obligate carrier daughters. They could choose how and when to integrate this knowledge into their daughter's lives. Girls growing up with fathers or brothers with haemophilia, attending haemophilia camps and other events, learned about haemophilia first hand. Girls and women in this situation reported that they grew up "always knowing", which they greatly preferred to the idea of suddenly having to learn about haemophilia and being a carrier at an older age.

Parents talking about this situation were in favour of the idea of incremental learning at the level the child was at over a number of years, and the opportunity to respond to particular enquiries from the child when she became interested in the implications for her. Their discussions were guided by the principle of the best interests of the child, according to contemporary ideas of educational theory and child development. Several people, including a woman who had been adopted herself, referred to current best practice about imparting information about adoption to adopted children, which was to create the situation where they always knew, as a model for imparting information about carrier status.

However, one of the young women attending the Young Women's Workshop was surprised to find a couple of carrier girls there who were about 13 or 14 who did not know much at all about haemophilia. She surmised that their parents had tried to hide it from them, although their attendance at the workshop suggested that perhaps the parents had just not been confident to educate them themselves. In contrast, one of our participants explained that his teenage daughter did not attend the workshop because his wife did not want other people talking to her about haemophilia. He said that she was well informed about her options from discussions at home.

Although there are guidelines about not testing carriers until the girl herself is old enough to give consent, many people, and apparently their health professionals, were blissfully unaware of them, or perhaps disregarded them. Some had recently heard about these informed consent guidelines and were not at all happy with them, whereas others agreed entirely that it was up to the girl to decide if and when she wished to be tested.

The discussion between Murray and Marie is a good example of the perspective favouring early testing.

Murray: I think it is important in so far as you take away the element of surprise, you are aware of it the whole time. So if you are aware of something, and perhaps information that will crop up along the way, like you'll put it in the back of your memory, rather than being dropped in it at the last minute, when you are not really educated about the whole deal.

Marie: I agree with carrier testing, and I reckon it should be done as soon as possible, before they are teenagers. You know, the whole issue of them only having Factor VIII level tested, as an indicator and then having it confirmed with genetic testing later on, when they are able to give consent, at 15 or 16, to me that is just a little bit late. 'Cause they are dealing with a whole lot of other issues and to have it sprung on them, "Yes, you are a carrier", you know!

Murray: It's a terrible time anyway.

Marie: And, who knows — hopefully not — they might get pregnant when they are only 13 or something!

Julie: You'd hope not, but it happens!

Marie: It does and then I guess, if we had a daughter and she grew up with a brother having haemophilia then she'd be already aware of some things and take notice of some things, probably like what me and my sister did, growing up with Dad. Knowing that whenever he hurt himself he'd have to go up to hospital, needing treatment.

Murray: I do think that it is a bit over-the-top PC, that thing. We're responsible bringing the children up to 15 or whatever and yet you are not allowed to have them tested and stuff, and yet you are responsible if anything goes wrong. I think it's PC gone too far, for, really, safety, in a way. 'Cause you know how things are with kids and stuff up to the age of 15. As well as the education thing, you know, whether they find out about it at a stage when they are not thinking whatever the parent says is wrong and they've got to find their own way in life.

Marie: Yes, that's true. I hadn't thought of that, like you know. You can go through a rebellious stage. If I hadn't known until I was 15 or 16 I was a carrier, at that point I used to have lots of arguments with Mum, and you know, ignored the things she said.

Murray: A little thing like that could cause a massive chip on someone's shoulder, a teenager's shoulder. "Why me?" and that sort of stuff. You know when, if they grow up with it, they don't know any different.

Julie: Well, it is interesting, 'cause all the daughters of the fathers know, from as soon as they know anything, about haemophilia, but the daughters of the mothers, that's where that issue is … [Because the couple appeared to be getting a little upset about this notion of informed consent, Julie explained the concept of Gillick's competence, where a child younger than 16 is deemed to be able to give consent if they are well informed and acting freely.]

Marie: I don't think they should know at the age of eight or nine, but 12 or 13.

Julie: It does seem to be an age when kids are curious, and intelligent.

Marie: And they can understand it, because we have a niece who is 11, she's at that age where she asks.

A minority of participants did not agree with testing being the parents' decision, and emphasised the importance of the daughter's informed consent and her privacy. However, Carolyn, who stressed that testing should be done only with the girl's consent, still thought that with going to haemophilia camps and likely having a brother with haemophilia, most girls would want to know by about 13 or 14.

Julie: Yeah, and just examining that a bit further, when thinking about the child needing to be able to consent, what do you think could be the downside for the child themselves, if parents have her tested at a young age? Because there is a range of practices in New Zealand, despite the guidelines.

Carolyn: I don't know. The child may be resentful, or may change the way you treat them, you know. If you've got a haemophiliac child whom you might protect a little bit more if you realise that your daughter may have issues as well. I'm not sure. I can't think of an answer to that. Yeah, I just think that sometimes, more people need to know than actually should. When something like that ... I know just with [son], that I try and be really particular who I tell, and I sort of feel it's out there a bit too much, 'cause I still don't know much and I want to keep it a bit closer, so yes, yeah, people who don't need to know, know more about a child who hasn't given consent for that information.

One mother who had had her daughter tested when she was about four years old, before there was privacy legislation or guidelines, wondered if her daughter might resent that now. On the contrary, her daughter, who was also a participant but who was interviewed in another town, was happy that she had always known:

> My point of view is, I think, 'the earlier the better', you know. The more you grow up with it and it's accepted, it does become something you just get on with, and it's just what it is and you just deal with, you know, rather than all of a sudden going, "Oh my God, is that what it is" … Yeah, the earlier the better.

Despite always having known her status, it was only when she had begun thinking about having kids in her early 20s that she really began identifying as a carrier, and seriously considering the implications.

People who had had cord blood tests had been offered the test, rather than requesting it, and some parents who had had their girls tested when they were quite young also reported that this had been suggested by a health professional. Other girls were tested as part of the effort to pinpoint the familial mutation when a brother was diagnosed with haemophilia or a mother's carrier status confirmed. Other parents had told their girls early on that there was a chance that they might have 'bleeding tendencies' and be carriers of haemophilia, and had told them to let them know when they would

like to be tested. Quite often it was around age 11 to 12 that girls requested their mothers to take them for a test, and at this age they presumably were deemed (under Gillick's competence) to be old enough and well-informed enough to be able to give informed consent, should anyone recall the guidelines.

A few girls probably did value their privacy. One mother told the story of her daughter going to the haematologist when she was 18 without telling any members of the family. However, by mistake the results were sent to the parents rather than the daughter, and she, understandably, was not happy.

The two men with haemophilia who commented on this issue held different views. Andy thought that you should "test as soon as possible but then as a parent, decide what is the best age to tell them. And I don't think the best age is once they are in a relationship and want children!" But Albe wondered what good it would do if the girl knew early on. He recalled that his sister had become very interested in babies around age 13, and thought that was the stage when it was best for a girl to know about her status.

Andy commented that parents of obligate carriers also needed to carefully judge how and when they tell their girls. But one of the obligate carriers described her parents "blasé" approach to this, about which she was unperturbed:

Frances: Like my mum didn't even tell my primary school and she got in trouble for it later when I had a three-hour blood nose. And the principal freaked out. No, I was just told, your father has haemophilia and this is why he goes into the laundry every morning and gives himself an injection, and one day you'll pass it on to your children. End of story. That was it.

I don't think they realised, I don't know when they realised [that I had bleeding problems]. Actually, it must have been when I was at primary school when I started having lots of blood noses. I don't know if I had lots of bruises. Oh yeah, that's right. Mum and Dad found out that the primary school had got a social worker to keep an eye on me because I had bruises up and down my shins and they thought my parents were beating me up, or something like that. And that's when Mum and Dad were contacted and "Oh no, she's actually a carrier." My mum's very … she doesn't like getting people worried about stuff, so …

Julie: So it wasn't to do with privacy, more to do with not making a fuss?

Frances: Exactly, exactly.

In contrast to our initial study when many women reported long delays in getting results from carrier testing, in this study that was a rare problem. Most people were quite happy with the way that they had heard the results. Several people explained that now DNA testing was being done in both Auckland and Christchurch that speeded things up. Auckland handled the more unusual mutations. Two women were waiting for results; and for one it was a long wait, as there were several complications relating to the diagnosis of her son's haemophilia and hence her own status. Another woman described a rather unfortunate way of hearing the result: when a laboratory person rang her to ask to arrange for blood samples to be taken from some female relatives. When she asked why, he told her that she was a carrier. Both of them were taken aback that she had not already been told by her specialist or GP.

Summary.

Guidelines for consent based on the characteristic 'western' concept of the autonomous, independent individual raises some difficult issues for people with genetic disorders, where, in order to get an accurate diagnosis for one person, several biologically-related individuals may need to be diagnosed. Testing of one person also provides information about others, who have not given consent. For example, some men with mild or moderate haemophilia are not diagnosed until later in life. That diagnosis inevitably provides a diagnosis for any daughters the man has or will have. Similarly, the unexpected diagnosis of a son with haemophilia points to a maternal carrier.

The majority of people interviewed believed that with the child's assent, if she were old enough to understand, it was within the parents' realm and the child's best interests for parents to arrange for girls to be tested early. Many parents wanted to build up an understanding for the child of her status as a carrier and its implications as she grew; certainly well before her 16th birthday. However, this view was not held by everyone, and it was one of the few issues on which we heard a brief but heated public debate, with one protagonist asserting the girl's right to choose whether she was tested when she reached the age of consent, and in the absolute rightness of this position, and the other person, much more tentatively, putting forward a child development perspective. This person argued it was better for the child to know her status, so that she would be in a comparable position to daughters of fathers with haemophilia.

Approaches to Carrying Haemophilia to the Next Generation

A participant in her 40s with a son with severe haemophilia told Julie that a contemporary whose father had severe haemophilia had had a total hysterectomy before she was 30 years old "for fear of having a son with haemophilia". Yet when our participant heard her own son's unexpected diagnosis, she thought, "Oh, is that all?" It is in this context of widely differing perceptions of haemophilia that men and women approach the topic of 'carrying haemophilia on' to another generation, and make sense of the technologies that are available to help prevent this.

Views on "How serious is haemophilia for boys with haemophilia?"

To an extent, decisions to carry haemophilia on or not are shaped by experiences with haemophilia, which form people's views of how serious a condition it is. For example, a pair of sisters, who have featured in the news media because of their determination not to have boys with haemophilia and their campaign for PGD, have explained that their position was based on their experience of the suffering that their late father had undergone as a result of haemophilia. Many of our participants had read this story and expressed sympathy with the women and what they had gone through with their father and would be going through in trying to have children. Yet several commented that, while it was true that men who would now be in their 40s to 60s had had a particularly difficult time — with inadequate treatment, HIV, hepatitis B, hepatitis C, arthritis and other complications, such as drug dependency from repeated pain relief — for children born now, things were different. Haemophilia was now a manageable condition with improving prospects.

Pam and her family were new to haemophilia when their son was born about five years earlier, and their discussion illustrates this latter perception:

> We don't have the stories of years ago, when the treatment wasn't as good as today and we can read about the ones that are portrayed in the media with haemophilia and their father had it, and I felt desperately sad for the family because I think, "Life's not like that." And I find it so desperately sad that their views are so tainted by their history that they make these decisions, but once again they are their decisions. But sometimes I think, "If only they knew what life was like now."

The sisters who appeared in the newspaper were not alone in their determination not to have boys with haemophilia. A daughter, with a father of a similar age to their father, was equally determined. However, she would not accept any of the available tests and therefore, at least for the moment, was intent on not having children.

But it was not just obligate carriers who had witnessed their fathers' encounter with haemophilia who had come to this decision. A woman in her late 20s, having watched her younger brother with haemophilia grow up and feeling the pain he had suffered, had started to research the options available to her to avoid having a son with haemophilia. The other main group of women who were determined not to have a son with haemophilia were those who already had an affected son or sons. In several cases, the boy had experienced serious complications (such as bleeds into the brain or the central nervous system), difficulties with treatment (such as inhibitors), or complications of diagnosis. In other instances, the boys were doing well and the women and their partners had decided that one son, or perhaps two, with haemophilia was enough. Two young women, both new to haemophilia — one because of a spontaneous mutation, the other because she was adopted and did not know her family history — determined to have no more children, or no more with haemophilia, after their first sons were born.

Because haemophilia runs in families and we interviewed several members of the same extended families, we could see that sometimes women who had observed the same group of men with haemophilia had come to different conclusions. For example, where one woman in a set of cousins determined not to have any boys with haemophilia, another, Nadine, was much more equivocal. Nadine, a young woman whose brother and cousin have severe haemophilia, spoke to Julie about prenatal testing in general.

> Nadine: I think it's great that we, maybe, prevent ourselves from having children with haemophilia but, I've talked to my brother about this before and I sort of agree with him, he wouldn't be who he is now if he didn't have haemophilia. It's made him who he is. I'm sort of worried if we're preventing people from getting haemophilia. There are still the spontaneous people, and treatment for them is just going to be very hard because it won't be as available and it will be a lot harder for them, and there probably won't be groups like there is now because not so many people will have it.
>
> Julie: Oh, that's a very interesting perspective.
>
> Nadine: So I think it is a good thing in a way, but I am just worried about the future, really.
>
> Julie: So it's the effects if a lot of people use these techniques?
>
> Nadine: And I reckon my brother is a much nicer person for it, and the experiences he's had. He understands a lot more.
>
> Julie: So you think that dealing with that has been a valuable learning?
>
> Nadine: It's been really hard for him. At the same time, it does make him who he is.

Nadine had two major concerns. One was the social, political and treatment implications if haemophilia became reduced to only those who have 'spontaneous mutations', reckoned to be about 30% of haemophilia in any generation at present. She was concerned that such people, confronted with their surprise diagnosis, would not have the strong and well-organised community to call on that people currently had, and that services and treatment would be even more difficult for such small numbers. That concern was seldom expressed by others.

However, Nadine's other concern — the value of people who have struggled with haemophilia, and the implicit message selecting against haemophilia would convey to them — was often mentioned and debated. It was a major factor for some parents in their decision-making about prenatal testing, especially when they already had a boy with haemophilia. Pam and Peter talked with Julie about their thinking when it came to their second pregnancy:

Pam: And what do we say to him in years to come? Is that saying to him that he's not good enough?

Peter: In some ways, it is. You know, if you deliberately terminate other children that you know are carrying what he's got, you're telling him that there's something wrong with him.

Pam: And yet we tell him "You're perfect. You're exactly the way God made you." And while we wished that he didn't have pain … he is the child he is because of haemophilia. He's an amazing little boy, and I think of the people that we have met in our journey with haemophilia and we've met some of the most amazing people, and we are the people we are now because of what we have been through.

Peter: I suppose it also talks about our values, because we are both carriers of some genetic disorder. Everyone is a carrier of something, so, you can't eliminate all those.

Ashley had two boys with haemophilia and felt that at her stage of life her family was complete. Julie had asked her to think about the uses of PGD.

Ashley: I wouldn't dismiss it out of hand. If you're looking at starting a family, if you don't already have children, then you'd be more likely to use it, I think. If you've already got a child with haemophilia, you get into that … "If I don't want to have any more children with haemophilia and I do this, what am I saying to that child?"

Julie: And what do you think about that question?

Ashley: It's not a question I would want to have to answer. I was willing to have another child with haemophilia if that was what happened.

Nadine, quoted above, would agree with Ashley. She said to Julie: "I think if people say, 'Oh, we can't have this child, it has haemophilia', that they are saying that they are not good enough. I don't think people should be seeing people like that." However, some other young women would disagree. They thought that this interpretation was more likely to be made by parents, rather than by their brothers with haemophilia. One of them told Deon that this topic had been discussed at the Young Women's Workshop, where several people had raised the point of view expressed by Nadine. However, she said:

Personally, I wouldn't have seen that as a problem, you know. I think if it was me, I would not take offence to that … . I think it's [haemophilia's] not something desirable … . If they could have a choice, I'm sure no one would pick to have haemophilia, and if, I think you just view it as IVF, you know, and you are gonna have some selection there and the rest are going to go somewhere else. I don't think that my brother would ever see it as "we would rather not have you", 'cause that would be ridiculous.

These differences of interpretation as to the meaning of reproductive choices all have to be accommodated by the haemophilia community. What one person sees as a 'ridiculous' interpretative extrapolation, another may experience as a very hurtful negation of value.

There were also differences in views between fathers and daughters. In one example, the father often sent his daughter information about new research on treatments and prenatal diagnosis, stressing that his experience was not going to be repeated by a little boy born in the 21st century, whereas the daughter was quite adamant that she would not risk bringing a boy with haemophilia into the world and perhaps would not have children at all. The haemophilia itself and how traumatic it has been is important, but this is modulated and shaped by the context in which it is experienced.

Views on "How serious is haemophilia for girls carrying haemophilia?"

On the topic of carrying haemophilia on into the next generation by having a carrier daughter, there was some diversity of opinion. In the light of anticipated improved treatments and prenatal technologies 20 or 30 years down the track when daughters were likely to be thinking about having children, plus the fact that most daughters did not have major problems themselves, most participants considered that the potential for having a carrier daughter was not sufficient cause to not have children, to have a prenatal test if a scan showed a female foetus, or to terminate a pregnancy. Some had thought about passing on to their daughter the same decision-making dilemmas that they had confronted.

Frances: And again, I guess it is fairly selfish. I'd be fully happy to have a daughter who is a carrier and know that she would have to do the same thing as me, 30 years on. I guess it is pretty mean of me, but hey, I'd be far happier to have a carrier daughter than have a termination.

However, quite a few participants were interested in the possibility of PGD in preventing the birth of carrier daughters. Sue had been part of a discussion of this at the first Young Women's Workshop, and reported a debate about it.

…. I found it interesting that they can identify the males with haemophilia and so they can select

> against that, but, because it is not considered a serious condition just to be a carrier, if there is a girl that carries haemophilia they won't select against that. They will just let that go through. And we had quite a debate about that — that yes, being a carrier is not so serious. I have no symptoms of haemophilia at all, I have nothing, so I'm very grateful for that, so whilst I can appreciate that, I feel that if you can actually stop it being passed on — I know it always comes out through [spontaneous] mutation — but if you can stop the family lines passing it on, why wouldn't you? It's an expensive condition to treat, apparently *the* most expensive.

Marty, a young man with no children as yet, had not heard about PGD. After Deon had given a brief description, he said that he might consider using it, once he had checked it out more thoroughly, whereas he would never consider abortion even for a boy with haemophilia. Deon asked Euan, an older man who had completed his family and had a carrier daughter, if he thought testing should be available to couples where the man has haemophilia. Euan had not thought about this, and, although he believed that it should be available to such couples, had he and his wife been offered it he thought it would not have made the slightest difference to their going ahead with the pregnancies. But a little later on in the interview, he said that at the time his children were being born it might have made a difference.

At the Young Families Camp, in a discussion on reproductive choices, the haematologist presenting noted that usually it was the women who were focused on, and they certainly did have more choices, but men with haemophilia also should consider whether they wished to have carrier daughters. Some of the options mentioned for men were sperm donation or adoption. One young father who had 'married in' to haemophilia registered for the first time that his son could pass haemophilia on to any daughters. Yes, he was assured — but it was not 'could pass', it was '*would* pass' it on.

Albe was in his early 30s, and did not want to have children. He said, "I've always wanted to live a Peter Pan lifestyle and never really grow up." Having nephews whom he saw quite often was enough child-rearing for him. He allowed that this decision could be "a bit" related to having haemophilia, and, later in the interview, in an extended discussion of PGD, he talked to Julie about this in more detail.

> But I wouldn't want to have children, in some respects, because I don't want to carry on the haemophilia thing as well. I have thought of it like that. If I did want to have children I would have to think about it, [sighs] especially with haemophilia. [Speaking slowly and thoughtfully.] You've got to be very careful of it, because it becomes a little overly selfish, because if you risk having children who are going to cost society a fair whack, and there are precautions you could take, you probably should take those precautions. And I think I would if I wanted to have a family.

Deon pointed out to Neil that they had heard people in the haemophilia community talking about PGD, and about the right to choose the gender of their child, so they do not create a carrier who passes on the responsibility to the next generation. Neil concurred.

> I've certainly heard amongst males with haemophilia talking about the fact that they only want males, so as they don't pass the disease on to, I suppose you could call it, the third-generation carrier females that they father.

Views on "Whose issue is reproductive choice?"

Within the haemophilia community there is the same wide range of views about reproductive choices as there is outside of it. These divergent and often deeply-held perspectives have to be accommodated so as to not disrupt either personal relations or the work of the Haemophilia Foundation. Issues surrounding carrying haemophilia on are handled very carefully in the community, with different views on the seriousness of haemophilia, prenatal testing and abortion being respectfully acknowledged, avoidance of the topic practised where people are aware of intractable differences, and the articulation of an often-repeated principle that 'what is right for the family is what is right'. In writing about these issues in a report that is intended for the community, we are aware that we are bringing views into juxtaposition that much of the time would be kept apart. These views were offered by our participants with compassion and respect, and we do the same. We show that it is possible for major differences in worldview to co-exist within a single community. That is certainly a positive message.

Many participants mentioned that most things are very open for discussion within the haemophilia community, such as carrier status or family history, but perhaps prenatal testing was a more restricted topic. It was something that you would discuss with certain friends in more private settings. As Dr Julia Phillips, presenting on reproductive choices at the Young Families Camp pointed out — and despite Neil and Deon's reference to men's wishes not to have carrier daughters — reproductive choices are not generally regarded as men's issues and are therefore not usually the topic of masculine conversation. When Deon and Neil had a conversation about amniocentesis and CVS, they remarked on the gendered nature of such talk, in the haemophilia community:

> Deon: This issue, though I mean, it's something that, it's not your usual topic of conversation [they both laugh], but have you talked about it much with other people?

Neil: Umm, only with people involved with haemophilia, and I think only women, that I'm aware of. In fact, yeah.

Deon: So you don't think it is really an issue that men with haemophilia would discuss?

Neil: I have never heard it discussed among men with haemophilia — either of those procedures.

Deon: And why do you think something like that hasn't been discussed, considering its implications for haemophilia?

Neil: Well, it, um ... I think that's very relevant to a remark that we heard recently, and that is that, you know, it's not solely the woman's role to determine … the future of a child, and yet in a number of circumstances those two tests that we talked about, it seems to me, have been sort of largely, largely an area of 'women's issues'. And, apart from the topic, you know, not being as interesting as rugby, and that fact that it's into the women's issues sort of sector, maybe that's why you don't get half a dozen people, men with haemophilia, sitting around talking about chorionic villi sampling. [Laughter ensues from both sides.]

Deon: And while watching rugby it wouldn't really work, would it!

The humour in this exchange comes from Neil's gentle self-ridicule as a rugby fanatic, as well as from the idea that this is a ludicrous topic of conversation among men. It was significant that, when Neil *has* discussed it, women have been his conversational partners. This seemed to be the case of the others, too. For example, Euan said that one of his women friends in the haemophilia community had talked with him about amniocentesis, and he and his young adult daughter have had considerable discussion about the merits of prenatal tests, and Ben was going to talk with his mother and sister about these issues when the time came.

Discussion about reproductive choices is largely left to carriers and their husbands and partners. As Andy (mid-20s) said, "See, we haven't discussed it much, simply because she is not the carrier and we know what the odds are with our children." While this is understandable — as carrier women and their partners do have many more choices and more urgency about those choices — the perspectives and experiences of men and boys with haemophilia are important to a full appreciation of the repercussions of these choices.

Men with Haemophilia Consider Prenatal Testing

Nine adult men with haemophilia took part in this update study, and Deon interviewed all but two of them. The three who were over 40 were all married with children. One couple had adopted both their children because of haemophilia, and the others had three daughters and a son between them. None of the younger men had had children yet, although all but one intended to in the future. Several men noted that by having daughters they were passing on these difficult choices to another generation, and, as one man pointed out, those daughters themselves also have a chance of bleeding problems. Significantly, men almost never refer to themselves as carriers of haemophilia, although they do talk about passing haemophilia on.

As members of the haemophilia community, men with haemophilia have views about the desirability and the acceptability of prenatal tests. We particularly wanted to tap into these views, as noted above, as often questions of prenatal testing are seen as 'women's issues', or perhaps issues for carrier women and their partners.

All the men were in favour of the tests being available to people with haemophilia. Nonetheless, there were a range of opinions about the desirability and use of testing. Marty, a young man, reflected on his own situation and believed these tests could be useful to prepare for early treatment of haemophilia:

> I reckon they [tests] should be available, because if you can test for it, you can start, probably treat it right at the start at least, instead of waiting, like I was probably about two before they knew I was a haemophiliac, so [it] could have saved a bit of pain and worry.

He added:

> I don't think they should abort if they are having a haemophiliac.

As noted above, Marty did not know about PGD, so Deon explained to him that it is "being able to test human embryos…. The embryos are created in-vitro and then cells are removed and genetically tested for haemophilia, then the unaffected one(s) would be implanted and the affected one discarded". Compared with amniocentesis or CVS, Marty thought PGD was quite a lot more desirable "because you don't really see a face". He did not disagree to the same extent if the embryos with haemophilia were discarded, in contrast to his belief that foetuses with haemophilia should not be aborted.

Eric felt strongly about his daughters being able to avoid having a child with haemophilia. He said that, as well as considering the child, and the impact on the health service, it was the impact on his daughters that a child with haemophilia would have that led him to his perspective:

> I know that it would dictate so much of their life for 20 years and beyond.
>
> We've got certain moral views and Christian beliefs and all that sort of stuff, but at the end of the day, you know, we've got daughters

> who are growing up and they're well informed and well able to speak their mind and they are brought up to speak openly about stuff. And I think I would be failing as a father or as a parent if I didn't tell them what the options were and give them every option that was available and let them make whatever decision they wanted to make … But, they need information, they need to make a good decision, and certainly with some input from someone who knows haemophilia for over 40 years, you know.

Others were less keen on prenatal testing because of their view that haemophilia was not such a serious condition, and a child can be tested at birth if haemophilia is suspected. Projecting himself into a woman carrier's shoes, but reflecting on his own life, Justin, a young married man, said,

> I mean I've had a great life, you know, like, you could say "Oh, I was so unlucky to get this thing, it's so rare!" And there's the difficulties that came along the way, but it just makes you who you are and I wouldn't, you wouldn't, change it, but you know, nuh, if I had a haemophiliac kid I wouldn't [be] bother[ed] at all, eh.

Albe took a different position, as he did not want to have children, partly because of haemophilia, as noted above. But he was not keen on PGD, saying, "I think the whole thing would be rather unsexy." He had considered the possibility of having children:

Albe: If I really wanted to have a family I would want it to happen naturally. It's 'designer babies'. It comes under the general topic of 'designer babies', in a way, which I was watching a doc[umentary] on, nothing to do with haemophilia. It was all about [puts on a vacuous, upper-class voice] "Should we have blue eyes or should we have brown?" [Laughs.] Big choices! [Ironic.] Which are in many ways kind of a bit freaky, I think. It's fashionable people. It's about fashion.

Julie: Oh, okay.

Albe: Yeah. I'm not too opposed. I think we've always got to be careful about —I've got some ideas, but are they right or wrong? And I'm not sure. And I think that some of the most scary people in the world are ones who really truly believe in a particular way.

A little later, having noted that there are always people like the Nazis around, he asked, "But okay, at what point do we (as a society) say 'No', to the choices that individuals want to make?" This comment was one of the few that suggested that there could or should be some limits as to 'what was right for the family', as Pam and so many others said.

Neil, Eric and Andy were all quite keen on PGD. Neil had been reading up on it. He found it attractive because it was not invasive in the sense that the cells were tested at a very early stage in the laboratory, although the embryo had to subsequently be implanted.

> So you've got the situation of being able to grow those single cells up to an eight-cell stage, and at that point determine not only the sex, but … whether those cells do actually contain haemophilia. And it seems to me that to have the opportunity to then discard those haemophilia cells and select obviously the healthy cells is absolutely wonderful.

Another advantage for Neil was that it would help carrier women and their partners avoid:

> … drama with respect to the first pregnancy, that mightn't end up as pregnancy at all. So I think there is all those sorts of psychological issues that both parties really have to grapple with, and to me, you know, it seems such a contrast with [amniocentesis or CVS] — the likes of PGD as an option that seems, you know, so much more, almost normal.

Andy believed that PGD would eventually supersede other forms of prenatal testing, and it would certainly be his choice, although knowing he was not going to have any boys with haemophilia he might not exercise that choice. He thought there was less risk for the woman and the foetus from PGD and it would be more acceptable because the child did not seem as tangible, being "at the cellular level". "It feels less like an abortion" because of this.

Eric thought that the Government would be crazy if it did not make PGD available for people with haemophilia, but he made the point that very good counselling needs to be available too, so that people are aware of the implications of their decisions before they make them. He added later, after stressing the personal benefits of having reproductive choice: "It makes economic sense."

Euan considered the availability of PGD desirable for the haemophilia community, but was not sure how acceptable it would be. He had read about PGD in *Bloodline* and had told his daughter about it. His view was that this technology was quite complex and that there could be some resistance to it because it was — he struggled to find the right word and was not totally happy with his choice of — "unnatural". He reported that his daughter did not find it any more acceptable than other forms of testing, because it still required a choice to nurture or destroy an embryo. Although he was hopeful that treatment improvements would mean that prenatal testing would not be necessary in the future, and testing at birth would suffice, he did concede to Deon.

Euan: Let's face it, none of our parents had an easy time bringing us up. Despite [clotting factor] concentrate and despite all the other benefits that we've accrued over the last 20, 30 years, bringing up a haemophilic child is a chore, it's an inconvenience. And bringing up kids in general is an inconvenience, so—!

Deon: Add that on too! [They both laugh.]

Men with haemophilia have a range of views about reproductive choices and the seriousness of haemophilia: from not passing it on at all by having no children, to being happy (imaginatively) to have a kid with haemophilia, to not agreeing with abortion for haemophilia. The topic of PGD has provoked considerable thought and its advent was generally welcomed. Although a couple of the younger men might possibly consider using it if they and their (future) partner were considering having a baby, they had not yet informed themselves about it or discussed it with their partner, or did not yet have a partner to discuss it with. Amongst those who had informed themselves about it, there were differences of opinion about its acceptability, and its 'normality' or its 'unnatural' characteristics.

Women Carriers and Their Husbands Talk About Prenatal Testing

The participating couples were all women carrying haemophilia and their partners. Three of the four already had children, and the fourth was hoping to become pregnant shortly. Two of the women were obligate carriers; one carrying severe haemophilia, the other mild. The other two women were diagnosed as carriers only after their first-born boys were diagnosed with severe haemophilia. One of these boys had inhibitors. Julie interviewed all of the couples.

We have presented the analysis of the interviews with couples separately, because, as the excerpts show, couple interviews are like small group discussions, in the sense that a lot of the conversation is between the partners. These excerpts give something of the flavour of the relationships between the partners as they describe the pathways that they have taken or plan to take through the maze of reproductive choice. Their views and decisions are not different from those described by the other men and women interviewed.

Murray and Marie had known that there was a chance that they would have a boy with mild haemophilia or a carrier daughter, as Marie's father had haemophilia. They vaguely recalled having been offered amniocentesis, but they opted to have "just the normal ultrasound" (Murray), which determined that they were having a boy. There is a risk to the foetus with the invasive tests, and they were going to have their baby "haemophilia or not", Murray explained. "So what was the point of having the procedure to find out, when you can find out when he is born a lot more safely?" Their baby, born within the past year, had mild haemophilia.

Julie asked them to consider what they might do if the haemophilia in their family were severe. Murray was impressed how well children with severe haemophilia did these days, with prophylaxis. He acknowledged that he would have second thoughts about it if the child's future had been going to be like that of guys over 35 who, he observed, have major joint problems. Marie was a bit more hesitant about this and thought that they might have a prenatal diagnosis if they were a severe family, but that they would be most unlikely to terminate: "I don't think I could possibly do that." Murray thought that if the haemophilia were severe, you would treat the whole birth procedure more cautiously, perhaps going for a Caesarean section.

A slightly older couple, Pam and Peter, had unexpectedly had a little boy with severe haemophilia, who had been diagnosed not long after his birth. When Pam was pregnant with their second child, they had gained the impression from their haematologist that prenatal testing was available only if you chose to terminate an affected foetus. This was explained to them as in terms of the risks of damage to the baby outweighing the risks of (not) knowing. They were told that they could just find out the information from a scan. However, as Pam was not an obligate carrier, a scan would only have given them partial information. They thought this was "in some ways reasonable" (Peter) and decided not to test, as "we knew that we wouldn't ever terminate" (Pam).

Even though testing was not for them, they valued it for the haemophilia community:

Peter: Yes, it's good to know. It's important that people have the ability to know what they're having so they can prepare.

Pam: I think that if it is important to the parents it should be offered, regardless of whether they are going to terminate or not. If it is important to the parents, it should be offered.

Pam and Peter acknowledged their joy and relief — and that of their whole family — when they found that their second child was a girl. It was only then that they realised what a burden they had been carrying, not knowing the diagnosis. A major consideration for them in the decisions that they took was the message they would implicitly convey to their son if they did terminate a pregnancy or use PGD, as described earlier.

Later in the interview they came back again to the issue of using technology to attempt to eliminate genetic disorders, and the moral and ethical, not to mention practical difficulties this raises because of the thousands of possible genetic conditions that humans carry.

Peter: You can't live in a risk-free world, and everyone's taking a risk, whoever is having a child. Every parent can have a child who turns out to be disabled, whether it's through genetics, or birth or an accident. And everyone is carrying that same risk. We just know it and they don't.

In this they were echoing Albe's concern about what the limits of testing were and how a society can arrive at some agreement on this.

Anna and Art, like Pam and Peter, also had a surprise diagnosis of severe haemophilia for their son, but this was not made until he was nearly a year old. When Anna became pregnant again, she had a scan and found she was carrying a girl — "a sigh of relief" for her — but Art had not wanted to know. Anna said that if it had been a boy:

> I would have been "Oh, what's going to happen?" I don't think I knew of any way ... at that stage. I only knew of amnio. So I think [testing] is a good idea, but for me it wouldn't change having the baby.

Julie asked about any downsides to testing.

Anna: It probably makes it harder, it does put pressure on you, then, if you do know there is something wrong, that you can, if you wanted to, do something about it. And that guilt. Sometimes it's best not to know, maybe. Hard one.

Julie: Yes, none of it is easy.

Art: Without being in that situation, it's hard to justify what decision you would make. I think it would be very tough, if we had been in that situation. It would be very tough for us, because Anna was going to have a second child and we were not going to worry about the testing side of things, and I was saying, "There is that risk there. Don't you think we should just put our resources into our son?" We had a fair bit of debate, before having our daughter. It's great having the second child, but if that testing had been done and then you had one of you saying "Look we've got to terminate", and the other one saying "No, I'm not", it could be a real toughy.

Anna: I couldn't see myself with just one child, I also didn't want my son growing up as an only child.

The potential dilemma outlined by Art was exactly the kind of difficulty that Neil thought the use of PGD could avoid. Anna and Art did not have to confront a male foetus with a positive result for haemophilia, but one of the other couples did.

The woman explained that she and her husband had made up their minds beforehand to terminate if that were the result, but "when push came to shove, it was very difficult. Never do it again and never have a CVS test again." She explained that if she got pregnant again, she and her husband had decided that they would not have any prenatal tests.

A woman who has grown up in the haemophilia community has had many years and much practical experience of haemophilia on which to base her decision. A husband or partner, relatively newly exposed to haemophilia, is in a different situation. The husband said:

> Yes, it was very hard for me. It was my son. Because of what was happening in his blood. Yeah, logically, when you used your brain it says, okay, you are not going to let him suffer. I don't know how much pain he went through. I don't really know, I don't know at that early stage. But it's needles all the time (once he is born), you know, with severe haemophilia, three times a week, and then they can't have a normal life — doing sports activities. So logically, your brain says that it's a good thing, you are doing the right thing, but your heart is the other way.

Friends of couples who have gone through CVS and terminations have witnessed their trauma and suffering. Whether the friends have agreed or not with the couple's decision, they have tried to provide support and, from the way they spoke, they clearly empathised with the pain of this decision-making and its consequences.

Marie had been to a talk about PGD at the New Families Camp in January and was familiar with it, but Murray felt he needed to read up about it as he had missed that talk. Marie explained it to him, and they chuckled over the possibility of multiple births. Although she was glad it was available for people with severe genetic disorders, Marie would not consider it for herself, even if she had more severe haemophilia.

Marie: 'Cause I hear the IVF side of things can be quite rough, but then I also sort of think that if you have an embryo that you find does have haemophilia and then you don't implant it, and then what happens to it? And it's, yeah, to me it's a life, even though it is at the cell stage, but still now that I've had children I don't think I could bear to [discard an embryo].

Julie: It would worry you?

Marie: Yeah, not that I'm religious and believe … you know, I don't believe in that type of thing [i.e., she is not a pro-lifer]. It's just my personal [belief] …

Murray: With the prophylaxis thing going on, and home treatment … it is more manageable, but I guess it comes down to how manageable and the amount of money to make it manageable, too. If that wasn't there it would definitely be difficult, you know.

Marie pointed out, too, that without any intervention she had a 25% chance of having a boy with haemophilia and a 25% chance of having a carrier daughter. Yet if she underwent PGD she would have only a 30% chance per IVF cycle of having a baby at all.

The views of this couple were at the opposite end of the spectrum to those of Debbie and Don, who were

seriously contemplating PGD. Debbie was an obligate carrier of severe haemophilia. She had witnessed her father's suffering and premature death due to a range of complications caused by haemophilia and its treatment. She explained: "From an early age I knew that I would not have a child with haemophilia. It didn't ever cross my mind that I would have a child with haemophilia." Julie interviewed this couple on the day that the funding for PGD had been announced, so it was particularly topical.

Julie: So from your perspective, what are the advantages and the drawbacks of this new technology?

Don: Drawbacks — is the whole procedure.

Deb: Injecting yourself every day.

Don: It has some risks for Deb … and the babies. So far, they say they are fine — so haven't heard anything really bad about it. There are some risks but really little, but the positive is deciding between some cells — well they are going to be a baby in the future, but they are still cells, not a real baby. So we think it is more painful for Deb to go through, but then it is easier to decide.

In contrast, Pam and Peter had discussed PGD with each other and had come to the conclusion that it was not for them, even though their son had severe haemophilia which was proving difficult to treat.

Pam: I struggle with … I look at our son now, and I think: so we wouldn't have had him. And I find that incredibly sad, because he is the light of our life!

Peter: In some ways it raises more issues because you have extra eggs, extra fertilised eggs and um …

Pam: And our belief is, for us, that there is a child from conception and we struggle with [that].

Art and Anna had been learning about PGD through *Bloodline* and a television programme. They thought it was a good idea, so long as "science doesn't go crazy" and specifically mentioned "the two girls in the *Sunday Times*" for whom it provided a solution.

Art: As long as the ethics are sorted. I think it's a great option to the extent that the likes of the testing [that] you [Anna] were talking about with the foetus. And that when we were looking at [having our] daughter, one of your big things about not doing it, I remember you commenting to me that "They said the risk is minimal, but it is still a risk." Why be invasive and go down that track as well? In this case, you are doing it early so it is not invasive. And there is still lots to come out about it, but it certainly seems that they're developing along the lines of what is required to address it, regarding the medical issues or as you [Anna] say, science gone mad, or the designer child or things like that. So as long as there are those checks and balances, I think it's a fantastic option to have available.

Anna: Save a lot of money, wouldn't it?

Art: Yes. I was going to say that the concern is that if they don't fund it, but it's available, only rich people are going to get that choice, or if you are talking about your daughter that's a carrier, you're limited to having one child or one child safely, because the budget doesn't allow you to spend whatever number of thousands of dollars it is every time.

Julie: So you'd be in strong support of having a minimum number of cycles publicly funded?

Anna: Yes, a couple publicly funded and then private, because there is only so much everyone else should be expected to pay for your … whatever—

Art: Subsidised system.

And a few days after this interview, this was exactly what was announced.

Women Consider Carrying Haemophilia On

Twenty women were interviewed alone, but only six of them did not have partners. In some cases, Julie and/or Deon also knew or had met their husbands or partners. Four women were sole parents. In one instance, the couple had separated mainly because the man had found it difficult to deal with his son's illness, although he did keep in regular touch and there was a possibility for family reunification. In the other cases, we were not told the circumstances and did not enquire. All but two of the partnered women had children. Deon interviewed five of the women, all of whom he knew already.

For most participants, the physical test itself — whether amniocentesis or CVS — was not a major drawback, although unpleasant. The main concerns were having CVS done too early and damaging a foetal arm or leg, and the chance of miscarriage. However, two women had experienced untoward events. One missed her CVS because when she went for the initial scan no baby was visible, then by the time she had her next scan the foetus was visible but her pregnancy was too far advanced for CVS. Another woman had such a bad experience with her CVS — with malfunctioning equipment, possibly incorrect timing and health professionals who did not appear competent — that she and her husband had made a complaint to the DHB. She said: "I would never have another CVS test done as long as I live, and I would never, ever go back and see that doctor." This greatly limited her choices, as she was not prepared to have a baby with haemophilia.

Although for most the test itself was not a big issue, deciding whether to have a test — or deciding what to do if the results were positive — was a major concern for nearly everyone, although on this topic, as on most others, there was a range of opinion. Ivy and her husband had made up their minds before they had the test what they would do.

Ivy: I mean it is easy to say, but I also said that we are so glad that we didn't have to make that decision because, when you get in there and have your CVS done, and the ultrasound and you see the wee heart beat, if that had have come back [boy with haemophilia] it would have been a hard thing to do.

Julie: Absolutely, a terribly tough decision.

Ivy: But luckily I didn't have to make it [because the baby was a girl].

Sue had been discussing CVS with Julie, and went on to point out that there was a big difference between thinking ahead about options and making an on-the-spot decision.

> I am very, very passionate about not having a child with haemophilia. If there's an option to have a child without it, I would certainly go with that. However, I know, not having any children yet, when it comes to the time, if that was an option to abort the child, I'm not sure. I think it is a whole other experience to go through, when it actually happens to you, and you need to make that decision in reality, not just in theory. That can change a lot of things.

Tui had unexpectedly had a son with severe haemophilia and some serious complications. She learned at that time that she could have an amniocentesis if she wished to have another baby. She did not recall being offered CVS, possibly because she lived a long way from a major hospital. She was in two minds about testing.

Tui: But otherwise, that is a hard one, I don't know. I can see the point, for myself with my son, and him being severe, and thinking that I don't want another child with haemophilia, with all the heartache, and what he'd have to go through, knowing that he's got haemophilia, yeah. I suppose in another way, we'd be more prepared for it.

Julie: Yeah.

Tui: So it works two ways. And then I would have my son to show the younger one how to do it, maybe. You know, but then, I thought to myself, "Do I want to put another child through that?"

However, on a visit to a matakite (a Māori spiritual seer and counsellor) who was helping her get through her son's health problems, she was informed that her next child would be a daughter, so she did not feel the need for a prenatal test and gave birth to a little girl who was not a carrier. After that she had a tubal ligation, as she did not want to go through another pregnancy "with something like that. A girl and a boy, that's me! And I've got my nephews, so that's something." The "something like that" was complex. Julie inferred: the possibility of haemophilia, the worry about the risks of testing, and, if the test were positive, knowing that "we'd really want to keep it, that's why I thought that was really hard" — the pain of deciding.

Ashley had two sons with severe haemophilia. When she was pregnant with her first son, she was not sure if she was a carrier as the family mutation had not been identified. She was offered amniocentesis because of her age, but decided against having it. She pointed out that it could only tell her if she was having a boy or a girl, and she and her husband felt that it was not worth the risk of miscarriage. By the time they were thinking about a second child, they knew about the possibility of haemophilia, but Ashley relayed, with a laugh, "As my husband said, 'One is not a family, and if we have two with haemophilia, well, it was the way it was going to be and the same house rules for both of them'!" Again, she did not have an amniocentesis, and she does not recall being offered CVS. She had the "normal scans", and another one at a later stage to confirm the sex to prepare for the birth.

Like Ashley, Carolyn was offered amniocentesis because of her age. However, as a scan had shown no abnormalities and age was the only "risk factor", she and her husband decided not to have the test. At this stage she did not know that she carried haemophilia. She was not offered CVS and had not thought it was used in New Zealand. However, she did say that if she had known that two risk factors had been present she would have had the test, and would probably have had a termination if anything serious had showed up. No one would have thought to test for haemophilia, though.

Trish was offered CVS in her third pregnancy. She herself was prepared to accept whatever came, but her husband was anxious to know if they were having a boy with haemophilia. She was a little cautious because they were not considering an abortion, whatever the test result, and she knew that there were risks to the foetus and the pregnancy. However, she felt that, given her husband's need to know, the test was warranted. They were completely frank with their haemophilia specialists about their reason and their decision, and this was accepted. However, when they attended the women's hospital to make the arrangements for the test, they found that the staff:

> … talked about the cost of the test and of course they wouldn't be offering it unless, if the child were affected, they would abort the foetus. So we had to pretend that that was what we would do, in order to get the test, which felt

> horrible, and I was a little angry about that, but I knew that we didn't have much choice really. So we just played along and said that was what we were doing, in order to get the test. And of course we discussed it beforehand and we said, well, if the results come back positive, we'll say, "We've changed our mind now, thanks very much." So it felt a bit underhanded, but that was what we had to do at the time.

As their third child, a boy without haemophilia, was 15 years at the time of the interview, this emphasis on the costs of testing might be dismissed as a thing of the past. However, in another part of New Zealand, a woman whose first child was a son with haemophilia had been offered CVS for her second pregnancy, under exactly the same strictures about cost and termination, except this time it was the haematologist who laid down the law. Again, she would not contemplate abortion for haemophilia so, as she said, she told a "blatant lie". Her now primary school aged daughter turned out to be a carrier so she did not have to confront her haematologist.

Women Talk Specifically About Preimplantation Genetic Diagnosis.

Not surprisingly, given the variety of responses to PGD by men with haemophilia and couples, women also had a range of responses, ranging from rejection to enthusiastic endorsement. Most, like Ashley who is quoted next, were somewhere in between:

> If you are going to do that, then you are not wanting to carry the gene on into the future for the benefit of your family and that's why you are doing it. Would I personally do that sort of thing? …I'm past my having children stage so it's a very hard one to answer. If I was a young one looking to start my family, I think I would want to know that it was available to me, and then I could make my decision to just take what was given to me, or have some choice in it. And I'm not — sitting here in this seat now — I'm not sure whether I would or whether I wouldn't, to be honest.

Lois, whose family had severe haemophilia, expressed a view that was very unusual among participants, namely that the Haemophilia Foundation, through its outreach workers particularly, should recommend prenatal testing, and especially PGD, about which she was enthusiastic.

> Lois: Well in principle I think they [the range of tests] are all a good idea. I think they should be strongly recommended to people rather than people just producing babies with haemophilia when it's possible to avoid them, avoid having children with haemophilia … But I think PGD is superb, should be recommended to our people by our outreach workers and by the Foundation and, you know, just strongly recommended.
>
> Deon: And what do you think makes PGD better than the previous ones?
>
> Lois: Well, I think there's more chance of avoiding a child with haemophilia, isn't there, than the other situation? Like one of them they used to do was if a child's got haemophilia, well you've got to have an abortion, don't you?
>
> Deon: Hm.
>
> Lois: So, yeah, I don't think that's such a hot idea.
>
> Deon: Yeah and—
>
> Lois: I'm not into abortion, but I haven't got any problem with selecting eggs, selecting the right egg that doesn't have the haemophilia gene.
>
> Deon: So you think that PGD should be something that we are, that the Foundation's promoting, and that people have lots of information about so that we can—
>
> Lois: Certainly. I know that people go "Well, you should just give people, give them information and give them a choice", but I think people are often looking for a recommendation, and I think it's the one that should be strongly recommended. I don't think there's any excuse, like as soon as it's funded I don't think there's any reason for people to produce people with haemophilia unless it's just the mutant gene just arrived in their family that they didn't know about, but I think any carriers should be clearly identified, should have all the facts and should be strongly recommended to go through PGD. I don't think, and people say "Oh well it's a big deal for women to go through." I think, "So what? It's not a big deal." It's a much easier deal for the woman to go through that than for a boy to be born with haemophilia all his life.

This last point — that PGD is a trial for women to go through, certainly, but is nothing compared to the pain and suffering of a boy with haemophilia — was made by several other women who had sons with haemophilia. They pointed out that it was easier to bear some pain and anxiety yourself than to stand by and watch your child suffer. However, none of them suggested that it should be a recommended procedure, and the outreach workers saw their roles as providing information and support, not recommendations.

Sue had been learning about PGD and, like several other participants, was initially excited by the idea, but as she learned more she became less enthusiastic:

> Initially, when I first just heard about it as PGD and they can take the embryo or whatever you call it, at a real cellular level, discard — terrible word. They can see, identify males with haemophilia, etc., and the first I heard about

> it, it sounded like a simple concept, fantastic, that solves all the problems. It's easy, brilliant, yep, I can do that. It's got a price tag — doesn't matter, that's fine. However, the more I have learned about it, it's quite … it sounds like quite an ordeal. It's a real process, and just the whole in-vitro — is that what you call it? — fertilisation, just that whole process and doing whatever they do hormonally to get you in the right cycle, etc. Even that concept, just doing something that's not totally natural, just goes against most of my philosophies … So initially I was a lot keener.

Despite these problems, Sue thought that morally and ethically PGD was preferable to prenatal testing and termination.

Frances was unable to get to the Young Women's Workshop, but she had been doing her own research by reading medical journal articles and articles in haemophilia magazines, and had come to similar conclusions about PGD, having started from a position of scientific scepticism:

> I think IVF is really, really invasive in the first place. But if people can't have children and it's their option, then great. But if I'm capable of having children, I'm not for it … I'm not for the idea of being pregnant and being married to an obstetrician for the entire pregnancy in the first place, and I think if I have IVF I would be. I'd be in the boat where you have to go to the obstetrician and do as you are told, and it's not really for me. You'd have to go to a hospital and you'd have to go and have scans and, yeah, I've read all the WHO research on ultrasound and Caesareans and induction drugs and everything, and I'm not for it.

Carolyn and her husband had had a boy with (unexpected) haemophilia and wanted to have another child. She was seriously considering PGD, although she realised that physically it was a tough process for a woman. A colleague of hers had gone through IVF for infertility, so she had had the opportunity to observe the physical and psychological effects.

> I don't have any moral objections to it, that sort of thing. I probably would have if we hadn't been in this circumstance of possibly wanting to have another child. … What I meant was that, just prior to having children, I probably would have said, you just sort of need to let nature be. I would have, that would have been my general opinion about, you know, stuff you see on TV, the technology and stuff like that, yeah. So. But now, I think that for a couple like us who may want to have another child, then it's [PGD's] a really good option.

To her, CVS sounded "a bit more natural, obviously you are conceiving naturally, there is less people in the mix, there's less sort of intrusion in your body, whereas the IVF, it doesn't seem so private". Nonetheless, because PGD takes place so early and does not involve abortion, for her it was preferable.

Although Karen was not as familiar with PGD as some others, to her also it was a greatly preferable option should she wish to have another child. Barbara, too, who could never have an abortion — not because she was against it, but because "there's nothing wrong with my son" — would opt for PGD, especially as her husband had recently had a vasectomy because they were not prepared to have another child with haemophilia. Coping with the child they had — emotionally, financially and employment-wise — was proving difficult. Others, believing that human life began at the moment of conception, did not find PGD an ethical advance on existing techniques, whereas some others did not believe that haemophilia was serious enough to warrant such interventions for themselves, but were pleased that it was available to others who did feel they needed it.

Summary of "Carrying Haemophilia on to the Next Generation"

This chapter has considered the responses of people with haemophilia and their families to haemophilia as an inherited genetic condition passed on by both men and women to the next generation. Because haemophilia is usually but not always 'silent' in women, most girls and women need to be tested if they wish to discover whether or not they are carrying the specific gene mutation for haemophilia. This testing was accepted by everyone we spoke to, but there were vigorous debates about when it should be done and who was responsible for initiating it. We also discovered considerable variation between health practitioners and clinics in their advice and practices on carrier testing. Compared with our initial study 10 years earlier, delays in receiving test results were very rare.

The specific demands of genetic testing for a familial condition means that the usual presumptions of individual privacy and autonomy are not met. This may be an area that haemophilia specialists and the HFNZ, perhaps in conjunction with the New Zealand Organisation for Rare Disorders, could usefully debate with the relevant ethics committees and the Privacy Commissioner to establish more satisfactory guidelines based on a presumption of human connectedness. We found it was causing a degree of angst among parents.

Prenatal testing using amniocentesis, CVS or PGD was also a topic which many people with haemophilia in their families had debated: for the most part in private. We found it was a discussion topic that women were much more likely to engage in. However, we discussed it with all the men with haemophilia, as well as with the husbands of women carriers and with the women

carriers themselves. Although most people with whom we engaged would not use these methods of preventing haemophilia in the next generation themselves, everyone with whom we spoke felt that there was a place for these technologies for people with genetic conditions. A frequently offered guiding principle was 'what is right for the family is what is right', although a few people noted that this principle could be questioned, as families might decide on trivial grounds and ultimately 'society' should have a say in such ethical matters which have eugenic potential.

Some people would not use these technologies themselves because they felt they could not personally countenance abortion or embryo selection. Very often, such people explained that this was not for religious reasons, rather, they just could not go through with it, but they accepted that other people could. Others did not think that haemophilia was a serious enough condition nowadays, with good treatment, to warrant the use of these techniques. While this judgement of seriousness was partly based on past experience, we found that people who had had similar past experiences came to different conclusions.

A further consideration, expressed by carrier women and their husbands, but not by men with haemophilia, was the effects on living boys and men with haemophilia if the birth of babies with haemophilia were prevented. Some women and their husbands felt that it conveyed a message of worthlessness. Other women, and all of the men, disagreed with this and saw it as simply aimed at preventing pain and suffering in the next generation, as well as limiting the demands on families, and in terms of the provision of expensive health services, on fellow citizens and the State. One woman expressed concern for the haemophilia community in the future if everyone who knew they had haemophilia prevented births of babies with haemophilia. This would then leave only the 'spontaneous' families, shocked with their surprise diagnosis, to cope on their own.

Only two of the nine men with haemophilia had taken a decision not to pass haemophilia on to the next generation. Women carriers whom we spoke to tended to limit the number of boys with haemophilia to one or two, and some were determined not to have a boy with haemophilia. Among younger women, a debate had arisen concerning the use of PGD to prevent the implantation not just of male embryos with haemophilia but also female embryos with the affected gene. While a few couples had used CVS or amniocentesis, or intended to use PGD to prevent the birth of babies (or another baby) with haemophilia, more used conventional birth control methods, including tubal ligation and vasectomy, to limit their family size.

In contrast to our earlier study, more people had thought about the costs of haemophilia to their health services and to the nation, and mentioned these considerations when they talked about limiting their family size. The expression of such considerations had been very rare in 1994 and 1995 (see Park and Strookappe 1996, Park 1998).

Chapter Five: "The Shadow on your Life": Living with Haemophilia and Hepatitis C

In Chapter Five we concentrate on the changing experiences of those participants and their families who contracted hepatitis C through clotting factor replacement products. The New Zealand blood supply was not free of this virus until the beginning of 1993, and consequently the majority of people with haemophilia who received treatment before that time contracted it. We outline the implications of this health disaster for the haemophilia community and how the HFNZ has sought to deal with it.

Personal Experiences with Hepatitis C and its Treatment

Well over one-third of our participants, including one woman, had hepatitis C themselves, or had an immediate family member who had contracted it: father, son or brother. In this part of the report, we outline some of the experiences of people with haemophilia and hepatitis C and its treatment. This RNA virus is usually called 'hep C' in this community, and we follow this practice as well as using the more formal nomenclature.

The particular significance of this research on the changing experience of people with haemophilia and hepatitis C lies in the comparative and cumulative nature of the research. Our New Zealand study of people with haemophilia and hepatitis C is part of a growing number of qualitative studies that focus on the individual experience of hepatitis C in other populations. Although each of these studies is small — e.g., 20 (Gifford 1999), 11 (Crossen *et al.* 1999), 22 (Garrett and Conrad 2001), 20 (Harris 2005) and 39 and 15 in the "Living with Haemophilia in Aotearoa New Zealand" studies — taken together and with the individual accounts of experiencing hepatitis C (e.g., McLean 1999), as well as accounts by support workers (e.g., Booth 1999a, 1999b), a rather more robust picture is being created. The more optimistic prospect created by new treatment regimens is an important part of the overall assessment of the effects of blood-borne hepatitis C on people with haemophilia and their families.

We begin with Justin's story, then outline how it differs from or is similar to the experiences of other affected people. Justin, a young married man when Deon interviewed him, learned that he had hepatitis C shortly after he started high school. He and his brother did not have severe haemophilia and so were treated at home with clotting factor on an infrequent basis; nonetheless, both became infected. For him, it was "the worst thing" about having haemophilia and "the shadow on your life" that meant he "did not know what [his] life expectancy was". His goal through high school was to live to age 26: a goal achieved.

Like many others, Justin and his brother found out their hepatitis C status in 1992, and were told by their doctor that no one knew much about hepatitis C, and it was possible the brothers could live their whole lives without it affecting them, but some people did get sick with it. Justin explained that no one really talked about hep C at home, the idea being that if it did start to affect them, they would talk about it then. He almost forgot about it, except it was "always there at the back of my mind", and the topic did sometimes come up, such as in discussions with close friends or during his visits to the haemophilia centre. In about 2000 or 2002, he recalled, he was contacted by the haemophilia centre who suggested he go and see a gastroenterologist. He had left home at this point, and he decided that he wanted to take ownership of this illness and find out what was going on, as he did not know much about hep C. For example, he had had a period of depression that he said was because he thought he could not have children, and he really wanted children. A big break-through occurred for him at the Hepatitis C Conference in Wellington early in 2005. He had not had much to do with the haemophilia community until then, and he found it very reassuring to meet so many people who shared his experiences.

In 2005, he started the new treatment regimen of pegylated interferon and ribavirin for up to 12 months. As he was still on it during his interview, the outcome was not yet known. Deon asked Justin about his experiences with the treatment, and specifically asked about symptoms, something that typically Justin tried to ignore himself and did not talk about with others. He revealed that he felt tired, had flu-like symptoms the whole time, burned easily in the sun, overheated easily, lacked energy, had lost his appetite, and had lost a lot of weight — the loss of three years' work in the gym on building up muscle. He had periods of being very short-tempered, suffered depression, and had skin problems, such as rashes and athlete's foot.

When he began his treatment, he had recently finished his university degree and had just started a new job. Although he was anxious about his lowered capacity for work, his boss was very understanding and encouraging. Reflecting on how he felt during his treatment, Justin said that, despite the symptoms listed above, "from what they've said it could be and how it has been, I've been really, really lucky". Later he added,

"As long as I can walk and stuff, I'm pretty happy, eh. I just look at that and say 'Well at least I can walk'!"

Justin's is one person's hepatitis C story. Although Justin's experience was relatively common, and some other men's stories differed in just minor details, everyone's experience was unique. Some people had been very sick with hepatitis C before their treatment began; others were not able to access treatment, or had had treatment reduced or interrupted because their white blood cell count had got too low. Some got rid of the virus early on, with just six months of interferon; others went through that treatment only to have the virus re-surface again after treatment stopped; while others paid for an extra six months of interferon treatment. Older people have often had hepatitis B as well, which has damaged their livers, and some, of course, also had HIV. Some people with haemophilia have died from hepatitis C. A few are not clear about their hepatitis C status and whether it will ultimately affect them. For one or two, neither hepatitis C nor the treatment had been very bad. The seriousness of the symptoms waxed and waned over the months of treatment, with a couple of people describing the last few months as being the worst, where the problems seemed to compound.

In the following paragraphs, we examine some of these different experiences, first outlining the historical sequence of available treatments, each one of which has been more effective than its predecessors.

Initially, from 1994, the funded treatment for hep C was six months of interferon. At this stage it was normally offered to people whose liver test results indicated liver damage. This 'monotherapy' was later extended to 12 months, if indicated, and eventually it was paid for by the State. The first combination treatment was interferon plus ribavirin, which was offered from the late 1990s onwards. In February 2004, funding of the more effective pegylated interferon with ribavirin was made available, again for up to a year. As time went on, research indicated that treating early, before the effects were felt or observed in tests, was more effective. A lower viral load indicated a better outcome from treatment. Several different strains of hep C have been isolated, and some of these are more responsive to the treatment than others. In addition to the pharmaceutical products, some herbal remedies have been helpful in reducing the symptoms of both hep C and treatment for it.

The most commonly mentioned symptom of hepatitis C (and treatment) is tiredness, often extreme tiredness: "dead tired". Three of the older men with the virus experienced two o'clock in the afternoon as the time when they needed to rest for a while or they would be knocked out for the rest of the day. This was very difficult to manage for those at work. Others were "knocked out" by about 7pm.

Often people experienced a range of symptoms, but only after reading more about hep C or attending a conference did they realise that they stemmed from hepatitis C. Eric and Deon discussed the difficulties of describing the effects of hep C.

Eric: I think looking back I can see, like I think about, around about the time I got it … and how unwell I was. And I'd been in hospital and come out of hospital and you know, I think about that time, and it was like I'd turned a switch off. And it was like that for ages, you know. I was really crook for a few months, and then, you know, sort of came right. And then there's been a couple of other times that it's been particularly noticeably difficult times, and other times when it's been quite good. I mean, I'm not — like I said, I know there was a lot of people who were yellow, you know, who brought their wives along because they didn't have enough energy to drive their car, … so from that perspective I think I've done pretty well, but yeah. Certainly, when we talk about "Have you felt fatigue and tired?", well, you know, it all sounds a bit, all sounds lightweight.

Deon: Yeah, I know what you're saying.

Eric: It sounds a bit lightweight, but yeah, basically, that's the sort of thing that I, I think has affected me.

Deon: I think perhaps it sounds more lightweight to people outside of it who don't really understand what tiredness could mean. It's like "well, you're tired, so what, we are all tired", you know …

Eric: [laughter] Dead tired, mate, it's the dead tired that gets you.

Access to treatment was an issue for some, even though treatment was theoretically available. One man had been pursuing treatment since 1998 and had contracted hep C well before this:

> I finally got to see a hepatologist who was prepared to do something about it in 2003. At that stage I was told that because I had hep B and hep C I was not entitled to treatment. My response was, "Well, surely it's good medicine to at least monitor, you know, what's happening." And I was told, "Well, no, we don't do that." We don't have a recall and follow-up system within our clinic, which I expressed amazement at, but, yes, they would arrange another appointment for follow-up, bearing in mind this was April 2003. Got home, in fact I wouldn't leave the clinic until I had the assurance that I would actually get a follow-up. Got home, and my follow-up appointment arrived and it was for November 2005. Um, their idea of follow-up is not my idea of follow-up. …

> I was not happy with that as a so-called follow up, and via my haematologist I asked for a further appointment, because as I far as I was aware Pharmac had changed the access provisions and frankly I was wanting to get on to treatment. So, I saw the hepatologist in February 2005. At that point he said, no, that hep B was no longer an impediment to access, ah, "We will start treatment immediately."… Bearing in mind that was a February appointment, here we are now into mid-June and I still haven't started treatment, but I certainly created a fury with the local clinician who's phoned to indicate he's pissed off — his phrase — in no uncertain terms. I've tried to ring him back to let him know that so am I — and that I don't intend to allow the systemic failures of the health system to impact on my liver. [His treatment started before our fieldwork was completed.]

This man's experience of difficulties of access was not typical in this study phase, although such experiences were very common in earlier years when criteria for access seemed to keep changing. Changing criteria for treatment have very direct effects on individuals, as this example shows. In addition, so many people were coming forward for treatment that the gastroenterology services in some DHBs seemed to be overwhelmed, including the DHB that this person attended, and there were some very long delays in getting the initial specialist appointment. However, once that barrier had been passed, most people started treatment quite promptly.

Only one person whom we interviewed in this update study appeared to be confused about his hepatitis C status. This was a major contrast with the earlier phases. This man, in his 30s, told us about having hepatitis B and the steps he had taken with his diet to reduce fat intake, how he had stopped drinking and given up smoking to improve his health and to give his liver a chance. Then he said in response to Julie's question about whether these changes were helping him feel better:

> Yes, really good. *Really* good! I missed out on hep C, I was really lucky, and so far have antibodies against it. I am a bit sceptical, I don't think anyone knows that much about it, they are learning from my situation, from others. I've had hep B and I still continue to carry that, and I reckon that that is just sitting there and, I wonder, is that doing damage? I think that if I am careful of my liver, as I am now, it should be okay. See how it goes.

Unlike some other infections where antibodies in the blood but no other symptoms show that a person has been exposed to a disease but does not have it, antibodies to hepatitis C more often show that a person has been infected with the virus and remains infected, although it is true that some people have got rid of the virus without treatment. The person quoted above was being monitored for hepatitis C, despite his hope that he had missed out on it.

One person, who got hepatitis C in 1990, had only just completed 11 months of treatment when she was interviewed. She was experienced in the use of herbal remedies and had used them to help with her treatment, as a complement. She had been offered a course of interferon in the early 1990s, but had turned it down because she was asymptomatic (apart from feeling tired and lethargic), and she thought that she could manage it with a good diet and herbal remedies. However, eventually she recognised that it was very hard to treat a chronic condition with the remedies that she was trying, and, at the haemophilia nurse's suggestion, she went on a course of pegylated interferon and ribavirin. Unlike many of the others, she did not experience many severe symptoms while on treatment. Initially she had flu-like symptoms, for a short period she had diarrhoea and stomach ache, she lost her appetite for a short period, and she experienced hair loss and insomnia. She had a skin problem — little pustules — which she did not realise was related to the treatment until she went to the Hepatitis C Conference, where one person spoke about it. She was able to find herbal remedies to reduce the symptoms, such as oat straw tea for the hair loss, and on her nurse's advice she took her treatment earlier in the day to reduce insomnia and camomile tea to get back to sleep. So for her, neither the hepatitis nor the treatment was very bad, and, although it was too soon to tell, the indications were looking positive for her being rid of the virus.

One of the difficulties with treating hepatitis C was timing the treatment so that the disruption to work and family life was minimised. For example, the woman who had just completed treatment had waited until her children were relatively independent, in case she reacted badly. Two recent graduates waited until their degree courses had finished. However, this meant that they were new in their jobs when they started treatment and both of them were quite nervous about this. One of them had had ACC agree that it would pay him an ACC allowance if he had to take time off work. However, with an understanding boss and by spreading out his sick leave, he did not have to have any substantial time off.

The other of these two young men talked about his drive to keep both of his part-time jobs going:

> Andy: But I didn't want to give it up because I didn't want a gap in my CV. That's how I felt about it anyway.
>
> Julie: Why was that?
>
> Andy: Because I think that in the sort of circles where I work, haemophilia is not a problem and I can openly talk about it and people even find it quite interesting, and have a bit of a joke about it. But

> you can never know for sure where you are going in the future and I want to be sure that it not be seen as a problem for me being able to work. And I think that sometimes, even though discrimination might not be openly talked about, if there are two candidates for a job who are equally good for the job, if there is a thought that this person's health might affect him coming in to work, there might be a question there. I think if you are better than the other candidate then you'll probably get the job, but if there's a few around that are equal, in some industries you won't get the job, and that was important to me.

Another who had treatment while attending university thought that he had probably wasted a semester's fees, as he had missed so much due to extreme tiredness.

People who were in casual work or self-employed had considerable difficulties. A sister described how her brother had been so ill he could not keep up his casual part-time work and was supported by family members, and we heard of other young people who had to give up their independence and return home to mum and dad, partly because of not being able to pay the rent, and partly to have someone to look after them. Eric, a man in his 40s was contemplating having treatment for his hep C during the time of his interview. He was busy with his business and was inclined to put it off.

> I'm kind of a bit busy to take a year off at the moment and, you know … talking more to a mutual friend having treatment, you know, about treatment ... Hell, I'm dog tired now just because of what I've got to do, let alone any treatment. So, it's not really a good time to do it. So, I'm just waiting for perhaps a better time.

The problem is, for this busy man with a family to support, there is unlikely to be a better time.

One of the older men had had six months' treatment with interferon in about 1994, one of the first to have treatment. Initially, it appeared to have been effective and his liver enzymes returned to near normal, but a few months after treatment he tested positive again. However, he did notice considerable improvement in his health. About this time he read an article in *Bloodline* about herbal remedies that can help with symptoms and he tried St Mary's Thistle. This was a great success and he felt that it almost normalised his health, although he still experienced exhaustion. In about 2000, at the prompting of his partner, he again went to see the gastroenterologist with a view to another course of interferon. However, in the tests preceding this he was found to have a life-threatening condition related to his liver condition, and instead was offered a liver transplant, which he accepted. He was amazed to be offered it, given that he had hepatitis C. The transplant was successful and cured his haemophilia, but it did not cure his hepatitis, although with a new liver he was not feeling the effects of the virus. As he said, "it was like turning the clock back 15 years". At the time of the interview he had been advised to wait for a couple of years for a different form of hepatitis C treatment which was more suited to transplant patients, to try to eliminate the virus.

This very dramatic event was not anticipated by him or the whole haemophilia community, who celebrated "the only ex-person-with-haemophilia" in New Zealand. It was a significant event and was mentioned by several people we interviewed. One friend of his had this to say:

> And of course since then, there's also been the liver transplant, which was actually quite important. For us, too, it was like, "Oh my God, there's something new. When all else fails, you have a liver transplant — and, oh my God, it gets rid of your haemophilia as well!" I mean, that was pretty amazing. Then to see the life that [my friend] was able to live after that. Just coming to life again, like he was given a new life, and that was actually very affirming, that was, although one would hope you'd never get to that point, that there was something else at that end of the scale as well.

Another mother, Karen, who did not know the man involved but who had heard about the transplant, had a son who had suffered brain damage at birth due to his haemophilia and who subsequently had had some serious spontaneous bleeds, but who did not have any liver infections. She wanted to know why he could not have a liver transplant to cure his haemophilia. So Julie suggested she speak to her haemophilia centre and outreach worker to hear about the pros and cons as well as eligibility criteria for transplants. Karen saw a liver transplant as a possible treatment for haemophilia itself.

Interferon treatment undoubtedly is difficult for the patient, and it can also be hard for those around him or her. A mother, whose son had had a successful 12-month course of interferon monotherapy when he was a teenager, talked about the difficulties the treatment had caused:

> The first couple of weeks were really grotty. It was really difficult for me to know what of his behaviour was interferon-related or what of his behaviour was … snotty teenage behaviour that you would not accept. So I didn't know whether to jump on it and say, "Hey, this is not okay in our house, you won't speak to me like that, you won't behave like that", or whether just to tolerate it … for it's only for the year and, you know what I mean? It was a really difficult year. And I wouldn't actually recommend to anyone else that they do it at that age. But then [voice brightens] because of his youth, and [that he] hadn't had the virus for long, actually was in his favour. So yeah, it was a very hard year, but I'm pleased it paid off.

Another mother, whose children were born late enough (i.e., after 1992) to be protected from hepatitis C, observed the effects of treatment on her friend's son, who was a few years older than her own:

> I think to myself, we were lucky we had a baby in the 'nineties and not in the late 'eighties, when the opportunity was there. You know we were married for six years before we had our son, we could have quite easily have had him, you know, in the mid-eighties and who knows what we would be facing today. I think I'm very lucky, because [my friend's] son did have hepatitis C. And you know I saw him at his worst, I saw him so cut up, so bloody sick from taking his interferon that you actually wondered what was in it for him at the end of it all. It was needles after needles, interferon after interferon, he was sick, sick, sick. He'd gone from a very healthy boy to as sick as a dog the next day. And I guess, years later we can reflect on that and say "He was bloody lucky", because he had a year's supply of interferon, that his parents paid the other six months for, because the Government would only pay for six months. So he was lucky because they were told that if he had a 12 months' supply he would have a better success, survival rate, so that's what they did. And look at him now, one fine healthy young adult. And he knows it. I love him to bits and I just saw him two weeks ago and just think, "By God, you are so fine", and I've often said to his mum that if [my son] grows up to be half as good as her's, I'd be absolutely delighted.

Interferon treatment is stressful for partners, and, as the outreach workers confirmed, they can need a lot of support to help them through the months of coping with and trying to support a tired, grumpy, depressed and irritable partner, especially if the patient also perceives the partner as a source of irritation. Some sisters also described the pain and fear that their brother's reactions to treatment had caused them: losing many kilos of weight, being too sick to get out of bed for your own birthday.

Several people mentioned that interferon treatment was expensive. Some thought that under-budgeting for the service as well as a lack of gastroenterologists had contributed to the treatment delays that occurred in some DHB regions. Hep C is at epidemic levels in New Zealand, with probably about 35,000 people infected (cited in Harris 2005:4). Those infected through blood transfusion and blood products are just a tiny fraction of this number, but the potential is there for hepatitis C treatment to overwhelm the health service. During the study period, patients were not paying the costs but they were aware of them, because each time they had their prescription filled, their small contribution (usually a few dollars) and the total cost were shown on the receipt. One person thought the dual course medication was $1600 for three months. Six months of monotherapy was reckoned at $3000, back in the mid-1990s, when some people had to pay for an extra six months themselves. The cost for pegylated interferon alone is approximately $2000 every three months. In addition, there are the associated costs of all the tests. Many participants had some knowledge and appreciation of the HFNZ's efforts over many years to get publicly funded and effective treatment for hepatitis C.

Privacy and disclosure for people with hepatitis C was an issue for some. Many people with haemophilia are quite open about their condition — although, as with everything else, there are always exceptions — but people were generally rather more cautious about disclosure of hepatitis C. This is in contrast to the well publicised travels of 'Jack' who kayaked and biked from one end of New Zealand to the other to draw attention to haemophilia and hepatitis C.

Marty was one who exercised close control over who knew about his hepatitis, for which he was receiving treatment, and he was also careful about whom he told about his haemophilia. He had told only his employer and close friends about haemophilia and he had spoken to no one about hepatitis. Deon asked him why this was.

> I suppose they might not understand it like you should. And like I remember once when I was a kid my mum told my neighbour, and I was best friends with their kid and, well, she said she just told them about me being a haemophiliac — they pretty much banned us from hanging out. I'm not … I guess she just didn't understand it. So that's why I don't talk about hepatitis C with other people, because they might react in the same way.

It is likely that the friend's mother's reaction was due to an association of haemophilia with HIV and AIDS. We came across several similar stories in our initial study in the mid-1990s. In contrast, everyone who had attended the Hepatitis C Conference spoke very positively about being able to discuss their illness and its treatment with other people who really understood their situation. This was especially the case for one or two people we interviewed who had not previously had much contact with the haemophilia community and therefore had had little opportunity to meet others with haemophilia and hepatitis C.

One of the men, with a high public profile because of his role in the Foundation campaigning for adequate treatment and a Government apology for hepatitis C, said that he was frequently asked by news media about his own hepatitis C status, which he refused to disclose. He believed that it was private and also that it was not relevant to his role in the Foundation campaign; however, his close friends in the haemophilia community knew what his status was. In contrast, another man who had

led parliamentary protests, and had been very active on the hepatitis C issue, did not mind being identified. He said, "Anyone who's a decent mate doesn't have a problem with it."

Generally, people who had undergone interferon treatment did not talk in great detail about the actual treatment process — injections and blood tests being just part of life for people with haemophilia. Most people treated themselves for hepatitis C at home with a subcutaneous injection of interferon once weekly and doses of ribavirin tablets twice daily. However, one young man gave an interesting glimpse of the outpatients clinic where he went for his check-ups.

> But also just going in to [the clinic], it was very good: the nurse that I had and the way that she handled it. But I also think that perhaps they quite like seeing the haemophilia patients. Like I was in the waiting room and there was somebody that was out of jail for the day strapped to someone else, and then there was somebody swearing to themselves and somebody else shaking, and I can only deduce from that that one of them was having withdrawal symptoms from drugs and one of them was having very bad side effects and one of them might have got it in prison or just from lifestyle. So us in the middle, we are still in a mess but we are more a voice of reason than a lot of these other people. And I think they are a bit more thankful in some ways for getting this treatment, though in saying that, I have heard stories that some people with haemophilia have been quite abusive to clinical workers because of the feelings of anger about hep C and the feelings of … They need to point the finger somewhere, and they haven't had the apologies from Government and they need someone to be angry with, so they are angry with the nurses.

His suggestion that the nurses may quite like seeing people with haemophilia because of the difficulties they have with some of their other hepatitis C patients receives some support from research with people with hepatitis C, such as Harris (2005), where she calls for a renewal of respect in the caring relationship and for a more contextual understanding of the lives of people with hepatitis C.

The Spread of the Shadow: Implications for Families Not Directly Affected

We asked everybody about hepatitis C who was willing to talk to us about it, because it has affected the whole haemophilia community: those infected and those not. In this section we briefly consider the personal implications of hepatitis C for those who do not have affected people in their family, and in the next section we consider the work of the Foundation in relation to hepatitis C.

"Hep C is a bit scary, in all honesty," replied a mother whose son was less than a year old to Julie's question about her views on the Foundation's efforts regarding hep C. After the mother had explained what product her son was on — it was plasma-derived due to certain difficulties of treatment — the following exchange occurred:

Julie: When you say hep C is a bit scary, is that still something that you are a bit frightened of?

Nita: No, not so much. Purely because of what [treatment products] my son would have, I suppose.

Nita knew that there had been no hepatitis C in the plasma-derived products since the end of 1992 and that all products were screened and tested, yet:

Nita: …it's just a fear, you know what I mean?

Julie: Yes, I can understand that.

Nita: It's like AIDS and that. I'm 99.99% sure that we'll never receive anything that is dodgy, but there is always that 0.01% that goes "Oh, I hope that's okay."

Julie: Oh, I understand that completely, a niggling thing that is always there?

Nita: Yes, little thing at the back of my mind that just goes "Oh God, it is blood products", you know, and I am putting it into my kid, trying to make him better and there is always going to be that little, little percent chance that it may do damage.

Another family with young children were careful to keep a systematic file (actually two large lever-arch files, even though their son was still young) of every piece of paper relating to his condition. When I commented on their systematic approach, they explained that that was what they had learned from the community's experience with hep C — always keep the paperwork. The father, who had been on the HFNZ Council, remarked at a different point in the interview:

> … we had a vCJD donor type thing pop-up in relation to Kogenate. The awareness of your medication possibly being compromised by viruses is still there. We don't sit and panic or stress about it, do we, but you sort of tend to look on teletext when you see something about bird flu or whatever.

His wife confirmed that they "feel pretty safe" about the recombinant product their son used, but "there is always a wee 'but' there … I am putting something into my child", and the husband repeated "we don't stress about it" and quoted a 0.0002% risk. However, they are vigilant if they have to go to Accident &Emergency. They always check that their son is being

given his normal product, as they have heard that only by parents checking the label have they prevented their children being given plasma-derived factor instead of recombinant product. They noted, nonetheless, that "those [plasma-derived] factors too have a safety factor as high as the recombinant".

Another mother of two young boys is reminded of the various viral threats to their health every time they go for a review. As well as having blood tests to establish the correct volume of clotting factor to be administered, their blood is tested for HIV, hepatitis B, and hepatitis C, standard practice for haemophilia reviews.

"They need all the support they can get"

No one in Hannah's family had hepatitis C, but through her involvement with her local committee and the Foundation's events she knew people who had. She was in total support of the Foundation's actions to try to get compensation, to assure best practice treatment continued into the future, and to obtain recognition from the Government. She owned that she probably was not as passionate about it as people who were directly affected, but she was prepared to help in any way she could. She believed the haemophilia community needed to band together on this issue as the people affected and their survivors "need all the support they can get".

Virtually everyone was in total agreement, and some expressed their agreement much more vehemently than Hannah. Mothers, especially, pointed out that the safety of the products they were using with considerable confidence for their children depended on the careful work the Foundation (and the Society before it), in conjunction with its Medical Advisory Council, had done, working with (and sometimes in conflict with) various government agencies, the Ministry of Health, and the relevant drug companies.

In fact, in only one interview did anyone express anything less than solidarity, although we discussed the topic in every interview. The wife in the dialogue below uses the same metaphor of casting a shadow as was used by Justin in the opening story in this hepatitis C section. But here it is not hepatitis C that casts the shadow, but the Foundation's efforts to bring the Government to account and the perception that, in this aspect, they are oriented to the past.

Julie: This is digressing a little, but how do you find living, for part of the time, like when you are at camps, in a community where there are a lot of people that have got that history? Not just the disablement of ankles and knees, but those past threats of hep C and so on.

Wife: Not our past.

Husband: We find it hard to relate to those families, especially around the hep C and the bad blood and all that sort of thing. They have quite a different outlook, even towards the health care and their health providers. It seems to be different from ours and our energies aren't sort of in that direction. The Haemophilia Foundation has still quite a strong [orientation towards] getting the Government accountable for hep C, and that's not an issue for us, so we are not driven in that direction so much.

Wife: I think there are other families within the Foundation, as well as us, that find it quite a burden, that to some degree, very insensitively, just want to say "Get on with life", eh?

Husband: Yes.

Wife: And that is very insensitive. But it casts a shadow over the whole.

This couple's focus is on trying to secure safe and satisfactory treatment for their children, now and into the future. Although they are very much part of the Foundation, their friends are mainly 'spontaneous cases' like themselves. Thus they have no family history with haemophilia. They are of course sympathetic towards people with hepatitis C, but they too have had serious problems to overcome, and the emphasis on hepatitis C is an additional burden for them. As noted earlier, hepatitis C is a scary concept for many people who have not been directly affected by it.

But the benefits of the Foundation's work on hep C are widely experienced, even if they are not specifically recognised as such. One young man with hepatitis C who was affected by his treatment at the time of the interview reported that some money had turned up from somewhere and he did not know whether it was some ACC money or some kind of settlement. He said that he wasn't particularly interested in getting money. What he was really concerned about was having his treatment paid for, and, of course, its success. He was also keen that everyone else should have access to the treatment. All these things — ACC recognition, paid treatment, and treatment for everyone — had been issues that the Foundation had campaigned on, or had assisted their members to access.

Another slightly older man, who did not (yet) have any symptoms of hepatitis C and was not on treatment, was relieved to find that now more effective treatment was publicly funded. He, too, was not interested in getting money, or in his caregivers being reprimanded, although he had a claim with ACC.

Participant: And I have been involved with the ACC Medical Misadventure Unit, recently, and they … It's been interesting, to me, because really [sighs] I don't like the term 'medical misadventure', because it is more like a medical adventure [laughs] rather than a medical misadventure. I mean 'cause nobody really knew. I knew I could contract diseases from this treatment. Medical misadventure, the whole

unit being part of it, suggests they are going to be hunting somebody down.

Julie: So you don't like that slightly punitive—?

Participant: No, that's right, that punitive feeling. And I made that very clear that I don't want any … I don't, I have, I'm very happy. I don't want my doctors to be hunted down like that. I just don't feel … How could they be blamed in any way? So I'm part of that and I have made a claim, but really all I want, I don't want any money 'cause I'm not sick, but I do, I wonder about, ah, the social future in the sense that maybe they will expect me to pay for care in the future, and okay if I do receive money, you hold on to that purely for your care for hep C, that's that, nothing else. And if you don't use [it] and nothing ever did [go wrong], if I was to be paid out — but see, "paying out", I don't like either, because see, "You're paid out, see ya!" [Laughs.] "Off you go". To me all of a sudden we've got this, a half-American-style medical system and half-social service type thing, and I'm very uneasy about the whole thing. I don't like the idea of money being bandied round. Um, yeah. I'm wary.

Julie: You feel: be on the safe side?

Participant: Yes, be on the safe side and be part of the experience and see what you can learn from it as well. And see how this system works. Because this is very much 'system' part of it. All I'd want, if I was bad, is a hospital bed to be in, a place to be if I was very sick from it.

One young woman who did have relatives badly affected by hepatitis C was also concerned about monetary payments. She agreed that compensation was required, but felt that those about to receive it would need education and guidance before getting a largish sum.

While compensation that would allow those affected to live as full lives as possible was an important plank in the treatment and welfare package being sought by the Foundation, it was clear from participants' comments that the security of having access to free treatment that was as effective as possible was probably of greater importance to those who had tested positive for hepatitis C. Most people thought that an apology for the injury they suffered through the medical system — especially because the Government did not take action on hepatitis C in the blood supply as early as they could — would help people to move ahead and some were passionate about this. Others were not so sure what good it would do. However, those who discussed this aspect did think that the Government should acknowledge its lack of action and the devastating consequences of this for the haemophilia community.

The Foundation leadership was aware of the potential for some division between those members who had lived through the HIV and hepatitis C epidemics and those who had joined later. However, they did not see it as a major source of division, just something that had to be recognised and managed. We talked with some of the past and present office holders about the situation.

> Well, that worries me, because I think new families look upon [hepatitis C] as a thing of the past, "Oh it doesn't affect me, it doesn't affect my family", but it does. It's just that the next issue that affects their family will have a different name other than hepatitis C. Now, that issue might be, you know, an international withdrawal of product, it might be some nonsense such as has already been suggested that the US should not export any product until the people of the United States have had, you know, adequate care. Well, on a product like this, I don't see that it belongs to the United States, it belongs to the world. It just happens to be being produced in the United States. So, those are the sorts of issues that I see still coming out of issues surrounding haemophilia. For the moment, it happens to be hepatitis C. In the 'eighties it happened to be HIV. Prior to the 'eighties it happened to be no treatment or insufficient treatment. I believe these issues are going to continue until such time as we can frankly avoid further haemophilia births in families knowing to be carrying the disorder.

Hannah had been at a talk given by the President of the Foundation, and related how he had squarely addressed the issue that new families did not get hepatitis C, yet the Foundation would continue its efforts to secure a settlement for those who did get it and would kept working towards it until it was achieved. She reported that he said, "We are going to support these people and we are going to see it through", and she added "and that was cool that he brought that up". In fact so many people mentioned that hepatitis C was just one in a long line of issues that stretch into the future, rather than something firmly located in the past, that there appeared to be little chance of this particular group of people forgetting the lessons for the future to be learned from their history. To some extent, the Foundation's efforts to secure publicly funded, best available treatment for both haemophilia and hepatitis C, rather than to concentrate on compensation only, can be seen as just such an effort: using the past to prepare for the future.

We also talked to leaders in the Foundation about recent events relating to hepatitis C. The past two years had been significant. From early in 2004, dual treatment was fully funded and available for people with haemophilia and hepatitis C, and many people began

treatment as a consequence in 2004 and 2005, including four in our study, and another was about to undertake the initial test for viral load.

The conference and workshop, to which two of the powerhouses behind the Irish settlement were invited, was rated a huge success and very supportive for those who attended. This event raised the public profile yet again, and the meetings surrounding it with the Irish delegates were very instructive for the New Zealand members working on the issue. Some intense negotiations with Government occurred, and a few weeks before the November 2005 parliamentary elections the Foundation members received a letter from the Prime Minister, the Rt Hon Helen Clark, which, we were informed, was at least a step towards meeting some of the Foundation's requirements. However, a Labour-led coalition would need to be returned for this to happen. After a nail-biting period of coalition negotiations, Helen Clark emerged again as Prime Minister, with a new Minister of Health. Those who were aware of the letter expected a speedy conclusion, after some quite difficult organisational matters were sorted out. Several participants in this study indicated their extreme disappointment with how this matter was prolonged, including this person, interviewed in February 2006.

> Okay, look, I mean the negotiations and the agreement at the end of September last year were very good and very positive and very practical for the Government. And certainly, you know, the letter came out really with a pretty played-down version of the excitement we felt about the whole thing I think. …Certainly around the table it was a very, very productive and exciting thing, but the wheels have fallen off it completely, as far as I am concerned. With a new Government, the instability over the election, and then we've got the bird flu pandemic, the possibility and the planning, and it's excuse after excuse. Personally, I think it's about time to go and visit the Ministry again. It's time for the lid on the privacy that we guaranteed the Crown to be taken off … The thing is, the public aren't aware of the offer, so the Government aren't accountable to do anything.

> You know, we haven't told anyone, so no one knows, and so we can't say "Hey you guys promised" and it's now been six months, been six month nearly, it's been six months, and nothing's happened! So … it's time for some action to take place, that's gonna come down through the channels [i.e., from the liaison person on the Hep C Working Group].

At the same time, some people with haemophilia — along with others affected by hepatitis C in the blood supply and people who had infected blood transfusions — were privately involved in a class action that appeared to be making little headway.

By the end of the fieldwork period, Foundation members who had been at the forefront of the hepatitis C campaign for Government accountability, welfare and treatment provision, which had appeared to be bearing results towards the end of 2005, were becoming disillusioned and believed that they would once again have to return to the hard work needed for a settlement. Six months later, as we completed the draft report, there had been no public announcement of settlement, and we understood that we were not at liberty to reveal the details. The looked-for announcement was made in December 2006.

Chapter Six: Conclusions

Over a decade has passed since the original study was completed, and over six years since a smaller update. New technologies for the treatment and diagnosis of haemophilia have had an impact on the experience of discovering that one has haemophilia, living with haemophilia, passing it on, and planning for the future. Despite the changes, there are some constant elements in the experience.

Discovering Haemophilia

The experience of discovering that there is haemophilia in the family has changed little. Being a rare condition, with 30% of cases occurring without warning, the diagnosis comes as a shock to both parents and health professionals alike. The occasional uninformed question of "How long have you had haemophilia?" from a few health professionals to people with haemophilia serves as a reminder of the rarity of the disorder. Not all health professionals are conversant with haemophilia; therefore, people living with it need to be well informed and 'bring out the big guns' (i.e, the haematologists), if necessary, when interacting with health services. In instances where there is no family history of haemophilia, initial medical investigations often begin with suspicions of child abuse and end with a diagnosis. The experience is, not surprisingly, both stressful and emotionally complex. Despite this interactions with health services were described by participants as positive, overall, although there was still a degree of variability between urban and rural services.

Changing Treatments

The treatment for many people with haemophilia has moved from on-demand to prophylaxis. This, along with the provision of recombinant clotting factors, has reduced the level of stress for parents Accordingly, for many participants, perceptions of the seriousness of haemophilia have changed too. Improved treatment for inhibitors has been a major step forward. The issue of restrictions in sports that people with haemophilia can play continues to be an important topic for discussion among the community. This is particularly true in rural areas where the question of playing a sport such as rugby invariably raises its head, as the sport is a central part of community life. We perceived a greater degree of freedom to live life as they wished among the participants in this study compared to our earlier studies when most people received blood products on demand and were very concerned about safety.

Over the years since our research began there has been a shift from treating a serious bleeding disorder with clotting factor products derived from blood donations to controlling a medical condition through recombinant products produced by pharmaceutical companies. Recombinant products are perceived by many as being safer, although as several older members of the community remarked: "Blood products have never been safer."

Considering the Costs

People were very aware of their treatment product brand, especially those using recombinant. The change for a community which once relied on blood donations and a non-profit blood service for its treatment to now relying on major pharmaceutical companies cannot be underestimated. They have made a transition from being recipients of donations to consumers of products of competing multinational companies, each of which wants to secure and increase their place in the market. Participants were aware of some of the complex issues surrounding their relationship to these companies. On the one hand, they relied on the firms' products and processes; on the other hand, they were wary of being used in ways that they cannot approve, and of the unintended consequences that might flow from their quests to find the best products or the most acceptable procedures. Many participants were aware that they are 'big business' for pharmaceutical companies. This is also an issue for the HFNZ, of which it is only too well aware.

Compared with our earlier studies, people were more aware of the direct financial costs of having haemophilia. Some expressed a degree of guilt about the costs of their treatment to the community. At the same time, for people who already had haemophilia and could not live normal lives without their treatment, repeated reminders about costs were experienced as a burden. These concerns were held against the backdrop of a complicated mosaic of DHB funding territories which had to cope with the random but clustered nature of this expensive and rare condition. Before completion of this update study, the formation of the National Haemophilia Management Group meant that funds for haemophilia would now be centralised. This is a positive step in our view.

The indirect financial costs of haemophilia — such as transport to and from the hospital or time taken off work — created some concerns. A number of people with haemophilia had needed to adapt their careers and even their place of residence to take account of their condition.

A Genetic Disorder

While the idea is still pervasive that 'women are carriers and men have haemophilia', there is an increasing realisation that men pass it on too and that women can also have bleeding problems. There is now more recognition of the complications arising from being a carrier of haemophilia; for instance, the reality of being a symptomatic carrier, which is equivalent to having mild haemophilia. A successful inaugural Young Women's Workshop Weekend for carriers and women affected with bleeding disorders was held during our study period. In contrast to our initial study when many women reported long delays in getting results from carrier testing, in this study that was a rare problem. Most people were quite happy with the way that they had heard the results. The issue of deciding when to tell girls that they are carriers remains fraught. The majority of those interviewed felt that it was both in the parents' and child's best interests for a possible carrier child to be tested early. This would allow the implications of being a carrier to grow with the child rather than to come as a shock.

Within the haemophilia community there is the same wide range of views about reproductive choices as there is outside of it. There is some contention around whether parents who have no intention of terminating a pregnancy are entitled to have a prenatal test. Couples where the wife is a carrier wanted to be able to make the decision themselves, after being informed of risks and costs and benefits from a medical perspective. They felt that even a health professional who knew them well was not in a position to be able to assess the costs, risks and benefits of not knowing and knowing the haemophilia status of their future child for them or their wider family. As several people remarked, they did not even realise themselves what strain a pregnancy was causing them until they felt that immense relief at learning that the foetus was a female or did not have haemophilia.

Issues surrounding carrying haemophilia on were handled very carefully in the community, with different views on the seriousness of haemophilia, prenatal testing and abortion being respectfully acknowledged. The principle that 'what is right for each family is what is right' was often quoted, and allowed conflicting views to exist side-by-side. However, the potential ethical problem of this principle was noted: at what point should a family's right to decide be limited? Many participants mentioned that most topics are very open for discussion within the haemophilia community (such as carrier status or family history), but perhaps prenatal testing was a somewhat more restricted topic, one that would be discussed in more private settings.

Gene therapy was described as being on an ever-distant horizon, possibly available in 10–20 years, much the same timeframe as in our initial study. Participants were wary of gene therapy, but not dismissive. They were more interested in other new technologies, such as preimplantation genetic diagnosis and its role in aiding reproductive decisions, and longer-acting recombinant products which would reduce the number of treatments required.

The topic of PGD has provoked considerable thought, and its advent has generally been welcomed. It was usually a topic thought about more by the women than the men, although we did ask both men and women about it. There were differing views about its normality or its unnatural characteristics. Many participants raised the issue of whether termination of an affected foetus or prevention by PGD of the implanting of an embryo with haemophilia conveyed a message of diminished worth to other people with haemophilia.

The men with haemophilia held different opinions about this. Most believed it conveyed no such message — it was just an attempt to prevent pain and suffering. Others believed that haemophilia was not so serious that such an action should be contemplated, while a minority were adamant that they would not have children at all, so that the haemophilia will stop with them. A couple of the men also noted that if these tests were available when their mothers were pregnant, then they might not have been born. However, they did not use this as an argument to limit the availability of prenatal or preimplantation genetic diagnosis.

The HFNZ and Hepatitis C

According to participants, the Haemophilia Foundation of New Zealand continues to be an important source of information and support, with particular reference to the services provided by the outreach workers. New families also appreciated the support provided by local branches. Some participants who had previously been involved with the Foundation noted that it appeared to have changed into a more formal and political group. This was particularly evident in activities around reaching a settlement with the Government over the issue of blood products being contaminated with hepatitis C in the early 1990s. The Foundation now has a hep C generation and a post-hep C generation. Nonetheless, hepatitis C affects the whole haemophilia community. It highlights the vulnerability of people whose survival and ability to live a normal life depends on the safety of the products that they use. Therefore, many people see the past experience of hepatitis C as relevant to the present and future. More effective treatment for hepatitis C was available during this update study than at any time in the past, and many of those who still had the virus were undergoing treatment, or planning to be treated, with pegylated interferon and ribavirin therapy. Anecdotally, people with haemophilia seemed to be having a promising response to treatment. As we were completing this report, the long-awaited and most welcome Government treatment and welfare package was announced in the media and through the HFNZ.

Afterword

Julie did the bulk of the analysis and drafting for this study while on University of Auckland research and study leave in Rarotonga, in the Cook Islands. While she was there, Radio New Zealand Pacific News reported that a high-school girl from an outer island who was staying in Rarotonga died as a result of 'haemophilia', despite being flown to Auckland for treatment. Although we are not certain of the accuracy of the specific reported diagnosis, the stark realities of lack of support for severe bleeding disorders and timely, effective, treatment could not be more apparent. As one of our participants said, with regard to haemophilia treatment in New Zealand: "It's the luck of being born in the right country." More, longer-term, outreach and partnership initiatives from New Zealand to close Pacific neighbours are highly desirable.

Recommendations

1. A **national service for haemophilia** for reasons of equity and efficiency. The need is even more pressing with 21 District Health Boards than it was with four Regional Health Authorities.
Status: The National Haemophilia Management Group (the NHMG) has now eventuated. The responsibility of ensuring equity of access to quality treatment is now placed with the NHMG.

2. Just settlement of the haemophilia claim in relation to **hepatitis C**.
Status: A treatment and welfare package was successfully negotiated by the HFNZ in 2006. Issues that are preventing payment for all affected members are being worked through by the Foundation.

3. Further exploration of how to resource primary health care providers to **reduce diagnostic delay** in babies and young children with haemophilia.
Status: This could be an option for the HFNZ to consider. More robust recommendations on continuing medical education could also be developed by the NHMG.

4. A specific study by and of **Māori with haemophilia** to describe the social contexts of Māori with haemophilia, to estimate the numbers of people with haemophilia who identify as Māori (not necessarily only as Māori), to establish their needs and how these may be met.
Status: Planning for this study by Māori members of the HFNZ, supported by the HFNZ is underway.

5. Haemophilia Foundation of New Zealand **outreach**.

a. The HFNZ continue to support and seek funding for haemophilia outreach workers to enable their work to continue at enhanced levels

b. Use of Internet discussion groups and/or web pages for special interest groups, including: Māori, Women with bleeding problems and other special interest groups as they become defined.

c. Consideration of ways to resource families who do not have internet access at home, or access to free public internet.

d. Continuing dissemination of information on prenatal testing to individuals and couples.

e. Initiating small telephone mutual support groups for families who are otherwise isolated.

f. Continuing debate on issues surrounding carrier testing.

6. Monitoring developments in "**the business of haemophilia**", particularly the roles of the large pharmaceutical companies in relation to the HFNZ, the National Haemophilia Management Group, and individuals.

7. Outreach to **Pacific neighbours**.
The HFNZ could make their quarterly newsletter available to affected people in Pacific nations and might consider arranging sponsorship for families to attend an annual camp in New Zealand.

Acknowledgements

We acknowledge the generosity of all of the participants in this study. We hope that our respect for you and the way you respond to the challenges of haemophilia is evident in the way that we have analysed your stories and written this report. We welcome your feedback.

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Research and study leave for Julie enabled her to analyse the field materials and write the first draft of this report in a concentrated period. Julie would like to acknowledge the provision of this leave by the University of Auckland. Our thanks to the two reviewers whose critiques have much improved our monograph. Finally, for the production of this monograph we thank Dr Melinda Allen, Chair of the RAL Committee, Hamish Macdonald for the book design and Tim Mackrell and Peter Quin of the Department of Anthropology for help with the illustrations.

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Glossary

Anti-D A product made from human blood that contains antibodies to Rho (D). It is offered to women who are Rh-negative if there is a chance their blood may have mixed with their Rh-positive baby's blood. It prevents the development of Anti-D antibodies. See http://www.nzblood.co.nz/site_resources/PDF_Documents/rcp_anti-d.pdf for more information.

Chorionic Villus Sampling (CVS) – DNA test which samples tissue from the placenta.

DDAVP Desmopressin acetate - A synthetic product derived from a hormone which may be taken as a nasal spray to reduce bleeding. It stimulates the release of FVIII.

Factor VII, VIII, IX Large proteins that aid blood clotting.

Inhibitors In haemophilia, substances that inhibit the actions of clotting factors. Treatment for this involves tolerisation.

NZORD New Zealand Organisation for Rare Disorders.

PGD Preimplantation genetic diagnosis — a procedure used to test early human embryos: (1) creation of embryo in vitro; (2) removal of one or two cells from embryo; (3) genetic testing of these cells for abnormalities; and (4) transfer of the unaffected embryo to a woman's uterus.

PND Prenatal diagnosis. Foetal cells are examined from the amniotic fluid, placenta or foetal blood to look for abnormalities before birth.

Portacath A device implanted under the skin that enables repeated venous access without the necessity for repeated needle sticks.

Target joint A joint which is the site of recurrent bleeding. Common target joints are the knees, elbows and ankles.

Tranexamic acid An antifibrinolytic agent used to prevent or treat bleeding. May be taken orally or by injection.

Tolerisation Treatment involving saturating the patient with large volumes of clotting factor to overcome inhibitors.

vWD von Willebrand Disease. An inherited bleeding disorder in which the so named clotting protein is impaired. It affects men and women equally.

For further information on all aspects of haemophilia, please see http://www.haemophilia.org.nz

Appendix A: Summary Report of "A Bleeding Nuisance" (1995)

Report To Participants In The "Living With Haemophilia In Aotearoa New Zealand" Research Project

Julie Park, Kathryn Scott, John Benseman and Elizabeth Berry – January 1996

In 1994 or 1995 you were kind enough to participate in this research project and were promised a summary report when the research was complete - here it is along with our New Year greetings.

This brief document summarises the main findings of the study and gives the recommendations in full. The complete research report (230pp) is entitled *A bleeding nuisance: living with haemophilia in Aotearoa New Zealand* (1995)….

We are grateful to the Health Research Council and to the Research Committee of the University of Auckland for the funds which made this research possible. The aims of the research, namely to document:

- the social effects of haemophilia,
- treatment services for haemophilia,
- and to produce a social profile of people with haemophilia

did not change during the project. However, certain aspects became more important than we had anticipated, e.g., the experiences of carrier women and the importance of hepatitis C (nearly 60% of those receiving blood products were infected). These two subtopics became the subject of students' enquiries (Ms B Strookappe and Mr R Hanssens) leading in each case to substantial reports.

The research was based on a questionnaire survey of the whole of New Zealand in which about half of the people whose current addresses were on the haemophilia register took part (193), on 80 interviews with families and people with haemophilia and various medical and nursing staff who specialise in haemophilia, on informal participation by the researchers in a range of `haemophilia' events, and on statistical and other reference information.

Although fair numbers of people with mild haemophilia took part (46%), we had higher proportions of those with severe (33%) and moderate (21%) haemophilia. We also had higher than average numbers of people with Factor IX deficiency (23%). However, we had a representational coverage of all parts of the country and an even representation of age groups. We feel confident of our results especially as we had the chance to have drafts of the report read by various experts including several people with haemophilia and parents: our sincere thanks for this.

The full report will be read by many people who know nothing about haemophilia, so in it we take the time to explain what the condition is, and the various complications and problems that have arisen through haemophilia itself and through the blood-borne viruses. This is not necessary for this audience! We can go directly to the **themes** which emerged from the research.

Because haemophilia is a rare and widely dispersed condition it poses severe challenges for regionalised health and blood transfusion services. People with haemophilia and their families become experts on the condition and its treatment (much of which is carried on at home). This creates special needs for support, and puts these people in the role of public educators - even for non-specialists members of health professions. Again this creates points of tension. The role of the Haemophilia Society in supporting and informing its members and in educating official bodies is crucial and with more resources it could be even more effective.

Overall, people with haemophilia show a high degree of satisfaction with their regular treaters and treatment services. Only A&E, physiotherapists, and hospital physicians, were rated as less than satisfactory, on average. However, our study showed regional inequities in treatment and indicated a need for increased treatment with clotting factors to minimise joint damage. Young people who were born after clotting factors were available are still reporting damaged joints in some geographical regions.

Uncertainty, variability and unpredictability are key features of haemophilia, of HIV, of hepatitis C and of the situation of carriers. The importance of prophylactic treatment in eliminating much of the uncertainty of severe and sometimes moderate haemophilia cannot be overestimated. For other sources of uncertainly, good information, good relations with treaters, good treatment and good support go a long way to improving quality of life.

The costs of effective treatment and the necessity to weigh these against other health needs create another set of issues, especially when there are rising numbers of people with haemophilia in the younger generations. In this context, genetic counselling and decisions about fertility by both men and women have an increasing importance.

Over one third of the study participants had some joint impairment. Prevention through adequate treatment is a priority but maximizing the mobility of those whose joints are already damaged and minimising the pain are also priorities. Only about half the people who replied

to questions about pain were satisfied with their pain relief. Using strong opiate-based drugs was a major concern, as was accessing adequate relief. Concerns with mobility and independence surfaced frequently.

A gender-focused analysis of aspects of haemophilia has raised questions about the socialisation of young New Zealand males, especially with regard to contact sports, has highlighted the experiences and needs of `expressed carriers' (i.e., women with haemophilia) and has indicated the importance of men being involved in decisions about having children.

People with haemophilia have been politicised through experiences with blood-borne infections, are deeply concerned with the safety and adequacy of the blood supply (71% were uncertain or unconfident about the safety of blood supplies) and have had a series of struggles to gain compensation. Blood borne viruses bring not only anxiety, illness and death but stigmatisation of the whole haemophilia community. The continuing safety and control of the blood supply is a major issue.

The haemophilia community has experienced enormous changes in recent years because of the reorganisation of nearly every health- and welfare-related institution. Change processes themselves have been disruptive, and some of the changes, particularly increased regionalisation, have created serious problems for the equitable provision of haemophilia services.

In summary, haemophilia is a genetic condition which has complications running the gamut from infectious diseases to disability. Comprehensive studies of the social effects of haemophilia are extremely rare. This research therefore contributes to an understanding of the consequences of haemophilia in people's everyday lives. Haemophilia offers a range of challenges to the health services. These are intensified by the small and scattered population, especially in the wake of reorganisation of the health and blood services. In addition to its special interest to the haemophilia community, this research provides a platform for evaluating many of the recent changes in the health system, ACC and associated services which are being examined in a series of shorter papers aimed at relevant audiences.

On the basis of this study we **recommend**:

1.1 A haemophilia service provided on a national basis and comprising:
   a] a national definition of a comprehensive needs-based service;
   b] a national register of patients;
   c] a national information and resource centre;
   d] a national and regional structure for service provision,
   e] funded nationally based on patients' needs-based funding provision.

1.2 A national blood transfusion service which would reduce the unpredictabilities and unevenness of demand for clotting factors currently experienced by those regions which have large numbers of severe and/or adolescent patients, and not financially disadvantage those regions.

1.3 An increased capacity for the Outreach service of the Haemophilia society to support people with haemophilia in all parts of New Zealand. This would have funding implications as further outreach workers and back up resources would probably be necessary.

1.4 Following an increase in the capacity of the Outreach service, support networks should be set-up and resourced in all appropriate parts of the country.

2.1 Men and women carriers of haemophilia, wherever they live, should have access to good information about genetic testing for themselves and their children, and support from a trusted other in making decisions about children.

3.1 Assertion training for new parents in dealing with doctors, hospitals and social services.

3.2 Resource packs for GPs and other health professionals, available on an `as needed' basis, such as when they have a first patient with haemophilia.

3.3 Guidelines for home treatment, including recommendations for when the home treater should seek outside advice and support.

3.4 Individualised agreed treatment guidelines, along the lines of the NZHS `Standards of Care', for the treatment of people with haemophilia and expressed carriers. Among other things, these guidelines should have recommendations about the speed of treatment and reiteration of the maxim: `If in doubt, treat'.

3.5 Involvement of people with haemophilia, and their families where appropriate, in decision-making partnerships with treaters.

3.6 Involvement of people with haemophilia in student medical and nursing training. More education for the general public to be available as needed and targeted to educators and employers.

4.1 The funding of prophylaxis and the provision of an adequate supply of NZ plasma-based and recombinant factor concentrates is a priority in the treatment of haemophilia, and should be available to those who can benefit from it and request it.

4.2 For those for whom prophylaxis is not indicated, adequate treatment to minimise bleeds and deal with related complication should not depend on geographical location.

4.3 Continued efforts to conserve factor concentrates, as at present, but not at the expense of adequate treatment

5.1 A safe blood supply is a precondition of safe management of haemophilia. The Blood Transfusion Trust appears to be the most appropriate body at present to be responsible for the safety of the blood supply.

5.2 Patients and their families should be kept fully informed by their treaters of potential risks as they become known.

5.3 Satisfactory compensation and the best available treatment at no cost to those infected, must be provided to those who may become infected through contamination of the blood supply.

5.4 Recombinant products should be increasingly integrated into treatment and funded by the health service. This may necessitate a change of emphasis for blood transfusion services.

6.1 Further recognition is required that both men and women carry haemophilia, and that some women as well as men suffer from the coagulation disorder, and that both men and women are concerned with parenting.

6.2 Educators, physical education specialists and young people's organisations should be encouraged to react creatively to the special recreational needs of children and young people with haemophilia.

7.1 Further representation of people with haemophilia as consumers on official bodies, and increased consultation by Ministry of Health, RHAs and related bodies with the NZHS when official reports and discussion documents are being prepared.

8.1 Organisations undergoing change should be required to minimise any adverse effects of that change on their clientele and take responsibility for informing their clientele of the changes and their implications. These requirements should be monitored as part of regular institutional audits and service evaluations.

8.2 Adequate consultation, research and discussion takes time. However important decisions affecting people with haemophilia should be made expeditiously according to an agreed timeline.

Where to now? As a colleague/participant pointed out: implementation is the problem! We plan to discuss these recommendations and our rationale for them with those individuals, organisations and institutions of State which are involved with the haemophilia community to ensure that they are understood and, if possible, viewed favourably by those who potentially may act on them. But implementation will not occur overnight, and not without many people's efforts.

Appendix B: Example of Participant Information Letter

New Technologies and Haemophilia: Individual and Social Implications

Introduction

You are invited to take part in a study of new technologies and haemophilia in New Zealand. The aim of the study is to find out what people with haemophilia and their families think about new technologies for diagnosis and treatment, and what impact these technologies may have on living with haemophilia and the provision of haemophilia services. Your participation is entirely your choice. If you choose not to take part you will receive the usual medical treatment and support from the Haemophilia Foundation. We will contact you in approximately one week to see if you are willing to take part in this research.

ABOUT THE STUDY

The main technologies that we wish to explore with you are pre-natal diagnosis, pre-implantation genetic diagnosis, gene therapy and new treatments for hepatitis C. However, we are also keen to hear from you about other changes that are having or will have impacts on your lives. This is the second update of our study, which began in 1994 and was published as *A Bleeding Nuisance*.

We are interviewing people with haemophilia, their partners and some members of their families, as well as some treatment and care providers. Approximately 60 participants will take part in this update.

We are based in Auckland, and the study will take place from there, but we will travel to various parts of New Zealand, including Waikato, Wellington and Christchurch and the regions around those cities, to conduct our interviews. Some interviews will also be by phone. People who were part of our original study will be invited to participate, as well as people who have joined the haemophilia community since then.

The time span for this update study is 16 months.

The study will consist of interviews of approximately one hour and a shorter follow up interview, usually by phone, to discuss matters arising from the initial interview and to receive the participant's feed back. The interviews will be audio-taped with permission, and transcribed. The tapes will be destroyed at the conclusion of the study. Transcripts will be retained with the rest of the study materials in a locked room at the University of Auckland. We plan to keep this material indefinitely, and if you permit it, to archive the transcript in a research archive.

BENEFITS RISKS AND SAFETY

The study aims to benefit people with haemophilia and health professionals working with haemophilia and its complications by providing current information about the reception and use of these new technologies in the haemophilia community.

The study poses no risk and the only inconvenience is the time taken for interview.

Participants will be offered a small gift as a token of thanks.

PARTICIPATION

If you do agree to take part in this research you are free to withdraw from the study at any time, without having to give a reason and this will in no way affect your health care.

GENERAL

Your GP will not be told that you are in the study. However, you may wish to inform him or her.

If you want more information about this study, please contact one of the researchers.

If you need an interpreter, one can be provided.

You may have a friend, family or whanau support to help you understand the risks and/or benefits of this study and any other explanation you may require.

During the interview you do not have to answer all the questions, and you may stop the interview at any time.

If you are a health professional taking part in this study, if you have any queries or concerns regarding your rights as a participant in this study, you may wish to contact your professional organisation.

If you have any queries or concerns regarding your rights as a participant in this study you may wish to contact a Health and Disability Advocate, telephone:
Northland to Franklin 0800 555 050
Mid and lower North Island 0800 42 36 38 (4ADNET)
South Island 0800 377 766 or 03 377 7501 in Christchurch.

For Auckland District Health Board Māori Health support, please contact Mata Forbes, RGON; Co-ordinator / Advisor, Māori Health Services, Auckland Hospital, Grafton Mobile 021 348432;
Tel (09) 307 4949, Extn 7292.

CONFIDENTIALITY

No material that could personally identify you will be used in any reports on this study. During the study the data will be kept in locked University of Auckland facilities, and all personal identifiers will be removed from all research materials.

RESULTS

This study is a phase in an on-going research project. A report of the study will be produced and copies made available through the HFNZ. Participants will be sent a summary report. This phase of the study will also be published in academic and health journals, and the information will contribute to a book on living with haemophilia in New Zealand.

STATEMENT OF APPROVAL

This study has received ethical approval from the Auckland Ethics Committee on behalf of the Wellington, Waikato, Canterbury and Otago Ethics Committees.

Please feel free to contact the researcher if you have any questions about this study.

Appendix C: Publications from the *Living with Haemophilia* Project in chronological order

Park, J., K. Scott, J. Benseman, and E Berry, 1995. *A Bleeding Nuisance: Living with Haemophilia in Aotearoa New Zealand.* Auckland, Department of Anthropology, University of Auckland, pp.228.

Park, J., and B. Strookappe, 1996. Deciding about Having Children in Families with Haemophilia. *New Zealand Journal of Disability Studies*, 3:51-67.

Howden-Chapman, P, J. Park, K. Scott, K, and J. Carter , 1996. `An intimate reliance: health reform, viral infection and the safety of blood products', in P. Davis (ed.), *Intimate Details and Vital Statistics*. Auckland, Auckland University Press. pp.168-84.

Park, J., 1996. "It's a hidden thing": Implications of the Rarity, Invisibility and Unpredictability of Haemophilia for Everyday Life. *Health-related papers from the Sociological Association of Aotearoa Annual Conference, Akaroa, December 1995.* P. Norris (ed). Christchurch, Sociology Department, Working Paper 16, University of Canterbury.

Scott, K., 1997. Haemophilia Care Not Just the Domain of Specialists. *New Zealand Family Physician,* 24(5):16-18.

Benseman J., and J. Park, 1998. A Bleeding Nuisance: the Implications of Haemophilia for Education. *Australasian Journal of Special Education*, 21(2):81-97.

Park, J., 1998. Technologies of Prenatal Testing: the Implications of an Inherited Condition for Women and Families. *Pacific Science Information Bulletin*, 49(3-4):33-37.

Park, J., K. Scott and J. Benseman, 1999. Dealing with a bleeding nuisance: a study of haemophilia care in NZ. *New Zealand Medical Journal*, 112:155-8.

Park, J., 2000. "The only hassle is you can't play rugby": haemophilia and masculinity in New Zealand. *Current Anthropology*, 41:443-453.

Park, J., 2004. Haemophilia and hepatitis C: Civil society and the State in New Zealand. Abstract for Poster presentation. XXVI International Congress of the World Federation of Hemophilia, October 2004, Bangkok.

Park, J., 2005. Beyond "his sisters and his cousins and his aunts": Discourses of haemophilia and women's experiences in New Zealand, in C.Gross, H.Lyons and D. Counts. *A Polymath Anthropologist*, RAL 6, Auckland, 97-104.

Park J. and D.York, 2006. "Carrying": Issues for women in the New Zealand haemophilia community. Abstract for paper presentation: XXVII International Congress of the World Federation of Hemophilia, May, 2006, Vancouver.

Appendix D: Interview outline, "New technologies and haemophilia" study.

"Bleeding Nuisance" Update Study 2005

Opening Explanation (applicable to everyone)

In our earlier study of living with haemophilia, we found a wide range of views on prenatal diagnosis and carrier testing. Since then pre-implantation genetic diagnosis has become available in several countries and there has been considerable debate about it.

We would like to ask you your views on these procedures and the issues that they raise. In addition, if you or family members have had experience with these processes, and if you are prepared to talk with us about them, we would very much like to hear about them.

Everyone over 16yrs

Prenatal Testing (Amniocentesis or CVS)

Are you familiar with this? (Explain briefly if not.) If familiar — how does the testing work?

Do you think it is accepted in the NZ haemophilia community?

Have you talked about it much with others?

What are your ideas about the desirability of PNT? [Reasons for and against, risks, situations where it is most desirable, least desirable.]

Can you tell us about your experiences with deciding to have it or not to have it (if relevant)?

If you have had it done, can you take us through your experience of the process (i.e., how did it work and also how did you feel about it, and also the other family members involved)?

Pre-implantation Genetic Diagnosis

Are you familiar with this new process? (May need to explain briefly.) Where did you learn about it? Have you talked about it with other people?

What are your views on the desirability of this procedure? (May need to probe for issues — its risks, ethical or religious viewpoints.)

Would you consider having this procedure yourself (for woman or couple) or advising a family member to consider it? Why, why not? (And there may be one or two people who have had it done.)

Females whose Mothers are known or possible carriers —

Carrier testing

Do you have any bleeding problems?

Could you tell us your ideas about the advisability (or not) of testing for carrier status.

[If they think testing is advisable] When do you think it is the best time for testing to be done?

Have you talked with other people about this issue? [Why?/Why not? Possible probe as to their relationship to person.]

How does testing work? Where did you learn about it?

Has carrier testing ever caused any tension for you or your family?

Have you had experience with the testing process? YES or NO

[If YES]

Can you take me through the process as it was for you and the other family members involved?

What were the effects of the process itself and of the outcome?

What did you (or they) do with the knowledge gained?

Everyone

Gene Therapy

As well as issues surrounding carrier status, pre-natal testing and so on, our previous study found that there was an interest in the possibilities of gene therapy among the haemophilia community.

What is your understanding of gene therapy? (explain if not familiar) How familiar do you feel you are with this topic?

Is this a topic that interests you? (Why?/Why not?) Have you discussed gene therapy with your family or others? What kind of issues did you raise?

Do you think this technology could offer a cure for haemophilia (or a related bleeding disorder)? Are you wary of gene therapy in any way? (Why?/Why not?)

If gene therapy could offer a partial cure for haemophilia (or a related bleeding disorder), would you consider having this treatment?

Hep C is another issue that was widely discussed in the earlier study that has affected the haemophilia population. Even if it has not affected you directly, we would still be interested in knowing your views on this topic. Would you feel comfortable discussing this topic?

People who are willing to talk about Hep C issues

Have you, or anyone in your family, been affected by HCV? (Ascertain who and their relationship.)

Did you/they have a Hep C positive diagnosis at any stage?

What kind of symptoms have you/they experienced/are you experiencing?

Have you/they been given the option for HCV treatment?

When did you first hear about treatment options available to you/them?

Did you/they accept treatment? (Why?/Why not?)

Was this combination therapy or monotherapy?

How long did treatment last?

Can you tell me about your/their experience of this treatment? (Be it mono or combo.)

Did you/they clear the virus via this treatment? (Probe further depending on YES or NO answer.) Do you talk much with others about your/their HCV status? (Why?/Why not?)

Possibly discuss dealings with ACC

Everyone

Are there any other issues that you would like to raise/ discuss?

Thanks to participants.

Research in Anthropology and Linguistics (*RAL*) is a refereed series dedicated to the five subfields of Anthropology (Archaeology, Biological Anthropology, Ethnomusicology, Linguistics and Social Anthropology) with a regional focus on New Zealand, the Pacific, and Australasia. *RAL* was initially established in 1995, an outgrowth of *Working Papers in Anthropology, Archaeology, Linguistics and Maori Studies* (discontinued in 1990).

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Preference is given to research undertaken by staff and students of the University of Auckland, Departments of Anthropology and Linguistics, but other manuscripts that deal with subjects of special interest to anthropologists are also considered. The issues in each series are numbered consecutively as independent contributions and appear at irregular intervals.

Prospective authors are referred to the *RAL* page of the Department of Anthropology web site for the current Monograph Style Guide: http://www.arts.auckland.ac.nz/departments/index.cfm?P=9711

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1 *Protecting Historic Places in New Zealand.* Harry Allen. 1997. Out of print.
2 *Consuming Identity: Modernity and Tourism in New Zealand*. John Taylor. 1998. $20.
3 *Raurimu Frontier Town 1900-1925*. Kate Hill. 1999. Out of print.
4 *Holiday Communities on Rangitoto Island*. New Zealand. Susan Yoffe. 2000. Out of print.
5 *Australasian Connections and New Directions: Proceedings of the 7th Australasian Archaeometry Conference*. Martin Jones and Peter Sheppard (editors). 2001. $35.
6 *A Polymath Anthropologist: Essays in Honour of Ann Chowning*. Claudia Gross, Harriet D. Lyons and Dorothy A. Counts (editors). 2005. $50.
7 *Oceanic Music Encounters — the Print Resource and the Human Resource: Essays in Honour of Mervyn McLean*. Richard Moyle (editor). 2007. $40.
8 *The Social Ecology of New Technologies and Haemophilia in New Zealand:* A Bleeding Nuisance *Revisited.* Julie Park and Deon York. 2008. $25.