

## Coping with joint pain in haemophilia

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### Summary

Some Rolling Stones fans might think that joint bleeds in haemophilia are now more a case of 'Its all over now' than 'Let it bleed', for just as Mick Jagger's song writing skills have changed since the 1960s and 70s, so has haemophilia care - treatment has improved dramatically over the last 30 years. Prophylactic clotting factor concentrates are now commonly used to prevent bleeding episodes among patients with severe haemophilia. Many patients, however, and especially those aged over 40, still live with chronic pain caused by arthritic complications of repeated bleeds into joints, leading to disability and reduced quality of life, and this aspect of the condition has not been widely recognised by health psychologists. Haemophilia patients' experiences of pain were the subject of a recent health psychology work placement and are the focus of an ongoing research project, collaborations with the UK Haemophilia Society, and this article summarises some aspects of that work.

### Introduction

Haemophilia is an inherited blood disorder in which natural clotting agents are absent or reduced, making sufferers vulnerable to spontaneous internal bleeding and bleeding caused by accidents or surgery. The incidence is between 1:5,000 and 1:10,000 males for haemophilia A, and between 1:35,000 and 1:50,000 males for haemophilia B (Haemophilia Alliance, 2001). Haemophilia affects only males, but von Willebrand's disease, a more common but generally milder bleeding disorder affects at least one in 1,000 women as well as men, and around 11,000 people with inherited bleeding disorders are registered with UK Haemophilia centres (Haemophilia Alliance, 2001). Prophylactic (preventative) treatments in the form of clotting factor concentrates can now prevent spontaneous bleeds in those with severe haemophilia but need to be administered intravenously two to three times a week. The most recent concentrates are genetically engineered, and these 'recombinant' treatments are not reliant on large pools of donated plasma and are much less likely to be contaminated with bloodborne pathogens. Many patients with moderate to severe haemophilia now keep factor concentrate at home, having been trained to self-administer by their haemophilia nurse.

Both acute and chronic pain are important issues in haemophilia because internal bleeding, especially into joints, can be extremely painful, and joints damaged by repeated bleeds can become arthritic, causing chronic pain (Choiniere & Melzack, 1987). However, very little systematic research has focused on cognitive and behavioural aspects of haemophilia-related pain, for most of the psychological research in haemophilia has focused on disability and quality of life (e.g. Solovieva, 2001; Triemstra et al., 1998; Szende et al., 2003), effects on partners and families (e.g. Miller et al., 2000; Triemstra et al., 1999) and consequences of HIV infection (e.g. Bordeaux et al., 2003; Marsettin et al., 1998).

With increased use of prophylactic treatments, pain will be a less important issue for younger patients in Western countries, except for those with inhibitors, which prevent clotting factor concentrates being effective. It is also more common nowadays for severely affected sufferers to have joints replaced, improving mobility and quality of life. Despite these developments, however, many patients aged over 40 are affected by chronic arthritic pain caused by past bleeds, and many patients, especially those in developing countries, suffer episodic acute bleeding pain because they do not have access to prophylactic treatments.

The Haemophilia Society is a national charity representing people with haemophilia and related blood disorders in the UK. It has a telephone helpline and offers information, advice and advocacy to people with haemophilia and related bleeding disorders, including those living with HIV and/or hepatitis C. The support it provides in relation to pain is presently based mainly on existing publications (e.g. Jones, 2002; Haemophilia Foundation, 2000). The Society keeps up to date with the latest scientific developments, for example the latest (third generation) recombinant treatments and advances in von Willebrand's diagnosis and care. It supports research and other initiatives to develop and improve patient care and quality of life, and publishes a magazine, *Haemophilia Quarterly*, which is sent to all members of the Society and all haemophilia clinical centres.

### **The work placement**

The Society felt that pain was an area of need and agreed to host a work placement as part of London Metropolitan University's MSc Health Psychology programme (Elander, Troop & Parker, 2003). Jacquie Bailey, then a student on the course, spent one day a week for 14 weeks working with people with haemophilia, with health professionals, and with data from a survey conducted by the Society the previous year. A sequence of activities was planned, each of which informed the development of the next:

1. Analysing data on pain management needs reported by respondents to the Society's 2003 Needs Assessment Survey, to identify factors associated with pain management problems.
2. Conducting a focus group on pain management to obtain insights into the experiences and needs of those affected and elicit suggestions about how those needs should be met by the Society and clinical services.

3. Interviewing haemophilia health professionals about their understanding of patient needs in relation to pain, and about their views on the best approaches to pain management.
4. Developing a pain management fact sheet to be published as an addition to existing informational resources of the Society.

The Needs Assessment Survey was completed by nearly 600 Society members, including 256 people with haemophilia A, 58 with haemophilia B, and 85 with von Willebrand's disease, with the remainder made up of those with other bleeding disorders and carers or parents of sufferers. Since the survey was sent to nearly all the 5000-strong membership of the Society, the response rate was not impressive, but we hoped the results would provide a preliminary picture of the ways in which pain management problems affected people with haemophilia.

Roughly equal proportions of sufferers with each type of bleeding disorder were affected by pain, with 30 per cent of those with haemophilia A, 26 per cent of those with haemophilia B, and 31 per cent of those with von Willebrand's reporting that they had problems managing pain. Similar proportions of sufferers reported other types of problem, with 21 per cent reporting feeling fairly or very unhappy, 32 per cent reporting sleep problems, 42 per cent reporting being disabled, and 45 per cent reporting mobility problems. Chi square tests showed that pain management problems were significantly associated with a number of factors, including feeling unhappy (all of the 'very unhappy' sufferers had problems with pain), having sleeping problems (all of those with pain problems had trouble sleeping, compared with just three per cent of those without pain problems), and mobility (68 per cent of those with pain problems also had mobility problems, compared with 35 per cent of those with no pain problems).

Respondents also provided open format, qualitative descriptions of the problems they experienced with pain, which were analysed using a thematic analysis. The themes that emerged were pain medication issues (including worries about dependency, side effects and adverse reactions to painkillers), arthritic problems (including joint damage and mobility restrictions), coping strategies (including resting, social support and cognitive strategies), and other issues related to pain, including sleeplessness and fatigue, depression and anxiety, and reports that viral infection and associated treatments were causes of pain (Bailey, 2004).

Those issues were explored in greater depth in a two-hour focus group of people with haemophilia recruited from the Society membership by invitation and through an advertisement placed in Haemophilia Quarterly. Six individuals aged between 30 and 65 took part, including five men with haemophilia and one woman with severe von Willebrand's disease who was interviewed separately to allow her to discuss potentially sensitive issues that may have been difficult to explore in an otherwise all male group. The questions were based on the findings of the Needs Assessment Survey and dealt with two broad areas: experiences and services. The focus group was tape-recorded, transcribed, and

subjected to a thematic analysis, which identified frustration, anger, and the need to manage a complicated life as the main themes in participants' experiences of pain. Several participants reported that they felt they had become addicted to painkillers for periods in the past when they were experiencing severe pain, and some of the anger and frustration they expressed was focused on the difficulties they had experienced with getting information and services for pain management. The focus group also highlighted inconsistencies in haemophilia care between different clinical centres, and the desire on the part of participants for an event organised by the Society to enable haemophilia sufferers to try complimentary therapies.

The results of the focus group were then used to inform the interviews with health professionals at two hospital-based haemophilia centres. At each centre, the director (a medical doctor in each case), a nurse and a physiotherapist were interviewed. Again, the interviews were tape recorded, transcribed, and analysed. The themes identified were differences between groups of patients, medication issues, physiotherapy and coping. Interviewees at both centres stressed the importance of physiotherapy and multidisciplinary team working. They saw many patients as very stoical in the way they approached living with haemophilia and were optimistic about the future of haemophilia management, viewing analgesic dependency as a small and diminishing problem. This optimism was tempered, however, by the knowledge that severe haemophilia sufferers with inhibitors will continue to have bleeds until the problem of inhibitors is solved, and that there are also issues in the care of patients with mild haemophilia. Those patients are not allowed to keep clotting factor concentrates at home, and those who do not live near a haemophilia centre sometimes encounter delays and difficulties when they present at a local hospital in an emergency because staff are less well informed about haemophilia care. There were different views among the health professionals about the latest generation of non-steroidal anti-inflammatory drugs. Staff at one centre were very cautious because of the risk of stomach bleeds, whereas those at the other emphasised the benefits in terms of reduced joint swelling, provided the drugs were used under careful supervision.

### **Links with health psychology theory and research**

Many of the themes and issues identified during the placement were consistent with theory about pain and pain coping. The gate control theory, for example, describes how emotional and behavioural factors, as well as nerve impulses from damaged tissue, affect the perception of pain. Participants in the focus group spoke about how their experience of pain was affected by anxiety, tension, and exercise, and described how distractions such as listening to music helped them to live with pain. This is consistent with the cognitive behavioural model of chronic pain, in which beliefs about pain influence behavioural and psychological functioning (Jensen et al., 1999). In chronic pain conditions, beliefs about self-efficacy have been shown to be especially important mediators of outcomes like disability and depression (Arnstein et al., 1999), and the focus groups showed that self-efficacy beliefs are also important in haemophilia. One focus group participant responded, for example, 'I have had this pain before and I know how to deal with it'. Participants who relied less on doctors to manage their condition had less severe problems with pain, and one participant

was actively involved in patient education and the self-management programme. Self-efficacy was also important in relation to the self-administration of factor concentrates, and participants who were more confident about self-administering factor concentrates also tended to have fewer problems with pain.

There was also evidence that patients benefited from making comparisons between themselves and less fortunate people with haemophilia, consistent with theories of cognitive adaptation to illness (Taylor, 1983). One focus group participant, for example, described his own situation in a more positive light after meeting people with haemophilia in Russia, where clotting factor treatments are much less available than in the UK and patients, therefore, have more pain from recurrent bleeds. Finally, the focus group also discussed concerns about dependence on analgesics, and several participants suggested that it was better to hurt a little than be addicted to painkillers. This was consistent with the findings of a recent national survey of haemophilia patients, which showed that the most common concerns about drug use in haemophilia were about dependence on prescribed analgesics (Elander & Barry, 2003).

### **Placement outcomes**

As part of the placement, Jacquie produced a pain management fact sheet for people with haemophilia that is now available from the Haemophilia Society ([www.haemophilia.org.uk/uploads/painmanagement.pdf](http://www.haemophilia.org.uk/uploads/painmanagement.pdf)). This included information about the need for coping skills and complementary therapies as well as medication for pain, with medical advice taken from an article written by a haemophilia consultant and including contributions from other haemophilia professionals. The first issue of the fact sheet will be distributed to Society members with a feedback form and freepost reply service so that it can be evaluated. Jacquie also wrote an article about her work for Haemophilia Quarterly and made a presentation to the staff at the Society, which she hoped would raise awareness about how health psychology can contribute to haemophilia care and help to inform staff who focus mainly on other aspects of the Society's work about issues raised by pain in haemophilia.

The placement also provided the impetus for a project to develop a video about pain coping, featuring people with haemophilia describing their experiences with pain. This will be distributed in the form of a DVD as part of a larger-scale prospective evaluation of interventions to promote more effective pain coping in haemophilia, using the haemophilia-adapted pain coping strategies questionnaire (Barry & Elander, 2002) as one of the outcome measures. A literature review revealed that home-based training in coping strategies has been relatively neglected among interventions for chronic pain (Turk & Rudy, 1991), and the DVD will be designed specifically for people with haemophilia to view at home, with information and encouragement that we hope will promote more effective pain coping. Other outcome measures include the Chronic Pain Acceptance Questionnaire (McCracken, 1998) and the Pain Stages of Change Questionnaire (Kerns et al., 1997). We hope that by examining the relationships between pain coping and those measures we will be able to

place pain coping in haemophilia in the wider context of theory developed from research in other chronic pain conditions.

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