psychosocial aspects of life with haemophilia

Literature review, prepared for Novo Nordisk as part of the HERO initiative
HERO – Haemophilia Experiences Results Opportunities

HERO is an international community inquiry initiated and supported by Novo Nordisk. This initiative will build a solid understanding of life with haemophilia, seen from the perspective of people with haemophilia, their families and their healthcare providers. It is our goal that their voices are heard to bring about action and change.

The inquiry has three main components:

- Literature review, researching existing knowledge of psychosocial factors in haemophilia
- Initial investigation, including 150 structured face to face interviews with an international cross-section of the haemophilia community to gather insight and provide guidance on development of:
- Full investigation, involving an on-line survey of around 1200 participants from 12 countries.
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1.0 Introduction

This report is a review of published research on the perceived value of addressing psychosocial factors in the treatment of haemophilia. From the first discussions in developing HERO, particularly at an international multidisciplinary expert meeting held in Montreal in 2009 (find more at www.changingpossibilities.com), it was clear that a great deal of knowledge and thinking exists on the subject of psychosocial support in haemophilia. This review was therefore conducted to provide a foundation of understanding upon which the HERO International Advisory Board and Novo Nordisk would develop the interviews and survey that make up HERO, ensuring that HERO provides new and valuable insights.

The report is set out in two parts. First, the literature on the impact of the psychosocial challenges on people with haemophilia is discussed. Second, the literature on how best to address these challenges is reviewed.

1.1 Summary

The review confirms the broad agreement that managing the psychosocial factors in the treatment of haemophilia is a worthwhile and necessary goal (Barlow, Stapley, Ellard & Gilchrist, 2008; Beeton, Neal, Watson & Lee 2007; Bottos, Zanon, Sartori & Girolami, 2007; Cassis, 2007; Ross, 2004; Bullinger, & Von Mackensen, 2003; Wiedebusch, Polman, Siegmund & Muthny, 2008). The benefits of active and multidisciplinary psychosocial treatment are seen as considerable (Cassis, 2007), not least in that they reinforce and support good medical care and adherence to medication (Geerts & Van de Wiel; Penica & Williams, 2008; Tamminga, 2008).

The review highlights that the ideal is for a comprehensive approach to treatment that incorporates the specialist skills of psychologists, social workers and counsellors in a complementary relationship with healthcare professionals.

Advances in medical treatment of haemophilia in terms of prophylactic and ‘on-demand’ treatment during the 1970s mean that a relatively normal life can be enjoyed by individuals with haemophilia in those countries with the necessary healthcare resources (Beeton, 2007; Dolan, Hermans, Klamroth, Madhok & Schutgens, 2009; Geerts et al, 2008; Koiter, Van Genderen, Brons & Nijhuis-Van Der Sanden, 2009; Lambing & Kachalsky, 2009). Moreover, the improvements in purification of blood and blood-screening and the availability of recombinant replacement factors means that the risk of associated diseases such as HIV and Hep C are now low (Dolan et al, 2009; Sulser, 2006). From a purely medical perspective, haemophilia can largely be controlled (Beeton et al 2007; Dolan et al 2009; Lambing & Kachalsky, 2009; Mauser-Bunschoten, Fransen Van De Putte & Schutgens, 2009).

The literature suggests that nowadays living a better life with haemophilia is both possible and common for parents and patients with access to the right medical care. Significant improvements over recent years mean that outcomes and quality of life for people with haemophilia are better than ever before (Beeton et al 2007; Dolan et al 2009; Gringeri et al, 2008; Lambing et al 2009; Mauser-Bunschoten et al 2009).

In fact, the lifespan of an individual with haemophilia has risen dramatically. In the US, for example, average life expectancy has risen by a decade, from 61 to 71 (Soucie et al, 1998). This welcome improvement brings a new range of considerations associated with aging, particularly in dealing with co-morbidities such as diabetes, hypertension and heart disease, which require careful medical treatment in, for example, the selection of drug therapy (Lambing et al, 2009).
It must be noted that the great majority of literature is focused on Western Europe and/or other developed countries. Therefore differences between developed and developing countries, or differences between countries where haemophilia treatment is readily available and those where it is not, are rarely commented on.

Many authors point out that improved medical treatment of haemophilia alone will not maximise quality of life without appropriate psychosocial support (Barlow et al, 2007; Beeton et al, 2007; Cassis, 2007; Wiedebusch et al, 2008). A great deal of psychological adjustment is needed at the point of diagnosis and beyond. Both parents and patients unquestionably benefit from a range of psychosocial support to deal with a formidable range of challenges.

In conclusion, this review shows a consensus arguing for more psychosocial support for individuals, their families and acquaintances and suggests that the investigation being undertaken by HERO, exploring the challenges identified in the literature, will add useful structured evidence to the debate. This research could help clarify the experience of individuals with haemophilia and their families in different countries, including the level of psychosocial support which is and has been made available.
2.0 The Psychosocial challenges of Haemophilia

These challenges have been divided into three parts, the first set of challenges relates to the issues faced as a result of diagnosis and treatment; the second, to social challenges and includes the impact on family life; and the third relates to specific psychological challenges for the individual.

2.1 Challenges of diagnosis and treatment

2.1.1 Early prophylaxis & in-home treatment

Improvements in medical treatment have removed much of the uncertainty surrounding the condition. For example, administering treatment in the home gives a greater sense of control and normality to life for all concerned.

“Prophylactic treatment has improved parents’ sense of control leaving them less anxious and less inclined to feel pressured to monitor every aspect of their child’s life.”

(Weidebusch et al, 2008).

Accessibility of medical treatment has also had a huge impact on quality of life for mothers – those who lived in areas where factor consumption was at its highest, report better quality of life than those living elsewhere (Tedgard, Ljung, Bullinger, & Von Mackensen, 2008).

The early stages of treatment of haemophilia are a significant area of concern. Media scares regarding the safety of blood products mean that nurses often need to reassure anxious parents about the purity of blood products at the onset of treatment (Walters, 1995). More recently, risk of Creutzfeldt-Jacob disease (CJD) in blood products has been a real concern for the population with haemophilia (Sulser, 2006).

In addition, in-home treatment can often seem a daunting responsibility for parents – uncertainty as to whether a fall will result in a bleed means parents are constantly anxious due to their lack of experience of the disease. Also, initial attempts to locate the veins of infants as young as 12 months can be traumatic (Beeton et al, 2007).

2.1.2 Acceptance of initial diagnosis and early management of disease

The disease can be frightening for those who haven’t previously encountered it in the family, and initial diagnosis tends to have a huge financial and emotional impact on the whole family. The literature highlights that diagnosis of the disease has a life changing effect on parents who feel shock and guilt, resulting in a lack of self esteem (Beeton et al, 2007). Studies reveal that the quality of life is most impaired at the initial stages of the diagnosis (Bullinger et al, 2003). Suspicions regarding child abuse due to severe bruising sometimes complicate diagnosis to make this situation even more traumatic for parents (Beeton et al 2007; Cassis 2007).

The majority of the literature suggests that mothers have a key role in the treatment of their sons and some papers refer to the ‘special guilt’ some mothers feel as carriers, who feel they are ultimately responsible for their son’s haemophilia (Ross, 2004).

This guilt can be overwhelming and can give way to passiveness and despair at a time when parents need to focus on getting the best medical treatment for the child (Ross, 2004). Guilt can result in depression and a rejection of their child (Cassis, 2007) or by contrast overprotection and overcompensation which can ultimately lead to the child using their haemophilia as a means to avoid challenges, responsibility and adulthood (Ross, 2004).

Not surprisingly, the psychosocial support on offer for parents at this time is crucial to help acceptance of the diagnosis and deal with the changes it will have on family life. Parents need help to manage their emotions in order for acceptance and learning to take place (Cassis, 2007). If parents are given the information and support they need at this stage it can positively affect...
their view of the condition by reducing both anxiety and the need to overprotect the child – things which can do long-term damage to the emotional health of the child (Beeton et al., 2007; Cassis, 2007; Remor et al., 2003; Wiedebusch et al., 2008).

Mothers and fathers can react very differently at the diagnosis stage. Disappointment, helplessness and frustration can lead mothers to want to talk; fathers may want to simply withdraw or alternatively regain their control of the situation through information (Cassis, 2007; Ross, 2004).

The challenge is to recognise these feelings in order to help parents deal with them and work together. Indeed much of the literature emphasises the importance of communication between parents – joint decisions regarding the illness help alleviate the strain of managing the condition.

“Supporting each other in daily care, talking about emotions and solving illness related problems together are adaptive coping strategies for managing the psychological burden of caring for a child with haemophilia.” (Weidebusch et al., 2008).

2.2 Social & Family Challenges

2.2.1 Impact on family life

Parents can feel overwhelmed by responsibility in the management of the life of their child with haemophilia. While most of the time a normal life is possible, the unpredictable nature of the disease means that setbacks can occur. After long periods of normality, these can make the patient and family feel vulnerable once again. (Beeton et al., 2007).

In addition, there is little respite from the pressure that parents feel, for example, the need to be constantly contactable. Pressures include ensuring that emergency measures are in place for the child in their transport and activities.

Therefore, despite having access to medical treatment, parents can feel the world is a dangerous place for their child. Much of the literature suggests that parents cope with the disease less well than their child (Beeton et al., 2007).

“Parents tend to overestimate problems in certain areas of their child’s life.” (Bullinger et al., 2003).

The tendency to fix all the attention on the child with haemophilia can lead to neglect of other family members and adverse effects on social and family life (Bottos et al., 2007; Evans et al., 2002; Goldstein & Kenet, 2002). Family life is often disrupted and activities abandoned at the last minute due to spontaneous bleeds (Shoenmakers et al., 2001). Siblings can become jealous and then feel guilty at being jealous (Ross, 2004).

2.2.2 Fostering independence

Parents can find it difficult to find a balance between autonomy and setting limits so as to foster independence and confidence in the child and avoid overprotection and dependency.

Children still need to learn management of the disease, such as recognising the onset of bleeds and the appropriate action to take, in order to develop acceptance, control and independence (Cassis, 2007).

Parents who are able to foster a sense of confidence and independence will see an improvement in their own quality of life, as they see their child managing their disease and enjoying life within the limitations set by the condition. (Beeton et al., 2007)
2.2.3 Adolescence – transition from paediatric treatment to adult healthcare

Adolescence presents a new challenge for parents and patients. A certain amount of rebellion is part of teenage life and studies suggest that, in common with other conditions, treatment compliance is lower at this stage. Adolescents want to do the same as their peers and so they tend to push the boundaries of what they can do. Many adolescents with haemophilia risk doing long-term damage to their joints which can result in disability (Geerts et al., 2008).

One study that looks at the transition from paediatric to adult healthcare reveals parents to be more concerned than the patients themselves. Mothers are more worried than fathers about the medical issues around transition. However, both parents rate the quality of life of their son to be poorer post-transition vs pre-transition. The study speculates that this is due to parents perceiving a relinquishing of control of their son’s medical condition (Geerts et al., 2008).

2.2.4 Secrecy through fear of social stigma

Secrecy about haemophilia can be found at all stages from diagnosis onwards. Unless haemophilia is very severe, making the disability outwardly visible, the condition can go largely unnoticed. This can mean that some parents and individuals choose not to tell people about it:

“Not knowing what to tell the family and wider social networks about the disease can make acceptance of it much harder for the family and the child with haemophilia. It is sometimes easier not to talk about it.”

(Beeton et al., 2007)

The theme of secrecy can be seen at each developmental stage. For example, parents may tell the schools about their son’s haemophilia but this will not always be communicated to the individual’s peers through fear that he may be treated differently; the adolescent individual may hide bruises from his family through guilt and not wanting to be a burden to his family; an adolescent with haemophilia may be concerned over how and when to tell someone about haemophilia, and this may impede the development of intimate relationships; in finding employment and also at work, where feelings about social stigma can stop the adult individual finding satisfying employment (Cassis, 2007).

Secrecy can impede the development of trusted relationships and thus acceptance of themselves and their condition for patients and family (Beeton et al., 2007). Most of the qualitative methods note social isolation as being a negative side effect of haemophilia for parents and patients. However, one Quality of Life (QoL) study found that individuals with haemophilia or their parents are no more isolated than their age and gender equivalent cohorts (Hartl, Reitter, Eidher, Ramschak & AY, 2008).

According to some studies that look at haemophilia from the perspective of carriers, many fathers with haemophilia manage their condition in secret, preferring not to talk about their haemophilia or explain the condition to their daughters who are themselves potential carriers. Given this, many daughters become mothers without knowing their obligate carrier status (McGregory, Boddington, Dimon, Atkinson, Clarke et al., 2007; Ross, 2004).

Misconceptions amongst the general public or colleagues regarding haemophilia such as all individuals with haemophilia are HIV and gay; that you can’t touch them or they’ll bruise; or that you should avoid close physical contact; are also documented.

“90% felt that there is a lack of understanding amongst the public regarding bleeding disorders. Over 50% were careful about informing others that they had a bleeding disorder.”

(Barlow et al., 2007).
2.2.5 Participation and isolation

Individuals with haemophilia can lead a normal life within set limits but these are not the same as for the rest of the population. This difference manifests itself in different ways at different life stages, but commonly involves a decision about when not to participate in an activity, which inevitably leads to a degree of isolation.

For example, children with haemophilia can now participate in school; enjoy a variety of activities and certain sports. However, they are not free to do exactly what they like, and tend not to participate in competitive or contact sports. This means that:

“Children must manage the frustration of not having impairments but having restrictions imposed regarding their energetic pursuits”

(Beeton et al, 2007)

There are examples throughout the literature of childhood experiences being denied to children with haemophilia. One paper cites a small child who could not participate in a friend’s birthday party because it involved a bouncy castle, yet he could see the party from his bedroom window. His mother “still cries about it now.” (Beeton et al, 2007).

Plainly, this kind of situation is psychologically demanding for both parent and child. The child with haemophilia must work within a restrictive framework which recognises the dangers of their condition. Practically this can mean that they must withdraw from sports as they age, and the games become more physical and the injury risk rises. This has been highlighted as a very difficult time for both the child and the parent who often has to enforce this decision, and contributes to the fact that adolescence is a crisis point in the life of many individuals with haemophilia (Cassis, 2007).

In addition, in a Quality of Life study, older children were reported to have higher impairments on ‘social’ dimensions which relate to ‘perceived support’ and ‘friends’ and also in ‘dealing’ which related to adaptation to the condition (Gringeri et al, 2004).

The area of work is touched on only lightly in some of the literature but seems to suggest that a need for support around employment is necessary for individuals with haemophilia. One study reported that compared to a cohort from the general population, people with haemophilia are less involved in full time paid work and suffer more from occupational disability (Plug, Peters, Mauser-Bunschoten, de Goede-Bolder, Heijen et al, 2008). Another, by Hartl et al (2008), found that adults with haemophilia are less likely to be in work or training than in the age-gender equivalent cohort studied, although positively, they are more likely to be married.

Isolation is likely to be even more significant in countries where prophylaxis is not readily available. For example, in a Polish study, a sample of individuals with severe haemophilia who had not received primary prophylaxis found that 38% were unemployed and received some form of social subvention (Windyga, Stefanska, Lopaciuk, Juszynski & Wozniak, 2005).

Adults with haemophilia sometimes give up work due to frequent time off resulting from their condition which has financial consequences for themselves and their families as well as feelings of dependency. Also they are more likely to take early retirement due to their condition and associated co-morbid conditions (Barlow et al, 2007). Cassis (2007) stressed the importance of careers counselling regarding training options and work for young adults in order to find work that is suitable for them. The need to make a contribution to society by working is essential for adult individuals with haemophilia and has a hugely positive impact on their self-esteem.
2.3 Psychological Challenges
The precise psychosocial impact of the disease varies according to age but general themes persist. They include: fear of what the disease can provoke and fear of chronic pain; denial; anger and frustration; depression or lack of motivation. These themes are often connected and as such, they are dealt with in two sections below.

2.3.1 Fear of pain and pain management
One of the major challenges for the patient themselves is learning to manage the stress of chronic pain and dysfunctional joints (Manco-Johnson, 2003; Valentino, 2007).

The importance of working with the child from an early age to enable them to understand their pain and the benefits of the treatment is reported by Cassis (2007). Cassis emphasises the importance of getting the child to report the bleed as quickly as possible; to adhere to treatment such as physiotherapy sessions and to manage their pain via visualisation techniques. A child who grows up with this approach will soon come to realise the benefits of treatment.

Another study by Penica & Williams (2008) endorses the use of distraction and counter-conditioning interventions which are reported to reduce stress and anxiety and therefore help adherence to treatment in young children, and therefore help to avoid joint damage in the long term.

It is not surprising then that many of the psychological problems experienced by individuals with haemophilia stem from pain and the physical impact of the condition. Arthropathy, repeated bleeds, arthritis and a history of orthopaedic surgery reduce the quality of life in people with haemophilia and result in frustration and lack of independence because of reduced mobility (Barlow et al, 2007; Beeton et al, 2005).

2.3.2 Anxiety and depression, anger and frustration
Depression is most commonly found in individuals with haemophilia who have contracted HIV or Hepatitis C through contaminated blood products. The social stigma that is attached to these diseases along with the side effects of treating these co-morbid conditions, can lead to depression and anxiety (Barlow et al, 2007).

Interestingly, a study by Golden et al (2006), found fewer feelings of stigma, depression and anxiety amongst people with haemophilia who had contracted Hepatitis C as a result of contaminated blood products compared with those who had contracted it due to injecting drugs or iatrogenic disease caused by transfusion.

Anger and frustration, anxiety and depression are common themes in the literature along with a feeling of uncertainty about the future both for themselves and for their families (Barlow et al, 2007; Beeton et al, 2007; Bottos et al, 2007).

However, while these themes tend to be recognised in the qualitative literature, where information is generated by the individuals themselves, they do not always appear in QoL studies, for example, Hartl et al (2008) which acknowledged greater pain problems and problems with physical functioning, general health, social functioning but not differences in mental health from an age-gender equivalent cohort. Stieltjes et al (2009) found that: “Older haemophiliacs tend to have the best mental quality of life, contrasting with their highly impaired orthopaedic condition and physical quality of life.”

To explain the higher reported quality of life in some studies, Beeton et al (2005) talks of the ‘disability paradox,’ a phrase used in a study by Albrecht & Devliegar (1999) to describe the interesting correlation between poor quality health and the development of adaptive coping strategies, which leads to good quality of life being reported. In the 2005 study by Beeton, adults with haemophilia said they had developed strength and resilience from the pain and isolation they had experienced. In addition, acceptance of haemophilia
as ‘an integral part of the self’ enabled them to find success vs. those who may suffer a sense of loss of ‘self’ due to the development of a chronic disease later in life.

Overall, there are very few studies that look at depression, anxiety and suicidal behaviour in individuals with haemophilia. However, it is an important issue to consider as one paper reports that suicidal ideation is high (Ghanizadeh, 2009).

2.4 Comparison with other chronic illnesses
A study by Wiedebusch et al (2007) reports that parents of children with haemophilia experienced a higher quality of life than parents with juvenile idiopathic arthritis and type 1 diabetes. The authors believe that this may be ascribed to improvements in medical therapy and specifically the use of prophylactic factor replacement. This study also reports that there are no differences in the quality of life between mothers and fathers of children with haemophilia; this appears to contradict previous studies that found mothers to be more depressed and anxious than fathers (Saviolo-Negrin et al, 1999)

Bullinger et al (2003), in a study covering six European countries, reported that children with haemophilia have a higher QoL than patients with asthma/atopic dermatitis and obesity when using the KINDL generic questionnaire. Again, the authors suggest that the high QoL is associated with state-of-the-art haemophilia care.
The literature agrees on the need for psychosocial support which begins either with the initial diagnosis or at the point of considering parenthood for couples with a family history of the disease. This need continues through childhood, adolescence and on into adulthood (Barlow et al, 2007; Beeton et al, 2007; Bossard et al, 2008; Bottos et al, 2007; Cassis, 2007; Ross, 2004).

However, with the exception of Cassis (2007), few authors suggest in detail what the psychosocial support should look like and specific guidelines are not referred to in the literature. However, it consistently mentions the ideal as being support offered by a multidisciplinary team, whilst also acknowledging that the ideal is seldom attained.

3.1 Multi-disciplinary team
Multidisciplinary teams are now a common feature in some areas of healthcare and bring a range of professionals and their complementary skills to bear on treatment and support. Such a team could include physicians (often with different specialist skills), specialist nurses, psychologists, physiotherapists, social workers and occupational therapists. They would therefore hold expertise in several areas identified in the literature as key to psychosocial support.

Currently, not all haemophilia treatment makes use of a multidisciplinary team, and so it is unlikely that individuals and their families will receive the full range of support identified in the literature. That said, where a multidisciplinary team is not available, healthcare professionals (HCPs) should have the knowledge to make referrals to all the other relevant parties who can offer patients and their families this support.

Key areas for psychosocial support:
1. Counselling
2. Communication
3. Support networks
4. Information and education
5. Participation in sports
6. Techniques to increase adherence to treatment

3.1.1 Counselling
With the exception of Cassis (2007), the literature around the benefits of counselling is limited. A number of authors, for example, Bottos et al (2007), have mentioned the positive change to or strengthening of the use of adaptive, problem solving coping strategies resulting from regular counselling programmes, where families are given information and the chance to share their experiences and feelings with other families with the disease.

In her 2007 paper Cassis sets out a comprehensive counselling and support system based on the developmental needs of the growing child with haemophilia which includes the participation of parents, family and other relevant individuals. She defines the specific needs of the child and family at different stages and gives ideas of how to meet these needs, whilst also empowering parents to try their own ways of meeting these needs.

The importance of good information and advice, understanding and empathy from HCPs involved from the start is also stressed by Cassis. This builds trust in the healthcare system for both parents and child, which is vital given that this is likely to be a long-term relationship.

Counselling adults allows them to express fully their emotions, fears and frustrations so as gradually to move them to a practical mindset that allows them to cope with the disease and parent the child in a
way that it develops good mental and physical health (Cassis, 2007; Beeton et al, 2007). Also, counselling young children to understand the benefits of their medical treatment so as to cope with their frustration will enable them to focus on their possibilities.

Counselling is also necessary for women who are potential carriers. According to Ross (2004) they are often left out of the equation and are in need of counselling support regarding the options open to them.

3.1.2 Communication  
A key finding from the review has been the need for guidance regarding how to communicate haemophilia to family and the wider social network. This is essential for emotional adjustment for family and individual.

A perceived lack of understanding of the disease by others and a fear of rejection often results in secrecy by parents or by the patient concerned, that can leave them socially isolated (Beeton et al, 2007; Cassis, 2007). Providing information and support to manage the condition will help feelings of control and confidence which in turn will make talking about the disease easier.

As mentioned previously, talking freely about haemophilia, knowing what to say and how to say it depending on the audience is commonly referred to in the literature (Barlow et al, 2007; Beeton et al 2007; Cassis, 2007). Therefore, equipping individuals and their families with information and support so they are comfortable speaking about the condition, if and when they choose to, is important for self-esteem and acceptance of the disease whilst also avoiding social isolation which can often result from secrecy and fear of rejection.

3.1.3 Support networks  
The benefit of sharing experiences and learning from others who are in the same situation by the formation of support groups for parents and individuals is mentioned in the literature. This can remove feelings of isolation and help give self-esteem back to the individual or their family.

Cassis states the importance of support groups from the diagnosis in order to help healthy adjustment for parents and children especially from age 7+ where they begin to develop a sense of self; pre adolescence and adolescence where they are pushing their limits and compliance issues are a problem; and for young adults where an increase in responsibilities may lead to depression, social isolation and sometimes drug and alcohol problems.

In a paper by Schooten & De Bruin (2008), where children spent 3 days in a camp with other children with haemophilia they reported that “kids explored their boundaries, gained confidence and knowledge, improved the relationship with healthcare staff and became less anxious about infusion therapy”.

Talking about emotions and experiences and hearing others expressing theirs will help provide a language around haemophilia for newly diagnosed patients and their families that can be empowering. This type of support may make speaking about haemophilia to others who may not always understand easier to do.

3.1.4 Information and education  
This is a very large area, encompassing the broadest possible audience – healthcare professionals, individuals with haemophilia, their families, friends, schoolmates, teachers and the population at large.

Various papers reveal the lack of knowledge amongst non-specialist HCPs, and identify a clear need for more education (Beeton et al, 2007), especially amongst hospital Accident and Emergency staff who need to be aware of the protocol regarding treatment of bleeds (Barlow et al, 2007). Teachers also need training in order to understand the needs of children with haemophilia and other chronic diseases, so that they can ensure the best experience for those individuals during their key school years (Nabors, Little, Akin-Little & Lobst, 2008).

Barlow et al (2007) suggests that the current lack of education in the workplace is a problem and that the general public also have little understanding of haemophilia. This is felt to impede relationships for those individuals with haemophilia.
“A public better informed about bleeding disorders will have beneficial effects on the psychosocial well-being of those with haemophilia.”

3.1.5 Participation in sports

Participation in sports is another area seen as beneficial by a number of authors. Von Mackenson, in her 2007 study, focuses on the positive effects of participation in sports or ‘sports therapy’ for individuals with haemophilia. When patients choose to participate in a sport they enjoy it tends to have a positive impact not just on physical well-being but also self-esteem, positive body image, building relationships with peers as well as less disability. This is echoed by other studies (Seuser et al., 2007; Wittmeier et al., 2007; Van CreveldKliniek et al., 2008).

Many of the above studies into sport as a psychosocial support to medical treatment do add the caveat that differences between developed countries in terms of ready access to treatment including prophylaxis should be considered in risk assessment of sports participation. However, in those countries where factor replacement is on-demand, physical activity is still recommended and can be monitored by a healthcare professional.

3.1.6 Techniques to increase adherence

Increasing adherence obviously has very positive benefits for children and individuals with haemophilia at any age. Including distractions such as objects or activities whilst treatment is taking place can help reduce the distress of a small child. Changing the established negative behavioural response to treatment via a counterconditioning process, which includes the use of topical anaesthetic creams, favourite DVDs, snacks and the use of circles sheets (which represent the stages of the process so the child can monitor their own progress), proved to be effective methods to increase treatment adherence as documented by Penica & Williams (2008).

Strategies recommended by Cassis (2007) for use in the early years in a child’s treatment are cited as: getting the caregiver to play the role of the patient receiving treatment and being brave while receiving an infusion; role playing in order to get an insight into the child’s feelings; and counselling parents to react calmly.

Interestingly, one study by De Moerloose et al. (2008), that spanned six European countries, reports that adherence to treatment is at its best at a younger age. In addition, “prophylactic treatment, time spent with a haemophilia treatment centre (HTC) and the quality of the relationship with the haematologist and nurse were also important factors for increased adherence”. In the same study, the reduction, fluctuation or disappearance of symptoms, forgetfulness, lack of time for treatment and convenience were factors quoted by the sample that impacted negatively on adherence, although these reasons differed according to age of the individual and by country. The study also reported the use of the internet and electronic patient diaries as helpful aids to adherence. Interestingly, the UK and Sweden had a dedicated multi-disciplinary care team supplied by the specialist HTC whereas in France, Germany and Italy a specialist team was available but not only for haemophilia treatment. In addition, while feedback was generally very good regarding the HTC, ‘treatment of the disease rather than the patient’ was one aspect where it fell down – suggesting that even within a multidisciplinary HTC context, psychosocial support is lacking.
4.0 Conclusion

A review of the literature shows that there is a clear need for more psychosocial support for individuals, their families and acquaintances. More research is necessary as to what this support should consist of although focusing on helping the individual and their family to cope, adjust and self-manage the condition, including complying with treatment is a priority.

Furthermore, some of the authors call for support to be carefully tailored to the specific socio-economic and cultural needs of the individual. International research, of the kind being undertaken in HERO, will help to clarify the experience of individuals with haemophilia and their families in different countries, in terms of medical treatment and psychosocial support currently available. Then the specific challenges of the different countries can be addressed.
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