

PSYCHOSOCIAL CARE FOR PEOPLE WITH HEMOPHILIA

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Psychosocial Care for People with Hemophilia

Frederica R.M.Y. Cassis

Introduction

Psychosocial support is an important part of comprehensive care for people with hemophilia. Beyond the medical condition, individuals with hemophilia commonly face a number of psychosocial challenges. Social workers, psychologists, and counsellors play a key role in helping new patients and their families adapt and learn to cope with their new reality.

It is possible to have hemophilia and still live a full life, even for those who face many social and economic challenges. However, medical treatment alone does not automatically translate into better quality of life. Beyond medical treatment, the responsibilities of the hemophilia treatment centre (HTC) include educating families about how to manage hemophilia and helping them find interventions for psychosocial issues.

The aim of this monograph is to shed light on some of the psychosocial issues encountered by people living with hemophilia, and guide healthcare professionals in the interventions and support that can be provided. It is intended for all professionals who may be involved in the support of patients from a psychosocial perspective. This may vary among treatment centres and countries.

Psychosocial aspect of comprehensive care

Psychologists, social workers, and counsellors have different expertise and strengths. In HTCs with the resources for a multidisciplinary team, there are individual staff members who specialize in these areas of expertise. However, in centres with limited resources, other healthcare professionals may be the ones to provide psychosocial support. Therefore, this monograph does not differentiate between psychologists, social workers, and counsellors, and HTC healthcare professionals who take on the psychosocial support role in their absence (often nurses).

Ideally, a hemophilia treatment centre should have a multidisciplinary team with a specialist for each aspect of care. While this is not always possible where

resources are scarce, there are still opportunities for psychosocial care by other healthcare providers. Regardless of the resources available, all HTC healthcare professionals should be aware of the psychosocial issues that may arise with hemophilia and other bleeding disorders. To provide optimal care, healthcare professionals need to be able to identify issues and challenges related to having a bleeding disorder that may be affecting their patients' cognitive and emotional development.

A bleeding disorder is a chronic condition that imposes limitations, but it can also represent an opportunity to bring about positive change through learning and self-awareness. There are different ways of experiencing and coping with pain and chronic illness. Some people with hemophilia may focus on the emotional challenges, while others find ways to better cope with their situation. Short-term psychotherapy, alternative therapies, and social services can help individuals with hemophilia cope with symptoms and limitations and develop a healthy sense of self.

The role of psychosocial support

Psychosocial support in the medical setting helps individuals gain a personal understanding of hemophilia. Healthcare professionals provide psychosocial support by helping patients and their families develop strategies to cope with physical, mental, emotional, and social challenges related to having a bleeding disorder. This includes providing education, individual and family counselling, resources, and community referral services. The ultimate goal is to empower people affected by hemophilia and other bleeding disorders to manage their circumstances and challenges autonomously. Quality of life depends largely on the ability to adjust to having a chronic disorder and the challenging circumstances that may arise.

In psychosocial care, it is as important for healthcare professionals to learn from their patients as it is for them to provide guidance. Cultural values and socio-economic factors affect how psychosocial issues are experienced and addressed. Other important issues for people with bleeding disorders include accessibility

of the HTC, availability of factor replacement products and prophylactic therapy, access to physiotherapy and corrective surgery, and support from hemophilia associations.

Support for different stages of psychosocial development

Psychosocial functioning and well-being are often specific to age and cognitive development. Issues and stressors arise at different stages. These stages can be classified as:

- Infant to toddler (newborn to age 5);
- Early childhood (6-9 years);
- Pre-adolescence (10-13 years);
- Adolescence (14-17 years); and
- Adulthood (18 years and over).

These age groupings, similar to those in child development literature, are based on the author's clinical experience in guiding families affected by hemophilia through the various phases of child development.

Infants and toddlers (birth to age 5)

Two key issues that often arise early on with hemophilia are coping with the diagnosis and balancing vigilance and overprotectiveness.

Helping families deal with diagnosis

Couples who know of hemophilia in their family history can make an informed decision about whether or not to have a child that may inherit the hemophilia genetic mutation. In some countries, when there is a family history of hemophilia, genetic counselling is available to evaluate family medical records, determine whether testing is advisable, and help prospective parents understand test results and their options. Psychosocial support is especially important for parents who decide to go ahead with reproduction. However, in about one-third of hemophilia cases, there is no previous family history.

A diagnosis of hemophilia can cause emotions ranging from acceptance to denial, confusion, anger, guilt, and fear for the future. These feelings can complicate or contradict the joy of the baby's arrival. The sooner these issues are confronted, the easier the adjustment will be—this is why the first years of interaction with the HTC are so important for families. For parents,

the HTC should be a place where trust is built, and reliability and mutual education are assured.

The diagnosis of hemophilia can be traumatic because the idea of a bleeding disorder raises anxieties about danger and death. Initial denial, anger, and sadness following a hemophilia diagnosis are well documented. It is important to help families put it into perspective.

Parents may sometimes feel guilt at having passed on hemophilia to their child; disappointment and anger that dreams for the child may not be fulfilled; anxiety over access to treatment or cost of treatment; concern about venous access for the delivery of factor replacement; anxiety about family disruption and sibling rivalry over the attention spent on the child with hemophilia; and fears about treatment and care. It is important to remember, however, that these feelings are likely accompanied by positive feelings such as the joy of having a new baby and the unique experience of being part of that child's emotional and physical development.

It is important to watch for signs of difficult adjustment, such as a parent rejecting or distancing oneself from the child, blaming the other (female) parent, shame, or intense conflict in family. With time and support from healthcare professionals, there can be acceptance.

It is also important for healthcare professionals to look out for persistent anxiety or signs of depression that may affect day-to-day life and family relationships, and provide appropriate psychosocial support.

Case study: Psychosocial support following diagnosis

A little boy, aged 3, was brought to see me in my small office at the hemophilia treatment centre in Sao Paulo, Brazil. His mother confessed that whenever they brought their son to the HTC either to be evaluated or for treatment, they felt frustrated and defeated by the realities of their son's hemophilia. This was their first session with a psychologist.

Building trust

Diagnosis is often first made at a hospital, in the emergency room setting. The interactions in this setting shape the parents' and child's initial impressions of the healthcare system and the treatment and care available for hemophilia. The toddler's diagnosis of severe hemophilia B was a very traumatic experience

for the family. Doctors had taken a very long time to diagnose the bleeding disorder. With no known family history of hemophilia, the parents had to endure questions about domestic violence and child abuse.

Not knowing what is wrong with one's child can be terribly hard for parents. Following diagnosis, it is important to help patients and their caregivers understand the services available at HTC. The role of HTC professionals is to provide information and support, and sometimes to help restore a sense of balance to family dynamics affected by hemophilia. This includes building awareness of the range of medical and psychosocial services and support available early on, and cultivating trust in the HTC.

Counselling parents

Although the child was the primary patient, in the early sessions I focused on the parents' emotional welfare and coping abilities. The mother needed to express her disappointment and frustration and learn to accept her son's chronic disorder. The father showed great interest in learning about hemophilia. His need was to gain control by understanding the disorder and taking an active role, while her need was to work through emotions that made her feel helpless. It is important to help parents understand that the diagnosis of hemophilia represents a new life challenge.

Counselling children

After addressing the parents' anxieties, I began to work with my little patient. The focus was his fear of treatment, especially of getting injections. This involved helping him understand basic facts about hemophilia and addressing his fear of pain. With experience, someone with hemophilia learns to know when a bleed starts, how to treat it, and when it begins to heal. It is important to help patients overcome the psychological barriers to accepting treatment.

After a year of weekly sessions, my young patient was at ease coming to the HTC. He has come to understand that infusion may hurt, but helps him heal better and faster. The child's mother has learned how to administer factor to her son and now does so at the centre. Eventually, the child will be able to enroll in a program on self-infusion for four- and five-year-old children.

Family dynamics

Hemophilia affects not just the individual but also the whole family. Siblings should be included in counselling

sessions and given a basic understanding of hemophilia. Have siblings express their feelings about their role and importance in the family. Answer their questions honestly and encourage parents to do the same.

In families with a known history of hemophilia, it is sometimes easier for parents to deal with the diagnosis. But there can also be challenges, such as generational differences and new developments in the understanding of hemophilia and its management. Past generations of children with hemophilia who did not have access to factor therapy were discouraged or prevented from taking part in sports or strenuous activities, and were often overprotected and somewhat isolated. Medical advances such as prophylaxis therapy and the availability of factor concentrates have allowed people with hemophilia to be more active and live more full and healthy lives.

Support networks

Encourage parents to join support groups or hemophilia organizations where they can talk to other parents affected by hemophilia about their concerns, and where children can meet others who have bleeding disorders.

Vigilance vs. overprotection

Caution and injury prevention are important for people with hemophilia. However, it is important to distinguish between prevention and overprotection.

Healthcare professionals can help parents and children learn the difference between normal and unnecessary risks. Parents should be counselled to encourage normal activities—certain risks must be taken for healthy development to take place. If toddlers are not allowed to crawl, walk, or run normally, they will become insecure and fearful and possibly even more prone to injury because of lack of experience. Inactivity and overprotection can often bring on issues such as isolation due to poor social interaction and weight problems due to lack of exercise. The healthcare professional can guide parents on how to balance giving their child room to discover minor risk while also setting limitations.

Case study: Overprotection

Not long ago, I met a single mother in her early 20s whose first child had been diagnosed with severe hemophilia A. The nurse at the centre had detected the mother's lack of knowledge about hemophilia and considerable anxiety about how to keep her son safe.

At our first session, she held her one-year-old baby close in her arms. Every time he tried to move around, touch an object, or venture even slightly beyond his range, his mother would automatically hold him firmer and tighter. She was clearly afraid of any new movements that he made. The child would usually stop moving at his mother's reaction, but would then resume the behaviour of typically curious children.

Body language

The mother's reaction to try to protect her son from bumping his head, falling, or injuring himself was instinctive and natural. However, such constantly restricting behaviour can be harmful.

Through body language, parents can unknowingly convey to the child a sense of what he can or cannot do. However, it is important to keep in mind that learning occurs in stages. For example, learning how to go downstairs, a child will begin by sliding or bumping down the stairs on his bum. When he begins to walk, he can hold onto his parents' hands or the banister until he has mastered the movement on his own. If a parent routinely forbids movement, the child will have a hard time learning these skills and understanding the restrictions on his impulses and curiosity. Or if movement is always forbidden, the child can become very dependent on his caregiver or overly worried about being cautious.

The health professional can help parents assess when they are being over-protective of the child and correct the behaviour. The goal is to help parents:

- Recognize their fears and discuss them;
- Be aware that parental influence on their children eventually diminishes and that the parents' role is to guide their child towards independence; and
- Understand that overprotection may hinder the child's emotional, social, and physical development.

In this case, I counselled the mother for six months and used role-playing techniques to draw out her feelings and anxieties and how she was expressing them. When she could see how others perceived her, she was able to perceive herself better. Empathy about the challenges of hemophilia also helped her strengthen her self-esteem and confidence as a mother. Different approaches will work for different individuals.

Psychosocial staff can help parents develop an appropriate balance between giving their children autonomy and putting some limits on their impulses. It helps to identify issues and goals in advance, and for parents to practice being firm, sensible, and patient. For example: "Today you have played a lot and exercised your joints and muscles. Now your body needs to rest and recover. You can watch some TV, read, or paint, or we can do something together like cooking or gardening."

Strategies for the early years

- Help parents come to accept the diagnosis and understand that hemophilia is just one aspect of an individual's life.
- Counsel parents to react calmly. This will set a healthy pattern of response, and will calm the child's fears and those of other family members. Overreaction to a bleed can cause the child to feel guilt and distress harmful to developing a healthy self-image.
- Help parents move beyond emotional reactions to a practical mindset.
- Encourage parents to develop confidence and coping strategies by learning as much as possible about the bleeding disorder, and about the resources and HTC healthcare professionals available to help families manage.
- Use flashcards with pictures representing situations related to hemophilia (getting hurt, bleeding, receiving treatment, etc.) to explain the disorder.
- Use role-playing to draw out the child's feelings. By playing and mixing up different kinds of emotions, the patient can work out his concerns and develop a sense of mastery of his condition. Activities and stories can be tailored to the child's interests.
- To get the child to lose the fear of being touched, the caregiver can play the role of the one being probed and being brave while receiving an infusion.
- Compare different kinds of pain, such as an injection to an insect bite.

Guiding parents on sensible protection

Protective measures and devices (such as elbow or knee padding) help protect joints from injury and bleeds. At the same time, a little boy must be able to feel his body, learn to trust his sensations, and learn how to manage his movements in the best way. Balance is key: in some situations, it can be a good idea to protect the joints to allow for further play while protecting against specific risks. For example, a family who lives in a home with concrete floors may be advised to protect the toddler's knees with padding when he crawls or to put a carpet on the ground. Parents will have to make their own decisions about whether environments need to be adapted and whether to use padding, weighing protection against the child's developmental and learning processes. It is important to keep in mind that a child needs to be protected but at the same time, he needs to learn gradually how to become independent.

Objectives for the first years

In the first years it is very important for health professionals to:

- Provide the family with basic information about hemophilia, familiarize them with the services at the HTC, and make sure that they know first-aid measures for treating a bleed.
- Help the family adapt to the fact that hemophilia is a life-long chronic condition.
- Guide the development of the child's confidence in basic activities such as crawling, walking, running, dancing, playing, etc.

Early childhood (ages 6 to 9)

The early childhood years are important for the development of cognitive skills, social competence, and emotional health. At around the age of six years old, children begin to read and write and are eager to learn and master new information. They begin to gain understanding of themselves within the social context. Gradually, patient education and counselling can be directed more toward the child. The key focuses during early childhood are cognitive development and socialization.

Cognitive development

Social interactions are important to children's learning and cognitive development. At this stage, they develop important social skills through relationships with peers and family. They also gain a sense of mastery and self-concept (understanding of who they are). It is important to provide a supportive environment as they begin to develop self-confidence and self-esteem.

Guiding parents on teaching children

Psychosocial staff can help parents develop techniques to answer difficult questions and encourage their children to stay positive. A key message to emphasize is that even when they are receiving treatment for a bleed they are still "like everyone else" in other ways—for example, in enjoying recreational activities or having to study or earn a living.

Helping children accept hemophilia

Children with hemophilia do not want to be seen as different and want to be treated as normal. At the same time, they need to understand that sometimes they will be patients, such as when there is a bleed and need treatment or rest to allow injuries to heal. It is important that as patients, children continue to learn about their hemophilia and their bodies and come to accept that they have a chronic life condition.

It is important for healthcare professionals to continually re-evaluate the extent of the patient's knowledge. Ask patients to review what they have learned. This method, called "teach-back," helps assess the effectiveness of their teaching techniques and ensure that the material is well understood.

Storytelling and imagery

Images and visual aids are very effective tools for teaching children about how blood functions and the clotting process. Storytelling, imagery, and comparisons to familiar concepts are also effective. For example, the clotting process can be explained as cells plugging a hole in a blood vessel, similar to plugging a sink to stop water. Another effective way to explain the coagulation process to children is to use a turtle to represent very slow healing, and a "turbo turtle" whose healing is accelerated following treatment.

Ultimately, it is important that parents understand that children with hemophilia go through the same normal emotional and cognitive development as other

children. Indeed, hemophilia also involves an ongoing developmental process for parents and families.

Socialization

Early socialization is important for children with chronic illnesses, who may encounter barriers to socialization later on (e.g., missed school, feeling different, etc.) Early social interactions help them develop self-confidence and a sense of belonging, and learn how to interact with others. Parents should be encouraged to take advantage of early opportunities for socialization, such as playing or sleeping over with friends and participating in community events for children and special activities at school.

Nonetheless, parents of a child with hemophilia can have some apprehensions as the school years begin. At the treatment centre in Sao Paulo, the pediatrician and nurse provide parents with an information manual about hemophilia and a letter for the child's teacher outlining key points such as:

- How the school can provide support to ensure that the child can engage in normal activities;
- Appropriate activities for children with hemophilia (this will depend on individual circumstances);
- What to do when a bleed occurs and who to contact;
- Risky activities and vulnerable areas of the body (e.g., head, abdomen).

Other interventions can include family members or HTC staff meeting with school staff and teachers, and making classroom presentations.

Informing schools about hemophilia

Whether and when to disclose a bleeding disorder, as well as how much to divulge, is something that the family will have to manage. In early childhood, the child's exposure to new physical activities and potential risks at school makes it prudent to advise the school about hemophilia and bleeding tendencies. As the child becomes older, he/she may choose not to disclose to all, while some situations require different decisions. The approach and degree of information to convey can be reconsidered each year. It is not necessary for children with hemophilia or their teachers to make a special point of telling the class about their hemophilia every year. However, it is certainly important for children with hemophilia to be able to explain their bruises or health factors that may cause them to miss school occasionally.

Families and school officials need to work out simple and practical solutions for schoolchildren with hemophilia. A "buddy system" with a classmate who lives nearby helps those who have to miss school get the lessons needed and keep up by doing assignments at home. It is important that the children remain in contact with their school so that they feel that they belong and are members of their class even when away. Flexibility helps – it is important to understand that children who miss school due bleeding episodes may not be able to complete all the assignments covered in full school days.

Case study: Building social confidence

The idea of starting school and learning how to read and write was very exciting for one of my young patients with moderate hemophilia A, aged 7. His parents were also very happy, but at the same time his mother worried about leaving him alone with all those other big boys. My patient himself was a little overweight from overprotection and lack of exercise – in fact, when I asked him for a self-portrait, he drew a whale. He had difficulty interacting with his peers, oscillating between bullying others or being teased about his physique. Boys with joint or muscle problems often face similar challenges.

Each child comes with individual challenges – the psychosocial worker's goal is to facilitate the child's overall competence and ability to deal with having hemophilia. My patient had a unique sense of humour and was also talented with games, computers, and drawing. I determined that working on these qualities would help him cope better with his body image and reduce his anxiety. At the same time, he needed to be encouraged to exercise to be more physically fit.

I also introduced him to other boys at the hospital, all his age and with hemophilia. The interaction and learning about other experiences with hemophilia helped him gain confidence in his capacities while also recognizing his limitations. Group therapy involving others with hemophilia is an effective approach for counselling children in this age group.

Objectives for early childhood

Basic Information

- Encourage parents to provide basic information about hemophilia to children but not overwhelm them with too much at a time.
- Encourage parents to be guided by their child's questions and use learning opportunities that arise naturally.
- Provide parents with information and educational materials and put them in touch with reliable resources and support groups.
- Remind parents that gaining knowledge about the medical, technical, and emotional aspects of hemophilia is a very gradual learning process.
- Provide parents with telephone numbers of HTC's, doctors, nurses, and ensure that the parents have a list with emergency phone numbers, and family members and friends who provide support.
- Facilitate discussions with school officials about the child's hemophilia.

Socialization

- Encourage parents to create opportunities for family and social interactions, which are important for building the child's self-confidence and sense of belonging.

Psychosocial Support for Parents

- Help parents sort out their reactions, emotions, and thoughts about hemophilia and its effects on the family.

Childhood or pre-adolescence (ages 10 to 13)

One of the goals during the pre-adolescence stage is to teach children how to manage their hemophilia. During this stage, children gradually gain confidence in their treatment, their bodies, and their own ability to manage their condition.

Beginning to self-manage

It is important to encourage children to take an active role in the treatment and management of their health. Doctors or nurses will not always know when patients are ready for self-management. A cognitive and

psychological assessment helps determine whether the child is ready to start taking control.

A "semi-directed" interview (alternating basic questions with questions that pick up on interesting comments by the patient) can detect problems and difficulties the child is having and capacity to take responsibility. However, psychological assessments are not commonly available, so treatment staff often need to assess readiness informally, using rough age guidelines and observation of the child's readiness for responsibility.

Peer identity

Pre-adolescence is a complex phase, with or without hemophilia. During this stage, peer identity becomes significant. Fitting in with peer groups and not appearing different from others become very important, as at this age children do not want to be excluded from social groups or stigmatized.

Body image

Body image gains importance during pre-adolescence. Aside from changes in voice, weight, and height as puberty approaches, pre-adolescence is also when children grow most rapidly and these growth spurts include changes in joints and muscles.

Physical or functional limitations can make young children feel shy and embarrassed or cause them to be teased by others. Pre-adolescence is a stage when boys and girls can come up with highly creative but sometimes hurtful nicknames, and children with hemophilia, like any others, may be called names.

Psychosocial professionals can help children learn to view their condition with some levity and be better prepared to handle teasing. Children cannot always avoid meeting people who tease or bully, but they can be taught how to respond to provocation and not to be afraid or intimidated by the bully. At the same time, it is important that they learn how to be firm about not getting into physical fights. It is critical that children with hemophilia learn how to take care of themselves and not put themselves at risk for bleeds.

The role of the psychosocial worker during this stage is to help patients deal with their feelings about their physical changes and learn to start taking responsibility for their own health. Remind children that despite their hemophilia and the associated physical health problems, they are full human beings

who just happen to have a bleeding disorder. Still, they must accept that some parts of their body may not work as well as others, which can be hard to accept. Psychosocial support is important to help children with hemophilia look beyond their physical limitations and identify with their whole being, as an individual with much to learn from and contribute to society.

Setting boundaries

During pre-adolescence, children begin to be less inclined to listen to their parents about their care, and also start to test their limits. The parents, child, and healthcare professional must work together to identify activities and the degree of liberties and limitations the child can handle, taking into consideration level of maturity and psychological readiness and benefits.

Just as in the general population, some children with hemophilia will be more outgoing than others; for these children, it is harder to deal with physical limitations. However, during this phase, parents must be able to set limits for their children because pre-adolescents have not yet learned how to maturely handle their impulses and desires. There will be rebellion and disagreements with parents as children seek to gain independence and their parents' trust. Encourage parents to give their children space to grow, while being firm about boundaries.

Case study: Hiding injuries to avoid stress

One of my patients with severe hemophilia A, aged 12, is the eldest of three brothers. The middle brother also has hemophilia but the youngest does not. Recent years were very difficult for the family due to financial challenges. The father was unemployed and the mother earned a very low salary as a maid. Living space had become very cramped with the birth of the third child and the arrival of an aunt from another province to help take care of the children.

I had worked with the eldest child when he was seven years old. He had always shown great interest in learning about hemophilia, and activities such as playing, drawing, and painting. However, after two years of coming to the walk-in clinic, the family stopped because of financial problems. This kind of interruption occurs frequently – distance, money, time, and transportation are all issues that can limit counselling interactions.

Recently though I received a call from the HTC nurse, who told me that the child and his mother had had a heated argument because he had not told anyone about a serious elbow injury for days. The bleeding into the joint of his left elbow resulted in severe joint limitation, and the right elbow was headed in the same direction. His mother was having a hard time with the conflicts they had over his care.

I met with the child again and gradually he told me about his frustration over not having a quiet place to study or rest, away from the shouting and teasing of his little brothers. He also felt guilty when he had a bleed because of the difficulties they created for his family in terms of time away from work, extra expenses to travel for treatment, etc. He had an underlying fear of causing financial and emotional stress to his parents. Furthermore, he had a high pain tolerance, allowing him to endure pain for days without anyone taking notice of discomfort. His reactions were clearly related to the day-to-day difficulties his family was experiencing: the stress, poverty, and struggle for survival. It was important to empathize with the parents, yet also convey that their feelings of powerlessness were affecting their son's attitude towards his hemophilia care.

It is important for parents to know that children may try to hide information to avoid getting into trouble. The child must be made to understand that going to the hospital is not a punishment, even if it sometimes provokes stress. When parents show stress or fear, it is important that they make clear that these emotions are not the child's fault. Such conflicts are normal and signs of change and development.

Objectives for pre-adolescence

Self-Confidence and Responsibility

- Encourage children to take an active role in the management of their bleeding disorder.
- Encourage children to take responsibility for their health and avoid dangerous situations and risks.
- Help children deal with their feelings about physical changes and health issues.
- Identify degree of liberties and limitations for children, taking into consideration level of maturity and psychological readiness.

Adolescence (ages 14 to 17)

Perceptions of age and what is normal at each stage can vary in different cultures. In some societies children reach adulthood quickly, marrying young or working from a young age to help support their families.

Adolescence quickly gives way to adulthood, which can result in limited educational opportunities. In other societies, there are very distinct pre-teen and teenage stages, marked by rites of passage that vary greatly from culture to culture.

Maturation and independence

Depending on the societal, economic, or cultural context, adolescents will mature in different ways and at different rates. For example, adolescent boys who have to be the main breadwinner due to cultural or socio-economic circumstances may not have the time or resources to properly treat their bleeds. In addition, they may not want employers to know about their hemophilia for fear of being seen as problematic or losing their jobs.

Compliance issues

Adolescents may have a tendency to ignore or dismiss bleeding episodes. At this stage developmentally, they are often focused on the present moment and may not consider future consequences. Treatment and therapy can interfere with activities, which can provoke resentment of the bleeding disorder and lead to non-compliance issues. Adolescents may also be non-compliant about physiotherapy or exercises if it takes them away from their favourite activities. Psychosocial professionals can help patients understand the importance of physiotherapy and taking care of their own bodies. They can also help patients understand and overcome their resentment of the therapy and tasks related to their bleeding disorder.

Transition of care

During adolescence, youths gradually move from the care of pediatricians to general practitioners. It is important at this stage to caringly encourage autonomy, self-management, and independence.

Education and career decisions

In the mid- to late-adolescent years, youths begin facing decisions about schooling and job preparation. During this stage, they will need guidance to make choices about academic programs and vocational pursuits that will satisfy their goals, make good use

of their talents—and not involve risky situations leading injuries or bleeds. Explore various career options with your patient and realistically assess the possible physical dangers that accompany each one.

Dating

Dating begins during the adolescent years. Youths with hemophilia and other bleeding disorders may be hesitant about dating because of self-esteem issues raised by having a chronic condition and the need to decide whether or not to disclose their hemophilia. Another challenge is that their date may not understand some of their limitations. For example, they may resent plans being cancelled at the last minute due to a medical emergency. Psychosocial professionals can help individuals develop strategies to help their dates understand hemophilia, such as bringing the date along to the centre for a visit.

HIV or HCV disclosure

Until the introduction of viral inactivation techniques for blood products, infection with HIV/AIDS or hepatitis C (HCV) via blood product transfusion was widespread. In countries where viral inactivation is not available, transfusion infection remains a substantial risk. People with hemophilia who also have HIV and/or HCV often benefit from psychosocial support to help deal with the social stigma. Psychosocial staff can also provide important support and coping skills for both the physical and mental challenges of having HIV and/or HCV. This includes sexual education, preventive safety measures, and genetic counselling, taking into consideration the individual's cultural and/or religious background.

Case study: Disclosure when dating

A patient of mine with hemophilia and HIV was going through difficulties because of a fear of relating to women. He felt that he did not have the right to fall in love or encourage love and affection from women. He was bitter, pessimistic, and sarcastic most of the time—attitudes, in fact, that would make it difficult for anybody to meet people and make friends.

Counselling provided a non-judgmental venue where he learned to accept his condition in a more constructive way. A major issue was his fear of rejection. We talked about how it is important to get to know the person that one is dating before opening

up about having HIV. Gradually, he learned that intimacy is something built slowly through effort and communication. Our sessions helped him talk about sensitive health issues and practice sharing the information with others. He also learned that he does not have to embark on a serious relationship right away – other kinds and levels of intimacy in relationships are still valuable.

Objectives for adolescence

Self-management and growing autonomy

- Emphasize to patients the importance of taking responsibility for their health.
- Encourage gradual autonomy, self-management in treatment and care, and independence.
- Guide adolescents towards academic and vocational pursuits that do not involve significant physical risk.
- Help patients through common adolescent challenges such as self-esteem issues and dating.

Transition to adulthood (18+)

During the transition to adulthood, life often becomes more complicated and people may face challenges on several fronts: work, interpersonal relationships, and financial stability.

Challenges for young adults

Cost of treatment is a major issue for patients entering adulthood who may no longer be covered by government health plans or their parents' medical insurance plans. Another issue is social isolation and depression, which often arise with having a chronic condition accompanied by pain and limited mobility. Alcohol and drug consumption are other risks. Specific therapeutic interventions may be needed.

Job interviews and disclosure

Everyone knows how important it is to convey a positive self-image at a job interview. Individuals with chronic medical conditions face additional issues, including disclosure. Different countries have different rules about disclosure. It is important that individuals be made aware of their civil rights and liberties. Some regions and countries have programs

or legislation that give workplace protection to people with physical disabilities and/or chronic illnesses, including people with hemophilia.

The issue of whether or not to disclose hemophilia or another bleeding disorder at a job interview raises a number of considerations. Hemophilia is not contagious and puts no colleagues at risk. An individual who has moderate or mild hemophilia and almost never has to go to a centre for treatment may choose to disclose the hemophilia while emphasizing that it does not affect job performance or attendance.

Disclosure could be necessary if:

- There may be frequent absences because of bleeding episodes.
- An individual is on prophylaxis and has to go to the hospital regularly for infusion.
- The job involves activities that could be risky for people with severe bleeding disorders.

In such cases, the job candidate can propose solutions such as working from home on days when there are bleeding episodes, sharing duties with co-workers, etc.

Workplace issues

Adults with chronic illnesses may face a number of issues in the workplace: medical absenteeism; insurance coverage and compensation challenges for work-related injuries; and possible limitations for advancement and promotion.

A multidisciplinary approach to helping patients manage workplace issues is ideal where possible. A social worker can help prepare young adults for the job market by directing them to training courses and funding for scholarships. A psychologist can help individuals explore different career paths through discussions about feasible job options and potential difficulties that may arise. A guidance counsellor can perform vocational tests and interviews. Where resources are limited, one healthcare professional may have to span these responsibilities.

Marriage and reproductive issues

Couples may have serious concerns about having children if a family history of hemophilia is known. While genetic counselling and antenatal diagnostic tests are available, they are not universally accessible or affordable. The decision to have children is a

family one. The HTC's role is to provide the range of information and support necessary to help the family make informed decisions.

Social networks

The importance of personal support networks in young adulthood cannot be overstated. Adulthood brings more and more responsibilities, including new challenges in the workplace, mobility problems if joint disease or muscle damage is an issue, and possible self-esteem issues. Support from family, friends, and psychosocial professionals are crucial to helping individuals face such challenges. HTC professionals can refer young adults to groups for adults with hemophilia or encourage the development of a new group.

Case study: Managing chronic illness and work

One of my adult patients, aged 40, has severe hemophilia A. In the 1980s, at age 17, he was diagnosed with HIV. At the time he thought he would die, but he has since survived many phases of the epidemic and its treatment.

My patient had originally come to see me for counselling because he had also developed HCV and felt he did not have the strength to undergo any more treatment cycles for HCV. The diagnosis of HCV a year earlier had been a shock – discovering that he could not tolerate the side effects of treatment was a second distressing surprise.

The patient was taking anti-depressants but medication alone was not enough. He wanted to review other aspects of his life, such as relationships with people at home and at work, and also try to better understand his perceptions about life and death, spirituality, etc. Our counselling sessions were a safe and comfortable place where he could explore these issues and develop ways to find solutions to his challenges.

When working with people living with HIV/AIDS, psychosocial professionals should be prepared to talk about very difficult and sometimes taboo topics such as death, faith, rejection, and rage. It is important that patients with HIV and/or HCV not regard themselves as victims. (At the same time, it should be anticipated that such thoughts might occur at times.) The focus of counselling is on psychosocial welfare and helping patients pursue normal and fulfilling activities.

Objectives for transition to adulthood

Independence

- Guide patients as they make the transition into adulthood and the workplace.
- Provide couples with information on the genetics of hemophilia and support to help them make informed decisions.
- Encourage patients to participate in social or support groups for people with hemophilia.

Socio-economic issues in some regions

In many countries there are not yet reliable tests for establishing a diagnosis or mechanisms for identifying and registering people with hemophilia. The absence of tools and data makes the treatment of hemophilia in some parts of the world a challenge. Even in regions where diagnosis is available, resources may be limited.

Lack of knowledge and information

Basic patient education is a major challenge in some parts of the world. In areas where there is no universal education and illiteracy is high, knowledge and understanding of hemophilia may be very minimal. Even doctors may not be trained in hemophilia care. In some countries, learning materials may be scarce or outdated, or treatment centres may be too far for patients to visit regularly for treatment and follow-up.

Remoteness

Some people with hemophilia must travel long distances for medical care. Sometimes it can be almost impossible to offer help other than lobbying governments to provide ambulance service. Often there is neither access to medication nor even ice available to treat pain.

Social stigmatization


In some cultures, people with hereditary bleeding disorders may be stigmatized or face difficulty being accepted as valuable and capable members of society. Social isolation can be detrimental to their social functioning, psychological well-being, and quality of life.

Early entry into workforce

In some societies, children as young as ages 8 to 12 already begin to work. Youths may be forced to engage in heavy manual labour or unsafe working conditions, where injury and serious bleeding are more likely. Issues associated with early entry into the workforce include increased dangers on the job, no time to treat bleeds during work hours, or job loss because of frequent bleeding episodes and absenteeism.

Conclusion

People with bleeding disorders live everywhere around the world. It is necessary to always consider the treatment approach in the context of the characteristics of each society and the resources available. Health professionals at hemophilia centres play an important and at times critical role in patients' lives, imparting key tools to help them improve their lives and actively participate in the treatment and management of their hemophilia, and ultimately lead interesting and productive lives.

Hemophilia and other chronic illnesses affect each person beyond the physical problems the condition can cause. In order to optimize their efforts to facilitate the health of their patients, it is important for treatment centres to provide psychosocial care as part of an integrated multidisciplinary approach. 

References

- Arranz, P et al. El apoyo emocional en hemofilia. Industrias Graficas Sprint, 1999.
- Bach, S. Images, Meanings and Connections. Daimon, 1999.
- Baer, E. Semiotics and Medicine. The Semiotic Web. Mouton de Gruyter, 1988; 215-267.
- Berger, M. The Missing Factor. Chipmunkpublishing, 2005.
- Byington, CA. Pedagogia Simbolica. Ed. Rosa dos Ventos, 1996.
- Cassis, F. Flowing and Clotting: A Biosemiotic and Hypermediatic Study of Hemophilia. Master Dissertation in Communication and Semiotics, Pontificia Catholic University, 2002.
- Dolto, F. La cause des enfants. Laffont, 1987.
- Dolto, F. La cause des adolescents. Laffont, 1992.
- Dolto, F. Les images, les mots, le corps. Ed. Gallimard, 2003.
- Hillman, J. O livro do puer. Paulus Ed., 1998.
- Jones, P. Growing Up with Hemophilia: Four Articles on Childhood. Oxford University Press, 2002.
- Kelley, L. Don't be victimized by hemophilia: How to be a more responsible, effective parent. Alpha Therapeutic Corporation, 1994.
- Kelley, L. Raising a child with hemophilia. Armour Pharmaceuticals, 1993.
- Kuriyama, S. The expressiveness of the body and the divergence of Greek and Chinese medicine. Zone Books, 1999.
- Nilsson, IM. Hemophilia. Pharmacia Plasma Products, 1994.
- Ramos, D. A psique do corpo – uma compreensao simbolica da doenca. Cultrix, 1994.
- Theron, A et al. Psicologia Evolutiva. Ediciones Piramide S.A., 1998.
- Whitmont, EC. The symbolic quest – basic concepts of analytical psychology. Princeton University Press, 1969.