Hemophilia, AIDS, and Occupational Therapy

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This article describes the blood clotting disorder of hemophilia, including its transmission, incidence, and physical and psychosocial effects. The epidemiology of persons with both hemophilia and HIV are discussed, as well as HIV's rapid spread due to contaminated blood products and the mechanisms commonly used to cope with HIV infection.

Specific suggestions on occupational therapy intervention are provided, including helping clients learn to cope with HIV by transferring skills previously learned for coping with hemophilia, teaching stress reduction, anger management, communication, and energy conservation skills; educating families and helping them set appropriate goals; designing adaptive systems for dealing with neurological deficits; and designing exercise programs.

HIV Infection

The incidence of HIV infection among persons with hemophilia is difficult to determine, due to this population's reluctance to be tested for the presence of antibodies. Nonetheless, the Centers for Disease Control estimates that up to 92% of those with hemophilia A and up to 52% of those with hemophilia B have been infected with HIV (Stehr-Green, Holman, Jason, & Evatt, 1988).

HIV infection can be represented in four stages:

- Stage 1—Infection with the virus.
- Stage 2—Seroconversion (antibodies to the virus are produced and are present in the blood) and asymptomatic reduction of immunity.
- Stage 3—More severe reduction of immunity and lesser opportunistic infections or the cluster of AIDS-related-complex symptoms.
- Stage 4—More severe opportunistic infections, wasting syndrome, or AIDS.

Treatment of Hemophilia

In persons with hemophilia, most of the major bleeding occurs internally, within muscles, joints, and the brain or other organs. Little cuts do not cause lethal bleeding. Bleeding may be caused by overuse of muscles or joints or by physical impact or may occur spontaneously, for no apparent reason (Salk, Hilgartner, & Granich, 1972). Bleeding into joints causes destruction of articular cartilage (Gilbert, 1975) and

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ults and adolescents with hemophilia and other blood clotting disorders constitute approximately 1% of the population of adults and adolescents with AIDS, and children with hemophilia constitute 6% of the population of children with AIDS (Centers for Disease Control, 1988). Infection with HIV has severely affected the population of approximately 20,000 Americans with hemophilia.

The two major types of hemophilia, hemophilia A and hemophilia B, result from a deficiency in the production of 2 of the 15 factors needed to clot the blood. Hemophilia A, also called classical hemophilia, is a Factor VIII deficiency and is the most common form of the disease. Hemophilia B, also called Christmas disease, is a Factor IX deficiency. Other varieties of hemophilia exist, but they are rare (Eckert, 1983). Most persons with hemophilia are men. Hemophilia is transmitted by a recessive sex-linked gene carried by the mother. Approximately one third of all hemophilia cases result from a spontaneous genetic mutation (Simon, 1984).

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and leads to orthopedic complications such as arthritis and joint fusion. Eighty percent of persons with hemophilia are estimated to have some form of orthopedic problem (Boone & Spence, 1976).

Until the 1960s, little could be done to stop severe bleeding. As late as 1972, the average age of persons with hemophilia was 11.5 years (Levine, 1985). A breakthrough in the treatment of hemophilia came with the development of freeze-dried (lyophilized) factor, which gained widespread use in the early 1970s. This factor is made in large batches, with each batch containing the blood of 2,500 to 25,000 donors (Levine, 1985). An adult with severe hemophilia, therefore, who may be infused with this factor three times a week or more, is exposed to the blood of millions of people over a lifetime. Thus, it is easy to see how the hemophilic population was so widely exposed to HIV infection.

The availability of effective treatment for hemophilia and the increasing life spans of hemophilic patients (Levine, 1985) popularized the approach of a comprehensive health care team for treating hemophilic persons and their families (Agle, Hilgartner, Lazerson, & van Eys, 1977). Such a team may include a hematologist, a nurse clinician, and an orthopedist, and possibly a physiatrist or physical therapist, a dentist or oral surgeon, a neurologist, a social worker, a psychologist, a genetics counselor, and a vocational counselor (Aledort, 1986; Wincott, 1977). Traditionally, occupational therapists have not treated persons with hemophilia. Most persons with hemophilia, however, are now infected with HIV, and occupational therapists treating persons with HIV will be seeing more of the hemophilic population. To provide the most effective therapy, occupational therapists should know the specific concerns of this group.

**Hemophilia and the Family**

As with any chronic disease, hemophilia affects the entire family. Parents may feel guilty that their genes may have caused the disorder; they may be overprotective in order to prevent excessive bleeding; and they may give more attention to the child with hemophilia than to their other children (Aledort, 1986; Jonas, 1977; Simon, 1984). Siblings are taught to treat the hemophilic family member gently, so as not to cause bleeding. This may lead the parents and siblings to feel overprotective, resentful, and guilty (Jonas, 1977). The child with hemophilia is taught to choose passive activities in order to avoid bleeding. This may cause problems, because boys in Western cultures are encouraged to be physically active and to play roughly. It can also lead to passive-dependent personality disorders (Aledort, 1986; Jonas, 1977; Simon, 1984). Teenagers with hemophilia may rebel against this passivity and choose more dangerous activities such as riding a motorcycle or engaging in contact sports, which greatly increase the risk of bleeding (Aledort, 1986; Jonas, 1977; Simon, 1984). Parents may feel that family relationships are negatively influenced by the hemophilia. As with any chronic disease, however, hemophilia's presence is generally accepted, and the family learns to adapt (Salk et al., 1972).

**Coping With HIV**

The hemophilic population is greatly troubled by the finding that HIV can be transmitted through blood infusions. Lyophilized factor, which may be infused at home, has provided freedom and independence to persons with hemophilia, allowing them to attend school regularly, live to adulthood, stay out of the hospital, and seek and maintain employment (Levine, 1985; Rabiner, Teifer, & Fajardo, 1972). Due to HIV contamination, however, this product is now threatening the lives of nearly all persons with hemophilia born before 1985. Since 1985, blood products have been screened for HIV (Petoom, 1988). Heat treatment of lyophilized factor to kill the HIV virus began in 1984 (Stehr-Green, Evatt, & Lawrence, 1988). From the time clotting factor concentrates became widely available to the time of widespread HIV infection, the hemophilic population has not had to face its own mortality so closely. Home infusion helped persons with hemophilia improve their attitudes about the disease, enjoy more feelings of freedom (Markova, Forbes, Rowlands, Pettigrew, & Willoughby, 1983), and feel more in control of their lives (Lineberger, Hernandez, & Brantley, 1984). But HIV infection may now be taking away that sense of freedom and control.

Hemophilic persons often cope with the threat of HIV infection by denying that it exists. Stehr-Green, Evatt, & Lawrence (1988) found that many hemophilic persons refused to be tested for HIV antibodies, as did their spouses and sexual partners, although spouses' and sexual partners' rate of infection estimates range between 10% and 60%. In addition, the offspring of an HIV-positive mother have a 50% to 60% chance of becoming infected perinatally (Hemophilia Information Exchange, 1988). Evatt (1987) reported that, even with these risks, the hemophilic population is reproducing at a higher rate than the general population. Few of these couples practice safe sex (Stehr-Green, Evatt, & Lawrence, 1988). In order to maintain an independent life-style, persons with hemophilia have traditionally been well informed about their own health care. The National He-
hemophilia Foundation has published and distributed much information on hemophilia treatment, AIDS, and other relevant issues for physicians, local hemophilia foundation chapters, and individuals. Information about the disease, therefore, is readily available, even if it is not widely used.

**Occupational Therapy’s Role**

Occupational therapists traditionally have not been part of the health care team treating persons with hemophilia, but the widespread rate of HIV infection in this population may change this situation. There is no documented evidence, however, that the functional deficits caused by HIV infection in this population differ from those of the general population of HIV-infected persons. Statistics from the Centers for Disease Control and the National Hemophilia Foundation regarding the course of HIV disease indicate that treatment recommendations should follow national trends for most infected populations.

Persons with hemophilia may face unique psychosocial issues. Occupational therapy can help persons with hemophilia with their denial of HIV disease. These clients have already learned strategies for coping with the life-threatening illness of hemophilia. They have learned to adapt to bleeding and chronic disability. Occupational therapists could help these clients learn to transfer these skills to coping with HIV and to focus on healing and wellness. An internal locus of control and a positive attitude have been shown to help bolster immunity (Siegel, 1986).

Persons with hemophilia may be angry about having been infected with HIV and may see themselves as victims. Additionally, they may be angry about being associated with persons who became infected with HIV due to behaviors deemed socially unacceptable or they may be angry at HIV-positive blood donors.

Occupational therapists can address these feelings by teaching stress reduction and anger management techniques. Stress not only lowers immunity (Kiecolt-Glaser et al., 1986) but also leads to more frequent and severe bleeding (Swirsky-Sacchetti & Margolis, 1986). Deep breathing, positive imagery, progressive relaxation, self-hypnosis, and gentle stretching are some of the stress-reduction methods that clients can learn.

Effective communication skills are important when dealing with patients with hemophilia and HIV infection. Family members, neighbors, friends, and co-workers must be educated about both diseases. The client must be able to express his or her feelings and listen to the feelings of others. Occupational therapists in both physical disabilities and mental health settings can help HIV-infected persons learn to communicate more effectively.

Differences in treatment for HIV-infected persons with hemophilia will likely involve biomechanical management. Exercise is important for maximizing health in persons with HIV disease. In designing treatment programs, the therapist must be aware of any muscle or joint bleeding before starting any treatment sessions. Even if an area has already stopped bleeding, recently formed clots can break loose. Bleeding must often be treated for 10 days to ensure recovery (Gilbert, 1975). If a client has severe bleeding in any limb or has any bleeding into the forearms, then these body parts can be splinted (Gilbert, 1975). Many adults will have some limited range of motion due to muscle scarring and degenerative joint disease, for which swimming is often recommended (Hodder, 1989). In addition to the prevention of bleeding, a treatment program should be designed to accommodate the lower energy levels experienced by persons with later-stage HIV infection.

Possible neurological complications of HIV infection include depression, withdrawal, apathy, ataxic problems, gross and fine motor latencies, balance problems, confusion, memory and attention deficits, and difficulties with mental flexibility (Boccellari, Dilley, & Shore, 1988). The therapist can enhance function through treatment based on a rehabilitative frame of reference. The techniques used could include (a) providing structure through creating schedules, (b) aiding memory by having the client carry paper or a tape recorder, (c) improving safety by posting relevant signs or pictures (e.g., turning off the stove), and (d) enhancing comprehension by breaking down information and directions into single steps (AIDS Health Project, 1987). Adaptive equipment such as raised toilet seats, plate guards, and reacher sticks may be used as well.

As with any client with later-stage HIV disease, energy conservation techniques can be useful. Occupational therapy focuses on assessing what is meaningful for clients and helping them achieve their goals. Therapists may need to help this population develop short-term goals that focus on the quality of life.

Some clients may still be working, at least part-time. Occupational therapists can help them adapt their job environment and help suggest easier ways to perform tasks. If a particular job is too physically or mentally stressful, the occupational therapist can help the person with hemophilia find ways to transfer his or her skills to another job.

As more children develop later-stage HIV disease, occupational therapists will increasingly have to help them and their parents explore life-style options.
Adolescents may need help making career choices and achieving independence. Persons with regressive development, lowered energy levels, and the prognosis of a shortened life span must learn to use their remaining resources as productively as possible.

Through family education, caregivers can help their loved one function as well as possible and can better cope with their own stress. Adapting home environments, showing caregivers mobility and transfer techniques, and encouraging family members to help the infected person contribute to the family are a few of the ways in which occupational therapists are already helping families care for their chronically disabled members.

Although the basic principles of occupational therapy do not change when one is providing services to persons with HIV disease and hemophilia, therapists must be informed about the specific needs and characteristics of this population. Therefore, a list of resources regarding hemophilia is included at the end of this paper.

Conclusion

The hemophilic population has gained its independence in the past 20 years due to the availability of concentrated blood clotting factor. Unfortunately, this life-giving treatment has threatened most of the hemophilic population with HIV infection. Unless a cure is found for HIV disease, most hemophilic persons born before 1985 will likely develop AIDS in the next few years. Occupational therapists can play a useful role in helping persons with hemophilia maximize their health and independence.

References


Salk, L., Hilgartner, M., & Granich, B. (1972). The psycho-social impact of hemophilia on the patient and his family. Social Science in Medicine, 6, 491-503.


Resources for Hemophilia and HIV Disease


The Northern California Chapter of the National Hemophilia Foundation, 7700 Edgewater Drive, Suite 710, Oakland, CA 94621-3017 415-568-NCHF. Publishes AIDS News, a quarterly newsletter on AIDS issues as they relate to hemophilia.


**AJOT Update**

Coming in April:

- Treatment approaches for adults with perceptual deficits
- A stress management program
- Effects of keyguard use and pelvic positioning on typing skills
- Quality of time use by adults with spinal cord injuries
- Improving feeding skills in a child with Rett syndrome
- Research attitudes and activities of clinicians

Turn to AJOT for the latest information on occupational therapy treatment modalities, aids and equipment, legal and social issues, education, and research.