A BRIEF HISTORY OF HEMOPHILIA

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Hemophilia is a sex-linked, hereditary bleeding disorder. The affected gene is carried by the female and expressed in male offspring. The first reported cases of uncontrolled bleeding appeared during biblical times when it was observed that some male infants bled to death following circumcision.

In recent history, the most familiar name associated with this disorder is Queen Victoria, who gave birth to a son with hemophilia and daughters who were carriers. No effective treatment to control bleeding was available during Victoria's time. However, as a result of medical advances, her hemophilic grandson was able to live well into his 50's.

By the mid-1800's, whole blood transfusions from unaffected persons to persons with hemophilia were known to check bleeding. Improvements in hemophilia treatment appeared in relatively rapid succession in the latter half of the 20th century. These advancements included the separation of plasma from whole blood, followed in the mid-1960's, by a serendipitous discovery by Dr. Judith Pool. She observed the formation of crystals at the bottom of a thawing bag of plasma. Testing the composition of these crystals, she learned they were rich in Factor VIII, the deficient clotting protein in persons with hemophilia. This material is called cryoprecipitate (cryo) and in the past was used in Hemophilia Treatment Centers to treat people with hemophilia and other clotting disorders, including von Willebrand's Disease. Cryoprecipitate is still used occasionally to treat specific cases.

Researchers then developed a laboratory methodology for concentrating the cryo into a dry, lyophilized form. The development of Factor VIII and IX concentrates is often considered the most significant scientific gain in hemophilia care. Prior to the availability of concentrate, persons needing treatment for hemorrhages were often forced to go to hospital emergency rooms, clinics, or their doctor's office where the plasma or cryo was infused under physician supervision. Concentrate, which could be stored at home in the refrigerator, made it possible for individuals and parents to manage infusions themselves. Thus began the era of home treatment, which made it possible for people with hemophilia to infuse concentrate as needed at home, school, or work. Home treatment enabled the person with hemophilia to attend school regularly, maintain employment, and function independently.

Private physicians provided most hemophilia care until the 1970's, when patients, families, and hematologists began to organize and lobby at a state and national level. By 1975, the Congress, through the Office (then Department) of Maternal and Child Health, enacted legislation to fund a network of Comprehensive Hemophilia Treatment Centers (HTC). The core clinical team at

these Centers continues to include a physician, nurse coordinator, and social worker. Consultants to the team typically include a physical therapist, genetics counselor, orthopedist, and dentist. Due to the creation of comprehensive care teams, medical management of hemophilia became easier. Care efforts were focused on normalization, increased life span, efforts to increase function (e.g., orthopedic surgery), dental care, and an increased emphasis on psychosocial care.

The future for persons with hemophilia was promising despite some well known complications of treatment such as inhibitors and hepatitis. Concentrate facilitated prompt, relatively simple, and effective treatment for most bleeding episodes. However, in 1983, the appearance of AIDS in the hemophilia population, spread by contaminated blood products, dramatically altered the perception of hemophilia as a chronic but manageable condition.

AIDS has both devastated and mobilized the hemophilia community. There have been hundreds of deaths, including those resulting from transmission to spouses and unborn children by infected men. On the other hand, self-help and other support efforts have been strengthened both locally and nationally. Increased funding in response to the AIDS crisis has increased the number of mental health specialists providing services at HTCs and enabled the National Hemophilia Foundation to develop educational and outreach efforts to the community of providers and consumers. Treatment Centers have developed individual and/or group support services in order to promote safer sex behaviors and to provide emotional support to families and individuals in crisis.

The progress toward improved hemophilia care continues. Increasingly sophisticated processes have been developed to eradicate HIV and other viral contaminants from factor concentrates. The most exciting research development involves the clinical trials, now underway, of a genetically engineered clotting material.

It remains a challenge to social workers to work effectively in collaboration with the multidisciplinary team, the HIV-infected population, and those who have not been exposed to HIV. It is hoped that the information in this orientation manual will help to foster your efforts.

For further reading:

Mason, P.J., Olson, R.A., and Parish, K.L., (1988),"AIDS, Hemophilia, and Preventive Efforts Within a Comprehensive Care Program". American Psychologist, vol.43, #11, pg.971-976.