

SCHOOL HEALTH MANUAL

HEMOPHILIA

MAINE SCHOOL HEALTH ADVISORY COMMITTEE

with input from

MAINE DEPARTMENT OF EDUCATION

MAINE DEPARTMENT OF HUMAN SERVICES

AND OTHER RELATED ORGANIZATIONS

COMMENTS

The School Health Manual is available electronically. Each section of the Manual is available as a separate electronic file from the Department of Education WEB Page at <http://www.maine.gov/education/sh/index.html> This will allow for sections to be updated on an ongoing basis.

Comments may be given to members of the School Health Advisory Committee or sent to the below.

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HEMOPHILIA

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Hemophilia is a complex, often misunderstood, expensive, congenital bleeding disease that affects 1.8/10,000 males. The number of hemophiliacs in the State of Maine is currently slightly in excess of 120 males, and the number in the United States is estimated to be about 95,000. Hemophilia is a disorder of the blood clotting mechanism. Because of an abnormality in one of the several plasma proteins necessary for clotting, the blood does not coagulate properly.

There are two major types of hemophilia: Hemophilia A (Classical Hemophilia resulting from an abnormality in Factor V111) and Hemophilia B (Christmas Disease resulting in an abnormality in Factor IX). The usual ratio of Classical Hemophilia to Christmas disease is 3:1, but in Maine the ratio is closer to 2:1.

This genetic defect results in uncontrolled bleeding from tissues, deep cuts, heavy blows, or tooth extractions. Recurrent bleeding may cause progressive joint damage and crippling. The hemophiliac does not bleed faster, but more easily and longer than the average person. Contrary to popular myth, he will not bleed from a scratch, but may limp or have a chronic gait disturbance as a sign of previous joint bleeds.

There is no cure for hemophilia. The bleeding episodes require treatment to temporarily replace the non-functioning protein factor. Traditionally these protein factors were present in fresh frozen plasma, FFP, for both Hemophilia A and B.

In 1965 a frozen preparation of Factor VIII was developed called cryoprecipitate. In the early 1970's, drug companies were able to produce the needed product of both Factor VIII and Factor IX in a powdered form referred to as concentrate. The infusion of these blood products and/or concentrates by injection may now be done at home or in the traditional medical or hospital setting; this has led to significant changes in the care and treatment of hemophilia, resulting in much less time in the hospital.

Hemophilia A and B are both "X-linked" inherited diseases. This means the disorder may be transmitted to male children by females who carry the genetic information, although the females themselves are not hemophiliacs. All the sons of hemophiliacs are normal, but all daughters of hemophiliacs are carriers. The female carrier has a fifty percent chance of transmitting the hemophilia gene to any of her sons or daughters, and thus has an equal chance of giving birth to a normal son, an affected son, a carrier daughter or a normal daughter.

The relevance of the Maine Hemophilia Treatment Center to Maine's citizens with hemophilia is threefold. First, the population of males with hemophilia is growing. One third of all new cases of hemophilia are spontaneous mutations, that is without a known history of the disease within the family. Of all genetic diseases, hemophilia has the fastest known mutation rate. In addition, the availability of concentrates is allowing hemophiliacs to live longer.

Second, the team approach to total comprehensive care for the patient with hemophilia and his family contributes to a higher quality of life and to a more independent life for the hemophiliac and his family.

And thirdly, through a system of clinical associates at several geographical locations throughout Maine, patients may receive the highly specialized care they require close to their residence without necessarily coming to the treatment center.

The Maine Hemophilia Treatment Center provides comprehensive care for persons with hemophilia, von Willebrand's disease, and other coagulation disorders. Comprehensive care means that the person, not just the condition, is the focus of concern. A major objective of comprehensive care is providing support and evaluation of the medical and social factors affecting the individual and family. Early and ongoing intervention helps prevent the occurrence of complications. The multidisciplinary team at the MHTC offers evaluation, counseling, education and advocacy to help the individual realize full life potential. The Comprehensive Care Team includes: the individual and family, the hematologist, the nurse coordinator, the social worker, the physical therapist and the orthopedic physician, the dentist, and the geneticist.

The Center offers many diverse services.

- • Hemophilia Education for persons with hemophilia and their families allows individuals the opportunity to make informed health care decisions.
- • The Home Infusion factor concentrate provides immediate treatment for bleeding episodes and prevents pain and disability. Individuals and parents are taught the skills necessary for home infusion, allowing greater freedom and independence in their lifestyle.
- • Summer Camp/Regional Retreats provide a fun and relaxing atmosphere, as well as an opportunity to meet peers with hemophilia, learn new skills and enhance self esteem. Professional resources and educational programs are available for community based physicians, hospital staff or health agencies.
- • Genetic counseling and carrier testing options, vocational referral and counseling, financial resource guidance, risk reduction education, hepatitis B vaccination, confidential HIV testing and counseling.
- • The Center is also the main contact for sickle cell anemia.