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# News

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**\*\*Team approach to hemophilia gives hope to children, adults.**

DALLAS--To the child with hemophilia the freedom to be like other kids may mean the difference between happy or unhappy childhood.

To his parents the freedom to have a normal family life may make the difference whether or not the home stays together as a loving, functioning unit.

To the adult with hemophilia the freedom to break the cycle of crippling social isolation and helplessness as a result of deterioration from the disease may literally save his life.

Hemophilia, sometimes called "the royal disease" or "the curse of the Hapsburgs," is a genetic, life-long chronic condition that prevents blood from clotting normally. And bleeding into the joints, especially at the ankle, elbow and knee, left unchecked lead to permanent crippling and a pain-ridden existence for the sufferer.

The victim of hemophilia - nearly all are male - does not produce one of the plasma proteins that are needed to form blood clots. The disorder is transmitted on a gene of the X chromosome. Since women have two X chromosomes, they are not affected since they always have one normal chromosome even though the other carries the defective hemophilia gene. The mother, however, may pass the defective gene on to her male children, and tests for carriers are only 80 percent accurate. There is a 50-50 chance that a carriers' sons will be hemophiliacs, and a 50-50 chance that their daughters will also be carriers. When there is no record of the disease in the family of a newly diagnosed case of hemophilia, it may be because of incomplete family history, or more rarely, it may represent a mutation.

Today, however, more than ever before, there is hope for the hemophilia patient, says Dr. George Buchanan, associate professor of Pediatrics and director of the North Texas Hemophilia Comprehensive Center. The North Texas Center is a cooperative effort of The University of Texas Health Science Center at Dallas, Children's Medical Center and Parkland Memorial Hospital, which cooperates through its blood bank and the hospitalization and treatment of adults. It works closely with the Texas Central Chapter of the National Hemophilia Foundation and encourages its young patients to attend the chapter's summer camp. In fact, Buchanan's whole staff helped out at the camp this year.

All these services are involved in opening up new freedoms to the hemophilia patients. Travis Roop of Dallas is such a patient.

Not only does Travis have a loving family with mother Gloria, father Gerry and older sister Ashley, but he lives in a city where he can have comprehensive care for his hemophilia. At three-and-a-half, Travis so far hasn't had so many bleeding incidents that his family has needed the training for home infusions. But as Travis grows older, says Gloria Roop, these bleeds could well become more of a problem. At that time both parents will take lessons on how to properly give home infusions. Then, says the mother, the family will feel freer to go camping in the wilderness of Colorado together because they won't have to worry about backpacking back to civilization before Travis could receive treatment.

Modern medicine, including home infusions, has certainly led to lengthening the life span of the patient with hemophilia. In addition, comprehensive care as it is being practiced in many medical centers today is making a distinctive contribution to the quality of life. Not only are physical therapists available as a part of the team to head off unnecessary crippling, but the orthopedic surgeon also monitors for permanent damage on a routine basis. In addition, there are the services of the dentist and the invaluable nurse/educator.

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hemophilia - add one

Another important role is played by the social worker. Because serious hemophilia - especially in the past - is often responsible for keeping children and young people out of school for long periods of time or an inordinate number of days each year, there may be learning and/or drop-out problems. Adding to the feeling of futility the student who is constantly behind may feel may be uncertainties about his masculinity. Restricted from participation in school athletics, the patient may begin to think of himself as "sickly" or "dainty."

Buchanan has been treating children with hemophilia for seven years at Children's and the hospital has made its resources available to children with the disorder and their parents for this time. Now, however, a concerted effort to provide on-going care and patient education to both children and adults is being made through the comprehensive North Texas program and at other similar centers around the country. Patients in the Dallas program make required visits either once or twice a year. However, they may make appointments to see health team members, who are also available in emergency situations, at any time.

The program at Children's brings together a whole health team to get acquainted with the hemophilia patient and his problems, whether these be medical, social or psychological. Team leader Buchanan is proud of the expertise assembled to combat this crippling illness. Members include the following: Dr. Marybeth Ezakik, an orthopedic surgeon; Nancy Richards, a pediatric nurse practitioner; Martha McKee, a social worker; Dr. Andy Harris, a pedodontist (dentist who specializes in children's teeth and gum problems); Dr. Mateel Graham, a physician who is a hematology specialist in internal medicine and works with adult hemophilia; Dr. Bob Sprinkle, a pediatrician and family practitioner who sees adults and young adults with the disease; Barbara Bailey and Alice Anderson, physical therapists; and Chris Holth, laboratory technologist.

Buchanan explains that while most people think that hemophiliacs are people who can bleed to death from any little cut anytime, this is not the case. The patient doesn't bleed any harder or faster than the normal person. However, he will continue to bleed long after the normal person because the blood cannot form clots that act as "plugs" to dam up the torn blood vessels.

Small surface cuts can usually be controlled with pressure bandages, but bleeding in the muscles and joints is prolonged. Often normal trauma, such as a fall or blow, may start the bleeding process, but the cause of some episodes, termed "spontaneous," may be unaccounted for. Just the normal activity that a baby goes through in learning to crawl, pull up or walk can start numerous bleeding episodes. Of course, not all hemophilia is of the same severity.

Serious problems are caused at any age by bleeding into joints and muscles. These "bleeds" may eventually cause a painful type of arthritis. Also, the bleeding causes weakened muscles around the joints and possibly severe pain. Sometimes the pressure on the nerves also causes numbness and a temporary paralysis in the area. Severe problems may lead to need for joint replacement or other types of expensive rehabilitative surgery.

While this description of the medical problems faced by hemophilia patients seems grim, Buchanan stresses that there is much that can be done. And, in fact, with the help of the whole team, the patient is in a better position to cope with his condition on a day-to-day basis, as well as having help available for the life problems that often accompany the disease.

On this front the team leans heavily on Richards, the nurse practitioner and member who spends the most time with patients, and McKee, the social worker. The two work closely together, one's role often supplementing the other or overlapping.

"Nancy," says Buchanan, "is the one who does all the stuff that makes me look good."

"Making the doctor look good" includes spending hours with patients - or in the case of children - with their families, too - in educating them about the disease, its symptoms and properly monitored care can help deter its progress.

The biggest breakthrough in care for patients with hemophilia has been home infusion of blood products, which has been in wide use for about the last 10 years, says the pediatric hematologist. Mild forms of hemophilia are usually treated with fresh frozen plasma. However, extracts of normal plasma, either freeze-dried concentrate or cryoprecipitate, sometimes called "cryo" for short, which is prepared from fresh donor blood and freezer-stored, are used in more severe cases. All are given by infusion whether in the home or the doctor's office or the hospital or emergency room.

(more)



hemophiliacs - add two

Not only does being able to start transfusions early help with the immediate bleeding problem - which is usually accompanied by a great deal of pain - but the quicker the blood products are given, the less the permanent effects from bleeding in the joints. Patients are also routinely monitored through blood tests for hepatitis, a constant threat to anyone using blood products on a regular basis, and AIDS, a new worry.

Not only does the UTHSCD/Children's program of regular medical care have a great effect on the physical freedom for hemophilia sufferers, for most there is a great gain in psychological freedom, says Buchanan. One Dallas patient, who neither thinks of himself as "sickly" or "dainty" is 14-year-old John Reeves of Plano. Reeves, who skis the "blacks" (tough slopes) in Colorado, scuba dives with his mother as his "diving buddy" in Florida and has recently taken up spear fishing. The teenager has been infusing himself for a long time and this contributes to his freedom.

"There are few kids his age who are better skiers than John - and certainly few hemophiliacs," says his mother Peggy. "The first year was terrible, though: the poor little thing would cry from arthritis pain on the slopes. And once he had to have the ski patrol take him down in a basket - but he wouldn't give up."

Peggy Reeves says that while it's hard to let John do some of the things he does, she learned an important lesson from her mother. "Don't lock him up in a cage," she says her mother told her, "Let him enjoy as much as possible the life he does have." The mother had learned this lesson herself the hard way: her infant son died from what is now thought to have been hemophilia.

When John's mother, a single parent, realized how much her son would enjoy scuba diving, she looked for "just the right teacher." She found one who was a paramedic himself and whose wife was a physician.

Still the teacher was skeptical. But the mother was determined. She pulled out her arguments on why diving is no more dangerous to a careful hemophiliac than to a careful non-hemophiliac - then pulled out the letter of permission from one of the doctors in hematology at CMC.

All the things John has learned to do well - including the skiing, scuba diving, spear fishing, playing a slow game of racquet ball, shooting skeet and playing pool - have worked together to give John good feelings about himself.

"I decided that was the most important thing, so I've really been working on it," says his mother. "When he was younger, I couldn't stand watching him sit and cry from a feeling of worthlessness."

Unfortunately, not everyone is as spunky as John. Not just dropping out of school but almost total social isolation may be the result of a patient's having bad feelings about himself. "Too often," says McKee, "patients in the past failed to educate themselves so they had no choice but manual labor. And that is the worst thing in the world for the hemophiliac with his bleeding problems."

One of the things that team member Richards does with young people is to get them thinking about what kind of job they want when they grow up. She sees part of her role as keeping them in reality about the kind of vocations that would be practical from a health-related viewpoint. She also says she often talks with adults who are making decisions about job changes or starting their own business because insurance that covers hemophilia care, including blood products, is of major importance. The financial strain on the family is often horrendous, says Richards. And the bills for products alone can run \$10,000 a year for one severe case in a family.

Sensitivity to what's going on in the family is also a major thrust for the social work component of the team, says McKee. "As with other major chronic health problems in a family, there is a high rate of divorce."

"We're proud of the team we've assembled and the way we work together," says Buchanan. "Our goal is to help the patients have as healthy and whole lives as possible."

And it seems they're succeeding - at least with patients like John Reeves.

On one of his ski vacations, an instructor was so impressed with John's skill that he asked if he would come by his classes the next week and give some pointers to a group of handicapped children taking lessons.

"Why?" asked John. "I'm not handicapped."

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