HIV's Startling Spread Among Hemophiliacs

By Randy Shilts
Chronicle National Correspondent

Americans afflicted with hemophilia have the highest HIV infection rate of any population in the world and face serious problems ranging from health care costs as high as $100,000 a year to the largely unabated spread of the AIDS virus to their wives and children.

An estimated 60 percent of the nation's 20,000 hemophiliacs carry the human immunodeficiency virus, which they unknowingly injected into their veins during routine treatments for the life-threatening blood disorder. Already, more than 1,100 hemophiliacs have been diagnosed with full-blown AIDS.

In spite of this astounding rate of infection, government agencies and private organizations have been slow to confront the AIDS problem, leaving hemophiliacs to suffer from substandard health care.

"It's so ironic," said Larkey De Neffe, 27, a hemophiliac and program chair of the Oregon chapter of the National Hemophilia Foundation. "Of all the groups who got AIDS, we were the ones who could have done the best. We are used to dealing with a medical system, relying on new medical techniques. And look what's happened."

What has happened is a chain of medical, psychological, social and economic problems that have converged to make hemophilia the darkest part of the AIDS epidemic:

- New technologies have enabled researchers to develop pristine blood products for hemophiliacs, but costs are so high that patients are quickly exhausting insurance limits and...
Deadly Spread of AIDS Virus in Hemophiliacs

From Page A1

sinking into poverty to pay for the products.

Some hemophilia centers have refused to prescribe new, purer blood products to HIV-infected patients in apparent violation of federal civil rights laws. At one center in Portland, Ore., a high-ranking physician has refused to prescribe an important new medical product because he is opposed to marketing techniques used by the firm that manufactures it.

Although AIDS clearly represents the most serious health problem for these patients, some hemophiliacs have refused to acknowledge the AIDS threat and have resisted gaining expertise on HIV, putting their patients’ health in greater peril.

Hemophiliacs, who are more assimilated into the general population than other high-risk groups, have been subject to some of the cruelest discrimination reported in the AIDS epidemic. Dozens of hemophiliacs have had to go to court just to be able to attend school. One family in Florida was burned out of its home.

Cleaner blood products stopped new HIV infections of hemophiliacs four years ago, but the disease continues to spread because the hemophiliacs—all of whom are male—generally have been slow to adopt safe-sex guidelines, infecting their wives, who then transmit the infection to their offspring during pregnancy.

“People are dying left and right,” said Dr. W. Keith Hoots of the Gulf States Hemophilia Center in Houston. “I can’t stand it.”

Young Victims

What makes the problems even more tragic is that they are so often played out in the lives of the young, as hemophilia strikes thousands of children and teenagers.

As these youths attend their hemophilia clinics around the country—with brightly colored posters of “Garfield” and “Teenage Mutant Ninja Turtles” hanging on the walls—the colorful decorations mask the severity of their predicament.

The Oyler family of Carmel, Calif., is one example. Benjamin Oyler had been jaundiced and infected with HIV in 1982, but was not infected himself. However, his son, Michael, was infected in 1985 when he was 10 years old.

Case Study

Lorkey DeNeef, a leader of the Oregon chapter of the National Hemophilia Foundation, injected himself with Factor VIII, which is used to treat hemophilia. He later tested positive for HIV.

The Oyler family of Carmel

The use of heat-treated Factor VIII largely stopped new infections by 1985, but huge numbers of hemophiliacs already were infected with HIV by that time.

Most estimates put the rate of HIV infection among the nation’s 7,000 to 9,000 severe hemophiliacs at about 95 percent. Among moderate hemophiliacs, who required fewer infusions, infection rates hovered between 40 percent and 50 percent. Few of those with mild hemophilia are infected with HIV.

The devastating tales of untimely death, particularly among young hemophiliacs, are often made worse by the social ostracism inflicted on these youths.

In Carmel, it was only when young Ben Oyler was about to begin the third grade that his mother learned the school board had decided to ban him from classes.

“We had moved to a new house,” said Mrs. Oyler, “so Ben had no friends around—he was so attached to going to school so he could have contact with other children.”

“We’re in the same room together,” she said. “We’re in the same room together.”

The Oyler family of Carmel

“We didn’t expect the rejection... We needed love and support.”

Benjamin Oyler was drafted to 34 pounds before he died of AIDS in 1986

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Portland Doctor Withholding New Version of Clotting Factor

Portland, Ore.

Barry Kurath was ecstatic when he learned that researchers had devised a pure new version of the clotting factor he needs to treat his hemophilia.

The new Factor VIII product, he knew, would drastically reduce his exposure to hepatitis viruses—a major development, given that liver disease is the second highest cause of death among hemophiliacs.

But when Kurath went to ask for the new treatment, his doctor told him he would not give him a prescription.

The physician, Dr. Everett Lovrien, medical director of Hemophilia Center at the Oregon Health Sciences University, concedes that the product is "very fine medicine" but refuses to give it to Kurath, in part because he does not like the company that makes it.

The company, Armour Pharmaceuticals, has been contacting patients directly to sell the new Factor VIII. To discourage such direct marketing, Lovrien is simply refusing to prescribe much of the product.

"We don't want to lose control of our patients," he says bluntly.

Complicating the issue is the high cost of Factor VIII and other treatments in general. Lovrien says he will not give expensive treatments to patients who, like Kurath, are HIV-infected.

Safer Treatment

"If you take some patient who's HIV-positive and put them on a medication that costs $50,000 a year—and the reason to do it is that it's safer, but they've already got HIV—thats doesn't make sense to me," he says. Lovrien controls financing for Oregon's Hemophilia Assistance Program, which is what pays for Kurath's Factor VIII treatments.

As for the treatment's effect on protecting immune-deficient patients from further liver disease, he says such benefits are yet to be definitively proved.

Jonathan Botelho, a hemophiliac and a staff member with the Office of Civil Rights of the Department of Health and Human Services, said such decisions violate federal laws banning discrimination against people with disabilities, such as HIV infection.

But Lovrien says, "I don't pay attention to the law. I try to do what's logical—what's logical, that's illegal.''

At the U.S. Department of Health and Human Services in Washington, Sharon Barrett, who administers the federal program that finances the Portland center, says she is "a little disturbed" by the problems patients face in obtaining monoclonal Factor VIII. "It is definitely an area I'm looking into this year," she said last week.

Many hemophiliacs say they feel restricted in their ability to protest, because they worry about losing their access to treatment altogether. Kurath is reluctant to pressure Lovrien because he is the chief of the only hemophilia treatment center within 600 miles of Kurath's Portland home.

"You don't want to sue your doctor when he's the only one who's got the medicine you need," says the computer programmer.

Lovrien's apparently capricious ill will toward Armour Pharmaceuticals has denied patients millions of dollars in free Factor VIII. He refused the company's offer of free monoclonal produced Factor VIII for his patients, saying he "doesn't believe in rebates." Patients who wanted the factor were simply refused a prescription.

To make matters worse, patients who have obtained prescriptions from other doctors often find themselves thwarted later when they ask insurance companies for reimbursement. Third-party payers often seek the advice of the region's foremost hemophilia expert before paying for the product. That expert is Dr. Lovrien, who advises them not to pay for the factor.

"You end up feeling like you're hitting your head against a brick wall," Kurath says.

— Randy Shilts

How Medical Success Story Turned Into a Nightmare

From Page A11

role in all policy related to HIV and hemophilia, sometimes to the frustration of public officials.

"I never felt it was the role of the U.S. Public Health Service to defer to a private foundation," said a staff member of the Centers for Disease Control who worked with hemophiliacs in the early years of the epidemic. "It really slowed things down."

Already Integrated

The NIH decision to temper its response to AIDS continues to haunt the organization today and is a subject of a new report from a central

AIDS AMONG HEMOPHILICS

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<th>Number of cases</th>
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there's a lot of Monday morning quarterbacking today about our actions then," Brownstein said. "We thought AIDS was a significant new event that would cause death, but we had no idea it would be as much of a problem as it was today."

Early AIDS denial also slowed the response of other populations, such as the gay community. In contrast to other groups, however, denial among hemophiliacs persisted long after the early, confusing years of the pandemic.

"Today, in no area is the continued denial of HIV more poignant than in the difficulty in persuading hemophiliacs to adopt safe-sex practices."

Psychological Reasons

Part of the resistance stems from deep psychological reasons, hemophiliacs say, because sex has traditionally been one of the few physical activities they could enjoy.

"I remember when I found out about sex. I thought, here's finally a sport I can do," said Botelho, of the Northern California Hemophilia Foundation. After years of having to avoid vigorous physical activity, he said, "it was a place where I could feel macho."

Carrying the burden of HIV infection in severe isolation, the unmitigated despair wrought from the loneliness has also led to growing reports of suicides among hemophiliacs, particularly among HIV-infected teenagers who feel they have nothing to live for.

In Portland, suicide is now the second leading cause of death among hemophiliacs—aids. One man jumped off a bridge, took overdoses of phenobarbital three times and was on his way to leap from the city's tallest freeway bridge except that his car ran out of gas before he could reach the central section of the span.

"The doctors are crazed by the gum and the good life," said Botelho. "It's the first time in a generation that we're seeing signs of AIDS in an otherwise healthy occupation."

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Already Integrated

The NHF decision to temper its response to AIDS continues to haunt the organization today and is part of the source of a central irony in the problem of HIV among those with hemophilia. In virtually every way, hemophiliacs were a population that was better prepared than any other to face the challenges of a new chronic disease.

They were already integrated into hemophilia centers, giving them unparalleled access to specialized medical care. With the NHF, they also had the first respected national organization with chapters in every part of the nation that could champion their interests. But by most accounts, rather than working to help hemophiliacs in the crisis, these factors sometimes impeded an effective response to the disease.

"The attitude from national (the NHF office) was not to get involved," said Jonathan Botelho, president of the Northern California Hemophilia. "They said, 'Don't talk about it out loud.'"

"To this day," he said, "there are chapters that won't print the word AIDS in their newsletters."

Along with fears of discrimination, national foundation leaders in New York City worried that too much attention on AIDS might frighten patients away from using Factor VIII, leaving them to suffer from the debilitating effects of untreated hemophilia.

However, NHF critics among hemophiliacs, researchers and health officials cite another, simpler reason for the lack of a strong NHF response: denial.

The Denial Factor

"The NHF wants to please its group, and the truth is not going to make them happy," said one federal official who worked extensively with the organization and asked not to be named.

This denial factor had disastrous effects nationally because most federal money for AIDS education and studies is coordinated through the National Hemophilia Foundation.

NHF officials counter that nobody could have predicted in the early years the grim new threat that HIV posed. Even critics credit NHF with being an effective lobbying group to Factor VIII producers and with wielding influence in more conservative quarters of Congress.

Six months ago, the group decided to begin to adopt a more forthright public relations policy in regard to the problem of AIDS among hemophiliacs.

"Yes, there was denial, but there's a lot of Monday morning quarterbacking today about our actions then," Brownstein said. "We thought AIDS was not such a significant new event that would cause death, but we had no idea it would be as much of a problem as it has today."

Early AIDS denial also slowed the response of other populations, such as the gay community. In contrast to other groups, however, denial among hemophiliacs persisted long after the early, confusing years of the epidemic.

Today, in no area is the continued denial of HIV more poignant than in the difficulty in persuading hemophiliacs to adopt safe-sex practices.

Use of Condoms

The CDC was first alarmed by the threat to hemophiliacs' spouses in 1984 when an early study showed that 10 percent were already HIV positive. Dr. Janine Jason of the CDC, who worked on the research.

Although the NHF began publicly recommending safe sex, some of the group's chapters continued to have children - even though they risk infecting both wife and unborn child.

One federal official, for example, was shocked when one of the NHF's most prominent spokesmen rose at a 1987 conference to point proudly to his pregnant wife.

"He said that they decided to take the chance," the official recalled. "It was appalling. On paper, the NHF is talking about safe sex, but at their meetings, they still tell people how to do it for ovulation."

When Pittsburgh's Dr. Ragni tested sexual partners of her patients, 13 percent were infected. Other studies have found that as many as 25 percent of hemophiliacs' spouses are now HIV positive. Some of these women have gone on to have children who were born with the virus.

A recent study by the CDC and Stanford University found that only about 47 percent of HIV-infected hemophiliacs consistently used condoms.

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This year, Brownstein said, NHF and federal officials designated the halting of sexual transmission as a top "national goal" for AIDS prevention efforts among hemophiliacs. He conceded that the programs met with "substantial resistance" at first, in part because many of the hemophilia doctors were uncomfortable discussing sexual issues.

Brownstein said that NHF chapters are now increasing their AIDS education. Last month, the group launched a Women's Outreach Network in chapters nationwide to educate sex partners of HIV-positive men with hemophilia.

Yet another problem continues to plague hemophiliacs, often with life-threatening consequences. Because some of the doctors who staff the hemophilia centers remain uncomfortable about AIDS, many hemophiliacs are receiving substandard medical care for the HIV-related ailments, patients say.

As blood specialists, few hemophilia experts had much experience with either immunology or the diseases that follow HIV infection.

"They'll be the first to admit they're lost with this," said Jason on CDC. "They don't have a clue as to what they should do. They're not giving appropriate care, but it's out of ignorance, not maliciousness."

With some doctors woefully uninformed about advances in new HIV-related treatment, hemophiliacs have been left without the benefit of the medical breakthroughs that have marked clinical management of HIV infection during the past two years.

Along with the medical problems are profound social problems being suffered by a group that has created few of the support networks that have marked the response to HIV among gay men.

"More than anything else, hemophiliacs wanted nothing more than to assimilate into American culture - to be just like anyone else," said De Neffe, who has AIDS. "We're isolated. We're afraid. We're afraid of being identified with something like AIDS. They don't want to be associated with gay people or the drug culture."

When De Neffe tried to organize a support group for HIV-positive hemophiliacs in Portland recently, only one other patient wanted to attend. The result is that many are carrying the burden of HIV infection in severe isolation.

The unmitigated despair wrought from the loneliness has also led to growing reports of suicides among hemophiliacs, particularly among HIV infected teenagers who fear they have nothing to live for.

In Portland, suicide is now the second leading cause of death among hemophiliacs - after AIDS.

One man jumped off a bridge, took an overdose of phenobarbital three times and was on his way to leap from the city's tallest freeway bridge except that his car ran out of gas before he could reach the central section of the span.

"The doctors are crazy by this," said Dr. Louis Aleldor, director of the Hemophilia Center at Mount Sinai Medical Center in New York. "We gave it to them. We didn't put the virus in their bottles, but we were the instruments of their death."

For the 500 babies born with hemophilia every year, HIV will not go away, but new technologies for treating Factor VIII and Factor IX through recombinant DNA and gene therapy processes will also provide pristine infusions.

Without effective, long-term treatments for HIV, however, the short-term prognosis for the hemophiliacs population is bleak.

In the end, for experts and people with hemophilia themselves, there is a lingering sense of betrayal over how a medical story that once was so bright has fallen into such disaster.

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Oyler family watched Ben, a once-active and husky 5-year-old, lose his battle to AIDS-related pneumonia before he died at the age of 9 in 1986.

"You couldn't live without the clotting factors, but then it turned out you couldn't live with it," his mother, Christine Oyler, said painfully. "I think about that a lot.

As with many families with the genetic blood disorder, the Oylers' tragedy did not end with Ben's death. A year to the day after his eldest son died, Christine's 29-year-old brother, also a hemophiliac, succumbed to AIDS as well.

Such dark endings are an unlikely occurrence that is among today's most brilliant stories. Hemophilia is linked to a defect in the gene that controls the production of Factor VIII or, in some cases, hemophilia A women can suffer from an even rarer clotting disorder known as von Willebrand's disease.

Hemophiliacs suffer from varying levels of clotting problems, with about a third of all cases being described as severe, a third as moderate and a third as mild.

Years ago, the waiting rooms in hemophilia treatment centers were crowded with kindergarten-size charts because patients rarely lived to be adults. Each week of hemophiliacs' existence was punctuated by frenzied visits to emergency rooms for the only treatment then available to stop bleeding--whole blood transfusions.

Even with this radical treatment, children are handicapped by an early age, especially for one body to absorb enough blood to get the necessary amount of clotting factor.

A bumped knee or a twisted ankle may extract just the clotting factor that the hemophiliacs lacked. Patients could get enough of the factor to stop and even prevent their uncontrolled bleeding.

Hemophiliacs are treated at only a handful of hospitals around the country, these regional centers face an enormous burden.

Hospitals Balking

"We've got a real threat to the bank," said Dr. Louis Alford, director of the Hemophilia Center of Mount Sinai Hospital in New York City. "The hospitals are beginning to balk because they say 'Why should we pay this price?'

When National Hemophilia Foundation spokesmen prodded pharmaceutical companies to reduce prices, they were met with threats of the United States.

In fact, months before the new factor VIII products were announced, several major manufacturers simply stopped producing their heat-treated versions of Factor VIII, creating a nationwide shortage of the treatments.

Some experts suggest the shortage was the result of an industry conspiracy to force hemophiliacs to pay the increased prices. Pharmaceutical companies argue that they could not risk the legal liability of selling the heat-treated product when they were on the verge of releasing a healthier monoclonal factor.

The property tax bill of four AIDs patients around the country are nearing the amount they can be reimbursed for. Some are preparing to quit their jobs and go on welfare so that they can qualify for Medicare.

The impact on care facilities has been severe. Because a patient may require many thousands of units of factor for the simplest of procedures, the hospital bills can be enormous, usually far beyond what either private or federal insurers will pay for.

A hemophiliac in the hospital for AIDs-related pneumonia for example can be reimbursed only at a rate set for all pneumonia patients. The hospital, in turn, may not get reimbursed for the Factor VIII it must prescribe to keep the patient from bleeding to death during a simple biopsy.

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Burden for Hospitals, Patients

A Wildly Expensive Disease

Dr. Keith Hoots eringes whenever he learns that one of his hemophilia patients is about to check into the hospital, even for a minor procedure.

"I just get so worried that one of these times the hospital is going to say, 'We won't admit him,'" said Hoots, an associate professor of pediatrics at the University of Texas who works with the Gulf States Hemophilia Center.

Hemophilia has always been an expensive disease, but new medical advances have driven the cost of treatment so high that some patient centers are on the verge of leaving hemophilia care altogether.

The chief reason for the surge in the cost rests with the new Factor VIII products now on the market, which are the result of advances in monoclonal antibody technology that creates much safer treatments.

They also are extraordinarily expensive. The original, heat-treated Factor VIII typically cost about 9 cents a unit and now costs about 45 and 60 cents a unit for the new monoclonal products.

An infusion of 2,000 units, therefore, has a $1,800 price tag. And some severe hemophiliacs that much as often as four times a week.

Insurance Limits

Because the insurance policies carry a $1 million lifetime limit, hundreds of patients across the country are nearing the amount they can be reimbursed for. Some are preparing to quit their jobs and go on welfare so that they can qualify for Medicare.

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Now, the pharmaceutical companies are preparing the next generation of concentrates using recombinant DNA technology—a process that should provide even safer products at even higher costs.

"It's going to be at least $1 a unit," said Dr. Marion Koerper, medical director of the Hemophilia Center at the University of California at San Francisco. "It causes much more despair, because you just don't know where the money is going to come from to pay for this.
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From Page A1...

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Although AIDS clearly represents the most serious health problem for these patients, some hemophiliacs have refused to acknowledge the AIDS threat and have resisted gaining expertise on HIV, putting their patients' health in greater peril.

Hemophiliacs, who are more than 50 percent foreign and who have been subject to some of the most severe discrimination reported in the AIDS epidemic, have not heard of the disease.

In some centers, hemophiliacs have been taught to avoid the disease by taking precautions such as avoiding contact with infected blood and not sharing needles, but many of them do not understand the nature of the disease or know what to do about it.

Cleaner blood products stopped new HIV infections of hemophiliacs years ago, but the disease continues to spread because the hemophiliacs do not know how to avoid transmission.

“People are dying left and right,” said Dr. W. Keith Hooks, director of the Gulf States Hemophilia Center in Houston. “I can’t stand it.”

Young Victims

What makes the problems even more tragic is that they are too often played out in the lives of the young, as hemophiliacs are most often children and teenagers.

As youths attend their hemophilia clinics around the country — with brightly colored posters of “Galahad” and “Teenage Mutant Ninja Turtles” hanging on the walls — the cheerful decorations mask the severity of their predicament.

The Oyler family of Carmel, Calif., has found itself increasingly isolated from others in the affluent community. “There were close friends; I didn’t hear from any more,” Oyler recalled. “It was too much. We didn’t expect the rejection... We needed love and support.”

Benjamin Oyler waited away to 34 pounds before he died of AIDS in 1986.