

Report on the current situation of Chinese Hemophiliacs Beijing Aizhixing Institute, 2006

I. Basic facts about hemophilia

1. What is hemophilia?

The etymological definition of hemophilia is love of blood and was a name suggested for the disease by a medical treatise in 1828.

Ironically, if you ask any hemophiliac, he will probably reply that his feeling towards blood is anything but love.

Blood is made up of two parts, a solid and a liquid one (plasma). The solid part is composed of red blood cells which transport oxygen, white cells which help fight infection and platelets which help coagulation. Platelets are necessary for clot formation and this phenomenon misled early nineteenth century scientists into thinking that they were linked to hemophilia.

In the 1930s, it was discovered that hemophilia was linked to plasma, the liquid part of blood. Plasma is composed of proteins, salts, sugars and water. Fourteen different proteins are responsible for clotting and are known as clotting factors. Seventeen recognized disorders, some milder, others more severe, result from some anomaly in these clotting factors, one of which is hemophilia¹.

2. Inheritance

Females have two X chromosomes and males have one X and one Y chromosome. When an egg is fertilized, the female contributes with one of her X chromosomes and the male with either his X or his Y chromosome. If the male contributes with his Y chromosome, the new born child will be a boy. If he contributes with his X chromosome, it will be a girl.

Hemophilia is hereditary, it affects mostly men, and is linked to a defect in the X chromosome. However, one third of hemophiliacs have no history of hemophilia and their disease is caused by a genetic mutation in one of the mother's X chromosome.

3. Symptoms

Because linked to a deficiency in blood clot formation, the symptoms of hemophilia are frequent internal bleeding and hemorrhage. Hemorrhage can occur spontaneously or as a result of a small bump or injury.

The frequency of bleeding varies from individual to individual and has led to the categorization into severe, normal and mild hemophilia. Some people bleed only once a month while others do as often as several times a week.

¹ De Prince, p.9

4. Physical, psychological and economic challenges

Hemophiliacs must face a series of physical, psychological and economic challenges which vary depending on the severity of the disorder, the economic status of the patient and the quality and availability of medical treatment.

a. Physical challenges

Although the number of deaths caused by hemorrhage has decreased dramatically compared to the 1960s, some hemorrhages, like the intracranial or abdominal, pose greater risks than others and can in some cases be fatal. A small bump in the head might be the cause of an intracranial hemorrhage. This risk is enormous for small children who start learning how to walk. Untreated intracranial hemorrhages may result in learning disabilities, paralysis and blindness. As for abdominal hemorrhages, apart from being extremely painful, they produce a great amount of blood loss and when occurring within small organs, may destroy them completely.

Bleeding weakens the patients by causing persistent anemia and damage to organs, joints and muscles. Joint deterioration eventually results in inability to walk. Frequent hospitalizations and periods of limited mobility are also extremely disruptive and often make it impossible for hemophiliacs to go to school or take up employment. Many parents of hemophiliacs choose home tutoring but this is available only to those who can afford it.

Bleeding is also accompanied by severe pain.

b. Psychological challenges

The psychological effects of hemophilia affect both hemophiliacs and their families. Often unable to attend school and find jobs, hemophiliacs may develop problems of social immaturity and isolation. They are also stigmatized and subject to social discrimination.

Close relatives often have to quit their jobs to look after hemophiliacs at home and this represents a heavy family and psychological burden.

Moreover, as symptoms are irregular, hemophiliacs live in a perpetual state of panic and anxiety as they don't know when to expect their next bleeding.

5. Treatment and care

Clotting factor concentrate is the substance, developed in the 1960s and 1970s, with which hemophiliacs are treated nowadays. It comes in little bottles and is freeze dried under sterile procedures. It therefore can be stored at home in the refrigerator and reconstituted with a small amount of sterile water.

Before the introduction of clotting factor concentrate, hemophiliacs ran to the hospital for emergency treatment whenever a bleeding occurred. They often faced the problem of having to convince the nurses of the urgency of the situation. There has always been much ignorance about hemophilia and little attention has been devoted to it in hospitals due to the low incidence of the disease and to a lack of public awareness. Patients often had to wait until they

were taken to the emergency room and this often took place when the bleeding had reached a critical state. In other cases, hospitals didn't know how to treat hemophilia patients.

Clotting factor concentrate meant the possibility of home treatment for hemophilia. It could be stored at home and this saved many emergency visits to the hospital, days lost from school or work and unnecessary bleeding complications as a result of time wasted from home to the emergency room. In other words, it made it easier to catch early bleedings in time and prevent the worsening of hemorrhages.

II. Chinese hemophiliacs

1. The problem of HIV, Hepatitis infection and compensation

China is estimated to have around 70,000 to 120,000 hemophiliacs. According to recent studies, there is a 3.5% (n=2/57) prevalence of HIV blood borne infection and a 49.1% (n=28/57) prevalence of Hepatitis virus among hemophiliacs². However, this study also reveals that only half of the hemophilia population in China has received testing for these diseases, thus making it impossible to give an accurate estimation of the exact number of infections.

It was in June 1983 that the United States Centre for Disease Control (CDC) announced that AIDS was a blood borne virus. In 1985, the first screening of HIV antibodies became available and this marked the beginning of blood supply screening in the United States. Later that year, it was suggested that laws on blood screenings be expanded to blood products and factor concentrates³.

It was also in 1985 that the Chinese Ministry of Health banned the import of blood products to prevent the spread of HIV. In 1987, Article 2 of the Provisions for the Monitoring and Control of AIDS stated that all blood products should undergo testing for HIV and that HIV carriers should not donate blood, tissue, organs or seminal fluids. In 1995, the Ministry of Health announced that all blood products produced before 31 December 1994 had high probabilities of being infected and should be immediately discarded. In 1998, the Ministry of Health requested all hospitals to keep records of blood donations and transfusions.

Treatment for hemophiliacs demands frequent blood transfusions and use of blood products and the lack of blood control previous to 1995 resulted in many cases of hemophiliacs infected with Hepatitis and HIV/AIDS.

Before 1995, the Shanghai Institute of Biological Products, the biggest center of production of the factor VIII, was responsible for producing and selling not reliably disinfected blood products. After the 1995 government regulation, the SIBP not only failed to inform its clients of the danger of using its products but also continued to sell them, thereby causing many more chain transmissions of HIV/AIDS and Hepatitis from hemophiliacs to their families. In spite of this, lawsuits filed against the SIBP have been dismissed for lack of evidence that infection occurred before 1995.

² Zhang L, Li H, Zhao H, Zhang X, Ji L, Yang R. Retrospective analysis of 1312 patients with haemophilia and related disorders in a single Chinese institute. *Haemophilia* 2003; **9(6)**:696-702.

³ Baral, Stefan, "Case Histories of Hemophiliacs infected with Bloodborne Viruses", manuscript

From 1987 to 1995, Gao, a hemophiliac, received transfusions from two government hospitals. He was diagnosed with HIV in 2002. He filed a lawsuit but failed to provide a record with evidence of these transfusions (since rules regarding compulsory medical records only came into effect after 1998). His case was dismissed and soon after Gao died. But Gao is only one among hundreds. Courts often refuse to hear litigations for compensation, and when they do, victims often have to wait too long for the trial. In other occasions, the justice fails to apply the sentence after the issue of the verdict. Moreover, hemophiliacs often refer to the indifference and brutality of court officials.

According to Article 106 of the General Principles of Civil Law of the People's Republic of China: *Anyone who contracts HIV/AIDS via blood transfusions in hospital with fault should be compensated by the hospital for damage and health violation. Any court house should accept their litigation and support their claims strictly based on what the law stipulates.* This is in clear contradiction with reality.

Hemophiliacs who continue fighting for rights and compensation often meet with government's suppression. On 24 October 2006, three hemophiliacs were arrested in front of a hospital in Shanghai. The following day, the well known leader of the Chinese Hemophilia Association Kong Delin was detained by the Shanghai authorities. All of them had recently raised issues regarding the protection of the rights of hemophiliacs. Kong Delin was released on 30 November, 36 days after his detention, but the remaining three hemophiliacs are still missing and no clue has been provided as to their whereabouts and health condition.

Since 2001, the Shanghai municipal government has offered compensation to hemophiliacs infected by products sold at the Shanghai Institute of Biological Products. They now have free access to HIV screening, treatment, factor VIII, 1000 RMB monthly subsidy as well as 10,000 RMB compensation for those hemophiliacs suffering from HIV/AIDS. Moreover, relevant departments have carried out a systematic follow up of the situation of the sufferers. However, while blood products produced by the Shanghai Institute of Biological Products were used by people from all over the country, this compensation was only made available to Shanghai residents.

3. Treatment for HIV and Hepatitis

In 2004, the Chinese government, in an effort to control the AIDS epidemic, implemented the national policy of "Four Free One Care". This meant free ARV (antiretroviral) drugs, free prevention of mother to child transmission, free voluntary counseling and testing, free schooling for children orphaned by AIDS and care for poor families with one of its members living with HIV.

However, this has not solved the problems of hemophiliacs. On the one hand, many hemophiliacs are also infected with Hepatitis C but no free treatment is provided for this disease. Besides, first line anti-retroviral therapy for HIV has strong negative side effects on individuals suffering from Hepatitis C and expensive or unavailable second and third generation medication

is needed.

4. Hemophilia treatment and insurance

If provided with adequate treatment, hemophiliacs may lead a life almost as normal as healthy people. The problem is that many hemophiliacs in China nowadays do not receive adequate or timely treatment. This leads to permanent physical disabilities which result in unemployment, difficulties to get married and high death rates, especially in rural areas.

Many places have basic health insurance policies in China but it is often hard for hemophiliacs to benefit from them since they are neither retired nor employed and therefore do not qualify for the package. Moreover, the most common form of hospital treatment for hemophiliacs is emergency treatment but in some places this is not covered by health insurance. Another complication is that children's insurance is provided by private companies in China but no insurance company is willing to accept children suffering from hemophilia.

Hemophiliacs have an average of 10 to 20 hospitalizations per year, each costing 2000 Yuan on average. Besides, the price of factor concentrate is above 300 Yuan per bottle, which puts a heavy burden on hemophiliac families. They receive no subsidy for these costs.

5. Hemophiliacs association

The Hemophilia Home of China describes itself as "a website that has become a space for mutual support and help. It is also a way for hemophiliacs to be understood by the outside world." The website, with contents both in English and Chinese, was created in 2000 and can be logged on to at www.xueyou.com.

The tasks of the "China Hemophiliac Association", a non profit organization founded by Chu Yuguang, Kong Delin, Guan Tao and other Chinese hemophiliacs are to register patients, organize activities, publicize information on hemophilia, seek donations and attract the government.

Both prove that many efforts have been made recently to join efforts to establish a common network for Chinese hemophiliacs, a place to share information, knowledge and experience as well as a powerful platform to raise public awareness and bring attention to a community which has been largely ignored.

6. Appeal

The cost of hemophilia treatment, medical insurance, HIV/HBV/HCV infection and compensation, physical, psychological and economic challenges, the need to set up a capacity building mechanism are in a nutshell some of the main difficulties and unsolved problems which Chinese hemophiliacs have to face. So far, a conference on "Blood Safety, AIDS, Law and Human Rights" was held in Beijing in 2005 and addressed all these issues. The second conference, scheduled to be held at the end of November 2006, was suppressed by police authorities and ended up with the three day detention of Wan Yanhai, leader of Beijing Aizhixing Institute for Health Education and responsible for organizing the conference.

The problems faced by hemophiliacs are many but the hemophilic population in China is limited and therefore these problems can be effectively addressed. It is not only necessary but also realistic to call for immediate action and request the help and cooperation of Chinese government departments, international institutions and organizations, private donors and the average individual to open their eyes and hold out their hands to the hemophilic community.

The most pressing issues which need to be addressed are as follows:

1. Target the particularities of the hemophilic community, consult other countries' practices and include those individuals with no access to clotting factor within the bounds of handicapped people.
2. Bring hemophiliacs into the social security system.
3. Establish strategic reserves and import clotting factor so that hemophiliacs, under crisis periods, can obtain clotting factor, and thereby protect their lives.
4. Establish a blood transfusion and blood product safety system to guarantee the safety of hemophiliacs using blood and blood products.
5. Raise social awareness and pay special attention to hemophiliacs, who, because of continuous use of blood products, are under a higher risk of contracting HIV/AIDS, HCV, and other viruses.
6. Timely treat and assist hemophiliacs who have contracted HIV/AIDS and hepatitis C as a result of using blood products.
7. Fulfill the obligation, along with relevant state departments, to inform society about the risks of contracting HIV/AIDS via blood or blood products. In addition, provide free testing for patients who have used blood or blood products and for their relatives and dependants, investigate the situation of hemophiliacs contracting HIV and seek liability. Create a medical treatment express path for hemophiliacs; solve hemophiliacs' general hospitalization problems.
 - a. Set up a full time polyclinic doctor to direct treatment for hemophiliacs
 - b. Supply public hospitals with clotting factor
8. Advise the government to expedite the introduction of suitable alternative medications for hemophiliacs with HIV/AIDS since the current range of medications available is very limited and produces many side effects.

