



Hemophilia and Thrombosis Center of Nevada

A Non-profit Center

AY 2001

SPRING NEWSLETTER M

Ricky Ray Trust Fund Update



The Ricky Ray Trust Fund received \$580 million for fiscal year 2001. Combined with the \$75 million received in fiscal year 2000, all petitions received to date will be paid. As of early February, 1800 payments distributed more than \$145 million. The office has received nearly 5600 petitions. At the current review rate, the office anticipates completing processing and payment on about 300 petitions per month.

The funds are available to anyone from the bleeding community who acquired HIV through treatment with factor concentrate. The petition must be completed by the patient, or surviving family member and a portion must be completed by the patient's doctor to confirm the diagnosis.

In order to expedite your petition and payment, there are several recommendations.

First, make sure the petition is complete. The office has received many incomplete petitions that delay payment. If you plan on sending additional or amended information, be sure to include your Ricky Ray file number on all documents. If your bank has changed or merged or you have moved or changed accounts, send a new payment form to the

Program Office to ensure payment. The payment and the petition form can be printed from the Program Office website at <http://bhpr.hrsa.gov/rickyray> or by calling the office at 1-888-496-0338. Information will continue to be updated on their website.

Inside this issue:

Message from India	2
Gene Therapy Trial Available	3
Questions and Answers	3
Acquired Hemophilia	4
Blood Test Results	5
Hepatitis C Treatment	5
Health Fair	6
Port Care	7
AB Direct Program	7

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REPORT FROM EARTHQUAKE AFFECTED RAJKOT CHAPTER AREA

Rajkot is located 250 km from the epicenter of the January 26th earthquake which registered richter scale 7.2. Chapter key persons helped to locate affected members and assess the damage.

Four families of Bhuj were missing after four weeks. It was hoped that they were with relatives, as they were not on any of the list of deceased persons filed with the police. The Gabu family were affected by the earthquake as they live in a remote village called Morsal, 82 km from Rajkot. They have three affected children between the ages of 8 and 14. All have severe hemophilia A. Because of recent drought conditions, they have had problems with crops and live below the poverty line. One of the walls of their house collapsed and major cracks were present in the remaining wall. One of his sons developed an intracranial hemorrhage and now has blurred vision. He needs surgery. One son is limping, however the family cannot afford to purchase crutches. Because of the damage due to the earthquake, many villagers must sleep in the open air.

Other families living in Rajkot had other difficulties because of the earthquake. Schools had not reopened and teachers had not reported to duty. Recurring mild tremors cause concern regarding the existing shelter.

Mr. Jayesh, a hemophiliac from the village of Morbi was studying engineering at Bhuj. On January 25, he came home to visit family. On January 26, his college hostel collapsed with twenty classmates buried under the rubble and debris. Because of the psychological stress, he refuses to go back to school when it finally re-opens.

I was amazed to note that most families

logic prediction. On January 13, an astrologer, Ambadas Patel, predicted in the leading dailies that due to most of the planets coming into alignment there would be a natural calamity between February 14, and 20. It was surprising to note that educated people also believe in these myths.

Sonal and Paresh Patel, have formed a women's group that makes embroidered wall hangings, tea cozys, and cushion covers in typical kathiawadi art. They plan to market these articles domestically and internationally in order to become economically self reliant. This group has brought a silver lining to difficult times.

I met with Dr. Ashwin Patel and was happy to know that there was no injury or house collapse to their members. However, anxiety runs high regarding the risk of future calamities. They expressed anger at the lack of empathy from the government quarters. Board examinations are taking place as scheduled but lower class examinations have not been scheduled. Children have trouble concentrating on their studies or being able to play.



Gene Therapy Study for Severe Hemophilia A

GemStar Therapeutics in conjunction with Baxter Healthcare Corporation is currently conducting a gene therapy research study for patients with severe hemophilia A. This study is being conducted at the University of North Carolina in Chapel Hill and the University of Washington in Seattle. This is a Phase I study of a MiniAd FVIII vector. A Phase I study evaluates the safety of a medication at different dosages. This study will also evaluate the ability of an intravenous infusion of a gene transfer vector to produce Factor VIII in the patient.

This study is open to nine adult patients over the age of 18 who have severe hemophilia (factor VIII level less than 1%). The patients must have no history of inhibitor or Hepatitis C virus.

The hope of gene therapy is to be able to insert a piece of nuclear material into the person with hemophilia. This piece of material will be able to incorporate into the patient's cells and produce factor that will be released into the patient's blood stream. If this is possible, additional infusion of factor concentrate may no longer be needed.

Gene therapy is very new and the risks involved are not yet known. However it holds promise for a cure for genetic diseases such as hemophilia.

If you or someone you know is interested in participating in this study and are able to travel to Seattle or Chapel Hill for treatment, please contact the treatment center for additional information and referral.

Questions and Answers

I have been told that I am anemic my entire life. Even though I have been taking a multivitamin with iron, it does not seem to help. Is there anything else that I can do to build up my blood?

Anemia is a common disorder where the blood count, measured as a Hemoglobin (Hb), is lower than the normal 12. There are many reasons for anemia and not all are caused by lack of iron in the body. Normally, the body reuses iron so that unless there is any bleeding or inability to absorb iron through diet, iron deficiency anemia does not occur. In persons with bleeding, either from the stomach and intestines, or through loss with periods in women, iron deficiency may occur. This can be tested by a blood count, and storage iron test known as ferritin level.

If the ferritin is low, iron may be replaced orally, by muscle injection, or by intravenous infusion. Usually the administration by vein is limited to persons who cannot tolerate the oral form of iron due to severe stomach intolerance or for treatment-

severe anemia in an effort to avoid the need for a blood transfusion. It is important that the correct oral dosage of iron be administered. It is recommended that Ferrous Sulfate, available without prescription, be administered three times daily. Because of the need for three daily dosages, it is easy to forget and therefore receive inadequate supplementation. An oral preparation that is a slow release iron, called SloFe is available and may be better tolerated and easier to administer since it is given only once daily.

If oral iron is inadequate or not tolerated, a single intravenous iron dextran dosage may be administered. Because of the risk of allergic reactions, a test dose under the supervision of a trained physician must be administered first. If this is tolerated, then the infusion may be administered. The dosage is calculated based upon the weight of the patient and the Hb level.

If your blood count is not normalized within several weeks after beginning iron supplementation, then further evaluation by a hematologist may be needed.

GET WELL WISHES TO:

ANNISE BULLOCK

Help us in wishing a speedy recovery to Annise Bullock, our laboratory supervisor. We're all looking forward to her return.

GOOD SAMARITAN:

DEREK WASHINGTON

The Good Samaritan Award goes to Derek Washington. Derek is a patient with an inhibitor on immune tolerance therapy. The therapy has been so successful that he no longer has any presence of the inhibitor and no longer needs activated factor concentrate to treat bleeds. After reading about the needs of patients in India, he generously donated several doses of FEIBA and Autoplex. The factor has been sent to a stewardess who regularly goes to India for delivery of this life saving medication to patients in India. Without this needed donation of factor concentrate, these patients may not receive any treatment.
Good Job Derek!!

CONGRATULATIONS TO:

MONA AND JOYEN VAKIL

Congratulations on the upcoming birth of their first child. Joyen is one of our patients, and his wife Mona has been our treatment center's physical therapist for the past two years. We wish them much luck and happiness.



Focus on Acquired Hemophilia

Many persons are born with bleeding disorders. How would you feel if you were in excellent health until one day you developed bruising and bleeding out of the blue. It is possible for persons who are in good health to develop an antibody that attacks the clotting elements in the body resulting in hemophilia. Often these patients require hospitalization to control life threatening bleeds. Because these patients have an antibody, they are not able to be treated with the usual factor concentrates administered to patients with inherited hemophilia as they will not work. Therefore, factor concentrates used to treat hemophilia patients with inhibitors are more effective.

While acquired hemophilia is an uncommon condition, we usually see several patients with this disorder each year. In contrast to inherited hemophilia, it may affect both women and men at any age. The treatment involves infusion of factor concentrate to treat bleeding complications while using immunosuppressive medication such as steroids to eliminate the antibody. Once the antibody is eliminated, the hemophilia goes away.

This condition, in contrast to the other bleeding conditions does not run in families. It may occur in cancer patients, patients with rheumatoid arthritis, associated with pregnancy, or for no reason at all. Because it is an unusual condition, the diagnosis may be delayed and the patient suffer complications before treatment is initiated.

Why We're So Picky About How Your Blood Tests are Handled

by Annise Bullock

Blood coagulates if left to sit for any period of time. This is because normal clotting elements are present in the blood to ensure that bleeding

"It is best that blood sample be drawn early in the morning during the week so that they may be transported to the specialty lab without delay."

will not continue. This is the wonderful way that your body protects you from injury. However, it poses a problem when we test a persons blood to see if clotting is

normal. If the blood is not drawn through the correct sized needle, if the person or room is cold, if the blood is not immediately put into the correct test tube, or if the blood is not put into a centrifuge to separate it's components, the blood will clot and an analysis will not be accurate. If all of these steps are correctly followed, then the blood must be carefully handled and kept cool until the analysis is complete. Therefore, it is important that the person who draws the blood and

prepares it before it goes to the laboratory for analysis be adept at managing this important task. When any of these steps are not done precisely, the laboratory testing will be inadequate and need to be repeated.

After the blood is obtained, spun, and prepared, it is analyzed. The analysis is best done in a laboratory specialized to do clotting testing. Two such laboratories on the west coast are located in Denver and Phoenix. Because all blood must be transported to the laboratory, drawing blood samples on a Friday may be problematic. It is best that blood samples be drawn early in the morning during the week so that they may be transported to the specialty lab without delay. With planned fund raising events at the HTCN, we hope to be able to purchase the necessary equipment to do clotting testing at our center. This will mean that the test results will be available the same day rather than waiting the two weeks it currently takes to obtain results. This will make it much easier for the diagnosis and treatment of all persons in southern Nevada who may require clotting testing.

New Agent for Hepatitis C by Dr. Rinah Shopnick

A majority of patients with bleeding disorders are infected with hepatitis C due to the use of contaminated blood products in years before sensitive testing was available. While many patients do not know if they are infected unless they receive screening testing, after 10-20 years of infection, continued liver injury by the virus may result in liver failure, cirrhosis, and even progress to liver cancer. Therefore, treatment to irradiate the virus is important. Unfortu-

nately, treatment is often expensive, associated with many side affects, and only effective in a minority of patients. A new agent, a long acting interferon (PEG Interferon), shows promise in improving the percentage of patients who respond to treatment. In the past, treatment with interferon injections had small response. With the addition of an oral medication, ribavirin, an improved response has been seen. Because PEG Interferon lasts in the system for longer pe-

riods of time, it may be more effective. In patients with hepatitis C, the use of PEG Interferon resulted in an improved response with complete irradiation of the virus. Whether this agent in addition to Ribavirin will improve the cure rate even more is not yet known.

PEG Interferon is associated with many of the side affects seen with Interferon. Because interferon is a natural substance that is released into the body in

Continued on Page 6

Health Fair

The HTCN is hoping to increase awareness of bleeding and clotting disorders. To do this, we have participated in several health fairs in Clark County. The first, a women's health fair, was not lucky enough to have good weather. Due to strong winds, we were forced to pack up early. The next scheduled health fair will be held on May 30 in Sun City, Summerlin. This fair is open to the public and is one of the largest health fairs in southern Nevada. It is organized by the HTCN's athletic trainer Jennifer Wells. The HTCN will participate by holding a booth with information about bleeding disorders, available studies, and will offer free coumadin testing. Come out to see us at the fair.



Hepatitis C *continued from page 5*

response to viral infections, it accounts for much of the muscle aches, fatigue, and malaise that is often associated with the flu. This is the biggest side effect seen with Interferon. Because PEG Interferon lasts in the system much longer, it may be administered as a shot once weekly instead of the daily or three times weekly injections prescribed with regular interferon. Interferon may decrease the ability of the bone marrow to produce blood cells. Therefore, it must be monitored closely. With care, the side effects can be easily managed in most patients. The best thing anyone can do is to be tested for the presence of hepatitis C before there is any sign of liver damage. This can be done by a blood test. If the presence of antibodies are demonstrated on the blood test, then a follow up blood test to detect whether and how much virus is present is needed. During treatment, this value is monitored to make sure that no virus is present after treatment. Anyone with signs of liver disease on blood testing may benefit from treatment. The options should at least be discussed with your doctor. If you do not opt for treatment at this time, make sure that you continue to be monitored on a regular basis. Much research is being done in this area and improved treatments are expected.

AB Choice Launched

Aventis Behring, a manufacturer of factor concentrates, and Stimate nasal spray, has announced a new program to provide customized access to resources and educational services for members of the bleeding community. The access is available to anyone regardless of the type or manufacturer of the factor concentrate used.

The program is centered around four features:

- Open Distribution
- Educational Resources
- Customized Support
- and a foundation for Research and Advancement of Patient Health.

The program provides educational information including patient diaries, a newsletter, events, and educational materials. Grants for financial assistance are available for persons who qualify.

For more information or to register for this program, log onto the web site at www.aventisbehringchoice.com

Port Care

by Myra Davis-Alston

Port-A-Cath or “ports” make it easier to administer intravenous medication or to withdraw blood samples for laboratory testing. A port is a small device that is placed under the skin either in the chest or the forearm. A catheter which is attached to the port tunnels into a large vein in the chest. There are several types and designs of ports. They may be inserted by a surgeon in the operating room or by a radiologist in a special procedures room. The type and design of the port is selected based upon therapy requirements, body size, and the location desired. There are both single and double lumen ports. The lumen refers to the soft portion in the center of each port. This is the area where the needle is inserted. A double lumen port has two areas for insertion of needles, therefore two IV’s can run at the same time.

Care of the port is important. The two most common problems that may occur are infection of the port with possible infection into the blood stream, and clotting of the port catheter. Routine port care is designed to prevent these problems from occurring. Ports are accessed with a special needle to prevent damage to the septum. They should be flushed with heparin at least once every other month to prevent the blood within the lumen from clotting. If resistance is felt when pushing the syringe, the catheter may be clotted. If this occurs, you should notify the treatment center as soon as possible. If a clot is identified promptly, it may be dissolved with special medication called tPA inserted into the lumen.

The more the port is accessed, meaning needle inserted into the lumen, the greater the risk of infection. It is important that sterile technique be used at all time when dressing or flushing a port to decrease the likelihood that infection will be introduced. If a port is used frequently, often the needle access will be kept in place for up to one week. If this is the case, a sterile dressing must be kept over the needle entry area. This helps prevent infection. Transparent dressings are recommended so that any irritation around the needle can be seen. The dressing and needle is changed once a week. If the dress-

ing pulls away from the skin, bacteria can get into the area. To prevent infection, the dressing should be changed as soon as possible. Early signs of infection include fever, chills, pain, redness, swelling, or drainage at the site. The single most important thing that can be done to prevent port infection is good hand washing before and after port care. Sterile technique at the port access site is also important to preventing infection. Sterile dressing change kits are available from home care companies. Many patients are taught how to access their own ports. With the proper care, ports can be used for many years.

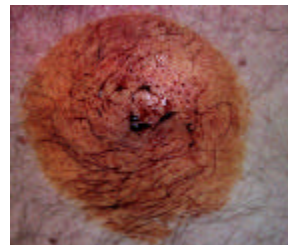
Other types of intravenous access devices such as PICC lines will be discussed in future newsletters.



The Port



Swab with Betadine



Clean with alcohol



Needle Access

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