



MEDICAL NEWS

April 1, 2004**Study Looks at Risk Factors for Range-of-Motion Limitations In Young Males with Hemophilia**

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A paper in the April issue of *Blood* detailed research on the prevalence and risk factors for joint range of motion (ROM) limitation among young males with hemophilia. Researchers collected data from more than 4,000 males with hemophilia between the ages of 2 and 19 who received care at 136 hemophilia treatment centers. ROM measurements were obtained on hips, knees, shoulders, elbows and ankles. These measurements, taken into account with several other factors, like age, race, hemophilia type, prophylaxis use and body mass index (BMI), allowed the researchers to see that in all disease severity groups, ROM limitation was greater for patients who were older, were non white and those with a greater BMI. For those with severe hemophilia, ROM limitation increased with the number of bleeds and the presence of inhibitors or recent orthopedic procedures. Those with factor VIII deficiency had a greater degree of ROM limitation than those who were factor IX deficient. The study's analysis focused on ROM limitation that was accompanied by asymmetric joint mobility. "We conclude that ROM limitations begin at an early age, especially for those with severe and moderate disease, and that BMI is an important, potentially modifiable risk factor," say the authors. The authors note that current health recommendations for people with hemophilia include regular exercise using low-impact activities to increase muscle tone and strengthen joints. "An additional component that may be considered is routine monitoring of BMI," the paper says.

The data used in the study was gathered from participants in a voluntary surveillance program called the University Data Collection project, which has been supported since 1998 by the Hematologic Diseases Branch of the Centers for Disease Control and Prevention. The study, authored by J. Michael Soucie, Christy Cianfrini, Robert Janco, Roshni Kulkarni, Julie Hambleton, Bruce Evatt, Angela Forsyth, Sue Geraghty, Keith Hoots, Tom Abshire, Randall Curtis, Ann Forsberg, Heather Huszti, Margaret Wagner and Gilbert White, can be found in *Blood* (103[7]:2467-2473).

March 28, 2004**Experts Team Up During National DVT Awareness Month**

On March 27, 2004, the National Alliance for Thrombosis and Thrombophilia (NATT) hosted an educational seminar in Denver to help patients better under-

stand risk factors, warning signs and treatments for debilitating blood clots. March is National Deep-Vein Thrombosis Awareness Month. In a NATT release, Marilyn Manco-Johnson, MD, the director of the Mountain States Regional Hemophilia and Thrombosis Center, said, "Blood clot prevention is much easier than treating them after they've developed. As many as 80% of patients with this condition experience life-threatening complications when clots reach the lungs, often without any warning signs. That is why patient education is the key in fighting this disease." Manco-Johnson was one of four other experts to speak.

Source: NATT release.

March 24, 2004

A Paradox Explains How von Willebrand Factor Works

A researcher at The Scripps Research Institute has found that "Type IIB is a variant form of von Willebrand disease in which a structural abnormality of von Willbrand factor (VWF) causes enhanced binding to the platelet glycoprotein IB receptor." In addition, "As a consequence of this functional alteration, there is a decrease in the concentration of the largest VWF multimers in plasma, and the platelet count may be episodically decreased as

a consequence of microaggregation." In *Biotech Week*, the author of the study, Zaverio Ruggeri, says, "The net result is an apparent paradox, since the presence of hyperfunctional adhesive molecule in blood causes a bleeding tendency." The study is published in the *Journal of Thrombosis and Haemostasis* (2004; 2(1):2-6).

Source: *Biotech Week*.

March 22, 2004

DNA Chip Can Detect HIV, HBV and HCV in Blood

Scientists at the Chinese Academy of Science have developed a DNA chip that enables simultaneous detection of HIV, the hepatitis B virus and the hepatitis C virus. The chip was developed by combining multiplex and nested polymerase chain reaction (PCR) with arrayed anchored primer PCR and a biotin-avidin alkaline phosphatase indicator system. The study appears in *Biosensors & Bioelectronics* (19(7):685-692).

Source: *Hepatitis Weekly*.

March 20, 2004

Peptide with Potential to Inhibit SARS Identified

Shibo Jiang, MD, PhD, and colleagues at the New York Blood Center have identified a peptide derived from the severe acute respiratory syndrome (SARS)-associated coronavirus (SARS-CoV), the virus that causes SARS. This peptide, which has inhibitory activity against SARS-CoV, was discovered using the same approach used when a potent anti-HIV peptide was discovered ten years ago. That anti-HIV peptide discovery led to the development of the first in a new class of anti-HIV drugs, HIV fusion inhibitors. Jiang compared the amino acid sequences of SARS-CoV spike protein with the HIV protein discovered earlier and noticed that they shared many common features. Using this information, the researchers will design and synthesize more potent anti-SARS-CoV peptides to develop drugs for the treatment of SARS that could be used to protect those working and traveling in SARS endemic areas. Jiang et al.'s current study appears in the March 29, 2004, *The Lancet* (363:938-947).

Source: *US Newswire*.

March 18, 2004**Noninvasive HCV Test Developed to Assess Liver Status**

LabCorp announced today the availability of FibroSURE, a noninvasive blood test for assessing liver status in patients with the hepatitis C virus (HCV). The test, developed in France, is designed to assess liver fibrosis and necroinflammatory activity. According to a company release, HCV FibroSure is recommended for use in assessing liver status following a diagnosis of HCV, as a baseline determination of liver status before initiating HCV therapy, as post-treatment assessment of liver status six months after therapy and for noninvasive assessment of liver status in patients at risk for complications from a liver biopsy.

BioPredictive, the Paris University-based laboratory that developed the test, is currently researching clinical use of FibroSURE for other disease populations, including hepatitis B, HIV-HCV and alcoholic and non-alcoholic steato hepatitis.

The test has poor performance characteristics in mid-ranges of fibrosis and adds little in subjects with clinical/laboratory evidence of cirrhosis. Its utility in the hemophilia population has yet to be determined.

Source: Company release.

March 8, 2004**New At-Home Device Would Check Blood Clotting Readings**

Scientists at Drexel University's School of Biomedical Engineering are developing a device that would allow patients on blood-thinning medication to take their own readings at home, much like the machines used by diabetics to check blood glucose levels. The researchers say their device would work by reading the amounts of specific kinds of proteins in the blood that are involved in clotting. Similar devices that are currently available measure the blood's thickness by how quickly it works its way through a mazelike structure.

Source: The Associated Press.

March 3, 2004**Some Anti-Depressants Can Cause Internal Bleeding**

According to the United Kingdom's Drug and Therapeutic Bulletin, a group of anti-depressants called selective serotonin re-uptake inhibitors (SSRIs) may predispose high-risk patients, like those who've previously suffered from gastrointestinal bleeding and the elderly, to internal bleeding. Researchers from the Consumer Association reviewed three studies that evaluated patients on SSRIs, which includes Prozac. The study found that those on SSRIs were more likely to suffer intestinal bleeding than those on other anti-depressants and reports have previously suggested a link between the drugs and easy bruising, nosebleeds and other hemorrhages. While the UK Department of Health suggests providing clearer warnings with the drug, researchers say doctors should avoid prescribing SSRIs to those over 80 years old and those who have a history of gastrointestinal bleeding.

Sources: The British Broadcasting Corporation and The Press Association.

March 2, 2004**Bayer Receives Approval for HIV Test To Be Used In Plasma Donations**

Bayer has announced US Food and Drug Administration approval for in-house nucleic acid testing for HIV in plasma donations. According to a Bayer release, this test will more directly ensure the integrity and efficiency of the testing process and the resulting safety of its plasma products prior to the manufacturing process. Using study data collected by Bayer, Roche Diagnostics received the approval for the COBAS AmpliScreen test.

Source: Bayer HealthCare Biological Products Division.

March 1, 2004**West Nile Virus Transmissions Thwarted by Blood Screening Procedure**

At the International Conference on Emerging Infectious Diseases, the Centers for Disease Control and Prevention (CDC) announced that up to 1,033 blood donors carrying a West Nile infection were identified through a screening process for the virus from late June through November 2003. According to a CDC researcher, "The number of cases of West Nile virus found in the blood supply is not surprising in that most

people who are infected through mosquito transmission never have symptoms." The screening test has helped health officials map the spread of the disease. Another CDC official told attendees, "The entire blood community went from the identification of a problem in the blood supply to the development and implementation of a solution nationwide within a year."

Source: United Press International.

March 1, 2004**Drug Combo Shows Highest Success Rates for HCV**

The leader of the largest study on patients with HIV and hepatitis C virus (HCV) coinfection announced this month a drug combination that showed the highest treatment response rates for HCV ever reported. The multinational study found that the combination of Pegasys (peginterferon alfa-2a) and Copegus (ribavirin) were much more effective than previous generations of hepatitis C therapy (which included standard interferon and ribavirin) and could treat patients for their HCV without compromising their HIV status. The study treated 868 HIV/HCV patients in 19 countries. Efficacy was defined by the absence of detectable HCV RNA for at least six months after treatment.

Source: Health and Medicine Week.

February 23, 2004**Study Shows Anti-HCV Therapy Should Be a Priority in HIV/HCV Patients**

Researchers in Spain recently published a study in *Clinical Infectious Diseases* detailing risk factors for severe liver fibrosis in patients with HIV and the hepatitis C virus (HCV). The investigation, which gave liver biopsies to 914 patients who had HIV, HCV and elevated serum alanine aminotransferase (ALT) levels, discovered that severe liver fibrosis was frequently found in these patients and that its severity increased with age. The study noted that the use of antiretroviral therapy was not associated with the severity of the fibrosis. According to one of the study's authors, "The rate of complications due to end-stage liver disease will inevitably increase in this population, for whom anti-HCV therapy should be considered a priority."

The study can be found "Incidence and Predictors of Severe Liver Fibrosis in Human Immunodeficiency Virusinfected Patients with Chronic Hepatitis C: A European Collaborative Study. " *Clinical Infectious Diseases*. 2004;38(1):128-133.

Source: *Hepatitis Weekly*.

LEGISLATIVE NEWS

March 29, 2004

CDC Issues New Hepatitis C Resource

The Centers for Disease Control and Prevention (CDC) has issued a new Hepatitis C pamphlet, *Living with Chronic Hepatitis C*. The pamphlet addresses questions that a newly infected person with hepatitis C might ask, including: "What is hepatitis C?" "How can a person get it?" and "What can be done to prevent passing it to others?" The pamphlet directs the reader to extensive information about the virus found at www.cdc.gov/hepatitis or by calling the Hepatitis Information Line at 1-888-4HEPCDC (1-888-443-7232).

March 16, 2004

USDA to Expand Testing For Mad Cow Disease

On March 16, 2004, the United States Department of Agriculture (USDA) announced it will greatly expand the number of cattle tested for bovine spongiform encephalopathy, also known as Mad Cow Disease.

The agency intends to spend an additional \$70 million to test as many as 268,000 animals a year, up from the current number of 40,000, to determine how prevalent Mad Cow Disease is in the American herd. The new testing will concentrate on cows most likely to be sick, and USDA believes the new testing has the statistical capacity to find an infected cow even if the incidence is as low as one in 10 million.

Until this announcement, USDA has maintained that the testing of 40,000 animals is sufficient to assure that mad cow disease is not a danger; however, at a briefing before the Congressional Biomedical Research Caucus on March 17, 2004, key scientist Stanley Prusiner recommended that the US follow Japanese practices on Mad Cow test-

ing. Prusiner is the University of California scientist who discovered prions, the infectious agent that causes Mad Cow Disease and its human form, known as new variant Creutzfeldt-Jakob disease (nvCJD).

Prusiner emphasized that the US should test every cow intended for human consumption for Mad Cow Disease. He called Mad Cow Disease "the greatest threat to the safety of the human food supply in modern times," and said its variant, nvCJD, "threatens the safety of the blood supply worldwide."

The expanded program is expected to begin in June 2004 and will last 12 to 18 months. USDA said it will test as many at-risk animals as possible during the 18-month period and expects to reach between 220,000 and 268,000 animals.

Under the expanded USDA plan, most of the animals tested would still be "downers," those that are unable to walk to slaughter. Downers are considered to be at highest risk of having the disease, which destroys brain tissue and leaves animals unable to walk or stand.

About 20,000 apparently healthy animals will be sampled from the 40 US plants that handle most of the older cattle slaughtered for human consumption. Older ani-

mals are at greater risk of having the disease because they are more likely to have eaten feed containing the ground-up beef tissue that was widely fed to cattle until 1997. The misfolded proteins that cause Mad Cow Disease are thought to be spread through such feed.

Prusiner reports that prions from cattle meat can infect humans. The body has no defense mechanisms for prions and no medicines have been invented to address the disease. Prusiner believes the feeding of body parts of cattle to chickens should be stopped because chicken manure is fed to cattle and can spread the disease. He also feels that even if feeding practices are changed, the testing of all cattle is necessary because the prions that cause brain-wasting diseases can form spontaneously.

The March 16, 2004, announcement relied heavily on the review of USDA's Mad Cow prevention strategies conducted by an international panel of veterinary experts. That review concluded that there was a "high probability" of additional Mad Cow cases in the United States, and it recommended significantly expanded testing, tougher food and feed regulations and a ban on using farm animal remains in cattle food

March 16, 2004

2004 Washington Days Program A Success

More than 140 members of the bleeding disorders community met March 10 to 12, 2004, for NHF's Annual Legislative Advocacy Conference in Washington, DC. One hundred twenty-six Congressional office visits were scheduled and 37 Congressional members met with NHF participants.

The program began Wednesday, March 10, with an evening reception and issue briefings for the following day's Congressional visits. Thursday began with the presentations of the Dr. L. Michael Kuhn Award to Congresswoman Carolyn McCarthy (D-NY) and Congressman Patrick Kennedy (D-RI) for their dedication to NHF and their leadership over the past year. Congressional Staff Appreciation Awards were also presented to Joel White of the House Ways and Means Committee and Charles Clapton of the House Energy and Commerce Committee for their work on Medicare reimbursement for clotting factor.

Members of the bleeding disorders community spent the day discussing hepatitis C prevention and research, women's bleeding disorders and health insurance reform with those in Congressional offices. The community successfully lobbied for seven additional members to cosponsor the Hepatitis C Epidemic Control and Prevention Act and twelve additional cosponsors to House Concurrent Resolution 314 on women's bleeding disorders.

On Friday, more than 90 program attendees stayed for NHF's expanded State Advocacy Workshop. Karen Pollitz and Joan Alker from the Georgetown Health Policy Institute presented information on insurance reform and access and the role of Medicaid. Stateside Associates, NHF's state advocacy partner, also presented on state lobbying and the fundamentals of advocacy as well as Medicaid.

All materials from Washington Day are available online and can be accessed at <http://www.hemophilia.org/events/washingtontoday.htm>.

March 10, 2004**Pennsylvania Sues Drug Makers**

The Commonwealth of Pennsylvania filed a lawsuit on March 10, 2004, against 13 large drug companies accusing them of price manipulation. The lawsuit, which seeks to recover hundreds of millions of dollars, was filed by Pennsylvania Attorney General Jerry Pappert. It accuses the drugmakers of artificially inflating prices to cheat taxpayers, employees and Medicaid recipients. It seeks relief and rebates going back to 1991.

The legal brief claims that the 13 companies "oppose[d] and avoid[ed] efforts to reduce prescription drug costs and/or to change the way in which payors reimburse for prescription drugs, and ... act[ed] to conceal and suppress their conduct to prevent detection by others." The state joins at least 12 others filing suit against the industry, and more state attorneys general are likely to join the legal battle.

Pennsylvania says that the companies have multiple prices—one that they charge physicians and the "average wholesale price," which is what the medical providers can then bill the government, insurance providers and patients at the much higher "average retail price." Pennsylvania claims that this scheme entices the medical providers to engage in negative price competition, where they have incentive not to prescribe the most cost-effective treatment available, but the most expensive one, since they can pocket the difference.

Named in the lawsuit are Pfizer (NYSE: PFE), AstraZeneca (NYSE: AZN), Amgen (Nasdaq: AMGN), Bristol-Myers Squibb (NYSE: BMY), Schering-Plough (NYSE: SGP), GlaxoSmithKline (NYSE: GSK), Bayer (NYSE: BAY), TAP Pharmaceuticals, Boehringer Ingelheim, Johnson & Johnson (NYSE: JNJ), Baxter (NYSE: BAX), Dey Pharmaceuticals and Aventis (NYSE: AVE).

Source: Associated Press.

NHF NEWS

March 26, 2004**Save the Date: New York Leadership Weekend and Gala**

The New York Leadership weekend will be held June 11 and 12, 2004, at the Waldorf=Astoria Hotel in New York City. The event will continue to build on the ideas discussed at the NHF Chapter/Association Caucus in Tampa on April 31, 2004. In addition, seven hemophilia foundations will be honored for having served their communities for 50 years or more at a gala event on Saturday night. Please mark your calendars and look out for invitations coming in the mail soon.

For more information, contact Rebecca Schoon, manager of Development, at rschoon@hemophilia.org.

March 15, 2004**NHF Teams Up for Celebrity Softball Games**

NHF has been fortunate enough to secure a partnership with Hollywood All-Stars (HAS) Celebrity Games, an organization that matches major league sports figures and a team of celebrities for a softball game benefiting a selected charity. In 2004, their 25th anniversary year, HAS has

chosen NHF and its local chapters to be the sole beneficiary of all six games.

Proceeds from ticket sales will benefit NHF and the local host chapters in each of the cities. In addition, the local chapters will provide the opportunity for children from their summer camps to attend the game and meet the athletes and stars.

Schedule (dates are subject to change):

May 15 and 16: St. Louis Rams vs Hollywood All-Stars softball game, golf tournament and party in St. Louis to benefit NHF and the St. Louis chapter;

June 5 and 6: New England Patriots vs Hollywood All-Stars softball game, golf tournament and party in Boston to benefit NHF and the New England chapter;

July 10: Texas Sports All-Stars vs Hollywood All-Stars softball game, golf tournament and party in Austin to benefit NHF and the Houston chapter;

August 21: Hollywood All-Stars 25th Anniversary Game either at Yankee Stadium or in Central Park to benefit NHF.

Stars playing on the Hollywood All-Star team will vary in each city, but some of the most likely participants are: Kevin James

("The King of Queens"), Dennis Haysbert (president on "24"), Richard Schiff ("The West Wing"), Esai Morales ("NYPD Blue"), Meat Loaf, Mark Consuelos ("All My Children"), Scott Wolf ("Party of 5"), Norm McDonald and many others.