Marfan's Syndrome Starting to Yield Secrets

A NEW ANTIBODY has led to the discovery of a previously unrecognized connective tissue protein. That discovery may help unveil some of the mystery of Marfan's syndrome, the extremely rare, but exceedingly notorious, hereditary disease, complications of which led to the untimely death of world-class volleyball star Flo Hyman two years ago and allegedly affected Abraham Lincoln, violin virtuoso Niccolò Paganini, and composer-pianist Franz Liszt.

The monoclonal antibody, developed by researchers at Oregon Health Sciences University, Portland (J Cell Biol 1986;103:2499), binds to a protein they have named fibrillin. According to one of the investigators, it may prove to be an aid in diagnosing this confoundingly pleiotropic syndrome.

Using the monoclonal and an indirect immunofluorescence technique in cell cultures of skin from patients with Marfan's, the researchers discovered an intricate scaffolding-like array of microfibrils, often attached to elastin fibers, that seem to be composed primarily of fibrillin. It appears that these extracellular microfibrils play some connective tissue role.

Marfan's is believed to be a disorder of the connective tissue because all the disparate, pathological abnormalities involve supporting elements. Marfanoid manifestations include disproportionately long limbs and fingers, displaced lenses, nearsightedness due to increased axial length of the eye, flat syndrome. In order to locate enough patients, the investigators contacted Reed Pyeritz, MD, PhD, the director of the medical genetics clinic at the Johns Hopkins Medical Institutions, Baltimore, a major center for Marfan's syndrome studies. Pyeritz sent skin samples from a total of 64 patients and controls.

According to the researchers, who presented their findings at the First International Symposium on the Marfan Syndrome, in Baltimore, 23 (88%) of 26 patients with Marfan's syndrome had diminished amounts of microfibrils, a finding apparent in skin or fibroblast culture, or both. The 13 normal individuals included in the study were all diagnosed as non-Marfan. However, the researchers misidentified as having Marfan's syndrome six of the 25 patients with a variety of other connective tissue disorders. Those disorders included epidermolysis bullosa, homocystinuria, cutis laxa, and ectodermal dysplasia.

In another study, the researchers looked at three Marfan's syndrome families, each with one affected parent and at least one affected child. "In all three kindreds," the researchers say, "all the affected members had a reduced accumulation of antibody-reactive, fibrous extracellular network in two-day fibroblast cultures, whereas all nonaffected family members displayed normal accumulation."

David Hollister, MD, senior investigator at Shriners Hospital for Crippled Children, rejected the position on ethical grounds. Another member, Rabbi J. David Bleich, PhD, of the Cardozo Law School in New York City, also is considered likely to dissent. At their most recent meeting, panelists responded to questions from Robert Windom, MD, assistant secretary for health (JAMA 1988;260:2012-2015).
Warning Label Required for Alcohol Containers

A YEAR from now, all beer, wine, and liquor containers will—for the first time—be required to bear a government warning label.

The label will point out that the surgeon general of the Public Health Service advises that women not drink alcoholic beverages during pregnancy because of risk of birth defects. It also reminds that “consumption of alcoholic beverages impairs your ability to drive a car or operate machinery, and may cause other health problems.”

The federal label should help to educate the public, predicts Donald W. Schiff, MD, president of the American Academy of Pediatrics: “Too many people do not know that fetal alcohol syndrome exists, much less that it is a preventable one. Although we must continue to give education about the many dangers of alcohol abuse, the new warning labels are a significant step.”

A number of public and private agencies are working together to combat the drinking-and-driving problem, which is compounded by the national dependence on the automobile. In the latest estimate, the American Coalition for Traffic Safety, Inc, Washington, DC, estimates that the national mean is 336 hours, about 14 days, behind the wheel each year:

The warning label legislation, considered in various forms since the 1970s, is part of the omnibus drug bill passed by Congress shortly before its adjournment.

—by Phil Gunby

Miscellanea Medica

David N. Sundwall, MD, has resigned as administrator, Health Resources and Services Administration, Rockville, Md (JAMA 199:260:1986). He has joined the American Healthcare Institute, Washington, DC, as vice president and medical director.

Jay Moskowitz, PhD, has been named acting director, Institute on Deafness and Other Communication Disorders, National Institutes of Health, Bethesda, Md. Congress has authorized $96.1 million for the new institute.

Seymour Diamond, MD, director, Diamond Headache Clinic, Chicago, is the first honoree of the Migraine Trust Lectureship, presenting the inaugural lecture in London at the Seventh Migraine Trust International Symposium.

Arno G. Motulsky, MD, professor of medicine and genetics, University of Washington School of Medicine, Seattle, has received the Sanremo International Award for Genetic Research. He is a member of the JAMA Editorial Board.

Byron J. Bailey, MD, Wiss Professor and chair, Department of Otolaryngology, University of Texas Medical Branch, Galveston, is the new president, American Academy of Otolaryngology—Head and Neck Surgery. He is chief editor of the American Medical Association's Archives of Otolaryngology.

Oliver H. Beahrs, MD, professor of surgery emeritus, Mayo Medical School, Rochester, Minn, is the new president, American College of Surgeons.

Editor's Note: Miscellanea Medica normally appears in the Medical News & Perspectives section several times each month. Items submitted for consideration should be directed to the attention of Phil Gunby, editor/director, Medical News & Perspectives Department.