News
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**Stormie Jones returns to Dallas for evaluation of transplanted liver.

DALLAS--Stormie Jones, the six-year-old child who underwent simultaneous heart and liver transplants in February at the University of Pittsburgh, may return Monday to The University of Texas Health Science Center for further testing.

Stormie will be admitted to the General Clinical Research Center here so physicians can monitor how well her transplanted liver is working to stabilize her disease. She suffers from a rare genetic disease called "homozygous familial hypercholesterolemia" (FH), which causes dangerously high cholesterol levels and subsequent heart attacks. Dr. David Bilheimer, who has studied patients with homozygous FH for 10 years, was the first to diagnose her condition last July.

"She is medically stable and recovering well from her operations. Although her plasma cholesterol level has dropped by more than 60 percent since her operation, the purpose of this admission is to determine more precisely how well her transplanted liver is functioning to remove the excess cholesterol and low density lipoprotein (LDL) from her body," explains Bilheimer, professor of Internal Medicine and Stormie's primary physician.

"This information is necessary to fully evaluate the benefits of liver transplantation in this rare disease and to determine if Stormie will need additional therapy to control her plasma cholesterol level in the future."

The studies require that Stormie eat a special metabolic diet for the estimated four- to five-week hospitalization. During that time the capacity of her new liver to clear cholesterol from her body will be measured. Says Bilheimer: "Since these studies are intricate and require the careful collection of precisely timed specimens of blood and urine according to specific protocol, she will be confined primarily to the metabolic ward, and only her family members will be allowed to visit her."

The preliminary results of her studies will not be available until the end of her hospitalization.

Dr. Scott Grundy, professor of Internal Medicine and Biochemistry, will be managing Stormie's care while Bilheimer is in Europe lecturing this month.

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The information doctors are able to gain from Stormie's case may have implications for at least one in 500 Americans who suffer from a less serious -- but nevertheless devastating -- form of the disease.

Drs. Joseph Goldstein, professor and chairman of Molecular Genetics and Michael Brown, professor of Molecular Genetics and Internal Medicine and director of the Center for Genetic Diseases, first discovered the genetic flaw in homozygous FH in the early 1970s at the health science center. Since that time, they and Bilheimer and Grundy have continued working to understand and treat the disease. They have learned that certain experimental drugs reduce cholesterol levels to nearly normal in the less serious form of the disease.

One in 500 Americans has "heterozygous familial hypercholesterolemia" -- a condition in which they inherit a mutant gene from one parent that produces higher-than-normal blood cholesterol levels from birth and heart attacks in early middle age. Patients like Stormie, who inherit mutant genes from both parents, have extremely high cholesterol levels and may develop heart attacks from atherosclerosis during the first decade of life.

Because of Stormie's condition, none of her body's cells contained "LDL receptors." Normally, these receptors recognize a form of cholesterol known as LDL and efficiently clear it from the blood. Without any receptors, Stormie had no way to remove cholesterol from her blood. As a result, her cholesterol levels were six to eight times normal for a child her age.

Studies by Dr. John Dietschy, professor of Internal Medicine, have shown that the liver is the major organ for clearing LDL. Therefore doctors predicted a normal donated liver containing receptors might save her life.

Bilheimer referred Stormie to Dr. Thomas Starzl in Pittsburgh for evaluation last December when it became clear the disease had ravaged her heart. Following massive damage to her heart caused by a heart attack and two coronary bypass open heart surgeries, doctors here feared Stormie's heart was too weak to withstand a liver transplant.

Starzl's evaluation confirmed their fears and the decision was made to attempt the dual transplant. Under Starzl's direction, Pittsburgh physicians performed the world's first combined heart and liver transplantation on Valentine's Day.

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EDITOR'S NOTE: B-roll and still photographs will be available via a pool system. For information about photos/videotape, contact: Ann Harrell at (Office) 214/688-3404, (Home) 214/369-2695 or Bob Fenley at (Office) 214/688-3404, (Home) 214/352-2502.

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