UTNews
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****New process "washes" excess LDL-cholesterol from blood

DALLAS -- Researchers at The University of Texas Health Science Center at Dallas have recently begun trials of a new experimental procedure that literally "washes" LDL, the harmful cholesterol-carrying lipoprotein, from the blood of children who suffer from familial hypercholesterolemia (FH). FH is an inherited genetic condition in which cholesterol builds up to dangerous levels in the bloodstream, clogging arteries and causing coronary heart disease. Its victims rarely live beyond their teen-age years.

LDL-plasmapheresis, a new procedure, is similar to plasmapheresis, another "blood-washing" technique that is sometimes used in neuromuscular autoimmune diseases and is a similar process to kidney dialysis, in which blood is filtered and cleansed. The new Pediatric Subunit of UTHSCD's General Clinical Research Center, located in Children's Medical Center, is the only pediatric medical center in the United States where LDL-plasmapheresis is being performed, said Dr. Ricardo Uauy.

Uauy, associate professor of pediatrics at UTHSCD and Children's Medical Center staff member, is directing the project with the collaboration of Dr. David Bilheimer and Dr. Scott Grundy, experts in lipid metabolism. Dr. Charles Ginsburg, interim chairman of pediatrics at UTHSCD and medical director at Children's, is in charge of the subunit. Director of the GCRC is Dr. Charles C.Y. Pak, UTHSCD professor of internal medicine.

Currently approximately 20 children are receiving treatment for cholesterol problems in the GCRC's Pediatric Subunit. Two of them are undergoing LDL-plasmapheresis, and the remainder are involved in other treatments. Uauy said that he and the other researchers working on the problem hope that new approaches, like the LDL-plasmapheresis, will help children with FH avoid or delay the necessity for liver transplantation in the future.

It was in the Dallas GCRC that Stormie Jones, the only person to survive a tandem heart-and-liver transplant, received her care before her life-saving transplant and where she continues her treatment today. The young patients receiving LDL-plasmapheresis have the same condition as Stormie Jones. It is called homozygous familial hypercholesterolemia, a rare genetic condition in which the person inherits abnormal LDL receptor genes from both parents. (When the abnormal gene from only one parent is involved, the condition is called heterozygous.)

LDL-plasmapheresis is being done on a limited basis in such countries as Germany, France, England, Norway, Italy and Japan, where it was developed, and at five centers in the United States where it currently is being used with 11 adults and three children. The goal of each treatment is to remove as much cholesterol as possible from the blood. Although standard plasmapheresis is sometimes used with these patients, Uauy said he believes that LDL-plasmapheresis will prove superior because other plasma proteins from the blood are returned to the patient rather than discarded, thus HDL (the "good" cholesterol) is preserved. The procedure's superior technology does not seem to damage the red cells.

Uauy said that LDL-plasmapheresis is currently awaiting FDA approval in the United States. The pediatric subunit is testing an LDL-plasmapheresis column called a Liposorber, which is produced by Kaneka, a Japanese company. The Liposorber is attached to a regular plasmapheresis machine, which is a part of Parkland Memorial Hospital's hemotherapy program.

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In the Liposorber a "binder" present in the column, in this case dextran sulfate, attracts and holds the "bad" LDL cholesterol so it can be removed. The blood flows through a hollow fiber membrane, or collection of small tubes. The red cells and platelets, which are too large to pass through the small holes, stay in the tubes as the plasma moves through. As the plasma passes out, LDL and VLDL (very low-density lipoproteins) are bound to the dextran sulfate inside the column, which takes on a yellowish color as the fat collects inside.

In regular plasmapheresis, the patient's plasma has to be discarded and replaced with a non-cholesterol-containing solution. In LDL-plasmapheresis the cleansed plasma is re-mixed with the red cells and sent back into the body of the patient via an intravenous line.

Working with UTHSCD and Children's Medical Center personnel on the new treatment plan are Mary Jo Phillips, R.N., apheresis supervisor, and Dr. Edwin Stean, UTHSCD associate professor of clinical pathology and head of Parkland's hemotherapy department. Phillips, who does the actual procedure, said that she and the physicians involved are very excited about the procedure that may provide an alternative to liver transplantation.

Yolanda Hurtado, mother of Ricardo and Jhoanna, both patients with hypercholesterolemia, said that the hope of avoiding transplant surgery for her children was a major reason she and her husband brought the children to Dallas for treatment. The two children were born in this country, and thus they were eligible for the federally funded program. However, it has taken the cooperation of immigration authorities to enable the family to stay since the parents have not yet established citizenship.

So far Ricardo, who was the second young patient to undergo LDL-plasmapheresis, has been responding well. His first treatment resulted in a drop from 916 to 147 in his overall cholesterol level (his normal count should be around 153), and his LDL went from 800 to 90 (under 100 is very good for his age, which is 10.)

"It should be understood that this treatment requires a long-term commitment," stressed Uauy. "It's a lifetime treatment -- not a 'fix.'" Currently researchers believe that it takes only about two or three weeks for the cholesterol and LDL levels to rise to a dangerous level, so treatment ideally occurs at two-week intervals. Also, it is somewhat painful for the children and takes one and a half to two hours each time.

"There's always the hope of a better way in the future through research, but we're still finding out the benefits of this treatment," said Uauy.

The health science center and its General Clinical Research Center program, which is funded by the National Institutes of Health, has become a major referral center for both pediatric patients with FH and those with other abnormal cholesterol metabolism, including obesity coupled with high cholesterol levels and unusually high cholesterol counts in otherwise normal children.

Other cholesterol-related studies are also going on in the GCRC in both the adult and pediatric units. In addition, UTHSCD researchers are looking at ways to crack the genetic basis of cholesterol problems, a puzzle that may turn out to have well over a hundred answers. While Drs. Joseph Goldstein and Michael Brown continue their Nobel Prize-winning work in the field, other health science center researchers are also looking for clues in collaborative efforts. These include Grundy, who is director of the Center for Human Nutrition; Bilheimer, professor of internal medicine and Stormie Jones's personal physician, and Dr. Helen Hobbs in the Department of Internal Medicine, who is also looking at the genetics of hypercholesterolemia.

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NOTE TO EDITORS: Photos are available on request. Please call for date of next procedure.

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