

Rehab and Transplant Best Options For Ped Small Bowel Disorders  
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Small bowel disorders can devastate pediatric patients—robbing them of nutrition, limiting their neural development, leading to infections, and damaging their livers. The answer, say specialists at the Morgan Stanley Children's Hospital of NewYork-Presbyterian, lies in an innovative program that combines intestinal rehabilitation with, if necessary, small bowel transplantation.

“One of the big pushes in the small intestine transplant field is to get involved with these kids early so that we can manage them with all our expertise,” said Steven Lobritto, MD. “We'd like not to get them as late referrals when their options are already really limited and all we can offer them is transplantation. We want to partner with their physicians and lend our expertise in how to manage them. Then, if the kids get into trouble, we're their safety net should a transplant be necessary.”

The intestinal rehabilitation team, which in addition to Dr. Lobritto includes surgical leader Dr. Dominique Jan and Drs. Lesley J. Smith and Robert A. Cowles, first works to optimize residual gut function by providing adequate nutrition and using medications to regulate motility and control acid output. They also employ surgical techniques to stop bleeding, to reconnect the colon and small intestine, to replace a damaged liver with a transplant, or to lengthen an intestine that has been shortened by previous surgery or disease.

The preferred method at the Hospital for bowel lengthening is the novel STEP (serial transverse enteroplasty) procedure, Dr. Lobritto noted: “Basically, it turns a highway into Lombard Street by cutting the bowel from the side in a zig-zag pattern. You change a straight, dilated bowel into a tapered zig-zag bowel with twice the length.”

Other treatments include physical therapy to overcome delayed walking or to teach swallowing. “A lot of these kids have never eaten so they have aversions to food. Giving someone food by mouth stimulates a number of hormonal reactions that favor adaptation,” said Dr. Lobritto.

Typically, pediatric patients arrive with either a shortened bowel (commonly caused by necrotizing enterocolitis or intestinal volvulus) or one of several bowel disorders, including Tufting syndrome or intestinal pseudo-obstruction.

They often rely on total parenteral nutrition to survive. Eventually, it becomes difficult to deliver nutrition, explained Dr. Lobritto: “You don't have an endless number of blood vessels that you can put I.V. lines into. When kids start to run out of those access points, it's time to start thinking about doing a transplant.”

Pediatric patients, usually in the neonatal period, are too small to receive a bowel from an adult donor. Instead, they must wait an average of one year for an appropriate pediatric donor. Then, the transplantation must be scheduled for a time when the

patient does not suffer from a systemic infection.

The primary challenge for small bowel transplant recipients, according to Dr. Lobritto, is not to merely survive surgery but to avoid long-term risks. Physicians must be alert to cytomegalovirus infection or exposure to the Epstein-Barr virus, which can lead to a high risk of post-transplant lymphoproliferative disorder. Immunosuppressants, while helping prevent rejection, also impart their own side effects.

“We’re constantly looking for a better type of immunosuppressant. What revolutionized liver transplants was when cyclosporin and tacrolimus came around and the medicines got better. So we’re looking at how we can manipulate the immune system in ways to benefit the patient, how we can minimize side effects of the medications by using combination protocols, and also how we can treat early viral complications,” he said.

Dr. Lobritto is confident that productive research is on the horizon. “A lot of questions can’t be answered by doing ten cases a year. The statistics won’t have significance with these small numbers,” he said. Each of the 1,200 small bowel transplants performed worldwide have been recorded in a registry; as the

Morgan Stanley Children’s Hospital expands its program, he hopes to pool data with leading institutions within the U.S. and internationally for clinical trials. “We have extensive clinical trial experience from our liver transplant program in pharmacokinetic, pharmacodynamic, multiple drug, and viral treatment studies.

“Five years from now,” he predicted, “what we’re doing now is going to seem like alchemy.”

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