

FOCUS ON PEDIATRIC UROLOGY

Affiliated with Columbia University College of Physicians and Surgeons and Weill Cornell Medical College

National Study Assesses Surgical Outcomes for Congenital Adrenal Hyperplasia

Part of the mission of the Comprehensive Center for Congenital Adrenal Hyperplasia (CAH) at NewYork-Presbyterian Hospital/Phyllis and David Komansky Center for Children's Health is to promote clinical research. Toward that goal, the Comprehensive Center for CAH is currently enrolling patients in the first-ever study to evaluate outcomes of modern genitoplasty techniques. This multicenter National Institutes of Health-funded study comes at a critical point to resolve the debate surrounding surgical approaches for children with nontypical genitalia. Findings from this study may mark a significant change in clinical practice.

"There has been so much interest in actual outcomes for patients with CAH who have undergone surgery, but no one has current data on the outcomes of patients whose care included modern surgical techniques. We need to look at psychological and physical outcomes in these patients to determine if the surgical decisions we are making today are appropriate," explains Dix Poppas, MD,



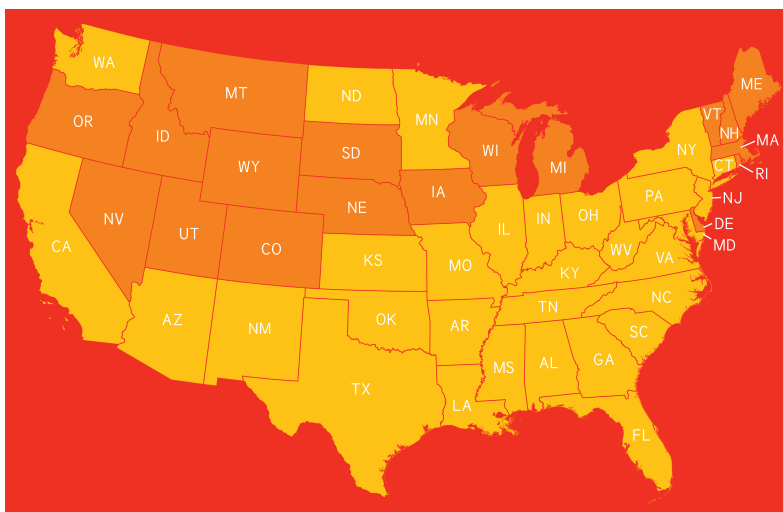
Chief of Pediatric Urology at NYP/Komansky Center, Richard Rogers Family Professor of Pediatric Urology at Weill Cornell

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Pediatric Urologic Surgeons Restore Form and Function

For children with complex congenital urologic disorders affecting bowel and bladder function, corrective surgery may not only restore quality of life, but can also be a matter of life and death. That was the case for Max, who was born without an anus and who had a dysfunctional bladder and a short spinal cord — all components of VATER association (Vertebral, Anal, Tracheal, Esophageal, and Renal abnormalities). After six surgeries over five years, he is now a first grader who enjoys Star Wars and Legos.

Novel techniques in the hands of a skilled pediatric reconstructive urologic surgeon have enhanced the lives of children like Max. The surgeon works as a member of a multidisciplinary team to restore function and cosmetic appearance, with the first operation often happening within the first days of a child's life. "The care of these patients requires a team approach so these



Patients come to Morgan Stanley Children's Hospital for urologic reconstruction from all over the U.S. (see yellow states) and the world, including the United Kingdom, Israel, United Arab Emirates, Ecuador, Dominican Republic, Puerto Rico, Nigeria, Egypt, and Saudi Arabia.

children can lead active, productive lives," says Max's surgeon Shumyle Alam, MD, Assistant Attending Surgeon at NewYork-Presbyterian/Morgan Stanley Children's Hospital and Assistant Professor of Urology at Columbia University College of Physicians and Surgeons.

The Center for Urinary Tract Reconstruction at NYP/Morgan Stanley Children's is among the world's leading centers for pediatric urologic reconstruction. Patients come

from across the country and around the globe for specialized care that is not available elsewhere, or with the same level of expertise. Dr. Alam does more of these procedures in one year than many pediatric urologic surgeons perform in a decade. He and his colleagues Pasquale Casale, MD, Chief of the Division of Pediatric

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Robotic Urologic Surgery Gets Kids Back on Track Faster

The benefits of robotic surgery have been well touted: smaller incisions, less anesthesia, less blood loss and postoperative discomfort, a magnified surgical field, and a speedier recovery compared with open surgery. For most children having robotic urologic procedures at NewYork-Presbyterian/Morgan Stanley Children's Hospital, those advances are translating into other benefits: a single night in the hospital, no need for narcotics, a quick return to sports and an even faster return to school, and often no need to return for a second operation.

"Using the robot, we can operate on a child on a Thursday or Friday, and he can be back at school the following Monday or Tuesday — and back on the soccer field two weeks later," explains Pasquale Casale, MD, Chief of the Division of Pediatric Urology at NYP/Morgan Stanley Children's and Professor of Urology at Columbia University College of Physicians and Surgeons. Ninety-eight percent of patients stay in the hospital less than 24 hours after a robotic procedure. The operations are performed without the need to insert stents, so patients don't have to return for a second stent-removal surgery.

The team is using the surgical robot to operate on patients as young as infants and adolescents up to age 18. NYP/Morgan Stanley Children's — home to the busiest pediatric urologic robotics service in the New York metropolitan area — has a robot dedicated to pediatric surgery and often receives referrals from other hospitals. Patients come to the service from all over the country and the world.

The pediatric robotic surgery program is part of NYP/Morgan Stanley Children's multidisciplinary approach to patient care. Any other specialists a child may need — such as orthopedists, neurosurgeons, gastroenterologists, nephrologists, pediatric gynecologists, and radiologists — are all located at the hospital. Key to optimal patient care is an accurate diagnosis; a diagnostic center for children with complex urologic disorders is therefore a



NYP/Morgan Stanley Children's is home to the busiest pediatric urologic robotics service in the New York metropolitan area.

vital component of the urologic clinic.

Says Dr. Casale, "Our goal is to accurately assess each child, put together a plan of care, perform whatever surgery is needed, and get the child back to the business of being a child."

Pediatric urologic surgeons use robotic surgery for complex procedures that require exceptional precision, such as:

- Pyeloplasty for ureteropelvic junction obstruction
- Partial and total nephrectomy
- Nephroureterectomy
- Ureteral reimplantation
- Bladder reconstruction
- Reconstructive procedures in children with spina bifida

Pediatric Urologic Surgeons Restore Form and Function (continued from page 1)

Urology and Professor of Urology at Columbia, and Sarah M. Lambert, MD, Assistant Attending Surgeon and Assistant Professor of Urology, trained with the pioneers of pediatric urinary and bowel reconstruction.

NYP/Morgan Stanley Children's pediatric urologic surgeons specialize in the:

- **Treatment of disorders of sexual development.** The team includes specialists from urology, endocrinology, and genetics. Care is also provided to perform vaginal reconstruction for girls with Müllerian failure and Mayer-Rokitansky syndrome (abnormal vaginal development).
- **Repair of cloacal exstrophy and bladder exstrophy.** NYP/Morgan Stanley Children's is one of only a few medical centers on the East Coast with an interdisciplinary program to treat exstrophy.
- **Bladder reconstruction in children with spina bifida and anorectal malformations born with neurogenic bladder.** Pediatric urologists have experience in appendicovesicostomy (creating a channel to enable patients to perform their own

bladder catheterizations) and the Malone procedure (to facilitate bowel emptying), using the appendix to create both channels — something not done in many other institutions.

- **Hypospadias repair.** Surgeons perform reconstructive surgery for hypospadias to restore cosmetic appearance and optimal function, especially re-operative hypospadias repair.
- **Surgery for children with end-stage kidney disease.** Twenty-five percent of children who need a kidney transplant or dialysis due to end-stage renal disease have urologic issues. Reconstructive surgeons perform pre- and post-transplant procedures to enhance graft survival, reduce infection risk, and improve urinary function.

The Division of Pediatric Urology is working with Hilda Fernandez, MD, Instructor in Medicine at Columbia, to develop a transitional care program for adolescents with chronic urologic disorders as they move into adulthood. "For many children, we need to see them throughout their lives," concludes Dr. Alam. "We are here for them as long as they need us."

Building a Better Bladder

A normal bladder expands to hold a growing volume of urine until a child urinates. But children with fibrotic bladders don't have that ability. The pressure that builds as the bladder fills with urine is transmitted to the kidneys, which can become damaged. Some patients' kidneys become so damaged that they require dialysis or a kidney transplant.

Researchers in the Institute for Pediatric Urology at NewYork-Presbyterian Hospital/Phyllis and David Komansky Center for Children's Health are launching preclinical studies to evaluate a synthetic bladder augmentation device for children born with very small fibrotic bladders. If successful, the synthetic bladder augmentation approach could help children with myelodysplasia and other causes of fibrotic bladder to achieve more efficient bladder function.

Pediatric urologists currently use an approach called ileo-cystoplasty to treat these children, inserting a piece of the ileum to open and enlarge the bladder. But this surgery is extremely complex, is associated with a lengthy hospital stay, and requires a large incision. Moreover, there is a risk of complications such as adhesions and reactions to the ileal tissue. Ileal cells inside the



A synthetic bladder augmentation device could potentially help children with fibrotic bladders. This device is being assessed in preclinical studies.

bladder transport chloride outside the bladder, causing hyperchloremic acidosis. And ileal cells inside the bladder secrete mucus that can cause infections and bladder stones.

Led by Dix Poppas, MD, Chief of Pediatric Urology at NYP/Komansky Center and Richard Rodgers Professor of Pediatric Urology at Weill Cornell Medical College, and Diane Felsen, PhD, Associate Research Professor of Urology and Pharmacology, investigators designed a nonreactive, synthetic bladder augmentation device with an exterior composed of polypropylene microfiber and

a hydrogel interior. In a study in which a patch of the material was applied under the skin of rats, it did not elicit a foreign body reaction. "Cells don't grow on it," says Dr. Felsen. "It's a novel synthetic which appears able to reside in the hostile environment of the bladder."

The synthetic bladder augmentation device is being evaluated in pigs with normal bladders to assess its function. If the results are encouraging, it will then be tested in a fibrotic bladder animal model, and eventually in clinical trials of young patients.

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Medical College, Surgical Director of the CAH Center, and principal investigator of this clinical research initiative. Maria Vogiatzi, MD, Chief of Pediatric Endocrinology, serves as the Medical Director of the CAH Center.

Surgical approaches for many patients with CAH and other types of nontypical genital development have been under intense scrutiny by patient advocates, based on poor post-surgical cosmesis and sexual dysfunction observed in genitoplasty outcome studies. However, these studies examined outcomes associated with outdated surgical procedures.

The largest group of patients to benefit from modern approaches is female patients with classical CAH, who will comprise the majority of the current study participants. Females with classical CAH are born with a spectrum of urogenital developmental issues. All patients in the multicenter study will be followed for five years.

The NIH grant was awarded to Amy Wisniewski, PhD, from The University of Oklahoma Health Sciences Center.

The study aims:

- To determine the effectiveness of modern genitoplasty for improving cosmetic appearance and functional outcomes in young children born with a spectrum of urogenital developmental disorders. Outcomes will be ascertained by surgeons who performed the procedures, as well as patients' parents and their other healthcare providers.
- To compare urinary tract infections between patients who have genitoplasty and those who do not, and to analyze postoperative complications among those who have surgery.
- To examine specific parental adjustment outcomes such as depression, anxiety, post-traumatic stress symptoms, perceived illness uncertainty, parental quality of life, and decisional regret by parents following their child's surgery.

The CAH Center's specialized multidisciplinary team makes NYP/Komansky Center a critical site for this kind of ground-breaking research. Designated by the CARES Foundation as the nation's first Center of Excellence for CAH, the Comprehensive Center for Congenital Adrenal Hyperplasia at NYP/Komansky Center has been at the forefront of medical and surgical care for affected children and is the main referral site for children with CAH from around the world.

For more information, to inquire about a patient's eligibility for this study, or to refer a patient, please contact Dr. Poppas' office at (212) 746-5337.

Enhanced understanding of genital neurovascular anatomy and better surgical techniques have provided surgeons with modern strategies to improve post-surgical outcomes.

Focus on Pediatric Urology

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