NewYork-Presbyterian Komansky Center Houses the Center of Excellence for Congenital Adrenal Hyperplasia

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Morgan Stanley Children’s Hospital Excels in Treatment of Endocrine Disorders

Endocrine disorders may cause significant medical and physical problems during childhood and adolescence that can have lifelong health and psychological consequences. Specialists in the Division of Endocrinology, Diabetes and Metabolism at NewYork-Presbyterian Morgan Stanley Children’s Hospital/Columbia University Medical Center are nationally and internationally renowned for their expertise in treating children with endocrine disorders that have the potential to disrupt normal pediatric development.

Pooling their expertise with that of their colleagues in other divisions of the hospital and applying the results of the latest basic science and clinical research — much of which is performed at NewYork-Presbyterian/Columbia — physicians enable many children to resume normal, healthy lives. The Division was ranked 10th in the country in the 2013-14 U.S. News & World Report “Best Hospitals” issue.

“Endocrinology is a field where we’ve seen tremendous growth over the last 40 to 50 years,” said Sharon Oberfield, MD, Director of the Division of Endocrinology, Diabetes and Metabolism and Professor of Pediatrics at Columbia University College of Physicians and Surgeons. “We now know the underlying causes of many disorders and can give specifically targeted treatments and medications to children to allow for normal growth and development.”

Members of the Division performed more than 7,900 outpatient and more than 250 in-house consultations in 2013. The team treats children with disorders of growth and puberty; obesity, insulin resistance, and diabetes; neuroendocrine dysfunction; metabolic bone disease and abnormalities of body composition; adrenal disorders; and endocrine problems related to pediatric cancer and its treatment.

Expertise in Type 1 and 2 Diabetes Management

The Naomi Berrie Diabetes Center is one of the largest pediatric diabetes programs in the country and focuses on both the prevention and treatment of this steadily increasing disease. It is one (continued on page 2)

Komansky Center Houses Center of Excellence for Congenital Adrenal Hyperplasia

Congenital adrenal hyperplasia (CAH) is one of the most common recessive birth defects in the world. The Comprehensive Center for Congenital Adrenal Hyperplasia at NewYork-Presbyterian Phyllis and David Komansky Center for Children’s Health/Weill Cornell Medical Center — recognized as the nation’s first Center of Excellence by the CARES Foundation (Congenital Adrenal Hyperplasia Research Education and Support) — provides exceptional care to patients with CAH from childhood to adulthood. The team also promotes research that will improve patients’ lives, and educates patients, families, and other healthcare providers about CAH and its management.

In patients with CAH, a deficiency in the 21-hydroxylase enzyme prevents the adrenal glands from producing cortisol. In the mild or “nonclassical” form of CAH, which is more common, the adrenal glands compensate, and cortisol production returns to normal. In the mild or “nonclassical” form of the disease, there is a complex lack of cortisol, which may be combined with a lack of aldosterone.

Children with classical CAH may experience feeding problems and severe dehydration, requiring prompt treatment. Chemical disruptions in CAH also lead to high levels of androgens. Girls with the severe form of CAH are often born with abnormalities of the external genitalia. Children with both (continued on page 3)
of three Diabetes Centers of Excellence in New York State and has been cited by the American Diabetes Association for its quality care. The Komansky Center offers multidisciplinary family-centered services, including home and school visits, an insulin pump program, and educational programs for parents and other caregivers. The Center’s services are tailored to young patients and their families, and the staff teaches them how to live a healthy life with diabetes. Patients also have the opportunity to participate in clinical trials of novel diagnostic and therapeutic approaches.

Pediatric DXA Testing

Morgan Stanley Children’s Hospital is home to a unique bone mineral density (BMD) testing program in New York City that operationally can assess children and adolescents. Using dual-energy x-ray (DXA) technology, the program also assesses patient’s body composition, including percentage body fat, lean body mass, and fat distribution.

For young patients with certain medical conditions and those taking long-term medications such as steroids, the risks of low BMD and subsequent fracture are very real. While BMD testing in adults is widely used, determining BMD in children requires specific expertise and training. At Morgan Stanley Children’s Hospital (New York Presbyterian Hospital [directed by John Bilezikian, MD]), our pediatric endocrinologist provides that expertise.

The program is especially useful for assessing bone health and monitoring response to therapy in children at risk for low BMD and fractures, such as those with:

- Rheumatologic, gastrointestinal and oncologic disorders who are treated with long-term steroid therapy
- Osteogenesis imperfecta, a rare genetic defect of bone formation
- Girls withamenorrhea nervosa or polycystic ovary syndrome
- Boys with Klinefelter Syndrome

BMD testing may also be useful for studying bone density changes in morbidity obese adolescents who have had bariatric surgery. The results of each patient’s DXA test are compared with reference values gathered as part of a multicenter NIH-funded Bone Mineral Density in Childhood study, in which the Division of Endocrinology, Diabetes and Metabolism participated.

Treatment of Disorders of Puberty and Development

The Division has a national and worldwide reputation for the management of pediatric disorders of growth and development. Furthermore, Morgan Stanley Children’s Hospital is designated site for the New York State Congenital Adrenal Hyperplasia (CAH) and Newborn Thyroid Screening Program. Endocrinologists work closely with specialists in pediatric urology to assess and treat children with CAH. The care of children requires a multidisciplinary effort that is best addressed by an integrated and experienced team. Specialized nursing of parents and children with thorough consultations and discussions of available options, including medical and surgical care.

The POWER Program: Tackling Adolescent Obesity

Obesity among teenagers has increased dramatically in the last several decades. The Adolescent POWER Program (Program for Overweight Education and Reduction) expands patient education for overweight teens in the community. POWER provides adolescents with medical, psycho-social, and nutritional evaluations and the opportunity to participate in either a group or individual treatment program. The program includes meetings with a nutritionist, an adolescent medicine provider, and an endocrinologist/obesity specialist. Pediatric endocrinology specialists also collaborate with surgeons in the Center for Adolescent Bariatric Surgery, offering a comprehensive, multidisciplinary weight management program that includes health and metabolic screening; nutritional guidance, dietary management, and education; individualized exercise programs; and bariatric surgical procedures.

The shared goal of these programs is to reduce obesity among teenagers, subsequently reducing their risk of type 2 diabetes, heart disease, hypertension, breathing and sleep difficulties, liver disease, and degenerative joint disorders.

An Integrated Approach to Your Patient’s Care

Physicians who refer their patients to the Division of Endocrinology, Diabetes and Metabolism at Morgan Stanley Children’s Hospital can be assured that all of the child’s medical needs can be met in one hospital. Any subspecialists needed to address the patient’s other health issues are on staff. Coordinated care is provided to ensure that the expertise of each patient and his or her family is comprehensive and seamless.

The team also makes sure that patients understand why they are there, delivering this information in an age-appropriate manner. “I enjoy watching their awareness become an understanding,” concluded Dr. Oberfeld. “They learn that they’re here to see a doctor who can help them.”

For more information or to refer a patient, call
(212) 305-6559 or visit nyp.org/kids/morganstanley.

New-York Presbyterian Morgan Stanley Children’s Hospital/ Columbus University Medical Center
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Komansky Center Houses Center of Excellence for Congenital Adrenal Hyperplasia

The Komansky Center for Children’s Health includes a multidisciplinary team of pediatric and adult endocrinologists, urologists, nurse practitioners, patient care coordinators, dietitians, genetic counselors, reproductive specialists, social work specialists, and psychologists. They collaborate to provide compassionate, individualized treatment for each patient, as well as support and education for their families.

“Treatment of disorders associated with CAH may also include ursodeoxycholic acid to promote bile flow and improve biliary excretion of bile acids, helping to prevent liver disease and malabsorption of fat and fat-soluble vitamins. Other treatment options include higher doses of glucocorticoids and mineralocorticoids to improve sodium retention and blood pressure control, and growth hormone therapy for patients who are short for their age. In some cases, medical treatment may not be enough for healthy growth and development. In these cases, surgical reconstruction may be necessary.”

Dr. Pappas, MD, Surgical Director of the Comprehensive Center for CAH and Chief of Pediatric Urology. “We spend a lot of time working with patients and their families to understand anatomy and function and to discuss available surgical options. Patients and families who come to us benefit from a team approach to care, where they can have all of their questions answered in one place.”

In addition to clinical care, the team at the Comprehensive Center for CAH conducts basic science and clinical research to advance the treatment of CAH and improve the lives of people affected by it. Physicians wishing to refer patients for enrollment in a clinical trial should contact Marta Aranda at (212) 746-3797.

Current research efforts are aimed at:

- Optimizing steroid dosing at all stages of a patient’s life (during well periods as well as times of illness, stress, and injury).
- Determination of the appropriate doses of steroid replacement in children with CAH who undergo surgery. The study is actively enrolling patients.
- Assessment of the psychological effects of CAH and its influence on quality of life and development. The study is actively enrolling patients.
- Examination of the roles of androgen receptors (AR) and estradiol receptors (ER) in gonadal development. NYP/Weill Cornell investigators characterized AR and ER in the genital tissue of females with CAH and found both AR and ER in erectile tissue from these patients, with a tendency toward higher AR expression. In Prader 1 patients compared with Prader 3 patients. These results provide insights into the development of both AR and ER in genitourinary tissue. The results of additional studies may lead to pharmacologic intervention as an adjunct to CAH treatment.

For more information or to refer a patient to the Comprehensive Center for Congenital Adrenal Hyperplasia at the Komansky Center, please call (212) 746-3462 or (212) 746-3075, or visit nyp.org/kids/komansky.

New-York Presbyterian Komansky Center for Children’s Health/ Weill Cornell Medical Center
525 East 68th Street, New York, NY 10065

The Division of Endocrinology, Diabetes and Metabolism was ranked 10th in the country in the 2013-14 U.S. News & World Report “Best Hospitals” issue.

The Berrie Center for Diabetes and Metabolism offers the American Diabetes Association for its quality care. The Berrie Center offers multidisciplinary family-centered services, including home and school visits, an insulin pump program, and educational programs for parents and other caregivers. The Center’s services are tailored to young patients and their families, and the staff teaches them how to live a healthy life with diabetes. Patients also have the opportunity to participate in clinical trials of novel diagnostic and therapeutic approaches.

Endocrinologists offer individualized treatment for each patient, as well as support and education for their families. "The challenges faced by patients with CAH are similar to those of patients with rare diseases," explained Maria G. Yugari, MD, Medical Director of the Comprehensive Center for CAH and Associate Professor of Clinical Pediatrics, Weill Cornell Medical College. "As they grow up, there are not many physicians who are well versed enough in CAH to provide their care. Our center has the expertise and experience to serve them throughout their entire lives — from the moment of diagnosis through adulthood." For example, through the center’s transitional care program, the team pays special attention to adolescents as they reach adulthood, providing them with information they need to advocate for their own health care as they enter their adult years and linking them with endocrinologists who treat adult patients.

The center attracts patients from around the world who come to access its services, which include:

- Medical treatment: Endocrinologists offer individualized treatments for patients with classical and non-classical CAH, including therapy with hydrocortisone or dexamethasone. Doctors need to tailor therapy to find the right balance for patients that is effective for treating CAH while ensuring normal bone growth, menstrual cycles, and stress levels as patients get older. Treatment needs to be modified regularly as patients age and their needs change.
- Surgical consultation: Endocrinologists consult with patients and their families and patients who wish to learn about genital reconstruction for girls with severe CAH.
- Psychosocial support: for children, adolescents, and adults with CAH and their families.
- Nutritional counseling by registered dietitians.
- Genetic counseling and prenatal testing for parents who know they carry the gene for CAH.
- Coordination of other medical care, including referrals to other New York Presbyterian/Weill Cornell physicians with knowledge about treating patients of all ages with CAH.
- "Our group is one of only two major referral centers in the country for CAH surgery," said Dr. P. Pappas, MD, Surgical