This past year was marked by significant expansion of the child health enterprise in the Mount Sinai Health System. This was due in large part to our integration with the Continuum Health Partners hospitals, and completion of state-of-the-art renovations of our Pediatric Intensive Care Unit in the Kravis Children’s Hospital at Mount Sinai, and in our pulmonary function testing, biobehavioral, and psychophysiology laboratories. Renovations are now under way of our Neonatal ICU and Center for Children’s Cancer and Blood Diseases.

Our reputation as a leader in clinical care is evidenced by the U.S. News & World Report ranking of the Mount Sinai Kravis Children’s Hospital among “America’s Best Children’s Hospitals” in 2013-14 in seven specialties: gastroenterology and GI surgery, nephrology, diabetes and endocrinology, pulmonology, cancer, cardiology and heart surgery, and urology. In 2015, the Department continued to maintain a robust extramural funding portfolio, ranking among the top ten pediatric departments nationwide in National Institutes of Health funding.

A major focus of the Department is to train the next generation of academic leaders in pediatrics: clinician-educators, physician-scientists, and those whose passion lies in public policy and advocacy. After a record-setting match season in 2013, we received more than 1,800 applications for our 21 PGY-1 resident slots in Pediatrics for 2014.

This report spotlights some of the exciting programs at the Kravis Children’s Hospital as we continue our long-and-distinguished tradition of excellence in all academic missions.

Studying p53 Link to Diseases in Protein Glycosylation

Jaime Chu, MD, Assistant Professor of Pediatrics (Hepatology) at the Kravis Children’s Hospital and Mount Sinai’s Recanati/Miller Transplantation Institute, leads a study investigating the novel intersection of a well-known tumor suppressor gene, p53, with congenital disorders of glycosylation (CDG).

CDG are rare—only an estimated 1,000 patients identified worldwide—and underdiagnosed genetic disorders. Children born with CDG have discrete defects in the processes that are essential for protein secretion and function, and present with debilitating, multisystemic disease. These patients develop signs of common liver and gastrointestinal diseases, making the study of CDG particularly relevant to the broader field of liver disease, as steatosis, fibrosis, and cirrhosis have all been associated with defects in protein glycosylation.

Dr. Chu has been the recipient of several National Institutes of Health (NIH) grants (T32, K12), and recently, a Mentored Clinical Scientist Development Award (K08) to further examine the role of p53 in CDG. Previously, Dr. Chu, with Kirsten Sadler Edepli, PhD, Associate Professor of Medicine (Liver Diseases), and Developmental and Regenerative Biology, has participated in a national consortium funded by an NIH American Recovery and Reinvestment Act grant, and were the first to develop zebrafish as a model to study CDG.

Dr. Chu is also a member of The Mindich Child Health and Development Institute, the translational research enterprise that works closely with the Department of Pediatrics, to advance knowledge and therapies for pediatric diseases.
New Clinical and Research Initiatives for Asthma

The prevalence of asthma has been rising over the past two decades, according to the U.S. Centers for Disease Control and Prevention. In many urban areas such as East Harlem—the New York City community immediately surrounding the Kravis Children’s Hospital at Mount Sinai—the prevalence is more than twice national rates, and utilization of emergency care and inpatient hospital services in the community is among the highest in the nation.

In 2013, Mount Sinai developed several new asthma initiatives. Point-of-care social work and educational services for patients admitted to the hospital were greatly expanded, and a newly funded Pediatrics Visiting Doctors Program was launched in conjunction with the Department of Pediatrics to minimize families’ needs for urgent asthma care and even outpatient clinic visits in some cases. Our goal is to streamline care, improve adherence to medical therapy, and ultimately decrease the need for emergency care and hospitalization.

Additionally, the Division, under the direction of Alfin G. Vicencio, MD, Chief of Pediatric Pulmonology, launched several new clinical and research initiatives to enhance care for the most severely affected patients. They include:

- a new state-of-the-art pulmonary function laboratory to monitor lung function noninvasively in children as young as 5 years of age;
- recruitment of patients for research protocols designed to better characterize specific “sub-types” of asthma that may benefit from individualized and novel therapies;
- the most comprehensive pediatric flexible bronchoscopy program in the region, capable of diagnosing and, in select instances, treating the most complex of airway diseases.

Ectopic Atrial Tachycardia Caused by a Large Right Coronary Artery Fistula

A 15-year-old girl presented with a one-month history of palpitations. Her ECG showed ectopic atrial tachycardia at 138 beats/minute. The echocardiogram showed a very dilated right coronary artery with a fistulous connection draining near the low right atrial wall.

Barry Love, MD, who serves as Director of the Pediatric Arrhythmia Service, and the Pediatric and Congenital Cardiac Catheterization Laboratory, suspected that the dilation of the coronary artery was irritating the atrial myocardium along the right atrioventricular groove and causing the tachycardia. Both the arrhythmia and fistula needed to be addressed: the arrhythmia, to treat the patient’s symptoms and prevent the cardiomyopathy that would inevitably result if she remained in this rhythm for a prolonged period, and the fistula, to eliminate the possibility of arrhythmia recurrence, the volume load on the heart, and the risk of fistula rupture.

The arrhythmia was mapped in the right atrium using a sophisticated 3D navigational system (Carto). The origin of the arrhythmia was found at the inferior right atrioventricular groove (Figure A). Simultaneous coronary angiography confirmed that the earliest atrial site was indeed contiguous with the dilated right coronary artery (Figure B). A radiofrequency application was made at the site of earliest activation and promptly terminated the tachycardia.

The patient was brought back six months later to address the fistula. Using tools designed for delicate neurointerventional procedures, Dr. Love threaded a tiny 0.021-inch microcatheter down the right coronary artery beyond the origin of the posterior descending coronary artery. He then delivered eight platinum coils at the site. After coil placement, there was no further flow in the fistula (Figure C).

The patient was discharged the following day. Six months after the final procedure, she remains asymptomatic with no tachycardia recurrence and has resumed competitive dance.
Managing a Rare Kidney Stone Disease

When a 14-month-old boy was referred to Kravis Children's Hospital at Mount Sinai, a sonogram confirmed multiple large stones in each of the kidneys. One kidney was already obstructed and the other was at risk. Jeffrey A. Stock, MD, Chief of Pediatric Urology, recommended and placed a ureteral stent, and jointly managed the patient with Jeffrey Saland, MD, Chief of Pediatric Nephrology and Hypertension.

Dr. Saland and Dr. Stock simultaneously met with the family in the pediatric stone clinic and coordinated a patient-centered management strategy with the goals of minimizing future nephrolithiasis and avoiding further surgery. Within days of the initial stent, some of the remaining kidney stones had migrated and caused obstruction of the opposite kidney, requiring another complex stenting procedure with laser lithotripsy. Given the severity of the presentation, Dr. Saland immediately suspected a rare kidney stone disease, primary hyperoxaluria (PH), and guided medical therapy around that working diagnosis.

The team first quantified the exact daily urinary oxalate concentration and initiated an aggressive plan of hydration to prevent further stone formation. Dr. Saland advised placement of a gastrostomy to facilitate the unusually large amount of hydration at home. Keith Benkov, MD, Chief, Division of Pediatric Gastroenterology, met with the family during the patient’s recovery from the second stenting procedure. Oxalate quantification and a molecular genetic diagnosis confirmed the diagnosis of primary hyperoxaluria and allowed early identification of a yet-unaffected but at-risk sibling.

Seven months later, no additional stones have formed. The remaining stones are being monitored closely: two have required endourological removal, but there is good reason to believe the greatest risk of kidney failure will be averted through ongoing therapy and surveillance.

Reducing Central-Line Infections In Neonatal Intensive Care Unit

The Division of Newborn Medicine, under the direction of Division Chief Ian R. Holzman, MD, has instituted a number of practices to improve quality, safety, and patient outcomes in the neonatal intensive care unit (NICU). Leading these initiatives was a successful effort to reduce the incidence of central line-associated bloodstream infection (CLABSI) by direct faculty observation of all catheter insertions. Adopting this practice helped the Division achieve 270 consecutive days in 2015 with zero central-line infections. For all of 2013, the NICU had two central-line infections, for an overall rate of 1.0 infections/1,000 line days. By comparison, the New York State benchmark is 1.9 infections/1,000 line days.

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The Mount Sinai Health System is one of only two centers in New York State to have a TSC Clinic recognized by the Tuberous Sclerosis Alliance for providing integrated multidisciplinary care. The clinic includes subspecialists in neurology, neurosurgery, nephrology, ophthalmology, dermatology, social work, genetics, and nursing. The epilepsy team is headed by Steven M. Wolf, MD, Director of the Comprehensive Pediatric Epilepsy Center, in partnership with co-director Patricia Engel McGoldrick, NP, MPA; and Saadi Ghatan, MD, Director of Pediatric Neurosurgery at Mount Sinai. To learn more, visit www.nycepilepsyteam.org.
Providing Care after Cancer

Cure rates now approach 80 percent for childhood cancer, and an increasing number of survivors are living into adolescence and adulthood.

“These survivors, however, are at risk for long-term medical side effects of chemotherapy, which may involve the heart, or result in obesity and endocrine problems,” says Birte Wistinghausen, MD, Clinical Director of the Jack Martin Division of Pediatric Hematology/Oncology. “Because of their experience with cancer, they also require extensive psychosocial support.”

The Division has created an innovative survivorship program to coordinate follow-up care between Dr. Wistinghausen, a nurse practitioner, social worker, and nutritionist in collaboration with a pediatric and adolescent psychiatrist, and endocrinology and cardiology staff. “The most common challenges involve school adjustment, concentration, inactivity, and obesity,” says Dr. Wistinghausen.

The survivorship program helped Jasmin, a 15-year-old middle school patient with B-lymphoblastic leukemia, fully integrate into high school. She enrolled in a summer research program that Mount Sinai offers to underserved high school students, for example, and participated in Girl Talk, a special support group for teenage girls with chronic illness offered by the Child Life and Creative Arts Therapy Department. The follow-up program continues to provide Jasmin, now in college, with psychosocial support, weight counseling, and monitoring for long-term medical and psychosocial sequelae of treatment.