Innovations in Pulmonology

Rady Children's Hospital San Diego

Rady Children's - A comprehensive system focused solely on children.

PEOPLE

Dr. Cernelc-Kohan brings expertise to bronchopulmonary dysplasia, severe asthma clinics

Mateja Cernelc-Kohan, M.D., serves on the Bronchopulmonary Dysplasia (BPD) Clinic team in the Division of Respiratory Medicine at Rady Children's Hospital-San Diego and on the multidisciplinary Severe Asthma Clinic team. She is also an assistant professor of pediatrics at UC San Diego School of Medicine.

Dr. Cernelc-Kohan's clinical and research interests include children with difficult-to-control asthma and wheezing disorders, premature infants with lung disease and children with primary ciliary dyskinesia. As a member of the BPD team, along with pediatric pulmonologist Julie Ryu, M.D., a dietitian and a social worker, she provides outpatient management of very low birthweight premature infants with chronic lung disease of prematurity who are on supplemental oxygen at home after discharge from the neonatal intensive care unit.

In her role at the Severe Asthma Clinic, she works with allergists/immunologists Sydney Leibel, M.D., M.P.H., and Bob Geng, M.D., to care for children with severe and persistent uncontrolled asthma. The team also includes a pharmacist, social worker, respiratory therapist and a registered nurse.

In addition to her clinical responsibilities, Dr. Cernelc-Kohan teaches medical students and residents, and will be mentoring a fellow on a study. The study is evaluating a novel education tool to promote proper use of asthma inhalers in children.

Dr. Cernelc-Kohan earned her medical degree from University of Ljubljana, Slovenia, and completed her pediatric residency at University Children's Hospital in Ljubljana. In 2005, she moved to the United States and completed another pediatric residency at the University of Hawaii in Honolulu, followed by a pediatric pulmonology fellowship at UC San Diego. She joined Rady Children's Respiratory Medicine division in 2014.

PROGRAMS

Aerodigestive clinic offers expert, collaborative care

At the beginning of the year, Rady Children's Hospital launched the multidisciplinary Aerodigestive Clinic, bringing together specialists in pulmonology, gastroenterology and otolaryngology to provide diagnosis and management of a variety of disorders involving the respiratory and digestive systems. These specialists are Daniel Lesser, M.D., and Annabelle Quizon, M.D., from the Division of Respiratory Medicine; Matthew Brigger, M.D., M.P.H., from the Division of Otolaryngology; and Hayat Mousa, M.D., from the Division of Gastroenterology, Hepatology & Nutrition.

The conditions treated at the clinic include aspiration syndrome, feeding difficulties, failure to thrive, genetic syndromes associated with airway and/or gastrointestinal morbidities, congenital airway abnormalities, chronic cough and recurrent pneumonia.

A comprehensive evaluation and treatment plan are provided to patients in a single visit. Typically, the treatment is a "triple endoscopy under single anesthesia" procedure – an upper and lower airway bronchoscopy plus a gastrointestinal endoscopy. The clinic also offers a dysphagia study, esophageal manometry, esophageal impedance and pH studies, functional endoscopic evaluation of swallowing and specialized imaging studies (e.g. chest CT scan, UGIS).

About 75 patients have been seen to date, with parents providing extremely positive feedback, especially regarding the collaborative care approach and ease of obtaining a treatment plan in one visit. Healthcare costs are also reduced by an efficient use of resources, performing procedures in a single visit and/or under a single anesthesia session.

Ongoing activities and upcoming plans include developing and refining clinic protocols, maintaining a patient registry/database that can provide resources for research questions, attending regional, national and international aerodigestive conferences, and including additional specialties in patient care, such as speech therapy, occupational therapy, radiology, surgery and anesthesia. The clinic seeks to collaborate with other aerodigestive programs in California, such as those at Children's Hospital Los Angeles and Lucile Packard Children's Hospital Stanford, particularly on research. Benchmarking will also be used to evaluate how the clinic compares to these programs.

The clinic is held in Otolaryngology twice monthly on the first and third Wednesdays of the month. Triple endoscopy procedures are also scheduled twice monthly on the second and fourth Thursdays of the month. To refer a patient, please contact the Otolaryngology

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division at 858-309-7701.

RECOGNITION

Division achieves top 30 ranking in well-known national survey

U.S. News & World Report's comprehensive survey of the "best children's hospitals" for 2017-18 ranks Rady Children's Division of Respiratory Medicine as the No. 26 pediatric pulmonology program in the nation.

Programs were ranked based on the care of serious respiratory problems. Successful management of asthma, cystic fibrosis and muscular dystrophy, along with other clinical data, accounted for 85 percent of each hospital's score. The other 15 percent reflects nominations from pediatric specialists and subspecialists who responded to the surveys in 2015-17 and recommended the hospital for serious cases in their specialty.

The Division scored "excellent" on the following measurements:

- Success with asthma inpatients (relative survival, length of stay and readmissions)
- Nurse staffing (average hospital-wide number of R.N.s over 24 hours relative to the average number of daily inpatients)
- Advanced clinical services offered (such as sleep center, certified asthma educators and staff participation in patient care conferences)
- Clinical support services offered (such as rapid-response team and pediatric pain management, infectious disease and anesthesia programs)
- Has full-time subspecialists available (such as pediatric pulmonologists and sleep medicine specialists)
- Commitment to best practices (such as consensus management plan for six conditions, including cystic fibrosis, pneumonia and bronchiolitis)
- Adoption of health information technology (such as collecting and analyzing data to improve quality of care)
- Help for families (such as through a family resource center, family support specialists and pediatric psychologists)
- Enlists families in structuring care (such as through a parent advisory committee that meets frequently)

For all nine years it has been surveyed, the Division has ranked as one of the nation's best.

INNOVATIONS

Division part of first national chILD registry

Rady Children's Division of Respiratory Medicine was among the first programs to join the National Registry for Childhood Interstitial and Diffuse Lung Diseases (chILD), the first multicenter prospective study of chILD in the U.S. The study's preliminary findings show substantial morbidity in this population, emphasizing the need for disease-modifying therapies.

In 2016, the Children's Interstitial Lung Disease Research Network (chILDRN) established this longitudinal observational study using the IRBChoice and SMART-IRB platforms for reliance agreements. Pediatric patients with chILD diagnoses are enrolled through participating sites across the country, and cases are classified according to clinical, genetic, radiologic and pathologic criteria, with data entry into a secure REDCap database. The registry aims to advance knowledge on the prevalence, clinical features, management and outcomes of children with these rare lung diseases and to facilitate scientific discovery and additional research in this field.

To date, 194 subjects have been enrolled from 11 chILDRN sites, with eight additional sites to participate soon. Median age at time of enrollment is 77 months (IQR 36-143), with a slight male predominance at 55.9 percent. Specific chILD diagnoses and frequency include neuroendocrine cell hyperplasia of infancy (35.2 percent), surfactant dysfunction (12.6 percent; 11 *SFTPC*, eight *ABCA3* and three with *NKX2.1/TTF1* mutation or disruption), bronchiolitis obliterans (11.5 percent), disorders related to systemic disease (9.9 percent), growth abnormality (4.9 percent), pulmonary hemorrhage (2.7 percent), pulmonary interstitial glycogenosis (2.7 percent), disorder of the immunocompromised host (1.6 percent) and others (7.1 percent). Another 11.5 percent of cases were of unknown etiology or unclassifiable.

Chest CT was used for diagnosis in 83.1 percent of patients, lung biopsy in 44.6 percent and genetic testing in 24.3 percent. Median age at diagnosis was 15 months (IQR 5.25-60). Seventy-three percent of patients had used home supplemental oxygen at any time, and of those patients, 66 percent are currently on home oxygen. Fifteen percent of patients had a history of chronic mechanical ventilation.

Severity of Illness scoring at study enrollment included 23.9 percent of patients who were asymptomatic, 22.7 percent who were symptomatic with normal oxygen saturations under all conditions, 33.5 percent with abnormal saturations with sleep or

exercise, 13.6 percent with abnormal resting room air saturation and 6.3 percent with pulmonary hypertension. Approximately 50 percent of patients have history of failure to thrive.

Cases lacking etiology or classification highlight opportunities for unique disease identification and genetic discovery.