Children's Mercy KANSAS CITY

ENHANCING CARE FOR PATIENTS WITH PULMONARY AND SLEEP DISORDERS

A MESSAGE FROM Christopher M. Oermann, MD

Division Director, Allergy, Immunology, Pulmonary and Sleep Medicine

The Section of Pulmonary and Sleep Medicine has continued to evolve through the new reality of the COVID-19 (and now 20 and 21) pandemic. Like colleagues around the country, we have adapted to a rapidly changing health care environment in which standard operating procedure one month is outdated and unrecognizable the next.

Throughout this challenging period, we have continued to work toward the Children's Mercy Kansas City vision of creating a world of well-being for children and fulfill our mission to be a leading independent children's health organization dedicated to holistic care, translational research, breakthrough innovation, and educating the next generation of caregivers. We continue to expand existing programs and develop new programs to serve the needs of the children and families we serve while supporting the growth and professional development of our faculty.

I am excited to provide you with this update highlighting some of our accomplishments over the past year.

2021 HIGHLIGHTS

Our Cystic Fibrosis Care Center ranks **#1 and #2 out of 90 centers** with the highest percentage of patients enrolling in studies through the CFF Therapeutics Development Network.

BEST CHILDREN'S HOSPITALS U.S.News & WORLD REPORT PULMONOLOGY & LUNG SURGERY 2021-22

Ranked #37 in Pediatric Pulmonology and Lung Surgery by U.S. News & World Report.

The Pulmonary Medicine Division has been able to expand services in a spectrum of multidisciplinary clinics, including the **Super Q Express Clinic**, which serves children with **22q11.2 deletion syndrome** and **22q11.2 duplication syndromes**.

Team members hold **25 leadership positions with national professional organizations and committees**, including the Cystic Fibrosis Foundation, American Thoracic Society, American Academy of Pediatrics, American Academy of Sleep Medicine, U.S. News & World Report Pulmonology Working Group, and others.

FY 2021 By the Numbers

1,103 new Pulmonary clinic patient visits and **3,338** follow-up visits

2,011 new Sleep Medicine patient visits and **1,934** follow-up visits

1,834 sleep studies conducted

9 new cystic fibrosis patient diagnoses

3 patient transfers from other cystic fibrosis centers

61 new visits at the Cystic Fibrosis Care Center and **917** follow-up visits

CLINICAL EXCELLENCE SETTING NEW STANDARDS OF CARE

The Division of Pulmonary Medicine expanded its services in a spectrum of multidisciplinary clinics:

- **Super Q Express Clinic** for children with 22q11.2 deletion syndrome and 22q11.2 duplication syndromes.
- **JAWS (Jaw, Airway and Sleep) Clinic** for infants and children with complex craniofacial syndromes.
- **Pulmonary-Sickle Cell Clinic** for children with pulmonary complications of sickle cell disease.
- **Muscle-Nerve Clinic** for children with respiratory complications related to a variety of neuromuscular diseases, including Duchenne muscular dystrophy, spinal muscular atrophy and others.
- **Congenital Diaphragmatic Hernia Clinic** for the management of the long-term pulmonary complications associated with CDH.
- **Pediatric Tracheostomy and Home Ventilator Clinic.**
- **Narcolepsy Clinic.**
- **Prader-Willi Syndrome Clinic** to address the multifaceted management of children with PWS.

Pulmonary Hypertension and Single Ventricle Clinic for children with complex congenital heart disease and idiopathic pulmonary hypertension.

This clinic allows pulmonology specialists to collaborate with Children's Mercy cardiologists to diagnose and treat increased pressure in the blood vessels between a child's heart and lungs. The clinic allows families to see two specialists at once, with a nurse practitioner and coordinator to manage the complex needs of the child, from medication pre-approvals to ordering in-home equipment. The pulmonary hypertension team also provides consultations and care for patients who are in the hospital as well as regular outpatient clinics and coordination with home care services.

Jade Tam-Williams, MD, and patient at the AAIR Clinic

AAIR (Advanced Asthma Interdisciplinary Respiratory) Clinic for children with difficult to control and life-threatening asthma.

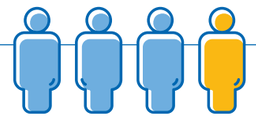
This clinic helps children 4 to 18 years old with severe asthma to better manage all aspects of their health. A multidisciplinary team of specialists (Pulmonary, Allergy, Pharmacy, Social Work and Environmental Health) collaborates to build an effective care plan for the patient.

In the past few years, the number of patients with severe asthma has steadily increased throughout the region, while access to care remained limited to Children's Mercy Kansas City hospital. However, in May 2021, the AAIR clinic expanded to provide telehealth visits at our Wichita and Joplin locations, allowing for greater frequency and more intense monitoring—especially for those requiring biologic therapies. These patients can receive spirometry testing and a full physical exam at their visit, facilitated by the on-site nurse, while the rest of the team participates remotely via Teladoc or Microsoft Teams.

CYSTIC FIBROSIS CARE CENTER

The Children's Mercy Cystic Fibrosis Care Center provides expert, family-centered care to infants, children and adolescents, and is fully accredited by the Cystic Fibrosis Foundation (CFF). Annually, **more than 270 children** from Missouri and Kansas come to our Care Center to receive specialized treatment. Our research team was recently recognized by the CFF Therapeutics Development Network for **ranking #1 and #2 among 90 centers** in two key outcome metrics, with the highest percentage of patients enrolling in studies. Additionally, we have several team members who participate in research aimed at developing new medications and therapies for people living with CF, including multiple publications in peer-reviewed journals and presentations at the 2021 North American Cystic Fibrosis Conference. The team also represents Children's Mercy by serving on CFF committees, participating in educational activities and mentorship programs, and engaging with the Therapeutics Development Network.

One out of every four families in our clinic participates in a research study.



LEADING THE WAY Through Research

Members of the Pulmonary and Sleep Medicine team are active locally, regionally and nationally. Our team applies for funding and conducts research into a wide range of clinical conditions, including asthma, sleep medicine/ narcolepsy, cystic fibrosis, children's interstitial and diffuse lung disease, primary ciliary dyskinesia and others.

In addition, our faculty holds **25 leadership positions with national professional organizations**, representing Children's Mercy and our profession with the following organizations:

- American Academy of Pediatrics
- American Academy of Sleep Medicine
- American Thoracic Society
- American Board of Pediatrics
- American Lung Association
- Children's Hospital Neonatal Consortium
- Cystic Fibrosis Foundation
- Gilead Sciences
- U.S. News & World Report Pulmonology Working Group
- Wake Up Narcolepsy

By the Numbers

35 published manuscripts

8 national presentations (3 oral and 5 posters)

16 invited lectures

17.39% of our cystic fibrosis patients participate in clinical trials.

Pulmonary and Sleep Medicine participated in **27 clinical trials** in 2021 and contributed data to **2 approved disease registries.**

Secured **$646,173** in external grant funding, including **$41,316** in NIH grants.

RESEARCH HIGHLIGHTS

Pediatric Pulmonary Medicine Workforce Concerns

In his role as the President of the Pediatric Pulmonology Division Directors Association (2019-2021), Christopher M. Oermann, MD, led an effort to understand and address the looming workforce crisis in Pediatric Pulmonary Medicine. This initiative resulted in the publication of a series of manuscripts describing the background of the crisis, efforts to increase interest in the field, the current state of fellowship training, and projections about the future of Pediatric Pulmonary Medicine. The task force also found significant geographic disparities in the availability of pulmonary medicine services for large parts of the country, noting a relative lack of pulmonologists compared to other pediatric subspecialties. Currently, there are widespread efforts underway to increase interest in the field, with many stakeholder groups involved.

- **Oermann CM**, Lahiri T, Peterson-Carmichael SL, Weiss P. The history of workforce concerns in pediatric pulmonary medicine. *Pediatr Pulmonol*. 2020 Sep 28. doi: 10.1002/ppul.25094.

- Nelson BA, Boyer D, Lahiri T, **Oermann CM**, Rama JA. A statement on the current status and future needs of the pediatric pulmonology workforce: Pipeline Workgroup. *Pediatr Pulmonol*. 2020 Oct 27. doi: 10.1002/ppul.25139. Epub ahead of print. PMID: 33107699.

- Esther CR Jr, **Oermann CM**, Ross KR, Weiss P. An assessment of fellowship training issues affecting the pediatric pulmonary medicine workforce. *Pediatr Pulmonol*. 2021 Jan 26. doi: 10.1002/ppul.25256. Epub ahead of print. PMID: 33497527.

Waking Up to Sleep Challenges in Children with Visual Impairment

Sleep problems are common in childhood, and children with visual impairment may be at increased risk given the potential disruption of normal circadian entrainment for light information processing. In a collaborative effort between Children's Mercy pediatric sleep medicine, ophthalmology and the National Federation of the Blind (NFB), researchers examined sleep challenges and interventions in children with visual impairment. Overall, most children exhibited sleep problems, with 72% having cycles of good and bad sleep, 69% having sleep patterns that caused significant stress for the family, and 89% scoring above clinical cutoffs on standardized sleep questionnaires. Those children with comorbid developmental delay were most at risk. Children with intact light perception still had a high prevalence of sleep problems, so clinicians should not presume normal sleep is assured with a normal light response. Helpful strategies for sleep problems included consistency in routine and supplemental melatonin.

- **Ingram DG**, **Cruz JM**, **Stahl ED**, **Carr NM**, **Lind LJ**, Keirns CC. Sleep challenges and interventions in children with visual impairment. *J Pediatr Ophthalmol Strabismus*. 2021 Aug 1:1-10. doi: 10.3928/01913913-20210623-01. Epub ahead of print. PMID: 34435902.

The Forgotten Pandemic: E-Cigarette and Vaping Product Use-Associated Lung Injury

The national attention garnered by the adolescent electronic cigarette (e-cigarette), vaping product use-associated lung injury (EVALI) pandemic has waned despite ongoing and pervasive use of these products by young people. Alvin V. Singh, MD, Director of the Pulmonology Fellowship Training Program at Children's Mercy, continues his efforts on multiple fronts to address this persistent but forgotten pandemic. He partnered with the American Lung Association and local school districts to educate teens about the dangers of e-cigarette use, vaping and nicotine addiction. Dr. Singh also provided commentary on the issue, especially regarding the lack of appropriate interventions by regulatory agencies. Additionally, he continues to develop a clinical database assessing the demographics of acute EVALI and post-EVALI recovery.

- **Singh A**. A commentary on adolescent electronic cigarette use and nicotine addiction. *Pediatr Pulmonol*. 2021 Dec;56(12):3580-3585. doi: 10.1002/ppul.25676. Epub 2021 Oct 4. PMID: 34520125.

Aminoglycoside Induced Ototoxicity Algorithm for People with Cystic Fibrosis

The Cystic Fibrosis Care Center Team implemented a standardized approach for monitoring people with cystic fibrosis (pwCF) exposed to ototoxic medications. Previously, less than 30% of pwCF exposed to ototoxic medications had an audiogram. After standardizing, patient monitoring increased to 98% of IV exposed and 58% of inhaled exposed pwCF. Hearing abnormalities were identified in 63% of pwCF that received IV ototoxic medications and 53% that received inhaled ototoxic medications. Fourteen pwCF received interventions including a referral to otolaryngology for hearing aid evaluation, modification of medications, or use of otoprotective agents. Most CF Care Centers lack standardized testing for hearing loss, which leads to it not being recognized. These discoveries were shared at national meetings and published to drive quality improvement.

- **Elson EC**, **Meier EM**, **Oermann CM**. The implementation of an aminoglycoside induced ototoxicity algorithm for people with cystic fibrosis. *J Cyst Fibros*. 2021 Mar; 20(2):284-287. doi: 10.1016/j.jcf.2020.08.002. Epub 2020 Aug 15. PMID: 32811788.

Genomic Answers for Kids Advances Rare Disease Research

The Children's Mercy Research Institute has released more than 2,300 pediatric rare disease genomes through its Genomic Answers for Kids (GA4K) program, which makes it one of the largest pediatric rare disease whole genomic datasets ever publicly shared.

To date, more than 3,700 patients have enrolled in the program, which has resulted in more than 18,000 new genomic analyses and more than 600 genetic diagnoses. In addition, the program has advanced research genomic analyses for children of 350 families with more common childhood diseases: cerebral palsy and Down syndrome.

The full pediatric data repository is shared in a real-time web interface through a comprehensive process, which gives researchers and clinicians low-barrier access to processed data with disease prioritized genetic changes.

"Giving access to our data allows researchers to link their own genetic findings so they can accept or reject hypotheses on their gene discoveries," said Tomi Pastinen, MD, PhD, Director, Genomic Medicine Center, Children's Mercy Kansas City. "Data sharing is the only way we'll make headway in the quicker delivery of results that are non-diagnostic today."

The GA4K program has helped hundreds of kids, like Celia, find a genetic diagnosis.

MEET THE TEAM

LEADERSHIP

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Children's Mercy KANSAS CITY

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LEARN MORE ABOUT THE SECTION OF PEDIATRIC PULMONOLOGY AND SLEEP MEDICINE.

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