May 2016

Innovations in Pulmonology

Rady Children's Hospital San Diego

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

PEOPLE

Dr. Akong leads Cystic Fibrosis Transition Program

Kathryn Akong, M.D., Ph.D, directs the Cystic Fibrosis Transition Program in the Cystic Fibrosis Center at Rady Children's Hospital-San Diego, in which young cystic fibrosis patients "graduate" to adult-centered care. With her background in both pediatrics and internal medicine, she is ideally suited for this role.

Dr. Akong has been recognized for many aspects of her work in cystic fibrosis, most recently receiving a Women Who Take Our Breath Away award from the local Cystic Fibrosis Foundation chapter, an honor that celebrates the important role that women play in the fight to cure and control the disease. She also received an award from the national Cystic Fibrosis Foundation for the care of young adult cystic fibrosis patients. For her research, she received a postdoctoral research grant from the UC San Diego Clinical and Translational Research Institute for her innovative work on neutrophil function in cystic fibrosis.

Recruited as the Division of Respiratory Medicine's first fellow in 2009, Dr. Akong subsequently earned a faculty appointment. She received her medical degree and a doctoral degree in genetics and molecular biology from the University of North Carolina and completed her residency training in both pediatrics and internal medicine at UC San Diego and Rady Children's.

Along with directing the transition program, Dr. Akong is the associate director of the Cystic Fibrosis Center, where she oversees all clinical and translational research activities.

RESEARCH

Discovery: Fluid maintenance mechanism in small airways

A study by Paul Quinton, Ph.D., and his colleagues is the first to provide a histo-physiological basis for a new concept to explain the mechanism of fluid maintenance in the small airways. This discovery should improve the understanding of both normal function and pathologies in small airway diseases, such as cystic fibrosis and asthma.

It is well known that the airways are coated with a thin layer of airway surface fluid (ASF) to keep the lungs clean and free of debris, but there is little understanding of the production and maintenance of ASF.

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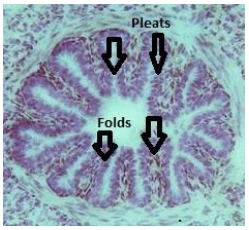
innovation belongs in every moment



For decades, it has been assumed that the airway epithelium could sense when it becomes too wet or dry and either secretes or absorbs fluid in response. It has also been assumed that the plicated structures of folds and pleats along the small airways (bronchioles) serve to ensure that the airways can more easily expand and contract as people inhale and exhale with each breath.

The researchers learned that the luminal epithelial cells do not reverse the directions of their fluid transport, but instead, cells in the folds constantly absorb, and cells in the pleats concurrently secrete fluid. As a result, the plications in the airway surface create a system that locally recirculates ASF. At every point along the small airways, ASF secreted in the pleats is reabsorbed as it covers the cells of the folds, recirculating ASF from pleat to fold, to pleat to fold, etc.

This simplicity of construct constantly maintains the proper level of liquid on the airway surface, giving air free passage through the airways without obstruction or dehydration, and without the unprecedented need for any cell to be able to acutely reverse the direction it transports fluid.

A disruption of fluid transport in either secretory or absorptive cells may result in pathologies. In cystic fibrosis, the airways have long been thought to be dry due to hyperabsorptive cells, even though in other CF tissues both secretion and absorption are inhibited by lack of CFTR (cystic fibrosis transmembrane conductance regulator). It now seems that airway dehydration in CF can be explained if secretion is inherently inhibited relatively more than absorption. In asthma or other "hypersecretory" conditions, the opposite may occur, in which stimulated secretion may exceed absorption to flood and obstruct the airways.

This improved fundamental understanding of how the airways function will result in improved care for a wide variety of airways diseases.

Read the abstract.

INNOVATIONS

Dr. Ryu partners with tech firms on personalized asthma care

Julie Ryu, M.D., clinical director of the Division of Respiratory Medicine and a physician informaticist at Rady Children's, has teamed up with Qualcomm and Propeller Health on an asthma pilot study to remotely monitor children with asthma and provide key data to families and physicians. The goal is to improve both doctor/patient communications and patient care.

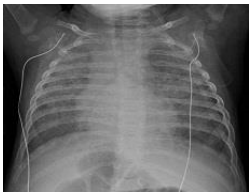
Propeller Health's sensor system

Patients will use inhalers equipped with Bluetooth-enabled sensors developed by Propeller Health. When paired with a smartphone or Qualcomm's 2net Hub device, the sensor tracks when and where the inhaler is used. The data is securely transmitted wirelessly via a Qualcomm connectivity platform to the patient's doctor.

PROGRAMS

Diffuse Lung Disease Program helps treat complex cases

Chronic cough, asthma, cystic fibrosis and pneumonia are familiar diagnoses to most pediatricians and pediatric pulmonologists. Some patients, however, present with vague respiratory symptoms, shortness of breath and unusual chest radiographs with abnormalities throughout both lungs (termed diffuse lung disease, or DLD). The differential diagnosis can be broad and daunting, and include rare disorders such as interstitial lung disease, alveolar proteinosis, ciliary dyskinesia and lymphatic disorders.



Diffuse lung disease

James Hagood, M.D., chief of the Division of Respiratory Medicine, has a career-long interest in rare lung diseases and currently devotes one clinic session monthly to solving and managing the medical mysteries of DLD. Along with participating in the national Childhood Interstitial Lung Disease (ChILD) Research Network almost since its inception, he is on the Scientific Advisory Board of the ChILD Foundation.

Dr. Hagood

To provide optimal care to children with DLD, Dr. Hagood has launched a comprehensive Diffuse Lung Disease Program, supported by a team of radiologists, pathologists and other subspecialists with expertise in rare lung disorders. The program will review prior records and imaging studies, provide a roadmap to getting to the correct diagnosis and appropriate treatment, and offer support for children and their families in navigating unfamiliar waters.

Because DLD cases are often complex, physicians are asked to call or email Dr. Hagood ahead of time with referrals so that the medical records and imaging studies can be reviewed prior to the first visit. He can be reached at 858-966-5846 or jhagood@ucsd.edu.

The team is developing a platform where patients and parents will have online access to the information through the Propeller Health mobile app as well as through a direct connection to their patient portal, MyChart at Rady Children's. MyChart at Rady Children's not only enables patients and families to access their medication usage and medical record online, but to also communicate with their medical team, refill medications and schedule visits.

Qualcomm Life's 2net™ Hub

With the sensor technology, physicians will have a more accurate picture of where medications are used and when they are needed. Dr. Ryu notes that younger children don't always keep track of their medicine usage, and teens may not tell their parents about it. The sensor eliminates the need to rely on memory or keep paper records. Patients will also be empowered, Dr. Ryu adds, by better understanding the relationship between their symptoms and medication usage in the context of their daily lives.

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