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Subject: OHSU Doernbecher Named Center of Excellence for Batten Disease
Date: Tuesday, January 18, 2011 10:56:51 AM

January 18, 2011

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OHSU Doernbecher Named Center of Excellence for Batten Disease

OHSU Doernbecher is recognized as national leader in diagnosis, treatment, research of this rare, currently fatal neurodegenerative disease

PORTLAND, Ore. — Oregon Health & Science University Doernbecher Children's Hospital has been named a Batten Disease Center of Excellence by North America's largest support and research organization for children diagnosed with a rare neurological disorder called neuronal ceroid lipofuscinosis (NCL), commonly referred to as Batten disease. OHSU Doernbecher now is one of four centers nationwide recognized by the Batten Disease Support and Research Association (BDSRA).

Batten disease is caused by genetic mutations, and children who inherit the defective gene are unable to produce enough of an enzyme, or protein, to prevent cellular waste build-up in the brain, which eventually causes the cells to cease functioning and die. Children with Batten disease appear healthy at birth, but as their brain cells die, they begin to suffer seizures and progressively lose motor skills, sight and mental capacity. Eventually, they become blind, bedridden and unable to communicate or function on their own.

The center of excellence at OHSU Doernbecher will provide a comprehensive multidisciplinary approach to children with Batten disease. Families and patients will experience a well-coordinated, organized team approach that will meet the specific needs of each child.

“Most physicians have never heard of Batten disease. We hope this first Batten Disease Center of Excellence west of the Mississippi will provide comfort to families in the region with affected children. When families contact the BDSRA Patient Advocacy Group for support after receiving the devastating news that their child has Batten disease, the BDSRA now will have a place to send the family for expert care,” said **Robert Steiner, M.D., F.A.A.P., F.A.C.M.G.**, an internationally renowned expert in rare metabolic diseases. Steiner is a Credit Unions for Kids Professor of Pediatric Research, and vice chairman for research in pediatrics at OHSU Doernbecher Children's Hospital.

As a Batten Disease Center of Excellence, OHSU Doernbecher will serve as a regional referral center for families in Oregon, Northern California, Washington, Idaho and beyond. Children referred to the center may be evaluated by specialists in metabolic disease/genetics, genetic counseling, neurosurgery, transplantation, neurology, epilepsy, gastroenterology, respiratory medicine, ophthalmology and social services, among others. The team also will educate and train other medical professionals in the evaluation and treatment of children with Batten disease.

“OHSU Doernbecher will provide the latest treatment advances and the opportunity to participate in clinical trials, some of which are unavailable anywhere else in the country. The team will work closely with families and their referring physicians to offer the best approach for managing their child’s plan of care, within their local communities whenever possible,” explained **Amira Al-Uzri, M.D., M.C.R.**, a nationally accomplished expert in pediatric kidney disease and transplantation; an associate professor of pediatrics (pediatric nephrology and hypertension); and the medical director of the pediatric kidney transplant program at OHSU Doernbecher.

This new designation further solidifies OHSU Doernbecher’s role as an international leader in pediatric rare diseases treatment and research.

In 2006 Steiner and the head of pediatric neurosurgery at OHSU Doernbecher, **Nathan Selden, M.D., Ph.D.**, co-led a landmark clinical trial to test the safety and preliminary effectiveness of StemCells, Inc’s purified human neural stem cells (HuCNS-SC®) as a potential treatment for Batten disease. Today Selden and others in the OHSU Doernbecher Batten Disease Center for Excellence are conducting the second phase of this groundbreaking study.

In 2009 OHSU Doernbecher was named one of 19 National Institutes of Health Rare Disease Clinical Research Consortia and is overseeing two major National Institutes of Health-funded clinical trials and six pilot research studies during the next five years on a group of rare diseases with a common biochemistry and health impact: sterol and isoprenoid diseases.

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The OHSU Doernbecher Batten Disease Center of Excellence comprises: **Steiner; Al-Uzri; Selden; Jessica Adsit, M.S.**, genetic counseling; **Dan Guillaume, M.D., M.S.**, pediatric neurosurgery; **Tom Koch, M.D.**, pediatric neurology; **Colin Roberts, M.D.**, epilepsy, neurology; **Peter Francis, M.D., Ph.D.**, ophthalmology, genetics; Ann Stout, M.D., pediatric ophthalmology, **Darren Janzen, Ph.D.**, neuropsychology; **Mina Nguyen-Driver**, neuropsychology; **Jeff Koh, M.D.**, pediatric anesthesiology; **Jeff Pollock, M.D.**, neuroradiology; **Holger Link, M.D.**, pulmonary and sleep medicine; **Yasemen Eroglu, M.D.**, gastroenterology, metabolism.

About the Batten Disease Support and Research Association (www.bdsra.org/)

The BDSRA is an international support and research networking organization for families of children and young adults with an inherited neurological degenerative disease known as Batten Disease began in 1987 by a group of parents looking for strength and support in a disease that only gives fear and uncertainty to those it affects. Our organization focuses on three things:

About OHSU Doernbecher Children’s Hospital (www.ohsудоernbecher.com)

OHSU Doernbecher Children's Hospital is a world-class facility that each year cares for tens of thousands of children from Oregon, southwest Washington and around the nation, including national and international referrals for specialty care. Children have access to a full range of pediatric care, not just treatments for serious illness or injury, resulting in more than 165,000 outpatient visits, discharges, surgeries and pediatric transports annually. Nationally recognized physicians ensure that children receive exceptional care, including outstanding

cancer treatment, specialized neurology care and highly sophisticated heart surgery in the most patient- and family-centered environment. Pediatric experts from OHSU Doernbecher travel throughout Oregon and southwest Washington to provide specialty care to some 3,000 children at more than 154 outreach clinics in 13 locations.

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