Visiting a medical exam room for the first time can be a daunting situation for a child. Such a room is often sterile, with large instrumentation that can appear threatening. They are asked not to touch things and to cooperate with processes that are unfamiliar to them. This setting can be especially intimidating to children with special needs.

For the pediatric ophthalmology department at UC Irvine, providing a comfortable environment is important in treating children with vision problems and neurobehavioral disorders. The new pediatric rooms at the Gavin Herbert Eye Institute will create an environment that sets an optimal standard of care for special needs children.

“The design for the new facility is going to accomplish two things,” says pediatric ophthalmologist Robert Lingua, MD, who currently practices at UCI Medical Center in Orange and on the main campus in Irvine. “It’s going to provide everything you would find in a pediatric ophthalmology facility anywhere in the United States—pediatric-sized instrumentation and objects that are age and ability appropriate—but it will also completely rethink the way you would structure an eye exam room.”

In contrast to the usually stark setting of a standard exam room, the pediatric rooms at the new eye institute will have a less intimidating, child-friendly setting that is focused mainly on a tabletop computer screen. Gone are the days of the big chair and giant microscope. With the use of computers and handheld equipment, concealable and nonthreatening in appearance, doctors will be able to gather needed information, all while the child watches a movie.

“The pediatric facility will mimic a setting that is familiar and comfortable to children and those with special needs, such as the classroom or home,” says Dr. Lingua. “A child with special needs and vision problems has an extremely blurry world to deal with on top of everything else. With the use of new advances in technology not previously employed in the pediatric age group, we are able to rapidly capture a vast amount of crucial information. The general approach is to appear as nonthreatening to the child as possible and to do whatever is needed to help ensure normal ocular development.”

ew pediatric facility at the Gavin Herbert Eye Institute takes kid-friendly approach.
How do you treat a disease so rare that it only affects about 2,000 people in the world? Jennifer Simpson, MD, Associate Professor of Ophthalmology at the Gavin Herbert Eye Institute, and Cystinosis Research Foundation (CRF) founder Nancy Stack are working toward finding treatments and a cure for cystinosis.

Cystinosis most commonly affects children and adolescents in the corneas, kidneys and the brain—causing blindness, renal failure and even death. The disease occurs when the body accumulates the amino acid cystine, a building block of proteins, which forms damaging crystals that appear during childhood and adolescence. Oral medication called cysteamine can reduce the crystals in the kidneys and other organs, but has no effect on reducing corneal crystals that cause pain, corneal scarring and loss of vision. No meaningful treatment currently exists for corneal cystinosis.

A Clear View
Dr. Simpson and Nancy Stack first met at a UC Irvine-sponsored cystinosis luncheon in 2007. Hearing Stack’s account about the frustration over limited corneal cystinosis therapies, and also about a newly developed cystinosis mouse model, Dr. Simpson realized how she could help. “Research and progress in rare diseases like cystinosis are often hampered by the difficulty in gathering enough patients to study and understand the disease. An animal model would give us an opportunity to test novel treatments and potentially accelerate research on cystinosis.”

This led Dr. Simpson to approach her Gavin Herbert Eye Institute colleague James Jester, PhD, the Jack H. Skirball Endowed Chair of Ophthalmology and Professor of Biomedical Engineering at UC Irvine. As a renowned expert in corneal imaging, Dr. Jester would be able to study cystine crystal formation in the mouse model using high resolution 3D photographs.

In a CRF-funded study using the animal model, Dr. Simpson and Dr. Jester were able to precisely show the progression of corneal cystinosis. The crystals form early in life and fill the corneas, which results in light sensitivity, eye pain, debilitating glare and inflammation that leads to corneal scarring and blindness. Establishing how corneal cystinosis progresses was a huge milestone that has been shared with other researchers around the world through the CRF International Symposium.

“A deep insight into how the disease acts in the mouse model, Dr. Simpson and Dr. Jester were able to work on the disease and make it accessible,” says Dr. Jester. “What we’ve learned is that we can continue to use the mouse model to test new treatments and potentially accelerate research on cystinosis.”

“We now have a more complete understanding of the disease process, instead of just a snapshot,” says Dr. Simpson. “Being able to visualize the crystals, see where they accumulate and follow them over time allows us to determine if a new therapy is actually working, and much more quickly than if we had to extract and extensively process the tissue.”

Setting Their Sights
Building on their findings, Dr. Simpson and Dr. Jester are currently investigating the effectiveness of new therapies including stem cells, which can potentially replace the corneal cells filled with cystine crystals. They are also working on new ways to deliver cysteamine eye drops, which currently have to be put into the eye every hour or every day.

“Long-lasting eye drops with a timed release could reduce this frequency, making cysteamine treatment more realistic for children and teenagers with corneal cystinosis,” explains Dr. Simpson.

“None of this would have been possible without the many opportunities for collaboration at the Gavin Herbert Eye Institute,” says Dr. Simpson. “By partnering with foundations, eye care companies and other institutions, we are working to deliver the best therapies for our patients. The freestanding eye institute that is now being built will further support these efforts by keeping physicians and researchers in close proximity. It will give us momentum when developing new therapies to treat people with both rare and common eye diseases.”

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By the time a child reaches the age of eight, a critical part of the visual development process has already been completed. If normal communication between the eye and brain has not been established, the child may suffer vision loss that can persist into adulthood or become permanent.

That is why it is important for parents to recognize the signs of conditions that may threaten normal ocular development. Strabismus and amblyopia are the most prevalent vision problems among children.

What is strabismus?
Strabismus is a misalignment of the eyes and commonly referred to as cross-eyed or wandering eyes. When misalignment occurs even in the slightest—in which one eye is turned in, out, up or down (Figure 1)—two separate images are communicated to the brain. This can be caused by weak eye muscle control.

A child may experience vision loss in the turned eye as the brain begins to select one dominant eye. Strabismus can be constant or intermittent. While intermittent strabismus may occur in a normally developing infant, it may require attention if it persists after six months. If the misalignment appears to be constant, it is best to seek an eye specialist immediately. Strabismus can result in other threats to a child’s vision, such as loss of depth perception.

What is amblyopia?
Amblyopia, often described as lazy eye, is the most common cause of vision problems among children in the United States. Three to seven percent of children suffer from this condition every year. It is the loss of one eye’s ability to make out clear details and results from a defect of visual information processing in the brain. In normal vision development, a clear image forms on the retina in the back of both eyes.

A child experiences anisometropic amblyopia when each eye focuses differently, one projecting a clear image and the other blurry. To avoid visual confusion from one eye projecting a blurred image, the child’s brain suppresses the image from the blurred eye as it does when strabismus occurs. If amblyopia is left untreated over time, this may lead to permanent loss of vision.

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An eye doctor treating amblyopia may employ some of the same strategies used when treating strabismus. Corrective spectacles, eye patching (Figure 2) or blurring drops over the stronger or dominant eye force the weaker eye to work harder at focusing and may help with vision recovery.

For amblyopia, however, there is no option for corrective surgery, which is why early detection and intervention is critical. A simple eye exam at an early age can help determine whether or not a child’s vision is developing normally.

How do you prevent or treat strabismus and amblyopia?
In strabismus, the type of corrective treatment an eye specialist suggests depends on whether the turned eye deviates all the time or just some of the time. For intermittent strabismus, corrective therapies that help the brain develop normal functioning of the eyes include judicious patching or special glasses. These treatments assist in strengthening the turned eye.

While early diagnosis and intervention is key in eliminating strabismus, restorative and reconstructive surgery for strabismus is an option at any age and is often covered by insurance.

CAUSES OF AMBLYOPIA

- **Refractive Amblyopia:** Nearsightedness, farsightedness or astigmatism that is very high in both eyes or different between the two eyes
- **Strabismic Amblyopia:** Misalignment of the eyes (in, out, up or down)
- **Deprivation Amblyopia:** Obstruction to vision, such as congenital cataracts or ptosis (droopy eyelid)

To make an appointment with a pediatric ophthalmologist, contact the Gavin Herbert Eye Institute at (949) 824-2020 (Irvine) and (714) 456-7183 (Orange), or visit www.eye.uci.edu to schedule online.

For more information on strabismus and amblyopia, visit the American Association of Pediatric Ophthalmology and Strabismus (AAPOS) resource center at www.aapos.org.
**RISING TO THE CHALLENGE OF RETINAL DISEASES: A DOCTOR’S EYE VIEW**

When a seemingly healthy male in his 50s came to the Gavin Herbert Eye Institute with blurred vision, Stephanie Lu, MD, could see there was something wrong with more than just his eyes. She ordered a carotid Doppler, which is an ultrasound image of arteries supplying blood to the brain. It showed severe blockage in his arteries that would have led to a major stroke, but could now be prevented.

Dr. Lu is an ophthalmologist, but concern for her patients reaches farther than their eyes; she also looks out for her patients’ overall well-being. “An hour-long surgery can make a difference in a person’s quality of life by giving back their vision,” says Dr. Lu. As Assistant Clinical Professor of Ophthalmology at the Gavin Herbert Eye Institute, she treats patients, performs sight-saving surgeries, researches treatments for age-related blindness and educates ophthalmology residents and medical students.

During medical school, Dr. Lu chose to specialize in diseases of the retina, which often cause permanent vision loss. She wanted to help people regain their vision through surgery using cutting-edge laser technology. This enables doctors to see blood flow in the eye while treating and diagnosing patients. Dr. Lu is also involved in clinical trials for new drugs that may last longer and be more effective than current treatments for retinal diseases. This could allow patients to receive fewer eye injections and spend less time at a clinic.

“The Gavin Herbert Eye Institute has unparalleled access to academic research and eye care companies in Orange County that help me bring the best and newest treatments to my patients,” says Dr. Lu. “Our faculty works hard to translate research into new sight-saving treatments.”

Working closely with the world-class clinicians, scientists and researchers at the Gavin Herbert Eye Institute to advance breakthrough technologies that can improve patient care...

**CHARITABLE GIVING KEEPS THE FUTURE IN SIGHT**

While a resident of San Clemente, Geneva Matlock, MD, was commuting to San Diego for treatment of macular degeneration, an age-related disease in the retina that affects central vision. She asked for a reference to the best care available closer to home and was pointed to Baruch Kuppermann, MD, PhD, at the Gavin Herbert Eye Institute.

“Dr. Kuppermann is internationally known for research on retinal diseases. He has been treating me for a decade and preventing my macular degeneration from progressing. I’ve also been treated at the institute by Dr. Roger Steinert, a true expert in ophthalmology, who performed my cataract surgery.”

After marrying her high school sweetheart, Dr. Matlock and her husband completed medical school, internships, residency and even practiced together before she moved to California in 1973 to practice anesthesiology at Hoag Hospital. She retired in 1985 and remains active in the medical and academic communities as a supporter and donor.

“I’ve watched the eye institute grow while living here in Orange County,” says Dr. Matlock. “I’m glad there will finally be a new building where they can consolidate under one roof. The institute helps everyone by creating a better life for those in danger of losing their sight—people like me.”

Dr. Matlock has committed $1 million to the Gavin Herbert Eye Institute via charitable gift annuity contracts, which will provide fixed income payments for the rest of her life. Afterward, the remainder of her gift will be used to benefit the eye institute.

“The doctors and researchers at the institute understand how important it is to continue learning how to treat eye disease,” says Dr. Matlock. “Donating to the Gavin Herbert Eye Institute allows me to help the entire community with new sight-saving treatments.”

**WORKING HAND-IN-HAND AND SEEING EYE-TO-EYE**

“We are honored to be associated with the day-to-day efforts of the UC Irvine Gavin Herbert Eye Institute,” says Kevin J. Buchler, Division Head of Alcon. “Alcon, the global leader in eye care, and the Eye Institute share a common goal—providing the best eye care. It is through the unequivocal collaborative efforts of both organizations that we are able to deliver quality eye care that helps improve people’s vision.”

“Bausch + Lomb is a world class eye center whose commitments to innovative thinking and integrity matched those of Bausch + Lomb—where our mission is helping people see better to live better,” says Robert Grant, CEO and President of Bausch + Lomb. “We wanted to support a world class eye center whose commitment to state-of-the-art patient care and their location in our community of Orange County, the international hub of ophthalmic research and technology, made them the perfect choice.”

“Alcon is pleased to work closely with the world-class clinicians, scientists and researchers at the Gavin Herbert Eye Institute to advance breakthrough technologies that can improve patient care.”

“DevicePharm is the perfect choice for an in-kind gift,” says Clay Wilemon, CEO and Chief Strategy Officer of DevicePharm. “The physicians and researchers at UC Irvine are already recognized worldwide for their leadership in vision research and patient care. The new institute will house all under one roof, in the nation’s most advanced environment for collaboration, and accelerate the rate at which sight-saving therapies move from an idea to a life-changing reality.”

We would like to thank our partners for their generous support. If you would like to help and make a gift of sight, please contact Janice Briggs at (949) 824-0091.
When Nancy Stack’s daughter Natalie was diagnosed with cystinosis as an infant almost 20 years ago, all the information she had on the rare, or orphan, disease was in a short pamphlet from the hospital. Cystinosis, a disease where cystine crystals develop and eventually destroy the major organs, was considered to be terminal with very limited treatment options.

Natalie’s 12th birthday wish was “to have my disease go away forever.” That wish prompted Nancy and her husband Jeff to found the Cystinosis Research Foundation (CRF) in 2003. “Our mission was to create a dynamic, ongoing cycle of cystinosis research, so this orphan disease would not be left behind,” says Stack. Today, CRF has raised nearly $16 million for research grants and funded more than 84 cystinosis studies, giving hope and support to families affected by the disease around the world.

Two grants have been awarded for studies on corneal cystinosis to Gavin Herbert Eye Institute doctors Jennifer Simpson, MD, Associate Clinical Professor of Ophthalmology, and James Jester, PhD, Professor of Ophthalmology and Biomedical Engineering.

“Together, the Gavin Herbert Eye Institute and the Cystinosis Research Foundation are helping create treatments for children with cystinosis here and all over the world,” says Stack. “Natalie is proud of the CRF grants that help bring treatment and hope to the entire family of people with cystinosis.”

For more information on CRF news and events, read the cystinosis research article (page 2) and visit www.cystinosisresearch.org.