Clinicians at UCSF Benioff Children’s Hospital San Francisco are excited by their recent move into a brand-new, freestanding, 183-bed facility at Mission Bay.

“We believe that moving into a space we’ve designed for the unique health needs of children – and working right next to our world-leading research enterprise – will enable us to improve the health of children in many more ways and for many years to come,” says Donna Ferriero, MD, chair of the Department of Pediatrics and physician in chief at UCSF Benioff Children’s Hospital San Francisco.

A Boon for Children Throughout the West
The new hospital increases the ability of UCSF Benioff Children’s Hospital San Francisco to serve patients throughout Northern California and the West with urgent, emergency and specialty outpatient care. The design of the facility:

- Optimizes technology with spacious operating rooms fully integrated with technology interfaces throughout the hospital.
- Helps prevent hospital-acquired infections because every room is a single-patient room, with hand-washing sinks and hand-sanitizer dispensers located near entryways. One-hundred-percent fresh air circulates throughout the hospital at all times.
- Reduces the risk of medical errors through the use of robotic technology and electronics to prepare, track and dispense bar-coded medications.
- Facilitates more attentive care with decentralized nurses’ stations that keep nurses closer to their patients.
- Fosters collaboration with telemedicine consults that can stretch from San Francisco to regions around the world.
Perspective

We learn early in medicine that when we spot opportunities to support and restore healthy lives for patients, we have to seize those opportunities. Now is the time for those of us at UCSF Benioff Children’s Hospitals.

As this issue’s cover story notes, the opening of our new facility at Mission Bay is the catalyzing opportunity – a genuine chance to usher in a new generation of care for children, both because of the way the design fosters better, safer care and because of the way the location makes real what we’ve always known to be true: a close, continuous, collaborative loop among physicians and scientists speeds important new discoveries to the lives of patients and families.

The other stories also speak to seizing opportunity when it knocks. They illustrate how we are leveraging breakthroughs in the treatment of pulmonary hypertension, easing the suffering associated with childhood headache, applying the latest evidence-based research to the treatment of undescended testes and expanding access to experts in rheumatology through close collaboration between our Oakland and San Francisco campuses.

As always, we are grateful for your partnership as, together, we embrace important changes to improve the health of our patients.

Donna Ferriero, MD  Hanmin Lee, MD
Physician in Chief  Surgeon in Chief
UCSF Benioff Children’s Hospital  UCSF Benioff Children’s Hospital

Discovering New Causes and Treatments for Pulmonary Hypertension

An infant with a severe congenital diaphragmatic hernia and pulmonary hypertension spent the first days of his life in the UCSF neonatal intensive care nursery. Eight years later – with the help of carefully calibrated medications and oxygen – he is able to do many of the activities most healthy 8-year-olds do.

With pulmonary hypertension, such success stories are rare, but they are becoming less so thanks to the efforts of clinicians and researchers like those in the UCSF Benioff Children’s Hospital pediatric Pulmonary Hypertension Clinic. There, pediatric physicians from neonatology, critical care medicine, cardiology, interventional cardiology and pulmonology – and a pediatric nurse practitioner and social worker – work together to care for children with primary and secondary pulmonary hypertension across all clinical settings. UCSF research supports the clinical work.

“Accumulating experience within the field has identified a number of diseases that can cause pulmonary hypertension,” says pediatric critical care specialist Jeffrey Fineman, MD, chief of Critical Care at UCSF Benioff Children’s Hospital San Francisco. “In fact, there is a spectrum of diseases, which means treatments must vary; but the good news is that our therapeutic options are increasing.”
Causes and Indicators
At present, potential causes and contributing factors include:

• Congenital heart defects
• Genetic predisposition
• Lung disease, such as bronchopulmonary dysplasia
• Abnormal lung development, such as congenital diaphragmatic hernia
• Left-sided heart disease, such as mitral valve disease
• Autoimmune diseases, such as lupus and scleroderma
• Sleep apnea and other breathing disorders
• Blood diseases, including clotting disorders and sickle-cell anemia
• Liver disease
• Certain drugs or toxins, such as diet pills
• HIV

“In teens or young adults, the condition can be associated with infections like HIV or drugs like we saw with the diet medication fen-phen,” says Fineman. “Infants with chronic lung disease are an increasingly recognized population.” He suggests that some populations — such as a premature infant who goes home on oxygen — should be proactively screened.

“The need for a Team Approach
The UCSF team develops care plans with families and referring physicians based on functional status, comorbidities and the family’s wishes.

“Because pulmonary hypertension is rare, patients tend to be spread out among different subspecialists,” says Fineman. “Each patient is different, the condition is chronic, and so the management is complex, requiring sophisticated continuous treatment across multiple specialties, which is why care coordination is so important.”

“We also participate in a Pediatric Pulmonary Hypertension Network, which UCSF began with seven other North American centers,” says Fineman, who co-founded an annual international conference that pairs patients and families with clinical experts and researchers.★

For more information, contact PNP Emma Olson at (415) 476-6795 or Dr. Keller at (415) 514-3192.

“Each patient is different, the condition is chronic, and so the management is complex, requiring sophisticated continuous treatment across multiple specialties.”
When to Refer

Early referral is essential. Lionetti says the American College of Rheumatology provides extensive referral guidelines for pediatricians, which include recommendations that pediatricians refer for:

- **Diagnostic evaluation of patients with unclear diagnoses**, especially those with prolonged fever, loss of function, inability to attend school and/or regression in physical functioning. “Lupus, other connective tissue disorders or vasculitis often present strangely and not always in the same way,” says Lionetti. “When we find these illnesses, it is important to start treatment in a timely manner. Collaboration with the primary care physician allows for improved care.”

- **Diagnostic evaluation and long-term management of juvenile arthritis, spondyloarthropathies and ankylosing spondylitis**. “We often can provide immediate relief by performing a joint injection. Other medications that are often prescribed have changed the face of rheumatology; we very rarely see long-term joint damage or disfigurement, which was previously a common sequela,” says Lionetti, who has significant experience in treating the condition, including with the use of immunosuppressive drugs when indicated.

- **Diagnostic or treatment plan evaluation for autoimmune disorders associated with other primary diseases**, such as immunodeficiency, neoplasm, infectious disease, endocrine disorders, genetic and metabolic diseases, post-transplantation, cystic fibrosis and arthritis associated with birth defects.

- **A second opinion or confirmatory evaluation for families requiring subspecialty input** to cope with the disease process, accept the treatment plan, allay anxiety and provide education.

“Early diagnosis and treatment of childhood rheumatic diseases is essential for minimizing their impact on a young person’s life,” says pediatric rheumatologist Geraldina Lionetti, MD. “Without early, aggressive treatment, many of these diseases can significantly disable children and, in some cases, even be fatal.”

UCSF recently expanded its pediatric rheumatology clinic to UCSF Benioff Children’s Hospital Oakland in an effort to provide early and expert care. With the Oakland opening, an expanding array of clinics – which include those already established at UCSF Benioff Children’s Hospital San Francisco, UC Davis Medical Center and the Shadelands Clinic in Walnut Creek – provide patients with suspected or confirmed disease convenient options for timely diagnosis and management.

“Aiming for a Lifetime of Prevention

Pediatric rheumatic diseases – from juvenile rheumatoid arthritis through systemic lupus, erythematosus, dermatomyositis and vasculitis – differ from their adult versions because of the impact these diseases can have on developing immune, neurologic and musculoskeletal systems. They can also have a significant psychosocial impact.

“Our goal is to control disease activity and, where possible, achieve remission so we can preserve a normal childhood for these patients and their families,” says Lionetti.

At the UCSF clinics, treatment teams include pediatric rheumatologists, physical and occupational therapists, ophthalmologists, social workers and nurses, who coordinate community-based follow-up services when needed. “It’s a multisystem approach,” says Lionetti.

“With the Oakland opening, an expanding array of clinics – which include those already established at UCSF Benioff Children’s Hospital San Francisco, UC Davis Medical Center and the Shadelands Clinic in Walnut Creek – provide patients with suspected or confirmed disease convenient options for timely diagnosis and management.”

“Without early, aggressive treatment, many of these diseases can significantly disable children and, in some cases, even be fatal.”
According to a 2009 study published in the *Journal of Child Neurology*, 17.1 percent of children experience frequent or severe headaches each year. While migraines are the most common, children can also suffer from periodic and refractory:

- Tension-type headaches
- Cluster headaches
- New daily persistent headaches
- Secondary headaches – e.g., idiopathic intracranial hypertension

Despite their prevalence, pediatric headaches are often misunderstood and undertreated, according to headache specialist and pediatric neurologist Amy Gelfand, MD, director of pediatric headache, UCSF Headache Center at UCSF Benioff Children’s Hospitals.

For example, she says there is little evidence supporting the use of restrictive diets in children with migraine, and that the potential for overuse and dependency with opioids and barbiturates outweighs the benefits of those treatments.

She also notes that catching periodic headaches before they become refractory may offer clinical advantages, so early referral for all types of pediatric headache is important.

**An Expanding Definition of Migraine**

One way to catch migraines early is to recognize that they can appear differently in young children than in older children or adults. The most recent criteria from the International Classification of Headache Disorders identify a family of disorders – from abdominal migraine to cyclical vomiting syndrome, benign paroxysmal torticollis and benign paroxysmal vertigo – as episodic syndromes that may be associated with migraine. There is even evidence that some infant colic is related to migraine.

“We see school-age children with recurrent episodes of abdominal pain accompanied by nausea and vomiting,” says Gelfand. “When GI testing is negative, we often find sensitivity to light or sound that is indicative of migraine…. But because each patient is different, a detailed patient and family history-taking by an experienced clinician is the main component of diagnostic testing.”

**Expanding the Options**

At the UCSF Headache Center, patients benefit from expert diagnostic services, as well as a full range of stepped treatment capabilities, including:

- Access to a pediatric sleep specialist and education and support for lifestyle changes.
- A selection of acute and preventive outpatient migraine therapies. “For babies, we look to behavioral interventions, such as swaddling or turning down lights and noise,” says Gelfand. “We can use Tylenol for children as young as age 4, and triptans are FDA labeled for ages 6 to 17.”
- Greater occipital nerve injections
- Intravenous (IV) dihydroergotamine (DHE)
- IV lidocaine

For many patients, these treatments will not entirely eliminate headache attacks, but they can dramatically improve quality of life by reducing headache occurrence to a few times a month.

If children don’t respond to first-line outpatient therapy, the next step is typically a greater occipital nerve injection, which has been effective in approximately two-thirds of adults treated at the Headache Center.

“We have a lot of experience doing these injections in children,” Gelfand says.

**Inpatient Procedures and Emerging Protocols**

If the nerve injection is not effective, neurologists at the Headache Center consider two possible inpatient treatments. IV DHE is a five-day procedure usually accompanied by antiemetics, while IV lidocaine is administered in the Pediatric Intensive Care Unit.

Finally, physicians at UCSF are developing an emergency-room protocol for children presenting with severe headaches. “Once the protocol is validated, we expect to disseminate it to any interested facility,” says Gelfand.

For more information, contact Dr. Gelfand at (415) 353-8393 or at GelfandA@neuropeds.ucsf.edu.
Important New Evidence-Based Guidelines for Treating Undescended Testes

“The American Urological Association recently issued important new guidelines for the diagnosis and treatment of undescended testes [cryptorchidism],” says Laurence Baskin, MD, chief of Pediatric Urology at UCSF Benioff Children’s Hospital San Francisco.

Experts at UCSF believe that one of the most important elements of the new guidelines is the recommendation that early referral and treatment improves the odds of success. Both diagnosis and treatment should optimally occur between 6 and 12 months of age.

“In addition, given the physical and emotional risks, it’s important that an experienced team implements the guidelines,” says Baskin.

Identifying the Problem

“When a physical exam on a baby identifies concern about undescended testes, we wait until at least 6 months of age before recommending intervention, because up to two-thirds of newborns with the diagnosis will have spontaneous descent after birth,” says Baskin. “This leaves a true incidence of undescended testes at approximately 1 percent of full-term newborn males.”

The contributors likely include genetic, hormonal and anatomical factors; children with severe hypospadias are at increased risk for acquired cryptorchidism and retractile testes. Procedures that can help with the diagnosis and treatment include:

• Ultrasound, which is the most common, but due to its poor sensitivity, it is not recommended, especially because it does not change patient management, which in most instances will be surgical correction, typically laparoscopic orchiopexy.

• Magnetic resonance imaging (MRI) has much higher sensitivity than ultrasound but requires general anesthesia and experienced interpretation – and has cost and availability challenges as well.

• “Since surgery to treat the conditions is the first-line approach – and diagnostic laparoscopy has 100 percent sensitivity – in most cases we recommend the laparoscopic approach because we can diagnose and treat undescended testes surgically at the same time,” says Baskin.

Why Surgery Is Still the Preferred Treatment

While researchers are exploring the use of hormonal therapies in an effort to avoid surgery, at this point, says Baskin, in randomized studies, human chorionic gonadotropin (hCG) has been effective no more than 25 percent of the time. “In contrast, surgical correction has proven to be 90 percent effective and so remains the preferred approach.”

He adds that experience matters for the surgical outcomes. “It’s definitely a volume issue, and so doing this in an expert center – that works closely with a pediatric anesthesia team, as we do here at UCSF – makes the procedure safer and more likely to achieve positive outcomes.”

Finally, because surgery reduces but does not eliminate the risk of long-term issues, such as infertility and testis cancer, appropriate counseling and follow-up of the patient and family are essential.

For more information, contact Dr. Baskin at (415) 353-2200.
• Eases the anxiety and fear that often accompany pediatric imaging because children undergo CT scans and MRIs in a specialized imaging suite that allows them to select a theme of sights and sounds, which transforms their diagnostic visit into an adventure tailored to their interests. This minimizes their anxiety and restlessness, resulting in better-quality scans.

• Empowers patients with multimedia walls in every room that facilitate everything from patient-clinician communication and patient education to access to entertainment and the Internet, as well as the ordering of housekeeping and food services.

Emergency Transport
"A major addition is our ability to bring an intensive care unit to all patients at referring facilities – and the ability to quickly and safely deliver those patients to Mission Bay," says Hanmin Lee, MD, chief of Pediatric Surgery and surgeon in chief at UCSF Benioff Children’s Hospital San Francisco.

Experienced teams trained in advanced practice procedures for pediatric and neonatal care staff a fleet that includes:

• A new medical transport helicopter, which is available 24/7 to respond to any hospital referral within a 150-mile radius of the Mission Bay facility.

• King Air fixed-wing aircraft and Learjet capability for transports greater than 150 miles. A dedicated ambulance meets the planes at San Francisco International Airport.

• Critical care monitoring equipment, including an ambulance that has ECMO (extracorporeal membrane oxygenation) capabilities, climate-controlled medication storage and rapid-cooling technology that supports protocols for keeping all patients at target temperatures.

“This really is a next-generation facility – a dream not just to work at, but one that truly helps improve patient care, patient safety and the patient experience,” says Lee. ★

For more information, visit ucsfmissionbayhospitals.org.
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