Cells from the first baby diagnosed with severe combined immunodeficiency through a unique newborn screening program. See story on Page 2.
Physician Referral Center (Consultations • Referrals • Transports)
Toll-free (800) 266-0366
chw.org/refer

When you want to contact a Children’s Hospital of Wisconsin specialist – for consultation, referral or transport – one number is all you need to know.

The 24-hour physician call center connects you to a nurse clinician who will expedite your request. We look forward to working with you to care for your pediatric patients.

Additional resources available online
chw.org/provider

• Referral form.
• Outpatient order and consent forms.
• Patient handouts and teaching sheets.
• Medical care guidelines.
• CME information.

Family Accommodations Program
Toll-free (800) 556-8090

We understand that traveling with a sick child to a new city can be stressful for families. To make a stay at our hospital as easy as possible, we have developed a program to help out-of-town families coordinate their travel and lodging arrangements in Milwaukee. Negotiated discounts are available through the program.
2 Primary immunodeficiency overview

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Primary immunodeficiency results from inherited genetic defects involving the immune system and immune responses. In aggregate they occur in as many as 1 in 2,000 live births. Primary immunodeficiency typically is classified according to the principle immunologic mechanisms affected. (See Table 1.) The early diagnosis of primary immunodeficiencies is essential and will markedly reduce the morbidity and mortality associated with recurrent infections. For example, severe combined immunodeficiency is uniformly fatal if undiagnosed, but long-term survival rates of more than 90 percent have been achieved with early bone marrow transplantation (at less than 3 months of age). Treatment with high-dose intravenous gamma globulin (IVIG) can prevent many of the infectious sequelae in patients with primary antibody deficiency states.

Many routine childhood vaccines use attenuated organisms. The use of such vaccines may lead to disseminated infections in patients with severe cellular immunodeficiencies and are contraindicated. Consultation with a clinical immunologist is indicated when attenuated vaccines are considered for patients with other forms of primary immunodeficiency. Irradiated, cytomegalovirus (CMV)-negative, lymphocyte-depleted cellular blood products should be administered to patients with cellular or combined immunodeficiency.

Several factors should influence the decision to evaluate patients for primary immunodeficiency. (See Table 2.) For example, a patient with a first pneumonia and a history of intractable sinus disease or recurrent gastrointestinal infections should be evaluated for primary immunodeficiency. The prevalence of autoimmune disorders is increased in many primary immunodeficiencies and autoimmunity in association with recurrent infections also warrants an evaluation for primary immunodeficiency.

There are several clues in the history and clinical presentation of patients with primary immunodeficiencies that suggest the type of immunologic defect present. (See Table 1.) The onset of diseases associated with cellular immunodeficiencies usually begins soon after birth or in early infancy. Infections with opportunistic or unusual pathogens, mycobacteria, disseminated viral infections and severe oral candidiasis may occur. Diarrhea and malabsorption are common and growth is delayed. In contrast, the onset of infections in patients with antibody deficiencies,

<table>
<thead>
<tr>
<th>Type of immunodeficiency</th>
<th>Example</th>
<th>Mode of presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antibody</td>
<td>XLA, CVID</td>
<td>Upper and lower respiratory tract infections (encapsulated and atypical bacteria), giardiasis</td>
</tr>
<tr>
<td>T cell</td>
<td>DiGeorge Syndrome</td>
<td>Abnormal facies, lymphopenia, recurrent infections</td>
</tr>
<tr>
<td>Combined B cell and T cell</td>
<td>SCID (multiple causes)</td>
<td>Opportunistic infections, thrush, intractable diarrhea, failure to thrive</td>
</tr>
<tr>
<td>Cellular/complex</td>
<td>IFN-g/IL-12 axis</td>
<td>Atypical mycobacterial and salmonella infections</td>
</tr>
<tr>
<td>Phagocyte</td>
<td>CGD</td>
<td>Recurrent abscesses</td>
</tr>
<tr>
<td>Complement</td>
<td>C5-C9</td>
<td>Recurrent <em>N. meningitidis</em> infections</td>
</tr>
</tbody>
</table>
such as x-linked agammaglobulinemia (XLA), may be delayed for several months until maternal antibodies no longer are present. Recurrent and severe upper and lower respiratory tract infections are the usual mode of presentation. Complement deficiencies may present in a similar manner as antibody deficiencies or with recurrent Neiseria sp. infections. In contrast to patients with cellular immunodeficiencies, growth usually is intact in patients with complement or antibody deficiencies.

One common misconception is that opportunistic pathogens are overwhelmingly the cause of most infections in patients with primary immunodeficiencies. In fact, many infections in immunodeficient patients occur with pathogens that are common in the community. However, these infections may be of unusual severity and respond poorly to therapy. Finally, it also is essential to exclude secondary immunodeficiencies (such as lymphoproliferative disorders and malignancy, malnutrition, immunosuppressive drugs, protein losing states) in patients that present with recurrent infection.
Diagnostic workup
There are a number of readily available and fairly inexpensive screening tests that should be used in the evaluation of a patient for possible immunodeficiency. (See Table 3.) Abnormalities found in these screening tests indicate the need for more sophisticated studies in collaboration with a clinical immunologist. Consultation with a clinical immunologist is imperative when there is any question regarding interpretation of screening tests. The goal in the evaluation of immunodeficient patients should be to define the specific genetic abnormality whenever possible.

Other considerations
Education is important for optimal outcomes for patients and families with primary immunodeficiencies. They need to be well informed about the inheritance, clinical manifestations and treatment of their specific primary immunodeficiency. Patients and families should establish long-term relationships with health care providers, including physicians, nurses and social workers, to obtain the best outcomes for their diseases. Important resources for families and physicians include the Immune Deficiency Foundation (www.primaryimmune.org) and the Jeffrey Modell Foundation (www.jmfworld.org).

Table 3

<table>
<thead>
<tr>
<th>Immune function</th>
<th>Enumeration/flow cytometry</th>
<th>Functional tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cellular immunity</td>
<td>• CBC with differential enumeration of T cells (CD3) and NK cells (CD16 and CD56)*</td>
<td>• Cutaneous delayed hypersensitivity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Enzyme assays (ADA, PNP)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• FISH for 22q11 and 10p11 deletion</td>
</tr>
<tr>
<td>B cells</td>
<td>• Enumeration of B cells (CD19+ or CD20+)*</td>
<td>• IgG, IgA, IgM levels*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Antibody response to immunization</td>
</tr>
<tr>
<td>PMN</td>
<td>• CBC with differential*</td>
<td>• Oxidase function (NBT, DHR, chemiluminescence)*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Enzyme assays (MPO, G6PDH)</td>
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<tr>
<td></td>
<td></td>
<td>• Phagocyte function</td>
</tr>
<tr>
<td>Complement</td>
<td></td>
<td>• AH50 (alternative pathway)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• CH50 (classical pathway)</td>
</tr>
</tbody>
</table>

* Preferred initial screening tests.
Device intervention in the pediatric catheterization lab
by Susan Foerster, MD

Susan Foerster, MD, is a pediatric cardiologist at Children’s Hospital of Wisconsin, an assistant professor of Pediatrics (Cardiology) at The Medical College of Wisconsin and a member of Children’s Specialty Group.

The last two decades have resulted in vast advances in the field of cardiac intervention, particularly with regard to nonsurgical closure of holes and shunts. Almost all device closure procedures involve less pain, give a better cosmetic result and result in less time in the hospital than with surgical closure. However, the degree of efficacy and risk also needs to be close to or better than those achieved with open techniques to gain wide acceptance. The cost often is similar between the two groups. Long-term data is needed to ultimately determine the better course of action, and this data still is being accrued.

The most common device intervention has been closure of the nonneonatal patent ductus arteriosus (PDA). Most neonatal ducts are treated medically first and then surgically if there are symptoms deemed related to the duct, particularly in the premature infant. Beyond the neonatal period, most ducts are associated with no clinical symptoms and do not respond to medical attempts at closure. They often present as a heart murmur or are found when imaging is performed for another indication. Rarely, patients have congestive heart failure symptoms, such as poor weight gain, frequent lung infections or failure to thrive. If there is dilation of the left heart on ultrasound, reflecting a sizable increase in pulmonary blood flow, almost all cardiologists will recommend device closure. This can be accomplished easily in patients weighing more than 5 kg in most circumstances, although it is somewhat easier in the larger toddler.

There is much more controversy about the need to close small ducts, particularly when they are not audible. While the argument for closure used to be based on the need for subacute
bacterial endocarditis (SBE) prophylaxis, this no longer is recommended based on the most recent American Heart Association guidelines. SBE in an isolated small ductus almost never occurs. The decision on whether to proceed often is based on the preference of the referring cardiologist given the very low risk of complications with device closure. Removing a disease label also is a consideration for future insurance concerns.

At this time, there are two broadly used options for ductal device closure. The first method involves placement of one or more vascular coils and is used for the smallest ducts (diameter less than 1.5 mm). Given the greater difficulty with implantation, as well as a higher risk of embolization or a residual shunt with larger ducts, most interventionalists prefer to use a preformed ductal device for larger vessels, usually an AMPLATZER® Ductal Occluder. (See Figure 1.) These come in a variety of sizes and generally are very easy to use, with excellent results. In very small children, it is important to ensure no obstruction to the left pulmonary artery or descending aorta after placement.

Another common indication for an occlusion device is for closure of atrial septal defects (ASD). In childhood, ASDs often are asymptomatic, with a soft pulmonary outflow murmur or a widely split S2. Patients with larger defects can have increased pulmonary blood flow and an increased chance of respiratory symptoms and frequent infections. Older patients with hemodynamically significant defects have higher rates of symptomatic heart failure, pulmonary hypertension and atrial arrhythmias if the defects are not closed by early adulthood.

At Children’s Hospital of Wisconsin, we generally look for echocardiographic evidence of dilation of right heart structures when we recommend closure. To qualify for nonsurgical closure, defects must have sufficient atrial septal margins to hold the device without interfering with the atrioventricular valves or systemic/pulmonary veins. Endocardial cushion (primum) defects and sinus venosus defects are not suitable. The AMPLATZER® Septal Occluder (See Figure 2.) is by far the most commonly used in the U.S. and worldwide, largely relating to its ease of delivery, relatively small sheath size for implantation and retrievability should repositioning be required.

Ultrasound guidance generally is used in addition to fluoroscopy to assist in assessing the candidacy of each defect for device closure. As with any procedure, there can be rare complications with use of this device, including erosion of the device into the aorta in about 1 in 1,000-5,000 patients and acquisition of EKG changes or even heart block in a small group. This usually is with oversized devices or those used to close relatively large defects and can be diminished through patient selection. The HELEX® device (W.L. Gore & Associates, Inc., Flagstaff, Ariz.) is another common alternative for smaller defects and is less prone to the above issues, although it is more challenging to place and requires a larger sheath.
Other mainstream devices have been developed to occlude various types of collateral vessels or other vascular anomalies and, less commonly, for closure of muscular ventricular septal defects (VSDs), especially when in a hard-to-reach area. Currently, devices are in development to close VSDs near the aortic valve (perimembranous) as well as a modification of the PDA device that may allow for its use in the neonate. Neither of these applications is “ready for prime time,” and they will have to be very good given the relative low risk of surgical interventions in these two areas.

References

Heart Matters
Children’s Hospital of Wisconsin’s Herma Heart Center is one of the nation’s top programs for medical and surgical treatment of congenital heart defects and heart disease in children. As one of the largest pediatric cardiac programs in the United States, we have set national benchmarks for surgical outcomes.
Early recognition and treatment of scarring skin disorders: Focus on acne vulgaris and morphea
by Yvonne Chiu, MD

Yvonne Chiu, MD, is a pediatric dermatologist at Children’s Hospital of Wisconsin, an assistant professor of Dermatology at The Medical College of Wisconsin and a member of Children’s Specialty Group.

Introduction
Scar formation is an unfortunate consequence of many skin disorders. Acne vulgaris is one of the most common skin conditions seen by pediatric dermatologists. While 85 percent of adolescents have acne, only 20 percent develop severe scarring disease. Perhaps the common nature of acne and the belief that it is a rite of passage prevents early diagnosis and referral.

In contrast to acne, morphea is a rare skin disease with yearly incidence rates of 25 per million and lifetime prevalence rates of 2,000 per million. Although rare, morphea is invariably scarring, and 10 percent of affected patients develop functional disability as a result of the scars. Because of its rare nature and insidious onset, morphea usually is not recognized immediately and substantial scarring already may have occurred by the time of referral.

Regrettably, the treatment of skin scarring is limited, often expensive and will improve but not reverse the scar. The most important management option is early recognition and treatment, before further scarring occurs. In general, any child with a scarring skin disorder should be referred in a timely manner to a pediatric dermatologist for management. (See Table 1.)

Acne vulgaris
Acne vulgaris encompasses a wide range of clinical features. Open comedones (whiteheads) and closed comedones (blackheads) form when keratinocytes and sebum accumulate within a hair follicle. Inflammatory acne lesions may be relatively small and superficial, 1-5 mm, erythematous papules and pustules. Severe inflammatory disease is characterized by large, deep, tender nodules and cysts.

Scarring from acne takes many forms. Most patients should be reassured that postinflammatory hyperpigmentation and persistent erythema are not permanent and slowly will resolve. Icepick scarring may result from superficial papules and pustules, and extremely rarely from comedones. Pitted scars typically are caused by nodulocystic lesions and are deeper and wider. Hypertrophic and nodular scars may develop from severe disease on the trunk. In addition to physical scarring, acne vulgaris and its subsequent scarring can have important effects on quality of life and self-image.
Comedonal and mild papulopustular acne vulgaris typically are treated with topical medications alone. More severe papulopustular and nodulocystic disease require treatment with systemic agents. Oral antibiotics, typically of the tetracycline class, have anti-
*Propionibacterium acnes* and anti-inflammatory effects. Hormonal therapy such as estrogen-containing oral contraceptives and spironolactone may be helpful for female patients. Isotretinoin is the gold-standard for severe nodulocystic disease that is refractory to other therapies and also may be beneficial for less severe but scarring disease. Isotretinoin results in complete clearance of acne in 85 to 90 percent of treated patients, and in contrast to other acne therapies that require continued treatment to maintain clearance, isotretinoin can induce a lasting remission of acne vulgaris. The typical dosing is 0.5-1 mg/kg/day over a 4-6 month course for a cumulative dose of 120-150 mg/kg. Isotretinoin has numerous side effects, including teratogenicity, and it is best managed by experienced practitioners. In an attempt to reduce fetal exposure to isotretinoin, the U.S. Food and Drug Administration has instituted a mandatory iPLEDGE registry for all prescribers, patients, wholesalers and pharmacies.

**Morphea**

Morphea, also known as localized scleroderma, is a rare connective tissue disease characterized by sclerosis of the skin and subcutaneous tissues. Children comprise almost half of all cases, and girls are more often affected than boys. Lesions typically start with the insidious onset of firm, violaceous or erythematous plaques. The plaques gradually enlarge with active, violaceous or erythematous borders and shiny, white, firm centers. Atrophic, sclerotic and depressed scars are the eventual sequelae. Progression to systemic sclerosis is exceedingly rare. As morphea evolves very slowly, parents and care providers may not notice the disease initially.
There are multiple types of morphea, with the linear form most common in children. Plaque morphea manifests as single or multiple discrete lesions, primarily on the trunk and ranging in size from 1-20 cm. Linear morphea most often occurs longitudinally along an extremity. As linear morphea can cause deeper sclerosis of subcutaneous tissue, muscle and bone, erythema and other signs of active inflammation in the skin may not be obvious. Flexion contractures or scoliosis are potential complications. Variants of linear morphea can occur on the frontal scalp and forehead (en coup de sabre) or cause atrophy of half of the face (Parry-Romberg syndrome). (See Figure 1.) Both en coup de sabre and Parry-Romberg syndrome may be associated with abnormalities on MRI or CT and neurologic symptoms such as headaches and seizures. Other less common presentations of morphea include guttate morphea, generalized morphea, bullous morphea, disabling pansclerotic morphea and eosinophilic fasciitis.

Treatment for limited plaque morphea consists of topical corticosteroids, topical vitamin D analogues and topical tacrolimus or pimecrolimus. Extensive plaque morphea and most cases of linear morphea are treated with systemic agents. Generally, long-term methotrexate is administered with an initial course of oral prednisone or pulse intravenous methylprednisolone. Therapy is used to halt disease progression but does not reverse existing scarring, emphasizing the importance of early treatment.

**Conclusion**

Many skin conditions, both common and rare, may result in scarring. Acne vulgaris is commonly seen in adolescents but encompasses a wide spectrum of disease ranging from mild to severe. Early referral and treatment should be considered for any patient with scarring acne. Morphea is a rare and insidious connective tissue disease, and thus, it often is unrecognized. Timely diagnosis and therapy prevents functional complications and disfiguring scars.

For more information, visit chw.org/dermatology.
A patient’s story: An interdisciplinary approach to a child with dysphagia

by Joan Arvedson, PhD; Robert Chun, MD; Richard Noel, MD, PhD; Cecille Sulman, MD; and Neelesh Tipnis, MD

A 16-month-old, full-term child presented to an outside institution with a lifelong history of choking with feeds and frequent respiratory illnesses requiring medical attention beginning in early infancy. He was initially bottle fed and after multiple formula changes his dysphagia continued. Solids were attempted at 6 months of age without improvement.

He otherwise met all other developmental milestones in speech and fine motor skills, had a normal brain MRI, but still was failing to thrive. He had one episode of RSV at 15 months and no symptoms of upper or lower airway obstruction.

Previous diagnostic testing
An upper gastrointestinal (UGI) contrast study was concerning for cricopharyngeal achalasia. Esophagogastroduodenoscopy (EGD) showed no abnormalities except a narrow upper esophageal sphincter that did not allow expected passage of a 9 mm gastroscope.

A bronchoscopy showed normal vocal cord function and normal glottic and tracheal anatomy. A video fluoroscopic swallow study (VFSS) was performed to assess the upper aerodigestive anatomy and dynamic function. It demonstrated a posterior cricopharyngeal bar and aspiration on thin and thickened consistencies, typically after swallows, on residue in the pharynx due to inability to clear the bolus through the upper esophageal sphincter (UES).

Cricopharyngeal musculature was injected with botulinum toxin with a repeat UGI showing resolution of the cricopharyngeal bar and no aspiration, but within 10 weeks his dysphagia returned and he resumed his NPO status. Due to continued dysphagia, aspiration and failure to thrive, a gastrostomy tube was placed at 8 months.

The patient’s care was then transferred to Children’s Hospital of Wisconsin.
Dysphagia

Differential diagnosis of dysphagia etiology

- Pyriform aperture stenosis.
- Choanal atresia.
- Adenotonsillar hypertrophy.
- Cleft lip and palate.
- Macroglossia.
- Retrognathia/micrognathia.
- Vallecular cyst.
- Saccular cyst.
- Laryngomalacia.
- Laryngeal cleft.
- Laryngeal web.
- Subglottic stenosis.
- Tracheomalacia/tracheostenosis.
- Vascular ring.
- Tracheoesophageal fistula.
- Gastroesophageal reflux disease.
- Eosinophilic esophagitis.
- Cricopharyngeal achalasia.
- Oral pharyngeal dysphagia.
- Vocal cord immobility.
- Poor gastric motility.

Interdisciplinary approach to patient care

Gastroenterology

Upper endoscopy (EGD) was useful in ruling out eosinophilic esophagitis and any other inflammatory problem that can present with infantile feeding disorders and dysphagia. Contrast studies and EGD identified a narrow cricopharyngeal luminal diameter, which subsequently improved after injection with botulinum toxin. Recurrence of symptoms may simply indicate cessation of the botulinum toxin effect. While positive response to botulinum toxin suggests cricopharyngeal achalasia, prior to a definitive operative treatment, manometric studies can functionally confirm a diagnosis of cricopharyngeal achalasia.

High-resolution esophageal manometry was performed and noted elevated resting pressure, poor relaxation of the cricopharyngeal musculature and poor function of the muscle. Pharyngeal contraction pressure and esophageal body function was normal. These findings confirmed the diagnosis of cricopharyngeal achalasia. Previous brain MRI failed to identify posterior fossa abnormalities that can be associated with cricopharyngeal achalasia.

Despite the feeding disorder, this patient was well nourished and growing appropriately. The benefits of supplemental gastrostomy nutrition in this case cannot be understated, as it allows the medical and surgical focus to remain strictly on resolution of dysphagia without worries about malnutrition and its impact on development.

Pulmonary

Children with aerodigestive pathologies almost always have poor respiratory health. The maintenance of respiratory health is a challenge and often involves diagnosing the airway and lung disease. A combined assessment of the airway by both otolaryngology and pulmonology are complementary. Rigid bronchoscopies are most useful in identifying anatomic abnormalities such as laryngeal clefts and subglottic stenosis. Flexible bronchoscopy is a useful dynamic assessment of the lower airway, especially for lesions such as tracheobronchomalacia.
Management often includes focusing on airway clearance in addition to inhaled and oral anti-inflammatory medication. Due to our patient’s history of aspiration and RSV bronchiolitis, there were long-term concerns for his respiratory morbidity with continued feeding difficulty and aspiration. In light of his resolving bronchiolitis, inhaled steroids and chest physical therapy were recommended.

**Speech-language pathology**

When our patient presented at 16 months, VFSS revealed intermittent silent trace aspiration events with very thin (juice) and thickened (nectar) liquid. Aspiration was seen more frequently, but very small volume per bolus, after swallows of smooth puree (pudding) because of the reduced opening of the UES noted by cricopharyngeal (CP) bar with increased pressure at the UES, consistent with manometry findings. Specific goals to advance oral feeding were established in light of the global physiologic findings.

The follow-up VFSS was completed about 24 hours after the myotomy. A CP bar was not prominent. Boluses moved through the UES with no restriction. Trace aspiration on very thin liquid was noted one time just prior to initiation of a pharyngeal swallow due to a timing problem, not a CP deficit. A small “pouch” was noted at the posterior UES with a very small amount of barium contrast seen in the superior retropharyngeal space. Overall, this child showed remarkable improvement in swallowing just 24 hours following surgery.
Communication with parents a few weeks after discharge indicated that functional oral feeding gains were occurring with gradual increases in volume of oral feeding and expansion of textures. Pharyngeal pressures had been reported within normal limits via manometry preoperatively, which was an important finding to support a positive prognosis for oral feeding gains anticipated following surgery.

Otolaryngology
Given that the botox temporarily resolved the child’s symptoms and the desire for definitive treatment, surgical options included external versus endoscopic cricopharyngeal myotomy. The cricopharyngeus muscle forms the upper esophageal sphincter and under tonic contraction prevents food passage.

External approach risks include external incision, restenosis or scarring, salivary fistula, esophageal perforation, mediastinitis, vocal cord paralysis and hoarseness.

Endoscopic approach risks include restenosis or scarring, retropharyngeal space infection and mediastinitis, thermal injury, fistula formation and subcutaneous emphysema.

Treatment and outcomes
The patient underwent a coordinated EGD, bronchoscopy and endoscopic cricopharyngeal myotomy. A postoperative VFSS demonstrated resolution of the cricopharyngeal bar and minimal aspiration. There was a pharyngeal defect secondary to the surgery, and the patient developed no signs or symptoms of infection. The patient was discharged to home with continued oral feeding and swallow therapy.

Key points:
• Differential diagnosis for dysphagia is broad and diverse.
• Children with cricopharyngeal achalasia typically have multiple issues in their swallowing difficulties, not a simple UES dysfunction.
• Botox injection can provide temporary alleviation of symptoms.
• Surgical management may be considered for long-term management.
• With complex patients with dysphagia, an interdisciplinary approach is best to evaluate and manage swallowing, feeding and breathing disorders. Professionals make the best decisions when interdisciplinary team members communicate directly among themselves and with parents at every step of the diagnostic workup, decision-making and follow-up treatment.

The Airway, Digestive and Voice Center at Children’s Hospital of Wisconsin is an interdisciplinary clinic dedicated to providing comprehensive medical and surgical care for children with complex aerodigestive needs. Same-day consultations are arranged with the pulmonary, gastroenterology, otolaryngology and speech and swallowing departments. This team approach provides convenience for families, facilitates communication between specialties, provides a unified treatment plan, minimizes sedation and general anesthesia, and improves patient care and clinical outcomes.
GENETICS

Patients diagnosed with a genetic disorder or those challenged by ongoing illness may benefit from an appointment with a genetic counselor. An accurate diagnosis may change medical management and clarify recurrence or familial risk. Diagnostic testing has increased in availability and now may provide clearer answers. New treatments continue to become available. Support groups provide reassurance to the family. A molecular diagnosis can offer many additional reproductive options such as chorionic villus sampling (CVS), amniocentesis or preimplantation genetic diagnosis for families with concerns about additional pregnancies. For more information or to make an appointment, call (414) 266-3347 to speak with a genetic counselor. Visit chw.org/genetics for more information.

SCOLIOSIS

The orthopedic team at Children's Hospital of Wisconsin believes in a simple approach to treating scoliosis. Surgery only is for the most severe cases. Detecting scoliosis early is very important for successful treatment. Specialists use cutting-edge technology like the EOS low-dose radiation scanner and Quantec to help diagnose and treat scoliosis. Children's Hospital is one of two pediatric hospitals in the nation with an EOS scanner. The goal of treatment is to stop the curve from getting worse and prevent future problems. Visit chw.org/orthopedics for more information.

TRACHEAL DISORDERS

Children's Hospital of Wisconsin is one of a few hospitals in the nation with the expertise to care for children with complex tracheal disorders. Operations on the trachea are complicated and take a well-coordinated surgical team. Children's Hospital's team includes an ear, nose and throat surgeon experienced in the upper trachea, a cardiothoracic surgeon who specializes in the lower trachea and lungs, and a pediatric anesthesiologist who keeps the patient breathing while the trachea is being repaired. Visit chw.org/trachdisorders for more information.
INTERVENTIONAL RADIOLOGY AND IMAGE-GUIDED THERAPY

Interventional radiology is a subspecialty of imaging used to diagnose and treat pathology with the least invasive technique possible. While some of these procedures are done for purely diagnostic purposes, more and more therapeutic applications are being developed. Image-guided therapy treats conditions using imaging techniques, such as CT and ultrasound, to insert needles and catheters. The images provide a road map that allows the interventional radiologist to guide these instruments. Interventional radiology offers an alternative to the surgical treatment of many conditions and often can eliminate the need for hospitalization and shorten recovery times.

The interventional radiologists at Children’s Hospital of Wisconsin are among a select group of physicians, fewer than 100 nationwide, who specialize in pediatrics. They have additional years of fellowship training and continue to learn the most innovative and effective techniques. Performing more than 3,000 cases a year, our volume is in the top six for pediatric hospitals nationally.

Conditions treated
• Aneurysmal bone cyst.
• Arteriovenous malformations/fistula.
• Painful bone tumors.
• Cancer.
• Lymphatic malformation.
• Venous malformation.

Procedures performed
• Angiography.
• Angioplasty.
• Blood clot filters.
• Catheter insertions.
• Embolization.
• Foreign body extraction.
• Gastrostomy tube placement.
• Injection of clot-lysing agents.
• Laser therapy.
• Radiofrequency ablation.
• Sclerotherapy.

Savannah Dews, now 2 years old, got another chance at life when she received a new liver at age 6 months. A year after the transplant, she experienced life-threatening complications that usually require surgery or another transplant. Using guided 3-D imaging, a blockage was opened and blood flow was restored. She went home pain free after just one night.

To view a video on use of radiofrequency ablation to treat osteoid osteoma, visit chw.org/rfablation.
Pediatric image-guided therapy CME webcast
Practical Pediatrics Grand Rounds CME webcasts provide lectures on recent advances and current knowledge in pediatrics. Each lecture is available on demand, allowing you to earn CME credit when it is convenient for you.

“Update on Recent Advances in Pediatric Image-guided Therapy” by Craig Johnson, DO.

Objectives
• To understand uses of radiofrequency ablation in pediatric bone lesions.
• To understand advancements in image-guided treatment of vascular anomalies.
• To understand advancements in direct percutaneous image-guided management of aneurysmal bone cysts.

This lecture is available online at chw.org/practicalpediatrics.

See Page 21 for CME information.

REFER A PATIENT
To speak with an interventional radiologist, call the Imaging Department at (414) 266-3100 or Children’s Hospital of Wisconsin Physician Referral Center at (800) 266-0366.

CALL FOR AN APPOINTMENT
To make an appointment, call Central Scheduling at (414) 607-5280 or toll-free (877) 607-5280.
To survive in today’s competitive market, health care providers must make informed decisions on cost reduction and resource optimization while maintaining the highest quality of care. Related health care improvement projects often require a long-term commitment. The following project is one example of quality and safety endeavors under way at Children’s Hospital of Wisconsin.

In 2003, Children’s Hospital’s Pediatric General and Thoracic Surgery team began an appendicitis quality improvement project to standardize the protocol used in the hospital, and that work continues today.

Appendicitis is the most common surgical emergency in the pediatric population. However, until as recently as 2003, there were no uniform guidelines on how to treat these patients at Children’s Hospital. The general and thoracic surgeons at the hospital recognized that clinical pathways improve patient care by reducing variability, increasing efficiencies and decreasing costs. Clinical pathways were created for the postoperative treatment of appendicitis. Accurate and operative classification of the degree of disease was mandated for all surgeons (acute vs. ruptured), and they agreed to follow a clinical pathway depending on the degree of disease. A predetermined length of antibiotic therapy (single agent antibiotic therapy for acute and triple agent antibiotic therapy for ruptured) and advancement of diet and pain control measures were delineated for each patient based on disease degree. Once the protocols were established, outcomes such as length of stay and postoperative morbidity were measured. The team then compared outcomes to other comparable hospitals within the Pediatric Health Information System (PHIS)*, looking for areas of improvement.

Children’s Hospital’s protocolized approach compared favorably with other hospitals and published literature in terms of postoperative complications. However, the team found longer length of stay for patients with ruptured appendicitis. New literature showed that single
antibiotic therapy was safe in ruptured appendicitis and that improvement in clinical status (no fever, normal heart rate and return of intestinal function) is a good indicator that intravenous antibiotics and hospitalization may be discontinued. In 2008, the team approach to ruptured appendicitis was modified in accordance with recommendations based on a systematic review of the literature. The team postulated that changing from multiple antibiotics to a single antibiotic agent for ruptured appendicitis also would decrease length of stay and overall cost.

The team continued to monitor outcomes for this disease. The change to single antibiotic therapy decreased length of stay for patients with ruptured appendicitis, but led to higher postoperative infection rates. In 2010, the antibiotic therapy for acute and ruptured appendicitis was changed to another single agent antibiotic that has more gram-negative and anaerobic coverage. This change decreased the postoperative infection rates while maintaining the decreased length of stay and decreased antibiotic and nursing costs.

* PHIS is a detailed comparative database of freestanding pediatric hospitals that gives participating hospitals and clinicians the ability to assess and improve the resources required for patient care. The external PHIS comparisons are based on 11 hospitals with characteristics similar to Children’s Hospital of Wisconsin. PHIS data also are used to compare hospital performance against clinical guidelines for the purpose of identifying opportunities that can lead to quality improvement and development of clinical benchmarks among peers.
REGISTER NOW for the Best Practices in Pediatrics fall 2011 conference

Our Best Practices in Pediatrics conference will provide the latest information about common problems encountered in pediatric practice and will benefit all health care providers who work with children, including pediatricians, family practice physicians, nurse practitioners and physician assistants.

When: Friday, Sept. 23, and Saturday, Sept. 24
Location: Grand Geneva Resort, Lake Geneva, Wis.
Cost: Friday, $55; Saturday, $70 (*There is only a cost if you are claiming CME or hours of participation.*)

Registration information and conference details can be found online at chw.org/bestpractices.

Early registration is recommended.

**Friday, Sept. 23**
Registration: Noon – 12:50 p.m.
Lectures: 12:50 p.m. – 5:50 p.m.
• “The Intimate Relationship with Feeding, Swallowing and Breathing.”
• “Dysphagia: Oral Feeding Decisions with Aspiration Risks.”
• “Review of Outcomes from Airway, Digestive and Voice Center Patient Satisfaction Survey.”
• “Eosinophilic Esophagitis.”
• “Navigating Nutrition and Eosinophilic Esophagitis.”
• “Congenital Aerodigestive Disorders.”
• “Panel Case Presentation of Aerodigestive Disorders.”

**Saturday, Sept. 24**
Registration: 8 a.m. – 8:20 a.m.
Lectures: 8:20 a.m. – 3 p.m.
• “What’s New in Diabetes Care? Pens, Pumps, Sensors and More.”
• “Immunodeficiency Case Studies.”
• “The Limping Child: Solving the Mystery Sooner Versus Later.”
• “Binge Eating Among Adolescents.”
• “Pediatric Acute Lymphoblastic Leukemia: Past Success and Future Challenges.”
• “Neuroradiology for the Practicing Pediatrician: Current Perspective and Future Directions.”
• “Basics of Pulmonary Hypertension.”

Accreditation
This activity has been planned and implemented in accordance with the Essential Areas and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of The Medical College of Wisconsin and Children’s Hospital of Wisconsin. The Medical College of Wisconsin is accredited by the ACCME to provide continuing medical education for physicians.

Designation of credit
The Medical College of Wisconsin designates this Live activity for a maximum of 10 AMA PRA Category 1 Credits™. Physicians should only claim the credit commensurate with the extent of their participation in the activity.

Application for CME credit has been filed with the American Academy of Family Physicians. Determination of credit is pending.

Hours of participation for allied health professionals
The Medical College of Wisconsin designates this activity for up to 10 hours of participation for continuing education for allied health professionals.

Children’s Specialty Group fully intends to comply with the legal requirements of the Americans with Disabilities Act. If any participant is in need of accommodation, please call (414) 266-3456.
TUNE IN to Practical Pediatrics CME webcasts
Lectures on recent advances and current knowledge in pediatrics and pediatric specialties now are easy to access, interactive and available when you are. Each month a new lecture is added online.

To view on demand, visit chw.org/practicalpediatrics. You can view these presentations from your computer when it’s convenient for you and earn continuing medical education credits.

Current lectures available for CME credit
- “Pediatric Musculoskeletal Exam: Shoulder, Knee and Ankle” by Kevin Walter, MD, FAAP (January 2011)
- “Overview of Primary Immunodeficiencies” by John Routes, MD (February 2011)
- “The What and the Why of the WHO Growth Charts” by Praveen Goday, MD (March 2011)
- “Distraction Osteogenesis in Pediatric Patients” by Arlen Denny, MD (April 2011)
- “Genetic Screening: What Should I Offer My Patients?” by David Bick, MD (May 2011)
- “Updates on Recent Advancements in Pediatric Image Guided Therapy” by Craig Johnson, DO (June 2011)

The Medical College of Wisconsin is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians.

The Medical College of Wisconsin designates this enduring material for a maximum of 1.0 AMA PRA Category 1 Credit™. Physicians should only claim credit commensurate with the extent of their participation in the activity.

The Medical College of Wisconsin designates this activity for up to 1.0 credit hour of continuing education for allied health professionals.
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**Residency:** The Medical College of Wisconsin, Anesthesiology.

**Fellowship:** The Medical College of Wisconsin, Pediatric Anesthesiology.

**Board certification:** Anesthesiology.

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**Board certification:** Surgery, Pediatric Surgery and Thoracic Surgery.

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Board certification: Dermatology.

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Fellowship: Children’s Memorial Hospital, Chicago, Pediatric Neurosurgery.

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Bachelor of Science: Baylor University, Waco, Texas, Health Sciences.
Master of Science: University of Nebraska Medical Center, Omaha, Physician Assistant Studies.
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Research fellowship: University of Colorado School of Medicine, Sports Medicine (Family Medicine).
Board certification: Pediatrics.

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Master of Science: University of Wisconsin-Milwaukee, Nursing.
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**Fellowships:** Medical University of South Carolina, Forensic Pathology, and Cincinnati Children’s Hospital Medical Center, Pediatric Pathology.

**Board certifications:** Forensic Pathology and Anatomic Pathology and Clinical Pathology.

Maya C. Evans, MD, is a pediatric physical medicine and rehabilitation specialist at Children’s Hospital of Wisconsin, an assistant professor of Physical Medicine and Rehabilitation at The Medical College of Wisconsin and a member of Children’s Specialty Group.

**Medical degree:** University of Medicine and Dentistry of New Jersey, Newark.

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**Board certifications:** Physical Medicine and Rehabilitation.

Kimberley L. Wong, MD, is a pediatric physical medicine and rehabilitation specialist at Children’s Hospital of Wisconsin, an assistant professor of Physical Medicine and Rehabilitation at The Medical College of Wisconsin and a member of Children’s Specialty Group.

**Medical degree:** The Medical College of Wisconsin.

**Residency:** The Medical College of Wisconsin, Physical Medicine and Rehabilitation.

Amy J. Wagner, MD, is a pediatric surgeon at Children’s Hospital of Wisconsin, an assistant professor of Surgery at The Medical College of Wisconsin and a member of Children’s Specialty Group.

**Medical degree:** The Medical College of Wisconsin.

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**Fellowship:** The Medical College of Wisconsin, Pediatric General Surgery.

**Board certifications:** Surgery-General.

**PATHOLOGY**

**PHYSICAL MEDICINE AND REHABILITATION**

**SURGERY**

Departures

Children’s Hospital of Wisconsin and The Medical College of Wisconsin would like to thank the following individuals for their contributions and wishes them well in future endeavors:

- **Ramin Alemzadeh, MD**, Pediatric Endocrinology.
- **David J. Brown, MD**, Pediatric Otolaryngology.
- **Shewanna Brown, MSN, FNP-BC**, Pediatrics (Adolescent Medicine).
- **Heena Desai, MD**, Pediatric Psychiatry.
- **Samantha E. Hill, MD**, Pediatric Dermatology.
- **Alan N. Mayer, MD, PhD**, Pediatric Gastroenterology.
- **Stephanie M. Merrill, MD**, Pediatrics (Neonatology).
- **Aparna R. Rao, MD**, Pediatric Pulmonary and Sleep Medicine.
- **Rohit P. Rao, MD**, Pediatric Cardiology.
- **Jean Rex, MD**, Pediatric Hospital Medicine and Critical Care.
- **Colin D. Rudolph, MD, PhD**, Pediatric Gastroenterology.
- **Uzma Sharif, MD**, Pediatric Neurology.
- **Andrea Winthrop, MD**, Pediatric Surgery.
- **Mary L. Zupanc, MD**, Pediatric Neurology.
Pediatric Rounds is published by Children’s Hospital of Wisconsin and The Medical College of Wisconsin. Comments should be directed to: Children’s Hospital of Wisconsin, PO Box 1997, Milwaukee, WI 53201 or e-mail csg@chw.org.

One of the BEST

National hospital rankings
According to a data driven survey by Parents magazine, Children’s Hospital of Wisconsin is the No. 3 children’s hospital in the nation. Children’s Hospital of Wisconsin was ranked in all ten pediatric specialties in the 2011-12 edition of U.S. News & World Report America’s Best Children’s Hospitals list.

Nursing excellence
For the second time, Children’s Hospital of Wisconsin has been awarded the prestigious Magnet Recognition Award from the American Nurses Credentialing Center. Magnet designation is given to health care organizations that are able to demonstrate excellence in nursing.

ECMO Program designated a Center of Excellence
The Extracorporeal Life Support Organization has named the Extracorporeal Membrane Oxygenation Program at Children’s Hospital of Wisconsin a Designated Center of Excellence. Children’s Hospital first received this award for Excellence in Life Support in 2006. The hospital now is designated through September 2012.

Best Doctors in America
More than 40 percent of Children’s Specialty Group physicians are listed in the 2010-2011 Best Doctors in America® database. This group practice is jointly owned by Children’s Hospital and Health System and The Medical College of Wisconsin. The 40,000 U.S. physicians listed in the Best Doctors in America® database represent only about 3 to 5 percent of the nation’s practicing physicians. The listing is a singular honor. Best Doctors in America® is a registered trademark of Best Doctors, Inc., in the U.S. and other countries.

The information presented in this publication is intended for educational purposes only, providing suggestions for helpful interventions in primary care settings. Each patient is different and these suggestions may not apply. This is not a substitute for professional medical or mental health advice or services, particularly in acutely ill individuals who warrant immediate evaluation in an emergency setting. Consult appropriate specialty providers with specific questions regarding your patients.

Pediatric Rounds is published by Children’s Hospital of Wisconsin and The Medical College of Wisconsin. Comments should be directed to: Children’s Hospital of Wisconsin, PO Box 1997, Milwaukee, WI 53201 or e-mail csg@chw.org.

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