Infants and children have specific vulnerabilities to seizures. If there is an ongoing "epileptic storm" in the developing brain, epilepsy becomes more difficult to manage.

The Case for Pediatric Epilepsy Surgery

Innovation at the Herma Heart Institute

CME Events Coming in 2018

Specialty Spotlight: Neurosciences Center

New Doctors Around the State
Making Strides
The Herma Heart Institute is committed to caring for the hearts of children

BY THOMAS T. SATO, MD

In this issue, you will read about how our Herma Heart Center recently became the Herma Heart Institute. We feel this transformation speaks directly to our expansive vision for learning, teaching and caring for the hearts of children.

As one of the leading clinical care institutions in the country, we are fortunate to have internationally recognized investigators and clinicians who are focused on an audacious goal: preventing congenital heart disease (CHD) from negatively affecting children’s lives.

Two of our researchers, Michael E. Mitchell, MD, and Aoy Tomita-Mitchell, PhD, are doing cutting-edge work to identify genetic factors for CHD. While achieving that goal will take time, they are already learning some of the genetic and environmental factors that contribute to the disease.

Our heart team is working on every developmental stage. Through the Fetal Concerns Center, we are gaining a much better understanding of CHD in the fetus and how it can be corrected or mitigated in utero. Our leadership team includes internationally recognized experts, including our chief of pediatric cardiac surgery, Viktor Hraska, MD, PhD, and our chief of pediatric cardiology, Jeanne M. James, MD.

“As one of the leading clinical care institutions in the country, we are fortunate to have internationally recognized investigators and clinicians who are focused on an audacious goal.”

In addition, clinicians at the Herma Heart Institute understand that care for the heart doesn’t end with structural repair—they are looking at every child’s quality of life after surgery, with comprehensive evaluation of neurodevelopmental milestones and the impact of concurrent congenital abnormalities.

Audacious goals—but not impossible.

Best,

Thomas T. Sato, MD, FACS, FAAP
CEO, Children’s Specialty Group
Senior Associate Dean of Clinical Affairs,
Professor of Pediatric General and Thoracic Surgery, Medical College of Wisconsin
Pediatric general and thoracic surgeon, Children’s Hospital of Wisconsin

What do you think?
Let us know what you like about Pediatric Rounds, what we could do better and what you’d like to see more of in these pages. Receive a $10 Starbucks gift card for taking our brief survey at surveymonkey.com/r/PedRounds2018.
What’s in a Name?

The newly designated Herma Heart Institute continues a tradition of innovation

In the fall of 2017, Children’s Hospital of Wisconsin recognized the achievements of its world-renowned Herma Heart program by designating it the Herma Heart Institute, the first such clinical institute at the hospital. In order to receive the institute designation, service lines must meet nationally accepted benchmarks in clinical outcomes, patient care quality, education and outreach, philanthropy, and research and innovation.

INNOVATIVE BEGINNING

The pediatric cardiology program at Children’s was innovative from the beginning. In the 1980s, S. Bert Litwin, MD, the first cardiac surgeon at Children’s, was dogged in his pursuit of a successful procedure to repair hypoplastic left heart syndrome (HLHS), a congenital condition that essentially leaves a baby with half a working heart. At the time, only 5 percent of babies born with the condition survived beyond their first month of life. Dr. Litwin and his team pioneered procedures and monitoring protocols that eventually pushed survival rates over 95 percent.

One family whose infant daughter was born with HLHS was so impressed by Dr. Litwin and the cardiology team at Children’s that they gave their name to the program. In 2003, the Herma Heart Center was created through the support of John and Susan Herma. Their continued support allowed the recent transformation to the Herma Heart Institute.

RENOVATED PRESENT

The Herma Heart Institute is one of the largest pediatric heart programs in the country, performing an average of 650 heart procedures a year with some of the best outcomes in the world. But for the Herma Heart team, driving excellent clinical outcomes is not enough. In the early 2000s, they broadened their focus to improving quality of life post-discharge.

Doctors, caregivers and researchers at the Institute have led improvements in fetal and

Victor Hraska, MD, PhD, performs surgery to correct a rare and complex heart defect in a 9-month-old boy.
pediatric heart care by:
• Developing the nation’s first home monitoring program
• Creating the first neurodevelopmental follow-up program to boost a child’s growth and development after heart surgery
• Implementing a school intervention program that advocates for children and their families in the educational setting, helping our patients achieve their highest potential in school

• Pioneering innovations to improve outcomes and the patient experience, including a less invasive blood test to detect if a patient is rejecting a donor heart

Other hospitals across the country are adopting these programs, which have become the highest standard of care.

**THRILLING FUTURE**

The launch of the Herma Heart Institute begins a new era of enhanced and focused investment in innovation and research. Herma Heart Institute researchers are currently focusing on three areas of pediatric and fetal heart care that hold significant promise for lifesaving advancements:
• Preventing surgical bleeding and blood clots
• Optimizing quality of life following heart surgery
• Using genetics to improve long-term outcomes for patients with congenital heart disease

The ultimate goal is no less than eradicating congenital heart disease. This is a lofty goal, and it won’t be easy. But the Herma Heart Institute team has never backed down from a challenge.

**Learn more about the Herma Heart Institute at chw.org/heart.**

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**Adult Congenital Heart Disease Program by the numbers**

**ONE IN 100** babies born in the United States has a heart defect.

Children’s Hospital of Wisconsin’s Adult Congenital Heart Disease (ACHD) Program is now the first and only accredited ACHD Comprehensive Care Center in Wisconsin. This designation recognizes the quality of care that the program delivers for this growing yet often underserved patient population.

12 accredited ACHD centers exist in the United States.

11 Children’s clinic locations in Wisconsin have ACHD specialists.

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This surgery by Dr. Hraska was broadcast live in 3-D during the Heart Summit.

Watch highlights and learn about the upcoming Heart Summit at chw.org/theheartsummit.
A Better Partnership

When referring providers give us their feedback, we work to meet their needs.

“Referring providers have choices in terms of where to send their pediatric patients,” says Carey A. Ehler, MD, a neonatologist at Children’s Hospital of Wisconsin, director of provider experience for Children’s Specialty Group and associate professor of Neonatology at the Medical College of Wisconsin. To learn how referring providers make those choices—and how Children’s can be an even better partner to our referring providers—Dr. Ehler and a team of physicians and administrators surveyed almost 2,000 providers in 2017.

What we learned from the survey results was both heartening and instructive.

MAKING A DIFFERENCE

More than 90 percent of referring providers who responded to the survey have an excellent or very good overall impression of Children’s. “We have a very positive reputation out there, and we’re really proud of those results,” Dr. Ehler says. But at the same time, she knows there is room for improvement.

The team identified three key factors that drive referring providers’ impressions of Children’s:

1. Courtesy and respect shown to providers
2. Patient satisfaction with specialists
3. Returning patients for follow-up care

These are areas where we have already been working to make improvements. For example, Children’s specialists are trained on guiding principles of working with referring providers, including recognizing providers’ expertise on their patients and responding in a timely manner.

The survey results show we are doing a better job of meeting your needs. “We heard loud and clear that our referring providers have noticed a difference,” Dr. Ehler says. “They have noticed that we are treating them with courtesy and respect, and we are including them as valued members of our care team. These will continue to be the areas of focus, because we know they are what matter most to our referring providers.”

COMMUNICATION AND ACCESS

In addition to improving partnerships with referring providers, Children’s is taking on real-world challenges of communication and access.

“We’re really trying to improve our central scheduling process, and we’re doing a lot of work internally on the physician referral and consultation line,” Dr. Ehler says.

In addition, we launched the CHW Refer app, which gives providers access to appointment availability for new patients, medical care guidelines, CME offerings and a physician directory from the convenience of their smartphone.

All these efforts stem directly from referring providers’ feedback, and Dr. Ehler hopes providers can see that their opinions matter. “It is a privilege to be able to partner with referring providers,” she says. “And we appreciate that they choose to send their kids to us.”

Download the CHW Refer app to search for Children’s specialists, check appointment availability for new patients, review medical care guidelines and much more. Find the app at chw.org/medical-professionals/make-a-referral/referring-provider-app.
Upcoming CME Events

Don’t miss these educational opportunities for medical providers offered by Children’s.

Connect with Children’s

There are no fees to attend these CME dinners.

APRIL 10
AUTOMOBILE GALLERY, GREEN BAY

APRIL 24
PECK CENTER, MILWAUKEE COUNTY ZOO

MAY 22
OUTEREDGE STAGE, APPLETON

SEPT. 25
KENOSHA

OCT. 23
PECK CENTER, MILWAUKEE COUNTY ZOO

NOV. 14
OUTEREDGE STAGE, APPLETON

REGISTER:
chw.org/connect

Fifth Annual Pediatric Cancer Symposium

Cellular and Immunologic Therapies for Solid Tumors

MAY 10
MILLER PARK, SKYY LOUNGE, MILWAUKEE
chw.org/medical-professionals/education/cancer-symposium

The Heart Summit 2018

Controversies, Debates and Discussions in Left Ventricular Outflow Tract Reconstruction

OCTOBER 11
MILWAUKEE
chw.org/theheartsummit

CHW 2019 Best Practices in Pediatrics

FEB. 28–MARCH 1, 2019
GLACIER CANYON LODGE, WISCONSIN DELLS
chw.org/bestpractices

“We sincerely hope you will join us for this terrific interactive visual learning experience.”
— Victor Hraska, MD, PhD

The Heart Summit: Exchanging Ideas on Complex Conditions

The first-ever Heart Summit, in October 2017 at Children’s Hospital of Wisconsin, brought together medical experts from across the country to exchange ideas on a complex cardiac condition to help improve treatments and build medical consensus.

The highlight of the event was a live surgery, broadcast in 3-D on an 18-foot screen in an auditorium where participants could hear and interact with Victor Hraska, MD, PhD, medical director of cardiothoracic surgery at Children’s and surgical director of the Herma Heart Institute, as he performed an anatomical “double switch” correction of congenitally corrected transposition of the great arteries. (See page 4.)

“The essential idea behind the Heart Summit was to create a platform for the exchange of ideas and information,” Dr. Hraska says. “We wanted to take the classic medical conference model and make it more interactive and engaging.”

The idea was so successful that Dr. Hraska and his team are repeating the event with a new topic in 2018. Register for the Heart Summit at chw.org/theheartsummit.
The Case for Pediatric Epilepsy Surgery

Determining when surgery can provide relief from the burden of epilepsy

BY SEAN M. LEW, MD

Epilepsy, or recurrent, unprovoked seizures, affects 0.5 to 1 percent of the population. It most frequently occurs during childhood, and every year approximately 25,000 to 40,000 children in the United States experience their first seizure that is not associated with a febrile illness or head trauma. Thirty to 65 percent of these children go on to have a second unprovoked seizure within the first year.
Fortunately, in most patients the seizures can be controlled with antiepileptic drugs (AED), and in some cases the epilepsy will resolve over time. However, epilepsy in roughly 30 percent of children cannot be controlled medically. In addition, even in children with well-controlled epilepsy, the morbidity associated with long-term AED use can be significant. Epilepsy surgery offers an alternative means of treating seizures and is an important weapon in the armamentarium of the epileptologist.

EARLY INTERVENTION IS KEY
Infants and children have specific vulnerabilities to even brief, recurrent seizures. If there is an ongoing “epileptic storm” in the midst of a developing brain, the brain undergoes aberrant synaptogenesis and apoptosis, as well as changes in the development of inhibitory neurocircuitry. Therefore, immature brains quickly become “programmed” to excitability with continued seizures. This leads to broad networks of epileptogenicity that can become progressively more difficult to manage. Frequent seizures result in a secondary encephalopathy with subsequent global impairment.

The consequences of uncontrolled seizures on children’s brains are severe: developmental delays, behavioral difficulties and irreversible cognitive declines are the norm. But if the seizures and ongoing epileptic encephalopathy can be stopped, whether with medication or epilepsy surgery, windows of developmental opportunity can open and lead to significant developmental gains.

Fortunately, it does not take years to determine if a patient’s epilepsy will be amenable to medical treatment. Epilepsy in 60 percent of children will be controlled with the initial AED prescribed. Of the 40 percent whose seizures persist, only 10 percent will achieve good control with a second AED. The remaining 30 percent constitute the intractable group, and subsequent medication trials are unlikely to be effective.

THE BURDEN OF EPILEPSY
The consequences of epilepsy can be severe, even in children whose seizures are well controlled. These children have a higher incidence of anxiety and depression, perform at a lower level academically, and ultimately suffer from a discrepancy in employment and income when

The Newest Member of the Neurosurgery Team: A Robot

The Children’s neurosurgery team has gained a powerful tool to help kids with complex cases of epilepsy. When antiepileptic medications are not effective, neurosurgeons at Children’s may perform stereoelectroencephalography (SEEG), a procedure in which electrodes are implanted in the brain to record the electric activity involved during seizures. Using the new Renishaw Neuromate®, an image-guided neurosurgical robot, surgeons can perform SEEG more quickly and more accurately. This allows them to place a network of electrodes deep within the brain, creating a 3-D grid that is used to determine the origin and spreading pattern of seizures and identify the epileptogenic zone. This valuable information can be used to plan tailored surgical treatments.
compared with peers. Adults with epilepsy are less likely to marry.

Finally, side effects from AEDs can be profound. Many older AEDs can produce sedation. Phenytoin causes osteoporosis with long-term use; it is suspected but unproven that the enzyme-inducing AEDs also cause osteoporosis. Carbamazepine and felbamate can produce aplastic anemia. Valproate increases the risk of polycystic ovarian syndrome and anovulatory cycles in women. Many AEDs are teratogenic, and some animal studies have suggested that long-term AED use has oncogenic potential.

For select patients, surgery can provide relief from the burden of epilepsy.

SURGERY IS NOT A LAST RESORT
There is a tendency to view brain surgery as a high morbidity/mortality endeavor that should be reserved for the direst cases. While there are certainly risks associated with epilepsy surgery, they are often far outweighed by the risks of uncontrolled epilepsy, as well as the quality-of-life benefits associated with attaining freedom from seizures or improved seizure control. Advances in diagnostic and surgical techniques have improved success rates and decreased complication rates.

Unfortunately, patients are often unsuccessfully managed for several years before being referred to a tertiary epilepsy center for surgical evaluation. Over that time, patients often suffer irreversible cognitive losses, while the epilepsy becomes more difficult to manage/cure due to the spread of epileptogenic networks. It is likely that many incurable adult epilepsy patients would have been more easily treated surgically at an earlier age with greater preservation of cognition.

The developing brain of a child is also much more plastic than that of an adult, so it’s possible for eloquent functions such as motor control and language to re-establish in other areas of a child’s brain—a feat not possible in a mature brain. Thus, it is important that medical providers and families consider surgical options early in the course of treatment, rather than as a last resort.

DETERMINANTS OF SURGICAL CANDIDACY
The decision of whether a child is a surgical candidate is typically made by a multidisciplinary team at a tertiary epilepsy center. Evaluations require expertise from a variety of fields, including neurology, neurosurgery, neuropsychology and radiology.

Questions we consider when determining surgical candidacy:

1. Is there a structural lesion that appears to be generating the seizures?
   Seizures associated with structural lesions, such as tumors, focal cortical dysplasias, vascular malformations, hamartomas, infarcts and mesial temporal sclerosis, are more often amenable to surgical treatments. In the majority of lesional cases, we can obtain complete seizure freedom through surgery.

2. What is the prognosis for continued nonsurgical therapy?
   If a patient has failed more than two AEDs, it is likely he/she will remain medically intractable. Some epilepsy
syndromes, such as benign rolandic epilepsy, are typically self-limited and resolve over time. Other syndromes have a predictable, progressive course leading to medically intractable seizures that can only be cured with surgery. If one can identify the specific epilepsy syndrome or etiology for the epilepsy, the prognosis can typically be defined. Rasmussen’s syndrome, epilepsy associated with mesial temporal sclerosis and large cortical malformations, such as hemimegalencephaly, are almost always associated with intractable seizures and a progressive course. These epilepsy syndromes should be evaluated for epilepsy surgery quite early.

3. Is it likely that an epileptogenic zone can be identified and safely treated? Much of the preoperative assessment is geared toward determining the likelihood that a focal epileptogenic zone can be resected or disconnected without generating unacceptable neurologic deficits.

If the epileptogenic zone is suspected but not certain, the patient may require intracranial EEG monitoring via implanted electrodes to better define the source of the seizures and define eloquent cortex, areas of the brain where important motor or language functions reside. Some patients have seizures without any evidence of such localization. They are typically less optimal candidates for epilepsy surgery. Others have epileptogenic foci that include eloquent areas of the brain, making surgical resection or disconnection a less palatable option.

4. Is there a role for palliative surgery? Some children have incurable epilepsy but still can benefit from surgery to decrease the frequency or severity of their seizures. For example, some patients with intractable generalized epilepsy gain substantial benefit from sectioning of the corpus callosum to eliminate drop attacks (tonic/atonic seizures), though they will continue to have other types of less-severe seizures. Other patients may have bihemispheric/multifocal epileptogenic zones. If one area is producing the predominant intractable seizures, resection of this area...
may provide a substantial reduction in seizure frequency and result in a significant improvement in quality of life.

**SURGICAL TECHNIQUES**

A variety of surgical techniques are used to treat pediatric epilepsy. The appropriate surgery is determined by the extensive presurgical evaluation. The initial phase of surgery is often diagnostic—electrodes are temporarily implanted intracranially to better localize the epileptogenic zone and to map areas of eloquent brain to be preserved.

Recently, we have adopted a minimally invasive technique of monitoring intracranially called stereoelectroencephalography (SEEG) (Figure 1). This involves placing electrodes via tiny individual incisions and holes in the skull via robot-assisted stereotactic guidance. (See “The Newest Member of the Neurosurgery Team: A Robot” on page 8.) By avoiding a large incision and craniotomy, there is decreased risk of infection and less postoperative discomfort. Once the area of seizure onset and eloquent brain are localized, attention is turned to therapeutic techniques to eliminate the seizures.

There are three broad categories of therapeutic surgical techniques:

1. **Resection.** Surgical resection of epileptogenic tissue is the mainstay of epilepsy surgery. Resections can be limited, as in the case of small lesions (lesionectomies) or areas of epileptogenic brain (topectomies). They can also include complete or partial resections of lobes of the brain (lobectomies). At the extreme end of the spectrum, the entire cerebral hemisphere can be removed (anatomic hemispherectomy) or disconnected (functional hemispherectomy) in the case of broad areas of pathology, such as Rasmussen’s syndrome, Sturge-Weber syndrome or perinatal infarctions.

2. **Disconnection.** If an epileptogenic zone is disconnected from the rest of the brain, the neurological and neurosurgical conditions, including highly sophisticated imaging capabilities and minimally invasive surgical options.

Our specialists treat the full range of neurological conditions from birth through childhood, and we have nationally recognized expertise in epilepsy.

The Epilepsy Center at Children’s is designated a Level 4 epilepsy center by the National Association of Epilepsy Centers. This is the only Level 4 pediatric epilepsy center in the state of Wisconsin. This designation means we provide the highest level of complex and specialized care for children living with epilepsy.

Learn more about the Neurosciences Center at Children’s and find hospital and clinic locations at chw.org/neurosciences.
Deep brain stimulation and cortical stimulation are techniques that show promise in adult human trials for managing some forms of epilepsy. Pediatric trials are forthcoming.

4. Ablation. A newer epilepsy surgery technique is stereotactic laser ablation (SLA). This minimally invasive technology involves placing a very small laser applicator into the targeted brain tissue or tumor. The laser is then activated with the patient in an MRI scanner to monitor the energy delivery to the targeted tissue. This allows for precise destruction of deep targets in the brain without the morbidity associated with open brain surgery (Figure 2). Only smaller targets can be treated with this technology, so it is not appropriate for most patients. Those patients who are candidates typically enjoy shortened recovery times. Most patients go home the day after the procedure with minimal postoperative discomfort. Children’s Hospital of Wisconsin is one of the few hospitals in the world where SLA is performed.

**SUMMARY**

- The immature, developing brain is particularly susceptible to the ravages of poorly controlled seizures, leading to permanent developmental/cognitive losses and the development of broader epileptogenic networks.
- It is imperative that children with epilepsy achieve maximal control in a timely fashion to prevent these long-term sequelae.
- A significant subset of children with epilepsy can benefit from epilepsy surgery as a curative measure or for palliation.
- The earlier these patients are identified and treated, the better the neurological/developmental outcome.
- The decision-making process for determining surgical candidacy and the appropriate surgical approach is complex and requires a multidisciplinary team at a comprehensive epilepsy program.

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**FIGURE 2.** Coronal MRI demonstrating stereotactic laser ablation of a hypothalamic hamartoma in a child with gelastic seizures.
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Douglas Zabrowski, MD, is a pediatric gastroenterologist at Children's Hospital of Wisconsin and an assistant professor of pediatric gastroenterology at the Medical College of Wisconsin.

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NEW ON STAFF
Specialists in our network ready to help
To refer a patient, call (800) 266-0366.

KEY TO SYMBOLS: DEGREE RESIDENCY FELLOWSHIP BOARD CERTIFICATION

Departures
Children's Hospital of Wisconsin would like to thank the following providers for their contributions. We wish them well in future endeavors.

Neil Connor, MD, Critical Care
Christopher Inglese, MD, Neurology
Carl Weigle, MD, Critical Care
Tracey Liljestrom, MD, is a pediatric hospitalist at Children’s Hospital of Wisconsin and an assistant professor of pediatric hospital medicine at the Medical College of Wisconsin. University of Virginia School of Medicine, MD Duke University, Internal Medicine and Pediatrics Internal Medicine, Pediatrics

Erin Preloger, MD, is a pediatric hospitalist at Children’s Hospital of Wisconsin and an assistant professor of pediatric hospital medicine at the Medical College of Wisconsin. Medical College of Wisconsin, MD University of Colorado/Children’s Hospital of Colorado, Pediatrics Pediatrics

Sachin Kumbhar, MD, is a pediatric radiologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric radiology at the Medical College of Wisconsin. Seth Gordhandas Sunderdas Medical College, Mumbai, India, MD Seth Gordhandas Sunderdas Medical College, Mumbai, India, Radiology; University of Washington School of Medicine, Nuclear Medicine University of Washington School of Medicine, Abdominal Radiology, Body Imaging, Pediatric Radiology

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Raquel Farias-Moeller, MD, is a pediatric neurologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric neurology at the Medical College of Wisconsin. Escuela de Medicina del Tecnologico de Monterrey, Mexico, MD Medstar Georgetown University Hospital, Pediatrics Seth Gordhandas Sunderdas Medical College, Mumbai, India, Radiology; University of Washington School of Medicine, Nuclear Medicine University of Colorado School of Medicine, Pediatric Infectious Diseases Pediatrics, Pediatric Infectious Diseases Pediatrics, Psychiatry, Neurology, Epilepsy

KEY TO SYMBOLS: Degree Residency Fellowship Board Certification
Alexandria Lutley, MD, is a pediatric neurologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric neurology at the Medical College of Wisconsin. 

Irene Kim, MD, is a pediatric neurosurgeon at Children’s Hospital of Wisconsin and an assistant professor of pediatric neurosurgery at the Medical College of Wisconsin. 

Joseph Amaral, PhD, is a pediatric neuropsychologist at Children’s Hospital of Wisconsin and an assistant professor of neuropsychology at the Medical College of Wisconsin. 

Alicia Lohwing, MD, is a pediatric orthopedist at Children’s Hospital of Wisconsin and an assistant professor of pediatric orthopedics at the Medical College of Wisconsin. 

Elizabeth Roth, MD, is a pediatric urologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric urology at the Medical College of Wisconsin. 

Pat Radoszewski, RN, MBA, retired this year after a distinguished career. Pat joined the Medical College of Wisconsin in February 2001 and went on to become chief operating officer for Children’s Specialty Group. “Pat is an institution unto herself and has been instrumental in the success of our practice,” says Thomas T. Sato, MD, CEO of Children’s Specialty Group. “Her grace under pressure, willingness to always push for improvement, ability to see the bigger picture and constant advocacy for our providers are just a few of the many talents she has brought to our practice.” Pat will be missed, and we thank her for her years of service.
The locations above are Children's Clinics. We also see patients in other clinics in the following cities: Fond du Lac, Green Bay, Oshkosh, Racine and Iron Mountain, MI. We also perform surgeries in Marshfield.