Mayo Clinic is pioneering advances in fetal surgery, allowing for earlier intervention to manage congenital anomalies. Although rare, these anomalies — including pulmonary hypoplasia in congenital diaphragmatic hernia (CDH) and fetal lower urinary tract obstruction (LUTO) — carry significant risk of mortality and disability.

“Typically, these problems have been treated after birth. But we are committed to developing surgical methods to reverse these conditions in utero and improve fetal development,” says Rodrigo Ruano, M.D., Ph.D., chair of Maternal and Fetal Medicine at Mayo Clinic in Rochester, Minnesota.

Fetal surgery requires a multidisciplinary team with specialists in maternal-fetal medicine, pediatric surgery, neonatology, radiology and anesthesiology, as well as pediatric otolaryngology, nephrology, urology and neurosurgery, as needed. As a major tertiary medical center, Mayo Clinic Children’s Center has the expertise to manage these complex patients, including providing both maternal and pediatric anesthesia. “We also work to establish collaborative relationships, so we can share care with the patient’s home physician,” Dr. Ruano says.

**Novel therapy for CDH**

Mayo Clinic has demonstrated the feasibility and safety of fetal endoscopic tracheal occlusion for severe CDH resulting in pulmonary hypoplasia, a life-threatening anomaly. The novel procedure (Figure) is performed percutaneously with ultrasound-guided uterine access and fetoscopic deployment of a detachable tracheal balloon at 26 to 30 weeks of gestation. The balloon is intended to remain in place until about 34 weeks of gestation, when it is removed, preferentially through a fetoscopic procedure. The treatment optimizes parenchymal growth, improving pulmonary hypoplasia and vascular remodeling.

In the June 2018 edition of Mayo Clinic Proceedings, a specialized multidisciplinary group including Dr. Ruano described significant improvement after the procedure in two recent cases: Lung growth essentially doubled in one fetus and increased by one-third in the other. After birth, when the infants left the hospital, the first was breathing normally and the second required minimal nasal cannula oxygen support.

“Our experience indicates that fetuses with severe CDH have higher survival rates when treated prenatally, and Mayo Clinic has established a successful multidisciplinary fetoscopic tracheal occlusion program,” Dr. Ruano says. “Although we have seen the beneficial effects of the tracheal balloon, we don’t yet understand the mechanism for lung regrowth as part of in utero regenerative medicine. Our research on the regenerative principles of the lungs continues.”

**Prenatal management of LUTO**

Mayo Clinic Children’s Center is one of the few centers with a multidisciplinary program to treat LUTO prenatally. Depending on the nature of the blockage, treatment might consist of cystoscopy,
The brachial plexus injuries experienced by newborns can range from a few stretched nerves to the tearing or rupturing of all nerves in the brachial plexus network. A multidisciplinary approach is beneficial to achieving optimal outcomes.

The brachial plexus clinic at Mayo Clinic, which manages care for children and adults, is one of the highest-volume brachial plexus clinics in the country. The treatment team includes two orthopedic hand and microsurgeons, as well as a pediatric orthopedic surgeon and a neurosurgeon specializing in peripheral nerve surgery. Subspecialized radiologists and anesthesiologists with experience in diagnostic and surgical procedures for these young patients also are available.

Appointments for patients with these injuries are generally made within a week or two of referral. Ideally, the required nerve surgery should be performed early, to avoid muscle degeneration due to the lack of nerve signals.

“We know that if you wait too long for that initial surgery — just observing for the baby’s first year — you can lose the ability to do primary nerve surgery. Typically, the nerves need to be operated on in the first six months of life,” says Alexander Y. Shin, M.D., an orthopedic hand and microsurgeon specializing in brachial plexus injuries at Mayo Clinic Children’s Center in Rochester, Minnesota.

The benefits of an integrated approach can be seen throughout the process of managing neonatal brachial plexus injuries. Diagnosis generally involves MRI and nerve tests. At Mayo Clinic, pediatric neurologists, radiologists and anesthesiologists coordinate to perform testing in one setting.

“Otherwise, a baby might end up having two or three major anesthesia procedures. We try to minimize anesthesia to one when possible,” Dr. Shin says.

Having a multidisciplinary team allows for surgical reconstruction that is tailored to the individual patient’s needs — nerve surgery, tendon transfers and shoulder-to-hand reconstruction — when necessary. Dr. Shin notes that brachial plexus

**Neonatal Brachial Plexus Injuries: An Integrated Approach**

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**Figure.** Illustration shows what occurs during neonatal brachial plexus injury surgery. A. The brachial plexus is found in the neck above the clavicle. B. The damaged segment and the level of injury are identified in surgery. C and D. The damaged segment is removed, then reconstructed by taking nerve graft segments from the leg and cabling them to match the size of the damaged nerve. E and F: The cabled nerve graft is sewn in place using microsurgery, allowing the nerve to regenerate to restore function to the paralyzed muscles.
nerve surgery (Figure) can restore 70 to 80 percent of shoulder and elbow function in certain types of injury. However, imbalances in growth from nerve injury around the shoulder can lead to secondary sequelae: typically, internal retraction contracture and the inability to externally rotate the shoulder.

“If those sequelae aren’t addressed before about 2 years of age, the child might experience long-term problems, especially with the shoulder,” Dr. Shin says. About 70 to 80 percent of patients who need secondary surgery have good outcomes.

“A baby having a brachial plexus injury can understandably be an emotional experience for parents,” Dr. Shin says. “Our brachial plexus team works together so that parents and baby can move forward with the best possible care.”

**Bronchoscopy: Highly Specialized Diagnosis and Care**

Mayo Clinic Children’s Center is one of the few centers in the United States where pediatric interventional pulmonology is part of daily practice. Mayo’s pediatric pulmonologists can provide expert diagnostic and interventional bronchoscopy spanning infancy into adulthood for patients with breathing problems ranging from common to extremely challenging (Figure). Collaboration with other Children’s Center specialists typically results in a treatment plan within days, even for children with complex conditions.

“In most centers, interventional bronchoscopy procedures are performed by adult specialists. However, as pediatric pulmonologists, we have enhanced understanding of the relevant indications for bronchoscopy in the pediatric population, and as part of a pediatric care team, we have a natural working relationship with other specialists,” says R. Paul Boesch, D.O., chair of Pediatric Pulmonology at Mayo Clinic Children’s Center in Rochester, Minnesota. “A solid, trusting relationship between the pulmonologist and anesthesiologist is essential for the safety and effectiveness of these procedures.”

Mayo Clinic Children’s Center performs between 400 and 500 pediatric bronchoscopies a year, for conditions including:

- Persistent wheezing that doesn’t respond to asthma medications
- Chronic cough and recurrent infections
- Aerodigestive problems
- Anatomic anomalies that might require surgery
- Severe scoliosis that affects breathing
- Recurrent chest infections
- Feeding and swallowing difficulties
- Airway masses
- Obstructive sleep apnea

“We find that a very high percentage of children with persistent or atypical wheezing or poorly controlled asthma have either infectious problems that can be treated or anatomic problems that exacerbate the wheezing,” Dr. Boesch says. “Sometimes anatomic problems can be addressed with surgery. If not, we might be able to scale back some of the asthma therapies that may not be having an effect — which is beneficial in terms of avoiding cost and side effects.”

Patients with complex airway problems often have both flexible and rigid bronchoscopy, performed in a single procedure by a pediatric pulmonologist and a pediatric otolaryngologist, respectively. “The two techniques are highly complementary and together provide the most complete evaluation of the airway. Both providers see the airway together but bring different perspectives. This provides a seamless team approach to airway reconstruction,” Dr. Boesch says.

Any additional testing, such as gastrointestinal endoscopy, can be done in the same procedure, avoiding multiple rounds of anesthesia, radiation exposure and clinic visits. “We prepare an itinerary for patients in which all tests and evaluations are completed within days. Our specialists then agree on a unified care plan,” Dr. Boesch says.

For children with early-onset, congenital or neuromuscular scoliosis, pediatric pulmonologists.
Identification and treatment of hearing impairment should occur as early as possible, including cochlear implantation for children as young as 6 months who have moderate to profound hearing loss. "Increasing amounts of data show that the earlier you implant, the better the speech and language outcomes are, in general. We usually implant both ears, and we’re implanting people with more residual hearing," says Colin L. Driscoll, M.D., an otolaryngologist at Mayo Clinic Children’s Center in Rochester, Minnesota.

A cochlear implant delivers sound signals directly to the auditory nerve, bypassing damaged portions of the ear (Figure). The sound signals are captured by a processor fitted behind the ear and transmitted to a receiver implanted under the skin behind the ear. The receiver sends the signals to electrodes implanted in the cochlea, which stimulate the auditory nerve.

Even in children who have some residual hearing, cochlear implantation can significantly improve the quality of sound, optimizing language development. “Although hearing aids may provide some sound for people with residual hearing, the sound quality isn’t nearly as good as what we can give with a cochlear implant,” Dr. Driscoll says. “If children aren’t meeting their language milestones with amplification, cochlear implantation should be strongly considered earlier rather than later. It’s an established technique with a very good track record.”

“Integrating pediatric pulmonologists into surgical teams is very beneficial and common here at Mayo,” Dr. Boesch says. “We can often identify new findings and perform novel interventions based on our wide experience with interventional bronchoscopy and a team approach to care.”

**Cochlear Implants: Early Intervention to Optimize Language Outcomes**

Early management requires early diagnosis. Yet despite increased screening, Mayo Clinic continues to see children with hearing loss that wasn’t diagnosed or treated early. Due to the critical time window for auditory language development, delays can have a lifelong impact. “If there’s any concern about a child’s hearing or speech and language development, the child should have a hearing test,” Dr. Driscoll says. “Recognizing hearing loss and referring the child early is very important.”

**Managing complex conditions**

Mayo Clinic Children’s Center has successfully performed cochlear implantation for children with a range of hearing-loss etiologies. Mayo Clinic was one of the first centers to use cochlear implants to treat children with auditory neuropathy spectrum disorder, which causes about 10 percent of cases of congenital sensorineural hearing loss. “Some kids with auditory neuropathy have a lot of hearing, but the sound quality is terrible. With cochlear implants we can often restore clarity,” Dr. Driscoll says.

Mayo Clinic Children’s Center also has experience treating children with progressive hearing loss due to conditions such as an inner ear malformation or cytomegalovirus (CMV). “We have developed new protocols for monitoring children with CMV,” Dr. Driscoll says.

About half of severe congenital hearing loss has a genetic cause. At Mayo Clinic Children’s Center, a pediatric genetist helps manage care for these children. Identifying a genetic cause for hearing impairment can spur investigation for associated vision and renal problems. “There’s great value in understanding why a child has severe hearing loss. Knowing the origin allows us to not worry about other potential causes,” Dr. Driscoll says.

For all children who undergo cochlear implantation, Mayo Clinic Children’s Center provides subspecialized care from clinical genomicists, pediatric neurologists, ophthal-
mologists and rheumatologists as needed. “We have a seamless system that allows a team of experts to communicate with one another and work together to really understand children and all their health problems,” Dr. Driscoll says. “That’s what allows us to provide optimal care for children with complex conditions.”

Avoiding Recurrence of Patellofemoral Instability

Understanding and treating all aspects of patellofemoral instability are key to preventing recurrence of instability and pain. “Patellofemoral instability is a multifactorial problem. Thorough diagnosis allows us to address all the anatomic issues and achieve an instability rate of less than 20 percent,” says Todd A. Milbrandt, M.D., a pediatric orthopedic surgeon at Mayo Clinic in Rochester, Minnesota.

Early intervention is important for optimal outcomes. “Patellofemoral instability can occur even in young kids, and we don’t want to wait until they’re skeletally mature to treat it,” Dr. Milbrandt says. “We have techniques to modify children’s growth. For severe knock-knee, we can straighten the leg with a small plate and then do the patella reconstruction. If we wait until the child finishes growing, the patella and the trochlea deform and the reconstruction is much harder.”

A quiver of treatment options

Most of the children seen at Mayo Clinic Children’s Center for patellofemoral instability have sports-related injuries. Dr. Milbrandt cites one patient who was unable to climb stairs without dislocating her kneecap.

“We go from that extreme to kids who tend to dislocate when they play a certain sport. Those kids could potentially just change sports — from playing soccer to swimming, for example — but they don’t want to do that,” Dr. Milbrandt says. “For a lot of young people, their sport is their peer group and their definition of themselves.”

To define all the components of an individual’s patellofemoral instability, a child undergoes:

- A thorough physical evaluation, including rotation of the tibia and femur
- X-rays from the ankle to the hip, to evaluate standing alignment and knee position
- CT scans or MRI to determine where the patellar tendon attaches to the tibia and to establish the tibial tuberosity-trochlear groove distance

Dr. Milbrandt notes that Mayo Clinic Children’s Center performs these tests, so testing before referral isn’t necessary.

A surgical plan is then developed. “We have a quiver of treatment options, all the way from minor tightening to very complicated osteotomies around the tibia and fibula,” Dr. Milbrandt says.

To reduce the risk of patella breakage, surgeons avoid drilling holes in the kneecap for ligament fixation (Figure). “We use metal suture anchors or screws in the kneecap, and we tension that on the femoral side. We tie the ligaments there so that they stay in place,” Dr. Milbrandt says.

A complete physical therapy protocol is provided after surgery. “Once strength in the vastus medialis oblique is apparent, we do a series of functional tests — assessing the knee’s movement when the patient hops and jumps down from a block. It’s important to see how that kneecap functions,” Dr. Milbrandt says.

To continually improve their technique, orthopedic surgery residents study and write up every patellofemoral instability case experience. Mayo Clinic’s team approach also allows pediatric orthopedic specialists to consult their colleagues in adult practice. “We discuss our difficult cases, and team up as co-surgeons when needed,” Dr. Milbrandt says. “We learn from one another and from our patient database, and that’s a huge advantage.”

Figure. On the left, MRI of a 16-year-old boy with patellofemoral instability shows lateral displacement of the patella. On the right, a postoperative image shows the patellar suture anchors and femoral fixation.
The Complexities of Pediatric Motility Disorders

Pediatric gastrointestinal motility disorders are common and sometimes seen as benign. Yet often these disorders have complex causes, and patients can benefit from specialized pediatric gastrointestinal management.

“Even a disorder as common as constipation is not as straightforward as some people think. Different types need to be managed differently,” says Mhd Louai Manini, M.D., a pediatric gastroenterologist at Mayo Clinic in Rochester, Minnesota. Children with constipation might have congenitally slow colons, anorectal malformations or pelvic floor problems.

Referral to a pediatric gastroenterologist may be beneficial for children with constipation and other motility problems, such as:

- Dysphagia
- Abdominal distension
- Poor feeding tolerance
- Chronic nausea and vomiting
- Acid reflux

“Although constipation is the most common issue, we see kids whose motility problems are complicated by underlying conditions, such as spina bifida, spinal cord lesions, tumors or Hirschsprung’s disease — a rare, congenital condition involving nerve cells in the colon that make it difficult to pass stool,” Dr. Manini says.

The available testing includes:

- Gastric small bowel and colonic transit studies
- Anorectal manometry
- Colonic manometry
- Esophageal manometry
- Gastroduodenal manometry

Esophageal manometry can detect ruminating syndrome, in which people repeatedly and unintentionally regurgitate food, then rechew and swallow it. “Rumination is an underdiagnosed condition, because the tools to diagnose and manage it aren’t common,” Dr. Manini says. Children with ruminating syndrome might be referred to a Children’s Center behavioral psychologist, who can teach diaphragmatic breathing techniques that are effective for most people.

Gastroduodenal manometry is used to assess the strength of stomach contractions and their coordination with the small bowel. “This testing can be challenging to do in kids,” Dr. Manini says. “But our team has expertise in caring for children and easing their concerns.”

Once testing is completed, the treatment plan might involve medication, surgery or behavior therapy. Heller myotomy may be performed in children with achalasia. Children with Hirschsprung’s disease generally require initial anorectal surgery and ongoing monitoring for future complications, such as strictures and incontinence.

For children with pelvic floor problems, physical therapists can provide pelvic floor therapy. Biofeedback might be used to teach children to relax and tighten pelvic floor muscles.

“Having these newer tools to evaluate children is a great benefit to guiding their management,” Dr. Manini says. “Testing and treating children are often not as straightforward as managing care for adults. But we have the expertise and experience to offer complete management for pediatric patients.”

Enhanced Pain Management for Pectus Excavatum Surgery

Mayo Clinic Children’s Center uses a novel system to manage pain after pectus excavatum surgery, typically leading to decreased use of pain medications, improved pain scores and shorter hospital stays. The system, developed by Mayo Clinic pediatric surgeons and anesthesiologists, involves the insertion of paravertebral catheters that pump a local anesthetic to the intercostal nerves for about a week after surgery (Figure).

“Since we started a multimodal pain management strategy in 2010, the use of opioids has gone down by more than half while the pain scores dropped significantly, which means children are having less pain. With this strategy, children are getting out of bed sooner and drinking and eating much earlier than before,” says Dawit T. Haile, M.D., chair of Pediatric Anesthesiology at Mayo Clinic in Rochester, Minnesota. “The major change to the pain management strategy was the introduction of paravertebral catheters. We use ultrasound to insert nerve catheters to block nerves innervating the chest wall while the patient is asleep, avoiding the pain and anxiety of placing the catheters while the patient is awake, such as when an epidural is placed.”

“The enhanced pain management has helped reduce the average length of hospital stay for the procedure from five days to about three,” says D. Dean Potter Jr., M.D., chair of Pediatric Surgery at Mayo Clinic’s campus in Minnesota. “Pain medication can be infused in the hospital and for two to three days after patients leave. The catheters..."
are removed at home. It’s as easy as removing a dressing,” Dr. Potter says.

**Optimizing outcomes**

Ideally, children with pectus excavatum should have an initial evaluation around the age of 8 to 10. Signs and symptoms may include the appearance of the pectus, shortness of breath and exercise intolerance. If the pectus is severe, signs and symptoms might include rapid heartbeat or palpitations, wheezing, and chest pain. Mayo Clinic Children’s Center can monitor the child for heart and lung complications before performing surgery if needed.

“The best time to do the surgery is ages 12 to 14,” Dr. Potter says. “Younger children generally don’t yet have the maturity to go through this procedure. By the time children turn 16 or 17, their chest walls are a little more rigid. While that’s not necessarily a problem for successful surgery, it may make the procedure more painful.”

To minimize radiation exposure, Mayo Clinic Children’s Center uses X-rays rather than CT scans to obtain the chest-cage measurements needed to plan surgery. Pulmonary function tests are done to exclude asthma or other pulmonary conditions as a cause of symptoms. Echocardiography may be performed to assess heart function. Additional monitoring and care are provided for children whose pectus excavatum is associated with a connective tissue disorder, such as Marfan syndrome or Ehlers-Danlos syndrome.

The minimally invasive surgery for pectus excavatum, which involves inserting a customized bar in the chest, takes about one to two hours. Patients generally can resume vigorous exercise three months after surgery and contact sports six months after surgery.

Hundreds of minimally invasive pectus surgeries have been done at Mayo Clinic Children’s Center since it became one of the first centers in the United States to offer the procedure. Dr. Potter notes that patients and their parents consistently report high satisfaction with the outcomes.

“We see kids who couldn’t run a mile and all of a sudden become runners after recovering from surgery,” Dr. Potter says. “Other children who were a bit embarrassed by their appearance come out of their shells and get involved in activities. These kids are much happier.”

**Strabismus: The Importance of Timely, Specialized Care**

Brian G. Mohney, M.D., a pediatric ophthalmologist at Mayo Clinic Children’s Center in Rochester, Minnesota, answers questions about Mayo Clinic’s approach to treating children with strabismus.

**How common is strabismus?**

Strabismus is relatively common. Between 2 and 4 percent of kids have esotropia, and 1 to 1.5 percent have exotropia. Almost 1 out of 20 kids has strabismus.

**When should children be referred to a pediatric ophthalmologist?**

Uncorrected strabismus can have far-reaching effects, including loss of vision and stereocuity, and adverse effects on psychosocial development and gainful employment. Esotropia, in general, is most urgent in terms of requiring immediate attention to manage any vision loss or adverse effects on stereocuity. Exotropia is generally a more intermittent and slowly progressive disorder, allowing for observation over time. However, a child with any form of strabismus should be seen as soon as possible by an eye specialist to rule out vision loss or a visually dangerous or unexpected cause of the deviation.

Sometimes esotropia isn’t present from birth. A child’s eyes might be fine up to age 3 or 4 years but then suddenly start to cross. If we catch that crossing early and straighten the eyes with surgery within three to six months, the 3D vision starts to work again. If a child lacks the potential for developing normal 3D vision, the eyes are prone to drifting again. Amblyopia or poor vision due to strabismus — esotropia or exotropia — that isn’t corrected before age 9 will result in a permanent loss of vision.

**How can you determine when esotropia began in an infant or child?**

Oftentimes the parents aren’t aware of when it started. They might think it’s been there months when it’s actually been there years, or vice versa.
We often ask parents to bring in pictures of the child so that we can determine the onset.

**How is strabismus treated at Mayo Clinic Children’s Center?**

For 99 percent of our patients, an office visit alone is enough for us to diagnose their eye conditions and provide a treatment plan. Each child undergoes a careful history and clinical evaluation.

Exotropia, depending on the severity, can be simply observed or may require treatment such as glasses, part-time patching or even eye muscle surgery. Children with esotropia are generally treated at the time of their first office examination with glasses and patching for those who have amblyopia. Esotropia that does not respond to glasses will require surgery, usually within several months, to preserve stereopsis.

If we treat the child with glasses, we have a follow-up visit about six weeks afterward.

We can usually determine by then if the glasses will be sufficient or if the child will need surgery. Seven out of 8 kids will successfully respond with one surgery. In more-complicated cases, 2 out of 3 will straighten with one surgery; the remaining third will need a second surgery and sometimes more.

**What signs and symptoms of other eye disorders should physicians look for when considering referrals?**

If one of a child’s pupils is a different color than the other — either lighter or darker — the child might have a cataract or retinoblastoma. Such children should be referred for evaluation within a week.

About 11 percent of newborns have a blocked tear duct. The vast majority of these blockages will resolve on their own. For those that don’t, we used to wait until the child was a year old before treatment. But we recently studied 2,000 of these kids and found that the rate of spontaneous resolution drops after age 9 months. We’re now advocating treatment at age 8 or 9 months since there’s no real reason to wait.