GUIDE TO THE TREATMENT OF CHIARI I MALFORMATION
About the Division of Pediatric Neurosurgery

The division of Pediatric Neurosurgery at Children’s Memorial Hermann Hospital specializes in the diagnosis and treatment of Chiari malformations as part of a complete diagnostic and treatment program for pediatric neurological disorders. The affiliated pediatric neurosurgeons at UTHealth Medical School provide comprehensive, coordinated care for patients with Chiari malformation in addition to conducting research that provides evidence-based care and potential new treatments for children with this condition.

What is a Chiari I Malformation?

Chiari I malformation, the most common and treatable type of Chiari malformation, is a neurological disorder in which the cerebellum – the part of the brain that controls balance – descends out of the skull into the spinal canal. Specifically, the portions of the cerebellum that descend lower than they should are called the cerebellar tonsils (not to be confused with the tonsils located in the throat). Normally, the cerebellum is located in an indented space at the lower back of the skull above the foramen magnum, a funnel-like opening to the spinal canal. Chiari I malformation is diagnosed, usually by MRI, when part of the cerebellum is located below the foramen magnum. Normally, only the spinal cord passes through this opening.

Chiari I malformations may develop when the indented space is smaller than normal, causing the cerebellum to be pushed downward into the foramen magnum and the upper part of the spinal canal. The result is pressure on the cerebellum that may affect functions controlled by this area of the brain and block the flow of cerebrospinal fluid (CSF) to and from the brain. CSF is a clear liquid that surrounds and cushions the brain and spinal cord.
Symptoms

Chiari I malformation may or may not cause symptoms and usually is first noticed in adolescence or adulthood, often during examination for another condition. The most common symptom is a specific type of headache which occurs in the back of the head and may involve the neck as well. Typically, these headaches are short-lived and worsened with “valsalva maneuvers” – when a person strains during exercise, coughing or other activities. They are often relieved when such activities are stopped.

Other symptoms may include:
• Balance problems
• Muscle weakness
• Numbness or other unusual feelings in the arms or legs
• Dizziness
• Vision problems
• Ringing or buzzing in the ears
• Difficulty swallowing
• Apnea (suspension or holding of breath)

Infants with Chiari malformation may have difficulty swallowing, irritability when being fed, excessive drooling, a weak cry, gagging or vomiting, arm weakness, breathing problems, developmental delays and an inability to gain weight.

Other Conditions Associated with Chiari I Malformation

Children and adolescents who have a Chiari I malformation may also have hydrocephalus and syringomyelia.

Hydrocephalus is the excessive accumulation of cerebrospinal fluid (CSF) in the brain causing increased pressure. If left untreated, loss of mental and physical abilities, brain damage and even death may occur.
Early diagnosis and timely treatment are important to the successful treatment and recovery of children with hydrocephalus. After birth, infants with hydrocephalus may exhibit growth of the head at a faster rate than normal due to the buildup of excess CSF. Older children may experience the sudden onset of severe headaches accompanied by vomiting as well as possible loss of bladder control, irritability, sleepiness, seizures and loss of balance, motor skills and memory.

**Syringomyelia** is a disorder in which a fluid collection, called a syrinx, forms within the spinal canal. When the syrinx grows, it expands and puts pressure on the spinal cord, causing pain, weakness and stiffness in the back, shoulders, arms or legs. Other symptoms may include headaches and a loss of the ability to feel extremes of hot or cold, especially in the hands. Some children and adolescents have severe arm and neck pain.

**Diagnosis**

If Chiari I malformation is suspected, the first step in diagnosis is a physical examination which includes a complete neurological examination. Your physician may also order an MRI, which uses radio waves and a powerful magnetic field to produce a detailed 3-D image of body structures, including tissues, bones and nerves. MRI scans include images of the brain to assess the Chiari malformation and rule out hydrocephalus or other conditions. They also include images of the complete spine to look for a syrinx. Special MRI scans to assess the flow of fluid around the brain and upper spinal cord, called CINE CSF Flow studies, may help your child’s pediatric neurosurgeon determine whether treatment is required.

MRI studies can take up to several hours and often require anesthesia in children. To minimize the length of the imaging study, the pediatric neurosurgery team at Children’s Memorial Hermann Hospital has instituted an innovative “Quick Brain” MRI protocol. Quick Brain MRI studies can be completed in less than five minutes, do not require sedation and have no radiation exposure. Your child’s pediatric neurosurgeon can tell you whether your child is a good candidate for this study.
Additional tests that your physician may order include somatosensory evoked potentials (SSEPs) and brainstem auditory evoked responses (BAERs). These tests assess the function of the spinal cord and brainstem, respectively, and may provide information to help your child’s physician determine the appropriate treatment plan.

**Traditional and Minimally Invasive Surgery**

Not all patients with Chiari I malformation require surgery. When Chiari I malformation is asymptomatic and does not interfere with daily activities, your physician will recommend observation without surgical intervention. Medications may also be prescribed to ease pain and other symptoms.

Surgery is the only treatment available to improve symptoms directly attributed to Chiari I malformation or to stop the progression of damage to the central nervous system. Posterior fossa decompression surgery creates more space for the cerebellum and relieves pressure on the spinal column. The procedure involves making an incision at the back of the head and removing a small portion of the bottom of the skull to provide additional space for cerebrospinal fluid (CSF) to flow normally.

The surgeon may also make an incision in the dura, a membrane that covers the brain and spinal cord. A duraplasty may be performed, in which a patch is sewn into the dura to create more space for the flow of CSF. Whether or not to open the dura during decompression surgery and perform a duraplasty is a source of controversy. A variety of different materials can be used to perform a duraplasty, including synthetic grafts and pericranium, a thick membrane that is attached to the skull. Pericranium can be obtained through the same incision required for the posterior fossa decompression.
When hydrocephalus occurs in association with Chiari I malformation, the ideal treatment is a minimally invasive procedure called endoscopic third ventriculostomy (ETV). This procedure enables normal circulation of fluid via a small incision on the top of the skull. An endoscope (a small camera with working ports) is inserted into the ventricles (the fluid spaces of the brain) and a hole is made in a membrane called the floor of the third ventricle which allows fluid to circulate normally. Some patients may not be candidates for ETV and may require a shunt, in which a flexible tube is inserted into the ventricle and fluid is drained into the abdomen or another part of the body.

**Treatment of Syringomyelia**

Typically, posterior fossa decompression results in improvement of symptoms and also causes either a decrease or stabilization of an associated syrinx. If a syrinx persists despite posterior fossa decompression, a shunt may be placed into the syrinx in order to drain the syrinx into the space next to the spinal cord (called the subarachnoid space) or into another compartment.

**Collaboration with Parents and Referring Physicians**

Parents are an integral part of the care team at Children’s Memorial Hermann Hospital. Every effort is made to keep you fully informed and involved in your child’s care and communication with your pediatrician or referring physician about your child’s progress throughout the evaluation, diagnosis and treatment process is a priority. After all initial patient consultations, referring physicians will receive a summary that includes the initial diagnosis, pending tests and treatment options. Additionally, updates are provided to the referring physician throughout all subsequent appointments.
To refer a patient or speak with a physician, please call 832.325.7234. For urgent consultations, please contact the page operator at 713.704.4000 and ask to page the pediatric neurosurgeon on call.

The Pediatric Neurosurgery Team

David I. Sandberg, M.D., FACS, FAAP
Director of Pediatric Neurosurgery
Mischer Neuroscience Institute
Associate Professor and Chief
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Stephen Fletcher, D.O.
Associate Professor
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Manish Shah, M.D.
Assistant Professor
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Lee Ann Conrad, PA-C
Physician Assistant
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Jacklyn Eyre, PA-C
Physician Assistant
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Lillian Sprague, PA-C
Physician Assistant
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Reyna Balderas
Medical Assistant
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital

Paula Maldonado
Medical Assistant
Department of Pediatric Surgery, Division of Pediatric Neurosurgery
UTHHealth Medical School • Children’s Memorial Hermann Hospital