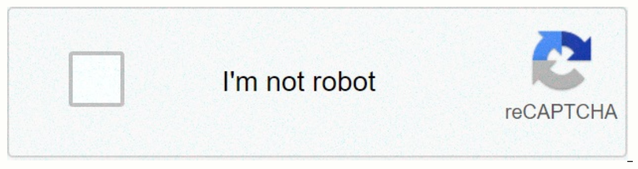

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Spina bifida occulta arnold chiari malformation

Chiari malformations are structural defects at the base of the skull and cerebellum, the part of the brain that controls balance. Usually the cerebellum and part of the brain stem sit above a gap in the skull that allows the spinal cord to pass through it (called a magnum foramen). When part of the cerebellum extends under the magnum foramen and into the upper spinal canal, it is called chiari malformation (CM). Chiari malformations can develop when parts of the skull are smaller than usual or misshapen, which forces the cerebellum to be pushed into the magnum foramen and spinal canal. This causes pressure on the cerebellum and brain stem that can affect functions controlled by this area and block the flow of cerebrospinal fluid (CSF)—the clear fluid that surrounds and cushions the brain and spinal cord. CSF also circulates nutrients and chemicals filtered from the blood and removes waste products from the brain. over What causes this malformation? CM has several different causes. Most often caused by structural defects in the brain and spinal cord that occur during fetal development. This can be the result of genetic mutations or the diet of mothers who are deficient in certain vitamins or nutrients. This is called primary or congenital Chiari malformations. It can also be caused later in life if spinal fluid is excessively drained from the waist area or spinal barracist either due to traumatic injury, disease, or infection. This is called the acquired or secondary Chiari malformation. Primary Chiari malformations are much more common than secondary Chiari malformations. what are the symptoms of Chiari malformations? Headaches are a typical sign of Chiari malformations, especially after a sudden cough, sneeze, or strain. Other symptoms may vary among individuals and may include: hearing neck pain or balance problems of muscle weakness or numb dizziness difficulty swallowing or talking vomiting ringing or buzzing in the ear (tinnitus) curvature of the spine (scoliosis) insomnia depressive problems with hand coordination and fine motor skills. Some individuals with CM may not show any symptoms. Symptoms may change for some individuals, depending on tissue and nerve compression and on CSF pressure buildup. Babies with Chiari malformations may have difficulty swallowing, irritability when fed, excessive saliva, weak crying, choking or vomiting, arm weakness, stiff neck, breathing problems, developmental delays, and inability to gain weight. how is the AGMS classified? Chiari malformations are classified by the severity of the disorder and the part of the brain that protrudes into the spinal canal. Type 1 Chiari malformations occur when the bottom of the cerebellum (called cerebellar tonsils) extends to the magnum foramen. Usually, only bone marrow that passes through this opening. Type 1—which may not cause symptoms—is the most common form of CM. Usually first first adolescence or adulthood, often inadvertently during examination for other conditions. Adolescents and adults who have CM but no symptoms can initially develop signs of the disorder later in life. Chiari Malformation Type II Individuals with Type II have symptoms that are generally more severe than in Type 1 and usually appear during childhood. The disorder can cause life-threatening complications during in-baby or early childhood, and treating it requires surgery. In Type II, also called classic CM, both cerebellum tissue and brain stem protrude into magnum foramen. Also the neural network connecting the two parts of the cerebellum may be missing or only partially formed. Type II is usually accompanied by myelomeningocele—a form of spina bifida that occurs when the spinal canal and spine do not close before birth. (Spina bifida is a disorder characterized by the development of the brain, spinal cord, and/or its incomplete protective cover.) Myelomeningocele usually results in partial or complete paralysis of the area below the opening of the spine. The term Arnold-Chiari malformation (named after two pioneering researchers) is specific to Type II malformations. Type III Chiari malformations are extremely rare and the most serious form of Chiari malformations. In Type III, several cerebellums and brain stems protrude, or hernias, through abnormal opening at the back of the skull. It can also include membranes that surround the brain or spinal cord. Type III symptoms appear in insancy and can lead to debilitating and life-threatening complications. Infants with Type III can have many of the same symptoms as those with Type II but can also have severe additional neurological defects such as mental and physical delays, and seizures. Type IV Chiari malformations involve incomplete or under-developed cerebellums (a condition known as cerebellar hypooplasia). In this rare CM form, the cerebellum is located in a normal position but parts are missing, and parts of the skull and spinal cord may be visible. what other conditions are associated with Chiari malformations? Hydrocephalus is an excessive buildup of CSF in the brain. CM can block the normal flow of this fluid and cause pressure inside the head which can result in a mental disability and/or an enlarged skull or misshapen. Severe hydrocephalus, if untreated, can be fatal. The disorder can occur with chiari malformations, but is most commonly associated with Type II. Spina bifida is an incomplete closure of the spine and membranes around the spinal cord. In babies with spina bifida, the bones around the spinal cord do not form properly, causing defects in the lower spine. While most children with disabilities it has a mild form so they have no neurological problems, individuals with Type II Chiari malformations usually have myelomeningocele, and the baby's spinal cord remains open in one area of the back and and Spine. The membrane and spinal cord protrude through the opening in the spine, creating a sac in the baby's back. This can lead to a number of neurological disorders such as muscle weakness, paralysis, and scoliosis. Syringomyelia is a disorder in which csf-filled tubular cysts, or syrinx, form inside the central tract of the spinal cord. The growing syrinx destroys the middle part of the spinal cord, resulting in pain, weakness, and stiffness in the back, shoulders, arms, or legs. Other symptoms may include a loss of the ability to feel extreme heat or cold, especially at hand. Some individuals also have severe arm and neck pain. Tethered marrow syndrome occurs when a child's spinal cord abnormally attaches to tissue around the lower part of the spine. This means the spinal cord cannot move freely inside the spinal canal. As a child grows, the disorder worsens, and can result in permanent damage to the nerves that control the muscles in the lower body and legs. Children who have myelomeningocele have an increased risk of developing tethered cords later in life. Spinal curvature is common among individuals with syringomyelia or CM Type I. The spine can bend left or right (scoliosis) or can bend forward (kyphosis). over How common is Chiari malformations? In the past, it was estimated that the condition occurred at about one out of every 1,000 births. However, the increased use of diagnostic imaging has shown that Chiari malformations may be much more common. Complicating this estimation is the fact that some children born with this condition may never experience symptoms or show symptoms only in adolescence or adulthood. Chiari malformations are more common in women than in male malformations and Type II is more prevalent in certain groups, including those of Celtic descent. top How was Chiari malformation diagnosed? Currently, no tests are available to determine whether the baby will be born with Chiari malformations. Because Chiari malformations are associated with certain birth defects such as spina bifida, children born with such defects are often tested for malformations. However, some malformations can be seen in ultrasound images before birth. Many people with Chiari malformations have no symptoms and their malformations are only found during diagnosis or treatment for other disorders. The doctor will perform a physical examination and examine the person's memory, cognition, balance (functions controlled by the cerebellum), touch, reflexes, sensations, and motor skills (functions controlled by the spinal cord). Doctors can also order one of the following diagnostic tests: Magnetic resonance imaging (MRI) is the most frequent imaging procedure to diagnose Chiari malformations. It uses radio waves and a strong magnetic field to painlessly produce detailed three-dimensional images or slices of two-dimensional body structures, including tissues, organs, bones, and nerves. X-rays using electromagnetic electromagnetics to produce images of specific bones and tissues in the film. X-rays of the head and neck cannot reveal cm but can identify bone abnormalities often associated with the disorder. Computed tomography (CT) uses X-rays and computers to produce images of bones and two-dimensional blood vessels. CT can identify hydrocephalus and bone abnormalities associated with Chiari malformations. how was Chiari malformation treated? Some CM shows no symptoms and does not interfere with a person's activity in daily life. In these cases, the doctor can only recommend regular monitoring with an MRI. When an individual experiences pain or headaches, a doctor can prescribe medication to help alleviate symptoms. Differentiating In many cases, surgery is the only treatment available to alleviate symptoms or stop the development of damage to the central nervous system. Differentiation can improve or stabilize symptoms in most individuals. More than one operation may be required to treat the condition. The most common operation to treat Chiari malformations is posterior fossa decompression. This creates more space for the cerebellum and reduces pressure on the spinal cord. The operation involves making an incision at the back of the head and removing a small part of the bone at the bottom of the skull (craniectomy). In some cases, the curved and bony roof of the spinal canal, called lamina, can also be removed (spinal laminectomy). Surgery should help restore the normal flow of CSF, and in some cases may be enough to relieve symptoms. Furthermore, surgeons can make incisions in dura, the protective cover of the brain and spinal cord. Some surgeons perform Doppler ultrasound tests during surgery to determine whether opening dura is even necessary. If areas of the brain and spinal cord are still crowded, surgeons can use a procedure called electrocautery to remove cerebellar tonsils, allowing more free space. These tonsils have no recognized function and can be removed without causing known neurological problems. The final step is to sew dura patches to expand the space around the tonsils, similar to removing the belt on a pair of pants. This patch can be made of artificial materials or tissue harvested from other parts of a person's body. Babies and children with myelomeningocele may require surgery to reposition the spinal cord and close the opening behind. Findings from the National Institutes of Health (NIH) show that this surgery is most effective when performed prenatally (when the baby is still in the womb) rather than after birth. Prenatal surgery reduces the onslence of hydrocephalus and returns the cerebellum and brain stem to alignment Normal. Hydrocephalus can be treated with a shunt system (tube) that drains excess fluid and relieves pressure inside the head. A sturdy tube, inserted into the head, connected to a flexible tube placed under the skin. This tube drains excess fluid into the chest or stomach so that it can be absorbed by the body. An alternative surgical treatment in some individuals with hydrocephalus is the third ventriculartomy, a procedure that increases the flow of CSF out of the brain. A small hole was created at the bottom of the third ventricle (brain cavity) and CSF was diverted there to relieve pressure. Similarly, in cases where surgery is ineffective, the doctor can open the spinal cord and insert a shunt to dry out syringomyelia or hydromyelia (increased fluid in the middle channel of the spinal cord). what research is being done? The mission of the National Institute of Neurological Disorders and Stroke (NINDS) is to seek out fundamental knowledge of the brain and nervous system and use that knowledge to reduce the burden of neurological diseases. NINDS is a component of the National Institutes of Health (NIH), a major proponent of biomedical research in the world. NINDS conducts research and provides research grants to major medical research institutions across the country. NIH genetics is examining genetic factors that increase the risk of developing Chiari malformations and related brain disorders. Recent research has identified gene mutations in the PI3K-AKT signaling pathway that cause brain overgrowth that can accompany hydrocephalus, CM, and other brain disorders. To better understand the genetic factors responsible for chiari I malformations, NINDS scientists looked for mutations of other genes that can act through PI3K-AKT signals and other pathways. This research could lead to new diagnostic tests and better treatment options for Chiari malformations and other developmental brain disorders. Brain mechanism Certain signals at the midbrain-hindbrain (MHB) boundary tell the brain to develop the cerebellum and other parts of the brain correctly. However, how this brain region is started, formed, and maintained is not well understood. NINDS scientists are studying zebrafish embryos to gain a better understanding of how MHB is formed. This will provide valuable insight into the development of the human brain, especially the cerebellum. Other investigators are studying the expression of different growth factors in the development of the brain, skull, spine, and spinal cord. Interference with normal gene function through gene mutations or environmental factors can affect the development of CM. Treatment The purpose of treating syringomyelia with surgery is to remove syrinx and prevent further spinal cord injury. Little is known about the effects of surgery over time on muscle strength, pain levels, and overall function. NINDS scientists are examining individuals who have syringomyelia or are at risk of being exposed to the disorder, including those with CM. By recording more than 5 years of symptoms, muscle strength, general level of function and MRI scan findings from individuals receiving standards for syringomyelia, the researchers hope to get more information factors that affect the development, development, and relief of symptoms. The results of the study may allow doctors to provide more accurate recommendations to individuals with syringomyelia regarding optimal surgical or non-surgical treatments. More information about research on Chiari malformations supported by NINDS and other NIH Institutes and Centers can be found using NIH RePORTER, a searchable database of current and past research projects supported by the NIH and other federal agencies. RePORTER also includes links to publications and resources from these projects. above Where can I get more information? For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at: BRAIN P.O. Box 5801 Bethesda, MD 20824 800-352-9424 800-352-9424