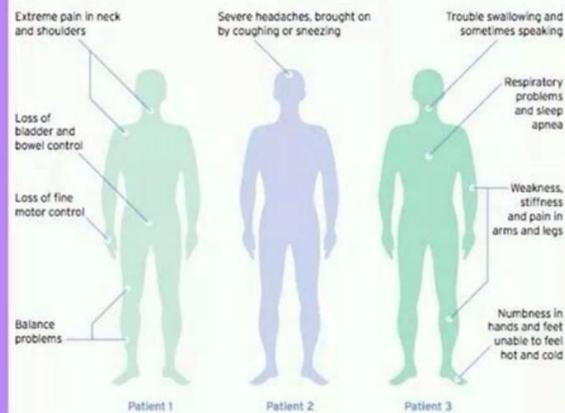


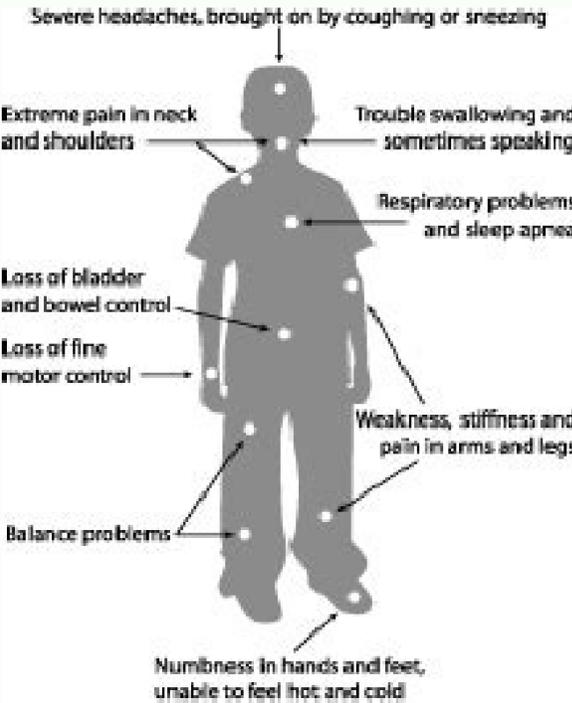
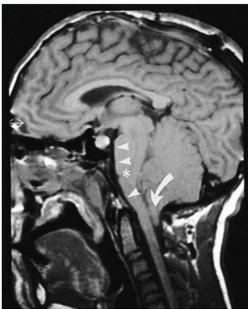
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# Chiari

Every case is different



# Malformation



Is chiari malformation rare. What happens if chiari malformation goes untreated. How serious is chiari malformation type 1. Can chiari malformation be caused by injury. What age is chiari malformation diagnosed.

ABOUT CAUSES DIAGNOSIS TREATMENT NEXT STEPS A Chiari malformation (CM) is a problem with how the brain sits in the skull. The brain normally sits fully inside the skull. With a Chiari malformation, the lower part of the brain (cerebellum) dips down through a normal opening (foramen magnum) at the bottom of the skull. In some cases, more brain tissue also dips down through this opening. This puts pressure on parts of the brain and spinal cord, and can cause mild to severe symptoms. These can include head or neck pain and trouble with balance or movement. In most cases, the problem is present at birth (congenital). There are 4 types of Chiari malformations: Type I (CM-I). This is the most common type. Part of the cerebellum dips down through the bottom of the skull. This type is most often congenital (also called primary CM), but is often not found until a child is a teen or young adult. In rare cases, this type may also develop later in life. This is known as acquired or secondary CM. It occurs from a loss of spinal fluid. This can happen because of an injury, contact with harmful substances, or an infection. Type II (CM-II or Arnold-Chiari malformation). Part of the cerebellum and the brain stem dip down through the bottom of the skull. This is most often seen in babies born with spinal myelomeningocele or spina bifida. A myelomeningocele is when a part of the spinal cord and backbone (spine) develop outside the body. A common problem with Type II CM is too much fluid on the brain (hydrocephalus). The extra fluid causes the pressure in the brain to increase and the skull bones to expand beyond normal size. Type III (CM-III). This type is the most severe. The cerebellum, brain stem, and possibly other parts of the brain dip down through the bottom of the skull. In rare cases, the brain and brain covering may poke out through the back of the head or neck. A baby with Type III CM may not live long. Children who do will have severe nervous system problems such as thinking problems, seizures, and muscle problems. Type IV. This is a very rare condition where the brain doesn't develop fully. Most babies with this type don't survive. Doctors don't know the exact cause of a congenital Chiari malformation type I. A problem during fetal growth may cause the defect. It may be caused by contact with harmful substances during pregnancy. Or it may be linked with genetic problems that run in families. An acquired Chiari malformation type I happens to a person after birth. It is caused by extra leaking of spinal fluid from the lower back (lumbar) or chest (thoracic) areas of the spine. This can happen because of an injury, contact with harmful substances, or an infection. Your child may not have any symptoms. Or symptoms may develop slowly over time. Most children don't have symptoms until they are teens or young adults. The most common symptoms are headaches or pain in the back of the head or neck. The headaches and pain are made worse by coughing, laughing, or sneezing. Your child may also have other symptoms of a Chiari malformation type I. These include: Hoarseness or trouble speaking Trouble swallowing Rapid, back and forth eye movements (nystagmus) Periods of not breathing during sleep (sleep apnea) Weakness or abnormal movements Trouble with balance Abnormal reflexes Abnormal shape of the spine (scoliosis) Your child may also have a pocket of fluid in the spinal cord or brain stem. This is called a syrinx. A syrinx can cause trouble walking or pain in the arms or legs. In a child with no symptoms, the problem may be found when imaging tests are done for other reasons. For a child with symptoms, the healthcare provider will ask about your child's health history and give your child a physical exam. He or she may refer your child to a specialist. Imaging tests are done to detect a Chiari malformation type I. Your child may have one or more of these tests: MRI. This test uses large magnets and a computer to make detailed pictures of the inside of the body. In some cases, a special dye is injected into a vein for the test. This dye helps show organs more clearly. CT scan. This test uses a series of X-rays and a computer to create detailed pictures of the inside of the body. A CT scan is more detailed than a regular X-ray. Your child may be treated by neurologists and neurosurgeons. These are experts in brain and spinal cord problems. Treatment will depend on your child's symptoms, age, and general health. It will also depend on how severe the condition is. With no symptoms. Your child's health may be watched closely. This may include frequent physical exams and MRI tests. Your child's healthcare provider may advise surgery to prevent problems. With symptoms. Your child's healthcare provider may prescribe medicines to reduce pain. Or he or she may advise decompression surgery. This is done to relieve pressure on the brain, or to restore the flow of spinal fluid. With few or no symptoms, but a syrinx. Your child's healthcare provider may suggest close monitoring of the problem with a special type of MRI called cine phase contrast. This helps look for blocked spinal fluid flow. Your child may need surgery, based on the MRI results or if symptoms get worse. With signs of sleep apnea. Your child may need a sleep study. In this test, your child will be monitored during sleep to look for problems. A sleep study can also help the healthcare provider decide on additional treatment. It is possible that your child may not have any symptoms in the future. But some children develop complications. These include: Long-term pain Development of syrinx Permanent damage to muscles or nerves Paralysis Carefully watching for changes in your child's health can help prevent complications. This helps to make sure that treatment is done early. It is hard for healthcare providers to predict how a Chiari malformation type I will affect a child's long-term health. Your child may not have any changes caused by the defect. Or he or she may have worsening nervous system problems. Your child's health will be closely watched. This will include with frequent physical exams and imaging tests such as MRI. There is ongoing research as to how to best manage Chiari malformations. Call your child's healthcare provider if you notice any changes in your child. Be sure to call if you notice problems with: Breathing Swallowing Feeding Speaking Walking or moving Key points about Chiari malformation type I in children With a Chiari malformation, the lower part of the brain dips down through a normal opening at the bottom of the skull. In some cases, more brain tissue also dips down through this opening. In most cases, the problem is present at birth (congenital). The most common symptoms are headaches or pain in the back of the head or neck. The headaches and pain are made worse by coughing, laughing, or sneezing. Treatments include careful watching, surgery, and frequent exams and tests. Carefully watching for changes in your child's health can help prevent complications. This helps to make sure that treatment is done early. Tips to help you get the most from a visit to your child's healthcare provider: Know the reason for the visit and what you want to happen. Before your visit, write down questions you want answered. At the visit, write down the name of a new diagnosis, and any new medicines, treatments, or tests. Also write down any new instructions your provider gives you for your child. Know why a new medicine or treatment is prescribed and how it will help your child. Also know what the side effects are. Ask if your child's condition can be treated in other ways. Know why a test or procedure is recommended and what the results could mean. Know what to expect if your child does not take the medicine or have the test or procedure. If your child has a follow-up appointment, write down the date, time, and purpose for that visit. Know how you can contact your child's provider after office hours. This is important if your child becomes ill and you have questions or need advice. 1. Chiari H. Über Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. Dtsch Med Wochenschr. 1891;17:1172-1175. doi: 10.1055/s-0029-1206803. 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Clinical summary of 16 infants and toddlers with Chiari I malformation CaseAgeSexSymptomsFonsillar ectopia1st operation techniquePostop outcome (after 1st op)Recurrence symptomsTime to 2nd operation (months)Reoperation techniquePostop outcome (2nd op)(months)LLevelDistance(mm)SymptomsMR (3-6 months)SymptomsMRI6MInconsonable cry with arching back, nighttime wakingC16Type 1MinorUnchangedGrabbing occiput, nighttime wakingC22Type 2\*ImprovedImprovedC27Stridor (left vocal cord paresis), spasticityC15Type 1TemporaryUnchangedGagging,vocal cord paresisType 2\* + PMCImprovedImproved311FNighttime waking with coughing/snooring, inconsonable cryC1-210Type 1ImprovedUnchangedGagging, snoring35Type 2\*ImprovedImproved417Head grabbing, coughing bout or cries, nighttime wakingC212Type 1TemporaryUnchangedNighttime waking24Type 2\* + PMCImprovedImproved519MEmesis with weight loss, occipital grabbing, irritabilityC14Type 1ImprovedImproved---621MBreath holding spells, seizures, irritabilityC1-210Type 1ImprovedMin. improved---725MBilateral estropiaC27Type 1TemporaryUnchangedHeadaches, emesis,hydrocephalus7Type 2\* + PMCImprovedImproved829MCrossing eyes with irritability, sleep apnea, holding back of headC28Type 1ImprovedUnchanged---933FNF1, holding head, emesisC1-212Type 1ImprovedUnchanged---1035FFailure to thrive, irritability, inconsonable cry with arching backC18Type 1ImprovedImproved---1111Increased head sizeC211Type 2\*ImprovedImproved---1219M Holding head with screamingC17Type 2ImprovedImproved---1319FFailure to thrive, poor gain of weightC2-316Type 2\*ImprovedImproved---1423MSleep apnea, inconsonable cry with arching backC2-310Type 2\* + PMCTemporaryUnchangedHead grabbing, cough, motor weakness7Type 2\*ImprovedImproved1527MDevelopmental delay, nighttime waking, head rubbingC1-213.5Type 2\*ImprovedImproved---1630FFailure to thrive, holding head expressing pain, gait ataxiaC1-26Type 2ImprovedMin. improvedHeadache, nighttime waking15Type 1ImprovedImproved

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