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The license may not give you all of the permissions necessary for your intended use. For example, other rights such as publicity, privacy, or moral rights may limit how you use the material. Chiari 2 malformation is a relatively common congenital malformation of the posterior fossa. Chiari 2 malformation is almost always associated withmyelomeningocele and is characterized by caudal displacement of the cerebellar vermis, brainstem, and fourth ventricle. Chiari 2 malformation can be associated with numerous additional findings, most commonly hydrocephalus and syringomyelia.This disorder can have severe morbidity and mortality if it is not identified and treated promptly. The natural history of patients with Chiari 2 malformation is quite severe, with medullary symptoms remaining the leading cause of death.Chiari 2 malformation may present a variety of clinical manifestations, which can include spinal symptoms secondary to myelomeningocele, tethered cord syndrome or syringomyelia, symptoms of secondary hydrocephalus, brainstem symptoms, or those due to lower cranial nerve dysfunction. Management of Chiari 2 malformation depends on the extent of the craniospinal malformation and its associated neurological impairments.This activity for healthcare professionals aims to enhance learners' competence in selecting appropriate diagnostic tests, managing Chiari 2 malformation, and fostering effective interprofessional teamwork to improve outcomes.Objectives:Identifythe etiology of Chiari 2 malformation.Implementrecommended evaluation strategies for patients with Chiari 2 malformation.Applyevidence-based management strategies for patients with Chiari 2 malformation.Collaborate withthe interprofessional teamto improve care coordination for patients with Chiari 2 malformation.Access free multiple choice questions on this topic.At the end of the nineteenth century, the pathologists Julius Arnold (1835-1915) and Hans Chiari (1851-1916) described a complex clinical and pathological condition involving deformity of the cerebellum and brainstem in children. Chiari malformations are now defined as a spectrum of hindbrain abnormalities involving the cerebellum, brainstem, skull base, and cervical cord. According to the type of herniation of the brain tissue displaced in the spinal canal and the characteristics of the anomalies of the brain or spine development, 4 types of Chiari malformations have been classified. Together with basilar invaginations, Chiari malformations represent the most common craniocervical junction malformations seen in adults.Chiari malformation type 2 or Chiari 2 malformation, commonly referred to as Arnold-Chiari malformation, is usually associated with spina bifida and is characterized by caudal displacement of the cerebellar vermis, brainstem, and fourth ventricle.[1][2]Despite its name, rather than a continuation of a spectrum with Chiari 2 malformation representing a severe version of Chiari 1 malformation, these malformations represent distinct disease processes with overlapping radiographic findings.(seeImage.Chiari Malformation Types 1 and 2). Chiari 2 malformation can be associated with numerous additional findings, most commonly hydrocephalus and syringomyelia.Clinically,Chiari 1 malformation can be an asymptomatic hindbrain hernia, while Chiari types 2, 3, and4 are congenital and clinically significant. Chiari 2 malformation may present a variety of clinical manifestations, which can include spinal symptoms secondary to myelomeningocele, tethered cord syndrome or syringomyelia, symptoms of secondary hydrocephalus, brainstem symptoms, or those due to lower cranial nerve dysfunction. The nosographic collocation of type 0 or 0.5 Chiari malformation or Chiari-like symptoms without tonsillar herniation,type 1.5 Chiari malformation between types 1 and 2, and complex Chiariis controversial and not universally accepted.[3]Table.Chiari Malformation Types. The precise etiology of Chiari 2 malformation is not entirely understood, with multiple proposed theories over the years. The most recent and accepted theory is the "unified theory," which assumes that the neural tube defect is the etiology from which the cranial features follow.[4]Earlier theories based on tethering of the spinal cord by the myelomeningocele resulting in caudal traction on the hindbrain and brainstem structures have fallen out of favor.Chiari 2 malformations are relatively rare, with no large-scale study accurately estimating their incidence. Experts have estimated that the incidence of Chiari 2 malformation occurs in 0.44 of 1,000 births without a gender predominance.[5] Given its association with myelomeningocele, perinatal folate supplementation can decrease its incidence.[6]The current best evidence for the pathophysiology of Chiari 2 malformation is the development of a pressure gradient across the foramen magnum, creating a craniospinal pressure gradient that will cause or hasten the development of a hindbrain hernia. The unified theory assumes that the neural tube defect occurs first, followed by all other manifestations.[4] Ultimately, multiple abnormalities ensue because the lack of mechanical support that the typically distended primitive ventricles provide prohibits normal development of the brain and overlying skull.Chiari 2 malformation may present a variety of clinical manifestations, which can include spinal symptoms secondary to myelomeningocele, tethered cord syndrome or syringomyelia, symptoms of secondary hydrocephalus, brainstem symptoms, or those due to lower cranial nerve dysfunction. Clinical findings related to the Chiari 2 malformation are often due to brainstem and lower cranial nerve dysfunction. The age of symptom onset is rare in adulthood. The clinical presentation in neonates also differs from that of the older child, who is more likely to experience insidious, rarely severe symptom onset.[7]The symptomatic neonate with Chiari 2 malformation is more likely to develop rapid neurologic deterioration over several days with profound brainstem dysfunction.[7]Other associated clinical findings may include:Facial weaknessWeak or absent cryNystagmus, especially downbeat nystagmusOpisthotonosAspirationArm weakness that may progress to quadriparesis[8]Apneic spells due to impaired ventilatory driveTransient stridor that may progress to respiratory arrest secondary to vagal nerve paresisSwallowing difficulties and decreased gag reflex that may manifestas poor feeding, prolonged feeding, nasal regurgitation, cyanosis during feeding, and pooling of oral secretions[7][9]Symptoms related to syringomyelia, if present, are often associated with the involved spinal cord level, size, and location of the syrinx. Syringomyelia classically presents with dissociated sensory loss (ie, preserved light touch and proprioception with loss of pain and temperature sensation), variable pain involvement, autonomic symptoms, sensorimotor dysfunction, Lhermitte phenomenon, neurogenic arthropathy, and musculoskeletal abnormalities.Scoliosis may present in associated with syringomyelia secondary to lateralized anterior horn compression manifesting as inequality of paravertebral muscle strength. Patients with scoliosis secondary to underlying syringomyelia often present at a young age with an atypical curve, rapid curve progression, and back pain symptoms.Diagnostic Imaging StudiesWhen evaluating a patient with Chiari 2 malformation, the diagnosis is primarily based on the neuroanatomy seen with imaging studies. Neuroimaging with magnetic resonance imaging (MRI) provides the highest resolution in evaluating patients with Chiari 2 malformation.For patients who cannot tolerate an MRI, a computed tomography (CT) can also aid in diagnosing Chiari 2 malformation and provide further detail on osseous anatomy. However, the disadvantages of CT include lower resolution, posterior fossa artifact, and ionizing radiation to the child. If fetal ventriculomegaly is present, Chiari 2 malformation can be detected with fetal ultrasound in some cases.In addition to the characteristic features of cerebellar vermis, brainstem, and fourth ventricle caudal displacement, the following findings may also be present:Lemon sign: describes the frontal bone indentation depicting a lemonBanana cerebellum sign: describes the cerebellum wrapping around the brain stemBeaking of tectumAbsence of the septum pellucidum with an enlarged interthalamic adhesionHydrocephalusSyringomyeliaCraniolacunia of the skull (ie, Luckenschadel)[10]Skeletal abnormalities (eg, platybasia, basilar impression, and Klippel-Feil deformity)MicrogyriaHeterotopiasMyelomeningoceleTethered cord syndromeAgenesis or dysgenesis of the corpus callosum[11][12]Overall, diagnosing Chiari 2 malformation requires careful evaluation if there is evidence of spinal myelomeningocele. MRI can help diagnose Chiari 2 malformation by showing a downward displacement of the medulla and cerebellum vermis, among other possible findings described above.In this particular patient population, hydrocephalus or shunt failure should be suspected as the etiology of presenting symptoms first and foremost; these should be excluded before attributing symptoms to a syrinx, tethered cord syndrome, or Chiari 2 pathology.Ancillary Diagnostic StudiesOther ancillary studies that may be considered in addition to diagnostic imaging include:Sleep studies: Involves sleeping overnight in a room where they can monitor breathing, snoring, oxygenation, and seizure activity to identify evidence of sleep apnea.Swallow studies: Fluoroscopy is used to demonstrate cricopharyngeal achalasia.Laryngoscopy:This study can visually verify the movement and function of the vocal cords.[13][14][15]Managing Chiari 2 malformation depends on the extent of the craniospinal malformation and associated neurological impairments. The natural history of patients with Chiari 2 malformation is quite serious, with medullary symptoms remaining the leading cause of death. Surgical intervention is often required to repairassociated open neural tube defects and provide cerebrospinal fluid (CSF) diversion ofhydrocephalus, typically via shunting. Early suboccipital hindbrain decompression surgery is accepted in an infant or child with symptoms found to be directly related to Chiari 2 malformation. However, there remain few studies exploring the role of decompressive surgery in this patient population, creating ongoing controversy. Those arguing against surgery propose that many of the neurologic findings may be due to intrinsic abnormalities for which surgical intervention cannot improve.[16][17]Proponents of expeditious brainstem decompression argue that the histologic lesions are due to chronic brainstem compression and concomitant ischemia and that surgery should be carried out if the warning signs of stridor, apneic spells, or neurogenic dysphagia are present.[7][18]Clinicians should consider excluding viral etiologies before attributing the presence or acute worsening of stridor and apnea to the Chiari 2 malformation.Certain medical problems, including neonatal feeding difficulties, apnea, respiratory failure, and neurogenic bowel, may require medical or surgical management.[19]Surgical intervention to repair myelomeningocele is commonly required. Some evidence has demonstrated the benefits of performing the myelomeningocele repair on the in-uterofetus.[20]For infants with a myelomeningocele diagnosis who did not have a fetal intervention performed, delivery should ideally occur in a hospital with a level III neonatal ICU. Initial management following the delivery includes providing prophylactic antibiotics and covering the lesion. Within the first 72 hours, the open neural tube defectshould be repaired surgically.[21]Following surgery, the patient requires close evaluation for the development of hydrocephalus.[22]Serial evaluation for abnormal neurological function is necessary throughout life. Nearly all patients with myelomeningocele will have a neurogenic bladder. This typically requires intermittent catheterization to decrease the risk of renal disease. Notably, a change in function may suggest an acute neurological complication.[23]Hydrocephalus is a common problem encountered in the management of Chiari 2 malformation. Neuroimaging findings and the severity of symptoms primarily determine the timing and need for surgical intervention in patients. Urgent surgical intervention is required in patients with rapidly progressive hydrocephalus. Another important indication for surgery is if a patient is symptomatic and hashydrocephalus. Some symptoms include vomiting, developmental delays, headaches, focal neurologic deficits, and papilledema. An additional indication includes progressive ventriculomegaly and clear evidence of an obvious obstruction on imaging. The clinician should be mindful that in patients with Chiari 2 malformation, ventricular size may not change in the setting of shunt malfunction.[24]The most common surgical intervention is the placement of a shunt. CSF diversion by shunting preventsCSF from accumulating and increasing intracranial pressure, which can ultimately be fatal.The treating clinician should know that presenting signs and symptoms may be associated with myelomeningocele, syringomyelia, scoliosis, and hydrocephalus or shunt failure. The differential diagnoses for Chiari 2 malformation also includes the following:Fourth ventricle ependymomaLhermitte-Duclos diseaseRhombencephalosynapsisOther Chiari malformation typesThe prognosis of Chiari 2 malformation is variable, dependent on the extent of the malformations and the patient's symptoms. The mortality rate is 71% in infants with vocal cord paralysis, arm weakness, or cardiopulmonary arrest within 2 weeks of presentation, in comparison to patients with a more gradual deterioration who had a 23% mortality.[7] Bilateral vocal cord paralysis is a poor prognosticator for response to suboccipital decompression surgery.[7]In patients requiring suboccipital hindbrain decompression, 68% had a complete or near-complete resolution in symptoms, 20% had no improvement, and 12% had mild to moderate residual deficits.[7]In a published series on patients with Chiari 2 malformation who underwent suboccipital hindbrain decompression surgery, respiratory arrest was the most common cause of mortality in 8 out of 17 patients who died, followed by meningitis or ventriculitis in 6 patients, aspiration in 2 patients, and biliary atresia in 1 patient.[9] There was a 37.8% mortality rate in patients who underwent surgery, with follow-up ranging as far out as 6 years.[9]Chiari 2 malformation, or Arnold-Chiari malformation, is usually associated with spina bifida and is characterized by caudal displacement of the cerebellar vermis, brainstem, and fourth ventricle.[1]Most patients with myelomeningocele also have Chiari 2 malformation in addition tohydrocephalus. Diagnosis of Chiari 2 malformation is primarily made radiographically by MRI. The treatment is typically centered upon a surgical intervention but is complex because of the variability of the malformation and associated clinical presentation. The prognosis of Chiari 2 malformation is dependent upon the extent of craniospinal malformation.While nothing can be done to prevent congenital Chiari 2 malformation, patient education plays a significant role in familiarising the patient's family with the signs and symptoms of the condition and helping to manage expectations. Patient education and counseling regarding conservative and surgical management strategies are also critical in the shared decision-making process.Chiari 2 malformation is best managed using an interprofessional team that includes primary care clinicians, neurosurgeons, radiologists, urologists, orthopedic surgeons, and occasionallyotolaryngologists. Trainednurses, physical therapists, and pharmacists should also assist in caring for patients with Chiari 2 malformation. The patients typically present early in their clinical course because of the associated abnormalities, including myelomeningocele. Utilizing clinical and radiographic findings, the diagnosis can be made. Treatment of Chiari 2 malformation often requires intervention from neurosurgery. Attention must also be given to neonatal feeding difficulties, apnea, respiratory failure, and neurogenic bowel. Rehabilitation is essential to regain mobility and function if there is residual impairment, as well as long-term follow-up to monitor for worsening symptoms. Open communicationamong interprofessional team members is indispensablefor improving outcomes.Review QuestionsChiari Malformation Types 1 and 2 Contributed by C Rowe 1.Sarnat HB. Disorders of segmentation of the neural tube: Chiari malformations. Handb Clin Neurol. 2008;87:89-103. [PubMed: 18809020]2.Fons K, Jnah AJ. 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Effect of Prenatal Repair of Myelomeningocele on Urological Outcomes at School Age. J Urol. 2019 Oct;202(4):812-818. [PubMed: 31075056]24.Piatt JH. Syringomyelia complicating myelomeningocele: review of the evidence. J Neurosurg. 2004 Feb;100(2 Suppl Pediatrics):101-9. [PubMed: 14758937]Disclosure: James Kuhn declares no relevant financial relationships with ineligible companies.Disclosure: Luke Weisbrod declares no relevant financial relationships with ineligible companies.Disclosure: Prabhu Emmady declares no relevant financial relationships with ineligible companies.Chiari malformation is when a part of the brain extends through an opening where the skull meets the spinal canal. It can happen if the skull is too small or misshapen. Depending on the severity, it can lead to neck pain, dizziness, and other neurological symptoms.It can cause pain, trouble swallowing, headache, and more. If the condition does not cause a person any problems, they will not require treatment. However, if it is affecting them, a doctor can recommend treatment options ranging from medication to surgery.Read more to learn about the different types of Chiari malformation, symptoms, how it is diagnosed, treatment options, and more.Share on PinterestLilkin/Getty ImagesChiari malformation is a condition where part of the brain tissue at the base of the skull pushes into the top of the spinal canal. The condition is present from birth. However, not all people with Chiari malformation know that they have it. If they do not have symptoms, it can go unnoticed.According to the American Association of Neurological Surgeons (AANS), slightly less than 1 in every 1,000 people have Chiari malformation. This is 0.1% of the population.There are four different types of Chiari malformation.This is the most common form of the condition. People with type 1 usually experience symptoms in late childhood and adulthood.They may have symptoms including:neck painbalance and coordination problemsdizzinessnumbness However, this type often causes no symptoms at all and is found incidentally later in life. For this reason, it is also called adult Chiari malformation.Type 2 is the second most common form of the condition and is also called classic Chiari malformation. In people with this type, a larger amount of tissue pushes into the spinal canal.It often happens alongside a type of spina bifida called myelomeningocele. This condition affects the spinal membranes and nerves, resulting in the spinal canal and backbone not closing properly. This type causes more severe symptoms than type 1, and it requires surgery. Some symptoms include:difficulty swallowingbreathing changesweakness in the extremitiesType 3 is very rare and happens when a lower part of the brain protrudes through the back of the head or neck. Unfortunately, this type is usually fatal and is diagnosed at birth. People with type 3 who survive infancy may experience:neurological defectsseizuresdevelopmental delaysThis is where the cerebellum, part of the back of the brain, is not fully formed. Type 4 is very rare and may result in parts of the spine and skull being visible.Tragically, children who have this type do not often survive infancy.The cause of Chiari malformation is unknown. It may be caused by a fetal developmental defect in the womb.Sometimes, a person may develop Chiari malformation later in life. This is known as secondary, or acquired, Chiari malformation.Although rate, this form of the condition occurs if a person loses an excess of spinal fluid from the lower back or thorax area. This may be due to injury, illness, or infection.The following are some symptoms of Chiari malformation:headacheneck paindizzinessswallowing or breathing problemsbalance problemsweakness of the musclestinnitus and hearing problemsnauseaexcessive vomiting or droolingscoliosispsychological concerns, including insomnia and depressioncoordination difficultiesHowever, some people with the condition have no symptoms at all. This is especially true of those with type 1.If a person has no symptoms, the condition can be tricky to diagnose. In some cases, it may often only be identified when a person is tested for another health condition. A doctor may see Chiari malformation on an MRI scan and order further tests to confirm a diagnosis.However, sometimes Chiari malformation can be diagnosed before a person is even born. Older research suggests that ultrasound in the second trimester may detect 7090% of abnormalities in fetuses. It is usually not possible to diagnose type 1 at this stage. However, if a fetus has types 2, 3, or 4, it may be visible.A 2019 review states that Chiari malformation is frequently misdiagnosed as another neurological or psychological condition. This is likely because symptoms can be generalized and linked to a wide range of more common disorders.If a persons Chiari malformation does not cause any symptoms, they may never need treatment. However, the condition would require regular monitoring by a doctor.For people with painful headaches and neck pain, a doctor may be able to prescribe some pain management medication. If this does not resolve symptoms, they may recommend surgery.The most common type of surgery to treat Chiari malformation is decompression surgery. This involves a surgeon opening up the base of the skull and removing small pieces of bone either from the skull or the top of the spine. It releases pressure and allows fluid to flow more freely.However, this type of surgery is not always guaranteed to work. It may ease some symptoms, especially headaches. In some cases, it may make symptoms worse or cause no change at all.This is another type of surgery that may help to treat Chiari malformation.During this operation, a surgeon will place a small tube called a shunt. This helps drain excess fluid, relieving pressure in the skull. It deposits the extra fluid into the chest or stomach so the body can safely absorb it.Some conditions frequently occur alongside Chiari malformation. They include:Syringomyelia: This is a condition where a growth-containing fluid grows on the spinal cord. If not treated, this can result in spinal cord damage. Spina bifida: This is where the spinal cord and backbone do not close properly.Hydrocephalus: This condition causes a buildup of fluid that sits in the brain, and it can be fatal if a person does not get treatment.Scoliosis: This condition is where the spine curves abnormally. It usually develops in children and adolescents.Tethered cord syndrome: This is a condition where the spine attaches to and stretches the spinal cord, reducing flexibility. This can result in severe nerve and muscle damage in the lower body.Sleep apnea: In some cases, sleep apnea can occur in people with Chiari malformation. Sleep apnea causes breathing disturbances during sleep, and a 2019 review suggests clinicians should test for this in children with Chiari malformation to allow for early diagnosis and intervention.For individuals with type 1 or 2, symptoms are not usually life threatening. However, some complications can occur and they may require treatment.Unfortunately, types 3 and 4 are often fatal.Chiari malformation is a condition where part of the brain tissue enters the spinal cord.There are four types of this condition. Types 3 and 4 are usually fatal. However, people with types 1 or 2 can go through life with minimal symptoms and may only find out if they have an MRI scan for another condition.Neurology / NeurosciencePediatrics / Children's Health Chiari Malformation Type 2 (also known as Arnold Chiari Malformation): Type 2 involves a herniation of the cerebellar tonsils and lower part of the brainstem (the medulla oblongata). Unlike in Chiari 1.5, in Type 2 the fourth ventricle is usually herniated, all/part of the cerebellar vermis (the tissue connecting both halves of the cerebellum) is missing or herniated, the corpus callosum (nerve fibers connecting both hemispheres of the brain) is fully/partially absent (agenesis), and it is almost always accompanied by a myelomeningocele (the most serious form of Spina Bifida, a congenital neural tube defect where the spinal canal does not close properly).[13][14][15] Diagnosis Requirements: While a myelomeningocele is usually evident and diagnosed at birth, a brain MRI should confirm the radiological aspects of Type 2. Treatment Options: Myelomeningocele is usually treated surgically at birth. If other related problems develop, such as hydrocephalus and/or tethered cord, they are often dealt with surgically as they become problematic. While some with Type 2 are decompressed, anecdotal evidence reflects a general trend of an increased failure rate with decompression surgeries as compared to those with Type 1. Because of this, some neurosurgeons choose not to decompress those with Type 2. Chiari malformations comprise a spectrum of congenital hindbrain abnormalities that affect the structural relationships among the cerebellum, lower brain stem, upper cervical spinal cord, and the foramen magnum within base of the skull (craniocervical junction). The foramen magnum is an oval-shaped opening in the skull base. Normally, only the medulla extends below the foramen magnum, where it merges seamlessly with the cervical spinal cord. In Chiari malformations, the cerebellar tonsils descend more than 5 mm below the level of the foramen magnum (Figure).PathogenesisThere is increasing evidence that Chiari malformations are due to underdevelopment of the posterior cranial fossa, resulting in overcrowding compared with the normally developed hindbrain.1,2 The posterior cranial fossa is the part of the cranial cavity, which contains the cerebellum and lower brainstem (ie, the pons and the medulla). A smaller cranial fossa leaves a typically sized cerebellum, pons, and medulla with less space and overcrowding causes herniation of the cerebellar tonsils below the level of the foramen magnum, distortion of the pons and medulla, and an alteration in the normal spinal fluid dynamics through the foramen magnum.ClassificationThere are 4 recognized types of Chiari malformation according to current classification.Chiari type I malformations (CM-I) are characterized by abnormally shaped cerebellar tonsils displaced more than 5 mm below the level of the foramen magnum. Normally, only the spinal cord descends through the foramen magnum (Figure). CM-I is the most commonly encountered Chiari malformation. The prevalence of CM-I in the US is approximately 0.1% to 0.5%, with the number of new cases diagnosed each year ranging from 5.5 to 78 cases per 100,000 people.3,4 CM-I has a slight female predominance of 1.3:1.Chiari type II malformations (CM-II), also known as Arnold-Chiari malformations, are characterized by downward displacement of the cerebellar tonsils and vermis as well as the medulla more than 5 mm below the level of the foramen magnum, a beaked appearance of the tectum on imaging studies, and a spinal myelomeningocele. Hydrocephalus is commonly present.Chiari type III malformations (CM-III) are the most rare. These malformations combine a small posterior fossa with a high cervical or occipital encephalocele. CM-III is often associated with displacement of the cerebellar structures into the encephalocele with inferior displacement of the brain stem into the spinal canal.Chiari type IV malformations (CM-IV) are characterized by cerebellar hypoplasia unrelated to other Chiari malformations.Proposed New ClassificationsTwo new types of Chiari malformation have been proposed. Chiari type 0 is characterized by normal position of the cerebellar tonsils and the presence of a spinal cord syrinx with signs and symptoms of CM-I.5,6 Chiari type 1.5 is characterized by increased cerebellar tonsillar descent with brainstem involvement.5,7It is generally accepted that extension of the tonsils up to 3 mm below the foramen magnum is considered normal, extension between 3 and 5 mm below the foramen magnum is considered borderline, and extension beyond 5 mm is clearly pathologic.8 Hence, the diagnosis of CM-I is made by imaging evidence of cerebellar tonsillar herniation more than 5 mm below the level of the foramen magnum. The 5 mm criteria has been challenged as being too nonspecific considering that many individuals who meet this criteria on MRI but are asymptomatic, whereas some individuals who do not meet the criteria have symptoms associated with abnormal spinal fluid flow and crowding of structures at the level of the foramen magnum.9Clinical PresentationPeople with CM-I are classified as being asymptomatic or symptomatic. Approximately 15% to 30% of people with CM-I are asymptomatic at the time of diagnosis.4,10-13 The asymptomatic group includes people who are oligosymptomatic with symptoms that are mild and nonspecific. It is difficult to classify malformation in individuals who are oligosymptomatic because their symptoms may be attributed to various etiologies other than CM-I.14 Several studies have analyzed the natural history of asymptomatic CM-I to determine how often people who were asymptomatic ultimately developed symptoms. Most people with symptomatic CM-I remain stable for years, whereas others deteriorate or, rarely, improve.4,15-18 From a clinical perspective, CM-I present most frequently in early childhood between age 8 and 9 years or in adults between age 41 and 46.19Clinical manifestations develop as a result of 3 pathologic consequences of the distorted hindbrain anatomy: 1) compression of the cerebellum; 2) compression of the of the brainstem and upper cervical spinal cord; and 3) abnormal cerebrospinal fluid (CSF) flow around and through the foramen magnum. The presenting neurologic findings of CM-I can be divided into 5 separate syndromes as follows, all explained by these 3 pathologic consequences.20-25Spinal cord syndrome occurs in 65% to 84% of cases and presents with motor weakness and sensory changes. Brainstem syndrome is seen in 22% to 38% of cases and presents with blurred vision, horizontal or downbeating nystagmus, and diplopia when the upper brainstem is affected and with hoarseness, dysphonia, dysarthria, dysphasia, vocal cord paralysis, palatal weakness, and tongue atrophy when the lower brainstem is involved. Cerebellar syndrome presentations occur in 11% of cases with incoordination, ataxia, and dysmetria. Cortical syndrome is rare, occurring in less than 3% of cases and presenting with brain fog, depression, generalized weakness, and fatigue. Even more rare is systemic syndrome in less than 2% of cases, presenting with chest pain, shortness of breath, pickups, postural hypertension, and syncope.The presenting signs and symptoms in CM-I are quite nonspecific.9,26 In a study of 244 adults and 74 children with CM-I, 82% to 90% had suboccipital headache, 78% to 81% had posterior neck pain, 60% to 67% had dizziness, and 49% and 48% had nausea, respectively.27 The presenting symptoms and signs in a case series of 71 people are shown in Table 1.28In summary, the most frequent signs and symptoms of CM-I include headaches with neck pain, dizziness, weakness, sensory changes, and hyperreflexia. The majority of signs and symptoms may be explained by hindbrain compression, although the headaches and neck pain are due to altered CSF dynamics.HeadachesPain is the most frequently reported presenting symptom of CM-I but is also among the most frequently encountered complaints in clinical practice. This overlap makes it difficult to evaluate whether a headache is a symptom of CM-I, secondary to another clinical problem, or a headache disorder (eg, tension-type headache or migraine). This is not a trivial dilemma, considering that persons with symptomatic CM-I are often referred for surgical intervention, whereas people with asymptomatic CM-I are often followed conservatively. Unfortunately, the frequency of migraine, tension-type headache, or chronic daily headache in population studies is similar to that of people with CM-I. In short, the presence of headaches in persons with significant tonsillar herniation does not automatically imply they have symptomatic CM-I.Cough headaches, triggered by coughing, laughing, sneezing, or any Valsalva maneuver, often play an important role in the diagnosis of CM-I. Cough headaches, by definition, have a mean duration of 1 minute within a range of 3 seconds to 4 minutes. Cough headaches are classified as primary, which are not associated with intracranial disorders, or secondary. Approximately 40% of cough headaches are secondary, and most of these patients have CM-I. Other reported causes of secondary cough headache include spontaneous intracranial hypotension, carotid or vertebrobasilar diseases, middle or posterior fossa tumors, midbrain cysts, basilar impression, platybasia, subdural hematoma, cerebral aneurysms, and reversible cerebral vasoconstriction syndrome. Secondary cough headaches are due to transient impaction of the cerebellum tonsils below the foramen magnum that causes compression of the first and second cervical nerve roots by the caudally displaced cerebellar tonsils.26,29,30In a study of 40 individuals with secondary cough headaches, 32 (80%) had radiographic evidence of CM-I.31 Another study of 34 people with CM-I, showed 59% complained of headaches, 34% had cough headaches, 41% had chronic daily headaches lasting from 3 hours to several days, and 24% had continuous headaches. The authors reported 12% of the participants met the diagnostic criteria for tension-type headache and 10% met the diagnostic criteria for migraine.32 Although the majority of people with cough headaches have CM-I, only a minority of people with CM-I have cough headaches. Ideally, the optimal way to understand the signs and symptoms associated with CM-I would be to correlate the findings to the degree of tonsillar herniation. Unfortunately, convincing evidence is conflicting. The degree of tonsillar herniation correlated with the presence of cough headache in only 1 of the case series discussed.Recently, the 3rd edition of the International Classification of Headache Disorders (ICHD-3) proposed diagnostic criteria for headaches due to CM-I and MRI criteria for the diagnosis of CM-I (Table 2).33 To help determine whether headaches in a person with cerebellar tonsillar herniation more than 5 mm below the foramen magnum are due to CM-I, advanced motion-sensitive MRI imaging techniques are necessary.Diagnostic Neuroimaging: Phase Contrast Flow MRIRoutine MRI studies can identify abnormal descent of the cerebellar tonsils below the foramen magnum, but do not provide information on CSF flow. Phase-contrast cine flow MRI is the imaging technique used to analyze CSF circulation at the level of the foramen magnum. Normally, during the cardiac cycle, there is bidirectional flow of CSF. During systole, there is an increase in the cerebral blood volume with displacement of the CSF caudally through the foramen magnum as a compensatory mechanism. During diastole, the elastic recoil of the dura propels the CSF in a caudal-cranial direction towards the intracranial compartment. In brief, CSF flows down through the foramen magnum on systole and up through the foramen magnum during diastole. Systolic CSF flow follows the dorsal subarachnoid space (ie, downward flow behind the brainstem), whereas diastolic CSF flow follows the ventral subarachnoid space (ie, upward flow in front of the brainstem). A movie-like cine MRI can determine if, and by how much, a Chiari malformation is blocking this typical back-and-forth flow of CSF between the brain and spine.Several studies have stressed the significance of altered MRI phase contrast cine flow at the level of the foramen magnum as a diagnostic tool for CM-I.34-36 A study has shown that the symptomatic presentation of CM-I correlates with both the absence of the normal CSF spaces at and below the foramen magnum and altered CSF dynamics at the craniocervical junction.13 Individuals with CM-I have significant elevations of peak systolic CSF velocity in the foramen magnum. In contrast, regular ebb and flow of CSF with relatively small variations in velocity throughout the subarachnoid space is evident in people without CM-I.37 In those with CM-I, jets of high velocity CSF were noted in some regions with little or no flow in other regions. CSF flow was also observed in a cephalad direction in a portion of the subarachnoid space and in a craniad direction at other times during the cardiac cycle. These spatial and temporal variations in flow suggested abnormal CSF pressure differentials and shear stresses with CM-I patients. In people with MRI evidence of significant tonsillar herniation who present solely with headaches, cine flow MRI studies have 2 significant benefits. First, cine flow MRI can determine whether the persons headaches are related to the Chiari malformation, and second, it can provide a noninvasive method for serial followup of people with asymptomatic CM-I.SummaryCM-I, the most common type of Chiari malformation, is characterized by cerebellar tonsillar herniation more than 5 mm below the level of the foramen magnum. The presenting signs and symptoms of CM-I are due to compression of the cerebellum, brainstem, and upper cervical spinal cord and disruption of the normal CSF flow around and through the foramen magnum. A significant minority of persons with CM-I are asymptomatic at the time of diagnosis. Those who are symptomatic typically complain of headaches, as well as neck pain, dizziness, and nausea. Unfortunately, the incidence of migraine, tension-type, and chronic daily headaches in the general population is similar to the incidence in CM-I. In a person with MRI evidence of significant tonsillar herniation, assessing whether migrainous, tension-type, or chronic daily headaches are symptomatic caused by CM-I requires MRI phase contrast cine flow studies. A Chiari mal