Chiari Malformation Decompression Surgery

Number: 0931

Policy

*Please see amendment for Pennsylvania Medicaid at the end of this CPB.

Aetna considers Chiari malformation decompression surgery medically necessary for Chiari malformation type II, III and IV.

Aetna considers Chiari malformation decompression surgery medically necessary for Chiari malformation type I (CMI) when members have radiographic findings of downward displacement of the cerebellar tonsils by greater than or equal to 5 mm (3 mm in children) with or without radiographic findings of a spinal cord syrinx and either of the following:

I. Presence of signs or symptoms that are related to the Chiari malformation given that:

   A. All other reasonable sources of signs or symptoms have been ruled out; and

   B. Symptoms have failed conservative treatment (e.g., analgesics, muscle relaxants, and/or soft collar; unless neurologic signs are severe or rapidly progressive); and

   C. If symptoms are isolated to headaches/neck pain, conservative treatment should be directed by a neurologist and failure of conservative treatment should be documented by a
neurologist. (if there is no local neurologist or pediatric
neurologist, a pain specialist is acceptable); and
D. If symptoms of sleep apnea or swallowing difficulty (with or
without headaches) are the basis for surgical consideration,
appropriate objective confirmatory testing should be
documented (e.g., sleep study and/or video swallow
evaluation).

II. Documented radiographic progression of either the downward
displacement of the cerebellar tonsils or of an associated spinal
cord syrinx.

Background

Chiari malformations (CMs) types I to IV are a heterogeneous group of
disorders that refer to a spectrum of congenital hind-brain abnormalities
affecting the structural relationships between the cerebellum, brain-stem,
the upper cervical cord, and the bony cranial base. The 4 types of CMs
are classified as follows: (Pakzaban, 2017; Khoury, 2018)

- CMI is characterized by abnormally shaped cerebellar tonsils that
  are displaced below the level of the foramen magnum.
- CMII (also known as Arnold-Chiari malformation) is characterized
  by downward displacement of the cerebellar vermis and tonsils, a
  brain-stem malformation with beaked mid-brain on neuroimaging,
  and a spinal myelomeningocele.
- CMIII is rare and combines a small posterior fossa with a high
  cervical or occipital encephalocele, usually with displacement of
  cerebellar structures into the encephalocele, and often with
  inferior displacement of the brain-stem into the spinal canal.
- CMIV is now considered to be an obsolete term that describes
  cerebellar hypoplasia unrelated to the other CMs.

The treatment of CMs depends upon the nature of the malformation and
the degree of associated neurological impairments. Asymptomatic
patients with an incidental diagnosis of CMI who do not have
syringomyelia can be managed conservatively with clinical and magnetic
resonance imaging (MRI) surveillance. For patients with CMI who are clearly symptomatic with lower cranial nerve palsies, syringomyelia, myelopathy, cerebellar symptoms, severe neck pain or occipital headache related to the CM, surgical decompression of the foramen magnum is recommended. For CMII and CMIII, surgical interventions may include closure of open neural tube defects shortly after birth, treatment for hydrocephalus (most often by use of a shunt), and decompression of tight posterior fossa structures. Medical issues involve management of neurogenic bowel and bladder, neonatal feeding difficulties, respiratory failure, and apnea. The objectives of surgery for CMs are to decompress the cranio-cervical junction (CCJ) and restore the normal flow of cerebrospinal fluid (CSF) in the region of the foramen magnum. The most common procedure is posterior decompression via sub-occipital craniectomy with or without duraplasty. Other procedures included anterior decompression of the foramen magnum by odontoidectomy and shunting (Khoury, 2018).

Milhorat and associates (1999) noted that CMI is defined as tonsillar herniation of at least 3 to 5 mm below the foramen magnum. These investigators examined a prospective cohort of 364 symptomatic patients. All patients underwent MRI of the head and spine, and some were evaluated using CINE-MRI and other neuro-diagnostic tests. For 50 patients and 50 age- and gender-matched control subjects, the volume of the posterior cranial fossa was calculated by the Cavalieri method. The families of 21 patients participated in a study of familial aggregation. There were 275 female and 89 male patients. The age of onset was 24.9 +/- 15.8 years (mean +/- standard deviation), and 89 patients (24 %) cited trauma as the precipitating event. Common associated problems included syringomyelia (65 %), scoliosis (42 %), and basilar invagination (12 %); 43 patients (12 %) reported positive family histories of CMI or syringomyelia. Pedigrees for 21 families showed patterns consistent with autosomal dominant or recessive inheritance. The clinical syndrome of CMI was found to consist of the following: headaches, pseudotumor-like episodes, a Meniere's disease-like syndrome, lower cranial nerve signs, and spinal cord disturbances in the absence of syringomyelia. The most consistent MRI findings were obliteration of the retro-cerebellar CSF spaces (364 patients), tonsillar herniation of at least 5 mm (332 patients), and varying degrees of cranial base dysplasia. Volumetric calculations for the posterior cranial fossa revealed a significant reduction of total
volume (mean of 13.4 ml) and a 40 % reduction of CSF volume (mean of 10.8 ml), with normal brain volume. The authors concluded that these data supported accumulating evidence that CMI is a disorder of the paraxial mesoderm that is characterized by under-development of the posterior cranial fossa and over-crowding of the normally developed hindbrain. Tonsillar herniation of less than 5 mm does not exclude the diagnosis. Clinical manifestations of CMI appeared to be related to CSF disturbances (which are responsible for headaches, pseudotumor-like episodes, endolymphatic hydrops, syringomyelia, and hydrocephalus) and direct compression of nervous tissue. The demonstration of familial aggregation suggested a genetic component of transmission.

Meadows and colleagues (2000) retrospectively reviewed the records of all brain magnetic resonance (MR) images obtained at their hospital over a 43-month period to ascertain the relative prevalence and MRI characteristics of asymptomatic CMIs. Of 22,591 patients who underwent MRI of the head and cervical spine, 175 were found to have CMIs with tonsillar herniation extending more than 5 mm below the foramen magnum. Of these, 25 (14 %) were found to be clinically asymptomatic. The average extent of ectopia in this population was 11.4 +/- 4.86 mm, and was significantly associated with a smaller cisterna magna. Syringomyelia and osseous anomalies were found in only 1 asymptomatic patient. The authors suggested that the isolated finding of tonsillar herniation is of limited prognostic utility and must be considered in the context of all available clinical and radiographic data.

Strahle and co-workers (2011) noted that CMI with an associated spinal syrinx is a common pediatric diagnosis. A better understanding of the relative age-related prevalence and MRI characteristics of these associated conditions may lead to improved treatment decisions. These investigators performed a retrospective review of 14,116 consecutive individuals 18 years of age or younger who had undergone brain or cervical spine MRI at the University of Michigan between November 1997 and August 2008. In the patients with CMI, demographic, clinical, and radiographic information was recorded. A total of 509 children (3.6 %) with CMI were identified. Among these patients, 23 % also had a spinal cord syrinx, and 86 % of the syringes were found in the cervical spine. The MRI prevalence of CMI with a syrinx was 1.2 % in girls and 0.5 % in boys (p < 0.0001). The severity of impaired CSF flow at the foramen
magnum was associated with the amount of tonsillar herniation \( (p < 0.0001) \) and conformation of the tonsils \( (p < 0.0001) \). Patients with CMI were treated surgically in 35% of cases; these patients exhibited more severe tonsillar herniation \( (p < 0.0001) \) and impaired CSF flow \( (p < 0.0001) \) as compared with those who did not undergo surgery. On imaging, 32% of all the patients with CMI were considered symptomatic by the treating physician. Patients were more likely to be considered symptomatic if they were female, had a syrinx, displayed abnormal tonsillar pulsations, or had a greater amount of tonsillar herniation. The authors described the age-related prevalence and MRI characteristics of CMI and its association with a syrinx and other abnormalities in a large group of children who underwent MRI for any indication. Syringes were more common in older children, in girls, and in patients with a greater degree of tonsillar descent and CSF flow impairment.

de Oliveira Sousa and associates (2018) stated that CM and basilar invagination are mesodermal malformations with disproportion between the content and volume of posterior fossa capacity and over-crowding of neural structures at the level of foramen magnum. Several alternatives for posterior fossa decompression (PFD) are available, including extradural (ED), extra-arachnoidal (EA), and intra-dural (ID) approaches. These investigators evaluated the effect of several surgical techniques for posterior fossa decompression in the outcomes of patients with CCJ malformations and evaluated complications in the techniques reported. They performed a systematic review on the effects of posterior fossa decompression using the Medline database and the Cochrane Central Register of Controlled Trials. The PRISMA statement and MOOSE recommendations were followed. A total of 539 articles were initially selected by publication title. After abstract analysis, 70 articles were selected for full-text analysis, and 43 were excluded. Ultimately, 27 studies were evaluated. The success rate (SR) with ED techniques was 0.76 versus 0.81 in EA technique and 0.83 in IA technique. The authors concluded that all PFD techniques were very successful. Results from observational studies were similar to that of the randomized trial. The main complications were CSF fistulas, most common in patients with IA approach. The overall mortality rate was 1%.
Singhal and co-workers (2018) noted that in 2003, pediatric neurosurgeons were surveyed under the auspices of the education committee of the International Society for Pediatric Neurosurgery (ISPN) to determine opinions regarding the management of CMI with and without associated syringomyelia. In the ensuing years, there has been further information from multiple CMI studies, with regards to indications, success rates of different surgical interventions, and complications. These investigators re-evaluated current opinions and practices in pediatric CMI. Pediatric neurosurgeons worldwide were surveyed, using an e-mail list provided by the ISPN communication committee chairperson. Respondents were given scenarios similar to the 2003 CMI survey in order to determine opinions regarding whether to surgically intervene, and if so, with which operations. Of 300 surveys electronically distributed, 122 responses were received (40.6% response rate) -- an improvement over the 30.8% response rate in 2003. Pediatric neurosurgeons from 34 different countries responded. There was broad consensus that non-operative management is appropriate in asymptomatic CMI (greater than 90%) as well as asymptomatic CMI with a small syrinx (greater than 65%). With a large syrinx, a majority (almost 80%) recommended surgical intervention. Scoliotic patients with CMI were generally offered surgery only when there was a large syrinx. There has been a shift in the surgical management over the past 10 years, with a bone-only decompression now being offered more commonly. There remains, however, great variability in the operation offered. The authors concluded that this survey, with a relatively strong response rate, and with broad geographic representation, summarized current world-wide expert opinion regarding management of pediatric CMI. They stated that asymptomatic CMI and CMI with a small syrinx are generally managed non-operatively. When an operation is indicated, there has been a shift towards less invasive surgical approaches.

Posterior Fossa Decompression With or Without Duraplasty

The role of duraplasty in posterior fossa decompression for the treatment of patients with CMI remain controversial. Zhao and colleagues (2017) performed a literature search using PubMed, CINAHL/Ovid, Cochrane library, and the Elsevier database to examine the most effective treatments of CMI. The key words "Chiari I malformation", "Chiari malformation type I", "surgery," and "treatment" were used for the search.
Articles had to be peer-reviewed and provided primary outcomes measured by clinical and radiographic outcomes after surgical treatments. Exclusion criteria included non-English-language articles, case reports, commentaries, information from textbooks and expert opinions, and articles that did not provide outcomes concerning specific surgical methods. Patients included were classified into 4 groups according the procedure: only bony decompression but not duraplasty (group I), bony decompression plus duraplasty (group II), bony decompression plus the resection of tonsils (group III), and shunt (group IV). A total of 18 studies were identified. Groups II and III had a significantly higher improvement rate (82.25 %, 86.10 %, p < 0.05) of outcomes with regard to clinical signs and symptoms than the other groups. Group IV showed a statistically higher rate (30.49 %, p < 0.05) of aggregating clinical signs and symptoms. In patients with syringomyelia, group III showed better clinical improvement (96.08 %). Group II displayed a significantly higher rate of decrease in the size of cavities (83.33 %, p < 0.05). Group IV had a statistically higher rate of increase in the size of cavities (33.87 %, p < 0.05). The authors concluded that only bony decompression could not achieve satisfactory outcomes. Bony decompression plus duraplasty showed the most favorable outcomes. They stated that resection of tonsils was not recommended because of the high rate of side effects; shunt may aggregate clinical signs and symptoms and increase the size of cavities.

Sabba and associates (2017) noted that CM is the most common and prevalent symptomatic congenital cranio-cervical malformation. Radiological diagnosis is established when the cerebellar tonsils are located 5 mm or more below the level of the foramen magnum on MRI. Surgical treatment is indicated whenever there is symptomatic tonsillar herniation or syringomyelia/hydrocephalus. The main surgical treatment for CM without cranio-cervical instability (such as atlanto-axial luxation) is posterior fossa decompression, with or without duraplasty. The authors described in details and in a step-wise fashion the surgical approach of patients with CM as performed at the State University of Campinas, emphasizing technical nuances for minimizing the risks of the procedure and potentially improving patient outcome.
Pakzaban (2017) stated that the main controversy regarding the treatment of CMI centers around the question of what surgical steps are necessary to achieve decompression of the cervico-medullary junction and restore CSF flow across foramen magnum. The most widely accepted approach consists of limited sub-occipital craniectomy, C1 laminectomy, duraplasty and arachnoidal dissection. On the other side of the spectrum are recommendations for bony decompression without opening the dura. Most surgeons who recommend this approach resect or incise the thick fibrous band that constricts the dura at foramen magnum without opening the dura. Some recommended serial incisions of the outer layer of the posterior fossa dura, with the expectation that this may expand the posterior fossa volume. Others recommended opening the dura, but not the arachnoid, and then performing a duraplasty. Still others recommended keeping the arachnoid closed but leaving the dura open, without duraplasty. Some recommended performing an occipito-cervical fusion at the time of decompression, citing up to 19% rates of cranio-cervical instability in pediatric patients after Chiari decompression. Those who advocated the more conservative extra-dural and extra-arachnoidal approaches seek to avoid CSF-related complications (CSF leak, pseudo-meningocele, aseptic meningitis), particularly in the pediatric population. However, other studies indicated that the incidence of such complications was too low to justify a positionally suboptimal extra-dural decompression. The author concluded that the neurosurgery literature is replete with studies that support all of these approaches, but, unfortunately, most of the studies provide only class III or limited class II evidence; prospective controlled trials are needed to compare the various techniques in adult and pediatric patients and in those with and without syringomyelia.

Xu and co-workers (2017) stated that the treatment of CMI with posterior fossa decompression without (PFD) or with duraplasty (PFDD) is controversial. In a systematic review and meta-analysis, these researchers compared the clinical outcomes (operation time, clinical improvement, and complications) between these 2 methods for the treatment of CMI. Randomized controlled trials (RCTs) or non-RCTs of PFD and PFDD were considered for inclusion. A total of 12 published reports of eligible studies involving 841 subjects met the inclusion criteria. There was significant difference in the operative time [mean difference [MD] = -74.63, 95% confidence interval [CI]: -83.02 to -66.25,
p < 0.05] in favor of PFD compared with PFDD. There was significant difference in overall complication rates [MD = 0.34, 95% CI: 0.19 to 0.60, p < 0.05] and rates of CSF leak [MD = 0.24, 95% CI: 0.07 to 0.78, p < 0.05] in favor of PFD groups. However, there was significant difference in the clinical improvement rate in favor of the PFDD group [MD = 0.85, 95% CI: 0.73 to 0.99, p < 0.05]. The authors concluded that although PFDD was related with longer operation time and higher CSF leak rate, it could still be considered as a preferable therapeutic option for most CMI patients for its higher improvement rate. They stated that more evidence from advanced, multi-center studies are needed to provide illumination for the surgical decision-making of CMI.

Li and associates (2017) compared the efficacy between PFD (n = 22) and PFDD (n = 34) in the surgical management for adult CMI. The clinical results were retrospectively analyzed to compare the efficacy of these 2 approaches. There was no death or severe neurological dysfunction case in 57 patients of the 2 groups. Patients undergoing PFD had shorter length of hospital stay (13.7 ± 3.5 days versus 16.2±4.1 days, p < 0.05); and surgical time (98.7 ± 22.1 mins versus 132.3 ± 39.6 mins, p < 0.05); CSF-related complications and intra-cranial infection were more common in patients undergoing PFDD (0/23 [0 %] versus 8/34, [23.5 %], p < 0.05). Clinical improvement was comparable in 2 groups (15/23, [65.2 %] versus 26/34, [76.5 %], p > 0.05) at the 1-year follow-up. The rate of syrinx regression in patients with syringomyelia was higher in patients undergoing PFDD (3/12, [25 %] versus 17/22, [77.3 %], p < 0.05). The authors concluded that for adult patients with CMI, PFD had the advantages of simple manipulation, short length of hospital stay and low incidence of CSF-related complications and intra-cranial infection, compared with PFDD; PFD was comparable to PFDD in clinical improvement, but the effect of PFD was not as good as that of PFDD in the aspect of syrinx regression.

Lu and colleagues (2017) noted that surgery is the definitive treatment of CMI. It entails PFD, which can be performed along with C1 laminectomy, reconstructive duraplasty, or tonsil shrinkage. These investigators provided an updated systematic review and meta-analysis of the latest available evidence regarding PFD versus PFDD in the treatment of CMI in children. These researchers carried out a literature search in compliance with the PRISMA (Preferred Reporting Items for Systematic
Reviews and Meta-Analyses) guidelines for article identification, screening, eligibility, and inclusion. Relevant articles were identified from 6 electronic databases from their inception to April 2016. These articles were screened against established criteria for inclusion into this study. From 12 relevant studies identified, 1,492 pediatric patients treated via PFDD were compared with 1,963 pediatric patients treated by PFD for CMI. PFDD was associated with greater overall clinical improvement ($p = 0.0009$), along with longer length of stay ($p < 0.0001$) and more post-operative complications ($p = 0.0001$) compared with PFD. No difference was observed between PFDD and PFD in terms of revision surgery incidence ($p = 0.13$), estimated blood loss ($p = 0.14$), syrinx improvement ($p = 0.09$), or scoliosis improvement ($p = 0.95$). The authors concluded that it appeared that the addition of duraplasty to PFD in the definitive treatment of CMI in children may alter surgical and performance outcomes. In particular, parameters of overall clinical improvement, length of stay, and post-operative complication may differ between children undergoing PFDD and those undergoing PFD. Moreover, they stated that current evidence in the literature is of low-to-very low quality that, as of yet, has not been able to completely control for inherent selection bias both in study design and surgeon preference. They stated that future, large prospective registries and RCTs are needed to validate the findings of this study.

Giammattei and co-workers (2018) described the surgical technique, indications and limits of surgical treatment for the treatment of patients with CMI. These investigators described the surgical technique, including: posterior fossa decompression, opening of the foramen of Magendie and duraplasty in case of CMI. The authors concluded that PFDD is a safe and effective procedure for patients with CMI.

In a prospective study, Jiang and associates (2018) compared the radiographic and clinical outcomes between PFD and PFDD in adolescent patients with CMI. A total of 90 adolescent patients with CMI were randomly assigned to undergo either PFDD or PFD. In both groups, a dissection from the occipital bone was performed. The dura was not opened in the PFD group, and the outer layer of dura was resected. However, in the PFDD group, the dura mater was opened and expanded. Data were analyzed for clinical outcome, complications, and syrinx resolution. The age, gender, and pre-operative neurologic status were
similar between the 2 groups. Compared with the PFD group, patients undergoing PFDD had significantly longer operation time, longer post-operative drainage time, and higher drainage volume. At the latest follow-up, no statistically significant difference was found between the 2 groups in terms of syrinx resolution. The clinical outcomes were similar in the PFDD and PFD group. Compared with the PFD group, patients in the PFDD group had a higher incidence of CSF leak. The authors concluded that compared with the more aggressive decompression with duraplasty, PFD without duraplasty produced comparable radiologic and clinical outcomes and was associated with a lower risk of complications.

Lin and co-workers (2018) performed a systematic review and meta-analysis of studies to evaluate the effect on clinical and imaging improvement, operative time, complications, and recurrence rate between PFD and PFDD in patients with CMI. These investigators systematically searched PubMed, Embase, Cochrane, Web of Knowledge, and ClinicalTrials.gov for retrospective or prospective studies comparing PFD with PFDD. The main end-points were clinical and imaging improvement, operative time, complications, and recurrence rate. They assessed pooled data by the use of a fixed-effects or random-effects model according to the between-study heterogeneity. Of 214 identified studies, 13 were eligible and were included in the analysis (n = 3,481 patients). Compared with PFD, PFDD led to a mean greater increase in operative time than did PFD (SMD, -2.35; 95 % CI: -2.70 to -1.99), a higher likelihood of clinical improvement in patients with syringomyelia (relative risk [RR], 0.70; 95 % CI: 0.49 to 0.98), no increased RR of clinical improvement in patients without syringomyelia, no increased RR of imaging improvement, but an increased RR of CSF-related complications (RR, 0.29; 95 % CI: 0.15 to 0.58), CSF leak, aseptic meningitis, pseudomeningocele, and a decreased likelihood of recurrence rate. The authors concluded that PFDD can be an optimal surgical strategy because of its higher clinical improvement and lower recurrence rate in the patients with syringomyelia. In patients without syringomyelia, PFD can be a preferred choice because of its similar clinical improvement and lower costs. Moreover, they stated that future randomized studies with large numbers and the power to provide illumination for surgical decision making in CMI are needed.
Grahovac and associates (2018) retrospectively reviewed 16 patients with CMI who were diagnosed and surgically treated between 2007 and 2014 during the first 3 years of life with minimum follow-up of 3 years. These investigators focused on the presenting symptoms, MRI findings, and surgical techniques used for posterior fossa decompression and their post-operative outcome. A total of 12 patients (75%) presented with signs of headaches such as irritability, inconsolable crying, head grabbing, and/or arching back; 10 patients (62.5%) presented with oropharyngeal and/or respiratory symptoms such as emesis, choking, gagging, snoring, sleep apnea, breathing pause, and/or vocal cord palsy. Only 1e patient had segmental cervical hydromyelia. At the first surgery, 10 patients had PFD with dural scoring (Type 1 procedure), while 6 others had PFDD (Type 2 procedure) with thermal reduction of the cerebellar tonsils in 4. Following the first operation, all initially had varying degrees of symptomatic improvement; however, 7 patients subsequently had symptomatic recurrence. Persistent crowding at the PFD site on the post-operative imaging indicated greater risk of recurrences in both Type 1 procedure and Type 2 procedure groups. Of 7 patients who needed a 2nd operation, 5 were after Type 1 procedure and 2 were after Type 2 procedure. The difference of recurrence rates between these 2 groups was not significant; CSF-related complications occurred in 4 out of 11 patients who had Type 2 procedure (1 after primary decompression and 3 after the 2nd decompression for recurrence). The authors concluded that young patients lacking effective verbal communication often present their CMI differently from older age groups. Behavioral changes indicative of headaches/irritability and oropharyngeal/respiratory symptoms were the primary presenting symptoms. The recurrence rate tended to be higher among the patients after Type 1 procedure (particularly those younger than 18 months) than after Type 2 procedure. These investigators observed that duraplasty at primary or at redo posterior fossa decompression provided for better decompression and long-term outcome. However, it should be noted that there is risk of CSF-related complications following duraplasty, particularly higher tendency after redo posterior fossa decompression.

Chai and colleagues (2018) compared the safety and effectiveness and safety of PFDD and PFD in treating patients with CMI. PubMed, Embase, and Cochrane Library were searched through May 2017. A total of 14 cohort studies comprising 3,666 patients with CMI were included. Studies
were pooled, and the RR and corresponding 95 % CI were calculated. The decrease in syringomyelia was better in patients in the PFDD group than in patients in the PFD group (RR = 1.57, 95 % CI: 1.07 to 2.32, heterogeneity, p = 0.042, I² = 56.6 %). The incidence of CSF leak (RR = 5.23, 95 % CI: 2.61 to 10.51, heterogeneity, p = 0.830, I² = 0 %) and aseptic meningitis (RR = 4.02, 95 % CI: 1.46 to 11.03, heterogeneity, p = 0.960, I² = 0 %) significantly increased among patients in the PFDD group compared with patients in the PFD group. When stratifying by age, a significantly reduced risk in the re-operation rate was observed in the adult group. However, the clinical improvement and the incidence of wound infection were not significantly different between the 2 groups. The authors concluded that the findings of this study confirmed that the decrease in syringomyelia was better for patients treated with PFDD than for patients treated with PFD alone. However, no significant difference was found in the clinical improvement and the re-operation rate between the 2 groups.

An UpToDate review on “Chiari malformations” (Khoury, 2018) states that “Given the absence of high-quality comparative studies, the utility of duraplasty for CMI is uncertain, and more rigorous studies are needed to resolve this issue”.

Furthermore, there is a clinical trial on “Posterior Fossa Decompression with or without Duraplasty for Chiari Type I Malformation with Syringomyelia” that is currently recruiting subjects (last updated March 15, 2018).

Leon et al (2019) noted that there are sparse published data on the natural history of "benign" CM-I (i.e., Chiari with minimal or no symptoms at presentation and no imaging evidence of syrinx, hydrocephalus, or spinal cord signal abnormality). These investigators reviewed a large cohort of children with benign CM-I and examined if these children became symptomatic and required surgical treatment. Patients were identified from institutional out-patient records using International Classification of Diseases, 9th Revision, diagnosis codes for CM-I from 1996 to 2016. After review of the medical records, patients were excluded if they (i) did not have a diagnosis of CM-I, (ii) were not evaluated by a neurosurgeon, (iii) had previously undergone PFD, or
(iv) had imaging evidence of syringomyelia at their first appointment. To include only patients with benign Chiari (without syrinx or classic Chiari symptoms that could prompt immediate intervention), any patient who underwent decompression within 9 months of initial evaluation was excluded. After a detailed chart review, patients were excluded if they had classical Chiari malformation symptoms at presentation. The authors then determined what changes in the clinical picture prompted surgical treatment. Patients were excluded from the multi-variate logistic regression analysis if they had missing data such as race and insurance; however, these patients were included in the overall survival analysis. A total of 427 patients were included for analysis with a median follow-up duration of 25.5 months (range of 0.17 to 179.1 months) after initial evaluation; 15 patients had surgery at a median time of 21.0 months (range of 11.3 to 139.3 months) after initial evaluation. The most common indications for surgery were tussive headache in 5 (33.3 %), syringomyelia in 5 (33.3 %), and non-tussive headache in 5 (33.3 %). Using the Kaplan-Meier method, rate of freedom from PFD was 95.8 %, 94.1 %, and 93.1 % at 3, 5, and 10 years, respectively. The authors concluded that among a large cohort of patients with benign CM-I, progression of imaging abnormalities or symptoms that warrant surgical treatment is infrequent, thus, these patients should be managed conservatively. However, clinical follow-up of such individuals is justified, as there is a low, but non-zero, rate of new symptom or syringomyelia development. Future analyses will determine whether imaging or clinical features present at initial evaluation are associated with progression and future need for treatment.

Furthermore, an UpToDate review on “Chiari malformations” (Khoury, 2019) states that "Decompressive surgery is indicated for patients with CM-I who are clearly symptomatic with lower cranial nerve palsies, syringomyelia, myelopathy, cerebellar symptoms, severe neck pain, or occipital headache".

Shen and colleagues (2019) summarized the clinical features, diagnosis, and treatment of CMI-associated syringobulbia. These investigators performed a literature review of CMI-associated syringobulbia in PubMed, Ovid Medline, and Web of Science databases. Their concerns were the clinical features, radiologic presentations, treatment therapies, and
prognoses of CMI-associated syringobulbia. This review identified 23 articles with 53 cases. Symptoms included headache, neck pain, cranial nerve palsy, limb weakness/dysesthesia, Horner syndrome, ataxia, and respiratory disorders. The most frequently involved area was the medulla. Most of the patients also had syringomyelia. Surgical procedures performed included PFD, foramen magnum decompression (FMD), cervical laminectomy, duraplasty, and syringobulbic cavity shunt. Most patients experienced symptom alleviation or resolution post-operatively. A syringobulbic cavity shunt provided good results in refractory cases. Physicians should be aware of the possibility of syringobulbia in CMI patients, especially those with symptoms of sudden-onset brain-stem involvement. The diagnosis relies on the disorder's specific symptomatology and MRI. The authors concluded that the findings of this review suggested that the initial therapy should be PFD with or without duraplasty. In refractory cases, additional syringobulbic cavity shunt is the preferred option.

Nikoobakht and associates (2019) noted that PFD, with and without duraplasty, are accepted treatments for symptomatic adult patients with CMI. However, there is still debate of the superiority of one technique over the other. These investigators determined the clinical and cranio-metrical imaging outcomes of a series of patients who underwent PFD with duraplasty. All adult patients with symptomatic CMI operated at a single institution with a minimum of 6 months follow-up were enrolled prospectively. Clinical outcomes and cranio-metrical parameters based upon MRI pre- and post-surgery were analyzed. A series of 33 consecutive patients who met the inclusion criteria were enrolled; mean age of 33.93 ± 10 years (range of 14 to 56 years). The most common pre-operative complaint was headache. The most common clinical sign was sensory dysfunction that was relieved or improved in 63% of patients. The mean syringomyelia size had a significant reduction after the surgery (p = 0.01). The mean tonsillar descent also had significant reduction (p = 0.00). The mean McRae line length before the surgery and after that were 33.4 and 53.1 mm, respectively; and this change was not statistically significant (p = 0.42). The odontoid process parameters had no significant changes following surgery. The authors concluded that PFD with duraplasty could improve both clinical and imaging outcomes
such as syringomyelia size and tonsillar descent for patients with symptomatic CMI. However, no significant difference was observed in cranio-metrical parameters before and after the surgery.

Intraoperative Neurophysiological Monitoring in Pediatric Chiari Surgery

Rasul and colleagues (2019) stated that the role of intra-operative neurophysiological monitoring (IONM) during surgery for CM-1 has not been fully elucidated. Questions remain regarding its utility as an adjunct to foramen magnum decompression surgery, specifically, does IONM improve the safety profile of foramen magnum decompression surgery and can IONM parameters help in intra-operative surgical decision-making. These researchers described a single institution experience of IOM during pediatric CM-1 surgery. The methodology comprised a retrospective review of prospectively collected electronic neurosurgical departmental operative database. Inclusion criteria were children under 16 years of age who had undergone foramen magnum decompression for CM-1 with IONM. In addition to basic demographic data, details pertaining to presenting features and post-operative outcomes were obtained. These included primary symptoms of CM-1 and indications for surgery. MRI findings, including the presence of syringomyelia on pre- and post-operative imaging, were reviewed. Details of the surgical technique for each patient were recorded. Only patients with either serial brainstem auditory evoked potential (BAEP) and/or upper limb somatosensory evoked potential (SSEP) recordings were included. Two time-points were used for the purposes of analyzing IONM data; initial baseline before skin incision and final at the time of skin closure. A total of 37 children underwent FMD with IONM. Mean age was 10.5 years (range of 1 to 16 years) with a male to female ratio 13:24. The commonest clinical features on presentation included headaches (n = 15) and scoliosis (n = 13); 24 patients had evidence of associated syringomyelia (24/37 = 64.9 %). A reduction in the SSEP latency was observed in all patients. SSEP amplitude was more variable, a decrease was observed in 18 patients and an increase was observed in 12 patients. BAEP recordings decreased in 13 patients and increased in 4 patients. There were no adverse neurological events following surgery; the primary symptom was resolved or improved in all patients at 3-month follow-up. Resolution or improvement in syringomyelia was observed in
19/24 cases. The authors concluded that these findings showed that FMD for CM was associated with changes in SSEPs and BAEPs. However, these investigators did not identify a definite link between clinical outcomes and IONM, nor did syrinx outcome correlate with IONM. They stated that there may be a role for IONM in CM surgery; however, more robust data with better-defined parameters are needed to further understand the impact of IONM in CM surgery.

### CPT Codes / HCPCS Codes / ICD-10 Codes

*Information in the [brackets] below has been added for clarification purposes. Codes requiring a 7th character are represented by "+":*

<table>
<thead>
<tr>
<th>Code</th>
<th>Code Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CPT codes covered if selection criteria are met</strong></td>
<td></td>
</tr>
<tr>
<td>61343</td>
<td>Craniectomy, suboccipital with cervical laminectomy for decompression of medulla and spinal cord, with or without dural graft (eg, Arnold-Chiari malformation)</td>
</tr>
</tbody>
</table>

| ICD-10 codes covered if selection criteria are met |
| G93.5 | Compression of brain [Chiari malformation type I] |
| Q01.0 - Q01.9 | Encephalocele [Chiari malformation type III] |
| Q04.8 | Other specified congenital malformations of brain [Chiari malformation type IV] |
| Q07.01 | Arnold-Chiari syndrome with spina bifida [Chiari malformation type II] |

The above policy is based on the following references:


AETNA BETTER HEALTH® OF PENNSYLVANIA

Amendment to
Aetna Clinical Policy Bulletin Number: 0931 Chiari Malformation Decompression Surgery

There are no amendments for Medicaid.