Chiari Malformation: A Research Study

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Abstract

Chiari Malformation Type 1 is a congenital defect in the base of the skull and the cerebellum, a portion of the brain that controls voluntary movement and balance. The radiological definition of Chiari categorizes it as a structural defect when the cerebellum extends below the foramen magnum and into the spinal canal. The condition arises in more than 200,000 people annually; however, numbers are increasing due to the availability of magnetic resonance imaging (MRI) that has increased the diagnosis of Chiari. Most radiological diagnosis conducted recently are in the asymptomatic (without symptoms) or oligosymptomatic (few or minor symptoms) patients. Decompression of the foramen magnum is the treatment of choice, as there is no role for conservative treatment or medications once symptoms appear. Nevertheless, the pathophysiology and natural history of this disease are still unclear, which reflect on the current absence of a standard technique for surgical treatment. Most of the studies available on the literature to date are with a small sample size of patients, with short follow up and retrospective in nature. Therefore, there is still much debate as to the amount of bony decompression and whether it should include duraplasty or just incising the outer layer of dura, and the need for arachnoid dissection, tonsillar coagulation or resection, shunting of csf and the choice of ideal type of graft for dural closure. Even though there is no level I or II evidence for recommendations, the recent trend has been toward a minimally invasive approach, since, with a few exceptions, most series report less complications and similar outcomes for extradural or extra arachnoidal procedures. Nevertheless, the recurrence rate is still relatively high for either

technique. This study reports on a consecutive series of 126 pediatric patients with Chiari I malformation. We intend to demonstrate cerebellar tonsillar resection is still a valid option for the management of these patients. Done meticulously and following strict principles, it can be performed safely and achieve improved outcome in selected cases.

Keywords: Chiari Malformation, cerebellar tonsillar, decompression, duraplasty, and arachnoid.

Introduction

Chiari malformations are structural defects at the base of the skull and the cerebellum, a portion of the brain responsible for motor functions and balance. In normal structuring, the cerebellum and the brainstem are positioned above an opening for the spinal cord to pass through known as the foramen magnum. When the cerebellar tonsils descend into the foramen magnum, it is radiologically defined as Chiari malformation ("Chiari Malformation Fact Sheet", 2018). Chiari tends to develop when the skull is smaller which ultimately forces the cerebellum into the foramen magnum. The compression of the brain leads to a buildup of pressure on the cerebellum and brain stem which can potentially affect the surrounding area and block the flow of cerebrospinal fluid (CSF). CSF holds the primary purpose of acting as a cushion for the brain and spinal cord by circulating nutrients filtered by the blood and removing any excess waste from the brain.

CM is a topic of intellectual interest primarily due to the lack of concrete knowledge known on the topic, especially with concern to the causes of CM. The proposed theory of how CM develops is due to structural defects during the developmental process of a fetus. The

mutations can be genetic or due to lack of vital nutrients in the mother's diet. Primary causes of CM tend to be categorized as congenital symptoms indicated that they were developed at or prior to birth, in the mother's womb. However, secondary symptoms occur when the patient develops Chiari later in their life. The possibility of secondary symptoms arises from the risk of the crianage of excess fluid if encountered with a traumatic brain injury, disease, infection, etc. Primary CM tends to be more common and less severe in comparison to secondary CM.

The primary symptoms of CM are generally headaches especially if they occur following a sneeze, cough, or anything that jerks the head slightly forward. Other symptoms may include vomiting, nausea, numbness, hearing or balance problems, ringing or buzzing in the ears (tinnitus), insomnia, scoliosis, neck pain, etc. In some cases, the patients may be asymptomatic indicating that they do not show any symptoms of Chiari. Symptoms generally vary based on the amount of CSF pressure in the child's brain and the effect of compression on the nerves. In infants, CM may indicate signs of irritability upon eating, difficulty swallowing, drooling, vomiting, apnea, slow development, lack of weight gain, stiff neck, arm weakness, etc.

Chiari malformation varied based on the type of CM. The grades range from one to four with one being more common and four being more rare, while CM-1 is less severe and CM-4 is extremely severe. CM is generally not a life threatening condition; however, in the later stages, there is a higher risk for fatality.

Chiari Malformation Type 1

Type 1 occurs when the cerebellar tonsils descend into the foramen magnum which is usually where the spinal cord enters. CM-1 is the most common type of Chiari and patients are

often asymptomatic, but they may develop symptoms as they age. Due to the lack of symptoms, most diagnoses for CM-1 are by accident during a regular neurological exam.

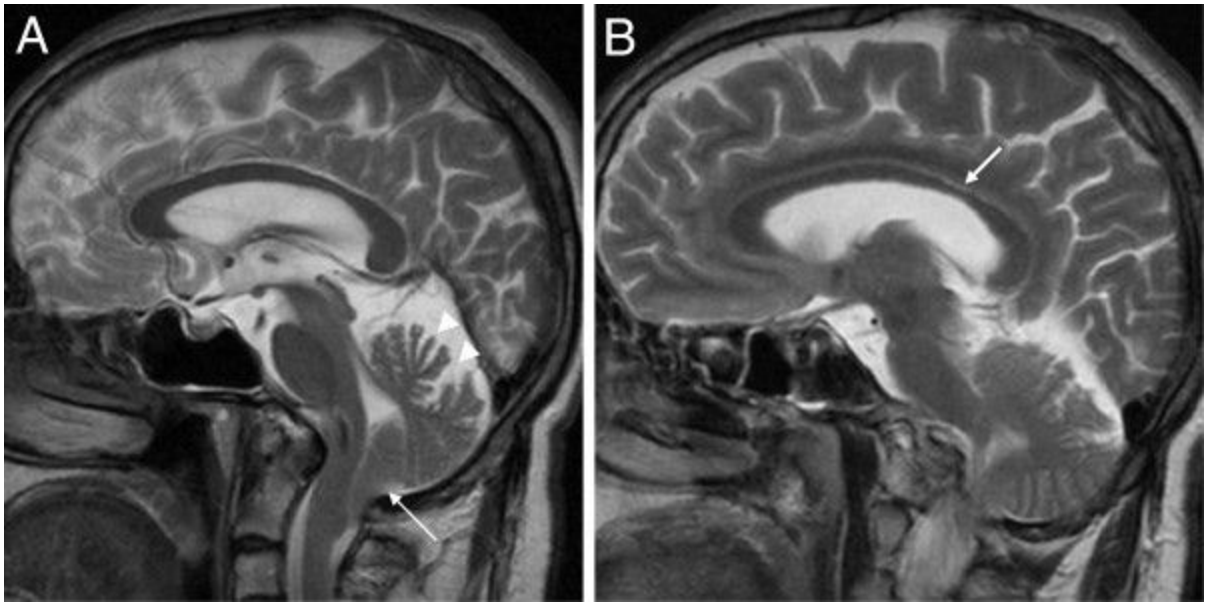

Figure 1 indicate Chiari malformation type 1 as indicated by the arrow. Scan A indicates a mid sagittal T2 herniation of the cerebellar tonsils indicating CM. Scan B indicated a para midsagittal depicting a slight short stretched thinning of the posterior mid body of the corpus callosum.

Chiari Malformation Type 2

Type 2 patients tend to indicate more symptoms than CM-1 patients and may oligosymptomatic. CM-2 can cause life threatening conditions that require surgery. In CM-2 the brainstem and cerebellum protrude out of the foramen magnum. It occurs when also when the nerve tissue that connects the two halves of the cerebellum is missing or partially seen. Type 2 usually indicated a myelomeningocele, a type of spina bifida when the spinal canal and backbone do not close before birth. A myelomeningocele may lead to complete or partial paralysis below the spinal opening

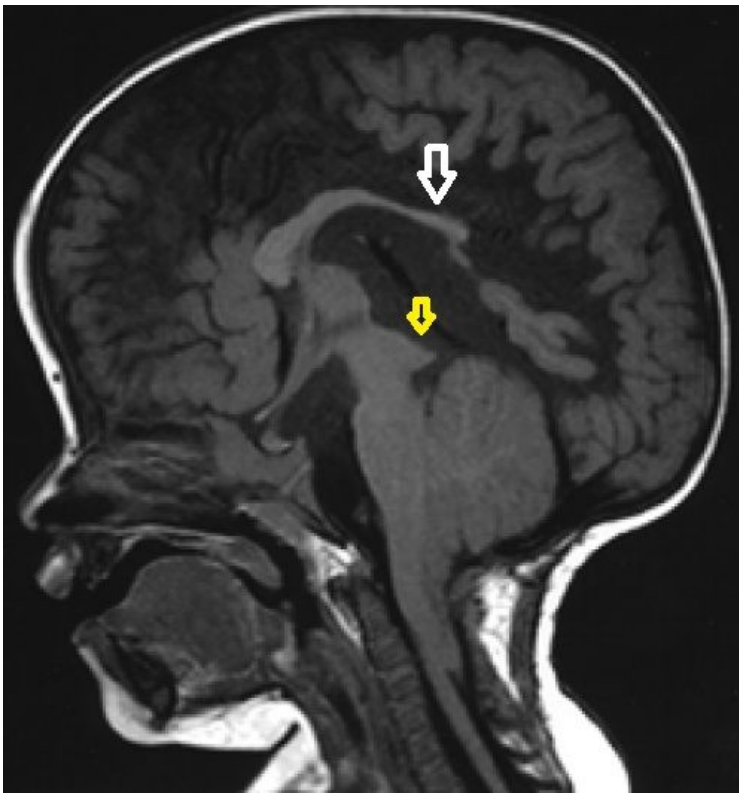

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Figure 1.1 indicates CM-2 with a portion of the cerebellum and brain stem protruding out of the foramen magnum. The MRI scan indicates lack of development in the spinal canal and backbone known as a spina bifida.

Chiari Malformation Type 3

In CM-3 some of the cerebellum and brain stem stick out or herniate through an abnormal opening in the back of the skull. The symptoms of CM-3 tend to appear during infancy and can be life threatening conditions. The child may face other neurological issues including developmental delays and seizures.

Chiari Malformation Type 4

CM-4 involves incomplete or underdeveloped cerebellum (a condition known as cerebellar hypoplasia). This condition is very rare in which the cerebellum is located in its normal position, but parts of it may be missing and portions of the spinal cord or skull may be visible.

Conditions Associated with Chiari Malformation

Hydrocephalus

Hydrocephalus occurs when there is an excess accumulation of CSF in the brain. This condition is responsible for the blockage of normal flow of fluid causing pressure in the head that can enlarge or misconfigured the skull in some regions. The condition is the most common in Type 2, but can be seen in CM-1. On some occasions, if hydrocephalus is left untreated it can be fatal. To treat hydrocephalus, reservoirs are placed in infants at a young age to remove cerebrospinal fluid manually. However, as the patients gets older a shunt is placed in the brain and is known as a ventriculoperitoneal shunt (VP) which relieves pressure in the brain by draining the CSF fluid into the abdomen underneath the fascia.

Spina Bifida

Spina bifida is defined as an incomplete closing of the backbone and membranes around the spinal cord. In infants, spina bifida yields a incomplete development of bones around the spinal cord. Most conditions associated with spina bifida in infants do not lead to neurological; however, in CM-2 patients develop myelomeningocele in which the spinal cord remains open in

one area of the back and lower spine. When the membranes and spinal cord protrude through the spinal opening, creating a bulging sac in the baby's back. The impairments caused from this can range from muscle weakness to paralysis and scoliosis.

Syringomyelia

The disorder occurs when a syrinx, a CSF filled cyst, forms in the central spinal canal. Once the syrinx begins growing, it can destruct the central part of the canal leading to muscle weakness. Development of a syrinx may also lead to the development of numbness and resistance to feeling hot or cold.

Tethered Cord Syndrome

This occurs when the spinal cord attaches abnormally to tissues around the bottom of the spine. Thus, tethered cord restricts the mobility of the spinal cord which leads . Children with myelomeningocele have an increased risk of developing an increased risk of tethered cord syndrome.

Spinal Curvature

Spinal curvature can occur in both CM-1 and with the occurrence of syringomyelia. The spine may bend left or right which would lead to scoliosis or bend forward leading to kyphosis.

Treatment

Treatment for Chiari malformation generally encompasses physical therapy for less severe cases or surgery for more severe cases. In asymptomatic patients, the patient can sometimes not need any treatment unless a syrinx develops or hydrocephalus, tethered cord, or spina bifida occurs. The most common form of treatment includes a posterior fossa decompression which created space in the cerebellum and relieve any pressure applied on the brain or spinal cord. The surgery usually consists of a craniectomy in which part of the skull is removed. In some cases the arched bony top of the spinal canal may be removes in a spinal laminectomy. While in some cases, the surgery is done to restore the flow of CSF, in others surgery is done to reconstruct the spine and fuse the spinal bone.

Infants with myelomeningocele may be done to reposition the spine and close any opening that may have occurred due to the lack of development known as spina bifida. However, surgeons usually recommend prenatal surgery as it allows the patient to be treated in the mother's womb which reduces the chances of a child developing hydrocephalus after birth.

Hydrocephalus may be treated with a shunt surgery in which a catheter is placed in the brain and goes down into the abdomen, underneath the fascia. The excess CSF fluid is drained into the abdomen and can be absorbed back into the body.

A procedure called third ventriculostomy is used in patients with hydrocephalus to restore the flow of CSF fluid. A small hole is drilled into the brain in the third ventricle and CSF fluid is restored to ensure that the fluid is flowing properly after surgery.

Material and Methods

A consecutive series of pediatric patients with Chiari I malformations all operated by the senior author between 1995 and 2013 was reviewed. Exclusion criteria were associated craniosynostosis or achondroplasia. Surgical indications were an expansile syringomyelia or the presence of symptoms attributed to the Chiari I itself or to the syrinx. All patients were evaluated with MRI and, in patients with few or unclear symptoms, also with cardiac gated cine MRI. We recorded pre operative signs and symptoms, MRI characteristics, sleep and swallow studies in the most recent patients, surgical technique, as well as clinical and radiological follow up results.

Surgical Technique

All patients were placed prone after proper padding. If they were older than 3 years, their head was fixed on the Mayfield. Otherwise, their head was firmly secured on a horseshoe holder. An incision was made midline from the mid scama occipitalis to just below C1. The musculature was elevated with a periosteal dissector assisted by bovie coagulation, and the foramen magnum was exposed. We did not routinely dissect the muscles down to the bone at the posterior arch of C1, but exposure was enough so as to allow visualization of the atlantooccipital membrane. A small suboccipital craniectomy was performed and the foramen magnum was enlarged. There was no specific measurement as for the size of the craniectomy, but it always included all the

bone turned inwards and indenting on the dura; it never exceeded more than 2 x 2 cm. The thick atlantooccipital membrane was cut and released. We did not routinely do a c1 laminectomy. The dura was opened in a Y fashion, and care was taken so as not to open the arachnoid. The arachnoid was then opened sharply in the midline and its lateral borders sutured to the dura. Attention was then turned to the intradural compartment with the aid of the microscope. The herniated tonsils were coagulated until they ascended back to the level of the foramen magnum. In case this could not be accomplished, tonsillar resection was carried out in a subpial fashion. Extra care was taken in the dissection so as not to injure the brainstem or arteries; this was accomplished by padding the arteries and pia with small rolled up moistened cottonoids. We ensured lateral decompression of the brainstem was sufficient by also removing the laterally herniated portions of the tonsils subpial. After visualization of the outlet of the 4th ventricle and outflow of CSF, attention was then turned to dural closure. Different materials were used for graft but, recently, the most frequent one has been a dermatome . We used a 6.0 pds double needle stitch starting from the bottom and moving up each needle on each side of the dura. The stitch ran from the inside to the outside of the graft and then from the inside to the outside of the dura in a way so that the graft would sit underneath and flat against the dura; this way, in case of high CSF pressure, it would push the graft towards the dura allowing maximum contact between the graft and the dura so as to avoid leaks. After the watertight dural closure, the remaining layers (muscle, fascia, galea) were sutured separately and the skin sutured with running stitches.

Discussion

Chiari malformation constitutes a continuum of hindbrain herniation, and Chiari type I occurs when only the cerebellar tonsils descend through the foramen magnum into the spinal canal. Diagnosis is made invariably when the descent is greater than 5mm or in a few situations less than 5mm when the tonsils are bilaterally herniated or when gated cine MRI demonstrates abnormal csf flow through the foramen magnum. The disease is related to a disproportion between a small posterior fossa and its neural contents Navarro, klekamp as well as to genetic predisposition klekamp. It is frequently associated with syringomyelia and, less often, with bone abnormalities such as achondroplasia, basilar invagination, Klippel Feil syndrome, hypophosphatemic rickets, among others. Syrinx is present in 30-70% of cases.

It is somewhat expected there is controversy regarding the optimal surgical treatment for Chiari I malformations since there is still debate regarding its pathophysiology. There are at least 3 major theories explaining the development of syrinx in Chiari I disease. According to Gardner, an incomplete opening of the outlet of the 4th ventricle is responsible for retained communication between the ventricle and the central canal. During systole, as csf pressure is increased, fluid can go through the obex into the central canal leading to formation and progression of the cavity. According to Williams, herniation of tonsils cause obstruction of flow from the cranial to the spinal subarachnoid space, creating a pressure gradient between the two, causing swings of venous pressure associated with coughing, sneezing and straining (Valsalva maneuvers) to move csf from the ventricle into the syrinx, as fluid is sucked into the central canal. Both theories depend on movement of csf through an opening between the syrinx and the 4th ventricle, which is infrequently seen. In fact, neither theory can explain the existence of a

syrinx located far from the 4th ventricle, down in the lumbar region, with a normal segment of the spinal cord in between. A more recent theory, by Oldfeld, tries to address this issue, as the obstruction of csf flow by the herniated tonsils makes them act as pistons, creating pressure waves that force csf into the cord through perivascular and interstitial spaces.

Indications for surgical treatment are usually not a matter of debate, and most surgeons will operate only on symptomatic patients kalb, caldarelli, kumar, klekamp, munshi, seok, yilmaz, regardless of the presence of syringomyelia. Only 9% of pediatric neurosurgeons in a recent survey recommended prophylactic intervention harou. Because all of our patients were pediatric and symptoms attributed to the cavity are less likely to improve hayhurst, munshi, el ghandour, klekamp, Williams in hayhurst, we also indicated surgery for asymptomatic patients when they had an expansile syrinx demonstrated on MRI. Some don't actually think this group of patients exists klekamp, even though we had 6 patients with incidentally discovered Chiari I and syringomyelia.

The most controversial issue regarding Chiari I malformation is to decide which is the most appropriate technique of surgical treatment navarro. There are several options available, but results. Foramen magnum decompression is standard, but a consensus has not yet been reached there are no prospective comparative studies available to determine which one produces the best on whether to open the dura, dissect the arachnoid, shunt the syrinx, coagulate or resect the tonsils. The indication for a specific technique lays on the surgeon's preference and experience hankison, munshi, kalb, galarza, yilmaz.

Recently, the trend has been toward a less invasive approach, either just bony decompression or only cutting the outer layer of dura or opening the dura while leaving the

arachnoid intact Hankinson, durham. Proponents to these approaches cite similar outcomes and fewer complications than the more invasive counterparts seok, caldarelli, imae in seok, such as dissecting the arachnoid, coagulating or removing the tonsils. In general, opening the dura increases the risk of csf related complications munshi while compromising long term effects. Perhaps, the most important issue is to determine which patient needs what type of surgery, so the procedure can be tailored. Since outcome is difficult to predict hayhurst, munshi, we still don't have an answer and despite initial encouraging results with any technique, a significant proportion will have high recurrence kalb, el ghandour, even as early as 12 months Hankinson. Tubbs reported the largest pediatric case series so far, with 500 patients. He performed posterior fossa decompression, C1 laminectomy and arachnoid dissection to establish spontaneous flow from the 4th ventricle. His complications and recurrences were low, that is 2.4% and 3%, respectively. His mean follow up was 5 years, one of the longest in all series reported to date. In his first case series, with 130 patients, he had a 36.3% rate of reoperations.

Klekamp looked at 359 patients, mean age 40 years, and defended duraplasty and inspection of arachnoid in all to make sure csf flow was present from the outlet of the 4th ventricle. Dissecting the arachnoid did not influence his complications; instead, they were related to the degree of arachnoid pathology he encountered. Even though he had few complications while opening the arachnoid (5.9% rate of csf fistula), his recurrence was significant (30 patients, or approximately 8.5%).

Dr. Gambardella proposed a large posterior fossa craniectomy but no intradural approach, just micro incisions in the outer layer of dura. Even though it is a short sample of 8 adult patients and 7 improved, one had symptom and syrinx progression (a 12.5% rate of recurrence).

Seok reported his findings in 25 adults that underwent arachnoid preserving duraplasty. One patient got worse and there was a 16% complication rate attributed to incidental dural opening. There is no mentioning of recurrences.

Dr. Munshi compared the results of posterior fossa decompression and C1 laminectomy with and without duraplasty in 34 patients between children and adults. He concluded there is a higher likelihood of improvement after duraplasty, even though there is an increased risk of minor complications (aseptic meningitis, csf leaks, superficial infections).

Kalb evaluated 137 patients, 17 of whom were children, that underwent posterior fossa enlargement with duraplasty, with few complications (2.8%). Outcome was not affected by whether arachnoid was opened or not.

Dr. Navarro studied 96 patients, with mean age of 9.8 years, and compared posterior fossa decompression and outer dura peeling with arachnoid dissection and tonsillar manipulation. Complications were much higher in those who underwent duraplasty (42.1%) than in those that did not have the dura opened (5.6%). Recurrence rate was reported to be 13.5%, and it occurred even in those who initially responded to surgery.

Dr. Kumar described the results of 87 patients with a mean age of 26y who underwent posterior fossa decompression, c1 laminectomy, duraplasty, arachnoid lysis of adhesions and tonsillar coagulation. He had 13 complications (14.9% complications, 8% were csf related). He did not discuss recurrences.

Dr. Caldarelli described the results of his serial incisions on the outer dura in treating 30 patients, with very good results (28 improved) and no csf complications. Nevertheless, his hospital stay was 6 days and there were 2 recurrences (6.6%).

Galarza reported on 60 pediatric patients and compared four techniques: duraplasty vs outer dura incisions vs coagulation of tonsils vs subpial resection of tonsils. He found better results with tonsil reduction procedures at no increased risk of complications.

Dr. Durham reviewed the literature in a 582 patient meta-analysis comparing duraplasty with bone decompression alone. There was significant outcome difference only in relation to csf complications and reoperation. Patients that underwent duraplasty had decreased reoperation rates (2.1%) and higher csf complication frequency (18.5%) compared to the ones who underwent bone decompression only (12.6% for reoperation and 1.8% for csf complications). Valentini studied 99 children that underwent different treatment techniques, divided into bone decompression only, duraplasty and tonsillar coagulation or resection. Csf related complications occurred in 14%, with a 3% incidence of neurological transient deficit. Interestingly, he reported improvement even in psychiatric symptoms, an observation we also share and were able to see in our series.

Dr. Mottolese reported on 82 pediatric patients that underwent bone decompression, c1 laminectomy and. duraplasty. He compared duraplasty with Goretex to leaving the dura open with an intact arachnoid. Tonsillectomy was also performed in case of symptomatic syrinx, in both groups. There were 7 complications (18%) only in the open dura group, with no complications in the ones that received Goretex. There is no information regarding recurrence. They reserved tonsillectomy just in case of failure of decompression with duraplasty.
Seon Park compared 57 patients that underwent posterior fossa decompression and duraplasty with or without tonsillar management. He found no effect of tonsillar manipulation on syrinx improvement, and recommended coagulation or resection of tonsils only in recurrences.

Nevertheless, he had four revision surgeries which all occurred in patients that underwent bone decompression and duraplasty alone.

Yilmaz studied 82 adult patients and compared duraplasty to non duraplasty. The results showed duraplasty was better when the degree of tonsillar descent was greater, but it had more complications, including neurological deficits and csf leaks.

Zhang reviewed 132 patients among children and adults, and compared large (5 x 6cm) with small (3 x 4cm) posterior fossa craniotomy; all patients underwent duraplasty and, depending on the degree of tonsillar herniation, also tonsillectomy. Complications were more frequent in the larger craniectomy group because of cerebellar sinking, excessive muscle dissection, limited cervical movement and adhesion to the subarachnoid space attributed to bleeding and scarring. There were no differences in syrinx or clinical outcomes and there is no information on recurrences.

Dr. McGirt reviewed 256 children with specific attention to symptom recurrence. His technique consisted of small suboccipital craniectomy, c1 laminectomy and duraplasty, though there was coagulation of tonsils in some patients. Frontal headache, increased duration of headache and vertigo pre operatively were associated with a higher risk of recurrence. Symptom recurrence was 22% and need of reoperation was 7%.

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