Arnold Chiari Malformation Type I in Conjunction with Idiopathic Scoliosis and Syringomyelia (Backgrounds, Patterns/Relationships, Treatments, and Contraindications)

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Senior Research Paper
Abstract

Objective: The aim of this literature review was to find clinical correlations between Arnold Chiari Malformation type I, Idiopathic Scoliosis, and Syringomyelia by combining and comparing various research data.

Design: The history of pathogenesis of Arnold Chiari Malformation type I along with Idiopathic Scoliosis and Syringomyelia has long been a mystery. Many hypotheses have been developed as to the origins of these various spinal anomalies. However, only a few distinct and conclusive evidence based studies have been published that attempt to clarify the origin of pathogenesis and link the three. Despite this fact, the studies are still not in complete agreement with one another on the origins, pathogenesis, or corrective procedures for the three anomalies. The bulk of this paper will present the backgrounds of Arnold Chiari Malformation, Idiopathic Scoliosis, and Syringomyelia along with any patterns or relationships, treatments, and contraindications that exist through literature.

Results: The results showed 96.54% of patients with SMI had some descent of the cerebellar tonsils (7). 72.97% of the SMI patients also had idiopathic scoliosis (7). 74.07% of the patients in whom the conus medullaris was seen had scoliosis and a low position of the conus medullaris (7). Twenty-two patients were identified and fittingly syringomyelia showed up in 50% of the cases, while the cerebellar tonsils were found to lie at C1 and C2 in 9 and 13 patients. No abnormalities or caudal descent of the midbrain or pons were identified.

Conclusion: Before any individual case of Arnold Chiari Type I, Syringomyelia, or Idiopathic Scoliosis is independently diagnosed, identification of secondary anomalies should be considered as possible contributing factors to the pathology at hand.

Key Indexing Terms: Arnold Chiari Malformation, Idiopathic Scoliosis, Syringomyelia, Chiropractic, cerebellar, brainstem, cervical cord symptoms, disturbances of cerebrospinal fluid dynamics
Introduction

The history of pathogenesis of Arnold Chiari Malformation type I along with Idiopathic Scoliosis and Syringomyelia has long been a mystery. Many hypotheses have been developed as to the origins of these various spinal anomalies. However, only a few distinct and conclusive evidence based studies have been published that attempt to clarify the origin of pathogenesis and link the three. Despite this fact, the studies are still not in complete agreement with one another on the origins, pathogenesis, or corrective procedures for the three anomalies. The bulk of this paper will present the backgrounds of Arnold Chiari Malformation, Idiopathic Scoliosis, and Syringomyelia along with any patterns or relationships, treatments, and contraindications that exist through literature.
Review of Literature

Arnold Chiari Malformation type I (ACM) is a projection of the medulla and cerebellum that extends through the foramen magnum and into the cervical spinal canal (1). It is widely presumed that during fetal life, such changes take place. A common theory is that fixations of the lower spinal cord or its nerve roots may exert traction on the upper cervical cord and brain stem, causing the medulla and cerebellum to herniate through the foramen magnum (1). Leong and Kermode add that Arnold Chiari may also be associated with displacement of the medulla and hydromyelia or syringomyelia (2). The history of individuals with type I Arnold Chiari Malformation is usually variable. In a reported series of 71 patients, 69% presented with pain, 56% had weakness, 52% had numbness, and 40% complained of unsteadiness (2). The effects of ACM type I can vary in presentation throughout different age groups. The entity of ACM type I may cause symptoms because of impaction of the tonsils at the foramen magnum leading to cerebellar, brainstem, or cervical cord symptoms, and also because of disturbances of cerebrospinal fluid dynamics (3). The diagnosis of ACM type I can be made with the use of magnetic resonance imaging (MRI). With the advent of MRI, the diagnosis of Arnold Chiari Malformations type I is being made in younger patients, often with minimal or no neurologic symptoms (4). This diagnosis is crucial in the event of certain treatment protocols, which could further worsen the existing problem.

Idiopathic scoliosis in adolescents is the most common type of scoliosis and may have link to a genetic predisposition. Most adolescent cases occur in girls and curves generally worsen during growth spurts. There are also infantile and juvenile forms that are less common and affect a similar number of boys and girls. Symptoms often seen
with scoliosis include fatigue and pain from the persistent irritation of ligaments. The greater the initial curve of the spine, the greater the chance for progression of the condition after growth is complete. Severe scoliosis (curves in the spine greater than 100 degrees) may cause respiratory problems. However, scoliosis is not limited to the adolescent age group. Once the spine has reached skeletal maturity or growth is completed, a patient who has been diagnosed with adolescent idiopathic scoliosis now has adult idiopathic scoliosis. The distinction between the two becomes important when considering the precipitating factors for treatment. Though the adult may still need treatment to halt the progression of the curve, accompanying pain is often the common chief complaint. Normal degenerative changes of the spine may be accelerated by curvature and the patient with adult idiopathic scoliosis may be at higher risk for skeletal pain or extremity pain due to nerve compression (5). If treated, adult idiopathic scoliosis should never lead to neurologic (paralysis) or cardiopulmonary (heart or lung failure) deterioration (5). Therefore, treatment protocols should be considered that will not exacerbate the current condition.

Syringomyelia (SM) is a condition in which a cyst forms within the spinal cord (6). Known as a syrinx (cyst filled with cerebrospinal fluid), this cyst expands and elongates over time, destroying the center of the cord. With destruction of the cord pain, weakness, and stiffness in the back, shoulders, arms or legs may be present. Other symptoms may include headaches and loss of the ability to feel extremes of hot or cold, especially in the hands and disruption in body temperature (6). SM may also adversely affect sweating, sexual function and bladder and bowel control (6). Spinal cord trauma or congenital developmental problems of the brain and/or spinal cord may result in SM (6).
With either case, the condition of SM may remain masked for many years where upon a variety of symptoms tend to become bothersome. With syringomyelia the normal flow of cerebrospinal fluid (CSF) is obstructed and is therefore redirected to the spinal cord itself. With redirection, the result is the syrinx. Remaining pressure differences along the spine cause fluid to move within the cyst (6). It is believed that this continual movement of fluid results in cyst growth and further damage to the spinal cord and connecting nerves (6). Generally speaking, there are two forms of SM. The disorder may be related to Arnold Chiari malformation. A syrinx may then develop in the cervical region of the spinal cord; this is referred to as communicating syringomyelia (6). Some people with this form of the disorder also have hydrocephalus (water on the brain), a condition in which CSF accumulates in the skull, or arachnoiditis, in which the arachnoid layer of the spinal cord is inflamed (6). The second major form of SM occurs as a complication of trauma, meningitis, hemmorhage or tumor (6). Here, the cyst or syrinx develops in a segment of the spinal cord damaged by one or more of these conditions. The syrinx may start to expand; this is sometimes referred to as a noncommunicating syringomyelia (6). The syringomyelia associated with ACM I however, is the form in which we are concerned within the context of this article.

Literature review on the correlations between Arnold Chiari Malformation type I, idiopathic scoliosis, and syringomyelia remain few. However, the literature available remains intriguing as to the possible connections. Royo-Salvador’s research looks into major theories of the pathogenesis of the three. A new theory involving a common pathology for ACM type I, idiopathic scoliosis, and idiopathic syringomyelia (SM) was proposed. Its confirmation depends on the presence of an abnormally low position of the
conus medullaris (CM) in patients with SM (7). Of 292 patients with SM, 231 patients with SM were selected, and 55 of these were chosen in whom the level of the conus medullaris could be determined, together with figures for SM, idiopathic scoliosis, and ACM I by MRI (7). The position of the conus medullaris in 50 patients who did not have SM, idiopathic scoliosis, nor ACM I on cervical and lumbar MRI was determined (7). The results showed 96.54% of patients with SMI had some descent of the cerebellar tonsils (7). 72.97% of the SMI patients also had idiopathic scoliosis (7). 74.07% of the patients in whom the conus medullaris was seen had scoliosis and a low position of the conus medullaris (7). It was shown in the control group that 6% had a conus medullaris at the level of the body of L1, while 84.21% with SMI had a partial or complete image of the conus medullaris at the same L1 level. In these patients an unusually low position of the conus medullaris was confirmed (7). The study concluded that the unusually low position of the conus medullaris in SM patients and its close relationship to ACM I and idiopathic scoliosis make it likely that the same pathogenic mechanism is shared by them all; a connection in which the study called “abnormal asynchrony of growth of the notochord and the spinal cord”.

Another study performed by Salvador along with Llenas, Domenech, and Gonzalez-Adrio reported the finding of a section of filum terminale in 20 patients with syringomyelia, scoliosis, and Chiari malformation. They reported that spinal cord traction caused by a tight filum terminale may be considered a pathogenic mechanism involved in the development of syringomyelia, the Chiari malformation type I and scoliosis (8). They stated that sectioning of the filum terminale is a proposed useful surgical approach for these conditions. The study reported that between April 1993 and
July 2003; a total of 20 patients (8 men and 12 women) with a mean average age of 33.5 years underwent section of the filum terminale with or without opening of the dural sac through a standard sacrectomy (8). Eight patients suffered from scoliosis, 5 from syringomyelia, 2 from Chiari malformation, and 5 with a combination of these conditions (8). The results showed that after sectioning of the filum terminale, patients with syringomyelia had an early clinical improvement of dysaesthesia, thermo-anaesthesia, hypo-aesthesia, and walking difficulties. The also stated that rising of the conus medullaris was observed. For the patients with scoliosis, back pain improved dramatically and a curve reduction was noticed, although progression of the curve was observed in one case (8). As for the patients with Arnold Chiari malformation, headaches, dysaesthesia, and paraparesis disappeared (8). Conclusions from the study state that sectioning of the filum terminale is a useful strategy in the treatment of scoliosis, syringomyelia, and Chiari malformation, and that it also offers a new aetiological basis for the understanding of these three disorders (8).

Two more studies explained results and connections between Arnold Chiari malformation and syringomyelia. The first study was performed by Mimura, Asajima, Saruhashi, and Matsusue (9). They explained a case of Arnold Chiari syndrome with flaccid paralysis and huge syringomyelia. The case report was of a 13-year old women who presented with progressive weakness in the lower extremities along with predominance on the right. Magnetic resonance imaging (MRI) revealed a huge syrinx (9). The patient also showed scoliosis, cleft palate, hearing impairment, excessive sweating, hairiness, dural ectasia, and malformation of the skull (9). After the treatment of Arnold Chiari with flaccid paralysis, both the syrinx and muscle strength were quickly
improved. The improvement stemmed from the placement of a syringo-peritoneal (S-P) shunt, after which the patient recovered the ability to walk (9). However, transient hypesthesia in the right hand occurred after the operation. They concluded that the syrinx around the conus was thought to have played a crucial role in the etiology of the patient case. The second study linking Arnold Chiari and syringomyelia was performed by Guillen and Costa (10). They reported a case in which spontaneous resolution of a Chiari malformation type I that was associated with syringomyelia took place. Summarizing the case, the child showed complete spontaneous resolution of a Chiari type I malformation associated with syringomyelia without any surgical intervention. The child was followed clinically by serial MRI and remained neurologically stable after 8 years of follow up (10). The mechanisms of the study however, were not disclosed and therefore provide no additional theories as to the association between ACM and syringomyelia.

Perhaps one of the more interesting studies was performed by Flynn, Sodha, Lou, Adams, Whitfield, Ecker, Sutton, Dormans, and Drummond (11). In their work, they presented with predictors of the progression of scoliosis after decompression of an Arnold Chiari type I malformation. The study was a retrospective study and the patients chosen were those with scoliosis and Arnold Chiari type I malformation that required operative management. The study sought to determine the factors that could predict whether a particular spinal deformity might progress despite neurosurgical management of ACM type I (11). Up to the time of this study (February 2004) Flynn, Sodha, Lou, etc. reported that few studies had been documented that showed the relationship between the diagnosis and treatment of ACM type I and associated spinal deformities. They stated that most studies tend to mix neural axis abnormalities and contain limited information
about the specific spinal deformity. The methods used to carry out this research involved medical records, radiographs, and magnetic resonant images of patients. The patients were evaluated focusing on age and findings at presentation, characteristics of presenting and follow-up spinal deformities, and the specifics of neurosurgical and orthopedic management (11). Patients were divided into two groups: those whose curves progressed >10 degrees or to surgical range (largest curve >45 degrees) after neurosurgical intervention (progressors) and those whose curves stabilized or decreased (nonprogressors) (11). The results of the study showed eight progressors presented at an average age of 11.4 years (range 2-19) and were followed for 6.3 years (range 2-15). Seven nonprogressors presented at 6.5 years of age (range 5-10) and were followed for 6.6 years (range 3.5-14) (11). Neurosurgical procedures were equivalent in both groups; however, surgical revisions were required in 3 out of 8 progressors and 1 out of 7 nonprogressors (11). All progressors had a double scoliosis curve; but only one nonprogressor had a double curve (11). Six out of 8 progressors and 0 out of 7 nonprogressors had a rotation greater than or equal to 2+ and 50% of progressors had a thoracic kyphosis >50 degrees compared to 1 out of 7 nonprogressors (11). The conclusion of the study stated that in this particular series, progression of spinal deformity after the neurosurgical management of Arnold Chiari type I malformation was associated with later age at neurosurgical decompressions and initial neurologic symptoms, double scoliosis curve patterns, kyphosis, rotation, and larger curve at presentation. From this study, a better appreciation for the correlation of scoliosis Arnold Chiari type I can be obtained. It seems that by altering the presentation of the ACM type I malformation, other spinal anomalies such as scoliosis are directly affected. The direct
effect between the two suggests that perhaps one causes the other. It is through literature such as this, that we are gaining more insight into the pathogenesis of ACM type I and idiopathic scoliosis and any correlations that may exist between the two.

There are other studies as well that elaborate on the connections between Arnold Chiari type I malformation, scoliosis, and syringomyelia. In fact, there are far more studies available in research literature, online publishing’s, etc. than can possible be covered through the context of this document. However, I feel that several more of these studies are worthwhile in explaining for they emphasize a few key points in the connections between the three. The first such study was done by Ono, Ueyama, Okada, Echigoya, Yokoyama, and Harata and talked about adult scoliosis in syringomyelia associated with Chiari I malformation (12). The study was done on adult syringomyelia associated with Chiari I malformation, the spinal deformity, the configuration of cerebellar tonsillar descent, the configuration of syrinx, and the clinical evaluation before and after surgery. The results showed the concomitant rate of adult syringomyelia with scoliosis to be 71.4%. As the scoliosis advanced, the kyphotic angle also increased (12). The findings showed that the more advanced the scoliosis was, the more aggravated the neurologic symptoms were, and the poorer the surgical outcomes tended to be (12). The conclusion from the study was that in adult syringomyelia with scoliosis, the morbidity period is long, the syrinx is long, the neurologic symptoms are aggravated, and the surgical outcomes tend to be poor (12). The next study done by Tubbs, McGirt, and Oakes, explained the surgical experience in 130 pediatric patients with Chiari I malformations. Their objective was to present the long-term findings of certain surgical procedures on pediatric patients with Chiari I malformations. One hundred thirty
symptomatic pediatric patients with Chiari I malformations underwent posterior fossa decompression (13). The study reported that of the most common presenting symptoms (headache/neck pain and scoliosis) 12 and 17% respectively, were not alleviated postoperatively (13). Complications occurred in 2.3% of the group and nine patients had to undergo repeated operations for continued symptoms or persistent large syringes.

Brockmeyer, Gollogly, and Smith’s study presented with scoliosis associated with Chiari I malformations and the effect of suboccipital decompression on the scoliosis curve progression (14). Their study was a retrospective review of the effect of suboccipital decompression and duraplasty on curve progression in 22 patients who presented with scoliosis, syringomyelia, and a Chiari I malformation (14). The experiment base showed 21 of the 22 patients who presented with scoliosis met the inclusion criteria of having a Chiari I malformation, scoliosis, and an unfused spine during the follow-up period after suboccipital decompression. Results revealed that 13 of the 21 study patients (62%) had curve improvement or stabilization during the follow-up period (14). Curve progression existed in 8 of the 21 (38%). Their conclusion was that age, gender, and initial size of the scoliotic curve influenced the results of suboccipital decompression on the behavior of scoliosis (14). The last such study on correlations amongst ACM type I, scoliosis, and syringomyelia pertained to the correlation between scoliosis and syringomyelia.

Ozwedemoglu, Denis, and Transdeldt completed a retrospective study in which they reviewed the statistical correlation of 112 patients having both scoliosis and syringomyelia. Their objective was to determine whether there was significant correlations between the type of scoliosis, location of the syrinx, size of the syrinx, clinical manifestations of the syrinx, and associated lesions such as cord tethering and
Chiari malformation both types I and II (15). Their conclusion was that anomalies of the spinal cord or spinal column coexisting with lesions of the central nervous system have significant effects on the syrinx and scoliosis (15). They stated that there is significant relation between the most caudal level of the syrinx and the locations of the scoliosis.

Much of the article to this point has focused on the correlations that exist between ACM type I, scoliosis, and syringomyelia. Some emphasis has been placed on the treatment protocols in several of the above cases. However, I want to elaborate on several other literature studies that demonstrate different treatment protocols and the specific results of each. The first study, which was mentioned earlier, demonstrates the use of applied kinesiology and its cranial evaluation in the treatment of symptomatic Arnold Chiari type I malformation with associated cranial nerve dysfunction. Cuthbert and Blum presented an overview of possible effects of Arnold Chiari malformation and offered chiropractic approaches and theories for treatment of a patient with a severe visual dysfunction complicated by ACM (1). They features a young woman who had complex optic nerve neuritis exacerbated by an ACM type I of the brain (1). They used applied kinesiology as the treatment of choice for the loss of vision and nystagmus. After the treatment, the patient’s ability to see, read, and perform smooth eye tracking showed improvement (1). The conclusion was that further study into applied kinesiology and cranial treatments for visual dysfunctions associated with ACM would be helpful to evaluate whether this single case study could be representative of a group of patients who present with similar problems.
Tubbs, Iskandar, Bartolucci, and Oakes's study on Chiari 1.5 malformation shows more of a surgical approach once again for the treatment ACM. They identify this type of ACM malformation as being ACM 1.5 based on the combination of brain-stem herniation through the foramen magnum in conjunction with a traditional ACM type I. Their attempt was to elucidate the best surgical strategy based on retrospectively analyzed cases from specific institutions in which hindbrain herniation was diagnosed. Each patient had undergone a posterior fossa decompressive surgery (16). MRI was used to evaluate the extent of caudal descent of the brainstem, amount of tonsillar ectopia, inclination of the odontoid process, and any brain or brainstem abnormalities (16). Twenty-two patients were identified and fittingly syringomyelia showed up in 50% of the cases, while the cerebellar tonsils were found to lie at C1 and C2 in 9 and 13 patients. No abnormalities or caudal descent of the midbrain or pons was identified (16). Eighteen patients experienced resolution of preoperative symptoms, while the presence of syringomyelia prompted a second posterior fossa operation secondary to progressive scoliosis in 13.6% of the patients (16). Conclusions from the study were that no single sign or symptom was found to be peculiar to the Chiari 1.5 malformation, although all patients in whom the diagnosis was established had undergone a posterior fossa decompressive surgery (16). 13.6% of the patients required repeated operation for persistent syringomyelia which prompted the examiners to state that neurosurgeons may wish to consider other means besides posterior decompressive surgery. This was based on the evidence that in conjunction with syringomyelia, posterior decompressive surgery to correct ACM 1.5 did not respond well in this study. Once again, the traditional surgical procedures for the correction of symptoms of ACM and related ACM
malformations (i.e. ACM 1.5) appear to have some flaws. Chiropractic manipulative procedures although non-invasive, may have to be considered for future use (i.e. symptom relief) if surgical procedures continue to show poor results.

On the note of chiropractic care, another study demonstrated the adjusting procedures of chiropractic in the care of Arnold Chiari malformation. Murphy, Goldstein, and Katz published a study on cervical adjusting in the ACM patient. Their study was based upon two cases in which patients with ACM type I were treated with adjustments to the cervical spine for conditions unrelated to the anomaly (17). The study reported that patients were adjusted multiple times in the cervical spine region and that no ill effects or complications were noted. Their conclusion was that asymptomatic ACM type I is not necessarily a contraindication to skilled adjustments to the cervical spine (17). Studies such as this allow the window of opportunities for chiropractic care to certain neurological conditions to remain open. Although the patients were asymptomatic, no exacerbations to the ACM were reported, which gives positive support in the direction of chiropractic care when compared to the exacerbations following surgical procedures.

When discussing the treatment options of scoliosis in association with syringomyelia but without ACM, more surgical literature comes to the forefront. Ozerdemoglu, Transfeldt, and Denis reported on the value of treating primary causes of syrinx in scoliosis associated with syringomyelia (18). Their retrospective study was based on analysis and values associated with the risks of neurosurgical procedures for syringomyelia and of fusion procedures for scoliosis. The 105 patients who had both
scoliosis and syringomyelia were subdivided into three groups. The group which consisted of patients without congenital scoliosis or myelomeningocele showed that suboccipital craniectomy offered curve improvement in seven, worsening in three, and no change in two, whereas direct syrinx shunting gave curve improvement in none, curve worsening in six, and no change in two. Yet another group showed a high rate of subsequent neurosurgery. The subsequent surgery was nearly always suboccipital craniectomy or detethering of the cord. Three of the 38 patients (8%) had neurologic worsening when scoliosis fusion surgery was done without previous syrinx decompression (18). Although the study did not focus on ACM, there were several patients who had ACM in conjunction with their other spinal abnormalities. The results showed that patients without myelomeningocele or congenital scoliosis, but with ACM and syringomyelia, suboccipital craniectomy gave the best chance for syrinx reductions and scoliosis improvement, particularly in children younger than 10 years (18). Syrinx shunting improved none of the scoliosis and for syringes in patients with congenital scoliosis or myelomeningocele, neither neurosurgical procedure resulted in curve improvement (18). Surgical procedures for scoliosis, like ACM don’t always prove to be the best treatment method as this study demonstrates. Other means of providing relief to the patient should be explored.

Certain studies have been done on Arnold Chiari type I malformation in conjunction with syringomyelia and the preferred treatment of choice. One such study was a single case research in which the authors, on the basis of current opinions presented in literature and in their own case talked about the considerations concerning pathogenesis, operative procedures and prognostic factures which were of value in
predicting long-term outcomes in patients with ACM type I-syringomyelia complex (19). They stated that the presence or absence of three preoperative signs (muscular atrophy, ataxia, and scoliosis) facilitate, with confidence of 95%, prediction of long-term postoperative result (19). Preoperative incidence of all three were said to be much less favorable for outcome. With this case study, all unfavorable signs were observed preoperatively and despite good MRI results, long-term outcomes were only fair (19).

Once again, another study which demonstrates the use of surgical procedures to help correct ACM in conjunction with syringomyelia. This particular study however, seemed to offer some further insight into the surrounding criteria for preoperative procedures. The study took into account certain factors preoperatively and still stated that the outcome was only fair. Even with surgical means, ACM is a very tough anomaly to gain complete control of.

The last three studies I want to elaborate on deal with ACM, scoliosis, and syringomyelia once again, but in a very special group; children. Up to this point, many of the studies focused on the relationships among the three but were composed of study groups of all ages and reported differences across a wide range of individuals. Children in any aspect of life, whether science related or not, need special attention for their needs. The first such case was done by Lazareff, Galarza, Gravori, and Spinks and focused on tonsillectomy without craniectomy for the management of infantile Chiari I malformation. The authors reported their experience with 15 pediatric patients who underwent resection or shrinkage of the cerebellar tonsils without craniectomy or laminectomy, for the management of ACM type I (20). The procedure was performed in six boys and nine girls with a mean age of 10 years (20). Of the patients, 13 reported with
the congenital form of this disorder and two patients with Chiari I malformation caused by lumboperitoneal shunting. The clinical complaints of the patients included: headaches, scoliosis, numbness of the extremities, and upper-limb weakness. Two other patients presented with failure to thrive and another presented with vocal cord palsy. Syringomyelia was seen in 8 patients. The patients’ symptoms had developed within a mean time period of 21 months (20). In all patients the cerebellar tonsils were exposed through a dura mater arachnoid incision at the occipitoatlantal space (20). The tonsils were resected in seven patients and in the eight patients remaining, the tonsils were shrunk by coagulating their surfaces. All of the patients improved postoperatively (20). In the resected neural tissue, gliosis with cortical atrophy was observed. In seven of the eight patients syringomyelia was reduced. The authors reported the mean length of follow up was seven months. Their conclusion was that removal of herniated cerebellar tonsils can be sufficient for alleviating symptoms in patients with Chiari I malformations (20). Therefore, surgical procedures can render aid to infantile subjects as well. In fact, infantile subjects may be the best patients because their results can be tracked for longer periods of time and can lend insight into whether or not the disease is self-limiting in childhood.

Another pediatric study focused on Chiari malformation in association with syringomyelia and scoliosis in which the review was done over 20 years and nonsurgical treatments were used. The retrospective study reviewed patients with ACM and scoliosis, and those without associated scoliosis. Their objective was to determine the effect of decompression of ACM type I with syringomyelia and stabilization or improvement of the associated scoliosis (21). All patients were treated with surgical
decompression of the Chiari malformation with or without drainage of the syringomyelia (21). Twenty-five patients were identified, ranging in age from 19 months to 16.5 years (21). Nineteen patients had associated scoliosis and the majority of them had sought treatment for the spinal deformity but only 6 had pain or neurologic symptoms (21). Eleven underwent fusion, eight had not gotten fusion and of those, 3 had progression, one remained in stable condition, and 4 experienced improvement of curvature since undergoing to decompression. The mean age of patients who experienced progress after decompression was 14.5 years compared to 6 years for patients who experienced improvement (21). Their conclusions were that early decompression of Chiari I malformation with syringomyelia and scoliosis resulted in improvement or stabilization of the spinal deformity in 5 cases (21). Of these 5 each underwent decompression before 8 years of age and before the curve was too severe. They stated, however, that this particular series represented a few patients demonstrating this trend and that follow-up and investigation was necessary. Despite the warranty of needing other studies on this matter, the research still provided great insight into the treatment methods of a problematic group showing correlation between ACM, syringomyelia, and scoliosis.

Chiropractic theory has long been that scoliosis can be helped with the use of adjusting procedures and other techniques to correct or at least stabilize and halt the progression of the curve. However, surgical procedures for scoliotic disorders continue to be a popular trend in our country and all around the world. The last study offers insight into the success rate of such surgical procedures on a group of scoliotic individuals. Sengupta, Dorgan, and Findlay offered a study asking if hindbrain decompression for syringomyelia can lead to regression of scoliosis (22). Their series
comprised of 16 patients with syringomyelia who presented with significant scoliosis in the absence of major neurological deficit (22). All underwent a hindbrain decompression, and follow-up ranged from 1 to 6 years (22). In eight cases subsequent deformity surgery was necessary but in only six (37.5%) was the scoliosis seen to improve or arrest its progression (22). They stated that improvement was found to be statistically more likely in children of younger age at the time of syrinx surgery and in those with left thoracic curves (22). At the time of hindbrain decompression however, those under the age of 10 had improvement occur at a rate of 71.4%.

Conclusion

As we see from this report, whatever the intervention treatment of choice for ACM type I, scoliosis, or syringomyelia, the decisions are not always easy. Each anomaly carries with it a certain entity that makes treatment protocol a challenge. The main factors determining whether or not to pursue certain treatment options lie within the contraindications. Although the literature did not list any specific contraindications, each abnormality has certain dangers that should be considered. With ACM type I, the chiropractic practitioner should be warned of the potential exacerbation of problems with upper cervical HVLA adjusting. The condition has several literature publications with symptoms worsening after upper cervical HVLA maneuvers. Scoliosis, as read above, has many unpredictable patterns even after treatment intervention. The main concern with this process is to make sure that no neurological symptoms become worse and to try and halt the progression of the curve. Syringomyelia on the other hand was reported in connection with ACM type I and scoliosis throughout this review. Other similar reviews
have shown similar connections with syringomyelia and ACM type I (23, 24, 25). Any contraindications for ACM type I or scoliosis should also be considered when deriving a treatment plan for syringomyelia. For specific contraindications for any of the three, further research literature should be read and reviewed.

Although it was stated that many literature reviews were available pertaining to ACM type I, scoliosis, and syringomyelia and the possible connections amongst the three, only a few, as explained above, really lay out a foundation for possible pathogenic connections. Other reviews are available and other theories are outlined, but it is clear that more research into the subject needs to be conducted to gain full understanding. The anomalies are of importance to not only the neurosurgical doctors, chiropractors, and associated medical world alike, but also to the patients who suffer from these. Ultimately it is they (the patients) who will benefit the most from further research and explorations into the possible connections between the three. Not only would a huge hole in science be filled with the discovery of the definitive pathogenesis, but those individuals who suffer from these anomalies would regain control of their health on a neurological level; something that would be immeasurable to any science!

References:


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