Association of Increased Body Mass Index with Chiari Malformation Type I and Syrinx Formation in Adults

Arnautović, Kenan I.; Splavski, Bruno

J Neurosurg 119:1058–1067, 2013

Association of increased body mass index with Chiari malformation Type I and syrinx formation in adults

Clinical article

Kenan I. Arnautovic, M.D., Ph.D.,[1,2] Dario Muzevic, M.D.,[3] Bruno Splavski, M.D., Ph.D.,[3] and Frederick A. Boop, M.D.[1,2]

[1]Semmes-Murphey Clinic; [2]Department of Neurosurgery, University of Tennessee Health Science Center, Memphis, Tennessee; and [3]Department of Neurosurgery, Osijek University School of Medicine, Clinical Hospital Center, Osijek, Croatia

Object. In this paper the authors describe an association between increased body mass index (BMI) and Chiari malformation Type I (CM-I) in adults, as well as its relationship to the development of syringomyelia.

Methods. In the period between January 2004 and December 2011, the senior author reviewed the data for all CM-I patients with or without syringomyelia and neurological deficit. Analyzed factors included clinical status (headaches and neurological signs), radiological characteristics of syringomyelia (diameter and vertical extent of syrinx), BMI, and relationship of age to BMI, syrinx diameter, and vertical extent of syrinx.

Results. Sixty consecutive adults had CM-I, 26 of whom also had syringomyelia. The mean BMI among all patients was 30.35 ± 7.65, which is Class I obesity (WHO), and was similar among patients with or without syringomyelia. Extension of the vertical syrinx was greater in overweight patients ($p = 0.027$) than in those with a normal body weight. Evidence of de novo syrinx formation was found in 2 patients who gained an average BMI of 10.8 points. After repeated decompression and no change in holocord syrinx width or vertical extent, a reduction in the syrinx was seen after BMI decreased 11.7 points in one individual. No correlation was found between patient age and BMI, age and vertical extension of the syrinx, and age and diameter of the syrinx.

Conclusions. An association between increased BMI and CM-I in adults was recognized. Gaining weight may influence the de novo creation of a syrinx in adults who previously had minimally symptomatic or asymptomatic CM-I, and reducing weight can improve a syrinx after unsuccessful surgical decompression. Therefore, a reduction in body weight should be recommended for all overweight and obese patients with CM-I.
(http://thejns.org/doi/abs/10.3171/2013.4.JNS122011)

Key Words • **Chiari malformation Type I** • **adults** • **syringomyelia** • **increased body mass index** • **obesity** • **syrinx vertical extension**

Chiari malformation Type I as a hindbrain disorder was first described by an Austrian pathologist, Hans Chiari, in 1891.[47,48] It is mainly characterized by a peg-like deformity and elongation of the cerebellar tonsils and their descent of more than 5 mm below the foramen magnum into the spinal canal. This malformation occurs in both pediatric and adult patients. One of the essential elements in the pathophysiology of clinical symptoms in patients with CM-I is an altered flow of CSF at the level of the foramen magnum and its frequent association with the subsequent development of syringomyelia. Decompression of the foramen magnum, reestablishment of CSF flow across the craniocervical junction, and resolution of the syrinx are well-recognized goals of operative treatment in symptomatic patients with CM-I. This treatment usually resolves or at least stabilizes symptoms in the majority of patients.

To the best of our knowledge, there has been no previous study of the association between increased BMI and CM-I or the influence of an increasing BMI on the de novo formation of a spinal cord syrinx in adults. We have recognized, investigated, and analyzed this phenomenon in a series of adults with CM-I over the past 8 years. The goal of this paper was to bring to light this interesting and underrecognized association that may contribute to a better understanding of the pathogenesis and treatment of CM-I in adults.

Abbreviations used in this paper: BMI = body mass index; CM-I = Chiari malformation Type I; COPD = chronic obstructive pulmonary disease; IQR = interquartile range; KPS = Karnofsky Performance Status; VAS = visual analog scale.

Methods

This study was approved by the appropriate hospital institutional review board. Inclusion criteria for this retrospective analysis were as follows: adult CM-I patients with or without syringomyelia and neurological deficit whose data were reviewed by the senior author (K.I.A.) in the period between January 2004 and December 2011.

Patients were divided into 2 subgroups according to the presence or absence of syringomyelia. For the purposes of this paper, the subgroup of patients having syringomyelia was additionally analyzed. Neuroradiological diagnostics consisted of MRI of the brain, craniocervical junction, and cervical spine. For patients with syringomyelia, the vertical extension and width (horizontal diameter) of the syrinx were measured by an independent neuroradiologist. Measurement of the vertical extension included the starting and ending vertebral level and the total number of vertebral levels involved. The width of the syrinx was measured relative to the percentage of the largest anteroposterior spinal cord diameter affected by the syrinx, as previously reported in the literature.[5,24,29,30,76] The diameter of the syrinx was graded according to the assigned percent value in our newly designed system: Grade I (0%–25%), Grade II (26%–50%), Grade III (51%–75%), and Grade IV (76%–100%).

Severity of neurological deficit was determined based on each patient's KPS score.[38] Those with a KPS score of 85–100 were considered to have a mild neurological deficit, those with a score of 65–84 had a moderate deficit, and those with a score < 65 had a severe neurological deficit.

Data concerning a patient's body weight and BMI were analyzed. The BMI, which represents the ratio between the body weight calculated in kilograms and the square of the body height calculated in centimeters squared, was used as an indicator of obesity. Using the WHO criteria,[81] we divided all patients into categories based on their BMI values: underweight (BMI < 18.5), normal body weight (BMI 18.5–24.9), overweight (BMI 25–29.9), Class I obesity (BMI 30–34.9), Class II obesity (BMI 35–39.9), and Class III obesity (BMI > 40).

In the subgroup of patients with cervical syringomyelia, we investigated the possible correlations between vertical extension of the spinal cord syrinx and BMI, as well as between the syrinx-to-cord ratio and BMI. Furthermore, we recorded data concerning patients' clinical status, including the presence and intensity of headaches (VAS), as well as the presence and magnitude of neurological deficits (KPS score).

Additionally, all patients were evaluated for the presence of diabetes mellitus, which may influence neurological findings. Patients were also evaluated for the existence of papilledema to rule out associated pseudotumor cerebri. The possible influence of smoking and COPD on syrinx formation was also studied. Finally, any correlation between the age of patients and BMI or syringomyelia was also tested.

The frequency in differences of variables between groups was obtained using the chi-square test or Fisher exact test for normal distribution. To analyze differences between 2 independent groups, the Student t-test was used. To investigate differences between 2 dependent samples, the Wilcoxon test was used for nonparametric analysis. Results were then correlated with the Spearman coefficient of correlation (ρ). A 2-sided p value < 0.05 was considered statistically significant. The Statistica for Windows 2005 program package, Version 7.1 (StatSoft, Inc.), was used for statistical analysis.

Results

Demographic Information

Sixty adults with CM-I included 8 men (13.3%) and 52 women (86.7%), with a mean age of 37.5 ± 11.3 years (range 18–63 years). There was no difference between the mean ages of men and women (p = 0.887, t-test).

Syringomyelia was diagnosed in 26 patients (43.3%), whose mean age was 38.8 ± 13.2 years (Table 1). There was no age difference between the 2 sexes (p = 0.440, t-test) or between those with and those without syringomyelia (p = 0.427, t-test).

In the subgroup with syringomyelia, a female predominance was noted (18 [69.2%] of 26; p < 0.001, Fisher exact test; Table 2).

Headache Data

The intensity of headaches was evaluated using the VAS. Among the 60 patients, 7 (11.7%) did not have headaches on presentation. The remaining 53 patients (88.3%) filled out a VAS headache questionnaire, 33 (62.3%) without and 20 (37.7%) with syringomyelia. The mean VAS score was 9.8 ± 0.81. In patients without syringomyelia the VAS score was 9.7 ± 0.88, and in patients with syringomyelia it was 9.85 ± 0.67. The relationship between the 2 groups was not established (ρ = 0.099, Spearman correlation coefficient; p = 0.677).

Neurological Deficits

A neurological deficit was present in all but 4 patients (6.7%). According to the KPS, disability was severe in almost half the patients (25 patients [41.7%]), moderate in 16 (26.7%), and mild in 15 (25%; Fig. 1). In the syringomyelia subgroup, all patients had a neurological deficit and more patients had more pronounced neurological deficits. Nonetheless, the difference in the severity of

TABLE 1: Basic measures in patients with CM-I, according to sex and diagnosed syringomyelia

Parameter	No. of Patients	Mean Age*	p Value†
sex			
M	8	40.4 ± 17.2	0.440
F	52	37.02 ± 10.3	
syringomyelia			
yes	26	38.8 ± 13.2	0.427
no	34	36.7 ± 10.1	
total	60	37.5 ± 11.3	

* Values expressed as the means ± SDs.
† Calculated using the t-test.

TABLE 2: Patient sex distribution in the syringomyelia subgroup

Sex	Syringomyelia (no. [%])		Total	p Value*
	Yes	No		
M	8 (30.8)	0	8 (13.3)	0.001
F	18 (69.2)	34 (100)	52 (86.7)	
total	26 (100)	34 (100)	60 (100)	

* Calculated using the Fisher exact test.

disability in patients with and without syringomyelia did not reach significance (p = 0.115, chi-square test). In the syringomyelia subgroup, 3 patients (11.5%) had diabetes mellitus Type II.

Radiological Diagnostics: Width of the Syrinx

All patients demonstrated descent of the cerebellar tonsils more than 5 mm below the level of the foramen magnum, including partial or total obliteration of the cisterna magna and lateral cerebellomedullary cisterns. In 26 (43.3%) of the 60 patients, syringomyelia was diagnosed. Among these patients, 18 (69.2%) had a syrinx width of 76%–100% (Grade IV), 6 (23.1%) had a syrinx width of 26%–50% (Grade II), and 1 patient each (3.8%) had a syrinx width 0%–25% or 51%–75% (Grades I and III, respectively; Table 3).

Body Mass Index

The average BMI among all patients was 30.35 ± 7.65, which is within the Class I obesity range according to the WHO scale (BMI 30–34.9). The mean BMI was similar in patients with and without syringomyelia.

Among the entire series, only 1 patient (2.9%) in the subgroup without syringomyelia was underweight (BMI < 18.5). Fourteen patients (23.3%) had a normal BMI (18.5–24.9) and 15 (25%) were overweight (BMI 25–29.9). The remaining 30 patients (50%) were obese: 16 (26.7%) in the Class I obesity group (BMI 30–34.9), 8 (13.3%) in Class II (BMI 35–39.9), and 6 (10%) in Class III (BMI > 40). Fourteen patients (23.3%) were of normal weight, and the remaining 45 patients (75%) had increased (above normal) values of BMI. There was no difference in BMI between the subgroups with and without syringomyelia (p = 0.514; Fig. 2).

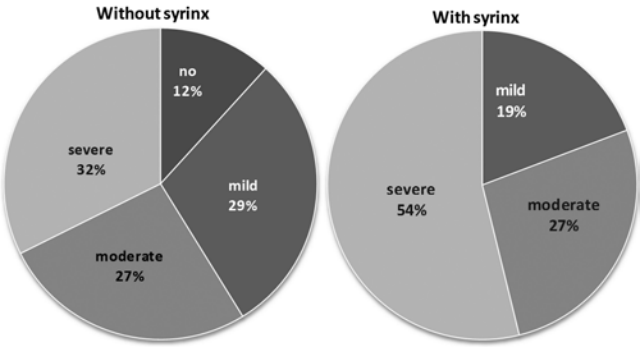

Fig. 1. Distribution of the severity of neurological deficits in patient subgroups (KPS scores: disability mild = 85–100; disability moderate = 65–84; disability severe = < 65).

TABLE 3: Distribution of patients based on the anteroposterior syrinx grade

Syrinx Width	No. (%)
Grade I (0%–25%)	1 (3.8)
Grade II (26%–50%)	6 (23.1)
Grade III (51%–75%)	1 (3.8)
Grade IV (76%–100%)	18 (69.2)
total	26 (100)

Relationship Between Increased BMI and Vertical Extent of Syrinx

The average vertical extent of the syrinx in our series was 8.2 vertebrae. In 13 patients (50%) the syrinx started at C-1, and it extended all the way to T-11 in 2 (7.7%) of these 13 patients. In 4 patients (15.4%) the syrinx began at the C-2 vertebra, and in 6 (23.1%) it began at C-3 (Fig. 3).

In the subgroup of patients with normal BMI values and syringomyelia, the most common vertical extent of the syrinx was 6 vertebrae (IQR 5–18). In the subgroup of overweight patients, the syrinx most commonly extended over 12 vertebrae (IQR 9.5–13.5). In the subgroup of obese patients, the syrinx most often extended over 5.5 vertebrae (IQR 4–10; Table 4). Patients with an increased BMI most often had a vertical extent of 12 vertebrae.

The difference in the vertical extent of the syrinx between patients with normal and those with increased BMI was significant (p = 0.027, Kruskal-Wallis test). The median vertical extent of the syrinx in overweight patients (12 vertebrae) was twice that in patients with a normal BMI (6 vertebrae). We used the Mann-Whitney test to analyze the differences in syrinx vertical extent between normal-weight and overweight patients; the obtained p value was 0.326. The difference between normal-weight and obese patients yielded a p value of 0.259, and the difference between overweight and obese patients yielded a p value of 0.005.

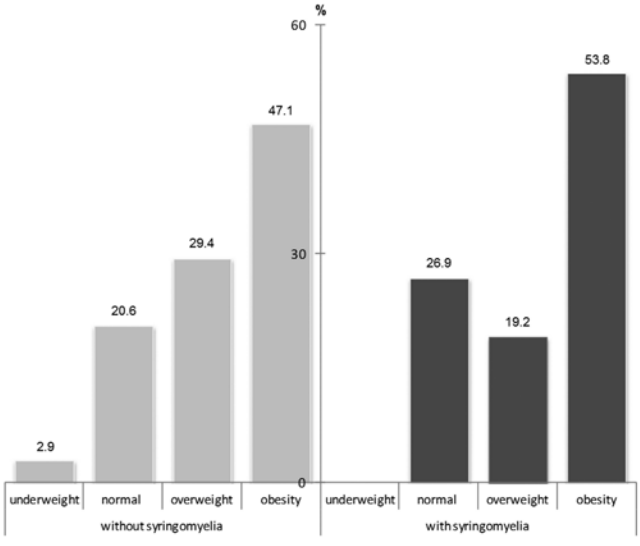

Fig. 2. The distribution of BMI in patients: subgroup without syringomyelia **(left)** and subgroup with syringomyelia **(right)**.

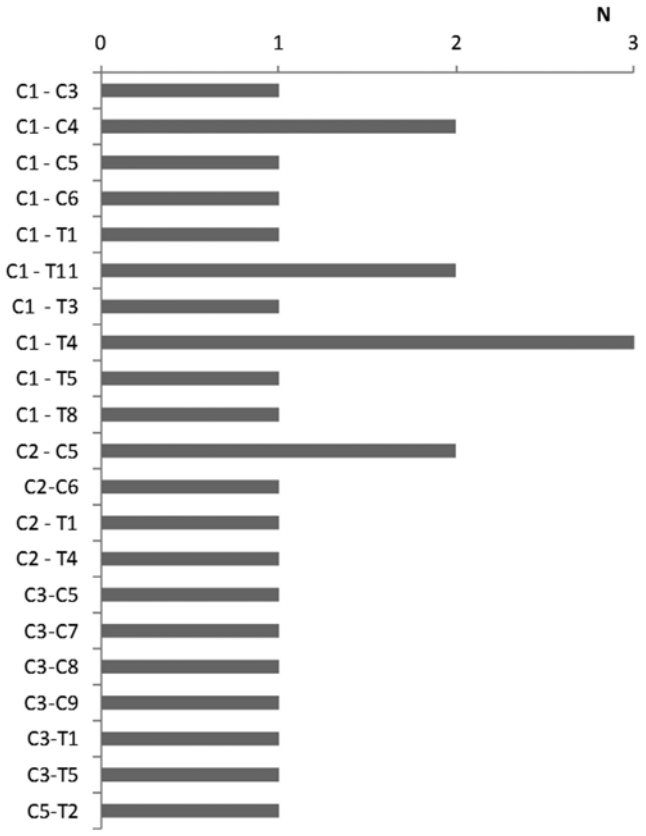

Fig. 3. Distribution of patients with syringomyelia, according to the vertical (vertebral) extent of the syrinx. N = number of patients.

Relationship Between Increased BMI and Width of the Syrinx

The mean width of the syrinx in the syringomyelia subgroup was 90% (range 50%–90%). A diagram of the correlation between BMI values and the width of the syrinx in the syringomyelia subgroup is shown in Fig. 4. The patients with a higher BMI had wider syringes. The correlational trend showed that with the increase in BMI, a simultaneous increase in the anteroposterior diameter of the syrinx followed. Using the equation anteroposterior diameter = (0.1414 × BMI) + 72.87, we determined that for every unit of BMI increase, there was a 0.14% increase in the width of the syrinx. In addition, there was significant dispersion of BMI values in the relatively small dispersion of the width of the syrinx in the syringomyelia subgroup as a whole. In the subgroup of patients with an increased BMI (BMI > 25), there was a noted increase in syrinx width. The correlation between the width of the syrinx and an increased BMI was not significant ($\rho = 0.018$, Spearman correlation coefficient).

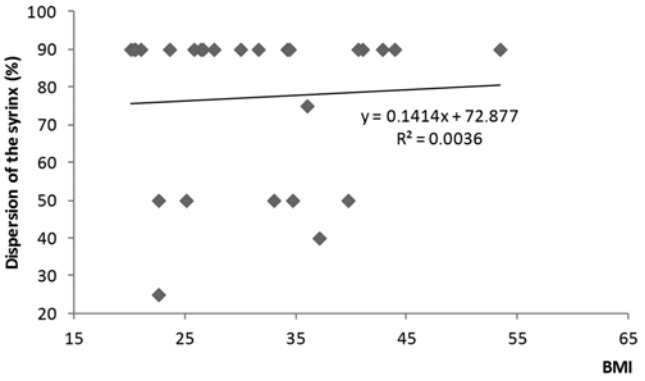

Fig. 4. Correlation between syrinx width and BMI values (syringomyelia subgroup).

Influence of Increased BMI on De Novo Appearance of Syrinx

In the syringomyelia subgroup, we identified 2 patients who had de novo formation of a syrinx upon a significant increase in their BMI. One patient had an incidentally found CM-I that was accompanied by mild headaches and mild neurological symptoms but no signs of a syrinx. Her BMI was 28.5, putting her in the overweight category (Fig. 5 left). Four years later, she still had a CM-I but she demonstrated a syrinx as well (Fig. 5 right). By that time, her BMI had significantly increased to 41 (12.5 BMI points), which is Class III obesity. Furthermore, her headaches and neurological function had worsened.

A CM-I without a syrinx was diagnosed in a second patient at a time when her BMI was 19 and she had only mild headaches. She had 3 subsequent pregnancies in 3 consecutive years, and her BMI increased to 28 (9 BMI points). During that time, her headaches worsened and her normal neurological status deteriorated.

An additional patient presented with a CM-I and holocord syrinx (BMI 45.3, Class III obesity; Fig. 6A). Two years later, she underwent classic suboccipital Chiari decompression. Her symptoms and syrinx persisted despite the decompression and even progressed when her BMI increased to 46.3 (Fig. 6B and C). She underwent a second operation a year later, and no significant residual compression was found. Postoperatively, her symptoms did not improve; nor did her syrinx reduce in size. Her BMI further increased to 47.7. At that time, she was counseled

TABLE 4: Vertical extent of the syrinx among different BMI groups

BMI	No. of Patients	Mean No. of Vertebrae (range)*	Median No. of Vertebrae	IQR (25%–75%)	p Value†
normal	7	9.5 ± 6.2 (4–18)	6	5–18	0.027
overweight	5	11.6 ± 2.5 (8–15)	12	9.5–13.5	
obese	14	6.4 ± 2.9 (3–11)	5.5	4–10	
total	26	8.2 ± 4.4 (3–18)	7	4.75–11	

* Values expressed as means ± SDs.
† Kruskal-Wallis test.

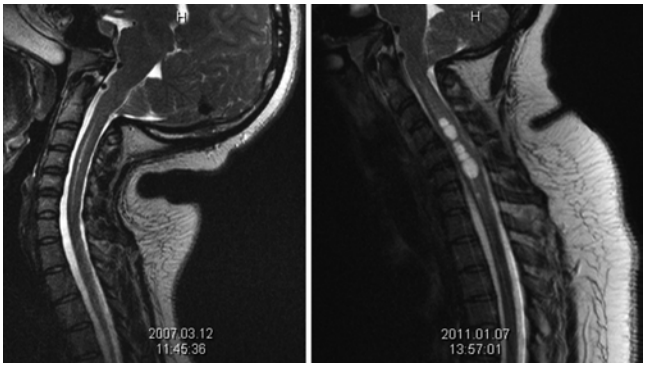

Fig. 5. Sagittal T2-weighted MR images of the cervical spine. The patient's BMI was initially 28.5 **(left)**; 4 years later, it was 41 **(right)**.

in a weight-loss program. Two years later she underwent gastric bypass surgery. Over the next 6 months, she lost significant weight, approximately 11 BMI points (BMI 36, Class II obesity). Follow-up MRI showed a noticeable reduction in the vertical extension and anteroposterior diameter of the syrinx, and her clinical status improved (Fig. 6D).

Correlation of Patient Age With BMI and Syringomyelia

We investigated the correlation between BMI values and patient age to determine whether BMI values increase with age (Fig. 7). No correlation between these 2 variables was identified (Spearman correlation coefficient, $\rho = 0.191$, $p = 0.82$).

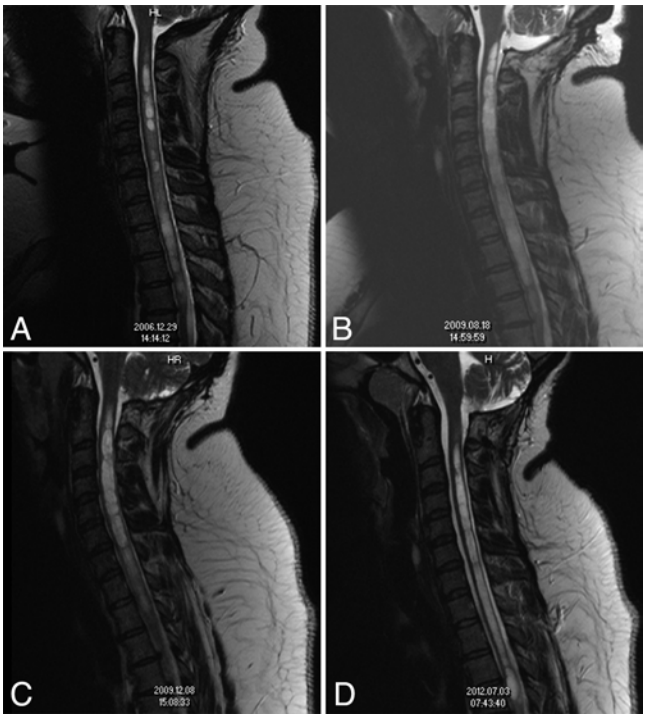

Fig. 6. Sagittal T2-weighted MR images of the cervical spine. **A:** BMI = 45.3. **B and C:** BMI = 46.3. **D:** BMI = 36. This patient presented with holocord syrinx, which, despite 2 occipital decompressions, persisted over 5 years. Within 6 months of weight reduction surgery, her syrinx diminished in size. The correlation between weight reduction and syrinx size decrease suggests a causal relationship.

We also evaluated whether any correlation existed between patient age and syrinx size to see if the width (anteroposterior diameter of the syrinx) and vertical extent of the syrinx increased with age. We did not find a correlation between patient age and vertical extent of the syrinx (Spearman correlation coefficient, $\rho = -0.059$, $p = 0.776$). In addition, we did not find any correlation between patient age and syrinx width (Spearman correlation coefficient, $\rho = 0.112$, $p = 0.585$).

Smoking and COPD as Possible Risk Factors for Syringomyelia

Smoking was recorded in 14 (23.3%) of the 60 patients, in 10 (29.4%) of the 34 without syringomyelia, and in 4 (15.4%) of the 26 with syringomyelia. There was no association between smoking and the presence of syringomyelia.

Discussion

Association of Increased BMI With CM-I in Adults

Chiari malformation Type I is a hindbrain disorder mainly characterized by the caudal descent of cerebellar tonsils more than 5 mm below the level of the foramen magnum into the spinal canal and the obliteration of dorsal and lateral cerebellomedullary cisterns. It is the leading cause of syringomyelia.[17,48,49,60] It occurs in both children and adults and has been reported frequently in the past 50 years.[2,4–6,9–11,13,14,16,20–22,24,25,28,30,33,37,39,43–45,58,60–66,70,71,74–79] Its incidence in the general population has been estimated at 3.5%,[47] making it a fairly common presenting diagnosis or incidental MRI finding. Approximately 30% of individuals with CM-I are asymptomatic, and approximately 25% of patients claim trauma (whiplash, spinal injuries, or direct blows to the head and neck) as the precipitating event.[48,49] Other precipitating factors that may convert an asymptomatic CM-I to a symptomatic one or create a de novo syrinx in previously asymptomatic patients are poorly understood and largely unknown.

To the best of our knowledge, the association of increased BMI with CM-I in adults has not been studied or reported before. Our series is quite similar to others[2,4–6,9–11,13,14,16,20–22,24,25,28,30,33,37,39,43–45,58,60–66,70,71,74–79] in all demographic parameters (prevalence of sex, mean age, association of syringomyelia; Tables 1 and 2), and the aver-

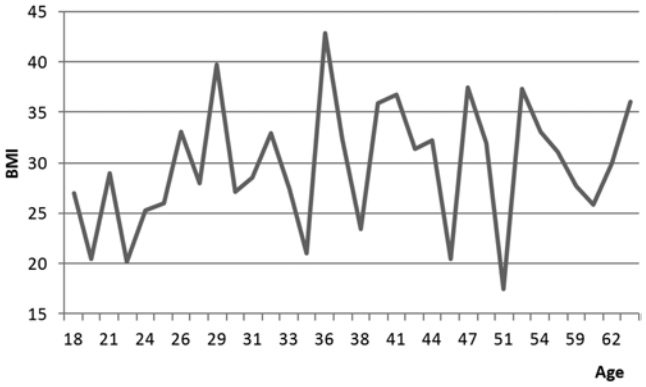

Fig. 7. Correlation between patient age (years) and BMI.

age BMI in all patients was 30.35 ± 7.65. This value was within the range of Class I obesity according to the WHO scale.[81] Furthermore, only 1 patient (1.7%) in the entire series was underweight, 14 patients (23.3%) had a normal BMI, and the remaining 45 patients (75%) were above normal weight, 30 (50%) of whom were obese. This increased weight proportion did not significantly differ between patients with or without syringomyelia (Fig. 2). Considering this, our data suggest an association between increased BMI values and CM-I in adults. In addition, among patients with an increased BMI, obese patients were more numerous than overweight patients by a margin of 2:1. One can speculate why an increased BMI and obesity in particular are associated with CM-I in our patients and possibly with all other adults in studies of patients with CM-I.

Increased CSF pressure has been associated with obese patients with idiopathic intracranial hypertension (pseudotumor cerebri)[23,40,50,56,57,68,69] and CM-I in children.[42] The obliteration of subarachnoid spaces around the foramen magnum is one of the essential elements in the pathophysiology of clinical symptoms associated with CM-I and its frequent association with syringomyelia.[48]

The classic works of Batson,[8] Hamilton and colleagues,[31,32] and Herlihy[34] established that there is a transmission of intrathoracic and intraabdominal pressures toward the spinal and cranial compartments and further to the CSF compartment at rest and with Valsalva maneuvering (coughing, sneezing, strenuous activities, posture changes, and so forth). This happens in real time because of the valveless connections between corresponding thoracic, abdominal, and vertebral venous compartments.[3] Based on studies of obese patients, Sugerman[68] and colleagues[69] coined the term "chronic abdominal compartment syndrome" after reporting on the increases in intraabdominal, intracranial, venous, and cardiac filling pressures in these patients. Therefore, one can hypothesize that in adult CM-I patients with increased BMI, the pressure transmission that normally exists between the intrathoracic/intraabdominal and vertebral valveless venous compartments is exaggerated because of the increased BMI. This exaggeration worsens the pressure gradient between the cranial and spinal compartments and their possibility of equalization, both of which are already altered by the CM-I syndrome. In turn, this phenomenon could precipitate the symptomatic presentation of a CM-I. Considering these possibilities, we advise warning patients with an incidentally diagnosed asymptomatic CM-I syndrome about the importance of strictly regulating their body weight. Furthermore, we think it advisable to warn female patients with minimally symptomatic or asymptomatic CM-I that increasing their weight in pregnancy may lead to a syrinx and progression to a symptomatic condition.

Unfortunately, it is unknown how the BMI data in our series compare with those in other studies of CM-I in adults.[2,4–6,9–11,13,14,16,17,20–22,24,25,28,30,33,37,39,43–45,58,60–67,70,71,74–77,82,83] In the past 50 years, most published studies have come from North America,[1,2,6,9,11,14,17,18,20,22,27,33,43,46,54,60,63,73] followed by Europe[5,10,12,15,21,25,26,30,39,44,45,58,59,61,62,65–67,71,74–77,82] and Asia,[4,24,28,35–37,64,70,72,83] but very rarely come from Africa[19] and South America.[13] One can speculate why such differences occur in the continental and regional frequencies of reported series of adults with CM-I; the level of medical services available and economic factors come first to mind. We wonder whether the existence of different BMI values among patients in different regions and continents has influenced more frequent symptomatic presentations of these patients and subsequently more frequent reporting.

It is clear that obesity is an increasing phenomenon in the US, and some regions may have a higher percentage of patients with obesity and CM-I. Further studies are needed to explore such correlations.

Effect of Increasing BMI on De Novo Syrinx Formation

Chiari malformation Type I is the leading cause of syringomyelia,[17,48,49,60] and the pathogenesis of syringomyelia formation was initially established by 2 classic hypotheses by Gardner[27] and Williams.[78–80] Over time, several authors have offered additional hypotheses.[7,41] Milhorat and colleagues[48,49] did experimental work on syringomyelia and the central spinal canal. More recently, Heiss et al.[33] and Oldfield et al.[55] proposed new insights into the pathophysiology of syringomyelia and its implications for the diagnosis and treatment of CM-I.

We have established clear evidence of the de novo formation of a syrinx in 2 patients in our series who initially had a verified CM-I and minimal or no associated symptoms. Over the course of 3 and 4 years, respectively, both patients had an increase in their BMI by 9 and 12.5 points, respectively. The first patient progressed from the overweight category to Class III obesity and the second from a normal to overweight as a result of pregnancy. A new syrinx, which had not been present on initial MRI, developed in both (Fig. 5). We believe that this phenomenon of an increasing BMI influencing the de novo formation of a syrinx further supports the pathophysiological mechanism proposed by Heiss et al.[33] and Oldfield et al.[55] According to these authors, with obstruction of the rapid to-and-fro movement of CSF in the subarachnoid spaces across the foramen magnum during systole and diastole, the cerebellar tonsils, which plug subarachnoid spaces posteriorly, move down with each systolic pulse and act as a piston on the partially isolated spinal CSF compartment. This movement then produces a systolic pressure wave in the spinal CSF that acts on the surface of the spinal cord, causing a progression of the syrinx from the pulsatile pressure waves forcing CSF into the cord through the perivascular and interstitial spaces. Therefore, we can hypothesize that an increasing BMI in patients can only aggravate and exaggerate this process and create or worsen clinical symptoms. Patients with an incidentally found CM-I who are contemplating pregnancy should be cautioned about the potential to experience symptoms and a de novo syrinx with an increasing BMI.

One may argue that a person's weight naturally increases with age. Consequently, because most CM-I are congenital, given enough time, a syrinx will eventually develop and progress in these patients. Therefore, we looked into the correlation between BMI values and age in our patients (Fig. 7). We also looked into the possible correlation between age and vertical extension of the syr-

inx as well as any correlation between age and the width (anteroposterior diameter) of the syrinx. No correlation was found in any of the 3 comparisons. This may further support our hypothesis of the influence of increasing BMI (gaining weight) on de novo syrinx formation and no influence of increasing age on this process.

In the treatment of idiopathic intracranial hypertension (pseudotumor cerebri), weight control via bariatric surgery has been reported to be successful.[23,51] Chiari malformation Type I patients with increased BMI and no or only minimal symptoms may also be advised to strictly control their weight as an initial treatment modality. We can also hypothesize that losing weight in adults with CM-I may influence the improvement of a syrinx. We observed this phenomenon in the patient featured in Fig. 7, who had a CM-I and holocord syrinx and whose BMI increased to Class III obesity. Her symptoms failed to resolve after the classic suboccipital decompression and subsequent reexploration. It was only through weight reduction of about 11 BMI points (from Class III to Class II obesity) that she started to experience noticeable improvement. In our experience, weight gain or loss of an average of 10.8 BMI points made a difference in 3 patients, 2 with de novo syrinx formation and 1 with improvement after failed decompression and reexploration.

One could argue that in the examples mentioned above, the syrinx oscillations reflect the natural history of the disease and are not related to the changes of the patients' weight. However, we have demonstrated a positive correlation between these variables with both increasing and decreasing weight.

Influence of Increased BMI on Vertical Extent of Syrinx

The average vertical extent of the syrinx in the patients with syringomyelia in our series was 8.2 vertebrae. Most commonly, the syrinx started at the C-1 level (in 13%–50% of patients with a syrinx; Fig. 3). Patients with an increased BMI most commonly had a syrinx whose vertical extent was 12 vertebrae. In half the patients with an increased BMI, vertical extension ranged from 9.5 to 13.5 vertebral bodies, while it was only 6 vertebral bodies in those whose BMI values were normal. The difference in the range of vertical extension between the groups with normal and increased BMI in our series was significant. Simultaneously, the range of vertical extension positively correlated with the increased BMI values. Based on results of the present study, we can establish the certain influence of increased BMI on the vertical extent of the syrinx. Because of the small number of patients, this influence is not statistically very strong, but it did exist (Table 4). This possible proportional influence of increased BMI on the vertical extent of the syrinx further supports the proposed mechanism; that is, that the origin and maintenance of syringomyelia is facilitated by pulsatile pressure waves forcing CSF into the spinal cord through perivascular and interstitial spaces.[33,55] We also looked into the possible correlation of age with vertical extension of the syrinx; however, no statistical correlation was found.

Influence of Increased BMI on the Width of the Syrinx

The mean width (transversal, or anteroposterior, diameter) of the syrinx in patients in our series was 90%. We introduced a new classification system for syringomyelia, consisting of 4 grades of syrinx width. This was done to more easily describe and assess the severity of syringomyelia and the size of the syrinx: Grade I (0%–25%), Grade II (26%–50%), Grade III (51%–75%), and Grade IV (76%–100%). We believe that this grading system simplifies the assessment of syrinx size and syringomyelia severity (Table 3). Furthermore, it makes possible the comparison of pre- and postoperative syrinx dimensions in each patient, between different syringomyelia patient subgroups, and between different patient series. We also looked into the possible correlation of age with syrinx width (anteroposterior diameter), but found none.

Patients with a higher BMI had wider syrinxes. The correlational trend shows that with the increased BMI, there was a simultaneous increase in the anteroposterior diameter of the syrinx. Using the regression equation anteroposterior diameter = (0.1414 × BMI) + 72.87, we can conclude that for every BMI unit increase, the width of the syrinx increases 0.14% (Fig. 4).

We also noted that in the syringomyelia subgroup as a whole, there was significant dispersion of BMI values in the relatively small dispersion of the width of the syrinx. In the subgroup of patients with a BMI > 25, there was a noted increase in syrinx width. However, the correlation between syrinx width and increased BMI was not significant ($\rho = 0.018$, Spearman correlation coefficient; Fig. 4).

Headaches and Neurological Deficit

In our series, as in other studies published in the literature,[2,4–6,9–11,13,14,16,17,20–22,24,25,28,30,33,37,39,43–45,58,60–67,70,71,74–79] headaches were present in the majority of patients (88.3%). Also as in other reported series,[2,4–6,9–11,13,14,16,17,20–22,24,25,28,30,33,37,39,43–45,58,60–67,70,71,74–79] a neurological deficit was present in 93% of patients in our series (88% of patients without and 100% of patients with syringomyelia). Moderate and severe neurological deficits were present in 59% of patients without and in 81% of those with syringomyelia. However, this difference between the 2 subgroups was not significant (Fig. 1). Furthermore, our results correspond with those of other authors[2,4,5,11,24,29,30,52,53,76] in that the anteroposterior size and vertical extent of the syrinx were not proportional to the neurological deficit.

Since only 3 patients (11.5%) in the syringomyelia subgroup had diabetes mellitus Type II, it is unlikely that the majority of neurological symptoms could be correlated to diabetic neuropathy. Furthermore, our results have also shown that smoking and COPD did not have a statistically significant influence on neurological findings. Finally, recognizing that patients with increased BMI also have increased CSF pressure, we did not find evidence of papilledema and pseudotumor in any of the patients in our series.

Conclusions

We have recognized a positive association between increased BMI and the pathogenesis of CM-I and syringomyelia in adults. An increasing BMI (gaining weight) may influence the de novo creation of a syrinx in adults

who previously had minimally symptomatic or asymptomatic CM-I. Patients should be counseled to control their weight, and asymptomatic or mildly symptomatic women who plan to become pregnant should be warned of the possibility of a de novo syrinx with weight gain. Furthermore, patients with CM-I, syringomyelia, and increased BMI and in whom classic suboccipital decompression has failed should consider weight reduction. Asymptomatic or mildly symptomatic patients may be counseled for a strict weight loss program as the initial treatment.

This is a retrospective study done at a single center with all pertinent limitations; therefore, our findings should be considered preliminary. Further studies are needed for many reasons: to determine results in a larger number of patients, to prospectively analyze the BMI association with CM-I, to perform multicenter studies, and to analyze the possible influence of increased BMI on the formation of syringomyelia, the width and vertical extent of the syrinx, and symptomatic presentation in adults with a CM-I.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Arnautovic, Splavski. Acquisition of data: Arnautovic. Analysis and interpretation of data: Arnautovic, Splavski, Boop. Drafting the article: Arnautovic, Splavski. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Arnautovic. Statistical analysis: Arnautovic. Administrative/technical/material support: Arnautovic, Muzevic. Study supervision: Arnautovic.

Acknowledgments

The authors thank Ms. Kristina Kralik for her biostatistics expertise and Julie Yamamoto and Andrew J. Gienapp for editorial assistance.

References

1. Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, et al: A novel scoring system for assessing Chiari malformation type I treatment outcomes. **Neurosurgery 70:**656–665, 2012
2. Alzate JC, Kothbauer KF, Jallo GI, Epstein FJ: Treatment of Chiari I malformation in patients with and without syringomyelia: a consecutive series of 66 cases. **Neurosurg Focus 11(1):**E3, 2001
3. Arnautović KI, al-Mefty O, Pait TG, Krisht AF, Husain MM: The suboccipital cavernous sinus. **J Neurosurg 86:**252–262, 1997
4. Arora P, Pradhan PK, Behari S, Banerji D, Das BK, Chhabra DK, et al: Chiari I malformation related syringomyelia: radionuclide cisternography as a predictor of outcome. **Acta Neurochir (Wien) 146:**119–130, 2004
5. Attal N, Parker F, Tadié M, Aghakani N, Bouhassira D: Effects of surgery on the sensory deficits of syringomyelia and predictors of outcome: a long term prospective study. **J Neurol Neurosurg Psychiatry 75:**1025–1030, 2004
6. Badie B, Mendoza D, Batzdorf U: Posterior fossa volume and response to suboccipital decompression in patients with Chiari I malformation. **Neurosurgery 37:**214–218, 1995
7. Ball MJ, Dayan AD: Pathogenesis of syringomyelia. **Lancet 2:**799–801, 1972
8. Batson OV: The function of the vertebral veins and their role in the spread of metastases. **Ann Surg 112:**138–149, 1940
9. Batzdorf U: Chiari I malformation with syringomyelia. Evaluation of surgical therapy by magnetic resonance imaging. **J Neurosurg 68:**726–730, 1988
10. Bidziński J: Late results of the surgical treatment of syringomyelia. **Acta Neurochir Suppl (Wien) 43:**29–31, 1988
11. Bindal AK, Dunsker SB, Tew JM Jr: Chiari I malformation: classification and management. **Neurosurgery 37:**1069–1074, 1995
12. Blagodatsky MD, Larionov SN, Alexandrov YA, Velm AI: Surgical treatment of Chiari I malformation with or without syringomyelia. **Acta Neurochir (Wien) 141:**963–968, 1999
13. Caetano de Barros M, Farias W, Ataíde L, Lins S: Basilar impression and Arnold-Chiari malformation. A study of 66 cases. **J Neurol Neurosurg Psychiatry 31:**596–605, 1968
14. Cahan LD, Bentson JR: Considerations in the diagnosis and treatment of syringomyelia and the Chiari malformation. **J Neurosurg 57:**24–31, 1982
15. Di Lorenzo N, Palma L, Palatinsky E, Fortuna A: "Conservative" cranio-cervical decompression in the treatment of syringomyelia-Chiari I complex. A prospective study of 20 adult cases. **Spine (Phila Pa 1976) 20:**2479–2483, 1995
16. Dones J, De Jesús O, Colen CB, Toledo MM, Delgado M: Clinical outcomes in patients with Chiari I malformation: a review of 27 cases. **Surg Neurol 60:**142–148, 2003
17. Dyste GN, Menezes AH, VanGilder JC: Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. **J Neurosurg 71:**159–168, 1989
18. Eisenstat DDR, Bernstein M, Fleming JFR, Vanderlinden RG, Schutz H: Chiari malformation in adults: a review of 40 cases. **Can J Neurol Sci 13:**221–228, 1986
19. El-Ghandour NMF: Long-term outcome of surgical management of adult Chiari I malformation. **Neurosurg Rev 35:**537–547, 2012
20. Ellenbogen RG, Armonda RA, Shaw DWW, Winn HR: Toward a rational treatment of Chiari I malformation and syringomyelia. **Neurosurg Focus 8(3):**Article 6, 2000
21. Ergün R, Akdemir G, Gezici AR, Tezel K, Beskonakli E, Ergüngör F, et al: Surgical management of syringomyelia-Chiari complex. **Eur Spine J 9:**553–557, 2000
22. Fischer EG: Posterior fossa decompression for Chiari I deformity, including resection of the cerebellar tonsils. **Childs Nerv Syst 11:**625–629, 1995
23. Fridley J, Foroozan R, Sherman V, Brandt ML, Yoshor D: Bariatric surgery for the treatment of idiopathic intracranial hypertension. A review. **J Neurosurg 114:**34–39, 2011
24. Fujii K, Natori Y, Nakagaki H, Fukui M: Management of syringomyelia associated with Chiari malformation: comparative study of syrinx size and symptoms by magnetic resonance imaging. **Surg Neurol 36:**281–285, 1991
25. Gambardella G, Caruso G, Caffo M, Germanò A, La Rosa G, Tomasello F: Transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression as treatment for syringomyelia with Chiari I malformation. **Acta Neurochir (Wien) 140:**134–139, 1998
26. Garcìa-Uria J, Leunda G, Carrillo R, Bravo G: Syringomyelia: long-term results after posterior fossa decompression. **J Neurosurg 54:**380–383, 1981
27. Gardner WJ: Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. **J Neurol Neurosurg Psychiatry 28:**247–259, 1965
28. Goel A, Bhatjiwale M, Desai K: Basilar invagination: a study based on 190 surgically treated patients. **J Neurosurg 88:** 962–968, 1998
29. Grant R, Hadley DM, Macpherson P, Condon B, Patterson J, Bone I, et al: Syringomyelia: cyst measurement by magnetic

resonance imaging and comparison with symptoms, signs and disability. **J Neurol Neurosurg Psychiatry 50:**1008–1014, 1987
30. Guyotat J, Bret P, Jouanneau E, Ricci AC, Lapras C: Syringomyelia associated with type I Chiari malformation. A 21-year retrospective study on 75 cases treated by foramen magnum decompression with a special emphasis on the value of tonsils resection. **Acta Neurochir (Wien) 140:**745–754, 1998
31. Hamilton WF, Woodbury RA, Harper HT Jr: Arterial, cerebrospinal and venous pressures in man during cough and strain. **Am J Physiol 141:**42–50, 1944
32. Hamilton WF, Woodbury RA, Harper HT Jr: Physiologic relationships between intrathoracic, intraspinal and arterial pressures. **JAMA 107:**853–856, 1936
33. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, et al: Elucidating the pathophysiology of syringomyelia. **J Neurosurg 91:**553–562, 1999
34. Herlihy WF: Revision of the venous system; the role of the vertebral veins. **Med J Aust 1:**661–672, 1947
35. Hida K, Iwasaki Y: Syringosubarachnoid shunt for syringomyelia associated with Chiari I malformation. **Neurosurg Focus 11(1):**Article 7, 2001
36. Hida K, Iwasaki Y, Koyanagi I, Sawamura Y, Abe H: Surgical indication and results of foramen magnum decompression versus syringosubarachnoid shunting for syringomyelia associated with Chiari I malformation. **Neurosurgery 37:**673–679, 1995
37. Isu T, Sasaki H, Takamura H, Kobayashi N: Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. **Neurosurgery 33:**844–850, 1993
38. Karnofsky DA, Burchenal JH: The clinical evaluation of chemotherapeutic agents in cancer, in MacLeod CM (ed): **Evaluation of Chemotherapeutic Agents.** New York: Columbia University Press, 1949, pp 191–205
39. Klekamp J, Batzdorf U, Samii M, Bothe HW: The surgical treatment of Chiari I malformation. **Acta Neurochir (Wien) 138:**788–801, 1996
40. Ko MW: Idiopathic intracranial hypertension. **Curr Treat Options Neurol 13:**101–108, 2011
41. Koyanagi I, Houkin K: Pathogenesis of syringomyelia associated with Chiari type 1 malformation: review of evidences and proposal of a new hypothesis. **Neurosurg Rev 33:**271–285, 2010
42. Kurschel S, Maier R, Gellner V, Eder HG: Chiari I malformation and intra-cranial hypertension: a case-based review. **Childs Nerv Syst 23:**901–905, 2007
43. Levy WJ, Mason L, Hahn JF: Chiari malformation presenting in adults: a surgical experience in 127 cases. **Neurosurgery 12:**377–390, 1983
44. Logue V, Edwards MR: Syringomyelia and its surgical treatment—an analysis of 75 patients. **J Neurol Neurosurg Psychiatry 44:**273–284, 1981
45. Matsumoto T, Symon L: Surgical management of syringomyelia—current results. **Surg Neurol 32:**258–265, 1989
46. McGirt MJ, Nimjee SM, Floyd J, Bulsara KR, George TM: Correlation of cerebrospinal fluid flow dynamics and headache in Chiari I malformation. **Neurosurgery 56:**716–721, 2005
47. Meadows J, Guarnieri M, Miller K, Haroun R, Kraut M, Carson BS: Type I Chiari malformation: a review of literature. **Neurosurg Q 11:**220–229, 2001
48. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al: Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. **Neurosurgery 44:**1005–1017, 1999
49. Milhorat TH, Nobandegani F, Miller JI, Rao C: Noncommunicating syringomyelia following occlusion of central canal in rats. Experimental model and histological findings. **J Neurosurg 78:**274–279, 1993
50. Mrejen S, Vignal C, Bruce BB, Gineys R, Audren F, Preechawat P, et al: Idiopathic intracranial hypertension: a comparison between French and North-American white patients. **Rev Neurol (Paris) 165:**542–548, 2009
51. Nadkarni T, Rekate HL, Wallace D: Resolution of pseudotumor cerebri after bariatric surgery for related obesity. Case report. **J Neurosurg 101:**878–880, 2004
52. Nishikawa M, Sakamoto N, Hakuba A, Nakanishi N, Inoue Y: Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. **J Neurosurg 86:**40–47, 1997
53. Nishizawa S, Yokoyama T, Yokota N, Tokuyama T, Ohta S: Incidentally identified syringomyelia associated with Chiari I malformations: is early interventional surgery necessary? **Neurosurgery 49:**637–641, 2001
54. Nohria V, Oakes WJ: Chiari I malformation: a review of 43 patients. **Pediatr Neurosurg 16:**222–227, 1990–1991
55. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ: Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. **J Neurosurg 80:**3–15, 1994
56. Ooi LY, Walker BR, Bodkin PA, Whittle IR: Idiopathic intracranial hypertension: can studies of obesity provide the key to understanding pathogenesis? **Br J Neurosurg 22:**187–194, 2008
57. Pakosova E, Timarova G, Krivosik M, Lisa I, Martinkova JK, Kukumberg P: Idiopathic intracranial hypertension: a retrospective clinical study. **Bratisl Lek Listy (Tlacene Vyd) 112:** 691–694, 2011
58. Paul KS, Lye RH, Strang FA, Dutton J: Arnold-Chiari malformation. Review of 71 cases. **J Neurosurg 58:**183–187, 1983
59. Perrini P, Benedetto N, Tenenbaum R, Di Lorenzo N: Extra-arachnoidal cranio-cervical decompression for syringomyelia associated with Chiari I malformation in adults: technique assessment. **Acta Neurochir (Wien) 149:**1015–1023, 2007
60. Pillay PK, Awad IA, Little JR, Hahn JF: Symptomatic Chiari malformation in adults: a new classification based on magnetic resonance imaging with clinical and prognostic significance. **Neurosurgery 28:**639–645, 1991
61. Pinna G, Alessandrini F, Alfieri A, Rossi M, Bricolo A: Cerebrospinal fluid flow dynamics study in Chiari I malformation: implications for syrinx formation. **Neurosurg Focus 8(3):**Article 3, 2000
62. Raftopoulos C, Sanchez A, Matos C, Balériaux D, Bank WO, Brotchi J: Hydrosyringomyelia-Chiari I complex. Prospective evaluation of a modified foramen magnum decompression procedure: preliminary results. **Surg Neurol 39:**163–169, 1993
63. Saez RJ, Onofrio BM, Yanagihara T: Experience with Arnold-Chiari malformation, 1960 to 1970. **J Neurosurg 45:**416–422, 1976
64. Sakamoto H, Nishikawa M, Hakuba A, Yasui T, Kitano S, Nakanishi N, et al: Expansive suboccipital cranioplasty for the treatment of syringomyelia associated with Chiari malformation. **Acta Neurochir (Wien) 141:**949–961, 1999
65. Sakas DE, Korfias SI, Wayte SC, Beale DJ, Papapetrou KP, Stranjalis GS, et al: Chiari malformation: CSF flow dynamics in the craniocervical junction and syrinx. **Acta Neurochir (Wien) 147:**1223–1233, 2005
66. Sindou M, Chávez-Machuca J, Hashish H: Cranio-cervical decompression for Chiari type I-malformation, adding extreme lateral foramen magnum opening and expansile duroplasty with arachnoid preservation. Technique and long-term functional results in 44 consecutive adult cases—comparison with literature data. **Acta Neurochir (Wien) 144:**1005–1019, 2002
67. Sindou M, Gimbert E: Decompression for Chiari type I-malformation (with or without syringomyelia) by extreme lateral foramen magnum opening and expansile duraplasty with arachnoid preservation: comparison with other technical modalities (Literature review). **Adv Tech Stand Neurosurg 34:**85–110, 2009

68. Sugerman HJ: Effects of increased intra-abdominal pressure in severe obesity. **Surg Clin North Am 81:**1063–1075, vi, 2001
69. Sugerman HJ, DeMaria EJ, Felton WL III, Nakatsuka M, Sismanis A: Increased intra-abdominal pressure and cardiac filling pressures in obesity-associated pseudotumor cerebri. **Neurology 49:**507–511, 1997
70. Takayasu M, Takagi T, Hara M, Anzai M: A simple technique for expansive suboccipital cranioplasty following foramen magnum decompression for the treatment of syringomyelia associated with Chiari I malformation. **Neurosurg Rev 27:** 173–177, 2004
71. Tognetti F, Calbucci F: Syringomyelia: syringo-subarachnoid shunt versus posterior fossa decompression. **Acta Neurochir (Wien) 123:**196–197, 1993
72. Tokuno H, Hakuba A, Suzuki T, Nishimura S: Operative treatment of Chiari malformation with syringomyelia. **Acta Neurochir Suppl (Wien) 43:**22–25, 1988
73. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC III, Rozzelle CJ, et al: Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. Clinical article. **J Neurosurg Pediatr 7:**248–256, 2011
74. Vanaclocha V, Saiz-Sapena N: Duraplasty with freeze-dried cadaveric dura versus occipital pericranium for Chiari type I malformation: comparative study. **Acta Neurochir (Wien) 139:**112–119, 1997
75. Vanaclocha V, Saiz-Sapena N, Garcia-Casasola MC: Surgical technique for cranio-cervical decompression in syringomyelia associated with Chiari type I malformation. **Acta Neurochir (Wien) 139:**529–540, 1997
76. Vaquero J, Martínez R, Arias A: Syringomyelia-Chiari complex: magnetic resonance imaging and clinical evaluation of surgical treatment. **J Neurosurg 73:**64–68, 1990
77. Versari PP, D'Aliberti G, Talamonti G, Collice M: Foraminal syringomyelia: suggestion for a grading system. **Acta Neurochir (Wien) 125:**97–104, 1993
78. Williams B: Cough headache due to craniospinal pressure dissociation. **Arch Neurol 37:**226–230, 1980
79. Williams B: The distending force in the production of "communicating syringomyelia." **Lancet 2:**189–193, 1969
80. Williams B: On the pathogenesis of syringomyelia: a review. **J R Soc Med 73:**798–806, 1980
81. World Health Organization: **Physical Status: The Use and Interpretation of Anthropometry: Report of a WHO Expert Committee. WHO Technical Report Series 854.** Geneva: World Health Organization, 1995
82. Yilmaz A, Kanat A, Musluman AM, Colak I, Terzi Y, Kayacı S, et al: When is duraplasty required in the surgical treatment of Chiari malformation type I based on tonsillar descending grading scale? **World Neurosurg 75:**307–313, 2011
83. Zhang ZQ, Chen YQ, Chen YA, Wu X, Wang YB, Li XG: Chiari I malformation associated with syringomyelia: a retrospective study of 316 surgically treated patients. **Spinal Cord 46:**358–363, 2008

Manuscript submitted October 23, 2012.
Accepted April 8, 2013.
Please include this information when citing this paper: published online May 10, 2013; DOI: 10.3171/2013.4.JNS122011.

Address correspondence to: Kenan I. Arnautovic, M.D., Ph.D., Semmes-Murphey Clinic, 6325 Humphreys Boulevard, Memphis, Tennessee 38120. email: kenanarnaut@yahoo.com.