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The Risk of Athletes with Chiari Malformations Suffering Catastrophic Injuries during Sports Participation is Low

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Abstract

Objective—To estimate the risk of athletes with Chiari malformations sustaining a catastrophic injury.

Design—Retrospective, descriptive cohort study.

Participants—All patients diagnosed with Chiari malformation at our institution between June 2008 and November 2011.

Assessment of Risk Factors—Participants were mailed a questionnaire regarding the number of seasons they participated in organized athletics. Magnetic resonance images were reviewed to describe the characteristics of respondent's Chiari malformations.

Main Outcome Measures—Whether or not the patient had sustained an injury resulting in death, coma, or paralysis.

Results—We had a 53% (N=147) response rate. Respondents were a mean age of 15 years (SD 2 years) at the time of diagnosis. The mean length of protrusion of the cerebellar tonsils below the foramen magnum was 11.2mm (SD 5.7mm). The majority of respondents had pointed cerebellar tonsils and some degree of crowding within the foramen magnum. During a total of 1,627 athletic seasons played by patients with Chiari malformation, 0 respondents (95% CI 0.0000, 0.0023) sustained an injury resulting in death, coma or paralysis. Likewise, during 191 collision sport

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athletic seasons, 0 (95% CI 0.0000, 0.0191) respondents sustained an injury resulting in death, coma or paralysis.

Conclusions—The risk of athletes with Chiari malformations suffering catastrophic injuries during sports participation is low. This estimate of risk should be considered when making return-to-play decisions. Given the variability of anatomical consideration for patients with Chiari malformations, however, each return-to-play decision must continue to be made on a case-by-case basis, considering all of the available information.

Key Terms

Athletes; death; coma; paralysis

INTRODUCTION

A Chiari malformation is a congenital disorder in which the cerebellar tonsils are displaced caudally, below the level of the foramen magnum.^{9, 10, 13, 17, 19} The prevalence of Chiari malformation has been estimated at 0.01%–3.6%.^{9, 10, 15, 24} Patients with Chiari malformations can be asymptomatic, but some, particularly those with further descent into the spinal canal, classically experience headaches or neck pain, particularly after Valsalva maneuvers associated with coughing, sneezing, straining, or physical exertion.^{10, 13, 14, 17, 19} Less commonly, patients may experience snoring, vision problems, dizziness, ataxia, and sleep apnea.^{13, 19} Chiari malformations are sometimes associated with other congenital craniocervical anomalies, including craniosynostosis, hyperostosis (particularly of the posterior fossa) atlantoaxial assimilation, and Klippel-Feil syndrome, among others.¹² Some authors have suggested that Chiari malformations diminish the normal capacity for the cerebrospinal fluid to buffer the brain during high-velocity impacts.^{4, 17} Furthermore, there have been several case reports of sudden death after seemingly minor trauma or rapid head movements in patients found to have Chiari malformations.^{1, 25, 27} This has led some physicians to consider a Chiari malformation a relative or absolute contraindication to participation in contact and collision sports.^{2, 9, 17, 19} Some authors recommend that athletes incidentally noted to have an asymptomatic Chiari malformation not return to contact sports,^{17, 21} with more stringent recommendations against returning to contact and collision sports for athletes who have a symptomatic Chiari malformation.¹⁷ Others, however, do not consider non-severe, asymptomatic, Chiari malformations a contraindication to return to contact or collision sports.¹⁹

Most authors recommend that athletes with Chiari malformations that are symptomatic,¹⁷ have unusually low tonsillar herniation,⁹ obliteration of the subarachnoid space,⁹ syringomyelia,⁹ and/or indentation of the anterior medulla,⁹ avoid returning to contact or collision sports. Most surgeons do not recommend preventive surgery for asymptomatic athletes with mild to moderate Chiari malformations engaged in athletics.^{19, 21}

There is, however, little data available to guide clinicians. Decisions to return athletes to play should be made on a case-by-case basis.⁹ The available literature consists mostly of case reports and expert opinion,^{1, 2, 9, 14, 17, 19, 27} making estimates of the risk of catastrophic injury for athletes with Chiari malformations who participate in contact or

collision sports difficult. In order to develop such an estimate, we conducted a survey of patients cared for in our hospital and diagnosed with a Chiari malformation.

MATERIALS AND METHODS

Patients cared for at our hospital between June 2008 and November 2011 and diagnosed with a Chiari malformation were mailed an introductory letter and questionnaire. For those under the age of 18 years, the letter was addressed to their parents as well. A website address to the questionnaire was offered in the introductory letter. Participants could decline by simply checking “no” on the questionnaire and returning it. Completion of the questionnaire constituted consent. Non-respondents received a follow-up phone call approximately 2 weeks after the initial questionnaire was mailed and were offered an opportunity to complete the questionnaire over the phone. The study was approved by the institutional review board of Boston Children’s Hospital.

The 10-item questionnaire recorded information regarding sports played by the respondents, the number of seasons of each sport played, and whether or not the patient had ever sustained an injury resulting in loss of consciousness, paralysis, coma, or death. Whether or not the patients had undergone decompression surgery and the dates of that surgery were also queried. Patients who underwent decompressive surgery were asked to separate out the number of seasons participating in sports before and after surgery. In order to be conservative in our estimates of exposure to athletics, when respondents endorsed participating in a given sport, but did not enter a number of seasons played, only 1 season was entered; when respondents reported playing in a given sport “all year round,” only 3 seasons were entered as opposed to 4; when respondents reported playing a given sport “for 6 years” or “from 7 years old to 15 years old” only one season per year was recorded. We were conservative so that we did not underestimate the risk of potentially catastrophic injury for athletes with Chiari malformation, as underestimating such a risk could result in devastating consequences.

When available, the magnetic resonance images of respondents were reviewed by 2 radiologists. Protrusion of the cerebellar tonsils below the foramen magnum (mm), the morphology of cerebellar tonsils (rounded vs. pointed), the absence or presence of brainstem compression, the absence or presence of skull base abnormality, the degree of foramen magnum crowding (none, mild, moderate, severe), the absence or presence of a cervical syrinx, the absence or presence of cerebrospinal fluid space attenuation, and the absence or presence of cerebrospinal fluid flow attenuation were all recorded, as defined below.

Tonsillar Protrusion

To measure the length of tonsillar protrusion below the foramen magnum, a line was drawn connecting the basion and the opisthion (sphenoid and occipital margins of the foramen magnum). The amount of inferior tonsillar protrusion beyond this plane (in millimeters) was determined using a measurement line perpendicular to the basion-opisthion line.

Tonsillar Morphology

Tonsillar morphology, i.e. rounded versus pointed tonsils, was a subjective determination made by the interpreting radiologist.

Degree of Foramen Magnum Crowding

The degree of foramen magnum crowding was graded from 0 (none) to 3 (severe), as detailed in table 1.

Brainstem Compression

Brainstem compression was defined as a separate variable, and noted as either being present or absent. In itself, brain stem compression also indicates severe foramen magnum crowding (Table 1), and if one of these parameters were present, the other was invariably present as well.

CSF Space Attenuation

As an independent variable, CSF attenuation was noted as either being absent or present. We used the effacement of the CSF space anterior to the brainstem, posterior to the cerebellar tonsils, or both as indicators of positive CSF attenuation. Per se, any degree of foraminal crowding would logically result in positive CSF space attenuation.

CSF Flow Attenuation

CSF flow attenuation was only noted as present or absent where a specific velocity-encoded phase contrast MRI sequence was performed. This was the case in the minority of patients. We therefore used this parameter only to reinforce CSF attenuation seen on structural images.

Additional Imaging Indicators

As an independent indicator of severity, specific note was made of the presence or absence of cervical cord syringomyelia. We furthermore assessed the skull base for any structural or morphological abnormality and mentioned these features specifically in our assessment. Examples of notable osseous abnormalities included an unusually short clivus, retroversion of the C2 dens, platybasia, basilar invagination and an unusual orientation to the basiocciput.

Continuous variables are reported as mean (standard deviation). The number of various outcomes (paralysis, coma, death) is reported per seasons of all organized athletics. Collision sports, as they represent perhaps the highest risk, were also analyzed separately. Collision sports are those in which purposeful blows to the body are delivered. For purposes of this study, collision sports were defined as American football, male ice hockey, male lacrosse, rugby, martial arts, wrestling, boxing, and male roller hockey. All analyses were performed using PASW Statistics 18 (formerly SPSS statistics, IBM, Armonk, NY 18).

RESULTS

From billing records, we identified 300 patients seen at our hospital and diagnosed with a Chiari malformation between June 2008 and November 2011. Each was mailed a

questionnaire. Of the 300 questionnaires sent out, 21 patients were no longer living at the address or reachable by the phone number on file at the hospital. Of the remaining 279, 147 (53%) completed the questionnaire, 26 (1%) declined to participate, 106 (38%) did not respond and could not be reached by phone. Of respondents, 61 (42%) were male. The mean age at the time of their hospital appointment was 15 years (SD 2 years) and ranged from 11–19 years. Two respondents did not answer the question regarding whether or not they had undergone decompression surgery. Of the remaining 145 respondents, 72 had undergone decompression surgery, 73 had not. The mean length of protrusion of the cerebellar tonsils below the foramen magnum on magnetic resonance imaging was 11.2mm (SD 5.7mm). The majority of respondents had pointed cerebellar tonsils and some degree of crowding in the foramen magnum (table 2). Soccer and basketball were the most commonly played sports (table 3). American football and ice hockey were the most commonly played collision sports (table 4).

There was no significant difference in the proportion of respondents who participated in organized sports before their decompression surgery and those participating in sports who had not undergone decompression surgery (88% vs. 92%; $p=0.13$). As we sought to determine the approximate risk of participating in sports with a Chiari malformation, further analyses were conducted using only participation in organized athletics that occurred prior to decompression surgery or by respondents who had not undergone decompression surgery.

One patient was deceased. Review of the medical record and confirmation by his treating oncologist confirmed that he died from metastatic osteosarcoma; his death was unrelated to his Chiari malformation. Although two respondents reported having sustained an injury that resulted in paralysis, review of the medical records confirmed that one suffered from facial paralysis, had an MRI revealing inflammation of the facial nerve, and was diagnosed with Bell's palsy. The other had episodes of right-sided paraesthesias, however, review of the medical records and confirmation with his treating neurosurgeon confirmed that he had not suffered an injury resulting in paralysis or weakness. No respondents reported sustaining injuries that resulted in a coma. Thirty three respondents reported that they had sustained a concussion while participating in sports. Twenty six respondents reported that they had sustained an injury resulting in a loss of consciousness, 21 of them during athletics.

The medical records of all non-respondents were searched. None of the medical records revealed an injury resulting in death, paralysis or coma. In addition, none of the non-respondents were found in a search of the state death registry.

Thus, in our study population, the risk of catastrophic outcome defined as death, paralysis, or coma was 0 (95% CI 0.0000, 0.0023) per 1,627 athletic seasons. The risk of catastrophic injury for those participants with Chiari malformation participating in collision sports was 0 (95% CI 0.0000, 0.0191) per 191 athletic seasons.

DISCUSSION

The risk of patients with Chiari malformations sustaining a catastrophic injury while participating in sports in general, and in collision sports specifically, is low. This estimate of

risk should be considered when making return-to-play decisions for patients with Chiari malformations.

Catastrophic injuries occur in sports, particularly collision sports.^{16, 28} While the risk of catastrophic injuries is real and measurable, it is fortunately low, even in those sports with the highest risk.²⁸ Athletes and their clinicians recognize that the benefits of participation in sports^{5, 11, 20, 23, 26} outweigh the potential risks, and therefore, athletes participate in sports, despite the risk.³ Certainly, the benefits of sports participation are no less important to patients with Chiari malformations.

Expert opinion and case reports in the medical literature have suggested that the risk of participation in sports may be higher for athletes with Chiari malformation, and therefore, many clinicians consider Chiari malformation a relative or absolute contraindication to sports participation.^{2, 9, 17, 19} Catastrophic injuries are known to occur, albeit rarely, during sports participation,^{6-8, 16, 18, 28} but whether or not the risk of catastrophic injury is higher for athletes with Chiari malformation than for athletes without Chiari malformation remains unknown. Until now, there have been limited data available, making quantifiable estimates of risk difficult. Our results suggest that the risk of athletes with clinically significant Chiari malformations suffering catastrophic injuries during sports participation is low.

Approximately 18% of respondents reported suffering an injury that resulted in a loss of consciousness, 14% during athletics. Unfortunately, we are aware of no comparable data with which to compare this result. In a study using the National Health Interview Survey (NHIS), Sosin et al report an annual incidence of traumatic brain injuries resulting in a loss of consciousness of 0.62% of the civilian non-institutionalized population.²² But we did not limit our question to the previous year, as the NHIS did. Therefore, we cannot make direct comparisons between our results and those of Sosin et al.

Our data must be considered in light of several limitations. First, our study included only 147 participants. Catastrophic injuries during athletics are rare events, making much larger sample sizes necessary in order to obtain more accurate estimates of risk. Ideally, a large, prospective cohort study would yield a more reliable estimate of the incidence. But since these injuries are rare, and most athletes with Chiari malformations are asymptomatic and unaware of their anomaly, such a study would require screening an enormous number of athletes with MRI, and would be impractical. Second, data were collected retrospectively, and therefore, are subject to inaccurate recall of outcomes. Our outcome, however, was catastrophic injury. Such events are unlikely to be forgotten or overlooked even with the passage of time. Third, with no outcomes in our sample and no similar survey of athletes without Chiari malformations, we cannot make estimates of risk attributable to the presence of Chiari malformation. Finally, as noted in our study, patients diagnosed with Chiari malformations have variable anatomy, differing in the degree of tonsillar descent below the foramen magnum, the amount of crowding of the foramen magnum, the degree of brainstem compression, and other factors that likely alter their risk of suffering a catastrophic injury. There may be concern that our population did not have clinically significant Chiari malformation. However, half of the respondents underwent surgical decompression for symptomatic lesions, the average descent of the cerebellar tonsils was 11.2mm, and the

majority of the tonsils were pointed, all characteristics that are consistent with significant malformations as described by most authors.

CONCLUSIONS

The risk of athletes with Chiari malformations suffering catastrophic injuries during sports participation is low. This estimate of risk should be considered when making return-to-play decisions. Given the variability of anatomical consideration for patients with Chiari malformations, however, each return-to-play decision must continue to be made on a case-by-case basis, considering all of the available information.

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Clinical Relevance

The low risk of athletes with Chiari malformations suffering catastrophic injuries in sports should be considered when making return-to-play decisions.

Table 1

Parameters used to grade foramen magnum crowding on MR imaging.

Grade	Description
0 = none	CSF Space is clear anterior to the brainstem and posterior to tonsils, no brainstem compression.
1 = mild	There is isolated dorsal or anterior CSF space effacement, or evidence of decreased anterior or posterior CSF flow on phase contrast studies. No brainstem compression. Isolated medullary kink is included in this category.
2 = moderate	CSF space effaced BOTH anteriorly AND posteriorly. No brain stem compression.
3 = severe	Brainstem and/or cerebellum physically deformed. CSF spaces obliterated.

Table 2

Characteristics of Chiari malformations among respondents

Cerebellar tonsil morphology	n/N (%)
Rounded	49/126 (39%)
Pointed	77/126 (61%)
Brainstem compression	
Yes	18/127 (14%)
No	109/127 (86%)
Skull base abnormality	
Yes	38/127 (30%)
No	89/127 (70%)
Foramen magnum crowding	
None	24/127 (19%)
Mild	45/127 (35%)
Moderate	39/127 (31%)
Severe	17/127 (15%)
Cervical Syrinx	
Yes	36/126 (29%)
No	90/126 (71%)
Cerebrospinal fluid space attenuation	
Yes	101/127 (80%)
No	26/127 (20%)
Cerebrospinal fluid flow attenuation	
Yes	18/24 (75%)
No	6/24 (25%)

* Imaging was performed at various hospitals using varying sequences. Thus, not all characteristics could be determined from each set of images.

Table 3

Sports played by respondents

Sport	Number of Participants	Total Number of Seasons Played
American Football*	13	39
Gymnastics	5	21
Equestrian/Horseback riding [†]	5	26
Ice hockey*	15	101
Lacrosse*	17	46
Baseball [†]	38	193
Soccer*	60	439
Softball [†]	26	161
Swimming/Synchronized swimming	9	29
Tennis/Squash/Racquetball	5	19
Track and Field	13	37
Basketball*	48	191
Cheerleading [†]	8	22
Cross country/road running	3	7
Field hockey*	8	18
Bowling	3	5
Dance	16	126
Martial arts ^{*‡}	8	36
Skiing/Snowboarding [†]	6	25
Volleyball [†]	11	22
Wrestling*	2	8
Springboard diving	1	4
Cycling [†]	1	1
Figure skating	1	6
Golf	9	41
Roller hockey	1	3
Boxing	1	1
Total	333	1,627

* Defined by American Academy of Pediatrics as a contact sport

[†] Defined by American Academy of Pediatrics as a limited contact sport

[‡] “Martial arts” includes respondents participating in mixed martial arts, kickboxing, tae kwon do, karate and jujitsu.

Table 4

Collision sports played by respondents

Sport	Number of Participants	Total Number of Seasons Played
Collision Sports		
American football	13	39
Male ice hockey	12	88
Male lacrosse	7	18
Martial arts	8	36
Wrestling	2	8
Boxing	1	1
Male roller hockey	1	3
Total	44	191

[†]“Martial arts” includes respondents participating in mixed martial arts, kickboxing, tae kwon do, karate and jujitsu.