A magnetic resonance imaging study of congenital Chiari malformations

PR Choudhury¹, P Sarda², P Baruah¹, S Singh³

Abstract

Introduction
Chiari malformations are developmental anomalies with the herniation of the brainstem medulla and the cerebellar tonsils and vermis through the foramen magnum. The patients manifest with headache, neck pain, progressive scoliosis and cerebellar dysfunction due to cervico-medullary compression. These malformations range from the simpler to the more complex varieties of presentation, signifying their stages of appearance during embryological differentiation and development. The aim was to study various types of Chiari malformations with the help of magnetic resonance imaging techniques.

Materials and methods
In this study, 45 cases of Chiari malformations were considered.

Results
Among 45 cases, 37, 6 and 2 were type 1, 2 and 3 Chiari malformations, respectively. 53.33% cases were male and rest were female cases. The hydrocephalus and syringomyelia were present in 28.89% and 48.89% of cases, respectively. Length of herniated cerebellar tonsil in 56.76% cases of type 1 Chiari malformations are in between 10 mm and less than 20 mm.

Conclusion
The magnetic resonance imaging technique gives better visualisation of Chiari malformations and thus this entity has rapidly evolved over the past decade.

Introduction
A complex range of anomalies of the hindbrain formation appearing at various stages of growth and development of the central nervous system are included under the group called Chiari malformations. Chiari malformation is the most common anomaly of the craniovertebral junction involving both the skeletal as well as the neural structures.

Professor Chiari first described the abnormalities that we now refer to as Chiari malformations around 1890. His observations were all made on stillborn babies or newborns, and he classified the abnormalities by the severity of tonsillar and cerebellar descent as type 1 being the least and type 4 the most severe.

The incidence of Chiari type 1 malformation by neuroimaging techniques range from 0.1 to 1% and it is least obvious clinically and may not be diagnosed until adult life. It consists of tonsillar herniation through the foramen magnum with or without varying degrees of elongation of the medulla oblongata and fourth ventricle. Chiari type 2 is virtually always with thoracicorom bulbar myelomeningocele and associated with caudal herniation of medulla, vermis and the fourth ventricle. Chiari types 3 and 4 have the major deformities with cerebellar hypoplasia and downward displacement of the brain stem and a high cervical or occipital encephalocele.

Today, we have a much clearer understanding of these conditions due to the advent of magnetic resonance imaging (MRI) scanning. As a result, this entity has rapidly evolved over the past decade with newer visualisation techniques, thus posing new challenges to diagnosis and management.

The aim of this work was to study an MRI study of congenital Chiari malformations.

Materials and methods
The study is based on the cases referred to the Department of Radiology for MRI from Departments of Neurology and Paediatric surgery with common symptoms of headache, swallowing problems, dizziness, nystagmus, etc. Informed consent of the patients and their attendants were taken. The study was approved by the Institutional Ethics Committee of the Medical College and Hospital, where the study was carried out.

Altogether 45 cases were considered for the study. The cases were classified into three groups as ‘A’, ‘B’ and ‘C’, according to Chiari type 1, 2 and 3, respectively (shown in Table 1).

The readings and evaluations were made by professors, demonstrators and postgraduate trainees.

Table 1 Different types of Chiari malformations with their number and percentage

<table>
<thead>
<tr>
<th>Groups</th>
<th>Number of cases</th>
<th>Percentage (out of 45)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group A</td>
<td>37</td>
<td>82.22</td>
</tr>
<tr>
<td>Group B</td>
<td>6</td>
<td>13.33</td>
</tr>
<tr>
<td>Group C</td>
<td>2</td>
<td>4.45</td>
</tr>
<tr>
<td>Total</td>
<td>45</td>
<td>100</td>
</tr>
</tbody>
</table>

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of Department of Radiology and Anatomy with 3–10 years of experience. The study was conducted from January 2008 to December 2012.

The data obtained from the MRI studies of 45 cases were tabulated, compared and calculated. The relative frequencies were calculated and p value was evaluated with the help of Student’s t-test probabilities.

**Results**

There were 24 males with a percentage of 53.33 out of 45 total cases and the rest were female. Distributions of male and female in different groups A, B and C (Figure 1) with percentages, mean, standard deviations and standard error of mean are shown in Table 2.

It can be concluded from Table 3 and Figure 2 that the maximum number of cases in groups A and B are in between the age groups of 15 years to less than 20 years and less than 5 years, respectively.

Relative frequency of Chiari malformations in between groups A, B and C can be studied from Table 4 and Figure 3.

The hydrocephalus and syringomyelia (syringomyelia shown in Figure 4) are present in 28.89% (13 out of 45 cases) and 48.89% (22 out of 45 cases) of cases, respectively, which can be concluded from Table 5. Lumbosacral meningomyelocele is found to be present in all the cases of type 2 Chiari malformations.

Length of herniated cerebellar tonsil (shown in Figure 4) in 56.76% (21 out of 37 cases) cases of type 1 Chiari malformations are in between 10 and less than 20 mm (Table 6). The smallest length of cerebellar tonsil in type 1 Chiari malformation is 5.2 mm and longest being 28 mm.

The level of significance of differences of Chiari malformations between

![Figure 1: Different types of Chiari malformations according to sex.](image)

<table>
<thead>
<tr>
<th>Groups</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>Percentage (out of 24)</td>
<td>Number of cases</td>
</tr>
<tr>
<td>Group A</td>
<td>20</td>
<td>83.33</td>
</tr>
<tr>
<td>Group B</td>
<td>3</td>
<td>12.5</td>
</tr>
<tr>
<td>Group C</td>
<td>1</td>
<td>4.17</td>
</tr>
<tr>
<td>Sum</td>
<td>24</td>
<td>100</td>
</tr>
<tr>
<td>Mean</td>
<td>8</td>
<td>33.33</td>
</tr>
<tr>
<td>SD</td>
<td>±10.44</td>
<td>±43.498</td>
</tr>
<tr>
<td>SEM</td>
<td>±6.028</td>
<td>±25.114</td>
</tr>
</tbody>
</table>

SD, standard deviation; SEM, standard error of mean

<table>
<thead>
<tr>
<th>Age groups (years)</th>
<th>Groups</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group A</td>
<td>Group B</td>
</tr>
<tr>
<td>Less than 5</td>
<td>2</td>
</tr>
<tr>
<td>5 to &lt;10</td>
<td>5</td>
</tr>
<tr>
<td>10 to &lt;15</td>
<td>6</td>
</tr>
<tr>
<td>15 to &lt;20</td>
<td>7</td>
</tr>
<tr>
<td>20 to &lt;25</td>
<td>6</td>
</tr>
<tr>
<td>25 to &lt;30</td>
<td>5</td>
</tr>
<tr>
<td>30 to &lt;35</td>
<td>4</td>
</tr>
<tr>
<td>35 to &lt;40</td>
<td>2</td>
</tr>
<tr>
<td>40 and above</td>
<td>0</td>
</tr>
<tr>
<td>Sum</td>
<td>37</td>
</tr>
<tr>
<td>Mean</td>
<td>4.11</td>
</tr>
<tr>
<td>SD</td>
<td>±2.315</td>
</tr>
<tr>
<td>SEM</td>
<td>±0.7718</td>
</tr>
</tbody>
</table>

SD, standard deviation; SEM, standard error of mean.
male and females is not significant (p = >0.05) whereas that of groups A and B, and groups A and C according to age groups is very significant (p = <0.01) and very highly significant (p = <0.001), respectively (Table 7).

**Discussion**

Arnold–Chiari malformation is a complex syndrome in which the brainstem medulla, and the cerebellar

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**Table 4** Distribution of frequency and relative frequency of Chiari malformations in groups 'A', 'B' and 'C'

<table>
<thead>
<tr>
<th>Class interval (years)</th>
<th>Group A</th>
<th>Group B</th>
<th>Group C</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Simple frequency (f)</td>
<td>Relative frequency (fr)</td>
<td>Simple frequency (f)</td>
</tr>
<tr>
<td>Less than 5</td>
<td>2</td>
<td>0.054</td>
<td>5</td>
</tr>
<tr>
<td>5 to &lt;10</td>
<td>5</td>
<td>0.136</td>
<td>0</td>
</tr>
<tr>
<td>10 to &lt;15</td>
<td>6</td>
<td>0.162</td>
<td>0</td>
</tr>
<tr>
<td>15 to &lt;20</td>
<td>7</td>
<td>0.189</td>
<td>0</td>
</tr>
<tr>
<td>20 to &lt;25</td>
<td>6</td>
<td>0.162</td>
<td>0</td>
</tr>
<tr>
<td>25 to &lt;30</td>
<td>5</td>
<td>0.135</td>
<td>0</td>
</tr>
<tr>
<td>30 to &lt;35</td>
<td>4</td>
<td>0.108</td>
<td>0</td>
</tr>
<tr>
<td>35 to &lt;40</td>
<td>2</td>
<td>0.054</td>
<td>0</td>
</tr>
<tr>
<td>40 and above</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Sum</td>
<td>37</td>
<td>1.00</td>
<td>6</td>
</tr>
</tbody>
</table>

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**Figure 2:** Different types of Chiari malformations in different age groups.

**Figure 3:** Relative frequency of groups 'A', 'B' and 'C'.

**Figure 4:** Magnetic resonance imaging showing tonsiller herniation (indicated by red arrow) and syringomyelia (indicated by white arrow).
The Chiari malformation is classified into four types. Type 1 is the most commonly seen clinical entity and characterised by tonsillar herniation greater than 5 mm below the foramen magnum. Syringomyelia is common with type 1 Chiari malformations. In this study, 82.22% cases fall into type 1 category which also contains the maximum number of syringomyelia cases (Table 5).

Type 2 Chiari malformation is characterised by caudal descent of cerebellar vermis along with the brainstem and fourth ventricle. This type of Chiari malformation is associated with myelomeningocele. Presence of hydrocephalus is a common finding in this type. In this study, 50% cases of type 2 is associated with hydrocephalus (Table 5). Type 3 is the rarest and most severe form, characterised by occipital or high cervical encephalocele containing herniated cerebellar or brainstem tissue. Marked cerebellar hypoplasia or aplasia is the main feature of type 4 Chiari malformation.

A discussion of the modern theories describing the aetiology of the congenital Chiari malformations begins with Gardner’s work. The ‘hydrodynamic theory’ relates the development of tonsillar herniation and other dysraphic states to disordered embryogenesis in and around the foetal hindbrain. The initial insult is the failure of pathways for cerebrospinal fluid egress from the embryological fourth ventricle to open normally at the foramina of Luschka and Magendie. This blockage at the normal exits of the fourth ventricle results in increased pressure. The cerebellar tonsils migrate caudally in response to this pressure gradient, causing the hallmark finding of crowding of the foramen magnum. This elevated pressure can also be transmitted through the obex to the spinal cord and continued pressure allows for persistence or dilatation of the central canal of the spinal cord, resulting in the formation of hydromyelic cavity.

According to Alden et al., the exact origin of Chiari type 1 malformation is unknown; however, it appears to be caused by a mismatch between the volume of the posterior fossa neural elements and the posterior fossa cranial content. Several theories have been proposed to describe the resultant pathophysiology of this mismatch. It is clear, however, that abnormal cerebrospinal fluid flow and velocity play a role in the symptoms and signs associated with this disorder.

The common symptoms related to Chiari malformation are headache, particularly precipitated by coughing, straining, sneezing, etc. (Valsalva maneuvers), dizziness, eye symptoms, most commonly nystagmus, swallowing problems, and sleep disturbances.

Type 1 Chiari malformations has been described in association with many different genetic disorders of established inheritance patterns, including Klippel–Feil syndrome,

<table>
<thead>
<tr>
<th>Table 5 Presence and absence of hydrocephalus and syringomyelia in groups ‘A’, ‘B’ and ‘C’</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type of Arnold–Chiari malformations</strong></td>
</tr>
<tr>
<td>------------------------------------------</td>
</tr>
<tr>
<td>Group A</td>
</tr>
<tr>
<td>Group B</td>
</tr>
<tr>
<td>Group C</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 6 Length of herniated cerebellar tonsils in type 1 Chiari malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Length of herniated cerebellar tonsil in mm</strong></td>
</tr>
<tr>
<td>------------------------------------------</td>
</tr>
<tr>
<td>5 to &lt;10</td>
</tr>
<tr>
<td>10 to &lt;20</td>
</tr>
<tr>
<td>20 to &lt;30</td>
</tr>
<tr>
<td>30 and above</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 7 The level of significance of differences of Chiari malformations in different groups</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Serial number</strong></td>
</tr>
<tr>
<td>------------------------------------------</td>
</tr>
<tr>
<td>1</td>
</tr>
<tr>
<td>2</td>
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<td>5</td>
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<tr>
<td>6</td>
</tr>
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</table>

Competing interests: none declared. Conflict of interests: none declared.
All authors contributed to the conception, design, and preparation of the manuscript, as well as read and approved the final manuscript.
All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.

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Carpenter syndrome and Hadju–Cheney syndrome. But syndromic type 1 Chiari malformations accounts for less than 1% of prevalence of type Chiari malformations, with most occurring as isolated phenomena.\(^9\)

**Conclusion**

MRI has become the imaging technique of choice to diagnose Chiari malformations. It is accurate, specific and noninvasive. In the exceptional case when MRI cannot be performed, computed tomography/myelography/cisternography with reformatted reconstructions can provide sufficient diagnostic information. In a number of cases, MRI alone may not be sufficient to clearly define the anatomy at the skull base or bony abnormalities in the cervical spine. The combination of MRI and reformatted computed tomography can define accurately the surgical anatomy and aid in surgical planning.

**References**