Decompression for Chiari malformation type II in individuals with myelomeningocele in the National Spina Bifida Patient Registry

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Abstract

Objective—The purpose of this study was to determine the rate of decompression for Chiari malformation type II in individuals with myelomeningocele in the National Spina Bifida Patient Registry (NSBPR). In addition, the authors explored the variation in rates of Chiari II decompression across NSBPR institutions, examined the relationship between Chiari
II decompression and functional lesion level of the myelomeningocele, age, and need for tracheostomy, and they evaluated for temporal trends in rates of Chiari II decompression.

**Methods**—The authors queried the NSBPR to identify all individuals with myelomeningocele between 2009 and 2015. Among these patients, they identified individuals who had undergone at least 1 Chiari II decompression as well as those who had undergone tracheostomy. For each participating NSBPR institution, the authors calculated the proportion of patients enrolled at that site who underwent Chiari II decompression. Logistic regression was performed to analyze the relationship between Chiari II decompression, functional lesion level, age at decompression, and history of tracheostomy.

**Results**—Of 4448 individuals with myelomeningocele identified from 26 institutions, 407 (9.15%) had undergone at least 1 Chiari II decompression. Fifty-one patients had undergone tracheostomy. Logistic regression demonstrated a statistically significant relationship between Chiari II decompression and functional lesion level of the myelomeningocele, with a more rostral lesion level associated with a higher likelihood of posterior fossa decompression. Similarly, children born before 2005 and those with history of tracheostomy had a significantly higher likelihood of Chiari II decompression. There was no association between functional lesion level and need for tracheostomy. However, among those children who underwent Chiari II decompression, the likelihood of also undergoing tracheostomy increased significantly with younger age at decompression.

**Conclusions**—The rate of Chiari II decompression in patients with myelomeningocele in the NSBPR is consistent with that in previously published literature. There is a significant relationship between Chiari II decompression and functional lesion level of the myelomeningocele, which has not previously been reported. Younger children who undergo Chiari II decompression are more likely to have undergone tracheostomy. There appears to be a shift away from Chiari II decompression, as children born before 2005 were more likely to undergo Chiari II decompression than those born in 2005 or later.

**Keywords**
myelomeningocele; spina bifida; Chiari II malformation; posterior fossa decompression; tracheostomy

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Chiari malformation type II is associated almost exclusively with myelomeningocele. Although not all patients with Chiari II malformation are symptomatic, a significant minority of patients do experience symptoms, which can be severe and life-threatening. Common signs and symptoms include dysphagia, apnea, stridor, aspiration, motor weakness, and opisthotonus.¹,²,⁴,⁶,⁷,¹³

The treatment of symptomatic Chiari II malformation in patients with myelomeningocele has changed over the past several decades. While direct hindbrain decompression was the historical treatment of choice, modern paradigms have suggested that the majority of symptomatic patients can be successfully managed with the treatment of hydrocephalus.¹,³,⁵–⁷,⁹,¹⁰,¹² Indeed, at most institutions, no patient undergoes Chiari II decompression without it first being determined that hydrocephalus is appropriately treated (i.e., CSF is adequately diverted).
Our and others’ experience suggest that some patients with hindbrain dysfunction and Chiari II–related symptoms who do not improve with treatment of hydrocephalus may also not improve with Chiari II decompression. One hypothesis for this phenomenon is that the underlying pathogenesis may not be extrinsic brainstem compression from caudal displacement of cerebellar and other tissues, but rather intrinsic disorganization of hindbrain development that may not respond to either CSF diversion or Chiari II decompression.\textsuperscript{9,12} This seems to be particularly true in infants who are born with signs of severe brainstem compromise, or who develop signs in the first postnatal days. Under this hypothesis, young patients who undergo Chiari II decompression for ongoing symptoms despite adequate CSF diversion would have a higher likelihood of requiring additional procedures, such as gastrostomy and tracheostomy.

The National Spina Bifida Patient Registry (NSBPR) was established in 2008 by the Centers for Disease Control and Prevention (CDC) to collect demographic, treatment, and outcome data on patients who attend selected spina bifida clinics in the United States. The NSBPR is a mechanism utilized for ongoing research with the goal of improving the quality of care and health outcomes for these individuals.\textsuperscript{8,11} Over 80\% of patients enrolled in the NSBPR have myelomeningocele.\textsuperscript{8}

The purpose of this study was to determine the rate of Chiari II decompression in patients with myelomeningocele entered into the NSBPR, thus providing a high-quality estimate of the national rate of this operation. Furthermore, this study examined the variation in rates of Chiari II decompression among the institutions that participate in the NSBPR, explore the rates of tracheostomy, and evaluate any temporal trends in rates of Chiari II decompression in patients with myelomeningocele.

**Methods**

Data collection for the NSBPR is performed at participating sites. A designated coordinator interviews patients and families, reviews the medical record, and records the included data on standardized case report forms. These data are then entered into the NSBPR electronic health record. Data quality checks are performed automatically. Queries are routinely sent to each center to validate questionable data entries. More complete details of the NSBPR data entry and verification process can be found in the Supplemental Appendix.

Per the established protocol for studying the aggregate data collected in the NSBPR, we submitted a data analysis proposal to the CDC Committee for Science and Publication. Once approved, we queried the NSBPR to identify all patients with myelomeningocele. We analyzed selected demographic and clinical variables and all operative procedures performed in these patients between March 2009 and October 2015. We identified individuals who had undergone at least 1 Chiari II decompression as well as those who had undergone at least 1 tracheostomy procedure. Chiari II decompression is considered an “if ever” surgery in the NSBPR, meaning that when an individual is enrolled in the registry, it is recorded if he or she has ever undergone Chiari II decompression.
We calculated the rate of Chiari II decompression among all individuals with myelomeningocele. For each participating NSBPR institution, we calculated the proportion of participants enrolled at that site who had ever undergone Chiari II decompression. For individuals who had undergone more than 1 posterior fossa decompression, analysis was performed of the first operation only. We then used univariate logistic regression to examine the relationship between Chiari II decompression and the functional lesion level of the myelomeningocele (more rostral functional lesion level, e.g., thoracic, corresponding to higher degree of neurological impairment than lower level, e.g., sacral). Finally, logistic regression was used to compare the rates of Chiari II decompression between children born before 2005 and children born in 2005 or later. The year 2005 was selected as a cutoff by our consensus opinion.

We subsequently examined the population of patients who had undergone at least 1 tracheostomy procedure. Tracheostomy was not initially an “if ever” surgery in the NSBPR, but it has been treated as such since September 2013. Therefore, to ensure accuracy, all analyses involving tracheostomy have included only individuals who have had an NSBPR clinic visit since September 2013. History of previous tracheostomy would have been noted at any visit after that time. Logistic regression was used to examine the relationship between the need for tracheostomy and Chiari II decompression, functional lesion level, and age at the time of Chiari II decompression.

Associations were considered statistically significant when \( p < 0.05 \). All analyses were performed with SAS (version 9.4, SAS Institute).

Results

A total of 4448 individuals with myelomeningocele were identified from 26 participating institutions in the NSBPR (Table 1); 2308 patients (51.89%) were female and 2140 patients (48.11%) were male. The mean age of included individuals at the time of their most recent clinic visit was 13.9 years (SD 10.3 years; median 12.3 years). The age distribution of included patients is shown in Fig. 1. Children younger than 1 year made up 12.5% of the total sample (558 infants).

With respect to functional lesion level, 882 patients (19.8%) had sacral-level (foot flexion present) myelomeningocele, 877 patients (19.7%) had low–lumbar-level (foot dorsiflexion present) myelomeningocele, 1325 patients (29.8%) had midlumbar level (knee extension present) myelomeningocele, 508 patients (11.4%) had high–lumbar-level (hip flexion present) myelomeningocele, and 856 patients (19.2%) had thoracic-level (flaccid lower extremities) myelomeningocele. Of all patients, a total of 407 individuals (9.15%) had undergone Chiari II decompression, 44 (10.8%) of whom had history of multiple posterior fossa decompressions (Table 2).

A total of 26 sites participate in the NSBPR, with a mean enrollment of 159 individuals (SD 11 patients, range 1–312 patients; Fig. 2). Among participating sites that enrolled more than 10 patients (23 of 26 institutions), the rate of Chiari II decompression ranged from 1.28% to 23.57%.
Data on the month and year of Chiari II decompression were available for 288 (71%) patients. Considering only these patients, the mean age at time of surgery for Chiari II decompression was 4.56 years (SD 7.46 years; median 1.5 years, range 0–47 years). One hundred eighteen (41.0%) children underwent Chiari II decompression when they were younger than 12 months.

Univariate logistic regression demonstrated a significant association between the functional lesion level of myelomeningocele and Chiari II decompression (Table 3). Chiari II decompression was performed in 146 of 856 patients (17.1%) with thoracic level myelomeningocele, 65 of 508 patients (12.8%) with high–lumbar-level myelomeningocele, 106 of 1325 patients (8.0%) with midlumbar-level myelomeningocele, 49 of 877 patients (5.6%) with low–lumbar-level myelomeningocele, and 41 of 882 patients (4.65%) with sacral-level myelomeningocele. More rostral functional lesion levels were associated with higher rates of Chiari II decompression (p < 0.0001). Using sacral functional lesion level as the reference, the OR for Chiari II decompression was 4.22 (95% CI 2.94–6.05) in thoracic-level, 3.01 (95% CI 2.00–4.52) in high–lumbar-level, 1.78 (95% CI 1.23–2.59) in midlumbar-level, and 1.21 (95% CI 0.79–1.86) in low–lumbar-level myelomeningocele. There was a statistically significant increase in the odds for Chiari II decompression for midlumbar or higher functional lesion level when compared with sacral-level lesions.

Logistic regression analysis demonstrated a significant difference in the rates of Chiari II decompression in children born in 2005 or later, with 281 of 2796 patients (10.05%) born before 2005 undergoing Chiari II decompression compared with 126 of 1652 patients (7.63%) born in 2005 or later (p = 0.0068; OR 1.32, 95% CI 1.08–1.61).

**Association of Chiari II Decompression and Tracheostomy**

All of the following analyses were conducted using only the 2974 individuals who had at least one NSBPR clinic visit after September 2013, of whom 270 (9.08%) had a history of Chiari II decompression. Of these 270 individuals, 32 (11.9%) underwent tracheostomy. Of the 2704 patients in the subset without a history of Chiari II decompression, 19 (0.70%) underwent tracheostomy (Table 4). Logistic regression analysis demonstrated a significant association between Chiari II decompression and the need for tracheostomy (OR 19.00, 95% CI 10.61–34.03; p < 0.0001).

Considering functional lesion level in the 270 participants with a history of Chiari II decompression, tracheostomy was performed in 11 of 92 patients (12.0%) with thoracic, 6 of 42 patients (14.3%) with high-lumbar, 7 of 71 patients (9.86%) with midlumbar, 5 of 38 patients (13.2%) with low-lumbar, and 3 of 27 patients (11.1%) with sacral functional lesion levels (Table 5). Logistic regression demonstrated no significant association between functional lesion level of myelomeningocele and the need for tracheostomy (p = 0.965).

The mean age at Chiari II decompression in children who had a tracheostomy was 1.26 years, compared with 4.94 years in children who did not require tracheostomy. Logistic regression demonstrated a significant association between age at Chiari II decompression and history of tracheostomy. Specifically, for every year of increase in age, there was a 24% lower odds of tracheostomy (OR 0.764, 95% CI 0.610–0.956; p = 0.0188).
Discussion

The overall rate of Chiari II decompression in patients with myelomeningocele in the NSBPR is 9.15%, which is consistent with reported rates in the published literature (8.7%–14.2%). However, there is significant variation in the rates of Chiari II decompression among participating NSBPR institutions. Unfortunately, the available data in the NSBPR do not contain enough detail to allow us to further explore this variation between sites. Moreover, the NSBPR do not record outcomes after Chiari II decompression with respect to improvement in Chiari II–related symptoms and hindbrain dysfunction. Of the patients who had undergone Chiari II decompression, 10.8% required one or more additional decompressions, suggesting that some patients either did not improve after surgery or had recurrence of their symptoms at a later date.

We demonstrated a significant association between the functional lesion level of myelomeningocele and the likelihood of Chiari II decompression. More rostral functional lesion levels were associated with higher rates of Chiari II decompression. In our review of the literature, this relationship has not previously been reported. One explanation for this may be that more rostral lesions or larger myelomeningoceles might have greater outflow of CSF in utero, leading to more severe Chiari II malformations. Since the severity of the Chiari malformation is not further described in the NSBPR, we cannot test this hypothesis.

The data demonstrate a significant difference in the rate of Chiari II decompression in children born in 2005 or later compared with those born before 2005. This is consistent with our hypothesis that symptomatic Chiari II malformations are now more often managed with aggressive treatment of hydrocephalus, and therefore fewer Chiari II decompressions may be necessary.

It is well established that some patients with symptomatic Chiari II malformation may require other procedures, such as gastrostomy or tracheostomy. Pollack et al. reported a series of 25 patients who underwent Chiari II decompression, 11 (44%) of whom underwent tracheostomy and 10 (40%) of whom required nasogastric or gastrostomy feedings. Akbari et al. reported that 9 of 33 patients (27.3%) who underwent Chiari II decompression required gastrostomy and/or tracheostomy. Talamonti and Zella reported that all neonates in their series who required Chiari II decompression also required transient tracheostomy and gastrostomy, but the authors did not elaborate further.

In the NSBPR, 12% of patients who underwent Chiari II decompression and 0.7% of patients who did not undergo Chiari II decompression underwent tracheostomy. We were not able to evaluate the timing of these procedures in relation to Chiari II decompression, and we were also not able to evaluate whether these procedures were temporary, and if the tubes were removed after symptomatic improvement or whether they were permanent.

Overall, individuals with myelomeningocele who underwent Chiari II decompression were more likely to undergo tracheostomy than patients with myelomeningocele who did not undergo Chiari II decompression. Although we found a relationship between functional lesion level and the need for Chiari II decompression, there was no association between functional lesion level and need for tracheostomy placement. This suggests that while
functional lesion level does seem to play a role in the need for Chiari II decompression, it may not reflect the severity of Chiari II–related symptoms.

Although there was no association between functional lesion level and tracheostomy, there was an association between age at time of Chiari II decompression and tracheostomy. Younger children who undergo Chiari II decompression are more likely to require tracheostomy placement. This suggests that patients who become symptomatic at a younger age may have more severe symptoms, including lower cranial neuropathies and disordered breathing, than those who become symptomatic at an older age. The need for tracheostomy could reflect the severity or irreversibility of their presenting symptoms and signs, or hindbrain dysgenesis.\textsuperscript{12}

Since it was established in 2008, the NSBPR questionnaire has been updated and expanded. Data are currently being collected in the third version of the registry questionnaire, which collects much more data than previous versions. More analysis is indicated in the future to better understand the management of Chiari II malformation throughout the NSBPR and hopefully to help establish treatment guidelines for symptomatic Chiari II malformation in the future.

**Limitations**

As noted above, there are some limitations in the data collected in the NSBPR. Most pertinent to the present discussion is the lack of data available about the timing of surgery. Therefore, we cannot determine if tracheostomy had been performed before or after Chiari II decompression. There is also no information about the indication for Chiari II decompression or any method to determine outcome. It is also not possible to determine if an individual had a shunt or other procedure to treat hydrocephalus before or after Chiari II decompression.

It is possible that individuals may move and thus leave the institution that is participating in the NSBPR. However, should this individual then become a patient at another NSBPR site, data can again be collected and attached to the original record, providing longitudinal follow-up. It is estimated that less than 1% of patients move each year.

The method that is used to assess functional lesion level has not been validated. Personnel at each NSBPR site determine lesion level according to the standardized definitions provided on the NSBPR case report forms.

**Conclusions**

The rate of Chiari II decompression in patients with myelomeningocele in the NSBPR is consistent with previously published literature. There is a significant relationship between Chiari II decompression and functional lesion level of the myelomeningocele, which has not previously been reported. Younger children who undergo Chiari II decompression are more likely to have undergone tracheostomy, possibly indicating disorganization of the brainstem rather than extrinsic brainstem compression. There appears to be a shift away from Chiari
II decompression, as children born before 2005 were more likely to undergo Chiari II decompression than those born in 2005 or later.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Abbreviations

NSBPR National Spina Bifida Patient Registry

References


Fig. 1.
Age of included patients.
Fig. 2.
Rate of Chiari II decompression by site. Numbers above each bar represent the number of patients included in this analysis at that site. Figure is available in color online only.
Table 1.

Patient Demographics.

<p>| | |</p>
<table>
<thead>
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<tbody>
<tr>
<td><strong>Age, years</strong></td>
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</tr>
<tr>
<td>Mean (SD)</td>
<td>13.9 (10.3)</td>
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<tr>
<td>Median</td>
<td>12.3</td>
</tr>
<tr>
<td><strong>Gender (%)</strong></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>2308 (51.9)</td>
</tr>
<tr>
<td>Male</td>
<td>2140 (48.1)</td>
</tr>
<tr>
<td><strong>Functional Lesion Level (%)</strong></td>
<td></td>
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<tr>
<td>Thoracic (flaccid lower extremities)</td>
<td>856 (19.2)</td>
</tr>
<tr>
<td>High-lumbar (hip flexion present)</td>
<td>508 (11.4)</td>
</tr>
<tr>
<td>Mid-lumbar (knee extension present)</td>
<td>1325 (29.8)</td>
</tr>
<tr>
<td>Low-lumbar (foot dorsiflexion present)</td>
<td>877 (19.7)</td>
</tr>
<tr>
<td>Sacral (foot plantar flexion present)</td>
<td>882 (19.8)</td>
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Table 2.
Number of Chiari II decompression operations

<table>
<thead>
<tr>
<th>Decompression Levels</th>
<th>Number</th>
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<tbody>
<tr>
<td>1 decompression</td>
<td>363</td>
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<tr>
<td>2 decompressions</td>
<td>33</td>
</tr>
<tr>
<td>3 decompressions</td>
<td>9</td>
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<tr>
<td>4 decompressions</td>
<td>2</td>
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<tr>
<td><strong>Total</strong></td>
<td><strong>407</strong></td>
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Table 3.

Relationship between functional lesion level of myelomeningocele and Chiari II decompression

<table>
<thead>
<tr>
<th>Functional Lesion Level</th>
<th>Chiari II Decompression</th>
<th>Total</th>
<th>Percentage (%)</th>
<th>Odds Ratio (vs. Sacral Level)</th>
<th>95% Confidence Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>146</td>
<td>856</td>
<td>17.1</td>
<td>4.22</td>
<td>2.94 - 6.05</td>
</tr>
<tr>
<td>High-lumbar</td>
<td>65</td>
<td>508</td>
<td>12.8</td>
<td>3.01</td>
<td>2.00 - 4.52</td>
</tr>
<tr>
<td>Mid-lumbar</td>
<td>106</td>
<td>1325</td>
<td>8.00</td>
<td>1.78</td>
<td>1.23 - 2.59</td>
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<tr>
<td>Low-lumbar</td>
<td>49</td>
<td>877</td>
<td>5.59</td>
<td>1.21</td>
<td>0.79 - 1.86</td>
</tr>
<tr>
<td>Sacral</td>
<td>41</td>
<td>882</td>
<td>4.65</td>
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<td>ref</td>
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<tr>
<td>Total</td>
<td>407</td>
<td>4448</td>
<td>9.15</td>
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Table 4.

Rates of tracheostomy by need for Chiari II decompression

<table>
<thead>
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<th>Tracheostomy</th>
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</thead>
<tbody>
<tr>
<td>Chiari II decompression</td>
<td>32 / 270 (11.9%)</td>
</tr>
<tr>
<td>No Chiari II decompression</td>
<td>19 / 2704 (0.70%)</td>
</tr>
</tbody>
</table>

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Table 5.
Rates of gastrostomy and/or tracheostomy placement by functional lesion level of myelomeningocele among patients who had Chiari II decompression

<table>
<thead>
<tr>
<th>Functional Lesion Level</th>
<th>Tracheotomy</th>
<th>Total</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>11</td>
<td>92</td>
<td>12.0</td>
</tr>
<tr>
<td>High-lumbar</td>
<td>6</td>
<td>42</td>
<td>14.3</td>
</tr>
<tr>
<td>Mid-lumbar</td>
<td>7</td>
<td>71</td>
<td>9.86</td>
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<tr>
<td>Low-lumbar</td>
<td>5</td>
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<td>13.2</td>
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<tr>
<td>Sacral</td>
<td>3</td>
<td>27</td>
<td>11.1</td>
</tr>
<tr>
<td>Total</td>
<td>32</td>
<td>270</td>
<td>11.9</td>
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