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Chiari II decompression in patients with myelomeningocele in the National Spina Bifida Patient Registry (NSBPR)

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Introduction

• ~1500 patients born with spina bifida in the U.S. every year

• National Spina Bifida Patient Registry (NSBPR):
  • established by the CDC in 2008
  • collect demographic, treatment, and outcome data on patients who attend SB clinics in the U.S.
  • goal of improving quality of care/health outcomes

• 21 currently participating institutions

• ~80% of patients in the registry have myelomeningocele
Chiari II Malformation

- A significant minority of patients with myelomeningocele experience symptoms due to Chiari II malformation, which can be life-threatening.

- Treatment of symptomatic Chiari II malformation has changed over past several decades.

- Anecdotal evidence suggests young patients who undergo Chiari II decompression are at high risk for gastrostomy and/or tracheostomy.

- The underlying problem may be intrinsic dysfunction due to disorganized hindbrain development.
Purpose

- determine rate of Chiari II decompression in myelomeningocele patients in NSBPR

- evaluate variability in rates of Chiari II decompression among participating institutions

- examine relationship between Chiari II decompression and functional lesion level of myelomeningocele, age, and need for gastrostomy and/or tracheostomy

- evaluate for temporal trends in Chiari II decompression
Methods

• NSBPR was queried from March 2009 to October 2015 to identify:
  
  • patients with myelomeningocele and all operations undergone by these patients
  
  • patients who had undergone at least one Chiari II decompression
  
  • patients who had undergone at least one gastrostomy and/or tracheostomy
Results—Demographics

• 4448 patients with myelomeningocele
• mean age 13.9 years (median 12.3 years)
• 2308 female patients (51.89%), 2140 male patients (48.11%)
• Functional Lesion Level

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
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</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>856 (19.2%)</td>
</tr>
<tr>
<td>High-lumbar</td>
<td>508 (11.4%)</td>
</tr>
<tr>
<td>Mid-lumbar</td>
<td>1325 (29.8%)</td>
</tr>
<tr>
<td>Low-lumbar</td>
<td>877 (19.7%)</td>
</tr>
<tr>
<td>Sacral</td>
<td>882 (19.8%)</td>
</tr>
</tbody>
</table>
Results—Chiari II Decompression

- **407 / 4448 patients (9.15%)** underwent at least one Chiari II decompression operation

<table>
<thead>
<tr>
<th>Number of Decompressions</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>363</td>
</tr>
<tr>
<td>2</td>
<td>33</td>
</tr>
<tr>
<td>2</td>
<td>9</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>407</strong></td>
</tr>
</tbody>
</table>

- **44 / 407 patients (10.8%)** had more than one Chiari II decompression surgery
Results—Institutional Variability

- mean 9.02%
- median 7.09%
- range 0-23.57%
Results—Age at Surgery

- Month/year of Chiari II decompression was available for 288 patients (71%)
  - median 1.5 years
  - mean 4.56 years
  - range 0-47 years
Higher functional lesion levels were associated with higher rates of Chiari II decompression ($p < 0.0001$).

<table>
<thead>
<tr>
<th>Functional Level</th>
<th>Chiari II Decompression</th>
<th>Total</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>146</td>
<td>856</td>
<td>17.1</td>
</tr>
<tr>
<td>High-lumbar</td>
<td>65</td>
<td>508</td>
<td>12.8</td>
</tr>
<tr>
<td>Mid-lumbar</td>
<td>106</td>
<td>1325</td>
<td>8.0</td>
</tr>
<tr>
<td>Low-lumbar</td>
<td>49</td>
<td>877</td>
<td>5.59</td>
</tr>
<tr>
<td>Sacral</td>
<td>41</td>
<td>882</td>
<td>4.65</td>
</tr>
<tr>
<td>Total</td>
<td>407</td>
<td>4448</td>
<td>9.15</td>
</tr>
</tbody>
</table>

No previously published data on the relationship between Chiari II decompression and functional lesion level of myelomeningocele.
We attempted to determine if Chiari II decompression is typically performed in conjunction with treatment for hydrocephalus.

- **13 patients (3.19%)** who had undergone Chiari II decompression had no recorded treatment for hydrocephalus at any time.
Results—Year of Birth

Fewer Chiari II decompressions were performed in children born in 2005 or later compared to those born before 2005 (p=0.0068)

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<table>
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<tbody>
<tr>
<td>born before 2005</td>
<td>281 / 2796 (10.05%)</td>
</tr>
<tr>
<td>born in 2005 or later</td>
<td>126 / 1652 (7.63%)</td>
</tr>
</tbody>
</table>
There is a strong association between Chiari II decompression and gastrostomy/tracheostomy ($p < 0.0001$)

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<table>
<thead>
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</thead>
<tbody>
<tr>
<td>Chiari II decompression</td>
<td>57 / 407 (14.0%)</td>
<td></td>
</tr>
<tr>
<td>No Chiari II decompression</td>
<td>74 / 4041 (1.8%)</td>
<td></td>
</tr>
</tbody>
</table>
No significant association between functional lesion level and gastrostomy and/or tracheostomy among patients who underwent Chiari II decompression (p=0.378).

### Results—Gastrostomy/Tracheostomy & Functional Lesion Level

<table>
<thead>
<tr>
<th>Myelomeningocele Level</th>
<th>Gastrostomy and/or Tracheotomy</th>
<th>Total</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>16</td>
<td>146</td>
<td>11.0</td>
</tr>
<tr>
<td>High-lumbar</td>
<td>14</td>
<td>65</td>
<td>21.5</td>
</tr>
<tr>
<td>Mid-lumbar</td>
<td>15</td>
<td>106</td>
<td>14.2</td>
</tr>
<tr>
<td>Low-lumbar</td>
<td>6</td>
<td>49</td>
<td>12.2</td>
</tr>
<tr>
<td>Sacral</td>
<td>6</td>
<td>41</td>
<td>14.6</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>57</strong></td>
<td><strong>407</strong></td>
<td><strong>14.0</strong></td>
</tr>
</tbody>
</table>

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.
Patients <1 year of age at the time of Chiari II decompression are more likely to also undergo gastrostomy and/or tracheostomy (p=0.0045)

<table>
<thead>
<tr>
<th>Age at Chiari II Decompression</th>
<th>Gastrostomy and/or Tracheostomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1 year of age</td>
<td>43 / 237 (18.1%)</td>
</tr>
<tr>
<td>≥ 1 year of age</td>
<td>14 / 170 (8.24%)</td>
</tr>
</tbody>
</table>

Mean age at Chiari II decompression:
- 2.15 years in children with gastrostomy and/or tracheostomy
- 5.02 years in children without gastrostomy and/or tracheostomy
Limitations

• retrospective review of prospectively collected data

• Data available in NSBPR limited
  • unable to explore/explain variation between institutions
  • unable to explore timing of Chiari II decompression in relation to hydrocephalus treatment

• More data currently being collected in 3rd version of NSBPR
Conclusion

• 9.15% rate of Chiari II decompression in myelomeningocele patients in the NSBPR

• Higher rates of Chiari II decompression are seen in:
  • higher functional lesion level
  • children born before 2005

• There is a significant association between Chiari II decompression and gastrostomy and/or tracheostomy, especially in younger patients
Thank You

Questions:
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References


