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Developing a Systematic Neurodevelopmental Monitoring Program for Infant and Toddlers with Spina Bifida: Why Is It Important?

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Boston Children’s Hospital
Case example

Patient 0
History

- Adam was born to a G2, P0, 35-year-old woman via in vitro fertilization
- Both parents are physicians
- At 18 weeks gestation L4 myelo + hydro was diagnosed
- Parents went to CHOP for MOMS trial- surgery
- Surgery took place at 22 weeks
- Delivery took place by c-section at 35 weeks after mild dehiscence of the uterine incision site
- Apgars were 9 at one and 9 at five minutes and he did generally well at the time of birth. He weighed 2.75 kg at birth (6.1 pounds)
- Lives in an affluent waterfront town in Massachusetts
MRIs at birth: absent septum pellucidum, mild residual stigmata of Chiari II malformation with a small fourth ventricle, mild reduction of posterior fossa CSF space, and minimal beaking of the tectum; no hydrocephalus.

These are from when we saw him later on… (10/08)
<table>
<thead>
<tr>
<th></th>
<th>05/07</th>
<th>11/07</th>
<th>1/09</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age in months</strong></td>
<td>10</td>
<td>16</td>
<td>28</td>
</tr>
<tr>
<td><strong>Composite Scores (mean100±15) (age equivalent)</strong></td>
<td></td>
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<tr>
<td>Cognitive Scale</td>
<td>75 (8:0)</td>
<td>90 (14:0)</td>
<td>105 (30:0)</td>
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<tr>
<td>Language Scale</td>
<td>83</td>
<td>83</td>
<td>106</td>
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<tr>
<td>Motor Scale</td>
<td>70</td>
<td>76</td>
<td>79</td>
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<tr>
<td><strong>Subscales (mean10±3) (age equivalent)</strong></td>
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<tr>
<td>Receptive Communication</td>
<td>8 (9:0)</td>
<td>8 (14:0)</td>
<td>11 (35:0)</td>
</tr>
<tr>
<td>Expressive Communication</td>
<td>6 (7:0)</td>
<td>6 (11:0)</td>
<td>11 (31:0)</td>
</tr>
<tr>
<td>Fine Motor Skills</td>
<td>9 (10:0)</td>
<td>9 (15:0)</td>
<td>8 (26:0)</td>
</tr>
<tr>
<td>Gross Motor Skills</td>
<td>1 (6:0)</td>
<td>2 (10:0)</td>
<td>5 (17:0)</td>
</tr>
</tbody>
</table>
Follow up

• He was referred to Early Intervention, and received PT, OT, and developmental supports.
• Given his progress and lack of symptoms of increased intracranial pressure, he was not shunted.
• Now, he is excelling in regular education classes, with a verbal IQ of over 120.
Management of hydrocephalus
Dr. Warf and the ETV

• In 2000, Dr. Warf moved to Uganda to work with CURE international as a neurosurgeon.
• Saw many children with hydrocephalus, many infectious.
• It was challenging to treat hydrocephalus with shunts, as patients were not able to return in time if there was an issue.
• Dr. Warf developed the procedure to treat the hydrocephalus with little need for ongoing care.
• 2012 MacArthur Foundation Fellow; although he returned to the US, his work is carried on through the Cure Hydrocephalus (CH) Surgeon Training Program.
Endoscopic third ventriculostomy

• Endoscopic third ventriculostomy reestablishes the flow of cerebral spinal fluid out of the ventricles through surgical intervention
• The choroid plexus cauterization decreases the amount of fluid that is produced, increasing the success of the procedure
• Failures are typically observed in close proximity to the initial procedure, we don’t tend to see it later in life (so far)
• Eliminates the need for a shunt
• Does not work for all patients; we have had several patients have an ETV and still require a shunt
General routine

• Infants who are followed in the Boston Children’s Hospital Spina Bifida Center are closely followed for the development of hydrocephalus.

• They are referred for neurodevelopmental assessments as a part of this monitoring.

• I meet with the families in clinic to establish a relationship.

• The families return around every 6 months in order to follow up, if not more frequently.

• Evaluations are scheduled after imaging, on the same day that they are seeing a neurosurgeon team member for follow up.
Neurodevelopmental Screening Program
Assessments

• Typical evaluations include:
  • 6 months: Bayley:3; BSID-3 Social Emotional/Adaptive; Vineland:II (transitioning to Vineland:3)
  • 12 months: same as 6 months
  • 18 months: same with CBCL, BASC-II (transitioning to BASC-3)
  • 24 months: same as 18 months

• Assessments are scheduled more frequently when there are concerns about developmental progression, regression, or neurological changes.

• A developmental survey and medical regimen questionnaire are also completed
Findings

- Information from the results of the evaluations helps to prioritize interventions and is provided to:
  - families
  - developmental pediatricians
  - neurosurgical team
  - community based early intervention and outpatient therapists
Interview

• We ask a lot of questions about:
  • developing routines, such as sleeping, eating, playing, and therapies
  • their adjustment as parents and their support networks
  • bonding with their children, what types of activities the child enjoys, and what the family does together
  • the child’s bonding with siblings and pets; general adjustment of the family members
What we have found
Current numbers

• To date, we have had 69 children participate in the program, with a range of 1 to 6 evaluations each
• There has been a total of 168 evaluations
• For data analysis purposes, the patients are separated into 4 categories (% for the current data)
  • No ETV/shunt (42.4%)
  • ETV (30.3%)
  • Shunt (10.6%)
  • ETV and shunt (16.7%)
### BSID:3 Domains (mean 100±15)

<table>
<thead>
<tr>
<th>Domain</th>
<th>No shunt/ETV (n=28)</th>
<th>ETV only (n=19)</th>
<th>Shunt only (n=7)</th>
<th>ETV &amp; Shunt (n=11)</th>
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</thead>
<tbody>
<tr>
<td>Cognitive</td>
<td>96.7</td>
<td>87.2</td>
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<tr>
<td>Language</td>
<td>90.2</td>
<td>90.3</td>
<td>83.3</td>
<td>78.8</td>
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<tr>
<td>Motor</td>
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<tr>
<td>Social-Emotional (parent report)</td>
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<td>Adaptive (parent report)</td>
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<td>81.5</td>
<td>82.6</td>
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<td>BSID:3 subscales (mean 10±3)</td>
<td>No shunt/ETV (n=26)</td>
<td>ETV only (n=18)</td>
<td>Shunt only (n=7)</td>
<td>ETV &amp; Shunt (n=10)</td>
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<td>-------------------------------</td>
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</tr>
<tr>
<td>Language</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive Communication</td>
<td>8.6</td>
<td>8.7</td>
<td>8.1</td>
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<tr>
<td>Expressive Communication</td>
<td>8.3</td>
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<tr>
<td>Motor</td>
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<tr>
<td>Fine Motor</td>
<td>9.4</td>
<td>7.9</td>
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<tr>
<td>Gross Motor</td>
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<td></td>
<td>No shunt/ETV (n=26)</td>
<td>ETV only (n=18)</td>
<td>Shunt only (n=7)</td>
<td>ETV &amp; Shunt (n=10)</td>
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<td>------------------------</td>
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<tr>
<td>Adaptive Behavior Composite</td>
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<tr>
<td>Communication</td>
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<td>84.1</td>
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<td>77.9</td>
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</tr>
</tbody>
</table>
Benefits to these visits

• It allows families to connect with a provider about their child’s abilities, rather than medical needs
• It helps to establish the role of clinicians as advocates
• Provides information to help families understand that their child will do many of the same behaviors and developmental milestones as other children
• Allows the clinical team to understand family dynamics, parenting styles, and concerns
Other topics of interest

• We discuss a variety of topics that are essential for early self regulation skills especially sleep and wake cycles, napping, and establishment of feeding and playing routines
• Latex precautions, particularly around toys and play groups
• Feeding and its relation to oromotor skill development, food allergies, and managing weight gain and early constipation
• The importance of reading to children to support language development
• Encouraging independent initiation in the absence of motor movement
Language

- We have found a number of children with slowly developing language skills
- Given restrictions in motor skills, we can find significant challenges in their ability to initiate on the environment
- We have begun to recommend the use of sign language for children early on
- We have found that language acquisition in ASL can progress rapidly, and that children are able to transition back when verbal speech is a possibility
Summary

• Our neurodevelopmental monitoring program has provided a vehicle for parents to focus on well child development and proactively working with our medical team to support developmental progress.

• We are hoping that early detection of issues (e.g., slow language acquisition) in early development can help to maintain expected skill progression when provided with rapid intervention through outpatient services.