Neuropsychological Findings in Adolescents and Young Adults with Hydrocephalus

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Shunt Technology

Spitz-Holter valve

Wade-Dahl-Till valve
Additional self-care burden
Examples of target populations

- Spina Bifida
- Aqueductal Stenosis
- Intraventricular Hemorrhage
- Neurofibromatosis
- Tectal/Midbrain Tumors
- Epilepsy
- ADHD
- Learning Disability
What we know

- Two years after high school, 70% of young adults who have disabilities are still living with their parents, and many depend on their family for functional assistance and personal care.

- The unemployment rate for the general population is ~10%; for adults with disabilities the rate is between 50% - 75%.
  - (Adolescent Health Committee, 2006)
Who’s living outside of the home? Gender

Chi-Square=10.355 (p=.001)

Zabel, Bellin, & DiCianno; First World Congress on Spina Bifida, 2009
Who’s living outside of the home?  Mobility

Fisher’s Exact Test 2-Tail P-Value=.003

Zabel, Bellin, & DiCianno; First World Congress on Spina Bifida, 2009
Who’s working? Gender

Chi-Square=5.979 ($p=.014$)
Males almost 4 times more likely to be employed

Zabel, Bellin, & DiCianno; First World Congress on Spina Bifida, 2009
Who’s working? Mobility

Fisher’s Exact Test 2-Tail P-Value = .006

Zabel, Bellin, & DiCianno; First World Congress on Spina Bifida, 2009
Transition to Adulthood

Adapted from Bernstein & Waber, 1990

Support

Demands

Diploma / College
"Moving Out"

AGE

6 10 14 18
Cognitive Issues

- Memory
- Executive Functioning
  - Inhibition
  - Planning / Organization
  - Working Memory
Processing deficits in Spina Bifida

Associative Processing vs. Assembled Processing
An interaction of AdFx and ExFx

<table>
<thead>
<tr>
<th>Executive Ability</th>
<th>Adaptive Demands</th>
</tr>
</thead>
<tbody>
<tr>
<td>Typical</td>
<td>Typical</td>
</tr>
<tr>
<td></td>
<td>Typically-Developing Adolescents</td>
</tr>
<tr>
<td>Impaired</td>
<td>High</td>
</tr>
<tr>
<td></td>
<td>Diabetes, SCI</td>
</tr>
<tr>
<td></td>
<td>ADHD, TBI, Hydrocephalus</td>
</tr>
<tr>
<td></td>
<td>Spina Bifida, Late-HIV, OSA</td>
</tr>
</tbody>
</table>

Tarazi, Mahone, & Zabel (2007) Rehabilitation Psychology
Persistent ExFx Deficits?

Only group was a significant predictor of MCI mean raw score

Tarazi, Zabel, & Mahone (2007), TCN
Intervention versus Accommodation

Gain some weight

Buy a belt
Accommodations vs. Expectations

How the Mouse Trap Works

Scribes
Readers
1-to-1’s
The 1980's
Try out new ways of doing things while the child still lives at home!

Don’t Try This At Home!
Math

- Math deficits have considerable impact upon functional independence (maybe more than reading problems).
- Key areas include:
  - Estimation
  - Automaticity of math facts
  - Consistency of math procedures
Math facts and strategies

• Efficient:
  – 3+4= “7”

• Less Efficient:
  – 3+4= “3, 4, 5, 6, 7”
  – 3+4= “3+3+1=7”

• Even Less Efficient
  – 3+4= “1, 2, 3, 4, 5, 6, 7”

Barnes, Wilkinson, Khemani, Boudesquie, Dennis & Fletcher, 2006
### Math Errors and Procedural “slips”

<table>
<thead>
<tr>
<th>Errors</th>
<th>Math Fact</th>
<th>Procedural 1</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>562</td>
<td>742</td>
</tr>
<tr>
<td></td>
<td>- 3</td>
<td>- 136</td>
</tr>
<tr>
<td></td>
<td>558</td>
<td>614</td>
</tr>
</tbody>
</table>

- Smaller from larger
- No decrement with borrow
- Problems borrowing across zero

Barnes, Wilkinson, Khemani, Boudesquie, Dennis & Fletcher, 2006
Procedures and decomposition

\[
\frac{14}{6} \times 288/48
\]

\[
\frac{2}{5} \times \frac{14}{28} = \frac{2}{5} \times \frac{1}{2} = \frac{1}{5}
\]

\[
\frac{2}{8} = \frac{1}{4}
\]
Math Accommodations

Is spending with these... the same as spending with these?
Math Accommodations
Math Accommodations

Is managing this... the same as managing this?

Click Update Now to view your latest bank, credit card, and other account activity.

See all your online accounts in one place.
Math Accommodations

Is buying groceries like this... the same as buying groceries like this?
Written Language Accommodations

Is writing with these…

the same as writing with this?
Notetaking

The Pulse smartpen records everything you hear and write, so you never miss a word.
Spelling

WordQ 2

WordQ™ is a software tool used along with standard writing software. It suggests words for you to use and provides spoken feedback to help you find mistakes. Users of all ages who have problems writing and editing can benefit from using WordQ.

Online Price:

<table>
<thead>
<tr>
<th>Quantity</th>
<th>Price</th>
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<tbody>
<tr>
<td>1 – 9</td>
<td>$199.00</td>
</tr>
<tr>
<td>10 – 24</td>
<td>$194.00</td>
</tr>
<tr>
<td>25 – 49</td>
<td>$189.00</td>
</tr>
<tr>
<td>50 +</td>
<td>$179.00</td>
</tr>
</tbody>
</table>

Qty: 1

Add to Cart
How to Use the Free Online Context Spell Checker

Entering text and running check spelling

- Make sure you are in edit mode. If not, press the Edit Text option.
- Enter your text. Currently up to 1000 characters are supported. Remaining numbers of characters are presented in the bottom of the spellchecker.
- Run Check Spelling.
- Wait till the Check Spelling process completes and until you get the reply: "Please make your corrections."

Or arm a god mann hoo doz not no hou to spel. Plese, reeda Who To Us the Spel Cheker nstructions.
Transition areas of concern

- Health insurance / access to health care
- Health care decision-making (Guardianship, Surrogate Decision-Maker, Durable Power of Attorney, Health Care Agent)
- Education: diploma track or certificate track (in school until 18 or 21 years of age)
- Vocation (career)
  - Adolescents “age into” eligibility services and many need guidance/direction (e.g., OVR)
- Income / Financial Planning (SSI)
- Residence (family home, apartment, group home, individual support services)
- Transportation / mobility
- Driving
- Recreation
Why focus on the transition gap?

• Adolescents with childhood-onset conditions are increasingly surviving and “aging into” adult-level expectations
  – Work/income
  – Higher education
  – Independent living
  – Parenting
  – Financial management
  – Medical care / self-care

• Longer life-spans have brought increased attention to medical decision-making ability, guardianship, estate inheritance, trusts, etc.

• There is a lack of programs and adult-focused providers equipped to assume care of individuals with childhood-onset conditions and provide consultation in adulthood.
What we know

- Two years after high school, 70% of young adults who have disabilities are still living with their parents, and many depend on their family for functional assistance and personal care.

- The unemployment rate for the general population is ~10%; for adults with disabilities the rate is between 50% - 75%.
  - (Adolescent Health Committee, 2006)
Ongoing areas of NP concern

• Many “pediatric/developmental” conditions remain potentially unstable in adulthood, creating a need for ongoing monitoring and periodic updates of intervention / accommodation.
  – Shunt failure in adults with Spina Bifida / Hydrocephalus
  – Stroke in adults with Sickle Cell Disease
  – Early aging processes?

• Unique self-care competencies may create additional “executive burden,” overwhelming intact or impaired executive abilities
Ongoing areas of NP concern

- Expectations for increased independence can lead to reduction in parental oversight of:
  - General medical care
    - Medication: dispensing, monitoring, purchasing, storage, and preparation.
    - Scheduling necessary appointments / Care coordination
  - Specialized medical self-care
    - E.g., diabetes management
    - E.g., use of a C-PAP
- Problems may emerge or intensify in these areas as support is reduced, even if neurologic status is unchanged.
- Transition from specialized multi-disciplinary clinics to potentially less integrated adult-focused medical care can result in “holes” in medical care delivery.
Transition to Adulthood

Adapted from Bernstein & Waber, 1990

Diploma / College
Employment
“Moving Out”

AMOUNT

6 10 14 18

AGE
### Executive Ability

<table>
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<tr>
<th>Typical</th>
<th>High</th>
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<tbody>
<tr>
<td>Typical</td>
<td></td>
</tr>
<tr>
<td>Impaired</td>
<td></td>
</tr>
</tbody>
</table>

#### Typical
- Typically-Developing Adolescents
- ADHD, TBI, Hydrocephalus

#### High
- Diabetes
- SCI
- Spina Bifida, Late-HIV, OSA

*Tarazi, Mahone, & Zabel (2007) Rehabilitation Psychology*
Examples of target populations

- Spina Bifida
- Sickle Cell Disease
- Muscular Dystrophy
- Myelodysplasia
- Congenital Heart Disease
- Epilepsy
- ADHD
- Learning Disability
- Pediatric Cancer Survivors
ETV and SB ≠
A secondary insult?

NF, Tectal Tumors, IVH, AS…
Different Conditions

- Gordon Vigilance Task (Gordon, 1983).
- 101 children with SBM, 17 with AS, and 40 normal controls.
- Children with SBM did not differ from AS or NC groups on measures of sustained attention, but they committed more errors and responded more slowly.
Outcomes 1: Parent Ratings of Adaptive Dysfunction in SB

Jacobson et al., First World Congress on SB, 2009
Outcomes 2: Parent Ratings of Executive Dysfunction in SB

BRIEF scales
- Initiate
- Working Memory
- Plan/Organize

Mahone et al., Child Neuropsychology, 2002
Processing deficits in SB

Associative Processing vs. Assembled Processing
Cognitive impact of shunted hydrocephalus

• Studies typically suggest lasting cognitive deficits in children and adolescents with histories of shunted OH, including:
  
  – **Visual perception and nonverbal reasoning** (Fletcher, Francis, Thompson, Davidson, & Miner, 1992b; Brookshire et al., 1995)
  – **Memory** (Dennis et al., 2007; Vachha & Adams, 2005; Scott et al., 1998)
  – **Attention and working memory** (Boyer, Yeates, & Enrile, 2006; Matson et al., 2005; Dennis et al., 2007).

• Pediatric studies typically include participants with myelomeningocele [MM] (i.e., Spina Bifida).
Neuropsychological Functioning following ETV

- **Burtscher, Bartha, Twerdy, Eisner & Benke (2003)**
  - Cognitive improvement following ETV in 6 adults with late onset idiopathic AS.
  - Pre-surgical deficits were noted in anterograde memory and executive functioning
  - Anecdotal improvement noted at the time of post-operative follow-up (an average of 9 weeks post-ETV) even though post-ETV ventricular volumes did not reduce to normal size.

- **Lacy, Oliveira, Austria, and Frim (2009)**
  - Aggregate memory and executive functioning deficits in ten adults (mean age 37.4) in the years following ETV treatment for OH.

- To date, there has been no publication of a standardized examination of post-ETV neuropsychological functioning in children or adolescents.
Patient A

- 14-year old boy who underwent ETV at age 13
- Tectal glioma with OH diagnosed following new onset of hand tremors.
- Neuropsychological test score obtained approximately 21-months post-ETV.
- Parent rated executive functioning and adaptive functioning were within age expectations.
- Academic skills were within normal limits, but academic fluency was impaired.

Figure 2: Axial MRI of a 14-yr old male ETV Patient: Patient A
Patient B

• 16-year old boy who underwent ETV at age 14
• Tectal glioma with OH
• Mild TBI around age 3.
• At age 14 reported visual difficulties
• Sustained a second TBI while sledding at age 14
• Neuropsychological test scores were obtained approximately 29-months post-ETV
• Parent rated executive functioning and adaptive functioning were within age expectations.
• Academic skills and fluency also fell within normal limits.
Patient C

- 10-y.o. girl with ETV performed at age 9
- Intracranial midbrain lesion with OH.
- NF was suspected.
- Strabismus surgery at age 8 (w/o improvement)
- Neuropsychological test scores obtained 1-year post-ETV
- Parent rated executive functioning and adaptive functioning were within age expectations.
- Sight-word reading was intact, but math skills and academic fluency were below age level expectations.
Verbally-based skills

Figure 5: Verbal Skills

- Verbal IQ estimate
- Naming
- Repetition
- Comprehension
- Fluency
- Verbal Learning
- Verbal Recall

Patient A
Patient B
Patient C
Nonverbal skills

Figure 6: Nonverbal Skills

- Nonverbal IQ estimate
- Visual Perceptual Processes
- Visual Motor Integration
- Visual Learning
- Visual Recall

-3 -2 -1 0 1 2 z-score

Patient A
Patient B
Patient C
DTI?

Fig 4. An axial slice from a 16-year-old female pre-surgical ETV patient showing OB in the third ventricle and lateral ventricles. Axial slices are examples of (A) a DTI fractional anisotropy map and (B) a DTI-based color map for the patient.
Table 1: Structural size and FA parameters for a pre-surgical ETV patient and three age matched control participants.

<table>
<thead>
<tr>
<th></th>
<th>Pre-Surgical ETV Participant (age 16)</th>
<th>Control Participants (n=3; mean age 16 +/- 1.73 years)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Size (mm²)</td>
<td>FA</td>
</tr>
<tr>
<td>Genu of Corpus Callosum</td>
<td>77</td>
<td>0.51</td>
</tr>
<tr>
<td>Splenium of Corpus Callosum</td>
<td>88</td>
<td>0.66</td>
</tr>
<tr>
<td>Right Cingulate</td>
<td>19</td>
<td>0.35</td>
</tr>
<tr>
<td>Left Cingulate</td>
<td>16</td>
<td>0.33</td>
</tr>
<tr>
<td>Fornix</td>
<td>19</td>
<td>0.26</td>
</tr>
</tbody>
</table>

* Size and FA values of the ETV patient that are > 2 standard deviations below the mean size and FA values of the control group are presented in bold font and shaded cells.
MR-ICP?

Fig 1: Slice position for PC cine MRI with retrospective ECG-gating on (A) sagittal T1 weighted image in the mid-sagittal plane, (B) sagittal and coronal angiographic scout images as indicated by orange reference lines corresponding to (C) axial anatomic PC image.
Table 1: Summary of Hydrodynamic Parameters

<table>
<thead>
<tr>
<th></th>
<th>ETV Patient</th>
<th>Healthy Control Subject</th>
</tr>
</thead>
<tbody>
<tr>
<td>tCBF (ml/min)</td>
<td>844</td>
<td>890</td>
</tr>
<tr>
<td>tCVO (ml/min)</td>
<td>392</td>
<td>509</td>
</tr>
<tr>
<td>CSF area (cm²)</td>
<td>2.14</td>
<td>1.64</td>
</tr>
<tr>
<td>Stroke volume (ml)</td>
<td>0.48</td>
<td>0.62</td>
</tr>
<tr>
<td>Sys CSF velocity (cm/s)</td>
<td>1.49</td>
<td>2.21</td>
</tr>
<tr>
<td>PTP-PG (mmHg/cm)</td>
<td>0.038</td>
<td>0.045</td>
</tr>
<tr>
<td>ICVC (ml)</td>
<td>0.5</td>
<td>0.61</td>
</tr>
<tr>
<td>ICCI (mL/[Pa·m])</td>
<td>6.14</td>
<td>8.26</td>
</tr>
</tbody>
</table>

tCBF: total cerebral blood flow; tCVO: total cerebral venous outflow; CSF: cerebrospinal fluid; Sys: systolic; PTP-PG: peak-to-peak pressure gradient; ICVC: intracranial volume change; ICCI: intracranial compliance index
Research Questions

• Outcomes
  – Adaptive Functioning
    • Are there specific areas of persistent self-care, social, or ADL deficit?
  – Executive Functioning Outcome
    • Is there evidence of EF deficits that may impact transition into adulthood?
  – Academic Presentation / Outcome
    • Is there a common processing deficit associated with all forms of OH, or do learning problems vary between different OH-etiologies?
    • What are there specific problems that underlie common learning issues in math, reading comprehension, etc?