Spina Bifida and Hydrocephalus across the lifespan
Part 1

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### Table 1: The Glasgow Coma Scale and Score

<table>
<thead>
<tr>
<th>Feature</th>
<th>Scale Responses</th>
<th>Score Notation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eye opening</td>
<td>Spontaneous</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>To speech</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>To pain</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>None</td>
<td>1</td>
</tr>
<tr>
<td>Verbal response</td>
<td>Orientated</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>Confused conversation</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Words (inappropriate)</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Sounds (incomprehensible)</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>None</td>
<td>1</td>
</tr>
<tr>
<td>Best motor response</td>
<td>Obey commands</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>Localise pain</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>Flexion – Normal</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>– Abnormal</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Extend</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>None</td>
<td>1</td>
</tr>
</tbody>
</table>

**Total Coma ‘Score’**

3/15 – 15/15
Hydrocephalus and heterogeneity

We’ve learned a lot about this one!
Hydrocephalus and heterogeneity

- Spina Bifida with shunted hydrocephalus is a good model of lifespan neuropsychological research
- Relevant for research investigating other etiologies of hydrocephalus
  - Aqueductal Stenosis
  - Tectal Tumors
  - IVH
  - Isolated ACC? Cysts?
- Various syndromes mentioned yesterday…
Increasing survivability of SB
Evolution of neuropsychological model

- **IQ**: Mawdsley et al., 1967
- **Presence of HYD**: Smith & Smith, 1973
- **Spinal lesion level**: Hunt et al., 1973
- **Perceptual-Motor**: Soare & Raimondi, 1977
- **Cocktail Party Syndrome**: Tew & Laurence, 1979
- **VIQ>PIQ**: Dennis et al., 1981
- **Reading**: Barnes & Dennis, 1992
- **Subtle language deficits**: Dennis et al., 1994
- **Nonverbal Learning Disability**: Fletcher et al., 1995
- **Executive Dysfunction**: Fletcher et al., 1995; Mahone et al., 2002
- **Math**: Barnes et al., 2002
- **Phenotype of SB**: Dennis et al., 2006
Identification of a Cognitive Phenotype

CRITICAL REVIEW

A model of neurocognitive function in spina bifida over the life span

MAUREEN DENNIS,¹ SUSAN H. LANDRY,² MARCIA BARNES,³ AND JACK M. FLETCHER⁴
Main points #1 and #2

• Look beyond IQ
• Appreciate the malformations that underlie the hydrocephalus
### Neural Phenotype: Primary CNS Insults

- **Core Deficits**
  - Attention
  - Movement

- **Assembly**
- **Associations**

### Neural Phenotype: Secondary CNS Insults

- **Environment**
- **Perception**
- **Language**
- **Literacy**
- **Numeracy**

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**Spinal Cord**

**Brainstem**

**Chiari-II**

**Callosal Dysgenesis**
Main point #3

- Emphasize functional relevance
  - Math
  - Executive Functions / Prospective Memory
  - Self-Management (Self-Care)
Math Deficits

Barnes, Wilkinson, Khemani, Boudesquie, Dennis & Fletcher, 2006
Math Deficits

\[
\begin{align*}
\frac{1}{48} & \div \frac{96}{192} \\
& = \frac{96}{192} \\
& = \frac{2}{48} \\
& = \frac{1}{24}
\end{align*}
\]
Executive Dysfunction

What rating did parents give most of the time?
Initiation, Working Memory, Planning, etc.

Often

Sometimes

Never

Tarazi, Zabel, & Mahone (2007), TCN
Extra adaptive demands
Self Management

Kennedy Krieger Independence Scales – Spina Bifida Version (KKIS-SB)

- Begin CIC on time
- Get out of bed on time
- Finish morning hygiene
- Take medication on time
- Keep room clean
- Finish chores
Main point #4

• Consider context
Correlates of Depressive and Anxiety Symptoms in Young Adults with Spina Bifida

Melissa H. Bellin,1 PhD, MSW, LCSW, T. Andrew Zabel,2 PhD, Brad E. Dicianno,3 MD, Eric Levey,2 MD, Kim Garver,4 LCSW, BCD, Ronna Linroth,5 OT, MA, and Patricia Braun,6 PhD, RN

1University of Maryland, 2Johns Hopkins University, 3University of Pittsburgh Medical Center, 4SUNY Upstate Medical University, 5Gillette Children’s Specialty Healthcare, and 6Loyola University Chicago
DEVELOPMENTAL DISABILITIES RESEARCH REVIEWS 16: 40–46 (2010)

Psychosocial and Family Functioning In Spina Bifida

Grayson N. Holmbeck* and Katie A. Devine
Department of Psychology, Loyola University Chicago, Chicago, Illinois
Main point #5

- Maintain a future orientation when assessing skills and planning interventions
Future Orientation

Is spending with these… the same as spending with these?
Future Orientation (Retro)
Future Orientation (Modern)
Future Orientation?