Neuropsychological Findings in Adolescents and Young Adults with Hydrocephalus

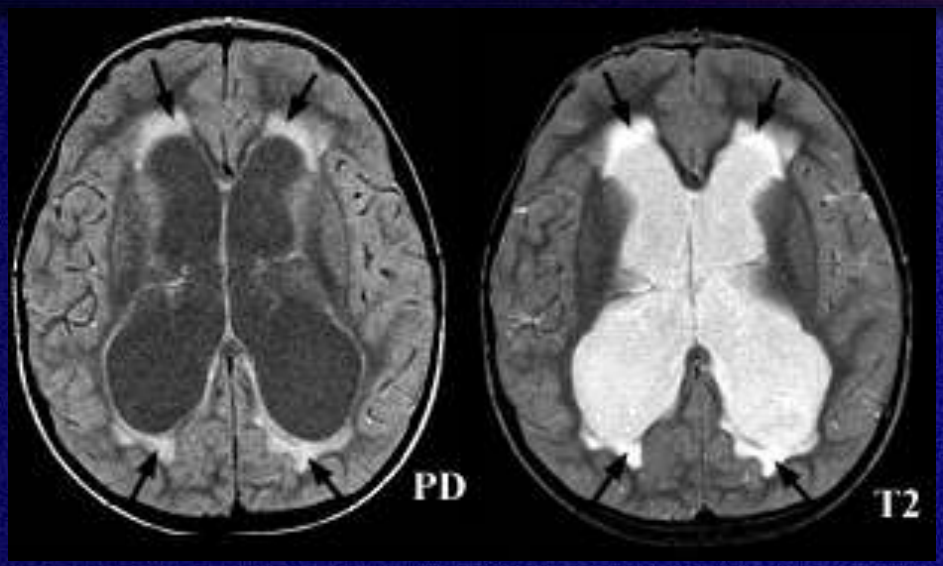
- T. Andrew Zabel, Ph.D., ABPP
- Kennedy Krieger Institute
- Johns Hopkins University School of Medicine

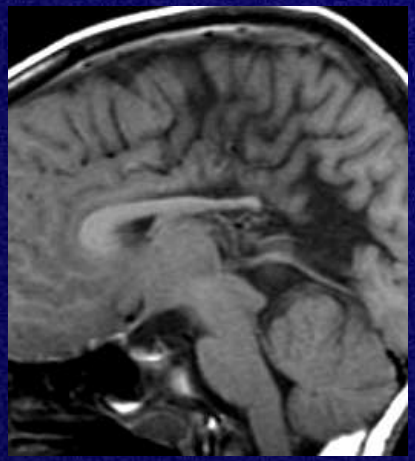
Shunt Technology

Spitz-Holter valve

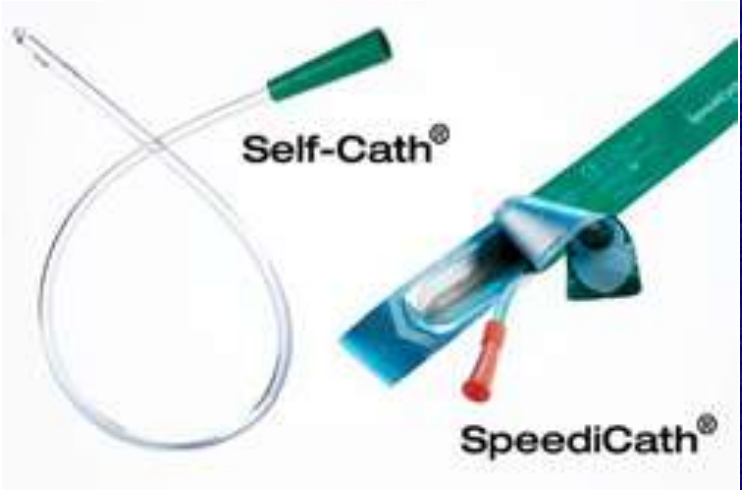
Wade-Dahl-Till valve

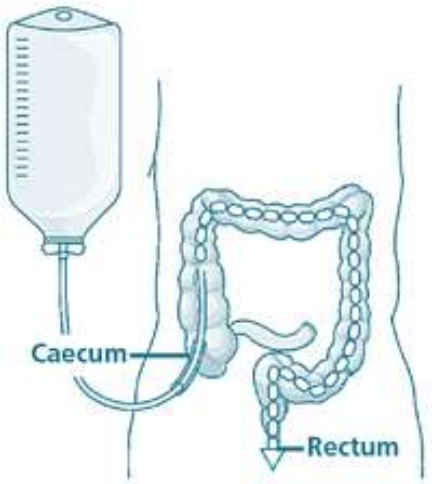
MRI

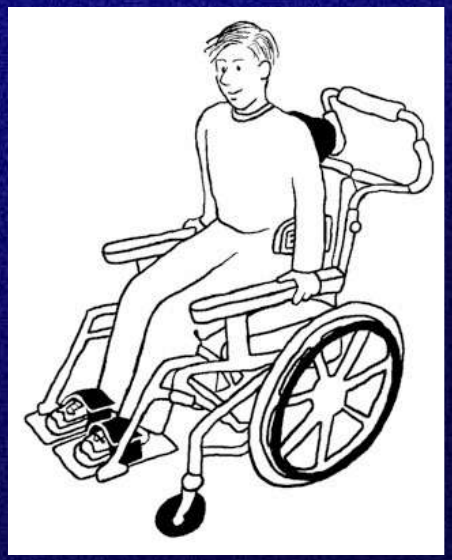




Additional self-care burden







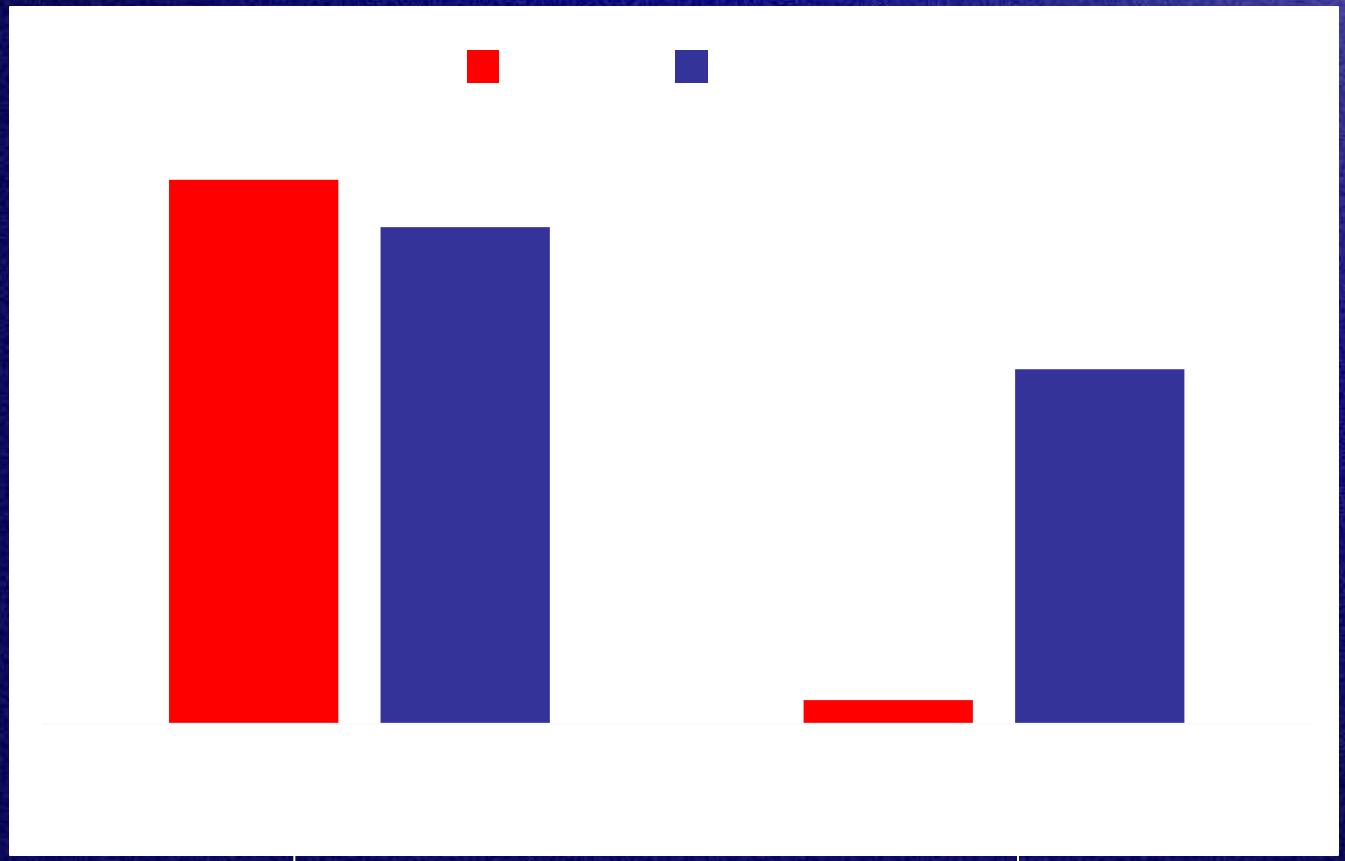
Examples of target populations

- 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
  - Geenen, S. J., Powers, L. E., & Sells, W. (2003).
- The unemployment rate for the general population is ~10%; for adults with disabilities the rate is between 50% - 75%
  - (Adolescent Health Committee, 2006)

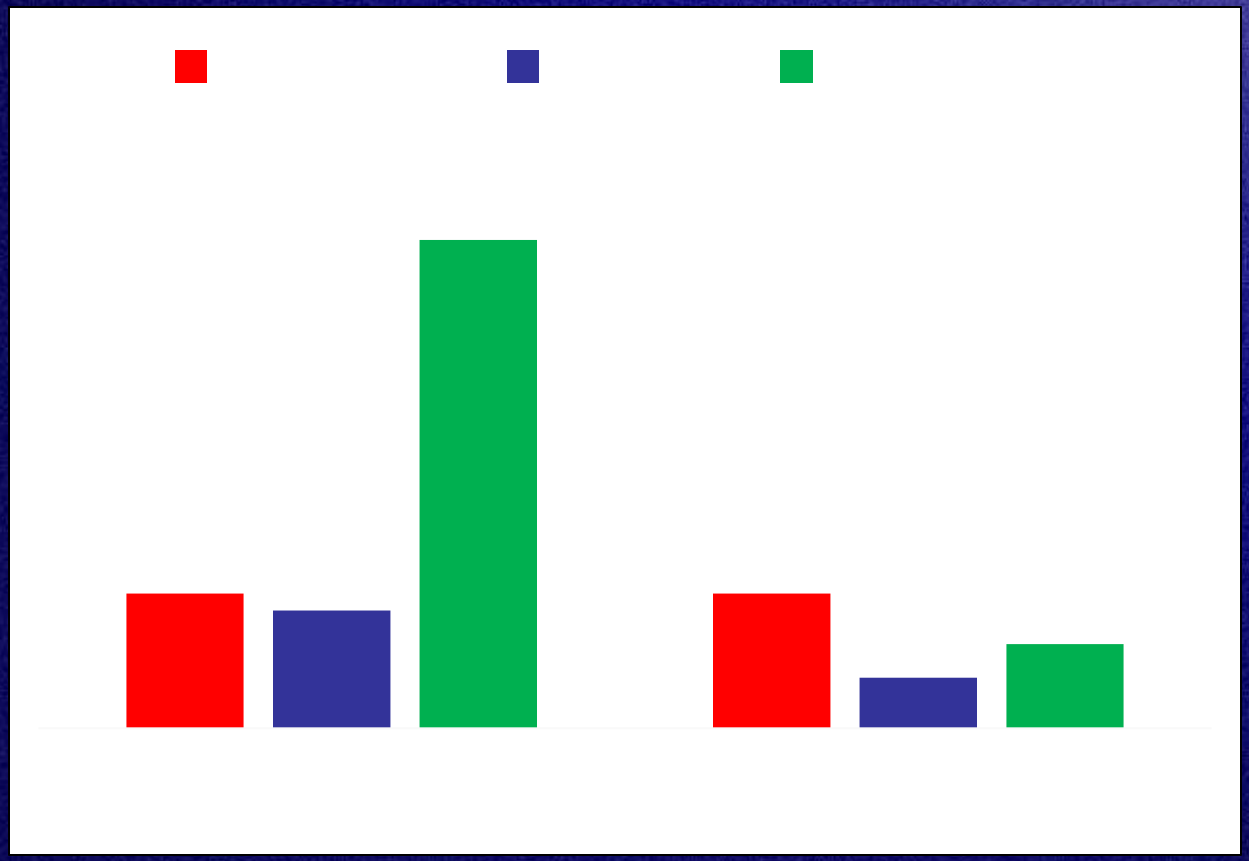
Who's living outside of the home? Gender



Chi-Square=10.355 (p=.001)

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

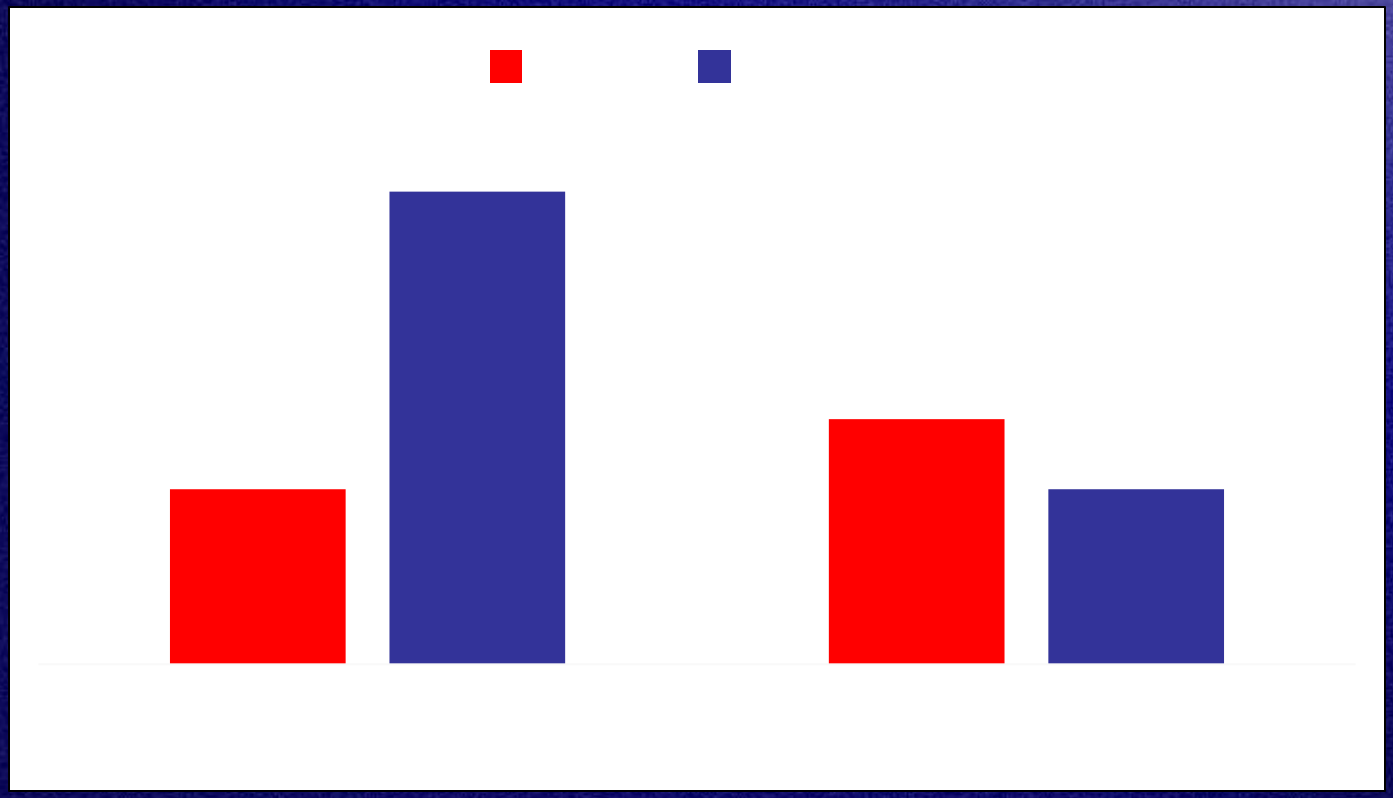
Who's living outside of the home? Mobility



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

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

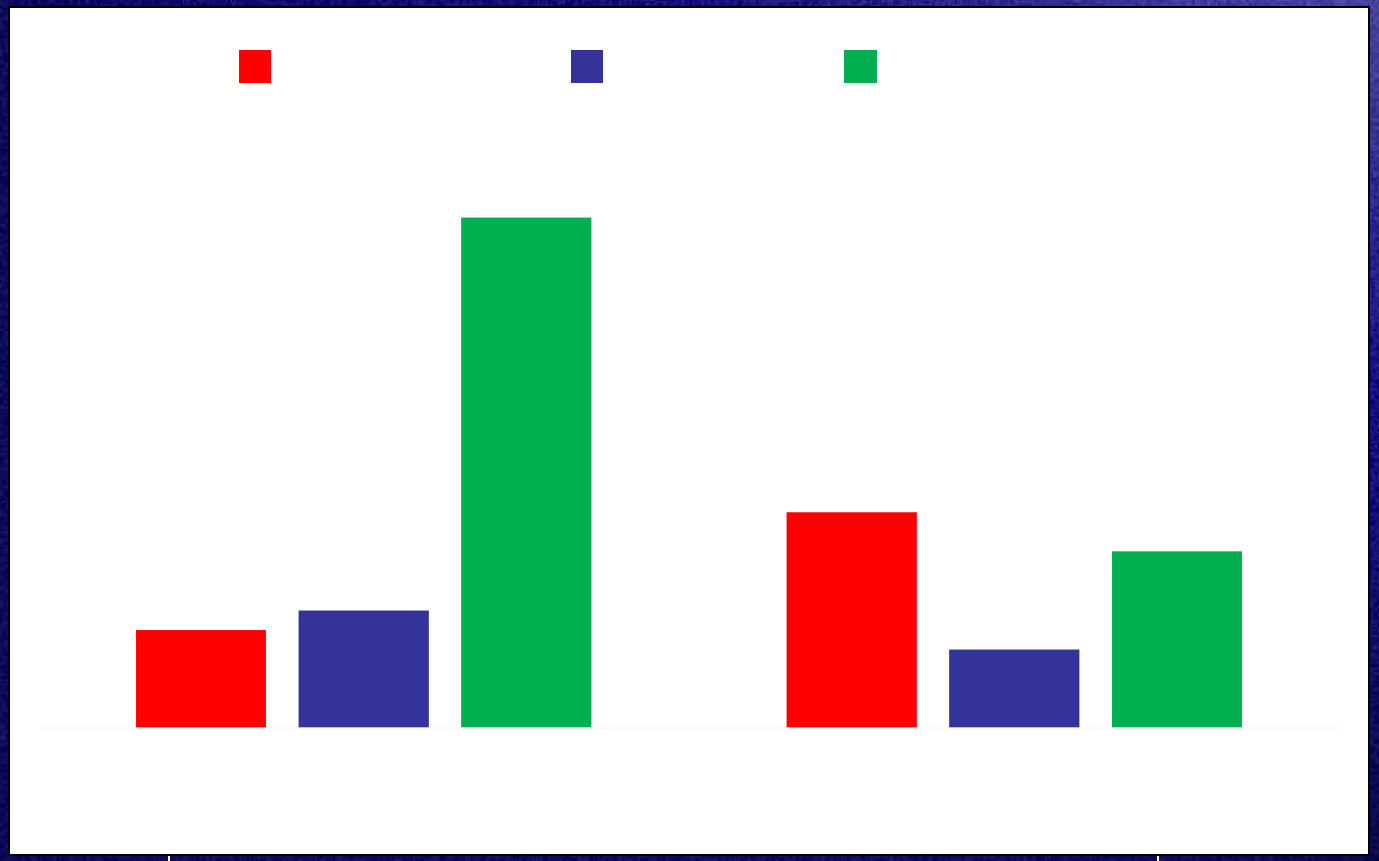
Who's working? Gender



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

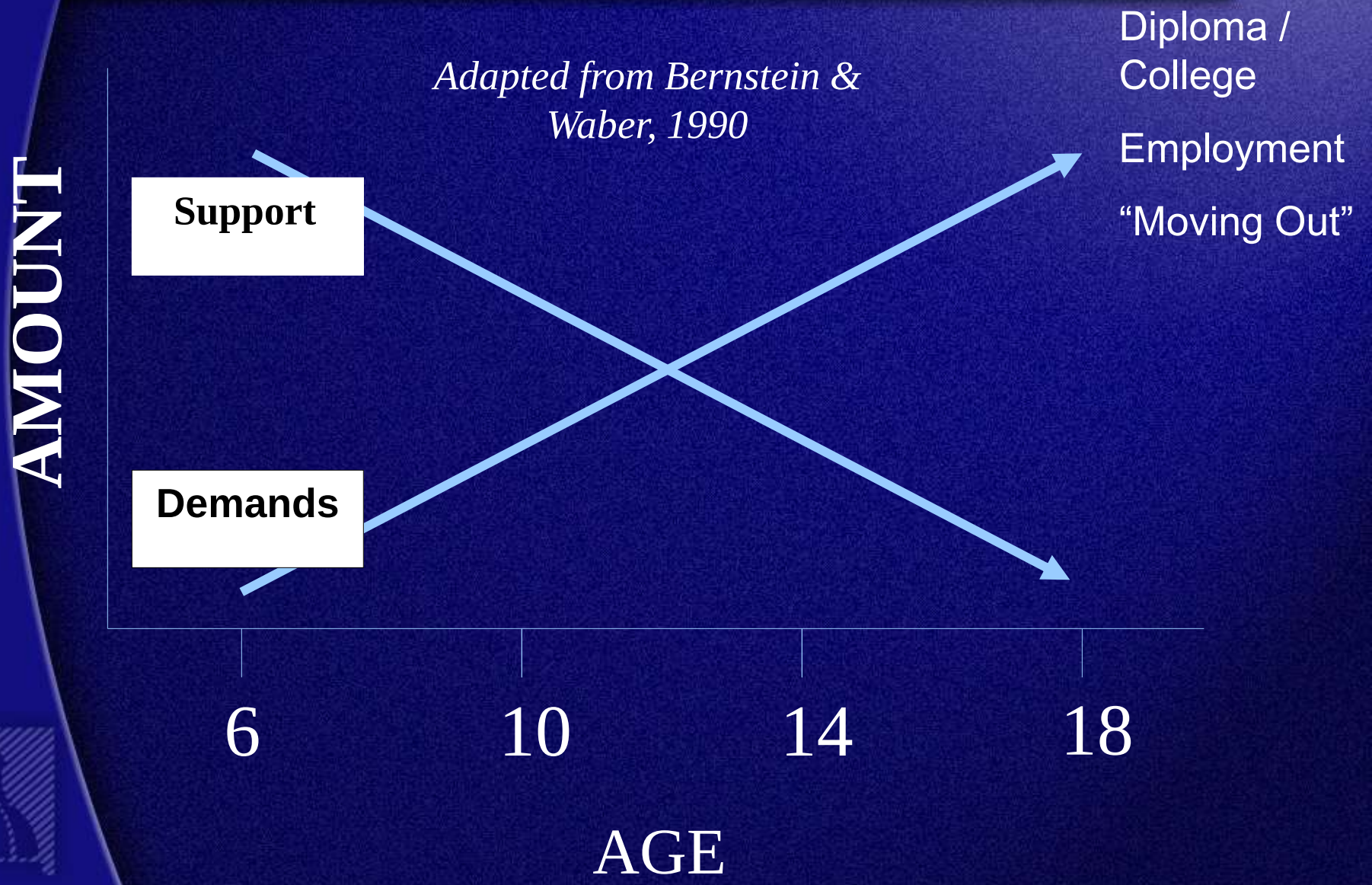
Who's working? Mobility



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

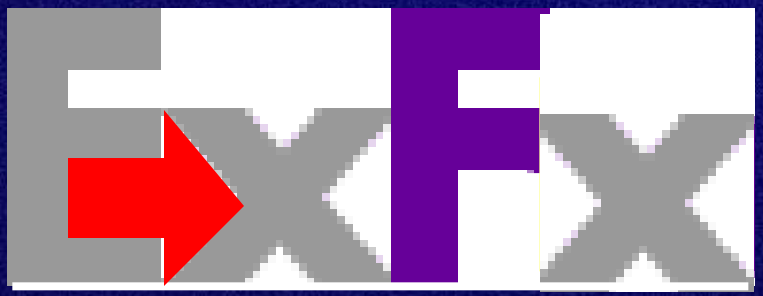
Zabel, Bellin, & DiCianno; First World Congress on Spina Bifida, 2009

Transition to Adulthood



Cognitive Issues

- Memory
- Executive Functioning
  - Inhibition
  - Planning / Organization
  - Working Memory

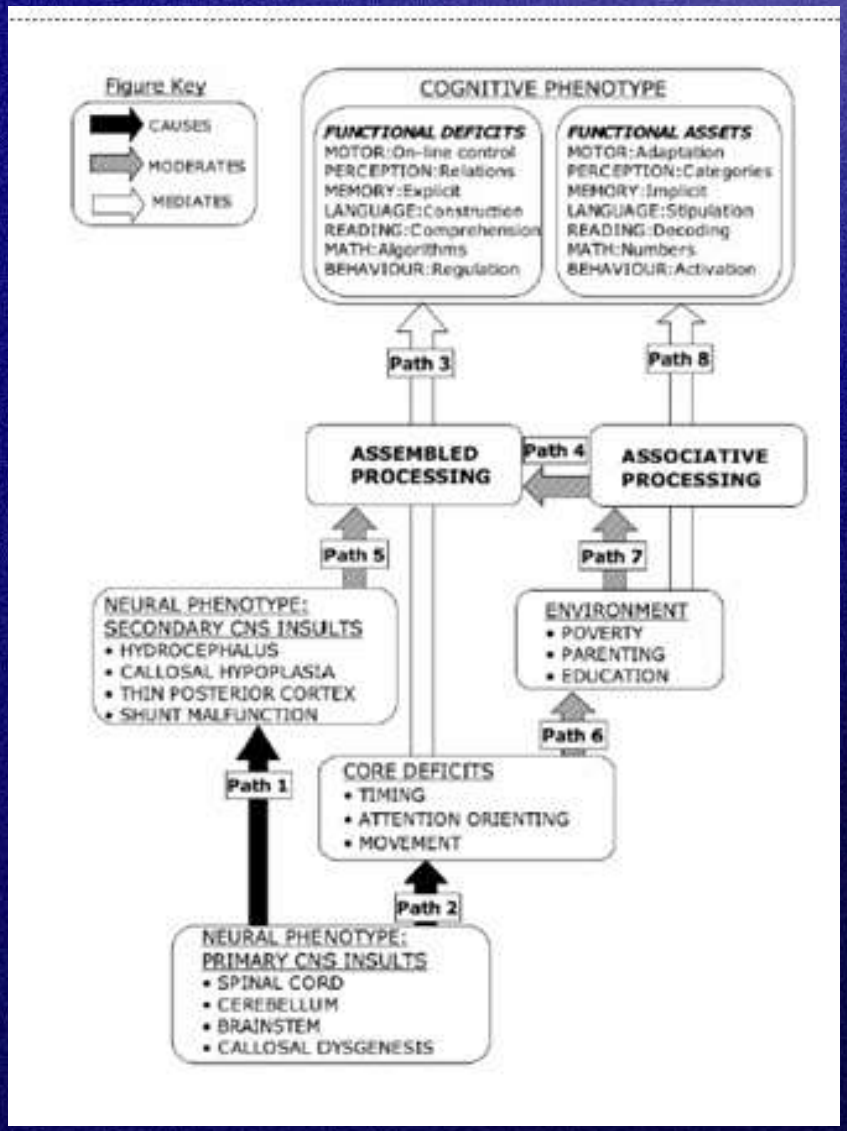


Processing deficits in Spina Bifida

Associative Processing

vs.

Assembled Processing



An interaction of AdFx and ExFx

Executive Ability	Adaptive Demands: Typical	Adaptive Demands: High
Typical	Typically-Developing Adolescents	Diabetes SCI
Impaired	ADHD, TBI, Hydrocephalus	Spina Bifida, Late-HIV, OSA

Tarazi, Mahone, & Zabel (2007) Rehabilitation Psychology

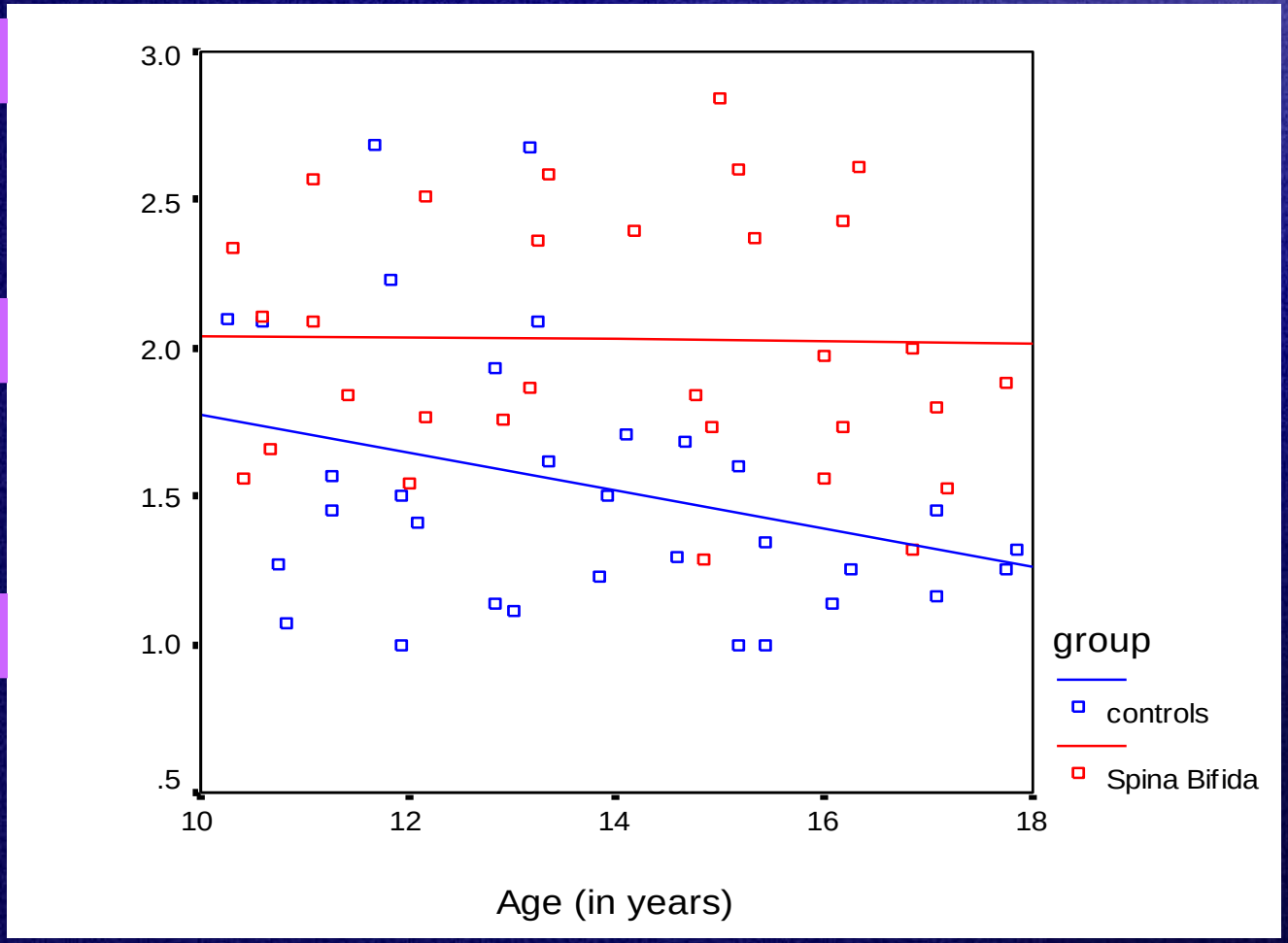
Persistent ExFx Deficits?

Only group was a significant predictor of MCI mean raw score

Often

Sometimes

Never



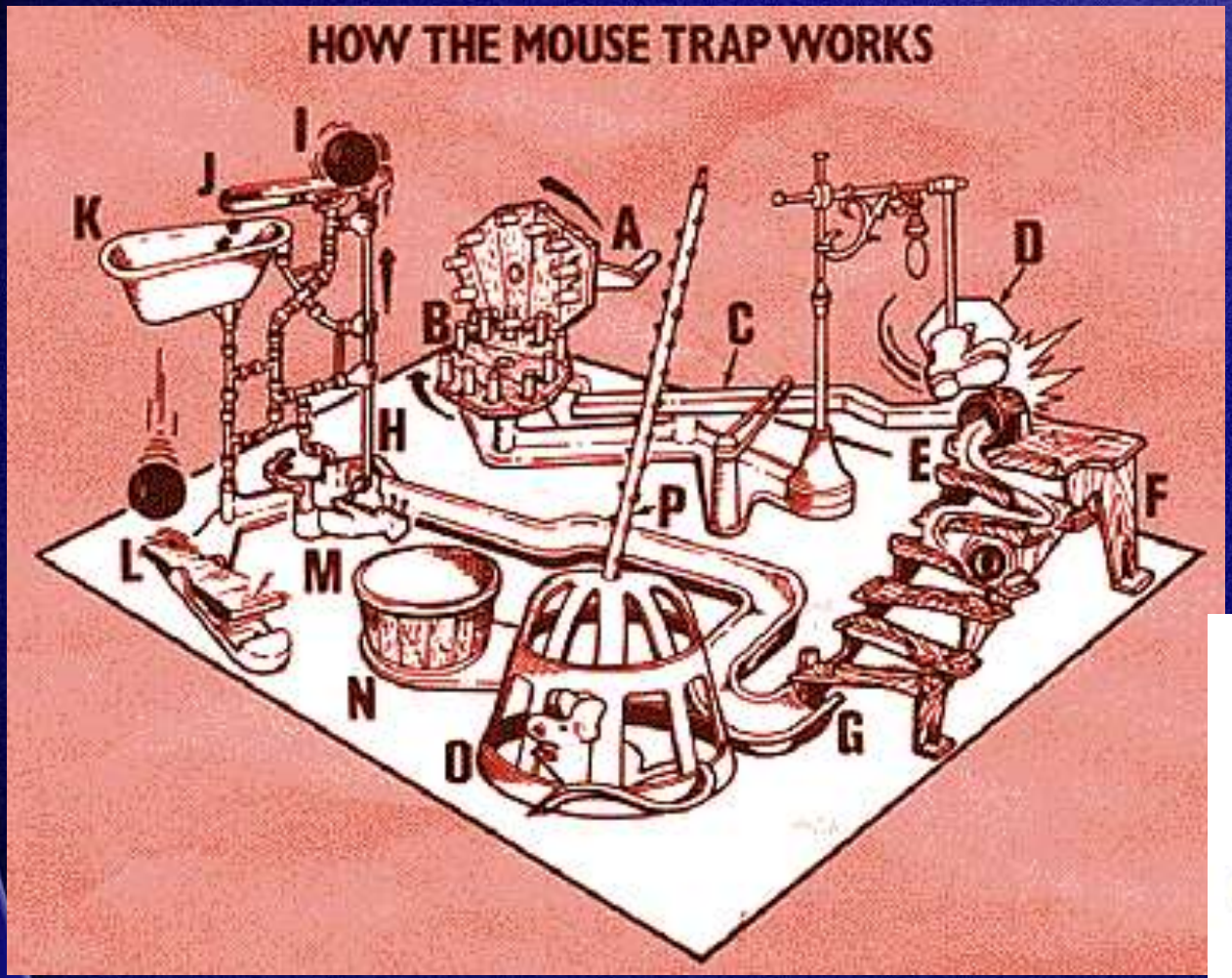
Tarazi, Zabel, & Mahone (2007), TCN

Intervention versus *Accommodation*

Gain some weight

Buy a belt

Accommodations vs. Expectations



Scribes
Readers
1-to-1's

The 1980's


CrazyAboutTV.com


Preview
1UP
HIGH SCORE
2UP
000000
HELP!
BONUS


NEW!
Coke

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

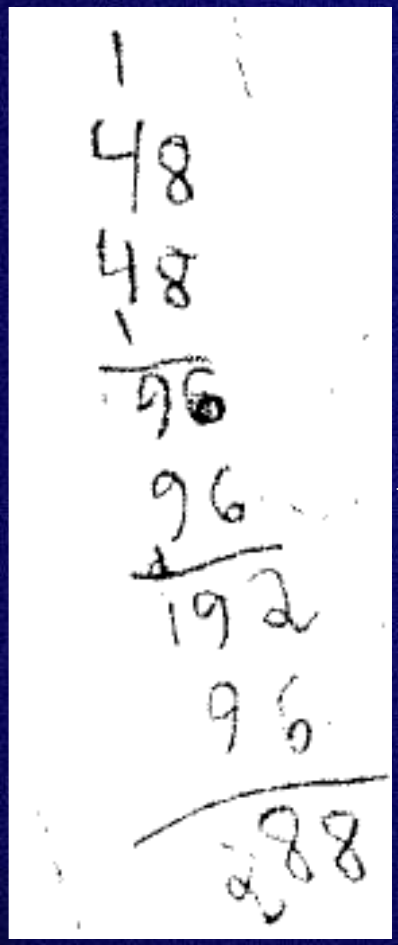
Math Errors and Procedural “slips”

Errors

Math Fact	562 - 3 558		
Procedural[1]	742 - 136 614	742 - 136 616	8007 - 5880 3227
	Smaller from larger	**No decrement with borrow**	**Problems borrowing across zero**

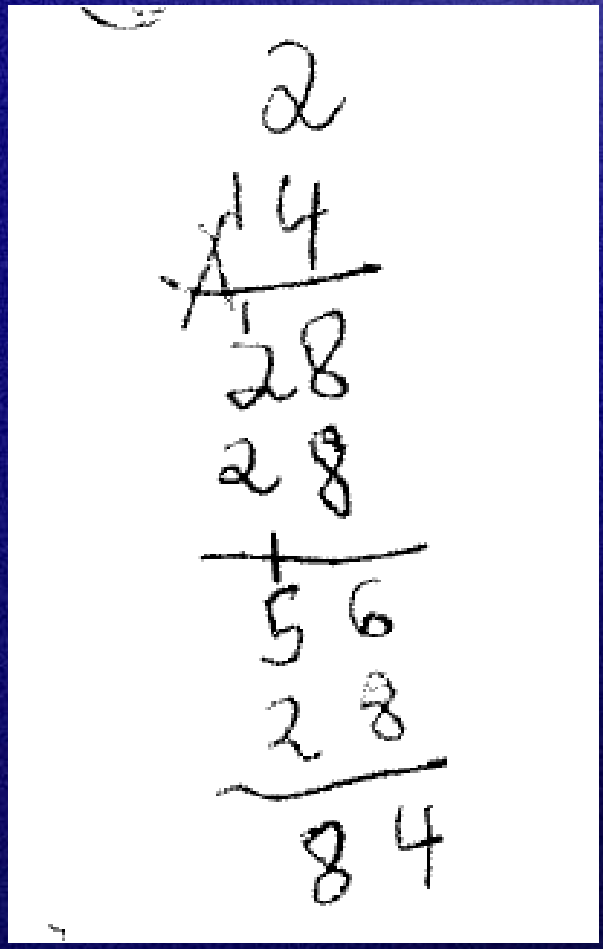
Barnes, Wilkinson, Khemani, Boudesquie, Dennis & Fletcher, 2006

Procedures and decomposition



14
x 6

288/48



Math Accommodations

Is spending with these…

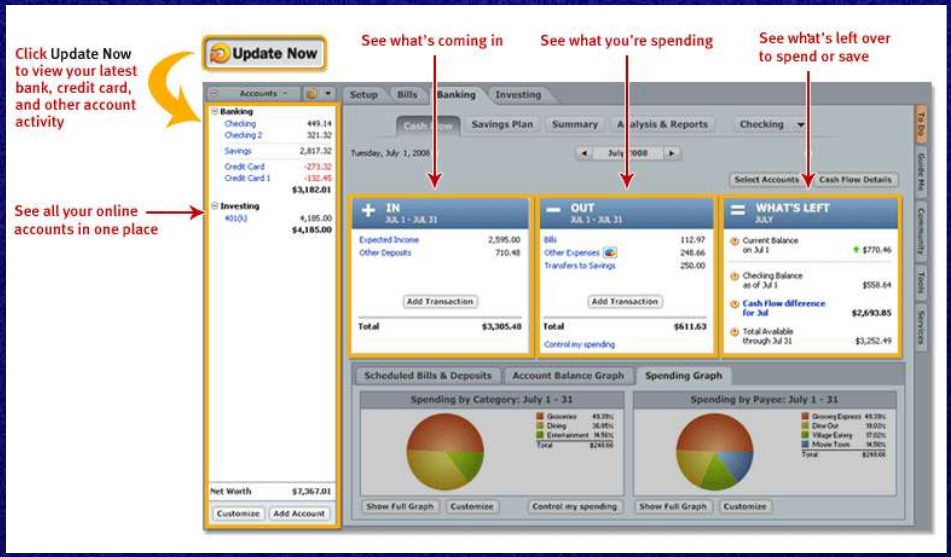
the same as spending with these?

Math Accommodations

Math Accommodations

Is managing this…

the same as managing this?



Math Accommodations

Is buying groceries like this…

the same as buying groceries like this?

Math/Organizational Accommodations

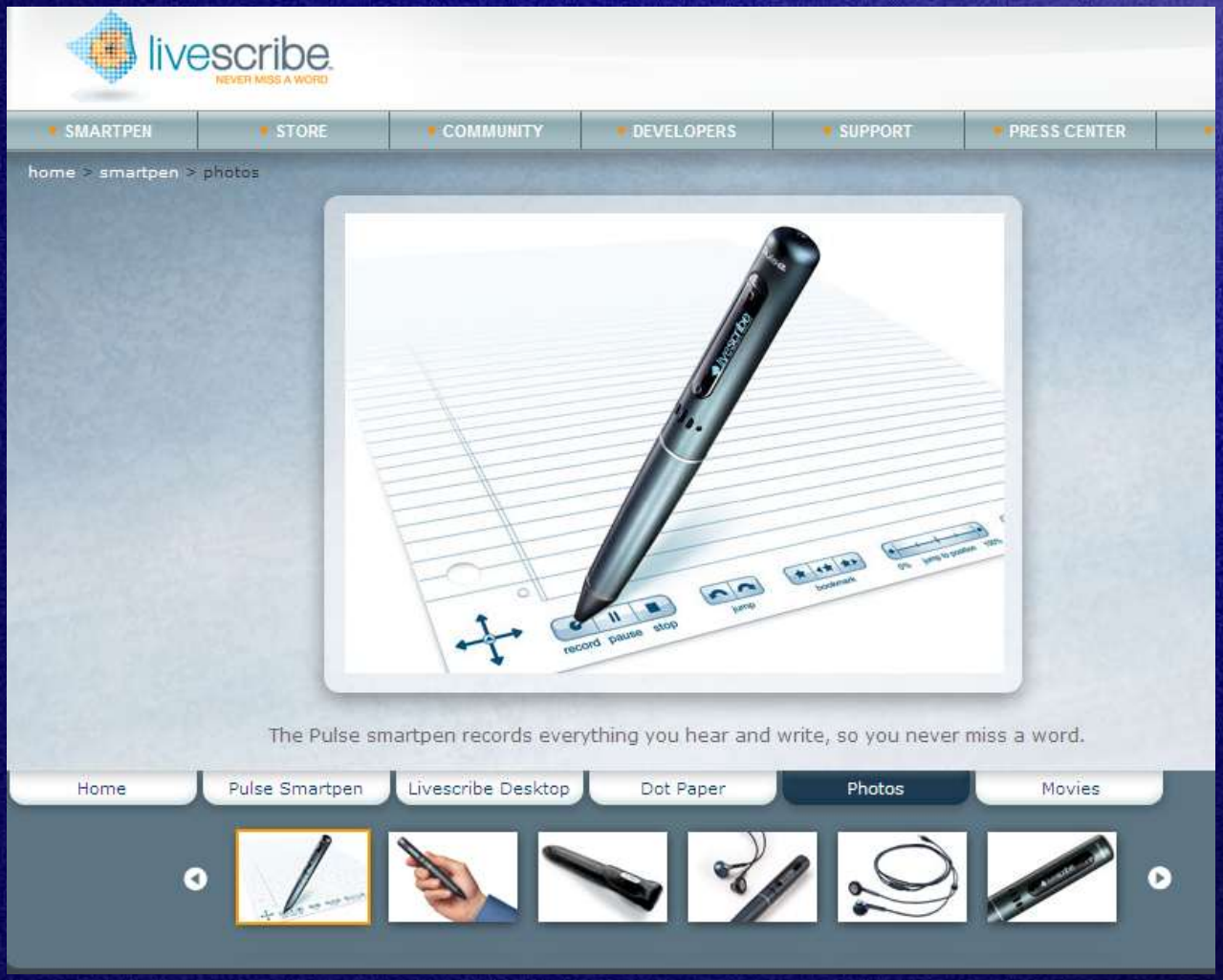
E-mail Notification of Spending

Written Language Accommodations

Is writing with these…

the same as writing with this?

Notetaking


livescribe
NEVER MISS A WORD
SMARTPEN
STORE
COMMUNITY
DEVELOPERS
SUPPORT
PRESS CENTER
home > smartpen > photos
record
pause
stop
jump
bookmark
The Pulse smartpen records everything you hear and write, so you never miss a word.
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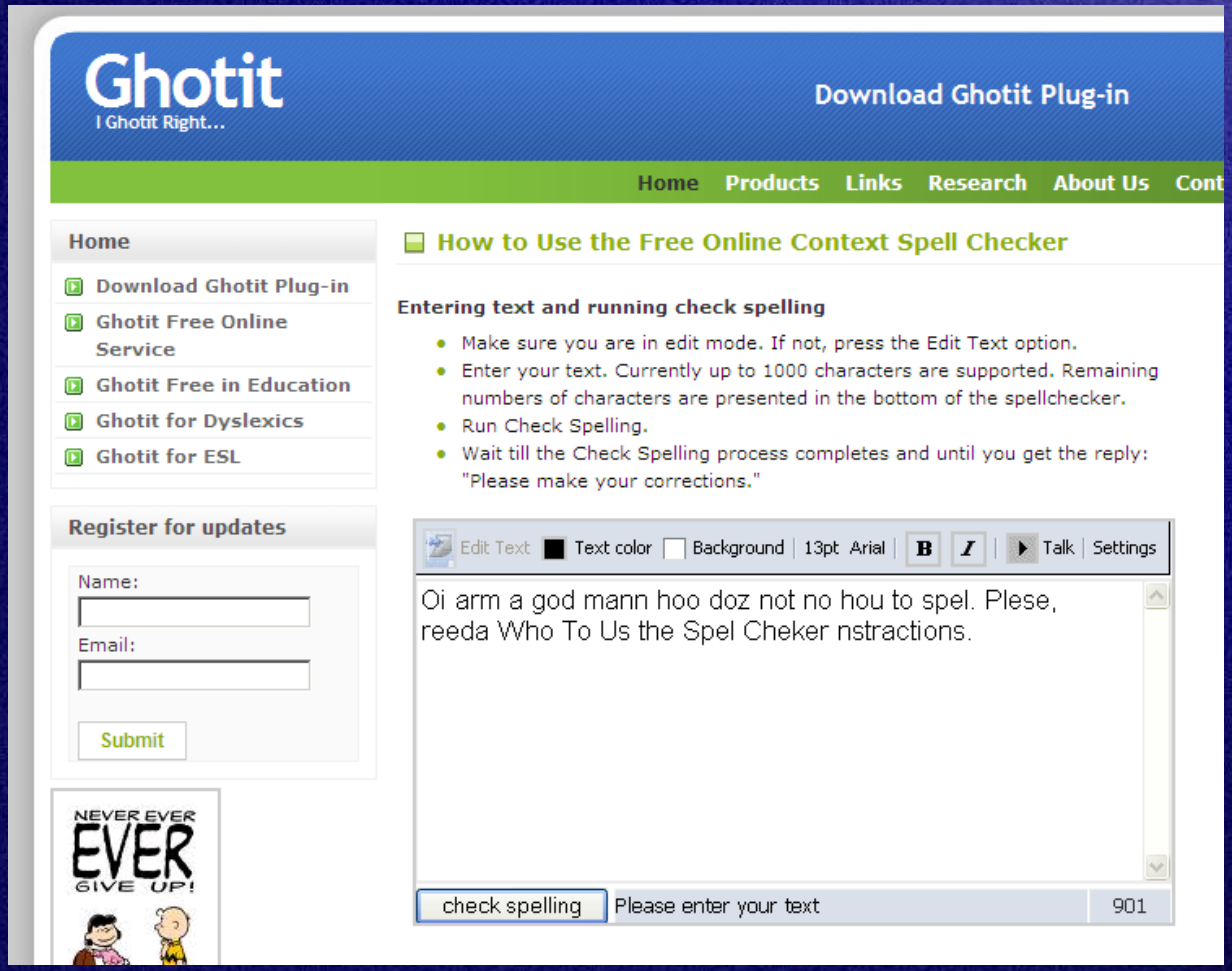
Grammar / Word Usage



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-

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
  - Geenen, S. J., Powers, L. E., & Sells, W. (2003).
- The unemployment rate for the general population is ~10%; for adults with disabilities the rate is between 50% - 75%
  - (Adolescent Health Committee, 2006)

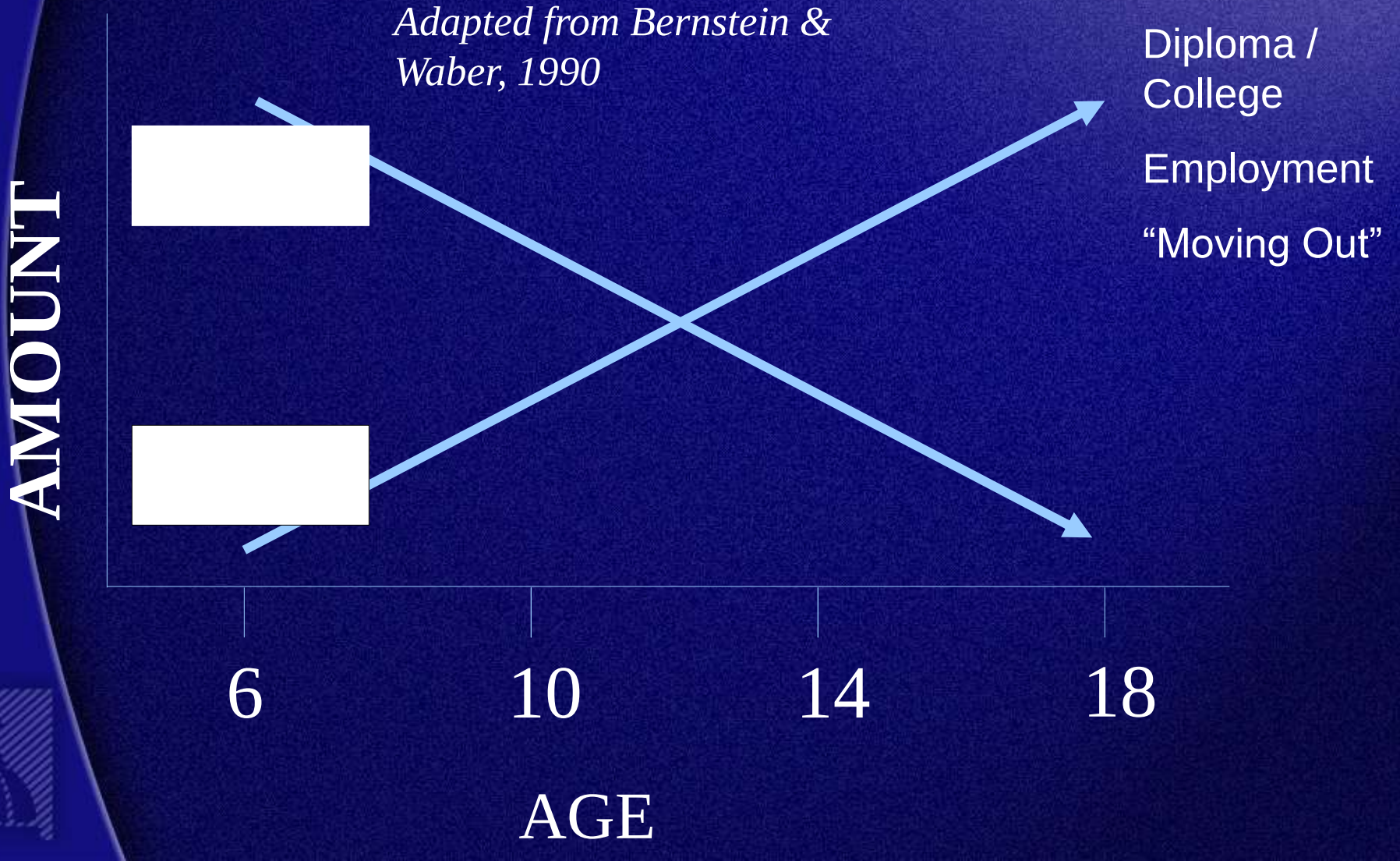
Ongoing areas of NP concern

- 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

Transition to Adulthood



An interaction of AdFx and ExFx

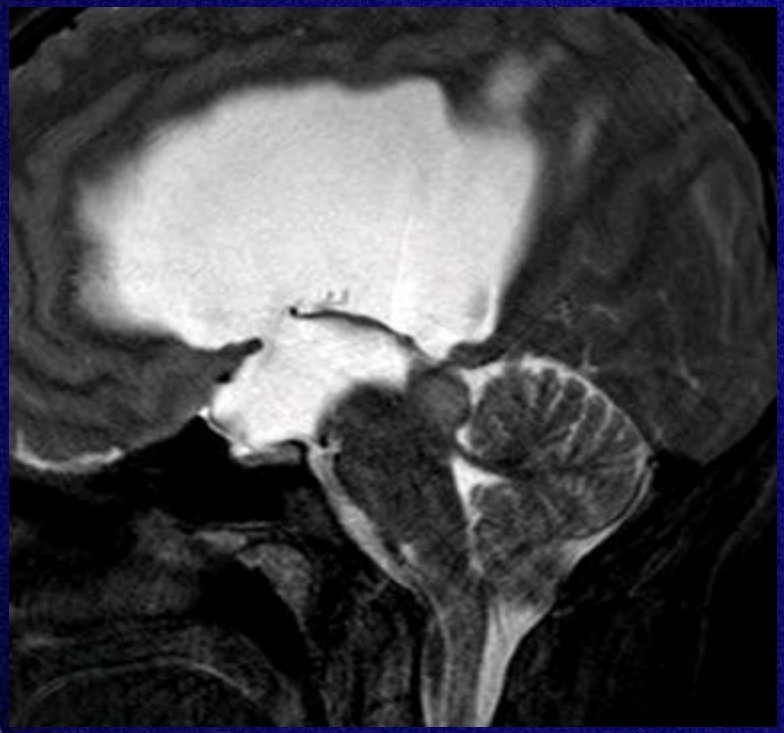
		Adaptive Demands: Typical	Adaptive Demands: High
Executive Ability: Typical		Typically-Developing Adolescents	Diabetes SCI
Executive Ability: Impaired		ADHD, TBI, Hydrocephalus	Spina Bifida, Late-HIV, OSA

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



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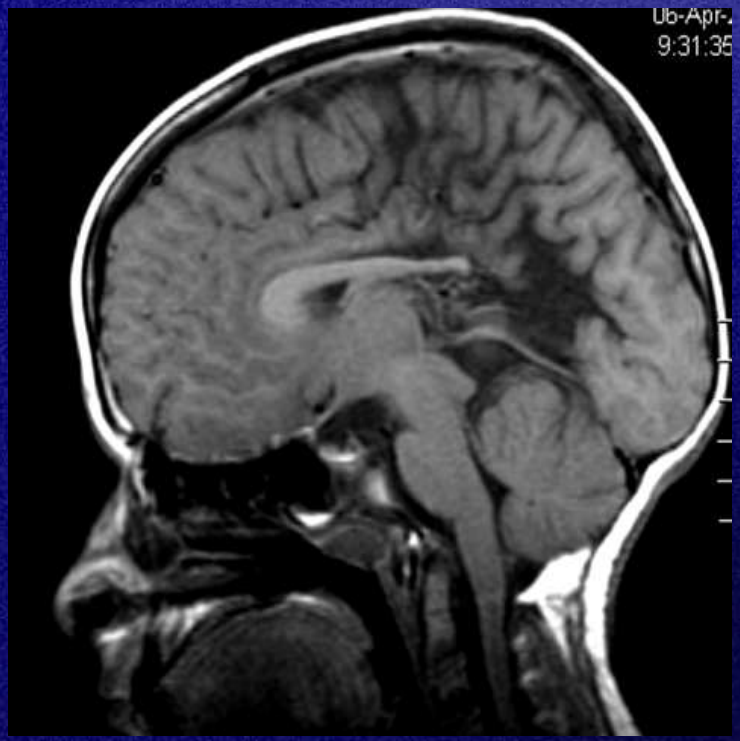


A secondary insult?

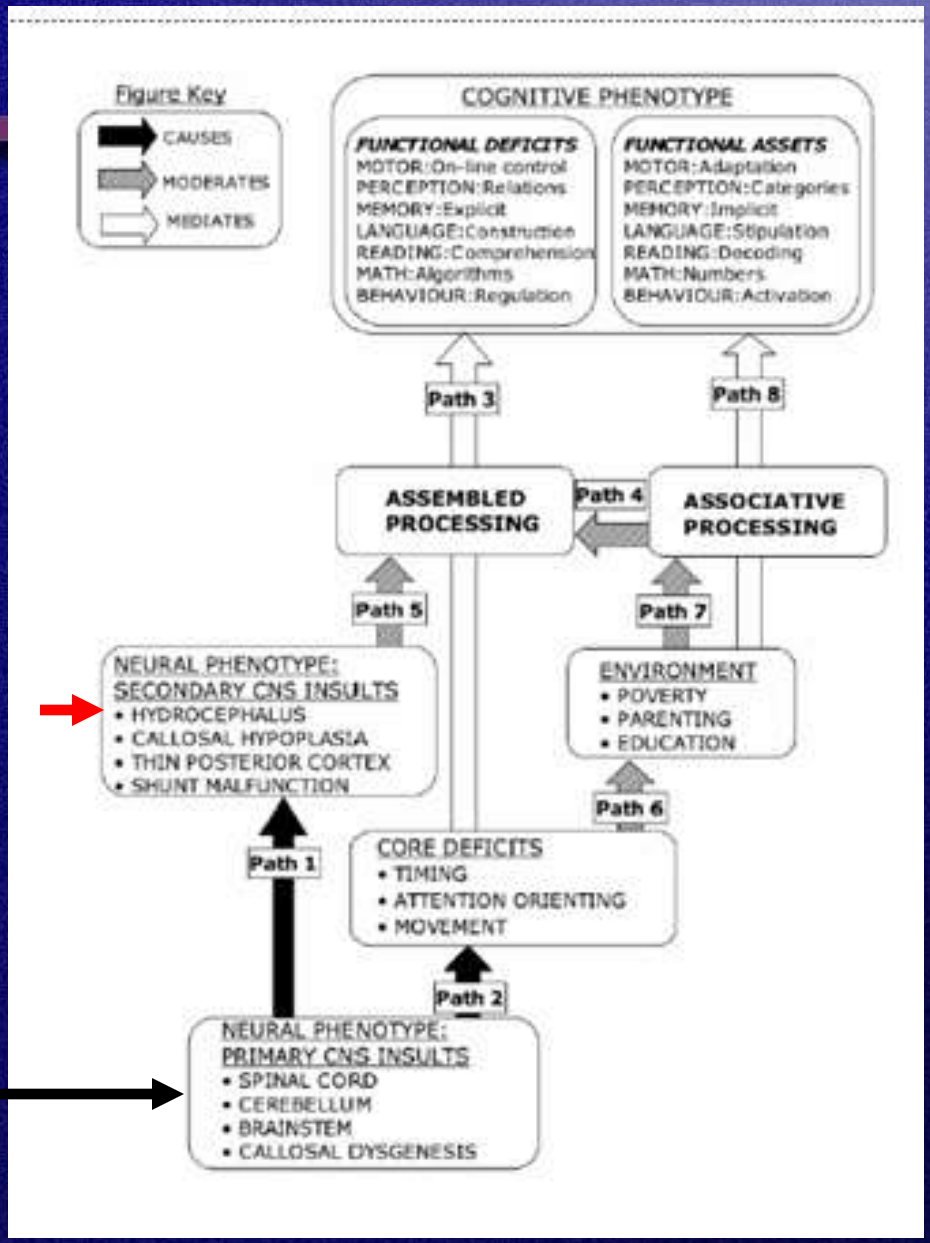
CRITICAL REVIEW

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. FLETCHER[4]

[1]Brain and Behavior Program, The Hospital for Sick Children and the University of Toronto, Ontario, Canada
[2]Department of Pediatrics, University of Texas Health Science Center, Houston, Texas
[3]Department of Psychology, University of Guelph, Guelph, Ontario, Canada
[4]Department of Psychology, University of Houston, Houston, Texas

(RECEIVED August 4, 2005; FINAL REVISION November 7, 2005; ACCEPTED November 10, 2005)



NF, Tectal Tumors, IVH, AS…

Different Conditions

Published in final edited form as:
Neuropsychology. 2008 November ; 22(6): 765–775. doi:10.1037/a0013373.

Sustained Attention in Children With Two Etiologies of Early Hydrocephalus

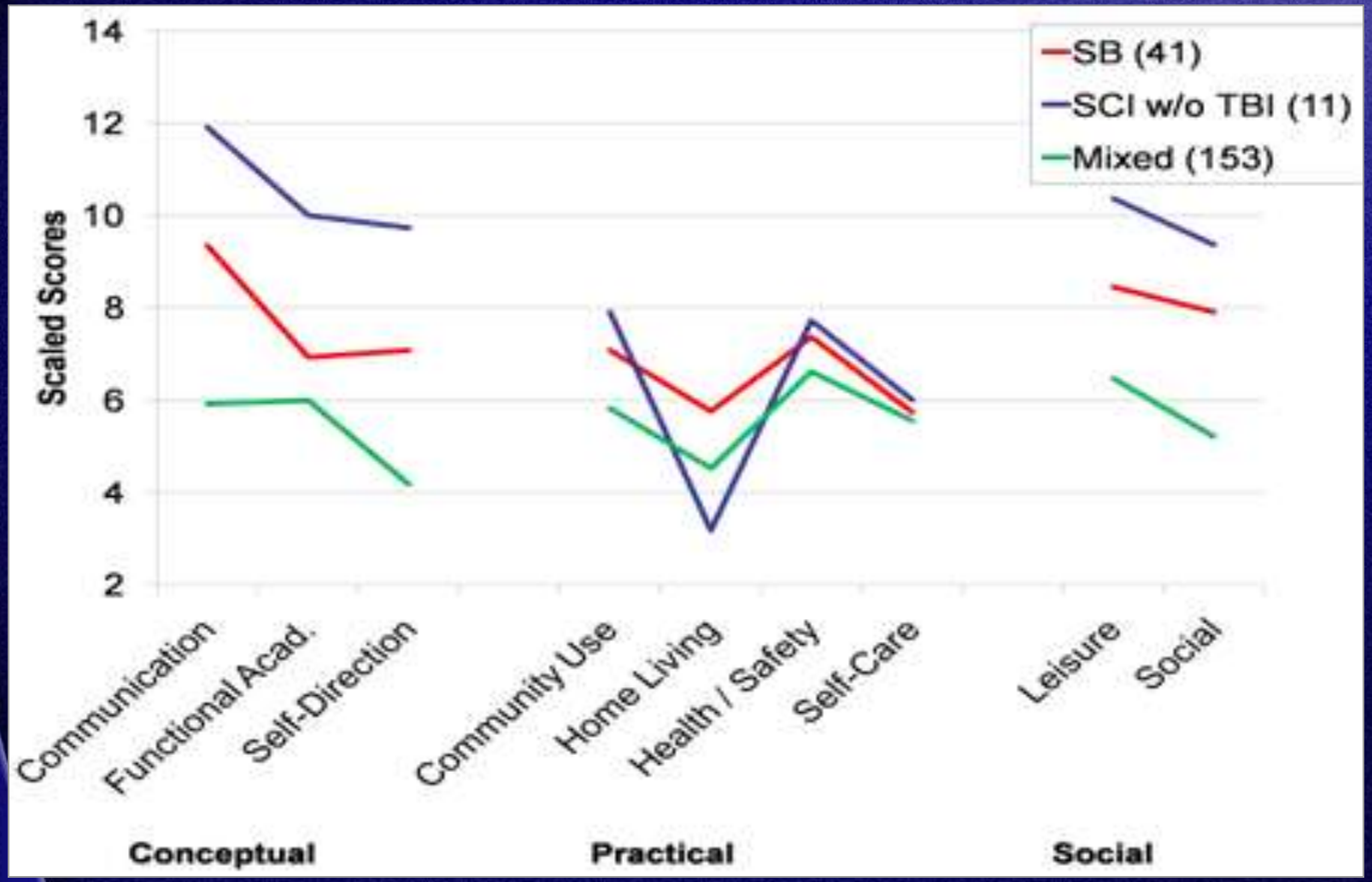
Maegan D. Swartwout, **Paul T. Cirino**, **Amy W. Hampson**, and **Jack M. Fletcher**
Department of Psychology, University of Houston

Michael E. Brandt
Center for Computational Biomedicine, University of Texas Health Science Center at Houston

Maureen Dennis
Program in Neurosciences and Mental Health, The Hospital for Sick Children, and Department of Psychology, University of Toronto.

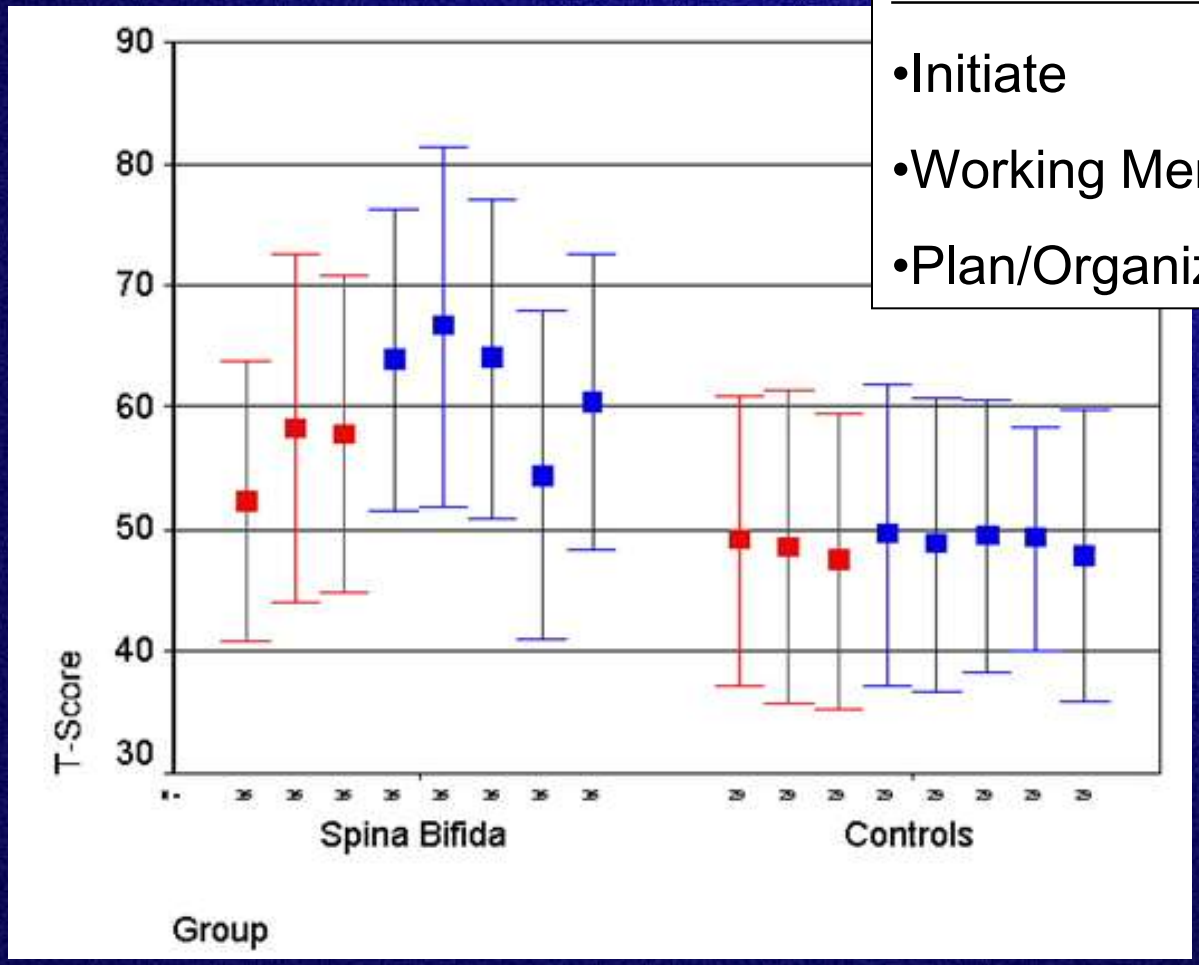
- Gordon Vigilance Task (Gordon, 1983).
- 101 children with SBM, 17 with AS, and 40 normal controls.
- Children with SBM did not differ from AS or NC groups on measures of sustained attention, but they committed more errors and responded more slowly.

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

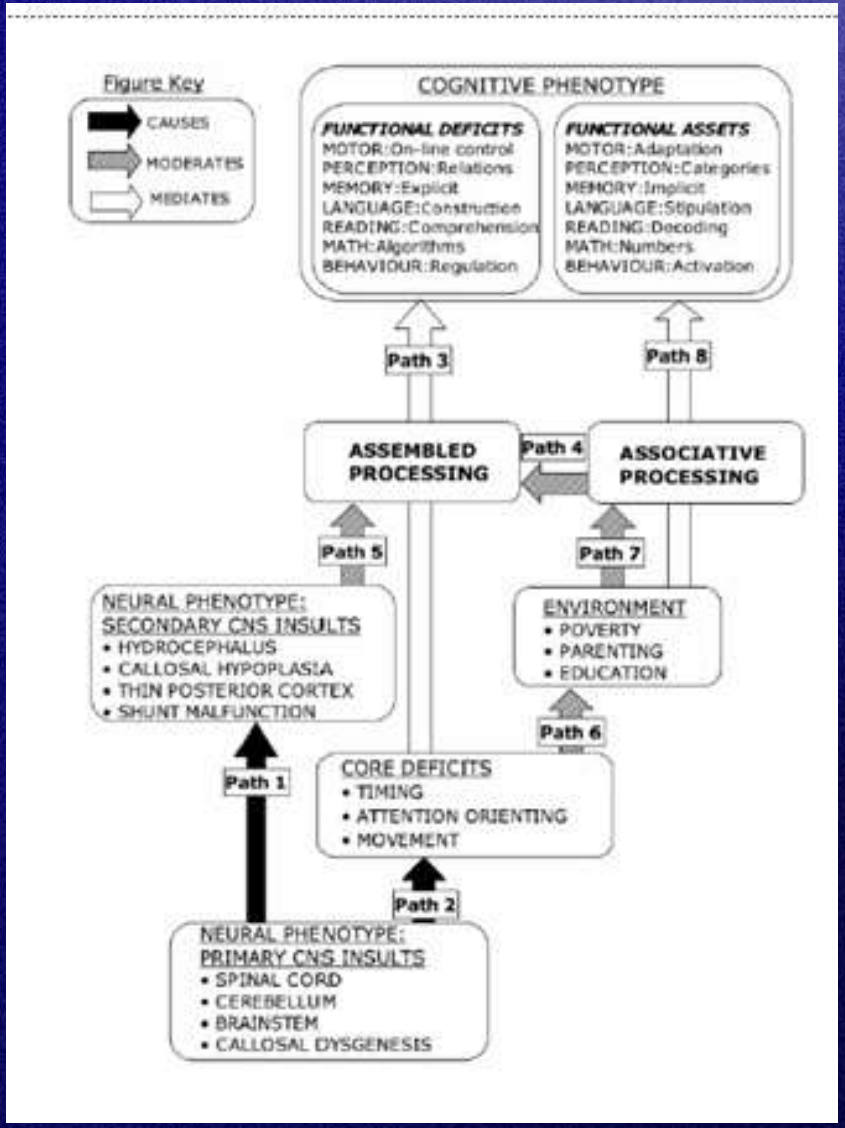
Mahone et al., Child Neuropsychology, 2002

Processing deficits in SB

Associative Processing

vs.

Assembled Processing



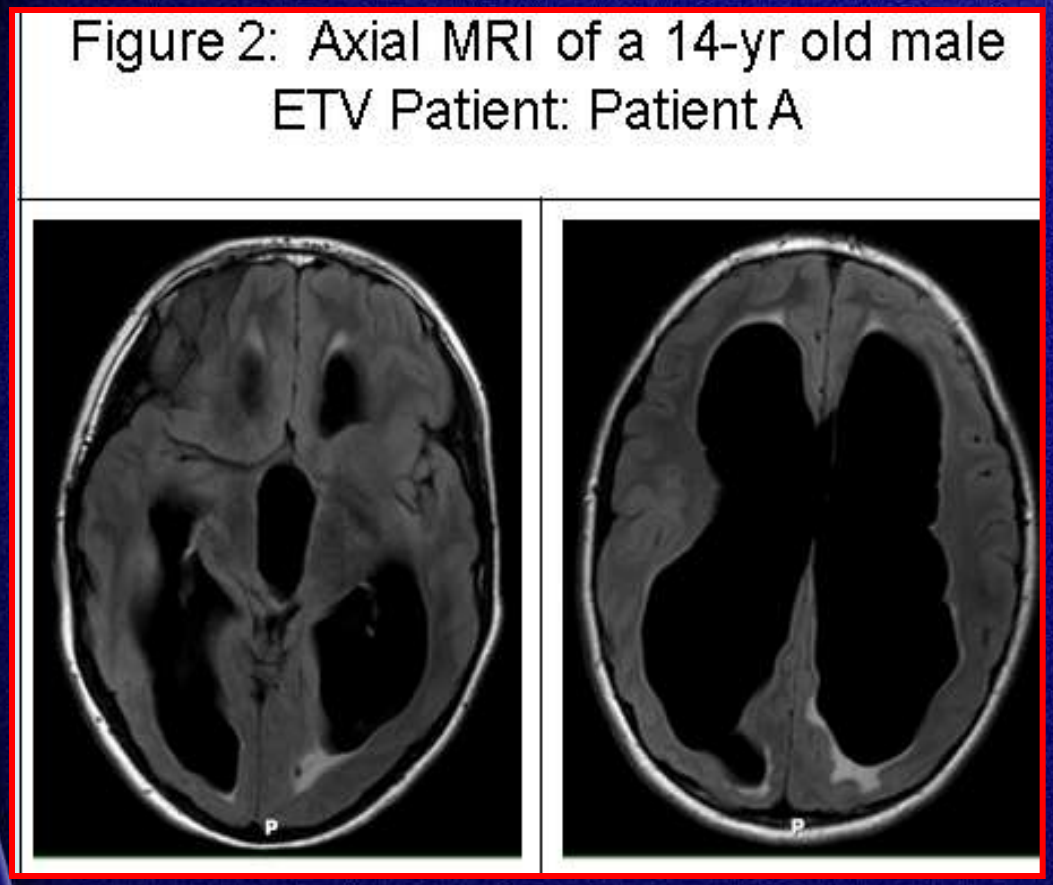
Cognitive impact of shunted hydrocephalus

- 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

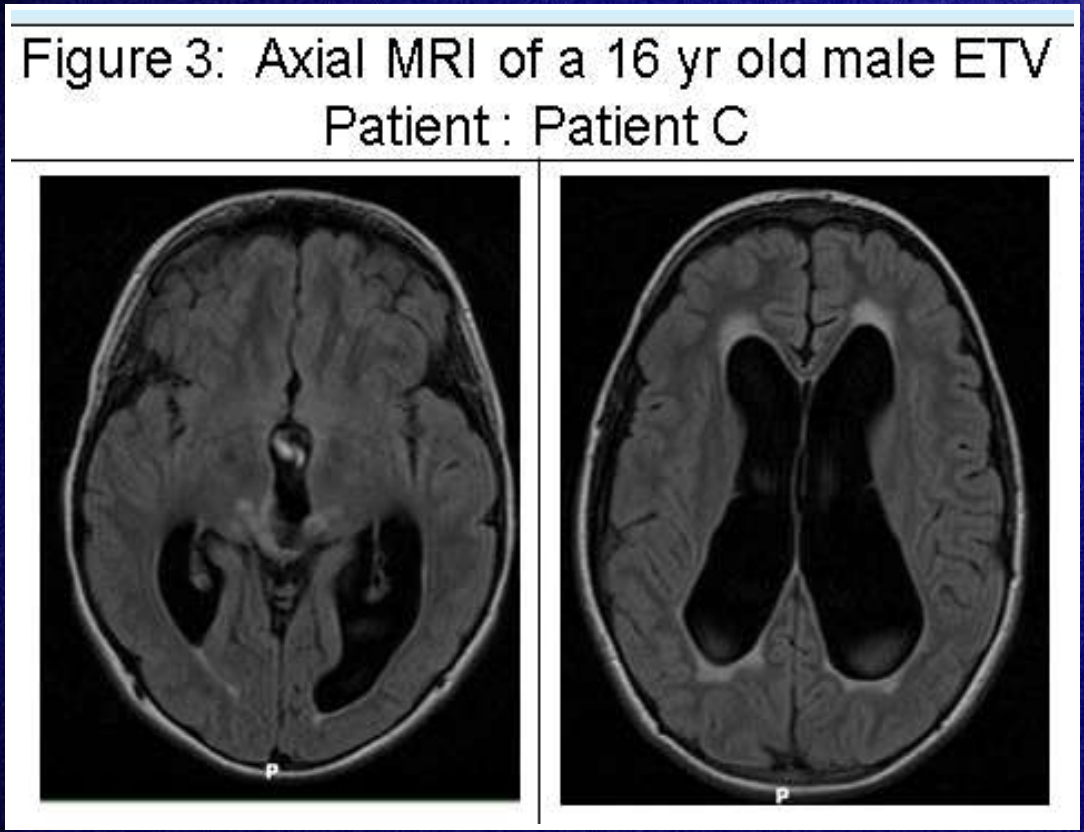
- Burtscher, Bartha, Twerdy, Eisner & Benke (2003)
  - Cognitive improvement following ETV in 6 adults with late onset idiopathic AS.
  - Pre-surgical deficits were noted in anterograde memory and executive functioning
  - Anecdotal improvement noted at the time of post-operative follow-up (an average 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.

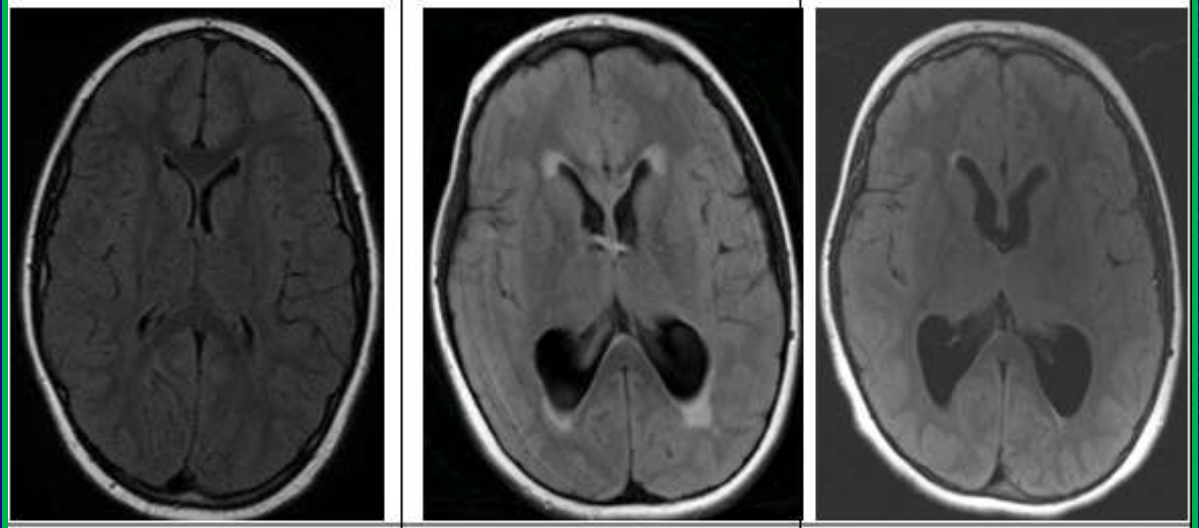
Patient B



- 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

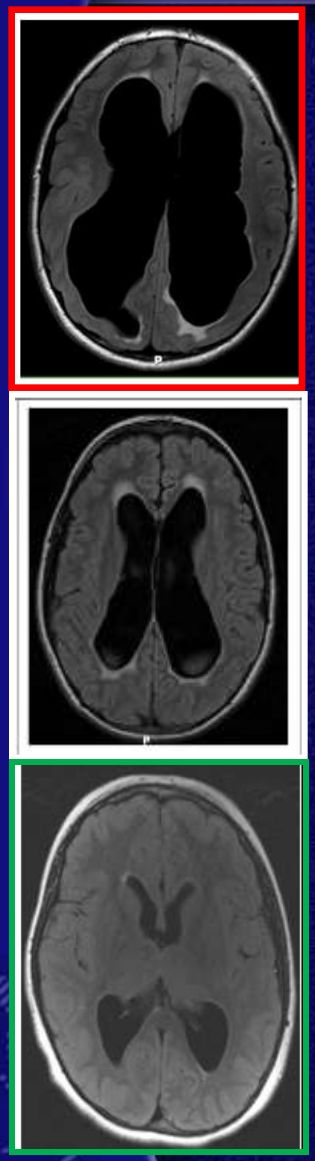
Figure 4: Axial MRI of Typically Developing Child (1) and 10-yr old female ETV Patient : Patient C (2, 3)

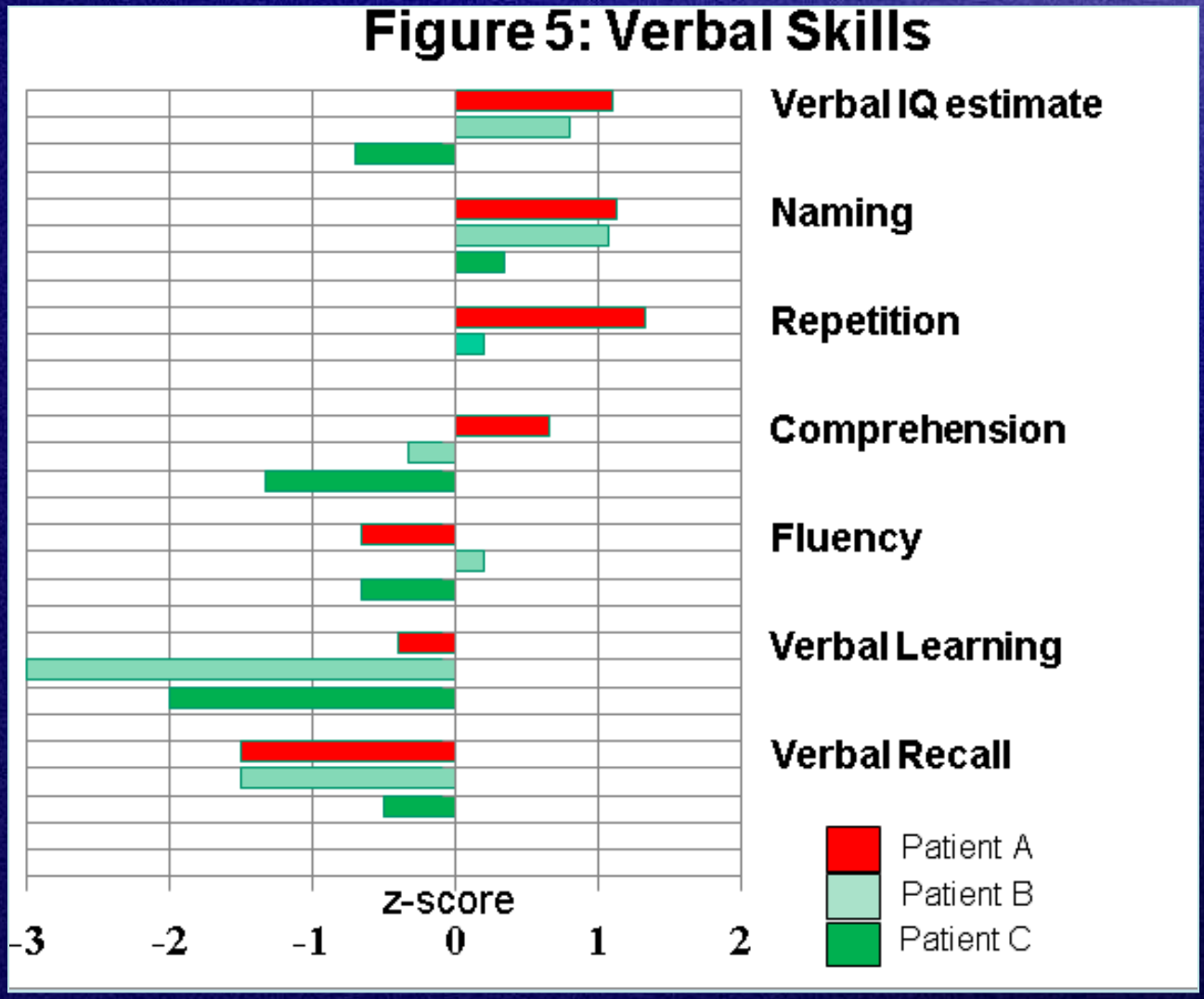


(1) Typically developing 10 Year old	(2) 10-yr old female 1 day post-ETV	(3) 10-yr old female 9 months post-ETV

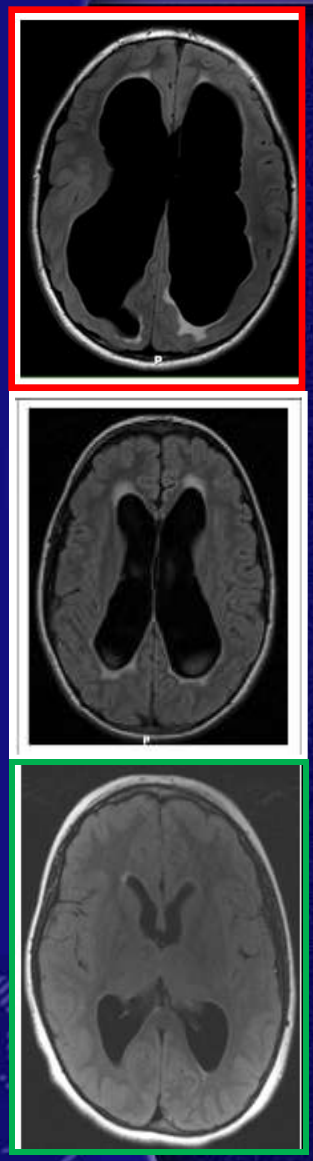
- 10-y.o. girl with ETV performed at age 9
- Intracranial midbrain lesion with OH.
- NF was suspected.
- Strabismus surgery at age 8 (w/o improvement)
- Neuropsychological test scores obtained 1-year post-ETV
- Parent rated executive functioning and adaptive functioning were within age expectations.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

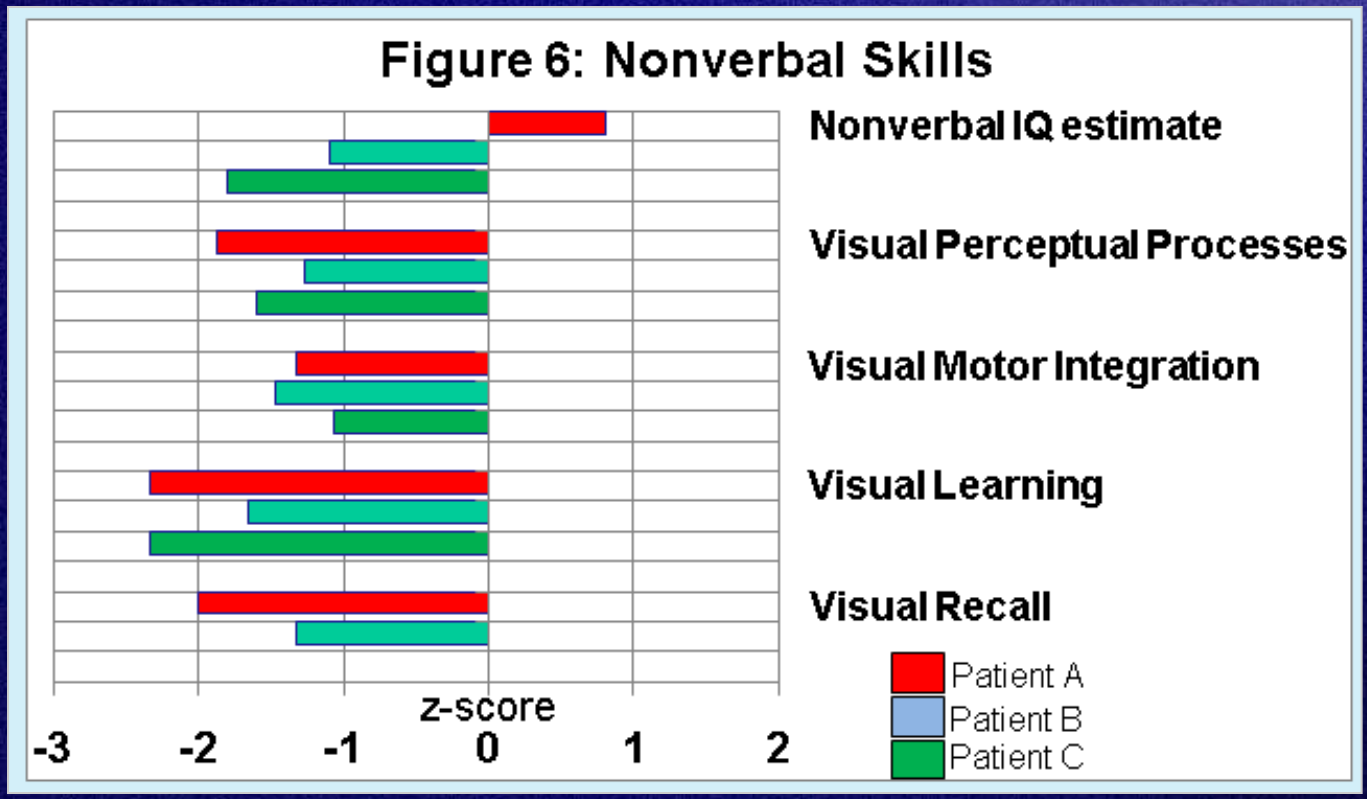
Verbally-based skills





Nonverbal skills





DTI?

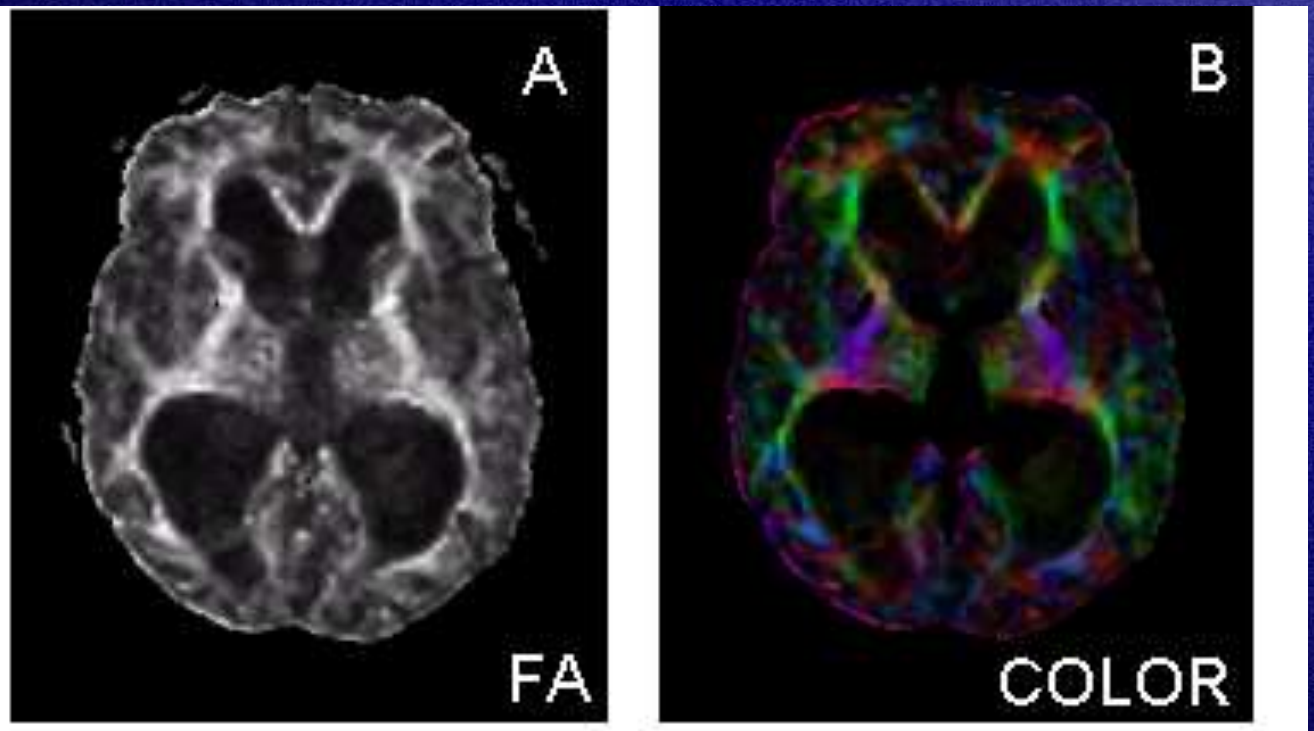


Fig 4. An axial slice from a 16-year-old female pre-surgical ETV patient showing OH in the third ventricle and lateral ventricles. Axial slices are examples of (A) a DTI fractional anisotropy map and (B) a DTI-based color map for the patient.

DTI?

Table 1: Structural size and FA parameters for a pre-surgical ETV patient and three age matched control participants.

	Pre-Surgical ETV Participant (age 16)		Control Participants (n=3; mean age 16 +/- 1.73 years)	
	Size (mm^2)	FA	Size (mm^2)	FA
Genu of Corpus Callosum	**77**	**0.51**	195 +/- 11.4	0.75 +/- 0.01
Splenium of Corpus Callosum	**88**	**0.66**	255 +/- 11.5	0.75 +/- 0.03
Right Cingulate	19	0.35	23 +/- 5.0	0.38 +/- 0.02
Left Cingulate	16	**0.33**	29 +/- 8.9	0.51 +/- 0.06
Fornix	19	**0.26**	27 +/- 4.3	0.50 +/- 0.08

* 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?

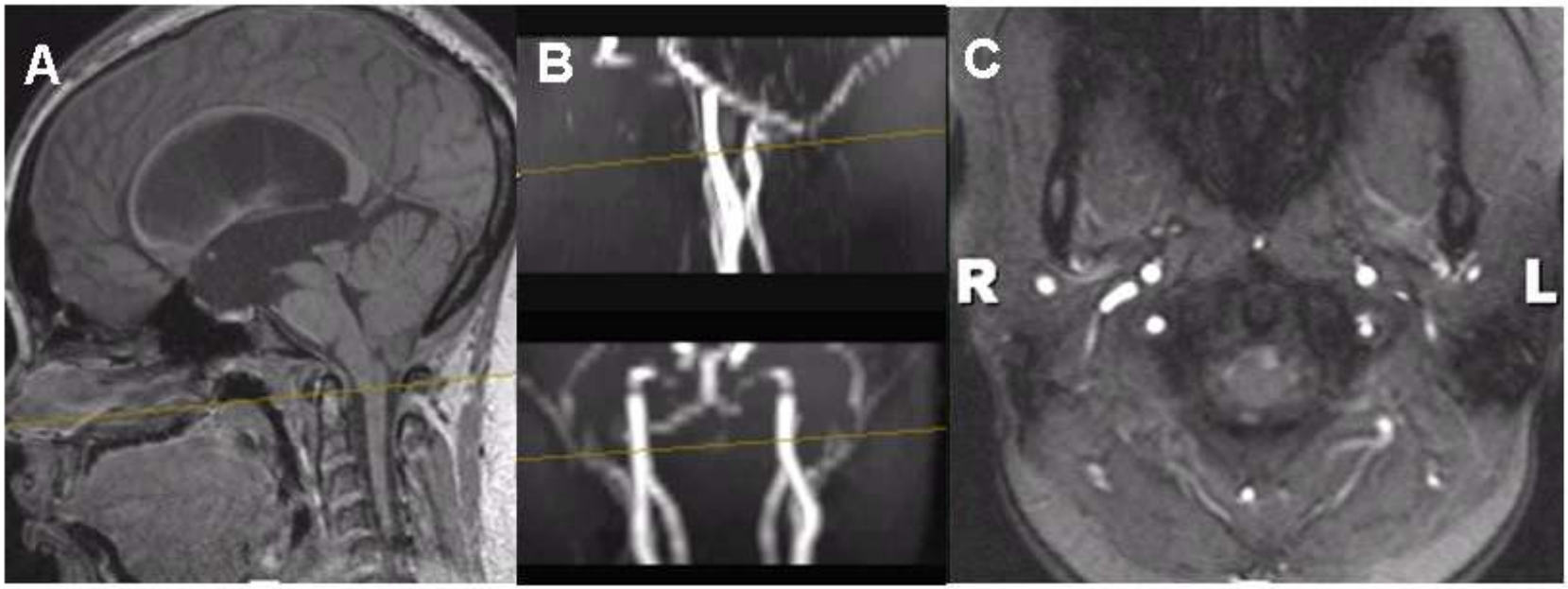


Fig 1: Slice position for PC cine MRI with retrospective ECG-gating on (A) sagittal T1 weighted image in the mid-sagittal plane, (B) sagittal and coronal angiographic scout images as indicated by orange reference lines corresponding to (C) axial anatomic PC image.

MR-ICP?

Table 1: Summary of Hydrodynamic Parameters		
	ETV Patient	*Healthy Control Subject*
tCBF (ml/min)	844	890
tCVO (ml/min)	392	509
CSF area (cm2)	2.14	1.64
Stroke volume (ml)	0.48	0.62
Sys CSF velocity (cm/s)	1.49	2.21
PTP-PG (mmHg/cm)	0.038	0.045
ICVC (ml)	0.5	0.61
ICCI (mL/[Pa·m])	6.14	8.26

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?