SPONSORED BY SEATTLE CHILDREN'S OUTREACH EDUCATION, THE DIVISION OF DEVELOPMENTAL MEDICINE AND THE SEATTLE CHILDREN'S CENTER FOR CHILDREN WITH SPECIAL NEEDS

39th Annual Duncan Seminar

Spina Bifida

Promoting Wellness and Preventing Pitfalls

Friday, April 20, 2018

7:15 AM - 4:30 PM

Wright Auditorium

Seattle Children's Hospital

4800 Sand Point Way NE, Seattle, WA 98105

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

39th ANNUAL DUNCAN SEMINAR

Spina Bifida - Promoting Wellness and Preventing Pitfalls

FRIDAY, April 20, 2018

2018 PARTNERS

Washington State Department of Health
Children with Special Health Care Needs Program

Center on Human Development and Disability

Leadership Education in Neurodevelopmental Disabilities Training Program
University of Washington

Seattle Children's
HOSPITAL · RESEARCH · FOUNDATION

39TH ANNUAL DUNCAN SEMINAR

SPINA BIFIDA – PROMOTING WELLNESS AND PREVENTING PITFALLS

April 20, 2018

Information for Participants

Welcome! Thank you for joining us today. We hope you will find many important resources at this conference to help you better care for children and youth with developmental disabilities.

Pagers and Cell Phones: In consideration of the presenters and other conference guests, **please silence your phones and pagers.**

Restrooms and ATM Machines: Restrooms and ATM machines are located outside the auditorium in the hospital lobby to your left, across the hall near the vending machines. Additional restrooms can be found in the lobby to your right near the River front entrance.

Continental Breakfast, Breaks and Lunch: Meals, snacks and beverages will be available in the auditorium lobby where the registration tables are located.

Name Badges: Your name badge allows you access to the auditorium, restrooms and other public areas within the hospital. Should you be interested in visiting patient areas, please stop by the Greeter's Desk and obtain a standard Children's photo visitor badge by presenting your driver's license or other photo ID. We appreciate your consideration of our efforts to protect the safety of our patients and families.

Resource Room and Poster Session: Please visit the resource room for local program information, resources and education for caregivers. Room is RC 905 located near the River Entrance.

CE Certificates: Enclosed in your syllabus are directions to access the online evaluation after the event has taken place. At the conclusion of the online evaluation you will be able to print either your **Certificate of Participation** from **your own computer**.

Clock Hours For School Employees: Puget Sound ESD Clock Hour registration forms are available at the registration table as you check in. Be sure to **sign the Clock Hour registration form** and pick up a Clock Hour registration packet. Complete the registration form and return it to the Puget Sound ESD with a check in the amount of $14.00. Do not sign section IV, verification of payment. Please follow the instructions in the Clock Hour packet on completing the Puget Sound ESD online evaluation from your own computer.

Evaluations: Please remember to complete the **Online Course Evaluation** within 5 business days. We value your feedback to help us plan future events. Directions to complete your online evaluation and print your continuing education certificate can be found in the following pages.

Questions: Feel free stop by the registration table if you have any questions or need assistance finding your way. We are here to help.

Thank You

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

39th Annual Duncan Seminar

Spina Bifida - Promoting Wellness and Preventing Pitfalls

Course Schedule

Friday April 20, 2018 | 7:45 AM – 4:30 PM | Seattle Children's Hospital, Wright Auditorium

7:15 am	Registration/Continental Breakfast (provided)
7:45 am	**Welcome and Opening Remarks** Jeff Sperring, MD, CEO
8:00 am	**Family Panel: Patient and Family Perspectives on Living Life with Spina Bifida** *Facilitator:* Timothy John Brei, MD, FAAP
9:00 am	**Days 18 – 28: Ten Days that Change Everything for Everybody** William O. Walker, Jr., MD
10:00 am	Break, Resource Room and Poster Browsing
10:15 am	**Keynote Speaker: Psychosocial and Family Functioning in Youth and Young Adults with Spina Bifida: What Have We Learned So Far?** Grayson N. Holmbeck, PhD
11:30 am	**2018 DUNCAN AWARD PRESENTATION** *Recipient:* **Cathy Graubert, PT, Ambulatory Rehabilitation Manager**
12:00 pm	Lunch (provided)
12:45 pm	**Physical Therapy and the Management of Children and Individuals with Spina Bifida (MM)** Solveig Hart, PT, MSPT, PCS
1:45 pm	**Fostering Academic Success: Identifying and Addressing the Learning Needs of Children with Spina Bifida** Kate Bowen, PhD; Emily Myers, MD; Hillary Shurtleff, PhD, ABPP
2:45 pm	Break, Resource Room and Poster Browsing
3:00 pm	**Promoting Optimal Feeding and Nutrition in Children with Feeding Difficulties and Spina Bifida** Kim Nowak-Cooperman, MS, RDN; Peggy Smith, OTR/L, BS
4:00 pm	**From Clinical Knowledge to Practical Application: Q&A with the Experts** Lisa Herzig, MD
4:30 pm	**Evaluation/ Adjourn** (15 minutes are included in CE hours)

2018 Duncan Seminar

Family Panel

Moderated by: Timothy John Brei, MD, FAAP
Co-moderated by: Sarah Coburn, RN, BSN, CPN
Date: April 20, 2018

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Disclosure Statement

- I Do not have any conflict of interest, nor will I be discussing any off-label product use.
- This class has no commercial support of sponsorship, nor is it co-sponsored.

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Objectives

- Develop an awareness of challenges and successes patients with Spina Bifida face in home, school, community and health care settings.
- Apply information gained from reported family experiences into your own practice.

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

10 Days That Change Everything (For Everybody)

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

William O. Walker Jr. MD
Robert A. Aldrich Professor, Pediatrics
Seattle Children's Hospital
University of Washington School of Medicine

Conflict of Interest

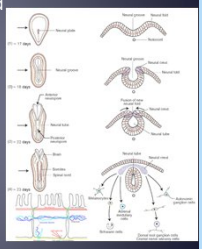
- I do not have any conflict of interest, nor will I be discussing any off-label product use.
- This presentation has no commercial support or sponsorship, nor is it co-sponsored.

Learning Objectives

- Know the etiology and epidemiology of neural tube defects with a specific emphasis on prevention
- Know the impact of genetics on the occurrence of neural tube defects
- Know the lifespan impact of neural tube defects on affected children and their families.

Neural Tube (NT)

- Develops into brain and spinal cord
- Closed by the 28th day post-conception (before knowledge of pregnancy)
- Defect occurs with failure of the neural tube to "zip up"
- Four separate sites along NT where closure occurs
- Most NTDs can be explained by failure at one or more closure sites



Epidemiology

- Based on 2009- 2011 data, the estimated average annual prevalence of anencephaly and spina bifida combined was 6.5 cases per 10 000 live births.
- Worldwide, approximately 300,000 infants are born annually with an NTD causing increased mortality, morbidity, disabilities, and economic burden.
- NTDs account for as many as 29% of neonatal deaths associated with congenital abnormalities in low-income settings.
- Currently, there are more adults with spina bifida than children with spina bifida.

ETIOLOGY

Folic Acid

- Half of all pregnancies in the United States are unplanned.
- The critical period for supplementation starts at least 1 month before conception and continues through the first 2 to 3 months of pregnancy.
- NTD prevalence declined from 10.7 cases per 10 000 live births before the implementation of food fortification (1995-1996) to 7.0 cases per 10 000 live births after fortification (1999-2011).
- Folic acid supplementation prevents about 1300 annual births from being affected by neural tube defects, according to recent estimates

Folic Acid

- About 50%to 72% of NTDs are preventable by adequate folic acid supply in the critical period of organogenesis.
- The precise mechanism through which folic acid prevents NTDs has not been fully defined.
- Not all NTDs are preventable through folate acid supplementation.
- Folate-resistant NTDs include those associated with poor glucose control in the first trimester, hyperthermia, maternal obesity, and aneuploidy or genetic disorders.

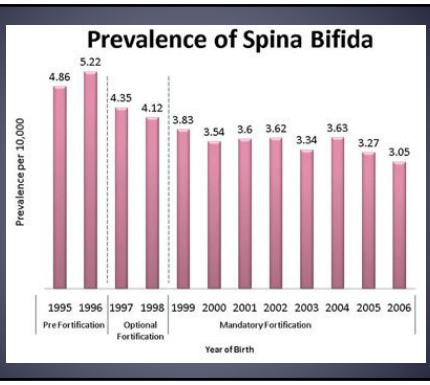
Folic Acid Recommendations (USPSTF Feb 2017)

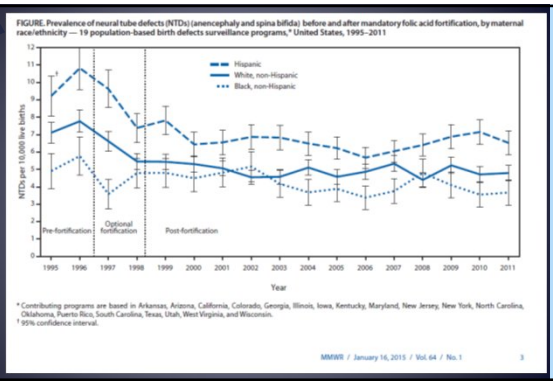
- Continue recommendation that all women planning or capable of pregnancy take a daily supplement containing 0.4 to 0.8mg (400 to 800 µg) of folic acid.
  - Reduce their risk of having a pregnancy affected by a neural tube defect.
  - Evidence Grade A. The USPSTF recommends the service.
  - There is high certainty that the net benefit is substantial.
- Found no new substantial evidence on the benefits and harms of folic acid supplementation that would lead to a change in its recommendation from 2009.

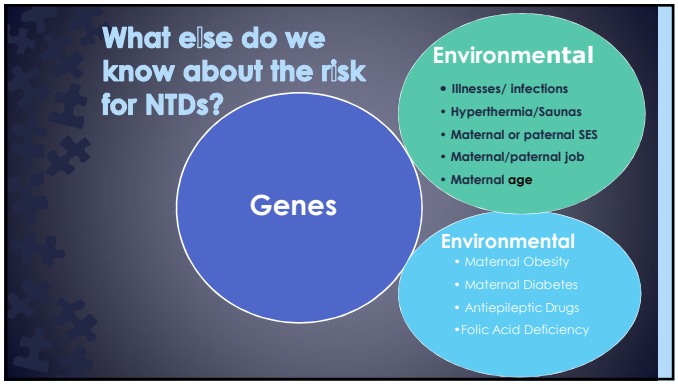
Folic Acid Recommendations (USPSTF Feb 2017)

- Similar recommendations have been in place since 1992, yet less than one-third of reproductive-age US women take a daily supplement containing folic acid.
- In some subgroups, the proportion of women who take a daily supplement is even lower
  - Hispanic women, about 23%
  - women with unintended pregnancies, about 16%

0.4 mg vs 4 mg







Genetics of Neural Tube Defects

- Strong evidence for a significant genetic component (estimated heritability of 60%) to the etiology of NTDs.
- >200 genes have been identified in animal models of NTD.
- Limited success identifying the responsible genetic variations in humans.
- Evidence for genetic causation includes
  - the high recurrence risk for siblings of index cases (2-5%), approximately 50-fold more than in the general population, together with a gradually decreasing risk in more distant relatives.
  - Women with two or more affected pregnancies have a very high risk (~ 10%) of further recurrence.
  - NTD prevalence is greater in like-sex twins (which are assumed to include all monozygotic cases) compared with unlike-sex pairs, consistent with a significant genetic component.

Genetics of Neural Tube Defects

- Isolated (nonsyndromic) NTDs are generally multifactorial, or attributed to a complex combination of genetic and environmental factors.
- Specific factors associated with NTDs:
  - Environmental exposures; certain medications; maternal medical conditions; geographic and ethnic associations; genetic etiologies, including chromosomal abnormalities and single gene disorders; and family history
- Understanding genetic susceptibility to NTD lags far behind that of other common structural birth defects.

Are Things Better Now Than They Were?

KEEP CALM
IT'S ONLY
EXTRA
CEREBROSPINAL
FLUID

"Drain for the Brain"

1956

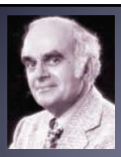
"The baby was suffering from a stubborn form of hydrocephalus (water on the brain): spinal fluid collecting in his skull cavity, caused his head to enlarge and threatened to squeeze the brain so that the child's mental development would be arrested.

…Neurosurgeon Eugene Spitz, 37, tried running a tube direct from baby Casey' head to his abdomen. It worked only for a few days at a time, then another operation was needed to clean it.

…That night (John) Holter went home (Philadelphia) and stared at his drawing board. He drew the design of a valve with two fins that opened and shut like the gates of a canal lock. But what to make it of?

…Finally Holter hit upon silicone plastic fins in a stainless steel body, and a plastic –molding company made up several sample valves. …"

Time Magazine – October 29, 1956



Jack Lapides, MD
(1914 – 1995)

CIC

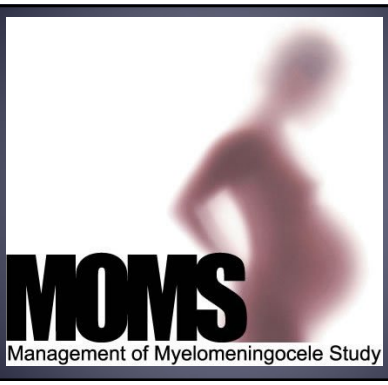
1972

CLEAN, INTERMITTENT SELF-CATHETERIZATION IN THE TREATMENT OF URINARY TRACT DISEASE

JACK LAPIDES, ANANIAS C. DIOKNO, SHERMAN J. SILBER AND BETTE S. LOWE

From the Section of Urology, Department of Surgery, University of Michigan Medical Center, Ann Arbor, Michigan

(Reprinted from J Urol, 107: 458–461, 1972)



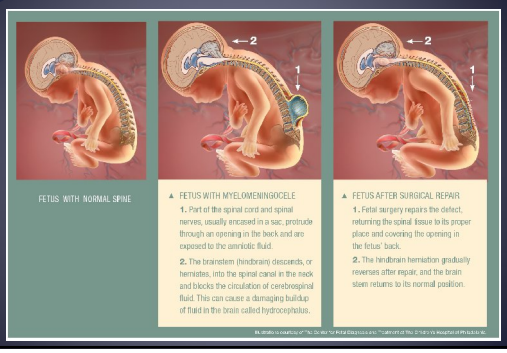
MOMS

Management of Myelomeningocele Study

2003 – 2010

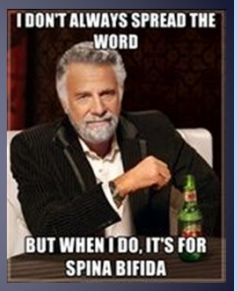
The "Two-Hit" Hypothesis

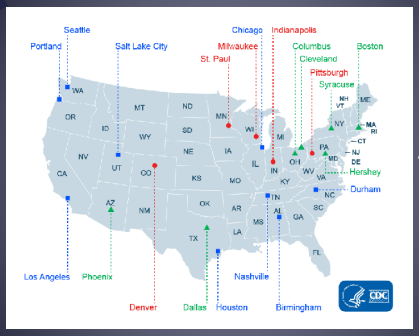
- The neural damage in MMC may be primarily the result of defective spinal cord development, a secondary event resulting from damage to the exposed spinal cord by the intrauterine milieu, or both – the 'two-hit hypothesis'.
- The two-hit hypothesis states that primary congenital abnormalities in anatomic development allow a relatively normal spinal cord to become secondarily damaged by amniotic fluid exposure, direct trauma, hydrodynamic pressure, or a combination of these factors.
- It is this secondary damage which may be ameliorated by early fetal surgical repair.



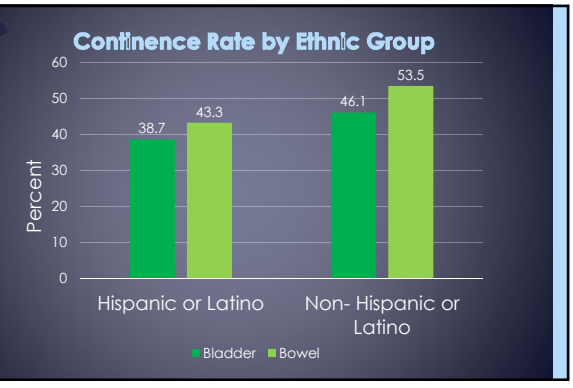
American Journal of Obstetrics & Gynecology FEBRUARY 2018

What's Next for NTDs?





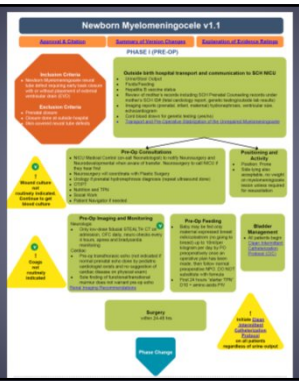
2009 – Current

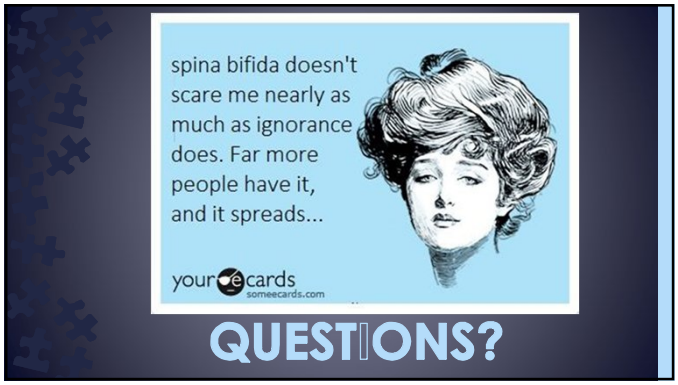


Clinical Standard Work Pathway
Newborn Myelomeningocele
September 2016

Objective:

To develop and standardize evidence – based approach to the care of the newborn with meningomyelocele during their initial hospitalization.




spina bifida doesn't scare me nearly as much as ignorance does. Far more people have it, and it spreads...
yourecards
someecards.com
QUESTIONS?

Psychosocial and Family Functioning in Youth with Spina Bifida: What Have We Learned So Far?

Grayson N. Holmbeck, Ph.D.
Professor and Director of Clinical Training
Principal Investigator, CHATS
Loyola University Chicago
Department of Psychology
1032 W. Sheridan Road
Chicago, IL 60660
773-508-2967
gholmbe@luc.edu

39th Annual Duncan Seminar: "Spina Bifida: Promoting Wellness and Preventing Pitfalls"
April 20, 2018

Disclosure Statement

- I do not have any conflict of interest, nor will I be discussing any off-label product use.
- This class has no commercial support or sponsorship, nor is it co-sponsored.

Objectives

- Explain relevant developmental issues for children, adolescents, and young adults with spina bifida.
- Describe the use of theoretical models for the study of youth and young adults with spina bifida.
- Describe the current state of knowledge regarding families, peer relationships, and psychosocial adjustment in this population.
- Discuss how a camp-based intervention may promote independence and self-management in youth with spina bifida.
- Identify key constructs in the study of self-management, the transfer of medical responsibility from parent to child, and the transition from pediatric to adult health care in this population.
- Discuss clinical implications of this research for youth and young adults with spina bifida.

chats

chats

Chicago Healthy Adolescent Transition Study

chats

Thank you

- The many families, teachers, and health professionals who have given their time
- Numerous current and past undergraduate and graduate students

Ann & Robert H. Lurie Children's Hospital of Chicago

ISBA
Illinois spina bifida association

Loyola University Health System
Loyola University Chicago
We also treat the human spirit.®

Shriners Hospitals for Children® Chicago

Riley Hospital for Children
Indiana University Health

chats

Thank you – Funding Sources

- NICHD (R01-HD048629)
- NINR (R01-NR016235)
- March of Dimes
- The Kiwanis Neuroscience Research Foundation (Kiwanis Illinois-Eastern Iowa District)

Overview

- Background: Spina Bifida (SB)
- Developmental issues: Why are we studying adolescents and young adults with SB?
- Models, Methods, and Procedures from CHATS-1 (1993-present) and CHATS-2 (2005-present)
- Research Findings
  - Psychosocial Adjustment of Youth with SB
  - Family Relationships
  - Parents and Parenting
  - Self-management
- Research Findings from Camp Independence (2006-present)
- Take Home Messages and Clinical Implications of Research Findings

What is Spina Bifida?

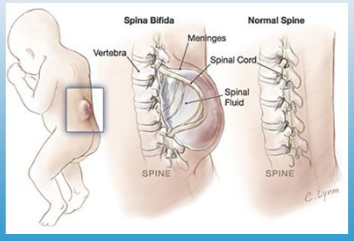
- Most common congenital birth defect that affects the CNS (18 per 100,000 live births, CDC, 2011)
- Also known as a neural tube defect
- Caused by a failed closure of vertebrae during early weeks of pregnancy
- Over 150,000 individuals with SB in the US with an estimated lifetime cost of $600,000 per child
- Produces urinary, bowel, orthopedic, educational, social, and neurological difficulties (hydrocephalus, Chiari II malformation)

What is Spina Bifida?

- Individuals with SB must adhere to a complex multi-component treatment regimen, while managing an array of cognitive and psychosocial comorbidities that hinder self-management and adherence
- Non-adherence to SB treatments can result in life-threatening health complications (pressure ulcers, UTIs)
- Up to ½ of hospitalizations are due to these preventable complications, with sepsis and renal failure being common causes of unexpected death

Spina Bifida

- Congenital birth defect



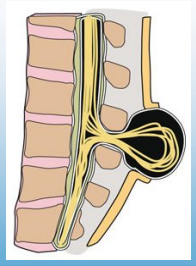
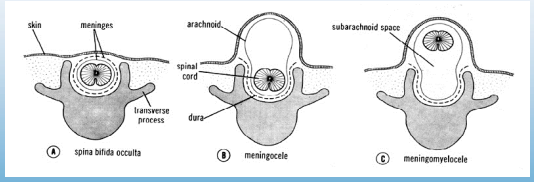


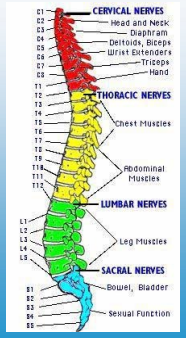
Illustration of spinal lesion

www.ich.ucl.ac.uk/.../F060452/index.html
and http://www.humanillnesses.com/Behavioral-Health-A-Br/Birth-Defects-and-Brain-Development.html

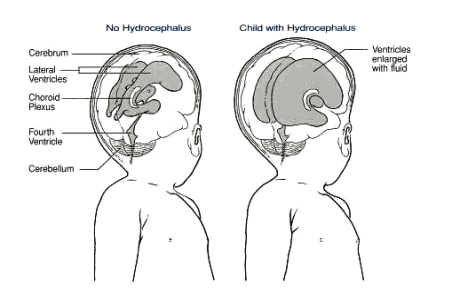
Spina Bifida - Subtypes



Spina Bifida - Sensory/Motor Defects



Spina Bifida - Hydrocephalus



Spina Bifida

Why Adolescence and Young Adulthood?

- Adolescence = transitional developmental period
- More biological, psychological, and social changes than any other developmental stage except infancy
- Two transitions: (1) Transition to early adolescence and (2) transition to early adulthood
- Health-behaviors are consolidated during adolescence
- The choices a child makes during adolescence have life-long effects
- An opportune time for prevention and intervention

Adolescent and Young Adult Development and Spina Bifida

Spina Bifida: Cognitive Development

- Low average range of intelligence
- Higher scores on verbal IQ than performance IQ (visual-motor integration)
- Academics are challenging (e.g., arithmetic)
- Abstract reasoning
- Executive functions
  - Planning and problem-solving ability
  - Goal-directed behavior
  - Ability to focus and shift attention

Adolescent and Young Adult Development and Spina Bifida (cont.)

Spina Bifida: Puberty and Social Development

- Onset of puberty may be early in adolescents with spina bifida
- Possible asynchronies between physical and social development
  - advanced puberty
  - delayed social development

Adolescent and Young Adult Development and Spina Bifida (cont.)

Spina Bifida: Autonomy Development

- Normative autonomy development is at odds with demands of medical adherence
- Parents of adolescents with chronic illnesses and physical disabilities are often faced with conflicting responsibilities:

(1) the responsibility to insure that the child remains healthy and adheres to treatment regimens

vs.

(2) the wish to facilitate independence in the child

Adolescent and Young Adult Development and Spina Bifida (cont.)

Spina Bifida: Other Developmental Issues during Adolescence

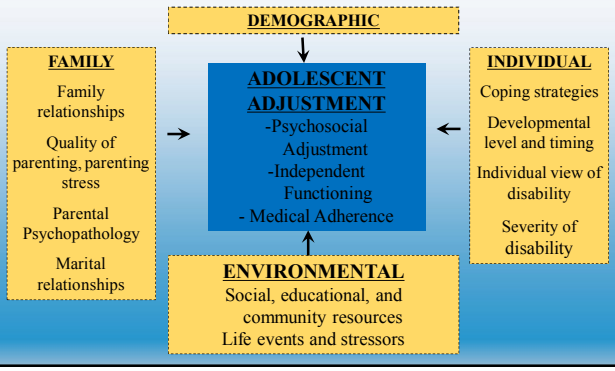
- Heightened sense of being different
- Development of same-sex and opposite-sex friendships
- Attainment of mature identity
- Development of sexuality in children with physical disabilities
- Planning for the future
  - Education, Vocation

CHATS-1: Longitudinal Study of Youth with Spina Bifida

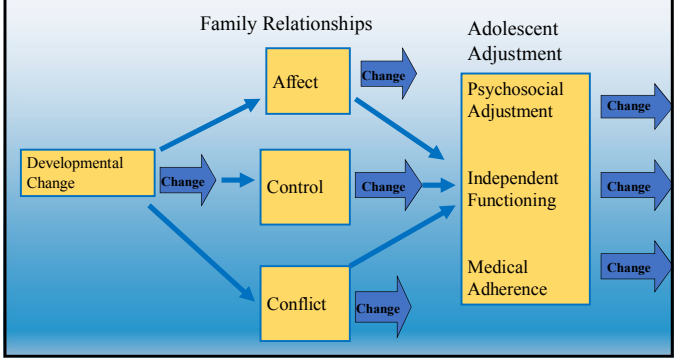
OUR INTEREST (early 1990s)

- Why do some children with spina bifida do well (with respect to medical outcomes and psychosocial adjustment) during the adolescent stage of development whereas other children are more challenged?

CHATS-1: Social-Ecological Model for Youth with Spina Bifida (1993)



CHATS-1: Longitudinal-Developmental Model (1993)



Strengths of CHATS-1

- Matched comparison sample of typically developing youth
- Home-based data collection with father included
- Longitudinal:
  - Families studied since 1993 (8-9 years old)
  - Completed Time 7 data collection
  - We began collecting 20-year follow-up data (Time 8) in November, 2013 when targets were 28-29 years old
- Information from multiple perspectives: mothers, fathers, children, teachers, health professionals, medical chart data, videotapes of family discussions

CHATS-1 (1993-present)

Participants at Time 1:

- Sample 1: 68 families with 8-9 year-old children with spina bifida (SB)
- Sample 2: 68 matched comparison families with 8-9 year-old typically-developing children (TD)
- Matched on 10 demographic variables
- All families studied in their homes
- In 80% of families, a father participated
- Time 7 included 54 families of children with SB (79%) and 61 comparison families (90%). Time 8 in process.

CHATS-1 (1993-present)

Spina Bifida sample (at Time 1):

- 32% sacral, 54% lumbosacral or lumbar, 13% thoracic
- 82% myelomeningocele (MM)
- 71% shunted
- 63% braces, 18% wheelchair, 19% no assistance
- average number of shunt surgeries = 2.50 (sd = 2.91)

CHATS-1 (1993-present)

Recruitment of spina bifida sample:

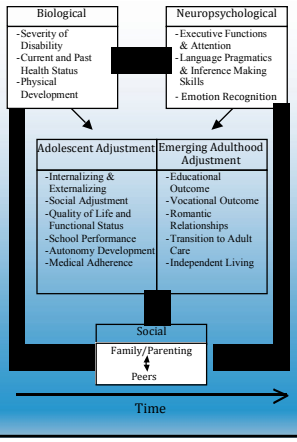
- 4 sources: 3 hospitals and state SB organization
- Recruitment letters to 310 children in 8-9 year old age range
- No differences between participants and those who declined on severity of SB

Recruitment of comparison sample:

- Recruited from a representative subset of schools where children with spina bifida attended

CHATS-2 (2005 to present)

- We learned from CHATS-1 that youth with SB have significant social and neuropsychological difficulties.
- We obtained funding from NICHD and began CHATS-2 to study these dimensions more rigorously (currently collecting T5 and T6 data with new funding from NINR)
- Within-sample study (*n*= 140)
- Includes observational data of children with SB talking with their parents and a session with a close friend (within ± 2 years of age)
- Over-sampling of Latino families
- Neuropsychological test battery



Bio-Neuropsychosocial Model of Psychological Adjustment in Children with Spina Bifida

Participants

- On-going longitudinal study of youth with spina bifida
- Inclusion criteria at Time 1:
  - Diagnosis of SB
  - 8-15 years old
  - Able to read and speak English or Spanish
  - Involvement of at least one primary caregiver
  - Live within 300 miles of Loyola University Chicago

Participants

- 246 families approached
- 163 agreed to participate
  - 21 could not be contacted or declined to participate
  - 2 were found to not meet inclusion criteria
- Final Sample
  - 140 families
  - Youth at Time 1
    - 54% female
    - M_{age} = 11.43 years
    - 52% Caucasian, 26% Latino/Latina, 12% African-American

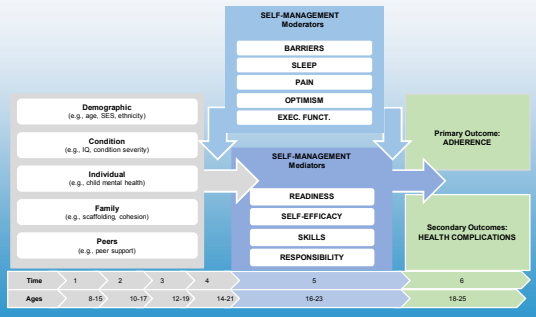
Procedure

- Data collected during 3-hour long home visits
  - Questionnaires (Youth, Parent, Teacher, Health Professional)
  - Neuropsychological test battery
  - Observational (separate sessions with family and close friend)
  - Medical chart reviews
  - Other methods: Daily Phone Diaries, actigraphy, structured interviews

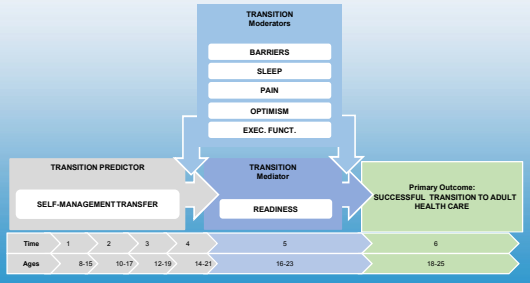
CHATS-2 (2005 to present)

- Sample was 8-15 years old at Time 1
  - Time 1: 8-15
  - Time 2: 10-17
  - Time 3: 12-19 (25% ≥18)
  - Time 4: 14-21 (50% ≥18)
  - Time 5: 16-23 (75% ≥18)
  - Time 6: 18-25 (100% ≥18; Emerging adulthood)
- As participants move into late adolescence/young adulthood (>18), the protocol changes to focus on the transition to adult health care.
- Given the predictable declines in health during young adulthood, at Times 5 and 6, we are interested in:
  - (1) early predictors of the transfer of medical responsibility from parent to child,
  - (2) associations between the success of this transfer and the transition to adult health care.

Longitudinal Conceptual Model of Medical Self-Management



Transfer of Self-Management Responsibility & The Transition to Adult Health Care



CHATS-1 and CHATS-2

Psychosocial Adjustment of Youth

CHATS-1 and CHATS-2

How well adjusted are the children with SB at ages 8-9 as compared to their TD peers?

Psychosocial Adjustment

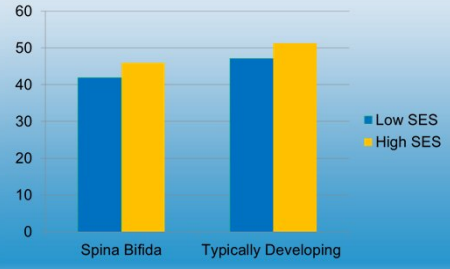
Holmbeck et al., 2003

- *Findings:* Children with SB tend to be:
  - socially immature and passive,
  - less likely to have social contacts outside school,
  - more dependent on adults,
  - less competent scholastically,
  - more likely to have attention and concentration difficulties
- *Additive risk factors* (with similar effect sizes):
  - Spina Bifida
  - SES

Psychosocial Adjustment

Holmbeck et al., 2003

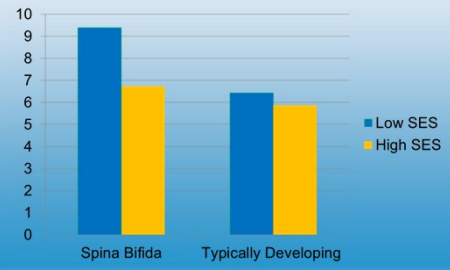
CBCL: Social Competence



Psychosocial Adjustment

Holmbeck et al., 2003

CDI: Depressive Symptoms



CHATS-1 and CHATS-2

Longitudinal Perspective: How do adjustment trajectories differ between youth with SB and their TD peers?

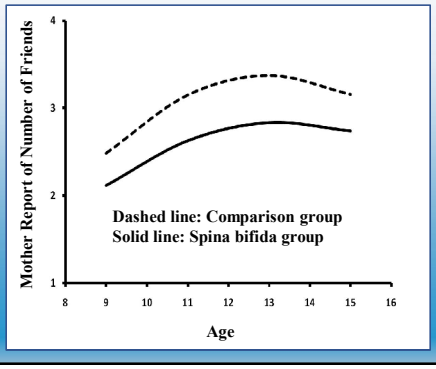
Psychosocial Adjustment

Holmbeck et al., 2010 (*JCCP*)

- *Objective:* 6-year, 4 wave, longitudinal follow-up of psychosocial adjustment in youth with SB
- *Method*: Growth curve modeling
  - 4 time-points (ages 8-15)
  - Predictor: Group status
  - Moderator: Gender
- *Findings:* Enduring difficulties for youth with SB:
  - Academic, attention, and social difficulties

Psychosocial Adjustment

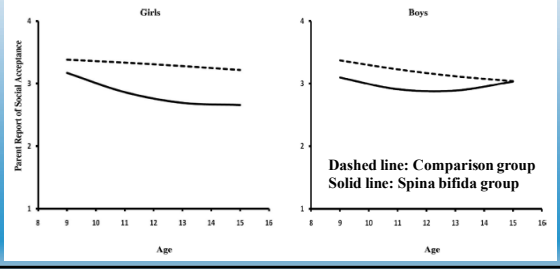
Holmbeck et al., 2010 (*JCCP*)



Psychosocial Adjustment

Holmbeck et al., 2010 (*JCCP*)

- Girls with SB at increased risk for:
  - social difficulties and negative perceived physical appearance



Psychosocial Adjustment

Holmbeck et al., 2010 (*JCCP*)

CHATS-1 and CHATS-2

Does a child's knowledge of spina bifida increase over time and, if so, in what domains?

Knowledge of SB

Greenley, Coakley, Holmbeck, Jandasek, & Wills, 2006

Table I. Group Means (*SD*) for Repeated Measures ANOVAs[a]

Question	Time 1	Time 2	Time 3	ANOVA
Etiology				
How old do you have to be to get SB?	.49 (.87)	.85 (.99)	1.05 (1.01)	T2>T1*, T3>T1**
Can people get SB as adults?	.72 (.97)	.59 (.92)	.82 (.99)	*ns*
How do kids get SB?	.00 (.00)	.00 (.00)	.13 (.50)	T3>T2*, T3>T1*
If someone with SB has a child, will the child have SB too?	1.11 (1.00)	1.05 (1.01)	1.44 (.90)	T3>T1*, T3>T2*
Functional differences				
What makes kids with SB different from other kids?	1.31 (.96)	1.02 (1.01)	1.18 (.99)	*ns*
How do kids with SB usually have to go to the bathroom?	1.08 (1.01)	1.41 (.92)	1.54 (.85)	T2>T1*, T3>T1**
What happens if they forget to catheterize?	.85 (1.00)	.95 (1.01)	1.08 (1.00)	*ns*
Why cannot kids with SB walk quite right?	.39 (.80)	.59 (.92)	.82 (.99)	T3>T1**
Shunt functioning				
Where is a shunt located?	1.76 (.66)	1.86 (.52)	1.90 (.43)	*ns*
What does a shunt do?	.48 (.86)	.43 (.83)	1.00 (1.01)	T3>T1*, T3>T2**
How does it feel if a shunt is not working right?	.71 (.97)	.71 (.97)	.90 (1.01)	*ns*
What do doctors do if a child's shunt is not working right?	1.52 (.86)	1.71 (.71)	1.71 (.71)	*ns*

ns, nonsignficant; SB, spina bifida.
[a]Scores ranged from 0 to 2. 0 = incorrect, 1 = inconclusive, 2 = correct.
*$p < .05$.
**$p < .01$.

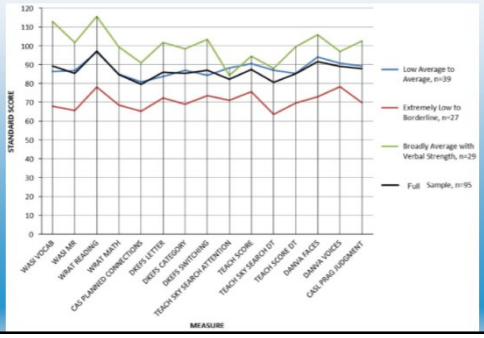
CHATS-1 and CHATS-2

Are there different subgroups of youth with SB based on neuropsychological functioning? How often and at what age are they diagnosed with ADHD?

Neuropsych Profiles

Wasserman & Holmbeck, 2016

- *Results:* Analyses indicated a 3-cluster solution



Neuropsychological Profiles

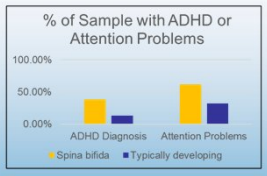
Wasserman & Holmbeck, 2016

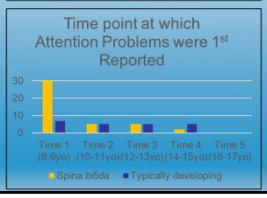
- *Results (continued)*
  - Neuro group=DV: SES, lesion level, and seizure history predicted group status
  - Neuro group=IV: Group membership predicted independence, academic success, expectations for the future, and child reported QOL

ADHD and SB

Wasserman, Stoner, Stern, & Holmbeck, 2016

- *Findings: Youth with spina bifida were…*
  - More likely to have ADHD or attention problems
  - More likely to be identified as having ADHD or attention problems at an earlier age
  - More likely to be treated with medication for attention problems/ ADHD





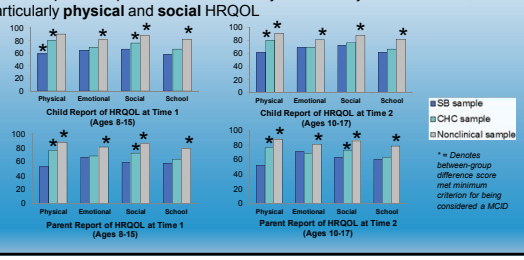
CHATS-1 and CHATS-2

How do the 2 samples (SB vs TD) differ across dimensions of quality of life?

Quality of Life

Murray, Holmbeck, Ros, Flores, Mir & Varni, 2014

- *Results: Group Differences*
  - Parent and child-reported HRQOL scores were clinically lower compared to a nonclinical sample, according to Minimal Clinically Important Difference scores (MCIDS).
  - Child and parent-reported HRQOL clinically lower than youth with CHCs, particularly **physical** and **social** HRQOL



CHATS-1 and CHATS-2

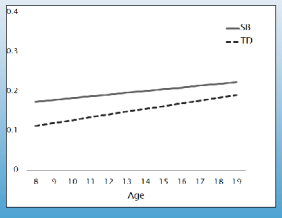
Is sleep a salient variable in this population? What is the developmental course of sleep in youth with SB? How do sleep-wake disturbances differ between youth with SB and TD youth?

Developmental Course of Sleep

Murray, Kirsch, Palermo, Holmbeck, Kolbuck, Psihogios, & Piggott (2016)

Results:

- Sleep disturbances increased over the 10-year period.
- Youth with SB had greater sleep disturbances during early adolescence.

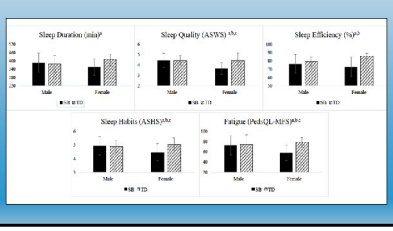


Case-controlled study of sleep in adolescents with spina bifida

Murray, Palermo, & Holmbeck, 2017

Results:

- Based on actigraphy data, adolescents with SB evidenced worse sleep quality, shorter sleep duration, greater sleep maintenance difficulties, and higher levels of fatigue compared with their TD peers.
- Exploratory analyses revealed females with SB were particularly vulnerable to developing sleep disturbances.



Note. Subscripts denote significant differences ($p < .05$) between [a]SB females & TD females, [b]SB females & SB males, and/or [c]SB females and TD males according to LSD post-hoc comparison tests

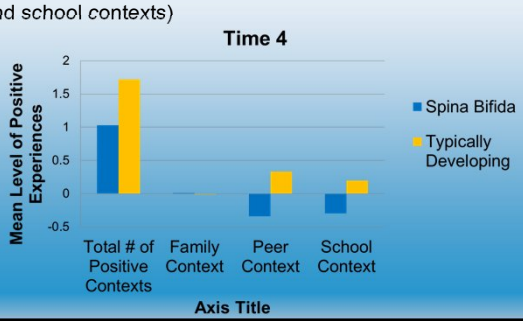
CHATS-1 and CHATS-2

Moving on to social adjustment…Do youth with spina bifida have fewer positive social contexts (i.e., family, peer, school) than their typically developing peers?

Social Contexts and Depressive Symptoms

Essner & Holmbeck, 2010

- Youth with SB have fewer positive contexts (particularly peer and school contexts)



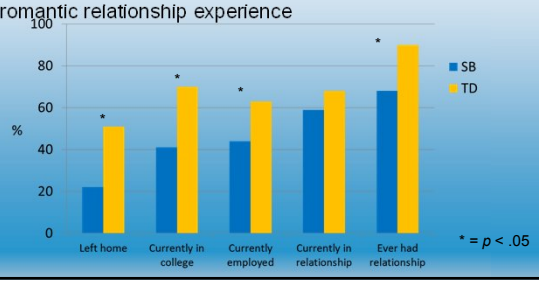
CHATS-1 and CHATS-2

Do the 2 samples differ on achievement of milestones in early adulthood (ages 18-19)? What about at 22-23? What are the longitudinal adolescent predictors (ages 14-15) of these milestones?

Emerging Adulthood

Zukerman, Devine, & Holmbeck, 2010

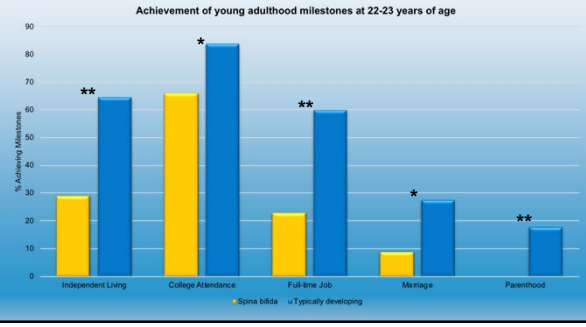
- *Findings at ages 18-19*
  - Emerging adults with SB were less likely to leave home, attend college, maintain employment, and have ever had a romantic relationship experience



Young Adulthood Milestones at ages 22-23

Holbein, Zebracki, Bechtel, Papadakis, Bruno, & Holmbeck, 2017

- *Findings/Implications*



Emerging Adulthood

Zukerman, Devine, & Holmbeck, 2010

- *Findings*
  - Factors associated with achievement of emerging adult milestones included:
    - Higher SES
    - Higher levels of executive functions
    - Higher intrinsic motivation
    - Lower maternal intrusiveness

CHATS-1 and CHATS-2

Do the 2 samples differ in their rates of normative and risky health behaviors during emerging adulthood and what is the influence of prior and current social adjustment on these health risk behaviors?

Health Behaviors

Murray, Lennon, Devine, et al. (2014)

- *Results*
  - Individuals with SB reported:
    - (1) lower initiation rates (i.e., ever used) and frequency of alcohol use, and
    - (2) less sexual activity (i.e., ever had sex) and fewer sexual partners compared to their TD peers.
  - Interestingly, better social adjustment during early adolescence (ages 12-13) predicted more frequent alcohol use and a greater number of sexual partners for all youth.
  - Thus…lack of social engagement is protective with respect to some risk behaviors

CHATS-1 and CHATS-2

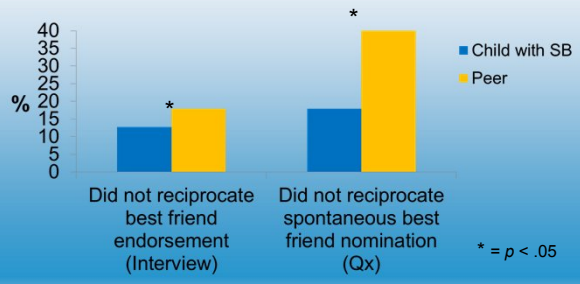
Does social functioning differ between youth with spina bifida and their selected close friends?

Are close friends less likely to reciprocate "best friend" nominations?

Friendships

Devine, Holmbeck, Gayes, & Purnell, 2011

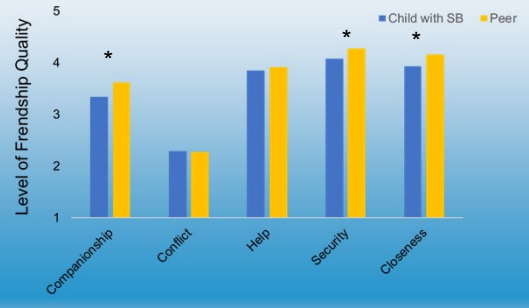
Reciprocity of Friendship



Friendships

Devine, Holmbeck, Gayes, & Purnell, 2011

Friendship Qualities



CHATS-1 and CHATS-2

Can we create reliable and valid observational scales for videotaped peer interactions?

Peer Interaction Macro Scales

Holbein, Zebracki, & Holmbeck (2014)

- *Findings*
  - Content validity: comprehensive literature review revealed 5 key constructs: Control, Prosocial Skills, Positive Affect, Conflict, Dyadic Cohesion
    - Panel of experts classified codes into these constructs
  - Internal consistency and inter-rater reliability: good-to-excellent (scale level)
  - Interscale correlations: low-to-moderate for four scales
    - Dyadic Cohesion scale dropped
  - Convergent and discriminant validity with questionnaire and interview items: encouraging evidence for the 4 scales

Peer Interaction Macro Scales

Holbein, Zebracki, & Holmbeck (2014)

Final Scale Composition

- Control
  - Dominance
  - Pressures others to agree
- Prosocial Skills
  - Confidence in stating opinions
  - Eye contact
  - Listens to others
  - Maturity
  - Promotes dialogue and collaboration
  - Receptive to statements
- Positive Affect
  - Anger (r)
  - Humor & laughter
  - Intensity/freq. of neg. affect (r)
  - Intensity/freq. of pos. affect
- Conflict
  - Able to reach agreement/resolution (r)
  - Attempted resolution of issues (r)
  - Level of conflict
  - Neg. escalation
  - Tolerates disagreements (r)

Macro Social Differences

Holbein, Lennon, Kolbuck, Zebracki, Roache, & Holmbeck, 2015

- *Findings/Implications*
  - Similar levels of basic social skills (e.g., affect, eye contact, listening skills)
  - Children with SB exhibited lower levels of the following social engagement skills:
    - Task involvement, clarity of thought, confidence in stating opinions, explanations for opinions, off-task behavior, maturity, dominance, and promotion of dialogue/collaboration

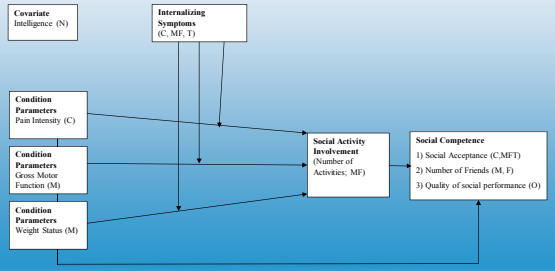
CHATS-1 and CHATS-2

Do we find support for proposed models of social functioning in youth with spina bifida?

Condition Parameters and Social Outcomes

Essner, Murray & Holmbeck (2014)

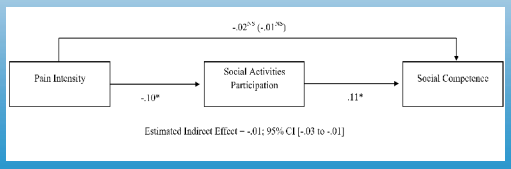
Study Model: Associations among Condition Severity, Social Activity Involvement, and Social Competence in Youth with SB



Condition Parameters and Social Outcomes Essner, Murray & Holmbeck (2014)

Study Model Findings

•Analyses confirmed that increased pain intensity predicted decreased involvement in social activities, which in turn predicted decreased social competence in youth with spina bifida (95% CI Lower to Upper = -.03 to -.01)

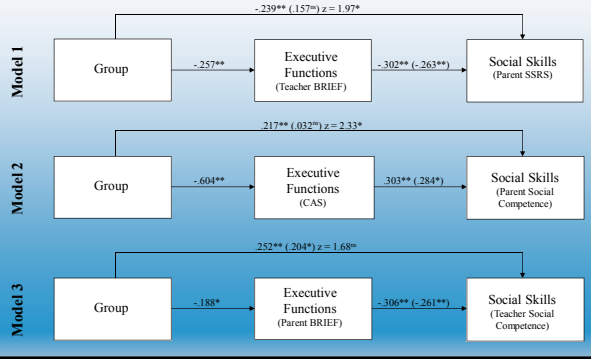


CHATS-1 and CHATS-2

Are measures of neuropsychological functioning (e.g., executive functioning) related to social adjustment longitudinally?

Executive Function

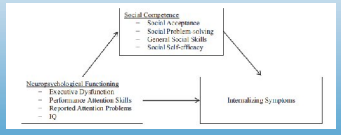
Rose & Holmbeck, 2007



Neuropsychological Functioning, Social Competence & Internalizing Symptoms

Lennon, Klages, Amaro, Murray, & Holmbeck (2015)

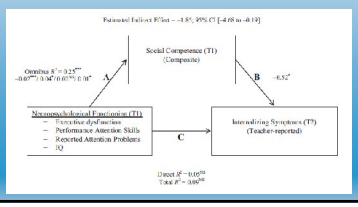
- *Objective*
  - To examine the longitudinal relationship between neuropsychological functioning and internalizing symptoms, as mediated by social competence in youth with SB.



Neuropsychological Functioning, Social Competence & Internalizing Symptoms

Lennon, Klages, Amaro, Murray, & Holmbeck (2015)

- *Results*
  - An indirect-only mediation model revealed that better neuropsychological functioning was associated with better social competence, which, in turn, predicted fewer internalizing symptoms (as reported by teachers) 2 years later.
  - Results did not vary based on age.



Social Skills Predictors

Holbein, Peugh, & Holmbeck, 2017

- *Findings*
  - Statistics: hierarchical linear regressions in Mplus

Dependent variable	Full model (all domains)	Neuropsychological base model (Family + Health)				Family base model (Neuro. + Health)				Health base model (Neuro. + Family)			
	R^2	R^2	$R^2\Delta$	F	f^2	R^2	$R^2\Delta$	F	f^2	R^2	$R^2\Delta$	F	f^2
Observed social skills	0.58	0.49	0.09	0.94	0.22	0.55	0.03	0.32	0.08	0.54	0.04	0.40	0.10
SSRS—Mother	0.70	0.51	0.19	2.75*	0.65	0.67	0.03	0.45	0.11	0.66	0.05	0.61	0.16
SSRS—Father	0.75	0.53	0.22	3.58*	0.84	0.69	0.05	0.82	0.20	0.60	0.15	2.27	0.57
SSRS—Teacher	0.54	0.39	0.15	1.39	0.33	0.49	0.05	0.44	0.11	0.52	0.02	0.16	0.04

*p<.01

  - Neuropsychological variables accounted for significant variance in parent-reported T2 social skills *beyond* family and health factors

CHATS-1 and CHATS-2

Family Relationships

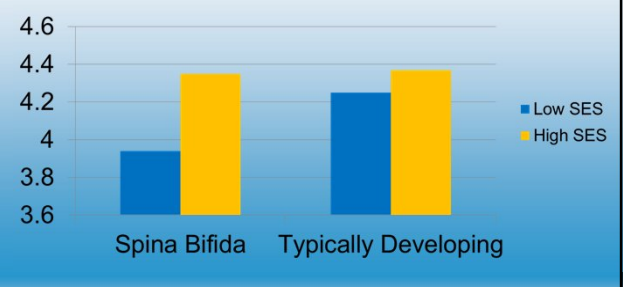
CHATS-1 and CHATS-2

Do the 2 samples differ on measures of family functioning?

Family Functioning

Holmbeck, Coakley, Hommeyer, Shapera & Westhoven, 2002

Observational Data of Family Cohesion



Family Functioning

Holmbeck, Coakley, Hommeyer, Shapera & Westhoven, 2002

- *Implications*
  - Across this set of analyses, findings support the *resilience-disruption hypothesis* for families of children with SB
    - SB produces some disruption (i.e. low family cohesion) but there is also resilience in some areas (i.e. comparable rates of family conflict and life stress)
  - As was found with adjustment, low SES families of children with SB may represent an at-risk group

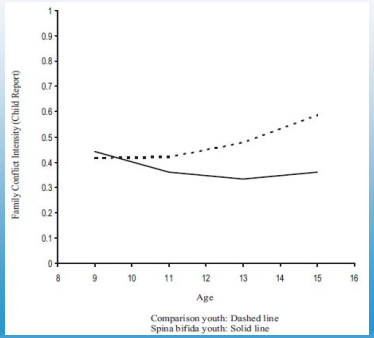
CHATS-1 and CHATS-2

Longitudinal Perspective: Do the 2 samples differ over time on measures of family functioning?

Family Processes

Jandasek, Holmbeck, DeLucia, Zebracki, & Friedman, 2009

- For youth with SB: changes in family conflict/ cohesion may be less dramatic



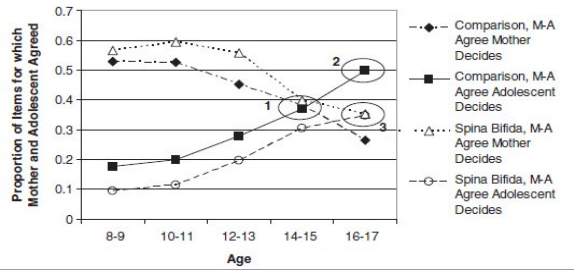
CHATS-1 and CHATS-2

Longitudinal perspective: Are there differences between the 2 samples when comparing changes over time in mother-youth agreement on who makes decisions in the family?

Decision-making Autonomy

Devine, Wasserman, Gershenson, Holmbeck, & Essner, 2011

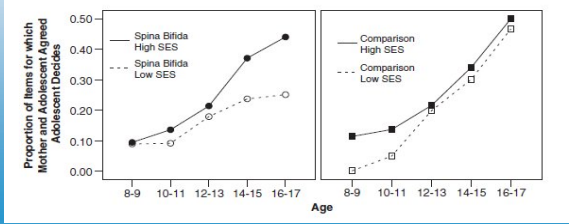
- For youth with SB: decision-making autonomy lags by about 2 years



Decision-making Autonomy

Devine, Wasserman, Gershenson, Holmbeck, & Essner, 2011

- Decision-making autonomy lower for youth with SB from lower SES backgrounds



CHATS-1 and CHATS-2

How does family functioning differ in families of Latino youth vs families of non-Latino youth?

Psychosocial & Family Functioning Among Latino Youth

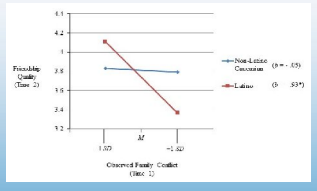
Papadakis, Acevedo, Ramirez, Stern, Driscoll, & Holmbeck (2017)

- *Results*
  - Latino youth had ***Less*** externalizing symptoms and ***Less*** family conflict, but also ***Less*** social competence
  - ***NO DIFFERENCES*** in internalizing symptoms, peer acceptance, friendship quality, family cohesion, family stress
  - RESILIENCE: Despite the greater number of challenges/stressors that are believed to be more prevalent for Latino youth, Latino youth with SB tend to fare similarly to (and sometimes better than) non-Latino Caucasian counterparts

Psychosocial & Family Functioning Among Latino Youth

Papadakis, Acevedo, Ramirez, Stern, Driscoll, & Holmbeck (2017)

- *Results (continued)*
  - Latino youth:
    - Greater family conflict predicted ***decreased*** friendship quality (supports salience of family relations for this group)



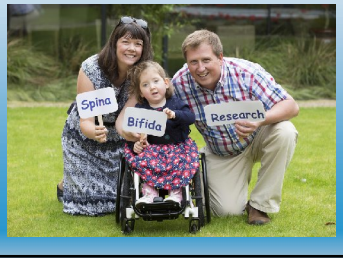
  - Non-Latino Caucasian youth:
    - Greater family cohesion predicted ***increased*** peer acceptance
    - Greater family stress predicted ***increased*** internalizing symptoms

CHATS-1 and CHATS-2

Parents and Parenting

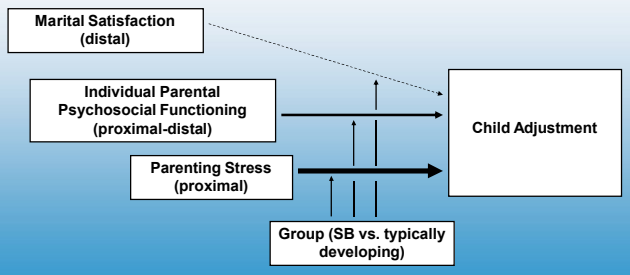
CHATS-1 and CHATS-2

Is the parent's own level of adjustment related to adjustment outcomes in youth?



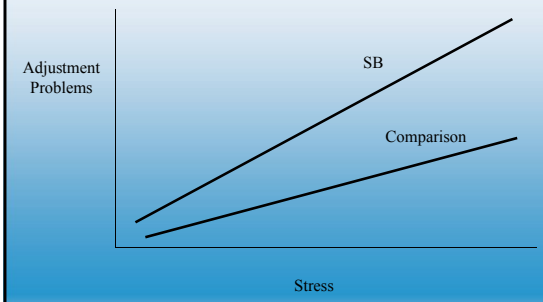
Parent Functioning

Friedman, Holmbeck, Jandasek, Zukerman, & Abad, 2004

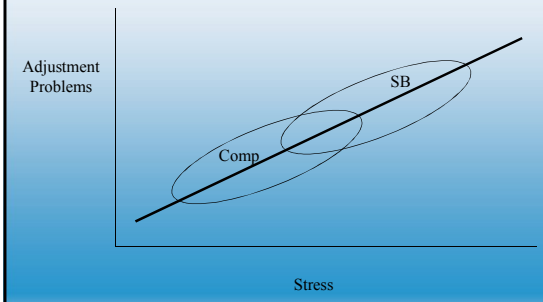


Strength-of-Association Moderator Model
Heavier lines indicate stronger predicted associations.

Model #1: Strength-of-association model



Model #2: Higher stress model



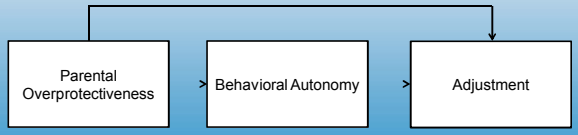
CHATS-1 and CHATS-2

Is parental overprotectiveness (i.e., intrusiveness) more common in families of youth with SB and is this type of parenting related to adjustment outcomes?

Parental Overprotection

Holmbeck, Johnson, Wills, McKernon, Rolewick, & Skubic, 2002

Mediational model of parental overprotectiveness, behavioral autonomy, and psychosocial adjustment



CHATS-1 and CHATS-2

Are there differences between the two samples in emotions expressed by parents (warmth, criticism, optimism) and are these emotions related to youth adjustment outcomes?

Expressed Emotion

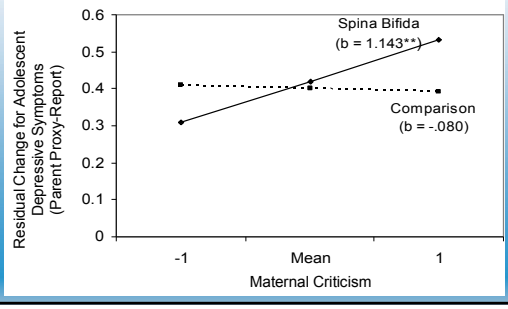
Kelly, Holmbeck, & O'Mahar, 2011 (*JPP*)

- *Objectives*
  - Examine validity for new measure of parental expressed emotion (EE) via audiotaped interview with mothers and fathers
  - Examine relation between parental EE (↑criticism, ↓warmth) and adolescent depressive symptoms

Expressed Emotion

Kelly, Holmbeck, & O'Mahar, 2011

Maternal criticism (youth=16/17) x group membership interaction for predicting residual change in depressive symptoms from 16/17 to 18/19 after controlling for child receptive vocabulary. (b=unstandardized regression coefficient; **p < .01)



CHATS-1 and CHATS-2

Do parents of youth with SB perceive their children to be "vulnerable"? Do these perceptions increase over time? What predicts these perceptions?

Predictors of Parental Perceptions of Child Vulnerability

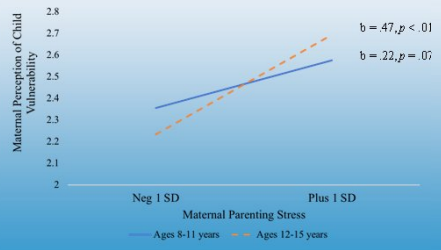
Driscoll, Stern, Ohanian, Fernandes, Crowe, Ahmed, & Holmbeck, 2017

Comparing Time 1 & Time 2 Reports

*p < .001
^ p = .05

Predictors of Parental Perceptions of Child Vulnerability

Driscoll, Stern, Ohanian, Fernandes, Crowe, Ahmed, & Holmbeck, 2017



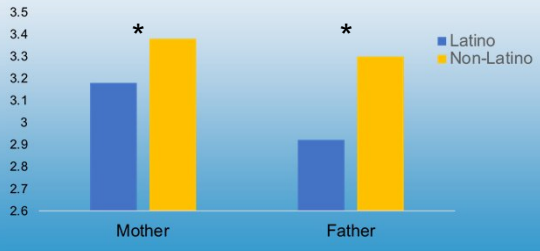
CHATS-1 and CHATS-2

Are there differences in parental functioning between Latino and non-Latino families of youth with SB?

Latino Parents

Devine, Holbein, Psihogios, Amaro, & Holmbeck (2012)

Parenting Satisfaction

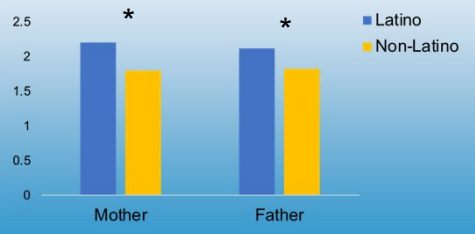


$p < .05$

Latino Parents

Devine, Holbein, Psihogios, Amaro, & Holmbeck (2012)

Perceived Child Vulnerability

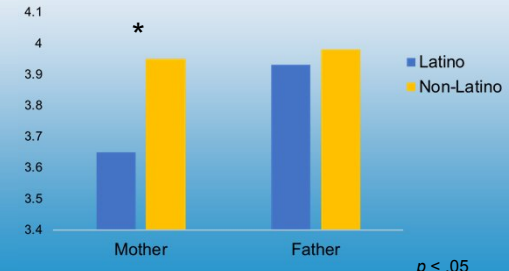


$p < .05$

Latino Parents

Devine, Holbein, Psihogios, Amaro, & Holmbeck (2012)

Parental Competence



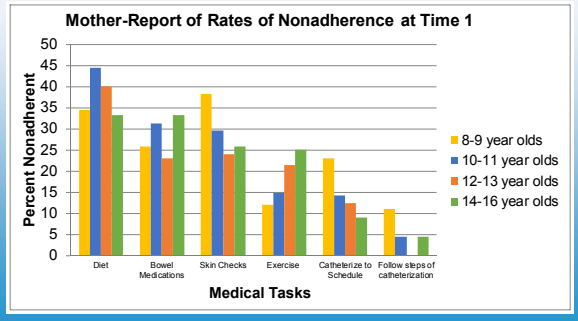
$p < .05$

CHATS-1 and CHATS-2

Self-Management and Medical Adherence

Condition Self-Management

Psihogios, Kolbuck, & Holmbeck, 2015

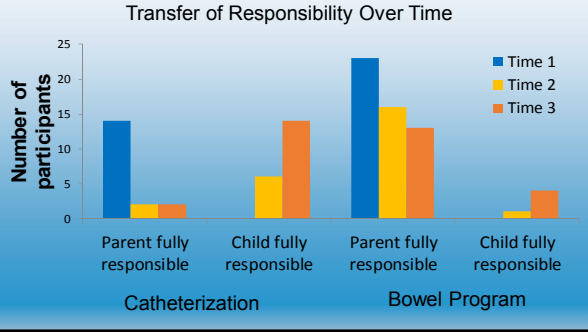


CHATS-1 and CHATS-2

At what age do parents transfer medical regimen responsibilities to youth?

Adherence

Stepansky, Roache, Holmbeck, & Schultz, 2010



CHATS-2 (2005-present)

Are mother-child disagreements over who is responsible for SB medical tasks associated with family conflict and medical adherence?

Medical Responsibilities

Psihogios & Holmbeck, 2013

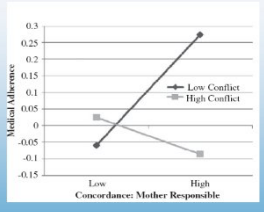
Table I. *Discrepancies Based on Child and Mother Reports of Who Is Responsible for Spina Bifida Medical Tasks*

		Parent report		
		Child responsibility	Equal responsibility	Parent responsibility
Child report	Child responsibility	1 Full concordance: child responsible *16.74% of responses*	2	3 Full discrepancy, "both report being responsible" *2.28% of responses*
	Equal responsibility	4	5 Full concordance: shared responsibility *12.01% of responses*	6
	Parent responsibility	7 Full discrepancy, "both report that the other is in charge" *7.28% of responses*	8	9 Full concordance: parent responsible *28.15% of responses*

Note. Only categories of full discrepancy or full concordance were used (i.e., cells 1, 3, 5, 7, and 9). The remaining four categories (i.e., cells 2, 4, 6, and 8) were not analyzed, as these categories represent only partial discrepancies.

Medical Responsibilities

Psihogios & Holmbeck, 2013



- Adherence is better when conflict is low and parents, rather than children, are responsible for managing the medical care of the target child.

Medical Adherence & Responsibility

Psihogios, Murray, Zebracki, Acevedo, & Holmbeck, 2017

Allocation of Responsibility and Adherence with Catheterization and Bowel Program

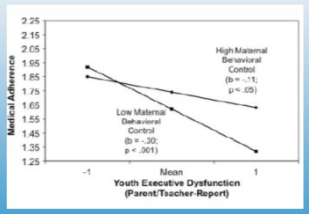
	Child Responsible	*Child Not Responsible*
Adherent	Group 1: *Adherent, Child Responsible*	Group 2: *Adherent, Child Not Responsible*
Non-adherent	Group 3: *Nonadherent, Child Responsible*	Group 4: *Nonadherent, Child Not Responsible*

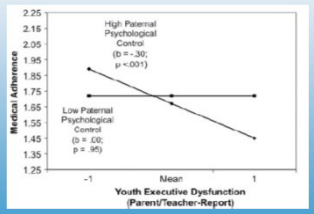
CHATS-1 and CHATS-2

- Are executive functions and parenting behaviors (acceptance, behavioral control, and psychological control) associated with medical adherence and autonomy?

Medical Adherence

O'Hara & Holmbeck, 2013





Impact of Depressive Symptoms and Neuropsychological Functioning on Medical Responsibility

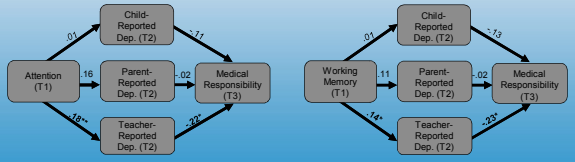
Stern, Driscoll, Ohanian, & Holmbeck (2018)

- *Objective*
  - To examine two longitudinal pathways through which depressive symptoms, attention, and executive functioning may be associated with medical responsibility in youth with SB.

Impact of Depressive Symptoms and Neuropsychological Functioning on Medical Responsibility

Stern, Driscoll, Ohanian, & Holmbeck (2018)

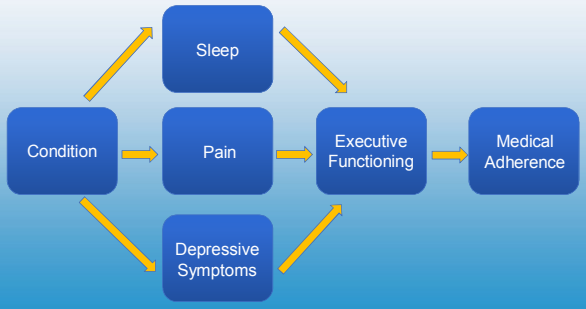
- *Results*
  - Results indicated no significant indirect effects of depressive symptoms on medical responsibility via cognitive functioning (all *p*'s > .05).
  - Teacher-reported depressive symptoms at Time 2 significantly mediated the relationships between attention and working memory at Time 1 and medical responsibility at Time 3 (all *p*'s < .05) in the expected directions.



Direct Effect[a]: *b*=-.09, SE=.03, *t*=-2.57, *p*=.01
Indirect Effect[b]$_{Teacher}$=-.04, LLCI to ULCI=-.09 to -.01

Indirect Effect[a]$_{Teacher}$=-.03, LLCI to ULCI=-.09 to -.01

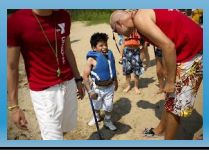
Where to Intervene?: Variables Upstream from Outcomes of Interest

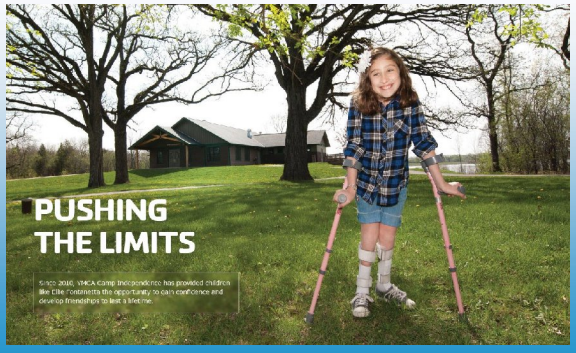


Camp Independence

Camp Study: 2006- present

Based on findings of CHATS-1 and CHATS-2, we designed a camp-based intervention for youth and young adults with SB





Challenges to Independence

- Spina bifida produces associated difficulties across several domains

Physical	Cognitive	Psychosocial
• Ambulation (may require a wheelchair or braces)	• Attention Problems	• Higher Rates of Depression and Anxiety
• Skin Health	• Executive Dysfunction	• Increased Dependence on Caregivers
•Orthopedic (for example, club foot or scoliosis)	• Learning/Memory Difficulties	• Decreased Peer Involvement
•Urinary & Bowel Function	• Visual-Motor Deficits	
	• Social Language Deficits	

Camp Intervention

O' Mahar, Holmbeck, Jandasek, & Zukerman, 2009 (*JPP*)

- *Objective*
  - Design and evaluate a camp-based intervention targeting independence among children, adolescents and adults with spina bifida; 3 manuals by age grouping
- *Intervention and Method*
  - Intervention embedded within a week long camp experience (Camp Independence, northern IL)
  - Three components:
    1. Parent and child goal-setting (SB-related and social)
    2. Goal monitoring by trained counselors
    3. Group sessions (i.e., psychoeducational, cognitive tools)
  - Participants assessed at three time points: pre- & post intervention, 1 month follow-up

Camp Independence

- Camp runs for 1 week, Sunday-Friday, for 8 weeks during the summer
- Different weeks for different age groups
  - 7-13 year olds
  - 14-19 year olds
  - Young adults (20+)
- Up to 16 campers attend each week
- Each camper is assigned a counselor

The Independence Program of Camp

GOAL = *help campers achieve developmentally and cognitively-appropriate independence*

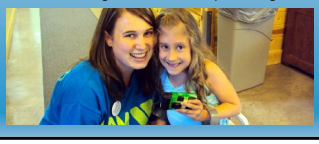
- Campers attend a workshop each day of camp (Monday-Thursday)
- Workshop lasts 1 hour, and is led by trained interventionist(s)
- Workshop promotes all aspects of independence
  - It highlights *typical and spina bifida related* independence
  - It is tailored to each age group

Workshop Day 1: Taking Care of Relationships

GOAL: Learn about creating and maintaining relationships

- Social Skills
  - ✓ Good listening skills
  - ✓ Making small talk
  - ✓ Asking a friend to do an activity
  - ✓ Asking for help
- Social relationships
  - ✓ Making and maintaining friendships
  - ✓ Getting along with siblings

Involves talking about skills and practicing skills!



Workshop Day 2: Taking Care of Yourself

GOAL: Learn about mental health and emotional wellness

- ✓ Developing positive self-esteem
- ✓ Recognizing how others feel
- ✓ Positive coping strategies
- ✓ Relaxation techniques

Workshop Day 3: Living with Spina Bifida

GOAL: Understand and reflect upon how spina bifida impacts daily life, and how to gain control

- ✓ Discussing feelings about spina bifida
- ✓ Responding to others and their reactions to spina bifida
- ✓ Handling embarrassing situations related to spina bifida
- ✓ Explaining spina bifida to a friend

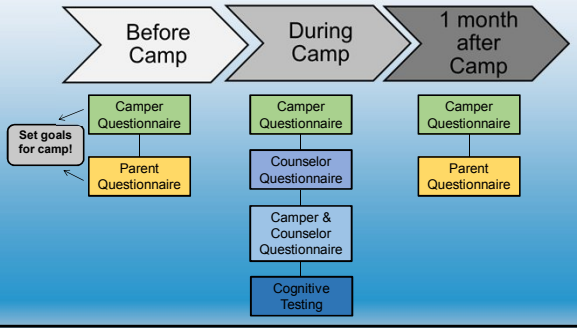
Workshop Day 4: Living with Spina Bifida/Preparing for the Future

GOALS: (1) understand features/ management of spina bifida (2) start thinking about the future

- ✓ Learning about spina bifida (Jeopardy! game)
- ✓ *Why* it is important to be responsible for spina bifida healthcare
- ✓ *How* to take responsibility for spina bifida healthcare
- ✓ Continuing to pursue goals after camp
- ✓ Becoming active in the community
- ✓ For older campers → applying for jobs (resume, interviews)

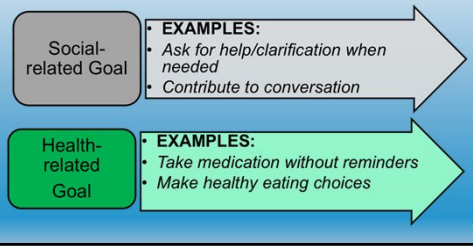
Independence Program: Evaluation

Data are collected at 3 time points



Independence Program: Evaluation

- Before camp starts, campers and their parents pick 2 goals
  - one social-related, one health-related
- **Counselors help campers work on their goals throughout camp**



Independence Program: Evaluation

Our research suggests that camp leads to significant improvements in:

* Individual social and health goals *

* Management of spina bifida-related responsibilities *

(Holbein, Murray, Psihogios, Wasserman, Essner, O'Hara, & Holmbeck, 2013; O'Mahar, Holmbeck, Jandasek, & Zukerman, 2010)

Camp Intervention

O' Mahar, Holmbeck, Jandasek, & Zukerman, 2009

- *Findings*
  - Significant improvement in:
    - Individually oriented spina bifida and social goals
    - Management of spina bifida responsibilities
    - Independence with general spina bifida tasks
  - Improvement in individual spina bifida and social goals maintained at 1-month follow-up (Figure 1)

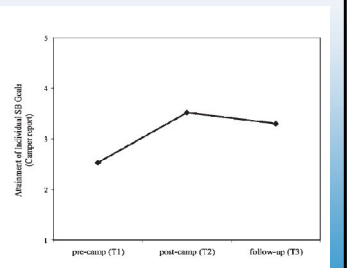


Figure 1. Mean level of goal attainment for individual spina bifida goals at times 1, 2, and 3

Take-Home Messages and Clinical Implications

Take home messages: What have we learned so far?

- It is important to study youth with SB during adolescence and young adulthood
- Youth with SB and their parents are confronted with a number of *psychosocial*, *neuropsychological, and familial* challenges that are just as important as their *medical and physical* challenges
- These challenges are related to their difficulties in managing the milestones of adolescence and emerging adulthood
- The challenges for youth and their families are particularly acute for Latino families and those in lower SES homes

Take home messages: What have we learned so far?

- We need to attend carefully to social difficulties in this population and study this more rigorously
- We continue to need research and interventions that focus on:
  - (1) The transfer of medical responsibility from parent to child
  - (2) The transition from pediatric to adult health care

Clinical Implications for Families of Youth with SB

Improve Psychosocial Adjustment

- Maximize time child has with friends,
- Address issues of privacy, teasing etc. with school
- Look for warning signs of depression
- Educate child about his/her condition
- Encourage child to express opinions
- Support intrinsic motivation (vs. dependency)
- Neuropsychological evaluation and interventions
- Meetings with school administrators and teachers
- Medication evaluation for attention problems
- Target low SES and Latino families
- Focus on "sharing" of adherence tasks during the adolescent transition
- Focus early on the transition to adult health care

THANK YOU!
Questions?

William R. Duncan, MD

June 16, 1912 – April 13, 2003

The Duncan Seminar was conceived and initiated in 1977 by Lynn T. Staheli, Director of Orthopedics, Children's Hospital and Regional Medical Center, as a living tribute to William R. Duncan, MD, Chief of Orthopedics at Children's Orthopedic Hospital from 1955 to 1961.

Dr. Duncan was born and raised in Seattle to the pioneer "Duncan and Sons" family. He graduated from Franklin High School and attended the University of Washington. After graduating from McGill University Medical School in Montreal in 1938, Dr. Duncan completed training in orthopedic surgery at the Hospital for Special Surgery and Presbyterian Medical Center in New York City. He served in the U.S. Army Medical Corps from 1942 to 1945.

Dr. Duncan returned to Seattle to practice and became nationally recognized as the founder of many programs in the Northwest providing care for children with cerebral palsy. He helped establish Boyer Children's Clinic and Preschool in Seattle, the Cerebral Palsy Clinic at Children's Hospital and Regional Medical Center, and programs for the disabled within the Seattle school system. He acted as a consultant to Lowell School and the Washington State Cerebral Palsy Center. As past consultant for the Washington State Department of Health and Crippled Children's Services (now Children with Special Health Care Needs), he instituted statewide diagnostic clinics.

Dr. Duncan was a Diplomate of the American Board of Orthopedic Surgeons and a member of the American Academy for Cerebral Palsy, Seattle Surgical Society, North Pacific Orthopedic Association, Western Orthopedic Association, American Academy of Orthopedic Surgeons, American Orthopedic Association and the Orthopedic Research Society. He founded the Orthopedic Research Foundation of Seattle. Dr. Duncan has a long list of publications on cerebral palsy.

Dr. Duncan emphasized a comprehensive approach to the needs of children with cerebral palsy and other disabilities. In keeping with his perspective, the Duncan Seminar Planning Committee has sought to bring fresh ideas into focus for an interdisciplinary audience, while always highlighting the "whole child."

The Duncan Seminar has thrived in part because of the support of Dr. Duncan's family and the following organizations: Washington State Department of Health; Maternal and Child Health Bureau (HRSA, DHHS); United Cerebral Palsy; Department of Rehabilitation Medicine, Division of Developmental Medicine, and Department of Outreach Education, Seattle Children's; and the Clinical Training Unit, Center on Human Development and Disability, University of Washington.

The Duncan Award

The Duncan Award was initiated as a companion to the Duncan Seminar to acknowledge those parents, professionals, and groups who have made significant contributions to the well-being of children with disabilities within the state of Washington. The Duncan Award is given each year to an individual or group that has:

1) Demonstrated Dr. Duncan's ability to keep the whole child in view.

2) Made a long-term commitment to children with disabilities.

3) Had an extraordinary positive social or scientific impact on the well-being of children with disabilities above and beyond usual career expectations.

The winner is selected by the interdisciplinary Duncan Award Committee. Past seminar topics and award recipients are:

Year	Award Recipient	Seminar Title
1977	**William R. Duncan, MD**, Orthopedist	First Annual Duncan Seminar on Cerebral Palsy
1978	**Rose Rhinehart**, Parent & Past UCP Board President	Families and Cerebral Palsy
1979	**Peggy Pomeroy, RN**, Nurse Clinician	Cerebral Palsy
1981	**Helen Russell, RPT,** Physical Therapist	Getting Along, Getting Around and Getting Through: Comprehe Care for the Physically Disabled Child
1982	**Ervin J. Larsen**, Past Executive Director, UCP	Transition into Independence: The Physically Disabled Adolescent
1983	**Pam Mullens, RPT,** Physical Therapist	Three Score and Ten: Lifetimes and Disabilities
1984	**Park W. Gloyd, MD**, Orthopedist	Problems That Won't Go Away
1985	**John E. Dunn, MD**, Orthopedist	National Scientific Meeting, AACPDM
1986	**Maplewood School Staff,** Edmonds School District	Feeding, Breathing and Communication in the Disabled Infant: An Interdisciplinary Challenge
1987	**Nora E. A. Davis, MD,** Pediatrician	Ambulation/Mobility: Improving Our Decision Making
1988	**Mary Shandorf**, Foster Parent	Cracks in the Yellow Brick Road
1989	**Lynn T. Staheli, MD**, Pediatric Orthopedist	Clinical Applications of Gait Analysis
1990	**David B. Shurtleff, MD,** Pediatrician	Spasticity: Measurement and Management
1992	**John F. McLaughlin, MD**, Pediatrician	Myelodysplasia, Part I
1993	**Elizabeth Ingman, RN,** Nurse Clinician	Myelodysplasia, Part II
1994	**Patricia Trulson, PHN** & **Isaac Pope, MD**	Arthrogryposis and Osteogenesis Imperfecta
1996	**Gay-Lloyd Pinder, PhD, CCC-S/LP,** Speech Pathology	Adaptive Technology: Choices, Priorities and Opportunities
1997	**Stella Lamoreaux, RN,** Public Health Nurse	New Perspectives in Pediatric Feeding and Swallowing
1998	**Children's Village, Yakima** **Special Recognition: John M. Neff, MD,** Pediatrician	Cerebral Palsy: Current Concepts
1999	**Jeanne L. Fischer, RPT,** Physical Therapist	Teens with Disabilities: Successful Transition to Adulthood
2000	**Betty L. Lucas, MPH, RD, CD,** Nutritionist	The Picture of Health for Children with Disabilities
2001	**Maxine Siegel, MA,** Exec. Director, Kindering Center **Judy Moore, MA,** Exec. Director, Boyer Children's Clinic	How Infants Learn: New Concepts and Implications for Children with Atypical Development
2002	**Robin G. Glass, MS, OTR,** Occupational Therapist **Lynn S. Wolf, MOT, OTR,** Occupational Therapist	What's New in Cerebral Palsy
2003	**Kiko Kimura Van Zandt, RN, BSN, CRRN,** Nurse Clinician	Spina Bifida: Issues Across the Life Span
2004	**Donald C. Gargas, MD,** Pediatrician	Behavioral Dilemmas in Kids with Developmental Disabilities

Year	Award Recipient	Seminar Title
2005	**Greg and Kathy Hull,** Foster Parents	Raising Special Needs Kids: Health, Growth, and Nutrition
2006	**Charles A. Cowan, MD,** Developmental Pediatrician	Sleeping Better in Seattle: Helping Children with Special Needs
2007	**Donald J. Meyer, MEd,** Director, Sibling Support Project	Pain, Pain, Go Away: Helping Children with Special Needs Manage Their Pain
2008	**James E. May, MA, MEd, LMHC,** Program Director (Ret.), Washington State Father's Network	Cerebral Palsy Today and Tomorrow: Updates in Evaluation and Treatment
2009	**Cristine M. Trahms, MS, RD, CD, FADA** Head, Nutrition Section, CHDD	"Give 'em a Hand" Improving Upper Extremity Function in Children with Disabilities
2010	**Margaret Jahn, ARNP,** CSHCN Coordinator, Whatcom County Health Department	Maximizing Mobility through Adaptation and Innovation
2011	**Steven Shores, MOT, OTR-L,** Occupational Therapist, Children's Therapy Unit **Special Recognition: Stanley Stamm, MD,** Pediatric Cardiologist	"I've Got Something To Say" Communication Strategies for Young Children with Physical Disabilities
2012	**Children With Special Health Care Needs Program,** Department of Health, State of Washington	**"**Adulthood - Here We Come" Smoothing the Healthcare Transition for Kids with Disabilities
2013	**William O. Walker, Jr., MD,** Chief., Division of Developmental Medicine, Seattle Children's Hospital	"What's New About an Old Diagnosis" Updates in the Care of Children with Cerebral Palsy
2014	**Iris Swisshelm,** Recreational Attendant, Specialized Programs, Seattle Parks and Recreation	Home from the NICU: Preparing for the Early Years
2015	**Pat Oelwein, MEd,** Coordinator, Down Syndrome Program, EEU, University of Washington	Down Syndrome – So Much to Talk About
2016	**Katherine TeKolste, MD, FAAP,** Developmental-Behavioral Pediatrics	The Heart of the Matter: Congenital Cardiac Conditions and Child Development
2017	**Beth Ellen Davis, MD, MPH, FAAP, Colonel (Retired), US Army,** Director, UW, Leadership Education in Neurodevelopmental and Related Disabilities (LEND)	The Juggling Act: Developmental Disabilities, Behavioral Challenges and Mental Health
2018	**Cathy Graubert, PT,** Ambulatory Rehabilitation Manager, Rehabilitation Medicine, Seattle Children's Hospital	Spina Bifida – Promoting Wellness and Preventing Pitfalls

2018 Duncan Seminar

Physical Therapy's Role in Promoting Physical Activity and Participation in Individuals with Spina Bifida

Presented by: Solveig M. Hart, PT, MPT, PCS

4/21/18

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

Disclosure Statement

- I Do not have any conflict of interest, nor will I be discussing any off-label product use.
- This class has no commercial support of sponsorship, nor is it co-sponsored.

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

Objectives

The following objectives will be addressed during this presentation:

- Discuss the role of physical therapy in the management of children with SB within NDV clinic.
- Describe equipment and bracing needs of children with SB.
- Consider strategies for maintaining function and mobility in children with SB as they grow and develop.

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

Neonatal Care

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Neonatal Care

- Positioning
- Muscle function and sensory testing
- Family education
- Assist with referral to early intervention
- Assist with early equipment needs (ex. Car bed)

Prone/Side lying Positioner

Neurodevelopmental Clinic Follow up

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

During yearly or twice yearly visits, assess child for:

- Pain
- Strength (0-5 scale manual muscle testing)
- Range of Motion with goniometer
- LE sensation testing with paperclip (sharp, light touch)
- Gait (video can be helpful)
- Postural alignment
- Skin integrity
- Functional mobility
- Equipment Needs

	Signs of Shunt Malfunction and Tethered Cord
Comorbidity	Symptoms
Shunt Malfunction	Headache, irritability, fever, nausea, vision or speech changes, changes in balance and postural stability (Marlin, 2004, Effgen, 2013)
Chiari Malformation	Child may present with changes in bowel and bladder control, muscle tone (increased spasticity), ataxia, and changes in upper extremity function (Stevenson, 2004)
Tethered Cord	Progressive changes in (loss of) strength or sensation, changes in bowel or bladder function, changes in functional mobility and gait pattern, back or lower extremity pain, changes in spasticity, loss of range of motion, or changes in foot positioning or progression of deformity, (Hinderer, 2006).

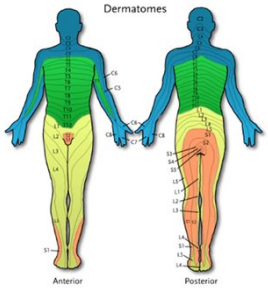
Use of Plantar Pressure and Observational Video in the Diagnosis of Tethered Cord in Spina Bifida: An Update.
Powell, Aaron J ., Bodkin, Amy, Elliot, Lindsay, Carollo, Jim, Wilson, Pamela
University of Colorado

- Tethered cord is often difficult to diagnose.
- Children at risk for or suspected of having a tethered cord were referred for comparison studies of gait and plantar pressures (PP). Digital video from side and front views, and PP were obtained at baseline, and upon referral after suspected tethered cord, and quantitatively analyzed tor changes.
- 75% of comparison studies did not reveal major changes, but **the majority of children who did reveal gait changes went on to have detethering procedures with good results.**
- Gait analysis and PP is relatively inexpensive but can add insight and sensitivity. Further analysis is needed.

Manual Muscle Testing – 5 year old boy

Manual Muscle Testing (Kendall, McCreary and Provance. Muscles Testing and Function, 4th Edition. Pp. 186-187)	
5	Can hold limb in test position against gravity with strong pressure
4	Can hold limb in test position against gravity with moderate pressure
3	Can move limb into and hold limb in test position against gravity but with little resistance
2	Can move limb through entire range of motion in a gravity eliminated position
1	Trace muscle contraction can be palpated
0	No muscle contraction can be palpated

Dermatome chart used for reference during sensation testing



Orthotics and Gait

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Examples of Orthotics

- Floor Reaction Ankle Foot Orthosis
- Fixed Ankle Ankle Foot Orthosis
- Supramalleolar Ankle Foot Orthosis
- Insert/arch support
- Knee Ankle Foot Orthosis (KAFO)
- Reciprocating Gait Orthosis (RGO)

Examples of Orthotics

Gait, with and without orthotics

- 4 year old girl with L5S1 MM
- 9 year old boy with lipomyelomeningocele, functional level L5S1

Orthotics and Gait

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Examples of Mobility Devices

- Posterior Wheeled Walker
- Lofstrand Crutches
- Wheelchair
- Power Mobility including Go Baby Go
- Standers and Mobile Standers

Things to consider……

- What is this child's lesion level and functional motor level?
- What can we predict regarding future mobility?
- Think beyond just independent ambulation and be open to MULTIPLE MODES of MOBILITY!
- What will insurance cover and how often will they cover it?
- Work closely with DME (durable medical equipment) therapists and vendors!

Considerations for introducing wheeled mobility

- Child's developmental level and age.
  - If the child is developmentally ready to begin moving around and exploring his or her environment on their own but is not independently ambulatory , consider a wheelchair.
- Joint Preservation, especially shoulders.

WW2

Motor Level	Criteria for Assigning Motor Levels (International Myelodysplasia Study Group Criteria for Assigning Motor Levels
T11 or above	Determined based upon sensory level and/or palpation of abdominals
T12	Some pelvic control in sitting or supine from the abdominals or paraspinals. Hip hiking from quadratus lumborum may be present.
L1	Weak (grade 2) Iliopsoas
L2	Iliopsoas, Sartorius, hip adductors all grade 3 or better
L3	Meets or exceeds criteria for L2 plus quadriceps are grade 3 or better
L3-L4	Exceeds criteria for L3 but does not meet criteria for L4
L4	Meets or exceeds criteria for L3 and medial hamstrings or anterior tibialis is grade 3 or better. May have weak peroneus tertius
L4-L5	Exceeds criteria for L4 but does not meet criteria for L5
L5	Meets or exceeds criteria for L4 and has lateral hamstring strength grade 3 or better, plus on of the following: gluteus medius grade 2 or better, peroneus tertius grade 4 or better, or tibialis posterior grade 3 or better.
L5-S1	Exceed criteria for L5 but does not meet criteria for S1
S1	Exceeds criteria for L5 plus at least two of the following: Gastrocnemius/soleus grade 2 or better, gluteus medius grade 3 or better, or gluteus maximus grade 2 or better (puckering of buttocks present)
S1-S2	Exceeds criteria for S1 but does not meet criteria for S2
S2	Meets or exceeds criteria for S1, plus gastrocnemius/soleus grade 3 or better, and gluteus medius and maximus are grade 4 or better
S2-S3	All lower extremity muscles normal strength (may be grade 4 in one or two muscle groups) and includes infants who appear normal and are too young to be bowel and bladder trained.
No-Loss	Meets criteria for S2-S3 and has no bowel or bladder dysfunction

Functional Motor Levels	Expected Muscle Function	Functional Mobility	Equipment Use	Orthotic Use
Thoracic	Abdominals Paraspinals Quadratus Lumborum	Non-Functional ambulation/standing during therapy, school or at home Wheelchair for Mobility	Standing Frame Wheelchair Parapodium	Trunk-Hip-Knee-Ankle-Foot Orthosis (THAFKO)
High Lumbar L1-L3	Hip Flexion Hip Adduction	Limited household ambulation, Wheelchair for mobility	Wheelchair Walker Forearm Crutches	Reciprocating Gait Orthosis (RGO) Hip-Knee-Ankle-Foot Orthosis (HKAFO)
Mid Lumbar L3-L4	Knee Extension	Household ambulation, Limited community ambulation Wheelchair for longer distances	Wheelchair, Walker Forearm Crutches	Knee-Ankle-Foot Orthosis (KAFO)
Low Lumbar L4-L5	Hip Abd Knee Flexion Ankle DF Ankle Inv. Toe Ext.	Household and Community ambulation Wheelchair for long distances	Wheelchair, Forearm Crutches	Ankle-Foot-Orthosis (AFO)
Sacral S1-S2	HipExt, Ankle PF Ankle Ev. Toe Flexion	Community Ambulation		Supramalleolar Foot-Orthosis (SMO), Foot Orthotic (FO)

Link to very useful chart on SBA website

- http://spinabifidaassociation.org/wp-content/uploads/2015/07/HOW-SB-LESIONS-IMPACT-DAILY-FUNCTION1-1.pdf

Slide 22

WW2 The number of slides may be too many, especially depnding on your videas. This might be one to delete - I think the same information is easier to follow for most folks in the next slide for functional levels.
Walker, William, 3/25/2018

2 year old girl with L3 MM

11 year old girl with L3 MM

Promoting Physical Activity

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Promoting Participation and Physical Activity

We know that motor level, presence/absence of shunt, and severity of contractures of other orthopedic deformity affects level of ambulation. (Dicianno et al, 2015)

But what about participation in physical activities, other than walking, for household ambulators and wheelchair users?

Personal and Environmental Factors affecting Physical Activity

Barriers
- Bowel/Bladder care
- Poor Fitness and obesity
- Equipment issues
- Lack of knowledge about opportunities.

Positive Factors
- Improving and advancing wheelchair skills
- Social influence of parents and positive role models
- Self Confidence
- Opportunities for Physical Activity

(Bloemen et al, 2015)

Unpublished Data from study: Mobility, Walking/Physical Activity, and Mobility-based Participation in Children with Spina Bifida

- Walker et al 2018 (Unpublished Data)

Opportunities for Physical Activity

- Outdoorsforall.org
- Shadowsealsswimming.org
- TOPS (The Outreach Program for Soccer) washingtonyouthsoccer.org
- Seattleadaptivesports.org
- Specialolympicswashington.org
- NCHPAD.org
- Sancaseattle.org

Summary

- Close monitoring of strength, sensation and mobility to detect tethered cord, shunt malfunction and Chiari II malformation
- Consistent follow up with medical team to ensure optimal bowel/bladder management.
- Optimal management of equipment needs
- Physical therapy intervention and home program to maximize strength and mobility, and prevent contracture
- Provide support and resources to maximize participation in physical activity

Questions?

solveig.hart@seattlechildrens.org

Seattle Children's
HOSPITAL • RESEARCH • FOUNDATION

Fostering Academic Success: Identifying and Addressing the Learning Needs of Children with Spina Bifida

Kate Bowen PhD
Neuropsychologist
Emily Myers MD
Developmental Pediatrician

Hillary Shurtleff, PhD, ABPP
Neuropsychologist
Seattle Children's Hospital
University of Washington

Disclosures

- **We do not have any conflict of interest, nor will we be discussing any off-label product use.**
- **This session has no commercial support or sponsorship, nor is it co-sponsored.**

Objectives

- Describe two general neuroanatomical differences that affect learning and cognition in individuals with Spina Bifida
- Identify common learning patterns and challenges experienced among individuals with spinal bifida
- Describe common therapeutic and educational approaches to address learning challenges

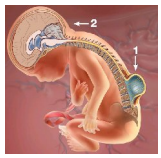
Overcoming Initial Perceptions

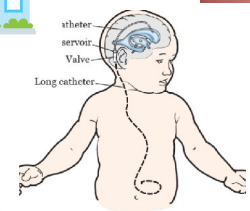
- Knowledge about Spina Bifida (SB) in the general population is scant
- We all tend to size people up quickly

Overcoming Initial Perceptions

https://www.healthination.com/health/overactive-bladder-how-much-water

Overcoming Initial Perceptions





https://www.mskcc.org/cancer-care/patient-education/patient-guide-pediatric-

Brain factors: The Reorganized Brain in SB

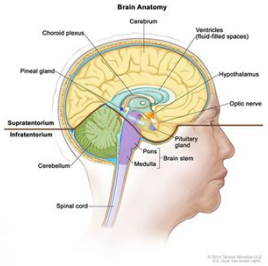
- Missing regions that should be present.
- Extra fiber tracts that should be absent.
- Thin regions that should be fat.
- Fat regions that should be thin.
- Normal sculpting that occurs too late.
- Abnormal sculpting that produces structurally dysmorphic regions.

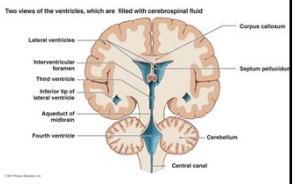
Maureen Dennis, 2010

Brain Differences that Affect Learning in Spina Bifida

White Matter Tracts
Corpus Callosum

Chiari II Malformation

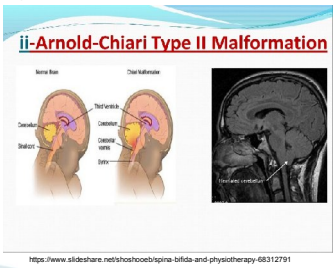




- **Cerebellum and brainstem tissue extend into the spinal canal**

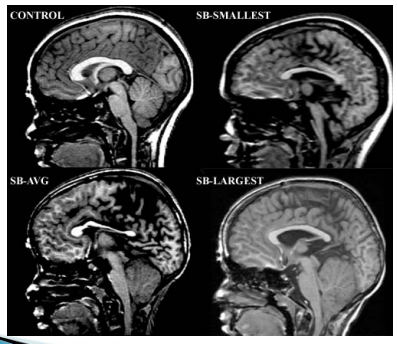
Brain factors: Chiari II Malformation

- Cerebellar abnormalities
- Midbrain abnormalities (i.e. tectal beaking)
- Potential hydrocephalus



https://www.slideshare.net/shoshooeb/spina-bifida-and-physiotherapy-68312791

Brain factors: Cerebellum



From Maureen Dennis 2010

Brain factors: Cerebellum

- Some behaviors affected by the cerebellum
  - Motor reaction time/regulation
  - ***Motor speed***
  - Speech fluency
  - Rhythm
  - Ataxias – of limb, trunk, and articulation
  - ***Attention*** *(and "on-line processing")*

Brain factors: Chiari II Malformation: Tectal Beaking

- Abnormality of the midbrain
- Eye Control: Nystagmus
  - Tubbs et al, *Childs Nerv Syst*, 2004
  - https://www.youtube.com/watch?v=YntJiBCz3pA

Images from www.imaging.consult.com

https://www.telegraph.co.uk/women/mother-tongue/familyadvice/8846737/These-perfect-parenting-classes-can-only-fuel-

Brain factors: Chiari II Malformation: Hydrocephalus

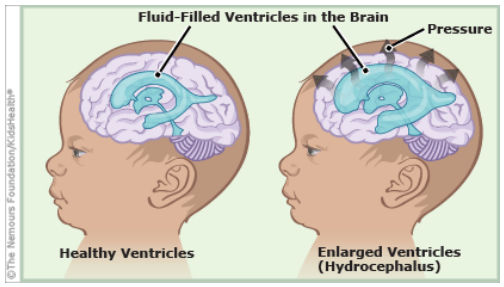


Image from: https://www.rchsd.org/health-articles/hydrocephalus/

Brain factors: Chiari II Malformation- Hydrocephalus

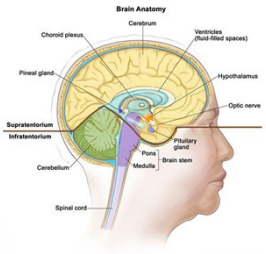
http://kimbiggio.com/2015/02/20/selective-ignoring/

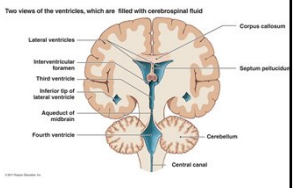
http://www.elperiodicodelafarmacia.com/articulo/ejercicio/deporte-clave-salud-cerebral/20151124105404002838.html

Brain Differences that Affect Learning in Spina Bifida

White Matter Tracts
Corpus Callosum

Chiari II Malformation



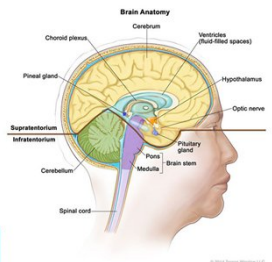


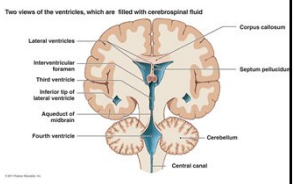
Cerebellar differences
Tectal Beaking
Hydrocephalus

Brain Differences that Affect Learning in Spina Bifida

White Matter Tracts
Corpus Callosum

Chiari II Malformation

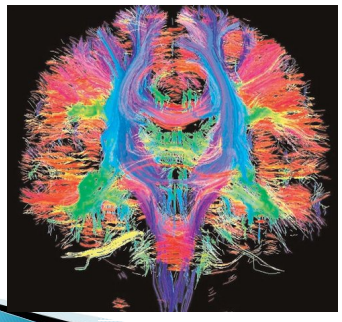




Arcuate fasciculus
Corpus Callosum

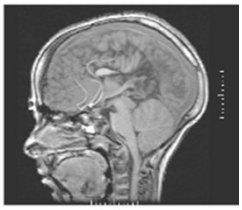
Brain factors: White Matter Tracts

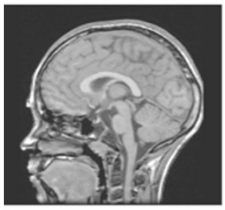
- White matter tracts in SB are shorter



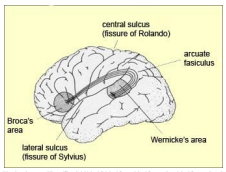
Brain factors: White Matter Tracks

- **Corpus Callosum** in SB may be:
  - Present, Absent or Partially Absent

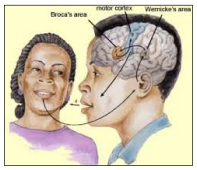


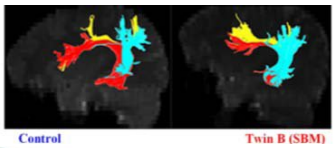


White Matter Tract Example: Arcuate Fasciculus



http://thebrain.mcgill.ca/flash/d/d_10/d_10_cr/d_10_cr_lan/d_10_cr_lan.html

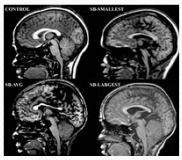


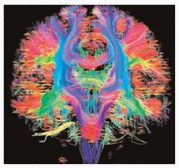


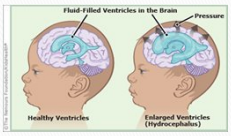
Glasan et al. *J Mag Res Imag* 2008

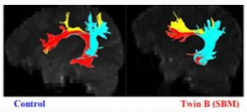
Slide Modified from Dennis, 2010

Recap: Pearls







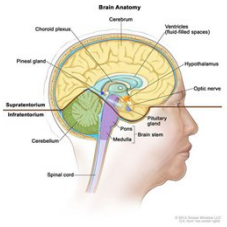


Chiari II Malformation

White Matter Tracks

Clinical Profile: Common Learning Patterns

3 Core Challenges



- Attention

https://www.appliedspirituality.com/books-with-online-courses/earthlings-quick-start-guide/eqsg-online-course-2/eqsg-online-course-pitfalls-distractions/

- Movement

- Timing

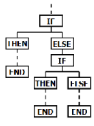
(Dennis & Barnes, 2010)

Broad Profile

- Strengths & weaknesses
  - Good at overlearned verbal expression
  - But weaker at perceptual reasoning/processing speed/working memory/executive functions
- IQ scores tend to show better verbal versus other abilities
- Usually chatty and sociable, so can "talk a good game"
- BUT, they may not meet others' expectations for productivity

Associative Processing

- Rule-based
- Concrete
- Routines
- Facts
- Sight words



Assembled Processing

- Linking information
- Generating new ideas
- Creating from scratch

(Dennis & Barnes, 2010)

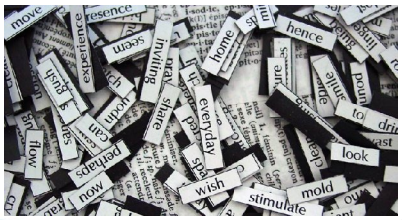
Potential Strength: Language

- Social, chatty, pursue social contacts

- Can “talk a good game”

Potential Weakness: Language

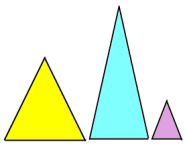
- Difficult to process
  - Wordy and off-topic
- “Cocktail party syndrome”

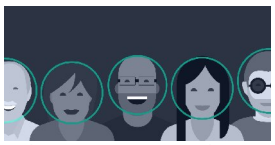


http://www.elemesos.com/index.php/itemlist/user/44.html?lang=en&limit=10&start=14480

Potential Strength: Visual Spatial

- Categorical classification
- Spatial relationships
  - Up, down, left, right





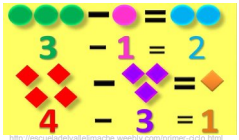
https://foxshack.wordpress.com/2016/12/02/web-based-face-tracking-webrtc-javascript/

Potential Weakness: Visual Spatial

- Mental rotation
- Organization
- illusions

Potential Strength: Academic

- Reading:
  - Word Reading
  - Decoding
  - Word Definitions



- Math:
  - Basic Calculation Skills

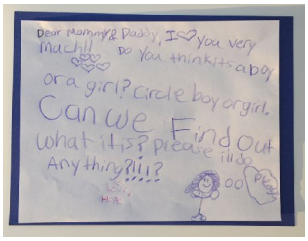
Potential Weakness: Academic

- Reading:
  - Context
  - Inferences

- Math:
  - Estimation
  - Problem Solving

Potential Weakness: Handwriting

- Rate, rhythm, and timing
- Motor control
- Spatial planning

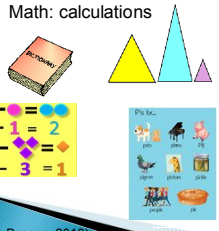


http://www.amandakern.com/blog/page/53/

Associative Processing

Strength

- Concrete:
  - Language: vocabulary
  - Visual Spatial: categories
  - Reading: sight words
  - Math: calculations



Assembled Processing

Weakness

- Integrated:
  - Language: context
  - Visual Spatial: mental rotation
  - Reading: context and integration
  - Math: estimation

(Dennis & Barnes, 2010)

Interventions and Supports

Overcoming Academic Challenges:
Interventions – **The first step is awareness –**
Have all issues been addressed?

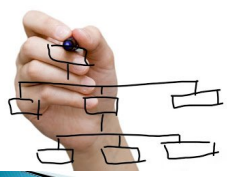
- Qualification?
- Medical? + appts.
- Toileting
- Social?
- Behavior?
- Academic?
- Team members?
- Communication?

Interventions – Specific Areas to Address

- Attention/Organization
- Socialization & Language
- Math
- Reading Comprehension
- Handwriting

Interventions – Classroom Productivity: Attention/Executive functioning

- Limit Distractions
- Need to be taught:
  - "metacognitive" strategies
  - Skills for organization, planning, monitoring



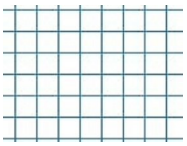
(Loveday & Edginton, 2010)

Interventions – Socialization and Language

- Speech/language therapy may help
- Social skills groups may help

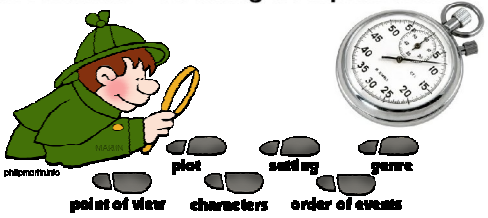
(Loveday & Edginton, 2010)

Interventions – Math



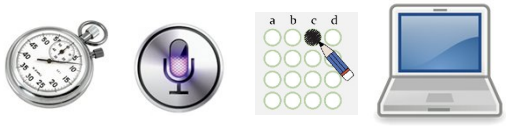
- Graph paper
- Calculator
- Times tables on the desk
- Strategies – i.e. checking work
- Provide notes instead of graphs or charts

Interventions – Reading Comprehension



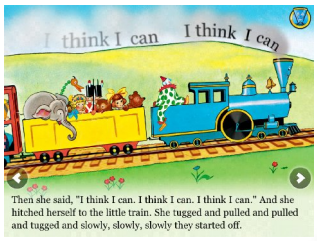
- More time
- Taught strategies for
  - Breaking down the information
  - Focusing in on the salient information

Interventions – Classroom Productivity: Handwriting



- Extra time
- Dictation programs
- Multiple choice tests
- Keyboarding
- *Handwriting without Tears* curriculum

Interventions - Awareness



- Good effort
- Motivated by social praise

Overcoming Challenges in the Academic Environment

We only scratched the surface…

The more that parents, teachers, and school personnel know about Spina Bifida in all of its complicated manifestations, the better they will be able to work together to create appropriate learning programs and foster healthy development into adulthood.

Thank you so much for your time.

Promoting optimal feeding and nutrition in children with feeding difficulties and Spina Bifida

Duncan Seminar
April 20, 2018
Kim Nowak-Cooperman, MS, RDN, CD and Peggy Smith, OTR

Disclosure Statement

- **We do not have any conflict of interest, nor will we be discussing any off-label product use.**
- **This presentation has no commercial support or sponsorship, nor is it co-sponsored.**

Objectives

- Participants will be able to describe the nutritional and feeding challenges of children with a diagnosis of spina bifida at various stages of development.
- Participants will be able to describe the factors that contribute to increased risk of overweight and underweight.
- Participants will be able to describe the signs and symptoms of dysphagia associated with Chiari II malformation.
- Participants will be able to develop interventions to manage nutritional and feeding challenges with spina bifida in their own practices

Pitfalls by life stage

- Newborn
- Infancy
- Toddlerhood
- Preschool
- School age
- Adolescence/early adulthood

Factors in feeding and nutrition problems

- Chiari II malformation and dysphagia
- Lower energy expenditure
- Lower lean body mass
- Reduced participation in physical activities
  - Increased risk of obesity
- Reduced sensory awareness, increased risk for wounds
- Bowel and bladder issues
- Increased risk for osteopenia and bone fractures

Newborn: initial hospitalization

- Surgery to correct lesion within 24-48 hours.
- Presence of symptomatic Chiari II malformation is monitored
  - Dysphagia, apneic spells, stridor, hoarse or high pitched cry
- Surgery to place shunt for hydrocephalus.(80-90%)
  - Delayed feeding
  - Increased needs to heal surgery
  - Need to provide nutrition support

Case example: Newborn

- Newborn who presents with a L5-S1 lesion.
- Chiari II Malformation is present shunt is placed at time of the back closure.
- Baby placed in prone or side-lying.
- Nutrition goals developed.
- Feeding strategies explored.

Routine clinic monitoring

- The goal for follow up in clinic is 1-2 weeks after hospital discharge
  - This visit includes a NDV nurse practitioner and OT/PT
- At 2-3 months routine clinic visit includes nutrition and feeding screening
  - Nutrition and growth
    - Growth parameters: overweight, underweight, short stature
    - Diet adequacy: variety, amount, meal routines
  - Feeding progression
    - Chewing, swallowing, sensory issues

Case example: Infant

- 2 month old infant who presents with poor weight gain and feeding challenges
- Assess growth on WHO growth charts: goal steady predictable growth and weight gain, avoid underweight or obesity
  - This infant has had slower weight gain for the past 4 weeks, weight/length has declined from the 75th %ile to the 25th
- Assess diet adequacy: breast or bottle feeding
  - Parents note difficulty starting each feeding
- Feeding ability, challenges: swallowing
  - Ongoing OT or Speech for regular feeding therapy and in depth assessment
- Feeding evaluation finds problems with sucking and recommended nipple, positioning
- Nutrition recommends increased caloric density of formula (or pumped breast milk) and feeding plan with optimal goals of intake
- OT recommends local feeding therapy to take place weekly
- Nutrition follow up is recommended in 1 month due to age.
- Goals at follow up: improved feeding sessions and appropriate weight gain and growth

Case example: Toddler

- Assess growth on WHO growth charts until age 2 years: goal steady predictable growth and weight gain, avoid underweight or obesity.
- Growth in linear height tends to slow around age 2, increasing risk of overweight/obesity
- Nutrition evaluates diet adequacy: variety, meal/snack routine
- Follow up: in 6 months with goal of age appropriate feeding, increased variety and slower weight gain velocity to match growth in length.

Case example: Preschool

- **3 year old with thoracic M&M and Chiari 2, presents with long term poor growth and poor weight gain**
- **Growth: weight gain velocity has crossed 2 %ile channels over the past year, and BMI is on the 5th %ile/.**
  - **Assess growth on WHO growth charts until age 2 years, then use CDC charts: goal steady predictable growth and weight gain, avoid underweight or obesity.**
- **Meals are unpleasant and last over 1 hour each**
  - **Feeding challenges include gagging, choking with eating**
  - **Assess diet adequacy: variety, meal/snack routine**
  - **Assess energy expenditure: ambulatory status/ability**
- **Feeding evaluation with significant concern for swallowing and sensory challenges. VFSS indicates significant oro-pharyngeal dysphagia.**
- **G-tube placement is recommended to supplement safe oral intake of purees and nectar thick liquids.**

Common signs and symptoms of dysphagia in Chiari II malformation

- Problems sucking and swallowing
- Difficulty positioning for feedings
- Difficulty forming a seal on nipple
- Refusal of cup, sippy cup
- Loss of food from mouth
- Nasal regurgitation
- Long feeding times
- High number of formula changes to improve tolerance

Common signs and symptoms of dysphagia in Chiari II malformation, con't

- Perceived lack of satiety
- Refusal to advance textures/increase variety
- Delays in self-feeding
- Choking with feedings
- Poor saliva control
- Aspiration
- Weight loss/growth failure
- Medications that can interfere further with eating/drinking
  - Glycopyrolate
  - Ditropan

Case example: School Age

- 10 year old with lower lumbar M&M who presents with overweight, picky eating, problems with chewing and feeling of some foods "getting stuck" when he swallows.
- Assess diet adequacy: variety, meal/snack routine
- Feeding ability, challenges: swallowing, chewing, texture issues
- Wants to lose weight and is motivated to change diet and increase activity

Sensory Processing Strategies "The Steps to Eating"

- LOOK AT THE FOOD!
- TOUCH IT!
- SMELL THE FOOD!
- PUT IT ON YOUR LIPS
- LICK IT! BITE IT!
- SPIT IT OUT!
- SWALLOW!

Case example: Adolescent

- 14 year old high lumbar M&M with history of decubitus ulcer on left heel. She is non-ambulatory and has short stature with fluctuations in her weight. Current weight is down 10 pounds from 1 year ago. She has ongoing issues with constipation and recurrent UTI's.
- Sedentary, prefers to play video games
  - Assess growth on the CDC growth charts: goal steady predictable growth and weight gain, avoid underweight or obesity. BMI goal 10th-85th %ile. Her BMI: 25th %ile .
  - Assess energy expenditure: fitness
- Has been "dieting" to control her weight. Avoids milk to cut back on calories. Skips breakfast.
  - Assess diet adequacy: variety, meal/snack/beverage routine including how choices address constipation. Inquire about dieting, weight loss history. May be at risk for an eating disorder.
  - At risk nutrients: calcium, Vitamin D due to lack of sunlight exposure, living in PNW, Vitamin D level checked and was 22 ng/ml (normal: 30-100 ng/ml)
  - Increased need for protein, vitamins and minerals (especially iron, zinc and Vitamin C)adequate fluid to address constipation and UTI's, fiber
  - Lower bone mineral density, increased osteoporosis and pathological bone fractures
- Nutrition recommendations: goals agreed upon using motivational interviewing:
  - Eat yogurt for breakfast and drink skim milk at lunch and dinner , drink 2 liters of fluid daily, consume 3 servings of raw fruit or vegetable daily, include 2 servings of whole grains (for fiber) add a multivitamin with iron
  - Increase physical activities: 30 minute "walk" 3 times per week

Nutrition challenges

- Diet adequacy in setting of lower energy needs
- Overweight
- Underweight
  - Medications that reduce appetite:
    - AEDs: topiramate, zonisamide
    - ADHD and behavior meds: concerta, Adderall, focalin, etc.
- Constipation/bowel management
  - At risk nutrients: calcium, Vitamin D
  - Lower bone mineral density, increased osteoporosis and pathological bone fractures
  - Lack of sensation, at risk for wounds
  - Increased nutritional needs after surgeries
    - Shunt issues
    - Orthopedic
    - Tethered cord

Feeding Challenges

- Chiari II Malformation affects most children with Spina Bifida
- Dysphagia is a common challenge affecting children from birth to adolescence due to the impact of Chiari II malformation.
- Oral motor function and swallowing challenges need to be monitored throughout their developing years.
- Sensory Processing challenges can impact these children well.

References

- Wittenbrook W, Best practices in nutrition for children with myelomeningocele. *ICAN*.2010;2:237-245
- Kreutzer C, Wittenbrook W, Nutrition issues in children with myelomeningocele (spina bifida). *Nutrition Focus*. Sept/Oct 2013; 28(5)
- DeVore J, Shotton A, eds. Pocket guide to children with special health care and nutritional needs. Chicago, IL: *Academy of Nutrition and Dietetics*. 2012 (take out if we don't use the early intervention slide)
- Silber TJ, Shaer C and Atkins D, Eating disorders in adolescents and young women with spina bifida. Int J Eat Disord. 1999;25(4):457-61.

References

- Kafadar I, Kilic BA, Yilmaz FK and Kilic M, Bone mineral density in pediatric patients with meningomyelocele. Childs Nerv Sysy. 2016;32(1): 111-9.
- Mazur LJ, Wilsford LD, Rosas L and Sullivan E, Low 25-hydroxyvitamin D levels in children with spina bifida. South Med J.2016;109(1):31-5.
- Martinelli V, Dell'Atti C, Ausili E, Federici E, Magarelli N, Leone A, et al.,Risk of fracture prevention in spina bifida patients: Correlation between bone mineral density, vitamin D, and electrolyte values. Childs Nerv Syst. 2015;31(8):1361-5.
- Ausili E, Focarelli B, Tabacco F, Fortunelli G, Caradonna P, Massimi L, et al., Bone mineral density and body composition in a myelomeningocele children population: Effects of walking ability and sport activity. Eur Rev Med Pharmacol Sci. 2008;12(6):349-54.

References

- Liusuwan RA, Widman LM, Abresch RT, Styne DM and McDonald CM, Body composition and resting energy expenditure in patients aged 11 to 21 years with spinal cord dysfunction compared to controls: Comparisons and relationships among the groups. J Spinal Cord Med. 2007;30 (Suppl 1):S105-11.
- Littlewood RA, Trocki O, Shepherd RW, Shepherd K and Davies PSW, Resting energy expenditure and body composition in children with myelomeningocele. Pediatr Rehabil. 2003; 6(1):31-7.
- Quan A, Adams R, Ekmark E and Baum M, Bone mineral density in children with myelomeningocele. Pediatrics. 1998; 102(3):E34.

References

Stevenson, KL. Chiari type II malformation : past, present, and future. Neurosurg, Focus. 2004;16:1-7

Mathisen BA, Shepherd K. Oral-motor dysfunction and feeding problems in infants with myelodysplasia. Pediatr. Rehabil.1997;1: 117-122.

Apkon,S.,Grady, R., Hart S. et al Advances in the Care of Children with Spina Bifida. Advances in Pediatrics. 2014;61:33-74

Toomey, Kay A. PhD, Kortsha, B. MA, OTR/L, Langerborg, D., CCC-SLP; SOS Approach to Feeding. 4 Day Basic Course. 9/14-17/17. Seattle WA

Thank

You!

Questions?

2018 Duncan Seminar

From Clinical Knowledge to Practical Application: Q & A with the Experts

Moderated by: Lisa Herzig, MD
Date: April 20, 2018

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

Disclosure Statement

- I do not have any conflict of interest, nor will I be discussing any off-label product use.
- This class has no commercial support of sponsorship, nor is it co-sponsored.

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION

Objective

- Discuss one way you will personally apply a relevant clinical management tip to your practice.

Seattle Children's HOSPITAL • RESEARCH • FOUNDATION