Pediatric Spinal Cord Injury Lifelong Outcomes

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Unique Features of Spinal Cord Injuries in Children and Adolescents

- Uniqueness of pediatric SCI is based upon:
  - the dynamic nature of growth and development in children and adolescents
  - the mutual interactions of growth & development with the manifestations & complications

Uniqueness of Pediatric SCI

- Children who sustain a SCI have a relatively long-lifespan
  - Susceptible to secondary health conditions over a longer period of time compared to adult-onset SCI
    - Premature aging
    - Live with secondary health conditions for a longer time
  - Children are also susceptible to unique complications

Uniqueness of Pediatric SCI

- Children are not small adults
- Must take into account growth & development from infancy through adolescence into adulthood
  - Physical
  - Physiological
  - Psychological
  - Cognitive

Developmental Considerations Physical

- Size
  - Weight
  - Physical size
  - Bladder volume
- Neuro-Musculoskeletal
  - Linear growth
    - Scoliosis

Developmental Considerations Physiological

- Heart rate
- Blood pressure
- Bowel and bladder function

Developmental Considerations Psychological

A SCI impacts/interacts with all the psychological changes characteristic of each developmental stage

Developmental Considerations Cognitive

Ability to communicate

- Symptoms of autonomic dysreflexia
- Expression of pain

Ability to learn/understand

- Self-catheterization
- Pressure ulcer prevention

Reasoning

Manifestations of SCI that are a consequence of young age

- SCIWORA = spinal cord injury without radiologic abnormalities
- Delayed onset of neurologic deficit

Complications of SCI related to young age at time of injury

- Hypercalcemia
- Scoliosis
- Hip subluxation and contractures

Incidence

Overall = 40 cases/1,000,000 or 10,000 new cases/year

<15 years of age 3-5% of SCIs
<20 years of age 20% of SCIs

Gender

	Male	Female
0-5 years	51%	49%
6-12 years	58%	42%
13-15 years	69%	31%
16-21 years	83%	17%
22+	81%	19%

Etiology

	0-5 yrs	6-12 yrs	13-15 yrs	16-21 yrs	22+ yrs
MVI	60%	57%	41%	52%	41.5%
Violence	5%	3.7%	12.2%	20.4%	9.4%
Sports	0%	9.4%	25.9%	17.3%	8.5%
Falls	7.5%	7.5%	7.6%	6.8%	31.2%
Med/surg	16.3%	18.7%	8.1%	0.8%	4.6%
Other	11.2%	3.7%	5.1%	2.9%	4.8%

Etiologies unique to pediatric SCI

- Lap-belt
- Birth injury
- Fibrocartilagenous emboli
- Transverse myelitis
- Acute flaccid paralysis
- High cervical lesions
  - Downs
  - Skeletal dysplasias
  - JRA

Transverse myelitis

- Diagnosis requires
  - Spinal cord inflammation
    - CSF pleocytosis
    - ↑ CSF IgG
    - Gadolininium enhancement on spinal MRI
    - Absence of CSF infection
- Bimodal distribution in peds
  - One peak in toddlers <3 years of age
  - Second peak 11-17 years of age

Transverse myelitis

- Better prognosis
  - Older age of onset
  - Shorter time to dx
  - Lower neurological level
  - Absence of T1 hypointensity acutely
  - Lack of CSF pleocytosis
  - Fewer affected segments

Transverse myelitis

- Prognosis
  - 1/3 completely recover
  - 1/3 some improvement but residual deficits
  - 1/3 have little to no improvement

Acute flaccid paralysis

- Acute flaccid myelitis
- Guillain Barre
- Toxic neuropathy

Acute flaccid myelitis

- Sudden weakness in one or more arms or legs
- Decreased muscle tone or absent reflexes
- Occasional involvement of cranial nerves
  - Facial weakness
  - Swallowing difficulties
  - Drooping of eyes

Acute flaccid myelitis

Confirmed case

- Acute onset of focal limb weakness AND
- MRI
  - Largely restricted to gray matter
  - Spanning one or more spinal segments

Probable case

- Acute onset of focal limb weakness AND
- CSF with pleocytosis

Acute flaccid myelitis

Etiologies

- Enterovirus D68
- West Nile virus
- Herpes

Acute flaccid myelitis

Clinical presentation

- Acute onset of asymmetric limb weakness

Median age=7.6 years (5months – 20 years)

Prognosis

- 2/3 some improvement
- 1/3 no improvement
- No one fully recovered

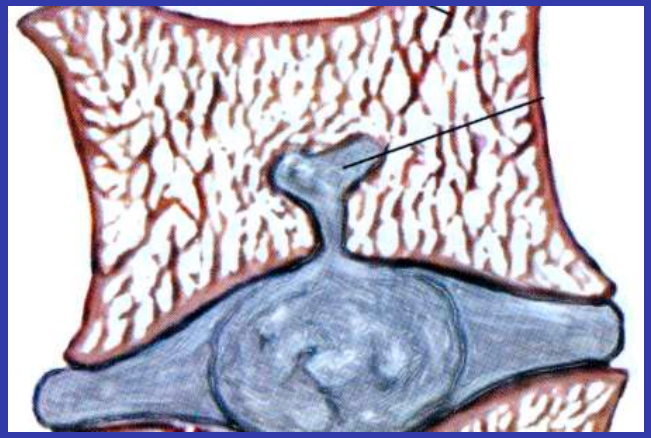
Fibrocartilagenous emboli

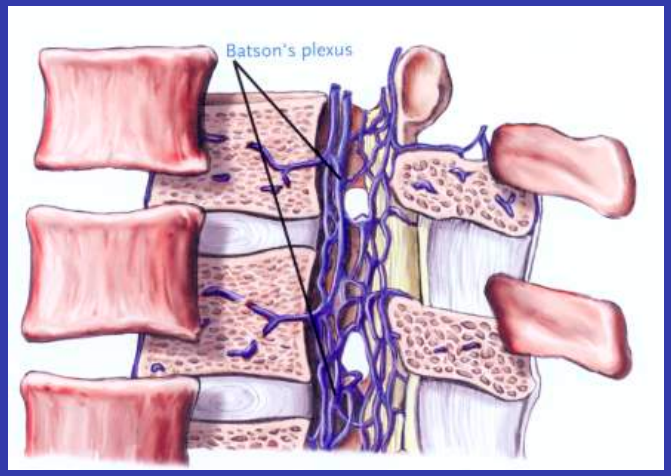
Acute vertical disk herniation ➔

Retrograde embolization of nucleus pulposus ➔

Vasculature supplying the anterior spinal cord ➔

Spinal cord infarct





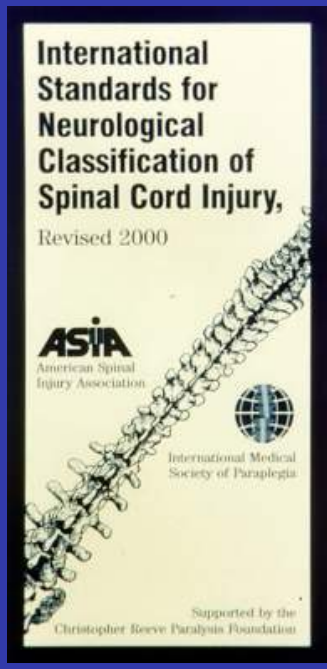
- Rapid onset of weakness after sudden onset of severe pain
- May be precipitated by physical exertion

Neurological Impairment versus Age at Injury

	Para	Tetra	Complete
0-5 years	55%	45%	80.7%
6-12 years	62.5%	36.5%	68.4%
13-15 years	47.6%	51.9%	55.6%
16-21 years	46.8%	52.9%	56.8%
22+ years	39%	60.3%	39.1%

Neurological Impairment versus Age at Injury

- Limitations of ISNCSCI in children
  - Especially in children ≤ 5 years of age
  - Safety pin and children's reaction
  - Discriminate sharp and dull
  - Validity of anorectal examination
    - Voluntary anal contraction



Mulcahey et al., Arch Phys Med Rehabil, 2011; 92:1264-9; Samdani, et al. Spinal Cord 2011; 49(3): 352-6.

SCIWORA = Spinal Cord Injury WithOut Radiologic Abnormality

SCI without evidence of fracture or dislocation on:

- Routine spine radiographs
- Tomography
- CT
- Myelography
- Dynamic flexion/extension studies

SCIWORA

0-5 years	64.2%
6-12 years	32.5%
13-15 years	22.3%
16-21 years	19.5%

Spine deformity

- Etiology
  - Muscle weakness/imbalance
  - Residual spine deformity following fracture
  - Iatrogenic: laminectomy

Prevalence of scoliosis

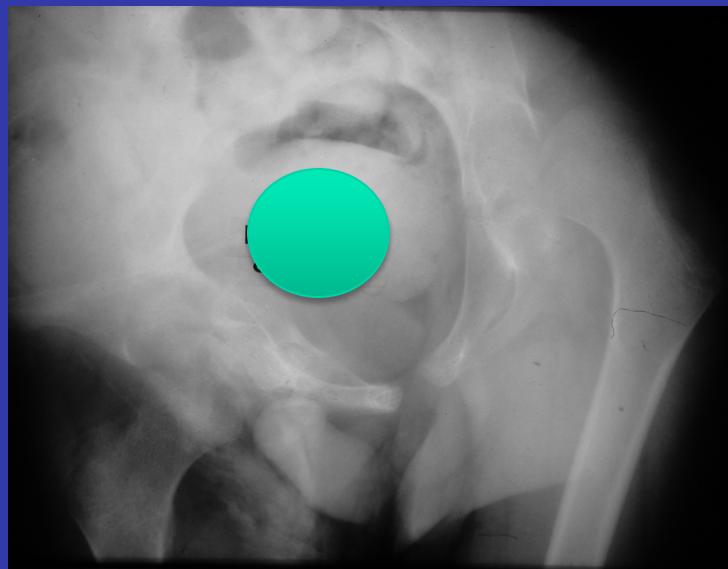
Dependent upon age at time of injury

	Prior to puberty	After puberty
Prevalence of Scoliosis	98%	20%
Need for surgery	67%	5%

Hip Subluxation/Dislocation

- Incidence = 30-40%
  - More common in children who are younger when injured

Age when injured	Incidence of hip instability
≤8 years	62%
≥9 years	10%



The ultimate measures of success in caring for youth with SCI

Optimal participation and satisfaction as they progress through childhood and adolescence

Bottom line

They become adults with productive and satisfying lives

Goals for adults with pediatric-onset SCI

- Healthy
  - Physical
  - Emotional
- Independent
  - Living
  - Mobility
  - Autonomy
- Employed
- Participate fully in their communities
- Satisfying lives

Challenges in caring for the child with a spinal cord injury

- Establish goals that evolve as the child matures and ultimately becomes an adult
  - Establish a sound foundation for a successful transition into adulthood that lasts a life-time
- Prevent complications
- Maintain focus on the big picture
  full participation and satisfying life
- Both innovative and standard interventions must support this overarching goal

Challenges in caring for the child with a spinal cord injury

- Need to have outcome measures that accurately assess key outcomes from injury as a child throughout adulthood
- Solid understanding of the natural history of pediatric SCI throughout their lifespan
- Identify risk factors for suboptimal outcomes throughout their lifespan

Outcome Instruments

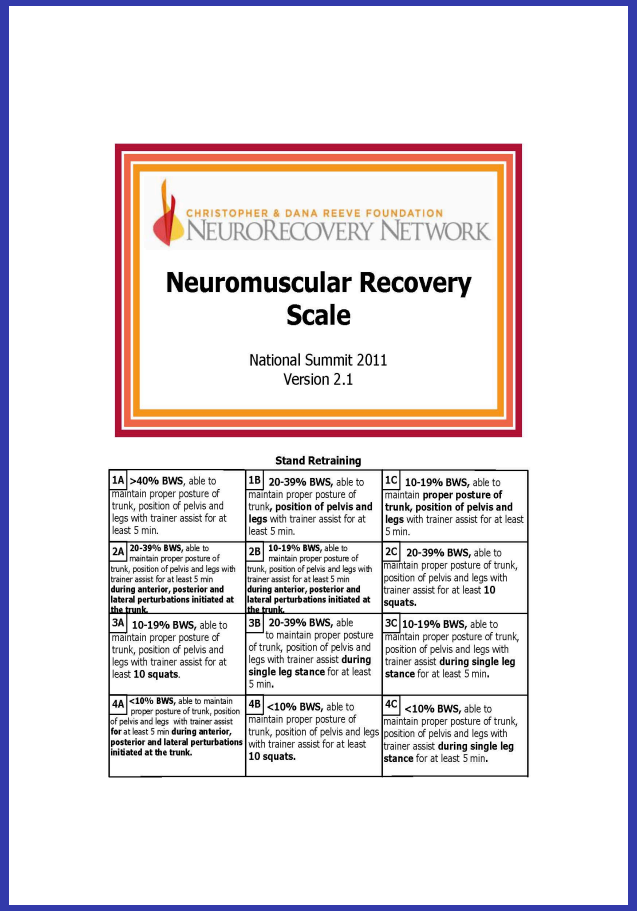
- Few instruments with psychometric support for children with SCI
- Difficulty in assessing outcomes and determining treatment effectiveness
- Priority for pediatric SCI rehabilitation
  - Establishing psychometric properties of existing tools
  - Developing and validating new tools, meaningful to children
  - Develop measures that cover the lifespan

Pediatric SCI Instrument: Computer Adaptive Tests

- Large item banks of mobility, activity and participation (Calhoun et al)
- Filter questions
  - Sex, age, use of equipment, type of chair
- Computer adaptive tests and short forms
- Linked with adult SCI CAT and short forms

Mulcahey et al 2012; Bent et al 2013; Tian et al 2014

Pediatric SCI Instrument: Pediatric Neurorecovery Scale



CHRISTOPHER & DANA REEVE FOUNDATION
NEURORECOVERY NETWORK

Neuromuscular Recovery Scale

National Summit 2011
Version 2.1

Stand Retraining

1A **>40% BWS**, able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least 5 min.	1B **20-39% BWS,** able to maintain proper posture of trunk**, position of pelvis and legs** with trainer assist for at least 5 min.	1C **10-19% BWS,** able to maintain **proper posture of trunk, position of pelvis and legs** with trainer assist for at least 5 min.
2A **20-39% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least 5 min **during anterior, posterior and lateral perturbations initiated at the trunk.**	2B **10-19% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least 5 min **during anterior, posterior and lateral perturbations initiated at the trunk.**	2C **20-39% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least **10 squats.**
3A **10-19% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least **10 squats**.	3B **20-39% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist **during single leg stance** for at least 5 min**.**	3C **10-19% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist **during single leg stance** for at least 5 min**.**
4A **<10% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist **for** at least 5 min **during anterior, posterior and lateral perturbations initiated at the trunk.**	4B **<10% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist for at least **10 squats.**	4C **<10% BWS,** able to maintain proper posture of trunk, position of pelvis and legs with trainer assist **during single leg stance** for at least 5 min**.**

Behrman A, Mulcahey MJ, Ardolino E
Craig H. Neilsen Foundation. Award Number: 260284
Neuromuscular Recovery Scale for Pediatric Spinal Cord Injury

Common Data Elements

- NINDS
- SCI CDEs
  - Pediatric modifications

International SCI datasets

- Need pediatric modifications
- Examples
  - Etiology
    - Birth injury
  - Bowel, bladder and sexual functioning

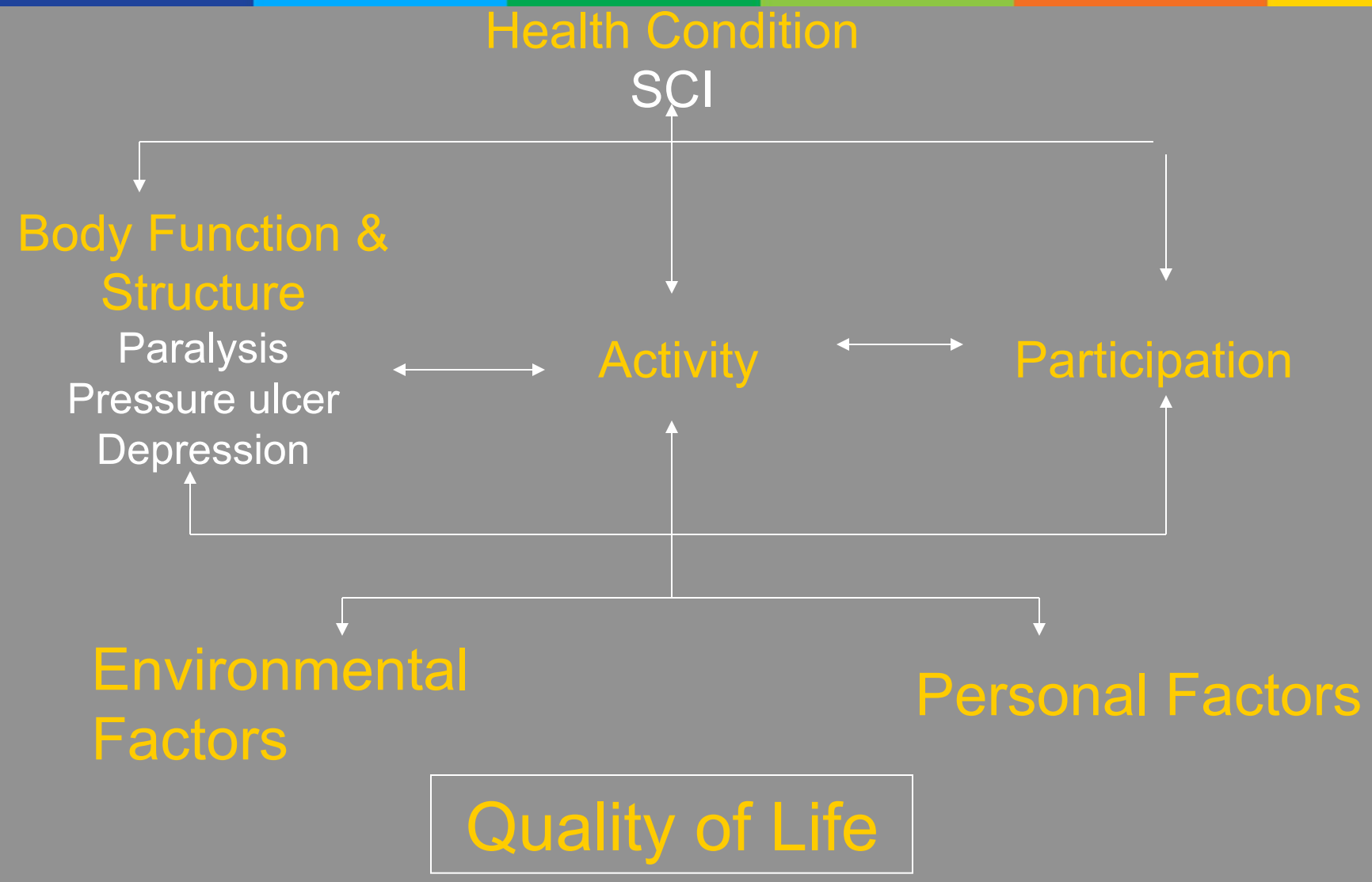
Profound change in our approach to rehabilitation & its goals

- A more comprehensive approach with incorporation of disability model
- Outcomes and different spheres of life
- The time-line
  - The lifespan
  - Developmental stages

Outcomes

- Education
- Employment/Occupation
- Living independently
- Mobility
- Participation
- Social development
- Sexuality
- Psychological functioning
- Health/Wellness
- Quality of life

WHO International Classification of Functioning, Disability, and Health (ICF) Model



Focus has changed from fixing impairments to focusing on participation and life-satisfaction

- Walking → Mobility
  - Full participation in school and community throughout the lifespan

Mobility versus Walking

Mobility is a critical factor that facilitates activity and participation

- The mobility modality must accomplish the task at hand
  - Efficient
  - Independent
  - Cosmetic
  - Socially acceptable
- Mobility for the entire lifespan
  - Issues of aging
    - Preservation of function
    - Prevention of complications

Walking

- Walking is integrated into a comprehensive rehab program where the goal is participation and life satisfaction versus “Walking" as the sole goal versus
- Walking versus the wheelchair
  - Is it one or the other?

Mobility
More Comprehensive Goals

- Ambulation
- Standing
- Wheelchairs
- Community
- The electronic world

Mobility - Transitions

Changes need to be viewed as transitions and not as failures

Transition of school-aged child from ambulation with orthotics to wheelchair

Exoskeletons

Activity-Based Rehab Locomotor Training

- Step Training using Body Weight Support on a Treadmill (BWST) and manual assistance
- Robot-assisted walking therapy
- Incomplete SCI

Walking Pitfalls

- Long-term complications
  - Upper extremity from assistive devices
  - Lower extremity from joint instability
- Guilt or sense of failure
  - Child/adolescent or parent
  - Failure to pursue “innovative programs”
  - Failure to continue to ambulate
  - Utilization of wheeled mobility

Mobility - More Comprehensive Goals

Wheelchairs

- Advances in wheelchair design
  - Ultra-light weight
  - Power assist
  - Advances in power chairs
- Upper extremity preservation
- Seating systems & pressure mapping
- Power versus manual

Mobility: More Comprehensive Goals

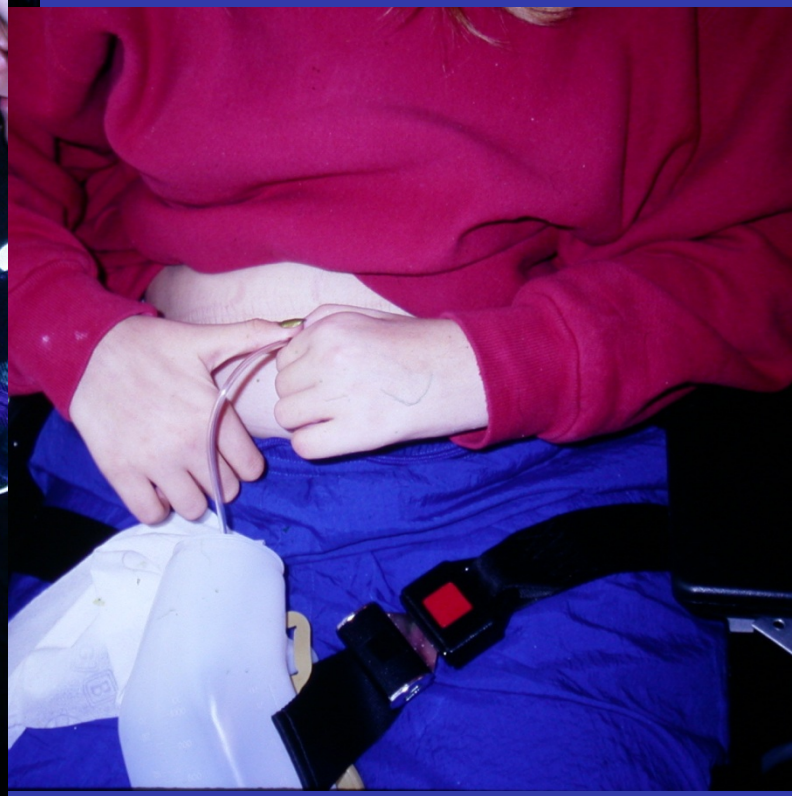
Community mobility

- Motor vehicles
- Public transportation
- The great outdoors

Went from fixing impairments to focusing on participation and life-satisfaction

- UE interventions → Self-catheterization

Continent Catheterizable Urinary Conduits
Upper Extremity Reconstruction



Psychosocial Outcomes

- Youth seem to be developmentally appropriate in terms of the content of their participation
- Youth seem to have lower levels of participation and quality of life ratings than typically developing youth

Psychosocial Outcomes

- Mobility and participation and quality of life of youth with SCI

- Ability to enter/exit independently was related to:
  - Participating in more informal activities
  - Participating in informal activities more often
  - Greater emotional, social, and psychosocial quality of life

Sexuality

- Issues common with all youth
  - Sexual development, functioning, and STDs
- Issues common for youth with all special needs
  - Self-esteem
- Disorder-specific issues
  - Sexual functioning
  - Fertility
  - Genetics

Caregivers

- Caregivers are critical to the well-being of youth with SCI
- The toll of caregiving
  - Physical
  - Emotional
    - 20% of caregivers reported experiencing moderate or severe anxiety
    - 22% moderate or severe depression

Caregivers

- **Direct relationships between caregiver education, mental health, burden, and problem solving**
  &
  mental health and quality of life of youth with SCI
- Caregivers having **more formal education** is related to:
  - Fewer symptoms of anxiety & depression among youth
  - Youth participating in more formal activities and experiencing greater enjoyment in formal activities

Caregivers

Increased caregiver anxiety & depression are related to:

- Decreased child psychosocial HRQOL
- Increased child anxiety and depression

Increased caregiver burden is related to

- decreased child psychosocial HRQOL

More effective problem solving is related to

- increased child psychosocial and physical HRQOL

Caregiver mental health influences how caregivers report about their child's outcomes

Approach shifted from emphasis on short-term goals to long-term goals

Our management plan expanded to a grander time-span → the entire lifespan

- Transition
- Issues of aging
  - Preservation of function
  - Prevention of complications

Transition for youth with SCI Challenges

- Challenges common to all youth
- Challenges common to most diagnostic groups
  - Environmental barriers
    - Physical
    - Attitudinal
- Disease/disorder specific
  - Physical
- Youth/Family specific
  - Personal factors
  - Socio-cultural

Goals for adults with pediatric-onset SCI

- Healthy
  - Physical
  - Emotional
- Independent
  - Living
  - Mobility
  - Autonomy
- Employed
- Participate fully in their communities
- Satisfying lives

The ultimate measures of success in caring for youth with spinal cord injuries

Optimal participation and satisfaction as they progress through childhood and adolescence

Bottom line

As adults they will lead productive and satisfying lives

Dynamic nature of growth and development throughout lifespan adds an additional dimension to the ICF

- Infants
- Toddlers
- Preschool-aged child
- School-aged child
- Early adolescence
- Late adolescence
- Emerging adults
- Adults
- Aging adults

What is an emerging adult?

- 18-25 year olders
- No man's land between the care-free adolescent and the adult with responsibilities
  - Job
  - Mortgage
  - Family
- May be the most tumultuous period of life after birth

A developmental perspective of the ICF model

	Mobility	Employment
Infant	Stroller / car seat	
Toddler	Walking	
Preschool	Walking / tricycle	+/- Chores
School-aged	Walking / bike	Chores/ Neighborhood jobs
Early adolescence	Walking / public transportation	Neighborhood jobs
Late adolescence	Motor vehicles	Community-based jobs
Emerging adult	Planes, trains & automobiles	Summer jobs First real job
Adult	Planes, trains & automobiles	Real jobs
Older adult	Loss of independent wheeled mobility	Retirement Volunteer

A developmental perspective of the ICF model

	Participation/Socialization	Where
Infant	Family	Home
Toddler	Playgroups	Home
Preschool	Playgroups	Neighborhood centers
School-aged	Sports and Scouts	Neighborhood
Early adolescence	Sports & hanging out	Community
Late adolescence	Dating & hanging out	Community & beyond
Emerging adult	Partying	Community & beyond
Adult	Marriage	Community & beyond
Older adult	Senior citizen groups	Community

How can we improve transition for youth with special needs

- Understand the natural history of youth with specific disabilities
- Evaluate factors associated with good and not-such good outcomes
  - Develop specific interventions when feasible
  - Target high risk groups for interventions

Long-term Outcomes of Pediatric SCI

- Caroline J Anderson, PhD
- Kathy Zebracki, PhD
- Kathy M Chlan

Long-term Outcomes of Pediatric SCI

- Identify long-term outcomes of adults with pediatric-onset SCI
  - Independent living and driving
  - Employment
  - Participation
  - Medical complications
  - Mental health
  - Quality of Life
- Identify factors associated with these outcomes
  - Demographics, impairment factors
  - Environmental factors

Participants

- 466 adults who sustained their SCI ≤ 18 y/o
- 63% males
- 54% tetraplegia
- 70% with AIS A
- Age of injury, mean = 13.9 (0-18)
- Age at follow-up, mean = 30.5 (24-45)
- Duration of injury, mean = 16.2 (6-38)

Long-term Outcomes of Pediatric SCI

	SCI	Census
College education	40%	32%
Employed	60%	90%
Married	21%	41-65%
Live independently	64%	88%

Long-term Outcomes of Pediatric SCI

• Pressure ulcers	33%
• Urinary incontinence	34%
• Bowel incontinence	13%
• UTI	69%
• Hyperhidrosis	15%
• Dysreflexia	50%
• Spasticity	44%
• Latex allergy	10.5%

Long-term Outcomes of Pediatric SCI

- Shoulder pain 59%
- Wrist pain 27%
- Elbow pain 19.5%
- Pain other sites 51.5%
- Fractures 5%

Long-term Outcomes of Pediatric SCI

• Hypertension/heart disease	6%
• Chronic medical conditions	22%
• Hospitalizations past year	22%
• Moderate-severe depression	8%
• Substance abuse	14%

	SWLS	CHART	Employment
Pressure ulcers	+	+	+
Spasms	+	+	+
Shoulder pain	+	+	+
Incontinence B/B	+	+	
UTI	+	+	
Dysreflexia	+	+	
Substance abuse	+	+	
Elbow pain	+		+
Wrist pain			+
Pain any site	+		
Hospitalizations		+	

	SWLS	CHART total	Employment
Marriage	+	+	+
SF 12 Physical	+	+	
SF 12 Mental	+	+	
Gender	+		
Race		+	
Income		+	
ASIA motor score		+	
Employment		+	
CHART total			+

Implications

- Medical complications are significantly associated with key outcomes of adults with pediatric onset SCI
- Therefore, preventing medical complications may improve outcomes

Implications

Establish expectations throughout the lifespan

- Healthy
- Independent
- Employed
- Participate fully in their communities
- Satisfying lives

Relationship between parenting & demographic and injury related factors

	Parent n=98	Not a Parent n=334
Female ***	66%	28%
Paraplegia*	54%	42%
Age @ follow-up	33.9 (5.8)	32.5 (5.9)

*p<.05, *** p<.001

Relationship between parenting & outcomes

	Parent n=98	Not a parent n=334
SWL***	27.7 (5.8)	23.0 (7.9)
CHART physical independence*	93.9 (9.7)	91. (11.9)
CHART mobility*	92.2 (12.4)	86.8 (19.5)
CHART social integration**	94.9 (10.0)	88.9 (16.7)
CHART Occupation***	90.8 (21.9)	70.6 (32.2)

*p<.05, ** p<.01, *** p<.001

The challenge in caring for children with SCI

- Prevent complications during childhood
- Because of long life span of individuals with pediatric SCI
  - Identify those at risk of complications
  - Develop strategies to prevent complications throughout their lifespan

Prevention of pressure ulcers

- Need to shift responsibilities from parent to youth
- Smoking prevention
- Nutrition
- Seating systems
  - Need to change with size of patient and their needs

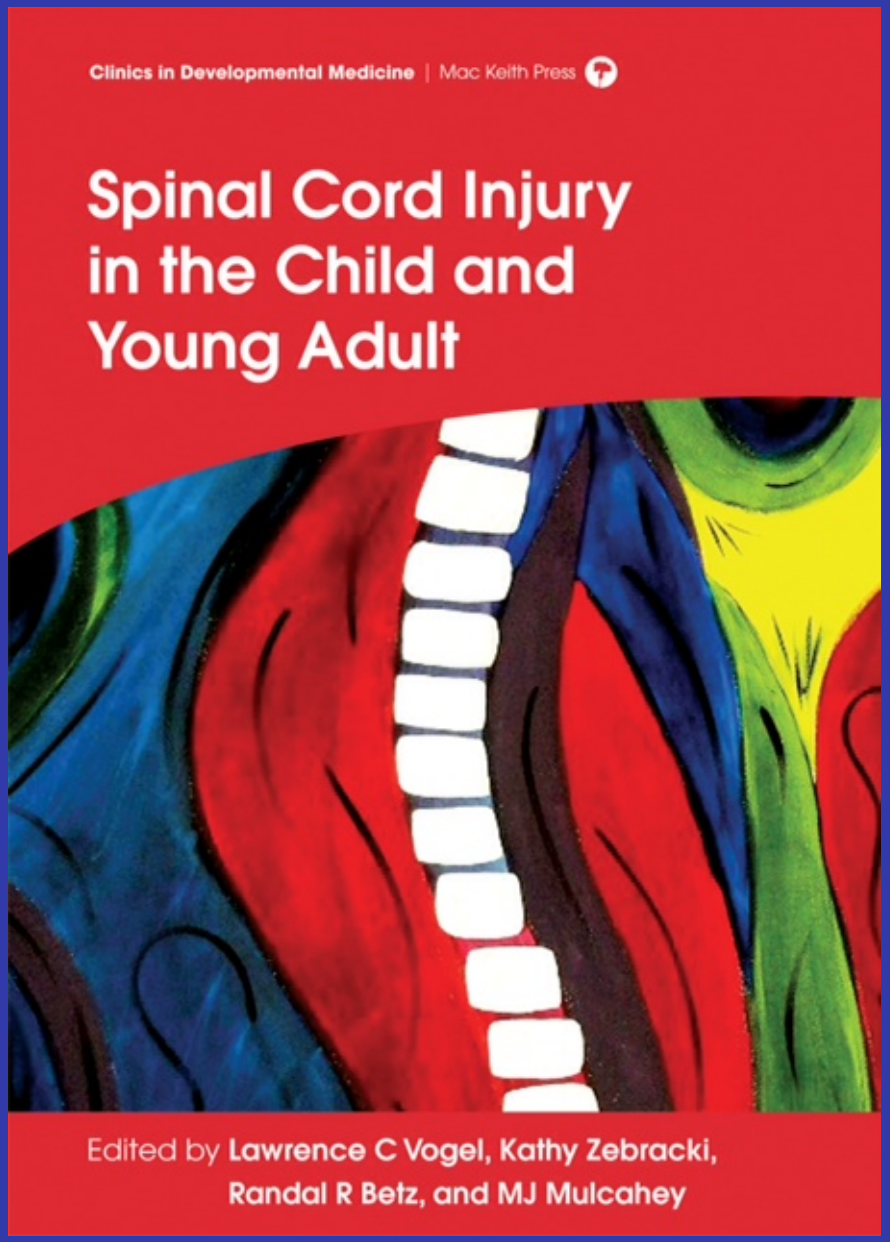
Prevention of upper extremity pain

- Critical role of wheelchairs
- Developmental implications
  - changing size of wheelchairs
  - different needs
  - assessing and teaching proper propulsion
- Power versus manual
  - How to save those shoulders for 60+ years in a 10 year older

Assure adequate preparation to ensure quality in adult life

The Future

- Prevention of SCI
- Wheelchair design
- Pressure ulcer prevention
- Implantable FES systems
- Robotics
- Brain Computer Interface
- Cure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