Cerebral Palsy throughout the Lifespan: A View from Both Sides

Hank Chambers, MD
David H Sutherland Chair of Cerebral Palsy Studies
Rady Children's Hospital San Diego
Professor of Clinical Orthopedic Surgery
University of California at San Diego

The secret to creativity is knowing how to hide your sources

What Is Cerebral Palsy?

- Is it brain damage due to obstetrical trauma?
- Was the baby too big or too small?
- Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, behavior, by epilepsy and by secondary musculoskeletal problems

Modified after Bax et al. DMCN 2005

Epidemiology: The Cerebral Palsies

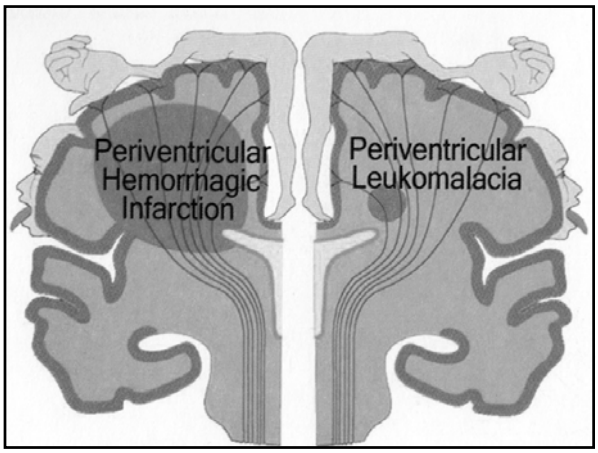
- Incidence: 3-4 patients/1000 live births (1 in 278)
  - 9000 new diagnoses each year
- Risk is 25-30 times in neonates < 1500g
  - 1 in 3 children with VLBW will have CP
- 87% 30-year survival rate
- Prevalence: ~765,000 Americans

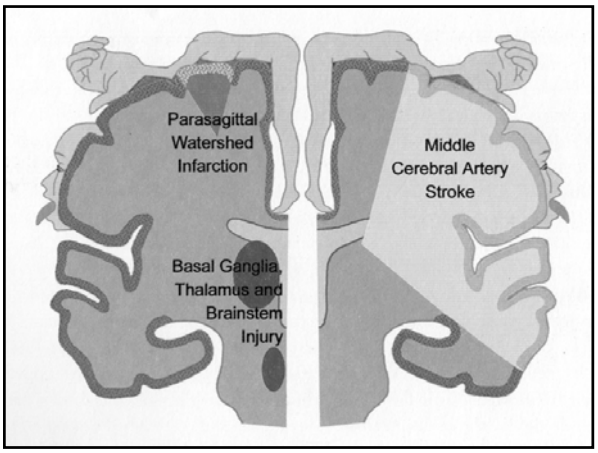
Some Statistics

- 1 in 3 very low birthweight children will have CP
- 72 percent of unemployed adults with disabilities would like to work
- 9000 infants are diagnosed each year
- 54 million americans have a disability
- 28 percent didn't have health insurance
- 29 percent work full time
- 72 percent would like to work
- 6 million children in special ed

Etiology of Cerebral Palsy

- ✶Prematurity
- ✶Chromosomal and Brain Abnormalities
- ✶Genetic Influences
- ✶Metabolic Influences
  - ✶Hormonal
  - ✶Heat
  - ✶Inflammation
- ✶Hemostatic Disorders
- ✶Infection
- ✶Trauma





Career Choices

- United Cerebral Palsy Telethon
- Pediatric Rotation
- Birth of my son, Sean in 1982 while I was an intern

Classification

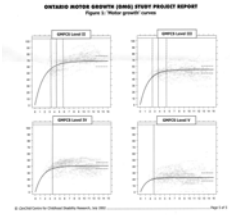
- Gross Motor Function Test
- Gross Motor Function Classification System (GMFCS)
- Functional Mobility Scale
- Gillette Gait Index
- Functional Assessment Test

Classification Systems

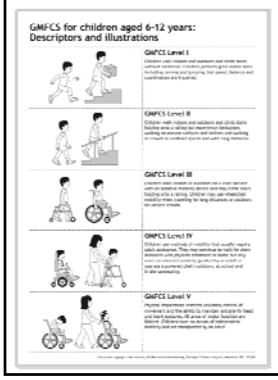
- The diplegia, quadriplegia, hemiplegia system has poor intra and interobserver reliability
- Levels of ambulation: household, therapy, community also has limitations
- Gross Motor Functional Classification System (GMFCS)

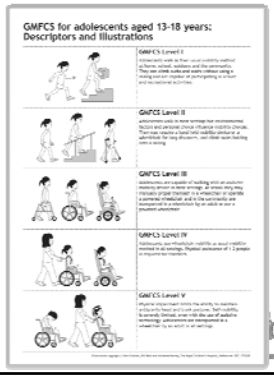
GMFM

- Series of tests given to ascertain the level of gross motor involvement in children with cerebral palsy.



GMFCS


GMFCS for children aged 6-12 years:
Descriptors and illustrations


GMFCS for adolescents aged 13-18 years:
Descriptors and illustrations

GMFCS and Musculoskeletal Issues

1. Hip displacement: incidence, type
2. Success of hip surgery: STR vs VDROs
3. Mortality & Morbidity
4. Contracture and bony deformity
5. Success of Gait Correction Surgery
6. Choice of procedures: Rectus Femoris Transfer, varus foot surgery

Functional Mobility Scale

Function at 5, 50 and 500 meters

Dimensions of Disability

- International Classification of Functioning, Disability and Health (ICF) WHO
  - Body Functions
  - Body Structures
  - Activities and Participation
  - Environmental Factors

The NCMRR Model of Disablement

National Center for Medical Rehabilitation Research. Bethesda, MD.

Treatment Paradigms

- Goal Setting
- Team Approach
- Management of Movement Disorders
- Physical Therapy
- Timing of Orthopedic Surgery
- Bony and Soft Tissue Surgery

First We Must Set Goals

Independence
Working
Communication
Activities of Daily Living
Mobility
Walking

Therapies

- Occupational
- Speech and Language
- Management of Drooling
- Visual Impairment

Physical Therapy

- Neurodevelopmental Therapy
- Hippotherapy
- Equipment

Speech Therapy

Communication Devices

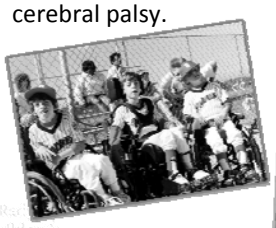
- **Simple**
- **Computer**

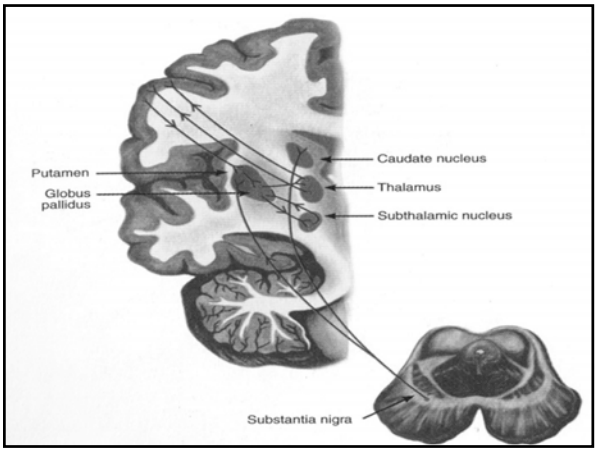
Mobility: Wheelchairs and Seating Systems

Role of Standing

Spasticity vs Dystonia

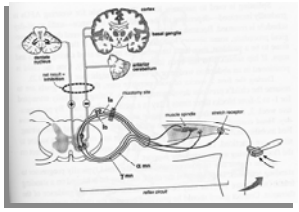
- New understandings of the definitions and therefore the natural history of children with cerebral palsy.



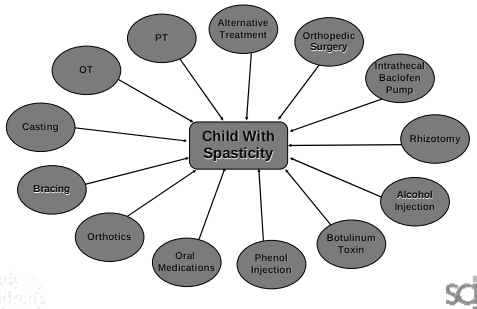


Motor Disorders

- Spasticity
- Athetosis
- Ataxia
- Dystonia



Integrated Treatment Approach in the Child with Spasticity



Other Important Problems

- Loss of Selective Motor Control
- Impaired Balance and Coordination
  - Ataxia
- Sensory Deficits
- Weakness
- Visual Disturbances

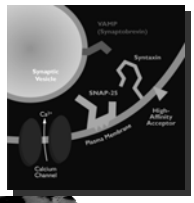
Current Treatment Options: General

- Exercise and physical modalities
- Systemic drugs
  - Valium
  - Baclofen
  - Artane
  - Etc.

Medical Management of Cerebral Palsy

- Anti seizure medications
- Antispasticity medications
- Antidystonia medications
- Management of Reflux
- Management of other GI issues such as Gall stones, constipation, dumping after bowel surgery

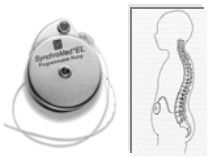
- Anesthetic and neurolytic injections
  - Phenol
  - Alcohol
- Chemodenervation injections
  - Botulinum Toxin A, B



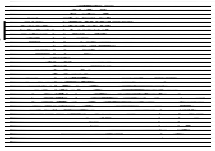
Orthopedic and Neurosurgical Methods

- Tendon lengthenings altering the muscle receptors
- Osteotomies
  - Lever Arm Syndrome
- Neurotomies
- Fusion especially spinal fusion stabilizes the trunk

- Intrathecal drugs
  - Intrathecal Baclofen



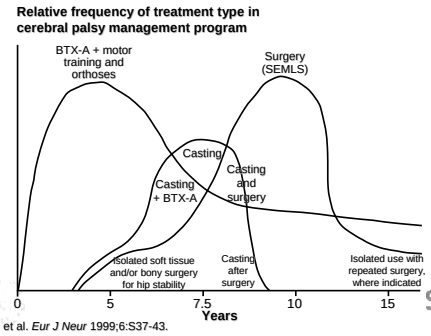
Selective Dorsal Rhizotomy



Principles of Orthopedic Surgery

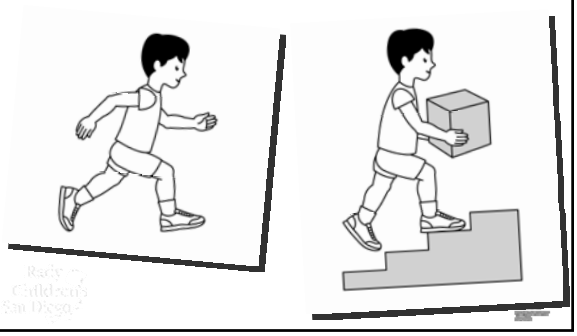
- Single event, multilevel surgery
- Delay surgery as long as possible (> 6 years)
- Use spasticity management as adjunct to surgery

Timing of Orthopedic Surgical Interventions



Boyd, et al. *Eur J Neur* 1999;6:S37-43.

GMFCS Level 1



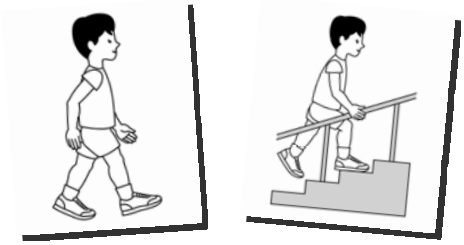
GMFCS Level 1

- High level physical functioning: spastic hemiplegia, mild spastic diplegia
- Seizures, occasionally
- Learning difficulties
- Behavioral problems
- Autistic spectrum disorders

GMFCS Level I

- Mild gait dysfunction
- Many benefit from Botox
- Few need any orthopaedic surgery
- Too mild for SDR or ITB
- No hip displacement, no scoliosis
- UL Surgery in Hemiplegia

GMFCS Level II



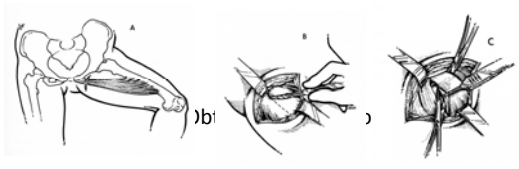
GMFCS Level II

- Mostly spastic diplegia of prematurity
- Some have severe hemiplegia
- Wide range of gait dysfunction
- Significant spasticity
- Significant deformities
- Mild hip disease, no scoliosis

GMFCS Level II

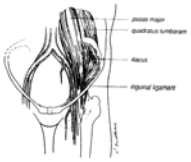
- Botox very useful
- SDR: a very few , highly selected cases
- No ITB
- Single level orthopaedic surgery: UL & LL
- Hip screening and preventative surgery
- SEMLS: Multilevel surgery

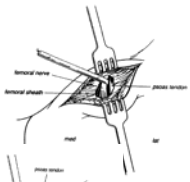
Adductor Lengthening

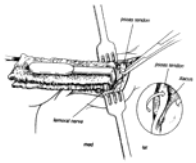


Canale and Beaty
Operative Pediatric Orthopedics

Psoas lengthening at the Pelvic Brim

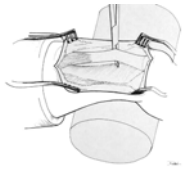




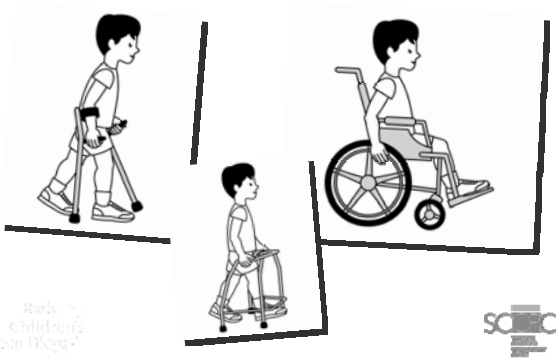


Sutherland and Chambers, JPO 1993

Rectus Femoris Transfer



GMFCS Level III



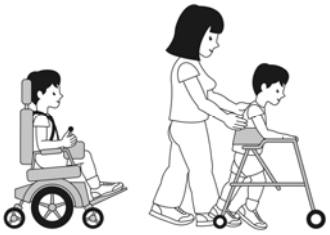
GMFCS Level III

- Severe diplegia, mild quadriplegia
- Spastic-dystonia
- Botox + Phenol are useful, some ITB
- Hip displacement common & important
- Screen and prevent hip displacement
- Gait correction surgery: hips and feet

Treatment of Lever Arm Syndrome

- Femoral Osteotomies
  - Proximal
  - Distal
- ? Gluteus Medius transfer
- Tibial rotational osteotomy
- Correction of foot valgus

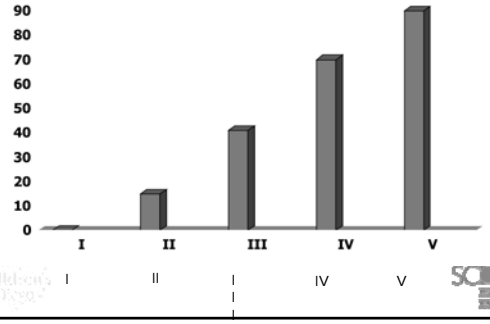
GMFCS Level IV



GMFCS Level IV

- Spastic quadriplegia: mild-moderate
- Spastic-dystonia
- Botox and ITB
- Hip displacement and scoliosis
- Screen and prevent hip displacement
- Orthopaedic surgery for standing, sitting
- May need hip and knee surgery

Hip Displacement (MP>30%) by GMFCS. Soo et al JBJS(A) Jan 2006



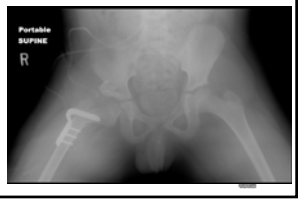
Hip Subluxation & Dislocation

Proximal Femoral Varus Derotational Osteotomies

Indications for surgery:

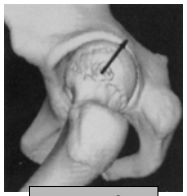
'd valgus

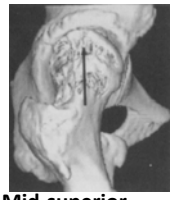
'd femoral anteversion



Acetabular Dysplasia

- Usually posterior dysplasia
- Should assess with CT Scan



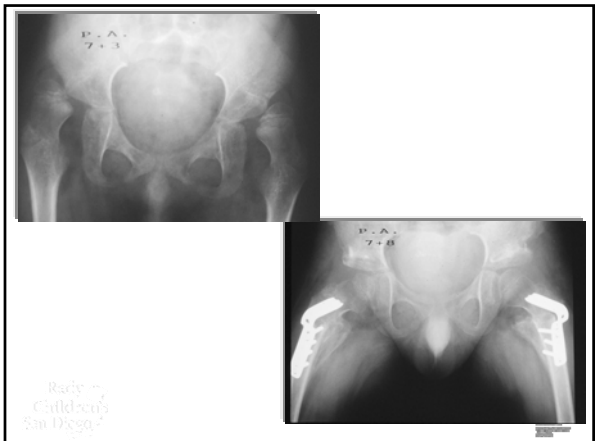


Anterior 29%

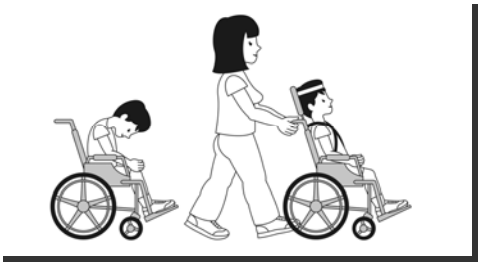
Posterior 37%

Mid-superior 15%

Kim and Wenger JPO 1997



GMFCS Level V



GMFCS Level V

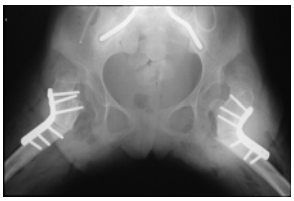
- Spastic quadriplegia
- Multiple medical co-morbidities
- Significant excess mortality in each decade
- Dystonia, spasticity: Botox, phenol, ITB
- 90% will develop hip disease and scoliosis
- Comfortable sitting

GMFCS Level V

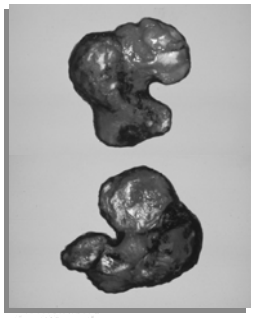
- Optimizing health
- Minimizing co-morbidities
- Goal setting
- Hip and spine surveillance
- Preventative, reconstructive surgery
- Child and care giver quality of life

VALGUS OSTEOTOMY

- Advantages
  - Increased motion
  - Pain relief
- Disadvantages
  - May be painful



PROXIMAL FEMORAL RESECTION



TOTAL HIP REPLACEMENT

- Advantages
  - Pain relief
  - Motion
- Disadvantages
  - Re-dislocation
  - Difficult
  - Infection

Botulinum Toxin for Pain in Dislocated Hips

- Current study at Rady Children's Hospital
- 16 patients with painful dislocated hips or previously surgically treated hips
- 400 Units of Botox in muscles about the hip (16 separate sites)
- Marked improvement in pain in 90%
- Must be repeated every 4-5 months.

Fuse Joints for Stability

Orthopedic Surgical Interventions: Spine

- Intervention to correct
  - Scoliosis
  - Spondylolisthesis
  - Hyperkyphosis
  - Hyperlordosis

Orthopedic Surgeries popularized from San Diego

- Iliopsoas Lengthening at the pelvic brim
- Rectus femoris transfer
- Pelvic Osteotomies
- Femoral Osteotomies
- Calcaneal Cuboid and Cuneiform Ostetomies
- Spinal Fusion Techniques

Unconventional or Alternative Treatments

- Hyperbaric Oxygen
- Adeli Suit
- Biofeedback
- Conductive Education
- Facilitated Communication
- Doman-Delicato
- *Many, many more*

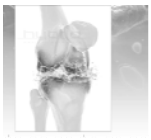
Caregiver Stress

- Increased physical strain
  - Higher incidence of back pain
- Increased mental strain
- More time to care for child, including feeding
- Increased marital stress
  - 85% divorce rate

Transition Issues

Adult Clinic

- 2200 Patients treated in last 15 years.
- Multitude of new problems including:
  - Increased pain
  - Arthritis
  - Difficulty sitting
  - Progression of movement disorder
  - Bipolar disease (45% of all my adult patients are on antidepressants)
  - Loss of ambulation
  - Cervical Spine Problems
  - Progressive Hydrocephalus



Musculoskeletal Problems of Aging

- Progressive weakness secondary to athetoid movements of the neck with cervical spondylosis
- Arthritic changes in joints which are not biomechanically sound because of malpositioning or abnormal stresses through a decreased range of motion.
- Muscle wasting of aging
- Poorer soft tissue coverage leading to skin ulcers and shear injuries

Chronic Pain

Psychosocial issues

- 75% are independent in ADL
- 30% live at home with their parents
- High incidence of depression and low self-esteem
- 65% of my adult patients are on antidepressant medication

Transition from Home to Community Living

- Home Care
- Residential Facilities
- Community Based Programs
- Intermediate Care Facilities
- Community Homes
- Supported Living
- Independent Living

Transition from School to Work

- Public Schools
- College
- Vocational Training

Economic Issues

- 60-70% unemployment
- Most receive government stipends

Work Options

- Competitive Employment
- Supported Employment
- Day Programs and Sheltered Workshops

Transition from Pediatric to Adult Health Care

- Health Care Insurance
- Mental Health Issues

Life's Goals

- Real Choices in all aspects of life
- Functional Skills
- Interaction with a variety of people
- Inclusion in generic services and activities
- Access to community resources
- Use of a range of community environments
- Living in typical neighborhood
- Meaningful daily activity
- Nonadversarial interactions
- Relationships with friends

Recreation, Leisure and Socialization

Issues of Sexuality and Self-Esteem

Competency as an Adult

A Global Call to Arms

- The aging population of people with disabilities is growing rapidly
- We know little of the natural history of each disability
- Research needs to be done in this area
- Education of medical students and other healthcare professionals needs to be instituted immediately
- Governmental payors and insurors need to be part of the solution: excellent role for health maintenance (HMO) approach

Solutions and Models

San Francisco Improving Transition Outcomes Project (ITOP) 2009

- Provider Competency
- Access to Quality Health Care
- Patient Support
- Quality Assurance and Oversight
- Public Engagement and Support

Provider Competence

- Train and mentor (clinical knowledge, cultural competency and communication skills) health care providers and staff
- Develop and implement use of electronic health records
- Remove physical and programmatic barriers (larger exam rooms, large print patient information, longer appointment times)
- Provide access to specialized health care providers and specialist consultation for primary care providers

Access to Quality Health Care

- Provide access to information about where to go for health services
- Finance/reimburse health care services adequately
- Provide access to transportation
- Limit the size of patient load for clinicians (100-300 patients per FTE)
- Provide health care in natural settings (e.g. home, school, community agency, neighborhood clinic

Patient Support

- Educate patients, health care and service providers, as well as family members and caregivers, of patient care rights and responsibilities, including support for self-determination
- Designate appropriate surrogate decision makers (i.e. informed consent, confidentiality, restraint)
- Proved training and resources for caregivers
- Identify an individual health care plan team member responsible for monitoring of the health care plan and ensuring implementation of the plan.

Quality Assurance

- Define health care outcomes and quality standards clearly
- Provide access to data necessary for evaluating quality of care and health care outcomes
- Provide access to information and research regarding promising models and practices
- Provide clear oversight and accountability for ensuring health care access and outcomes
- Loop back to provider training and curriculum

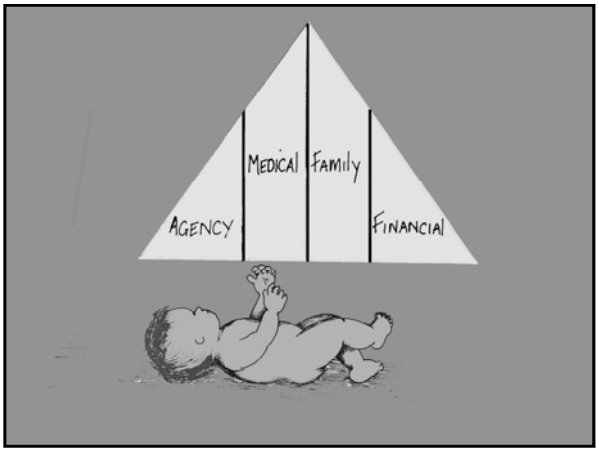
Public Engagement and Support

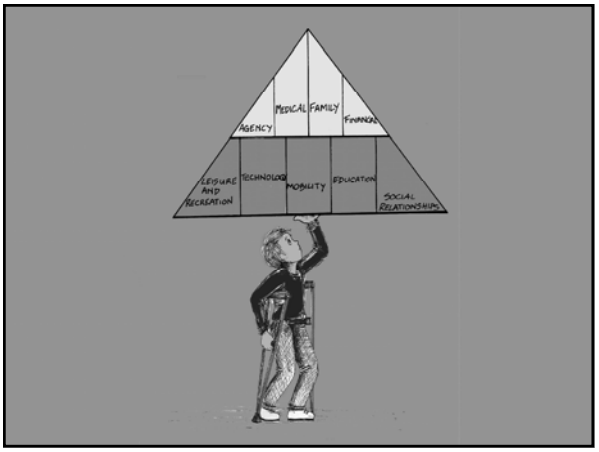
- Develop a shared vision and sense of responsibility
- Identify and engage a champion/leadership
- Educate policymakers and public officials

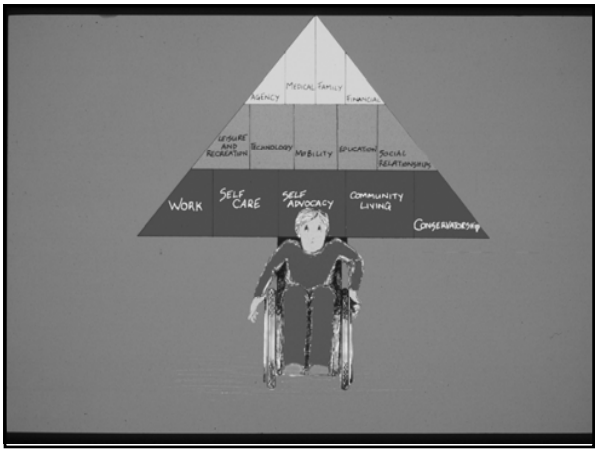
Current and Proposed Models

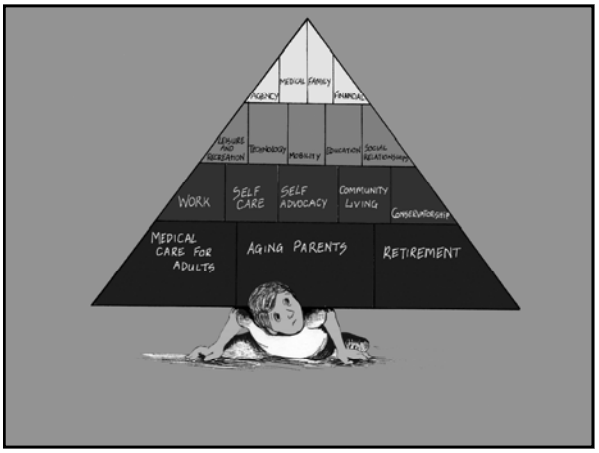
- Educate Primary Care physicians and have them work with pediatric subspecialists
- University models where transition is planned and adult providers take over care (specialty model)
- Lifespan physicians who care for the child and then adult with the disability

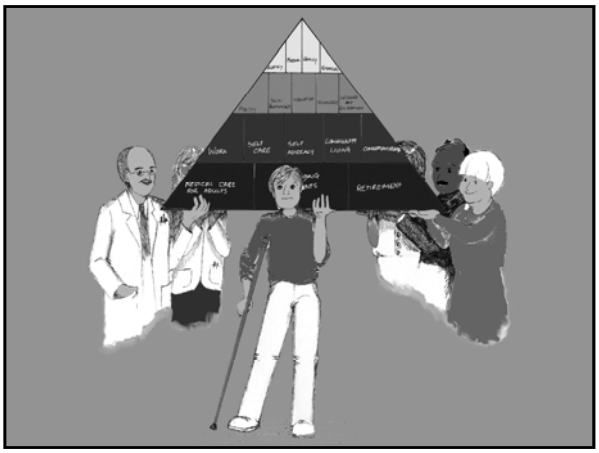
Transitioning through the Stages of Life and Disability











So, What have I learned in 27 years

- Parents are always seeking a cure for their child
  - Hyperbaric Oxygen
  - Stem Cells
  - ?????? And $$$$$$
- Simple insights lead to great changes in care
  - Definition of dystonia
  - GMFCS
- Treatment is important, but prevention is the real hope
- There are true heroes who have no vested interest other than the care of children who have dedicated their careers to the understanding and treatment of this disorder

Thank you