LIFE CARE PLANNING FOR CHILDREN WITH CEREBRAL PALSY

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WHAT IS CP?

- Cerebral palsy is a general term and not a specific disease
- 4 criteria for the dx of CP
  - Brain injury (prematurity with bleeding, anoxia, malformations, encephalitis, genetic, etc.)
  - Non-progressive (NOT leukodystrophies or other progressive brain disorders)
  - Onset before the age of 2 (early developmental period)
  - Affects physical function (most often have other associated problems, but only physical part of dx)
- Although non-progressive encephalopathy is part of dx, the impairments can change over time
- Associated problems (cognitive, GI, resp., vision, bowel and bladder, etc.)

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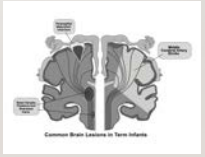
TYPES OF CP

- Diplegia
  - Involves legs more than arms
  - Usually from prematurity
  - Ambulatory
- Hemiplegia
  - Involves one side
  - Usually a term stroke
  - Ambulatory
- Quadriplegia
  - Involves all 4 limbs significantly
  - Various severe encephalopathies
  - More severe systemic problems

Common Brain Lesions in Preterm Infants



Common Brain Lesions in Term Infants

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GMFCS LEVELS

- Levels associated with severity and also related to associated impairments and care needs
- I
  - Fully ambulatory w/o aides (may use braces), can run and jump, can navigate curbs w/o a rail
- II
  - Ambulates w/o aides, difficulty with running, jumping, steps w/o a rail
- III
  - Ambulates with aides (crutches, canes, or walker) for household distances
- IV
  - Mobilizes self with manual or power W/C
- V
  - Cannot mobilize self even with W/C

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GMFCS LEVELS

GMFCS I

GMFCS II

GMFCS III

GMFCS IV

GMFCS IV

GMFCS IV

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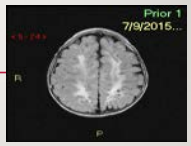
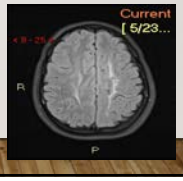
PERIVENTRICULAR LEUKOMALACIA (PVL)





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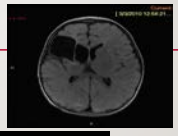
HEMIPLEGIC CP



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LIFE EXPECTANCY

- Huge financial issue but not the typical life care planner's fight
- A few good articles that simplifies calculations
  - Day, et al DMCN 2015
  - Brooks, et al DMCN 2014 part I & II
  - Crichton, et al DMCN 1995
  - Katz NeuroRehab 2003
- Gross motor function closely related to life expectation
- Cognition and ability to socially interact a big indicator
  - Non-ambulatory, good cognition: 50 y/o; non-amb, severe cognition deficits: 30 y/o

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MEDS

- Seizure meds
- Pain meds
- Spasticity
- Drooling
- Constipation
- Other

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HYPERTONIA

- Spasticity – velocity dependent, pyramidal tracts (spinal cord, cortex, periventricular)
- Dystonia – variable posturing and hyperkinetic movement, extrapyramidal (basal ganglia)
- Causes pain
- Pain causes increased spasticity
- 77% report problems
- 80-90% have contractures

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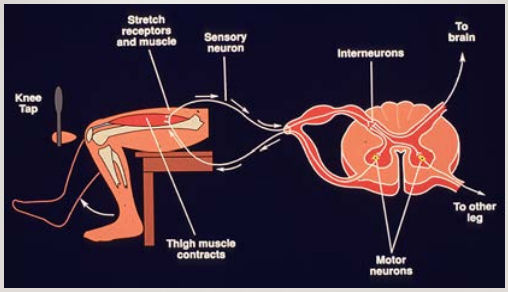

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SPASTICITY MEDS

- Baclofen
- Diazepam
- Tizanidine
- Dantrium
- CBD minimal effect

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TREATMENT: HYPERTONIA – INJECTIONS

- Botox/phenol
- focal treatment
- focal effect may generalize
- Botox lasts 4-6 months
- phenol lasts 6-12 months (may be shorter)

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SPASTICITY MANAGEMENT: MEDS

- Oral baclofen
- Botulinum toxin
  - Botox Adult dosing 400 units, pediatric 12 U/kg
  - Dysport adult 1,200 U, pediatric 30 U/kg max 1,000
  - Every 3-8 months until adult, then every 6-12 months afterwards
  - Sedation with nitrous oxide, OR, or sedation in clinic common
- Phenol neurolysis
  - Every 6 months until adult, then every 1-2 years afterwards
  - Requires OR usually
- Baclofen
  - Oral v. ITB pumps
  - ITB refills every 3-6 months

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PRE- & POST PHENOL/BOTOX

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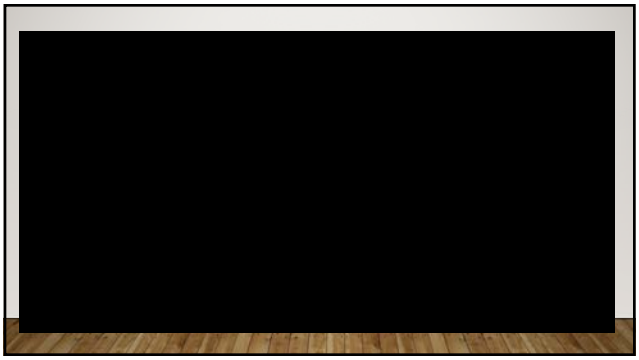

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ORTHOPEDIC SURGERY

- Good for bony deformities
- Helps prevent hip dislocation
- Optimizes base or support and joint position
- Doesn't treat spasticity long-term
- Limited effect for dystonic posturing

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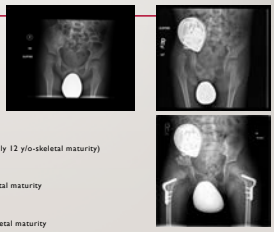
DIAGNOSTIC/EDUCATIONAL TESTING

- Hip x-rays
  - Schedule depends on GMFCS level
  - Frequency can change to yearly if progressing
  - AACPDM guidelines
- GMFCS I
  - Routine x-rays not needed
- GMFCS II & hemiplegic severe
  - 1st x-ray at 2 y/o, then 6 and 10 y/o (severe hemiplegia biannually 12 y/o-skeletal maturity)
- GMFCS III
  - 1st x-ray at 2 y/o, annually until 8 y/o then biannually until skeletal maturity
- GMFCS IV & V
  - 1st x-ray at 2 y/o, annually until 12 y/o then biannually until skeletal maturity



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ORTHO SURGERIES

- SEMLS
  - Femoral derotational osteotomies
  - Gastroc recessions
  - Hamstring lengthening
  - Tibial derotational osteotomy
- Risks
  - GMFCS I: 20%
  - GMFCS II: 40%
  - GMFCS III: 50%
  - GMFCS IV: 75%
  - GMFCS V: 90%

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PRE- & POST-ORTHO

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DIAGNOSTIC/EDUCATIONAL TESTING

- Scoliosis
  - >20° brace, >40° surgery
  - Associated with GMFCS level
  - Rare before 8 y/o
  - If has spine fusion mean age 14 y/o
  - Get x-ray if noted on exam then annually until >20° then semiannually
  - MAGEC rods (magnetic growing rods) preserves growth, <10 y/o, fusion later
- Risk of >40°
  - GMFCS I-II: near 0%, III: 10%, IV: 50%, V: 75%

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TREATMENT: SPASTICITY- SURGERY

- selective dorsal rhizotomy
  - selectively cuts lumbar sensory roots
  - affects spasticity, but not dystonia
- Ideal candidates
  - Spastic diplegia
  - PVL from prematurity
  - Ages 3-8
  - GMFCS I – III
  - Bright, good social support

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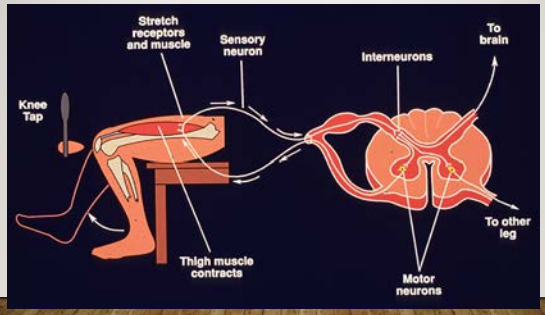

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PRE- & POST-SDR/ORTHO SURG

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SDR

Pre-SDR

1 yr post-SDR

2 yr post-SDR

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with SDR

without SDR

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TREATMENT: HYPERTONIA – SURGERY

- intrathecal baclofen pump (ITB)
  - treats spasticity and dystonia
  - catheter tip from lower thoracic to ventricles
  - Refill every 3-6 months
  - Change pump every 5-7 years

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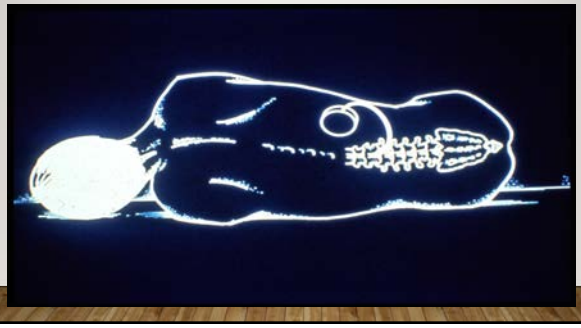

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TREATMENT: DYSTONIA – ORAL RX

- mostly trial and error
- oral medications may improve tone some but not resolve hypertonia
- "cocktails" common
- side-effects high
- treat symptomatically

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TREATMENT: DYSTONIA – ORAL RX

- trihexyphenidyl (Artane), anticholinergic
- risperidone (Risperdal), antipsychotic
- tetrabenazine (Xenazine), antichorea
- L–dopa (Sinemet), dopaminergic
- baclofen (Lioresal), GABA agonist
- diazepam (Valium), GABA agonist
- clonazepam (Klonopin), GABA agonist
- cannabis?

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TREATMENT: DYSTONIA – SURGERY

- Deep Brain Stimulator
  - suppresses irregular overactive neuronal patterns, stimulates inhibitory pathways
  - electrodes in basal ganglia
  - 10% caudate strokes
  - 10% infection rate
  - mixed results with secondary dystonias

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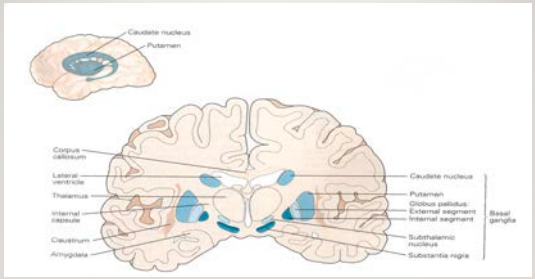

Kandel, Schwartz, and Jessell 2000

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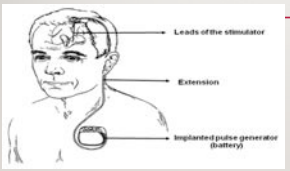
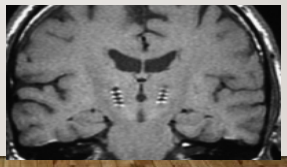
DEEP BRAIN STIMULATOR





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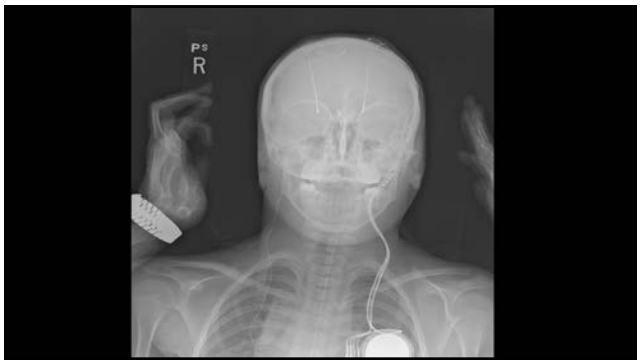

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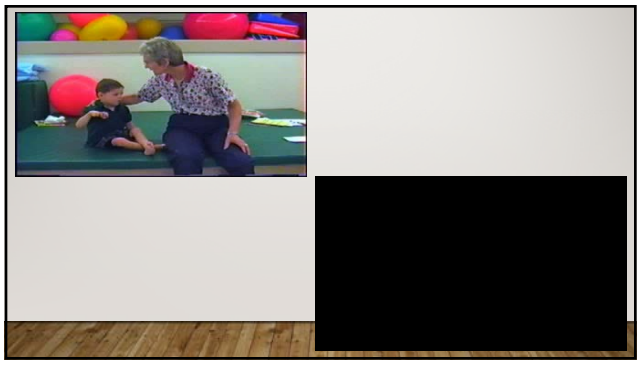

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VENTRAL DORSAL RHIZOTOMY

- Usually for severe intractable hypertonia with limited functional motor skills
- Both a ventral (motor) and dorsal (sensory) rhizotomy
- Non-selective
- Lumbar and cervical
- About 50% dorsal rootlets and 50-70% ventral rootlets

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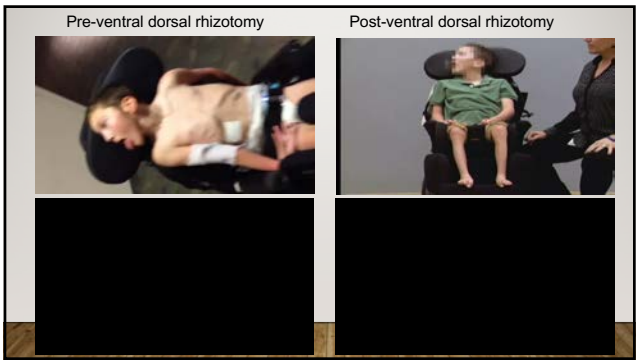

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DIAGNOSTIC/EDUCATIONAL TESTING

- Neuropsychological testing
  - 1x middle school, 1 high school, 1 young adulthood
- Bone density testing
  - Provider and results dependent

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DIAGNOSTIC/EDUCATIONAL TESTING

- Gait lab assessments 2-4 in ambulatory patients
- Seizures - 30% risk
  - Risk factors: neonatal sz, low APGAR, MR
  - Usually , 2 y/o
  - Hemiplegic: 60%, quad: 50%, diplegic: 20%, dystonic: 20%
  - EEGs
    - For those with seizures
    - One-hour v. 48 hour
- MRI
  - Symptom dependent: treater input needed, usually at least 1 in lifetime
- Hydrocephalus
  - Usually within first 2 years, 13% risk overall, if have VPS 85% need revision, ave. 3 VPS revisions
- Labs
  - Vitamin D, others

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FUTURE MEDICAL CARE ROUTINE

- Primary care
  - complex care
- PM&R
- Ortho
- Neuro
- Pulmonology
- Palliative care
- Sleep medicine
- Numerous others

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PROJECTED THERAPEUTIC MODALITIES

- Eval v. treat
  - Eval complexities
- Units of treatment
  - CPT codes
- Episodes of care
- Lifelong treatment v. evaluation
- Psychology services/behavior analyst
- Audiology
- Nutrition evaluations 2-4/year in growth years

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FUTURE MEDICAL CARE NON-ROUTINE

- ER, urgent care
- Hospital days
- Dental procedures in the OR
  - 1 per year or every 2 years

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WHEELCHAIRS

- Pediatric v. adult replacement
- Stroller
- Manual
  - tilt in space
- Power
  - Attendant control
- Pricing for wheelchairs
- Seating: custom v. off the shelf
- Internet pricing such as Spin life etc.
- W/c maintenance costs

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DME

- How things are really ordered when there is a therapy evaluation
- Shower chairs
- Standers and gait trainers
- Beds
- Toilet equipment
- Positioning equipment
- Grasshopper system, versaform pillows
- Feeding chairs
- Lifts

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ORTHOTICS

- Trunk orthosis
  - SPIO has 3 levels of support with different stays
- Thumb splints
- Hand splints
- Elbow splints
- Knee immobilizers
  - Needed with stander typically
- AFOs
  - 1 pair per year in growth years, start with stander use, may start with SMO to allow for functional improvements
  - 1 pair every 3-5 years after growth years
  - AFO repairs

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AIDS FOR INDEPENDENT FUNCTION

- AAC, wheelchair mount
- Switches
- Environmental control
- Adapted eating utensils
- Adapted writing aids

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SUPPLIES

- Toilet training
  - Typical children age 3
  - GMFC Level and intellectual ability strongly associated with continence
    - GMFCS scale of IV or V, severe impairment of motor and communication skills, dyskinesia or combined spasticity and dystonia negative predictors
    - For children with severe impairment likelihood of continence after age 8 is very slim
  - Those who achieve continence typically do so more slowly and less completely than controls
  - 88% of children with GMFCS I and II are day and night continent by age 5.5 years

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HOME CARE

- Home care
  - Childhood
    - School hours, sleep, parental responsibility
  - Adulthood
- Level of care: PCA, HHA, LPN, RN
  - Family provided care
- Case manager
- Group home
- Day program, supported employment
- Heavy housecleaning in adulthood

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HOME MODIFICATIONS

- Home accessibility specialists v. OT v. contractors
  - Rental v. ownership history
- Costs
- Ability to use kitchen
- Supply and equipment storage
- Caregiver space

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TRANSPORTATION

- AAA or equivalent
- Car seats
- Portable ramps
- Van accessibility features
- Maintenance and insurance
- Medical mileage/parking

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FITNESS

- Critical for successful aging
- Health club v. home gym
- Personal trainer v. PT
- Equipment
  - Adapted bikes
  - Adapted swings
  - Raised exercise mat

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CONSERVATOR

- Cost depends on amount of funds being managed
- Typically start at age 21

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TYPICAL AGING

- Maximum biological/physical function at age 18-25
  - In general 1% decline/year after this age (Adkins, 2004)
  - At age 70 functioning at 45% of reserve capacity
  - At age 80 functioning at 35% of reserve capacity
- Maximum psychological/cognitive function at age 35-45
- Maximum social function at age 50-60

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AGING WITH CP: PAIN AND CP

- Musculoskeletal pain in the 30's & 40's (Andersson et al, 2001, Arcand et al, 2000, Crawford, 1996, Murphy 1999, Murphy et al, 1995, Rapp et al, 2000, Report from the Roundtable… 1997)
- Less severe CP/more pain
- Back and legs most common
- Knees and hips by age 40
- Hands, wrists, neck
- Spasticity can increase pain and vice versa

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CP/AMBULATION

- Most pain in the group that walks (Turk et al, 1997)
- 40% fall 1x/month, 75% fall every two months (average age of 44) Kemp & Mosequeda, 2004/Rancho Los Amigos
- Some stop walking in their 20's because of efficiency
- Some stop in 40's because of pain
- Walking is lost to 25%
- For those who continue to walk, speed and distance decrease (Bottos et al, 2001)

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PHYSIOLOGICAL BURNOUT

- Overuse of arms/shoulders (wheelchair/crutches)
- Abnormal postures
- Constant switch use
- Employed and living without assistance most vulnerable
- Ergonomic assessments

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AGING WITH CP

- Most functional = most difficulty later
- Changing assistive technology needs
- Fatigue and pain
- Inactivity
- Loss of strength
- Loss of endurance and balance
- Need for more assistance

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LESSONS LEARNED

- Young parents; email, voicemail, text
  - HIPAA
- PM&R, neurology
- Discounted pricing
- Scope of practice

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BIG LIFE CARE PLANNING ISSUES

- Life expectancy
- Amount of care needed
- Level of care
- Parent provided care
- Group home v. care in the home
- Employability

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THANK YOU FOR ATTENDING

- Questions?

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REFERENCES

- Bladder and bowel continency in bilateral cerebral palsy: A population study (2016) Journal of Pediatric Urology 12(6).
- Risk factors for daytime or combined incontinence in children with cerebral palsy (2017) Journal of Urology May 19.

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