

The child with neuromuscular or muscular dysfunction

Unit XVII – Chapter 32

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Cerebral Palsy (CP)

***Umbrella-like term describing a group of chronic disorders impairing movement and posture**

***Not contagious, nor inherited, no cure, & nonprogressive**

***Most common permanent physical disability of childhood**

***1.5 – 3/1,000 live births in US**

***30-50% are MR, many have normal intelligence**

- Vocational rehab & higher education possible
- Adult: 30% cared for
 - 50% independent

Risk Factors of developing CP

***Prenatal, perinatal & postnatal:**

- Breech presentation, complicated L&D
- Low Apgar score, low birthweight
- premature birth: < 36 weeks or < 2000g, multiple birth
- Intrauterine exposure to maternal infection
- Periventricular leukomalacia
- Kernicterus

***Shaken baby syndrome (SBS)**

4 types of dysfunction: box 32-1

***Spastic (70-80%)**

- Hypertonicity with poor control

***Dyskinetic or athetoid (10-20%)**

- Slow, wormlike, writhing movement

***Ataxic (5-10%-rare)**

- Wide-based gait
- Poor rapid, repetitive movements

***Mixed**

CP etiology

***Acquired (10-20%)**

- Brain damage
- Brain infections
- Head injury

CP etiology

***Congenital**

- Infections during pregnancy
- Jaundice in the infant
- Rh incompatibility
- Severe O₂ shortage in brain or trauma to head during L&D

– Stroke

CP clinical manifestations

- ★ Vary; may change over time
- ★ Difficulty with fine motor
- ★ Balance or walking
- ★ Involuntary movements
- ★ Persistence of primitive reflexes

– Moro reflex

– Crossed extensor reflex

Early signs of CP (Box 32-3)

- ★ Usually appear before 3 years of age
- ★ Parent 1st to suspect
- ★ Delayed in meeting developmental milestones
 - Roll over, sit, crawl, smile, walk
- ★ Abnormal muscle tone
 - Hypotonia or hypertonia
- ★ Unusual posture
- ★ Favor one side of body

CP diagnosis

- ★ Based on clinical findings & medical hx
- ★ Difficult in early months
- ★ Suspicious if:
 - Small for age
 - Hx of prematurity
 - Low Apgar score (0-3 at 5 minutes)
 - Inflammatory, traumatic or anoxic event
 - Abnormal positions
 - Developmental delays

Diagnostic tests

- ★ Dx: MRI
 - Other: hearing, vision, EEG, genetic eval
- ★ Hand preference
- ★ Rule out other disorders
- ★ CT, US
- ★ Intelligence tests
- ★ Ophthalmologist, Otologist

CP mgt

- ★ Early recognition & optimal development
- ★ Individual plan
- ★ Incorporates Health Care Team & various settings
- ★ Preventive & symptomatic

CP Meds:

★ Spasticity

- Baclofen, diazepam, dantrolene: work short term
 - SE's: drowsiness, fatigue, muscle weakness

★ Valium: older child & adolescent

★ Botulinum toxin (Botox)

★ Baclofen infusion pump

- Fewer systemic SE's, reversibility

★ Antiepileptics: Tegretol & Depakote, Dilantin

- Dilantin: gum hyperplasia

CP assessing & dx

★ Assess all children at each health care visit for developmental delays

★ Note any orthopedic, visual, auditory or intellectual deficits

★ Assess newborn reflexes

★ Record dietary intake, ht & wt %

Nursing care of CP

★ Provide adequate nutrition

★ Maintain skin integrity

★ Promote physical mobility

★ Promote safety

★ Promote growth & development

★ Teach parents how to care for child

★ Provide emotional support

Nutrition

★ High-calorie diet or supplements

★ Small amount of food

- Semisolid foods

★ Proper positioning

- Jaw control (fig 32-3 &4)
- Encourage lip & tongue techniques

★ Tube feeds

★ Assistive utensils

Skin integrity

★ Protect bony prominences

★ Maintain proper body alignment

★ Support while in bed or chair

★ Support head & body of floppy infant

★ Prevent drooling

★ Prevent incontinence

Physical mobility

★ PT: prevent contractures

- Braces: Ankle-foot orthoses (AFO's)
- Wheeled scooter boards or go-carts

- ★ **Incorporate play**
- ★ **Teach hygiene**
- ★ **OT & Adaptive appliances**
- ★ **Surgery: contractures or spasticity**
 - Achille's tendon, Hamstrings, wrist, or hip
 - Dorsal root rhizotomy

Ankle-foot orthoses (AFOs)

Safety

- ★ **Safety belts**

- ★ **Helmets**

Promote growth & development

- ★ **Use appropriate terminology**

- ★ **Behavioral therapy**

- ★ **Promote positive self-image**

- ★ **Stimulation programs**

- ★ **Communication methods**

- ST: early
- American sign Language
- Provide audio & visual activities
 - Computer, TV, video & music
 - Special hand controls or pointers, sensors

Foster parental knowledge

- ★ **Teach parents about CP**

- ★ **Teach mx**

- Administration
- Desired effects & SE's
 - Dental care with anticonvulsants

Provide support & coping

- ★ **Counseling if needed**

- Parent groups

- ★ **Listen to concerns & clarification**

- ★ **Encourage expression of feelings & questions**

- ★ **Explain what to expect**

- ★ **Utilize other HC professionals**

- ★ **Siblings: behavioral problems**

Nursing care in the community

- ★ **New adaptive devices, ongoing developmental assessment, ? Surgery**

- ST, routine eye exams

- ★ Assistive technology
- ★ Early intervention programs
- ★ Mild to moderate: attend regular classes
- ★ Finances

Nursing care in the community

- ★ Referrals: UCPA & Shriners Hospitals, other parents
- ★ Recreation or after-school activities: National Disability Sports Alliance
- ★ Transition programs: living, transportation, employment
- ★ Counseling: adolescence
- ★ Vocational training

Spina Bifida = cleft/split spine

- ★ Congenital neural tube defect of spinal column, one or more vertebrae fail to form properly
- ★ CNS & spine develop between 14th & 28th day after conception (1st month)
- ★ US rates declined to 0.3/1000 births
 - after food fortification of folic acid in 1998
 - ^ prenatal dx & pregnancy termination

Cause: unknown

- ★ ?environmental
- ★ Drugs, chemicals
- ★ rx

- ★ Maternal health
- ★ Genetic mutation

Who is at higher risk for SB

Women who:

- ★ Have a child with spina bifida
- ★ Have spina bifida themselves
- ★ Have already had a pregnancy affected by any neural defect
- ★ Pregnancy maternal obesity
- ★ Use of antiepileptic (AED) drugs in pregnancy
- ★ 80-90% have hydrocephalus too

Types of Spina Bifida

- ★ Spina bifida occulta (hidden)
- ★ Spina bifida cystica/manifesta (cyst-like)
 - Meningocele: meninges & spinal fluid
 - Myelomeningocele: meninges, spinal fluid & nerves

Clinical manifestations (box 32-5)

- ★ Muscle weakness or paralysis
- ★ Loss of sensation
- ★ Loss of bowel and bladder control
- ★ Hydrocephalus
 - Learning problems
 - If untx: Brain damage, seizures, blindness

SB: Detection

★Tests to pinpoint bony defect

- CT scan
- ultrasound
- MRI
- Flat films of spinal column
- Myelography
- Labs: I.d. organism

★Prenatal: offered to all women

- Fetal US
- AFP: 16-18 weeks
- CVS: p 10 weeks

Surgery: close & repair

★? fetal surgery

★12-18 hrs to reduce infection & protect spinal cord

★Shunt procedures

★Abx

★Other surgeries done if child is symptomatic

★Assess:

- Family & finances
- long-term specialized school & health care

Orthopedic mgt

★Prevention contractures, correct deformity, prevent skin breakdown, optimal function of LEs

★Common problem areas: knee, hip, feet, spine; Later: kyphosis, scoliosis

★Orthoses: braces, walking devices, wheelchairs

★Infant: passive ROM, position & stretching

★Surgery done early if needed for mobility & ADLs

SB GU mgt:

★Neuropathic (neurogenic) bladder & urinary system distress

★Treatments:

- Regular care & prompt tx of infections
- Regular bladder emptying: CIC
- Mx: Ditropan, Detrol
- Surgery: vesicotomy, urinary diversion

SB Bowel control mgt

★Diet modification

★Regular toilet habits

★Prevent constipation/impaction:

- fiber, laxatives, enemas, suppositories

★Malone antegrade continence enema (MACE)

- <http://pediatrics.aappublications.org/cgi/content/full/109/3/484>

Prevention: folic acid

***No hx of SB: 0.4 mg daily**

***With FH of SB: 4 – 5 mg daily**

- At least 1-3 months prior to conception and through the 12 week of pregnancy (to p breast feeding)

***Additional sources**

- MVI, fortified breakfast cereals
- Foods
 - Dark green leafy vegetables
 - Egg yolks
 - Citrus fruits

SB – care of Sac preoperatively

***Cover w/sterile saline nonadherent dressing: change Q 2-4hrs**

***Incubator or warmer**

***Prone position: hips flexed, legs abducted**

***Assess: infection, leaks, abrasion, irritation**

***Prevent stool contamination: cleanse if soiled or use surgical drape**

***Comfort with tactile stimulation**

SB postop care

***VS, I&O**

***S/Sx of infection: no rectal temps**

***Watch for hydrocephalus or ^ICP, CSF leakage**

***Prone or side-lying position: no pressure on site**

***Feed when alert: turn head to one side**

***Promote mobility:**

- Splints, ROM, Special devices, PT

Home care & discharge: Teach all family members

***Skin:**

- Pad infant: no diapers until full healing
- Urinary retention: CIC
- Special mattress
- Cleansing, lotion, gentle massage

***S/Sx of ^ed ICP, hydrocephalus, shunt infection or malfunction, UTI**

***Affection: cuddle, stroke**

***Home care nurse or long-term care, schooling**

***Resource groups: Spina Bifida Association of America**

SB in the community

***Promote safety & independent mobility**

- Crutches, braces, wheelchairs

***Teach parents & child how to catheterize**

***Surgery**

***Nutrition**

***Emotional & social development**

Monitor secondary conditions associated with SB

- ★ **Latex allergy**
- ★ **Tendonitis**
- ★ **Obesity**
- ★ **Skin breakdown**
- ★ **GI disorders**
- ★ **Learning disabilities**
- ★ **Attaining & retaining mobility**
- ★ **Depression**
- ★ **Social and sexual issues**

Latex allergy

★ **Latex: manufactured from a milky fluid derived from the rubber tree, *Hevea brailiensis***

★ **Reaction if comes in contact with:**

- mucous membrane
- Wet skin
- Bloodstream
- Airway

★ **Diagnostic Labs: RAST**

What contains or is relates to latex reactions?

★ **Catheters**

★ **Elastic bandages**

★ **Baby bottle nipples**

★ **Pacifiers**

★ **balloons**

★ **Food cross reactions to: banana, avocado, kiwi, chestnut, milk**

Latex allergy symptoms

★ **Mild: skin redness, rash, hives, itching, swelling**

★ **Severe anaphylaxis (respiratory sx):**

- runny nose
- Sneezing
- Itchy or watery eyes
- Scratchy throat
- Asthma (difficult breathing, coughing spells, wheezing)
- Shock

Latex allergy

Who's at risk? Anybody

***Frequent exposure to latex**

- Those with SB (80%)

***Health care workers**

***Housekeepers**

***Hairdressers**

***Latex manufacturer workers**

***School nurses**

***Teachers**

***Child care workers, babysitters**

Prevention of latex allergy

***Detect symptoms early & I.D. those at risk**

***Latex free environment**

- Prohibited in food handling
- Labeling “latex-containing”

***Choose powder-free gloves with reduced protein content (vinyl)**

- Do not use oil-based hand creams or lotions
- Wash hands with mild soap & dry

Latex allergy prevention

***Teach/avoid latex containing products (box 32-7)**

***Sx of hypersensitivity**

***Emergency Tx**

- Wear or carry medical identification

***Educate high risk groups**

Spinal Muscular Atrophy (SMA)

***Affects voluntary muscles used for activities; Mainly proximal muscles**

***Weakness in legs > arms**

***Abnormal movements of tongue**

***Normal: senses/feelings, intellectual activity**

Diagnosing SMA

***Genetic analysis**

***Electromyography (EMG) with nerve conduction velocity (NVC)**

***Muscle biopsy**

***Prenatal dx:**

- Survival Motor Neuron (SMN) gene test

Progressive Infantile SMA (Werdnig-Hoffmann Disease): Type I

***Progressive weakness and wasting of skeletal muscle**

***Onset: at birth & before 2 years**

***“floppy infant syndrome”**

***General weakness of intercostal & accessory respiratory muscles**

***Poor prognosis: varies; death usually by 2 years**

- Some may live to 3rd or 4th decade or no progressive loss of strength or function

Werdnig-Hoffman

Werdnig-Hoffman nsg: No cure; Symptomatic and preventive:

***Prevent deformities & contractures : PT & ROM**

- Frequent position changes
- Water therapy
- Powered chairs, lifts, pressure mattress, environmental controls, orthoses

***RT: CPT, suctioning**

- Ventilator assistance
 - Negative pressure or bi-pap
- Tracheotomy
- O2 & pulse ox

***Infection (URI): Abx**

Werdnig-Hoffmann tx

***Prevent aspiration: feed slow**

- NGT or Gastric tube

***Promote stimulation of all senses & independence**

***Social interaction:**

- transport & change of environment
- school

***Parental support & encouragement**

***Genetic counseling**

Juvenile SMA (Kugelberg-Welander Disease): Type III

***Diagnosis p 18mos - adolescence**

***Can stand alone & walk; difficulty with getting up**

***proximal muscle weakness & muscular atrophy; fine tremor**

***Slowly progressive loss of function**

- Lose walking ability: 8 – 30 years

***Good prognosis: Normal life expectancy**

Kugelberg-Welander

Kugelberg-Welander tx:

***Symptomatic & supportive: maintain mobility & prevent complications**

- Walker, bracing, wheelchair, electric scooter
- PT/OT
- Diet

***Scoliosis: custom seating, seating aids, body jacket, spinal fusion**

Muscular Dystrophies (MDs)

***Inherited diseases c/b muscle fiber degeneration & symmetrical muscle wasting**

***Begin early or late in life**

***Chronic disability, progressive, incapacitating**

***Sx: generalized muscle weakness (lower), use upper body to compensate**

Muscular Dystrophy diagnostics

★ **DNA analysis**

- Blood or Muscle bx: measure dystrophin

★ **genetic testing**

★ **Prenatal dx: chorion villus biopsy (11-12th week Serum enzyme assay**

★ **Electromyography**

★ **Serum creatine kinase (CK): elevate early**

★ **Researching gene therapy**

Muscular dystrophy mgt

★ **Prevent complications**

- Infection & Spinal deformities, contractures
- Flu & pneumococcal vaccines

★ **Promote independence**

★ **Monitor cardiac & respiratory function**

- PFT, ECG, EKG
- Ventilation: noninvasive, pulse ox, trache, IPPV, suction

★ **Parental support**

- Genetic counseling

Pseudohypertrophic (Duchenne) Muscular Dystrophy (DMD)

★ **Defect in dystrophin protein in muscle fibers**

★ **most severe & most common MD in childhood**

★ **1 in 3600 live male births**

- 30-50% have no family hx
- Onset: 1st 3-4 years of life

★ **Death normally from respiratory or cardiac involvement**

Clinical manifestation (box 32-10)

★ **First: delayed walking, frequent falls, easily tired if walk, run or climb stair, bicycle**

★ **Difficulty rising from a sitting or supine position**

★ **Pseudohypertrophy: Enlargement of muscles d/t infiltration of fatty tissue**

★ **Toe walking, Waddling gait; lordosis; positive Gower's maneuver**

★ **Mental retardation frequent**

DMD boys

DMD Therapy

★ **Supportive care: Referrals**

- SNF or respite
- MDA of America

★ **Education:**

- language communication, manual skills

★ **PT/OT: braces, ROM**

- Stretch & breathing exercises, I/S

★ **Corticosteroids**

ACQUIRED NEUROMUSCULAR DISORDERS

★ **Guillain-Barre Syndrome (GBS) (Infectious polyneuritis)**

★ **Tetanus**

★ **Botulism**

★ **Spinal Cord Injuries**

Guillain-Barre Syndrome (GBS) (Infectious Polyneuritis)

★ **Acute inflammatory demyelinating polyneuropathy->deteriorating motor function & paralysis**

★ **Rapid onset; peak 4weeks**

★ **Symmetrical ascending progression of weakness, often paralysis; loss of reflexes**

★ **Affects 1-2/100,000**

★ **At any age, gender or ethnicity**

- Men>women

Guillain-Barre

GB Cause:

★ **Unknown**

★ **Viral or bacterial infection: Immune response to infectious organism (GI or resp illness 2-3 weeks prior)**

★ **Autoimmune**

★ **Immunizations**

★ **CMV, *Campylobacter jejuni*, Epstein-Barr**

BG Sx

★ **Infant Sx:**

- rapid progressive severe hypotonia
- ?resp distress
- feeding difficulty

★ **Children Sx:**

- rapidly progressive symmetric weakness & muscle pain

GB diagnostics

★ **Sx: progressive motor weakness & areflexia**

★ **Tests: Lumbar puncture**

- CSF: ^ protein & decrease lymphocytes
- Electrical test of nerve & muscle function
 - Electromyography

GB Therapy

Hospitalized

★ **Communication: explain procedures & txs**

★ **plasmapheresis**

★ **IV immune globulin (IVIG) & steroids**

★ **Immunosuppressive drugs**

Guillain-Barre mgt

★ **Respiratory: Trache, ETT or vent, pulse ox, ABGs, suction**

★ **Nutrition: IV or NGT**

★ **Prevent immobility complications: posture, strength, skin, DVT, alignment, ROM**

– PT: splints, aids, rehab, gait training, brace

★ **Bowel & bladder: prevent UTI, catheter**

GB mgt

★ **Emotional support: parent & child**

– GBS support groups

★ **D/c planning & teaching: refer to HHN or SW**

– Guillain-Barre Syndrome Foundation Intl

★ **Community care: outpt rehab**

– Positive self-image

★ **Recovery: 2-3 weeks**

– Most regain full strength; progresses in reverse

Tetanus "lock jaw"

★ **Acute, preventable, often fatal**

★ **generalized rigidity & convulsive spasms of skeletal muscles of jaw & neck**

★ **Cause: *Clostridium tetani***

★ **Found in: soil, dust intestinal tract (human & animal), skin & contaminated heroin**

★ **Entry: break in skin, umbilical cord**

Clinical manifestations (Box 32-12)

★ **Initial: stiffness & tenderness of muscles & neck & jaw**

– Infant: difficulty sucking

★ **Progresses to all voluntary muscles: opisthotonos, rigidity, tetany of respiratory muscles**

– Infant: inability to suck, excessive cry, irritability, & nuchal rigidity

Tetanus infant

Routine DTaP Vaccination

Tetanus care

- ★ICU: monitor & respiratory
- ★F/E & calorie intake
- ★Tetanus immune globulin (TIG) & tetanus toxoid
- ★Abx: penicillin G or erythromycin or tetracycline
- ★local care
 - Surgical debridement & cleansing
- ★Sedatives or muscle relaxants: diazepam (Valium), lorazepam (Ativan)
 - Neuromuscular blocking agent

Tetanus: nursing

- ★VS & neuro assess
- ★Decrease stimulation
 - Quiet room: sound, light & touch
- ★Monitor respiratory status
 - EET, vent, trache, O@ sat & ABG's
- ★Nutrition & hydration
 - NG, IV or gastrostomy feeding
- ★Reduce anxiety
 - Communicate beforehand
 - Anticipate needs
 - Encourage parents to room in

Botulism

- ★Acute flaccid paralysis caused by Ingestion of *Clostridium botulinum*
- ★Common <6 mos
- ★Source: restaurant foods; honey or light or dark corn syrup to infants; soil or dust
- ★Sx vary: constipation, poor feeding, weak cry, dehydration, lethargy, FTT->progressive resp paralysis & death

Infant botulism

Dx & Tx

- ★Onset: few hours to several days
- ★Dx: blood or stool specimen, EMG, food source
- ★nutritional & respiratory support
- ★Tx if suspicion:
 - ICU, NGT, PT/OT
 - Botulism Immune Globulin Intravenous (human) (BIGIV)
- ★Prognosis: good if tx

Infant Botulism

- ★Report signs of muscle impairment
- ★Recovery: slow, few weeks; 2% fatality
- ★Teach: timing feedings d/t fatigue
 - Stool softeners for several weeks
 - Aspiration risk
- ★Prevention: no honey under age 1 year

Spinal Cord Injuries

★ Causes: MVC's (leading cause)

- Older children: Sports related
- Toddlers & children: Falls, child abuse
- Birth injuries
- Penetrating: stab or gunshot wounds
- Congenital defect: myelomeningocele

The higher the injury the more neurologic damage

Spinal cord injuries Dx: observation

★ neuro exam

★ X-ray

★ CT

★ MRI

★ Fluoroscopy

★ Myelography

Tx: aggressive

★ Preop teaching with props

★ Monitor complications: DVT & PE

★ Skeletal traction or halo device

★ Surgery: fusion

★ Penetrating: Debridement & decompression

★ Corticosteroids

★ Functional electrical stimulation (FES)

Emergency interventions

★ Immobilize, stabilize before transfer

★ Calm, reassure, don't move; move when stabilized

- Cervical collar, rigid backboard
- Monitor airway & neuro function

★ If conscious: supine on flat surface

★ Infants & children: remove in car seat

WEB Sources

★ UCP.org

★ Ninds.nih.gov

★ Sbaa.org

★ Asbah.org

★ Osha.gov

WEB sources

★ Cdc.gov

★ Fsma.org

★ Mdausa.org

★ Muscular-dystorphy.org

★ Ncbi.nlm.nih.gov