Special Care Dentistry for the General Practice Resident:
Practical Training Modules

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Funded by the NYS Developmental Disabilities Planning Council

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Special Care Dentistry
For the General Practice Resident
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• This educational modular series consists of eight evidence based Power Point presentations designed to give the general practice resident a global view of dental treatment for people with special needs. Approximately 300 references are listed throughout this work. The eight modules address the most important aspects of clinical medicine and dentistry required for treating a patient with special needs. Discussion of access and barriers to dental care, the need for special care dentistry in the pre and post doctoral dental curricula, along with assessment of the competency of participants are included in the modules. Upon completion of the modules, the participant should have the knowledge to assess a patient with special needs.

• The educational package is a previously piloted pre and post test exam. The modules are accompanied by “teacher’s notes” which are visible in each Power Point presentation. This format alternately allows the instructor to assign the series as a self-study project.

continued
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• A description of each module follows below:
• **Introduction to Special Patient Care:** discusses the definition of disability, the prevalence and incidence of disability, aspects of “normalization”, and the barriers to care. A list of resources is provided for the individual and family.
• **Special Care Dentistry/ Legal and Ethical Issues:** discusses informed consent and various other types of consent, comprehensive medical history documentation, appropriate use of desensitization and restraint, communication/human rights issues, case law and detailed literature review of restraint.
• **Treatment Modalities/Treatment Planning for Patients with Special Needs:** discusses reasons for sedation, hospitalization OR cases, general anesthesia, pharmacological techniques, IV and enteral drugs.
• **Learning Disabilities/ Mental Retardation and Down Syndrome:** discusses the causes and risk factors, diagnosis and intervention, physical findings and medical concerns, dental and craniofacial characteristics of people with learning disabilities, mental retardation and Down syndrome.
• **Neuromuscular Disorders/Cerebral Palsy and Muscular Dystrophy:** discusses types of cerebral palsy, risk factors, oral and dental findings, various forms of muscular dystrophy and treatment planning considerations.
• **Autistic Spectrum Disorders:** defines and describes the spectrum of autistic disorders including Pervasive Developmental Disorder and Asperger’s. A recent review of the literature regarding proposed etiologies (i.e.: genetic links, vaccines) is presented, as well as suggestions for behavior management and treatment strategies.
• **Oral Manifestations/Genetic and Congenital Disorders:** discusses syndromology definitions, gene and chromosomal abnormalities, craniofacial disorders, dental and orthopedic conditions.
• **Seizure Disorders:** discusses definitions of seizures and epilepsy, risk, incidence and prevalence of seizures, classification and treatment of seizures, choice of medication therapies and practical considerations for dental treatment.
• Pre and post tests and the answer sheets are not included in the module series. Please contact Annette Shafer in the Office of Investigations and Internal Affairs at annette.p.shafer@omr.state.ny.us to request a copy and we will forward it to you electronically.
Neuromuscular Disorders
Cerebral Palsy & Muscular Dystrophy

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Cerebral Palsy
What is cerebral palsy?\textsuperscript{1,2}

- Cerebral palsy (CP) is a chronic disorder of movement or coordination, caused by injury to the immature brain during the prenatal or perinatal period.

- CP is a \textit{static encephalopathy}; that is, the original brain lesion does not progress or enlarge.
CP can be classified according to the type of movement disorder involved:\textsuperscript{2}

- **Spastic** (70-80%): characterized by increased muscle tone, tightness, stiff, jerky movements
- **Dyskinetic or athetoid** (10-15%): Low muscle tone, loose, uncontrolled body movement
- **Ataxic** (<5%): Affects balance and depth perception
Cerebral Palsy

Types of Cerebral Palsy

1. **SPASTIC** - tense, contracted muscles (most common type of CP).
2. **ATHETOID** - constant, uncontrolled motion of limbs, head, and eyes.
3. **RIGIDITY** - tight muscles that resist effort to make them move.
4. **ATAXIC** - poor sense of balance, often causing falls and stumbles.
5. **TREMOR** - uncontrollable shaking, interfering with coordination.

United Cerebral Palsy: www.ecaucp.org/images/types_of_cp.gif
CP can further be classified according to the topography of the neuromuscular involvement:²

- **Hemiplegia**: affects one side of the body
- **Paraplegia or Diplegia**: affects both legs (sometimes slight involvement in other extremities)
- **Quadriplegia**: affects all 4 extremities equally, as well as the trunk
- **Monoplegia** (very rare): Involvement of only 1 extremity
**ARM AND LEG ON ONE SIDE (HEMIPLEGIC)**

- arm bent; hand spastic or floppy, often of little use
- She walks on tiptoe or outside of foot on affected side.

**BOTH LEGS ONLY (PARAPLEGIC)**

- this side completely or almost normal
- child may develop contractures of ankles and feet.

**BOTH ARMS AND BOTH LEGS (QUADRIPLEGIC)**

- upper body usually normal or with very minor signs
- child may develop contractures of ankles and feet.
- When he walks, his arms, head, and even his mouth may twist strangely.
- Children with all 4 limbs affected often have such severe brain damage that they never are able to walk.
- The knees press together.
- legs and feet turned inward
Risk Factors (prenatal)²

- Intrauterine infections
- Maternal exposure to toxic or teratogenic agents
- Maternal abdominal trauma
- Multiple births
Risk Factors (neonatal)²

- Premature birth (<32 weeks)
- Low birth weight (<2500g)
- Intracranial hemorrhage
- Hypoxia and/or bradycardia
- Infection
- Trauma
- Hyperbilirubinemia
- Seizures
Associated disorders seen in individuals with CP³

- Intellectual disability (ID)
- Seizures
- Failure to thrive
- Vision and hearing impairment
- Speech impairment
- Dysphagia
- Hip dislocation
Oral and dental findings associated with CP$^{3,4,5,6,7,8,9,10}$

- Malocclusions
- Enamel defects
- Increased incidence of dental trauma
- Bruxism
- Sialorrhea (drooling)
• Individuals with CP have a higher rate of dental enamel defects than the general population

• Bruxism in a patient with neuromuscular and Intellectual impairment can be a very difficult behavior to extinguish
Malocclusions, especially anterior open bite and posterior crossbites, are secondary to hypotonia of oral-facial musculature and subsequent poor development of the palatal shelves\textsuperscript{5}
• Increased risk for dental trauma can be attributed to:⁷
  – Problems with balance
  – Muscle weakness in legs
  – Malocclusions
  – Seizures
Sialorrhea (drooling) in individuals with CP is not usually due to excessive production of saliva, but can be attributed to a compromised swallow reflex\textsuperscript{8,9}.
Dysphagia\textsuperscript{11,12}

Many individuals with neuromuscular disorders (including cerebral palsy and muscular dystrophy) that involve the oral/pharyngeal muscles develop dysphagia (altered swallow reflexes) or aphagia (an inability to swallow).
Dysphagia\textsuperscript{11,12,13}

- Any individual with dysphagia is at risk for developing aspiration pneumonia. Use of a rubber dam and diligent high speed suction are a must when providing treatment to these patients.
- Some individuals with aphagia may receive nutrition through a gastric tube. A side effect of pooling saliva can be extensive calculus deposits throughout the dentition.
These calculus deposits are typical of those that may be seen in a patient who receives nutrition via a gastric tube. In the posterior dentition, the calculus may completely cover all surfaces of the teeth.
The Muscular Dystrophies
What is muscular dystrophy?

Muscular dystrophy (MD) is an umbrella term used for a group of genetic diseases characterized by progressive weakness and degeneration of skeletal or voluntary muscles which control movement. Cardiac muscle and other involuntary muscles may also be involved.¹
Major forms of MD include:\(^2\)

- Duchenne
- Becker
- Fascioscapulohumeral
- Emery-Dreifuss
- Limb-girdle
- Myotonic dystrophy
- Congenital
Duchenne Muscular Dystrophy

- Onset: Early childhood (usually 2-6 yrs of age)
- Symptoms: Generalized muscular weakness and wasting. Limb and trunk muscles affected first
- Cognitive impairment is seen in some individuals with Duchenne MD
- Progresses fairly rapidly; eventually affects all voluntary muscles. Survival beyond late twenties is rare
- Incidence: 2/10,000
- Inheritance: X-linked recessive
Symptoms of Duchenne MD

Great effort is involved in getting to a standing position.

- Shoulders and arms are held back awkwardly when walking.
- Swayback.
- Weak buttocks and muscles (hip straighteners).
- Knees may bend back to take weight.
- Thick lower leg muscles (the 'muscle' is mostly fat, and not strong).
- Tight heel cord (contracture); child may walk on toes.
- Belly sticks out due to weak belly muscles (child is poor at sit-ups).
- Thin, weak thighs (especially front part).
- Poor balance; falls often.
- Awkward, clumsy if walking.
- Weak muscles in front of leg cause 'foot drop' and tiptoe contractures.
Symptoms of Duchenne MD

The *pseudohypertrophy* commonly seen in Duchenne MD is actually caused not by muscle enlargement, but by fatty infiltration of muscle tissue.
Becker Muscular Dystrophy

- Symptoms resemble Duchenne MD, but progresses much more slowly
- Symptoms usually appear by age 12, course is more variable than that seen in Duchenne MD
- Ability to walk may continue to age 40
- Incidence: 3-6/100,000
- Inheritance: X-linked recessive
X-linked recessive inheritance pattern: Duchenne and Becker MD
• The underlying problem in both Duchenne and Becker MD is a defect in the gene that produces dystrophin (a muscle protein).\(^1\)

• In Duchenne MD, little to no dystrophin is produced

• In Becker MD, some dystrophin is produced, but it is not completely functional
Diagnostic Tests for Duchenne and Becker MD³

- **Serum Creatine Kinase**: will be elevated in both disorders
- **Electromyography**: will see reduced electrical activity in muscles affected by MD
- **Muscle biopsy**: can examine dystrophin activity in muscle – can help distinguish between Duchenne and Becker MD
- **DNA testing** prenatally is only about 70% accurate in determining if a male fetus is affected
Fascioscapulohumeral MD²

- Onset: Childhood to teens
- Symptoms: Slow progression of facial muscle weakness (eyelid drooping, difficulty pronouncing words), progressive weakness of shoulders and upper arms
- Incidence: 5/100,000
- Inheritance: Autosomal dominant
Emery-Dreifuss MD^2

• Onset: Childhood to early teens
• Symptoms: Slow progression of muscle weakness in shins, upper arms and shoulders. Joint deformation commonly noted.
• Cardiac involvement is common
• Inheritance: X-linked recessive (female carriers, males affected)
Limb-Girdle MD²

- Onset: Childhood to middle age
- Symptoms: Slow progression of muscle weakness; first noted in shoulder and pelvic regions.
- Cardiac & pulmonary complications are common in later stages of disease
- Inheritance: Can be either autosomal recessive or x-linked recessive
Myotonic Dystrophy

- Most common adult form of MD (onset usually by 20 yrs), rarely seen in a congenital form
- Symptoms: Delayed relaxation of voluntary muscles (myotonia) is usually first symptom. Muscle wasting first affects face, neck, hands, feet.
- Inheritance: Autosomal dominant
Myotonic Dystrophy

- Has a very variable course; can be fairly benign or progress to severe disability
- A true multi-systemic disease. Complications include:
  - Cardiomyopathy
  - Ocular cataracts
  - Respiratory impairment
  - Dysphagia
  - Pancreas (excessive insulin output)
Autosomal Dominant Inheritance
Myotonia

- Characterized by prolonged muscle spasms and stiffening.
- For example, percussion of the palmar muscles will cause the thumb to move into opposition and adduction. Return to the initial position will be very slow.
Head & Neck Symptoms of Myotonic Dystrophy

- Frontal balding
- "Hatchet" facies due to atrophy of temporalis muscle
- Ptosis and drooping mouth due to weakness of facial muscles
- Wasting of sternocleidomastoid muscle
- Cataracts
- Gynecomastia
Congenital MD²,⁶

- **Onset:** Present at birth
- **Symptoms:** Very variable
  - **Classical CMD:** slowly progressing muscle weakness, changes in white matter of brain, usually no cognitive deficits
  - **Fukuyama CMD:** Severe progressive muscle weakness, mental retardation
- **Incidence:** Rare. Classical form more common in USA, Fukuyama form more common in Japan and Europe
- **Inheritance:** Usually autosomal recessive (affects males & females equally)
Congenital MD

Muscle weakness typically seen in infants with congenital muscular dystrophy
Oral findings in MD$^{7,8}$

- Oral findings are variable and progressive, depending on severity of disease. These can include:
  - Anterior open bite
  - Expansion of maxillary and mandibular arches
  - Macroglossia
  - Loss of facial muscle strength
  - Sialorrhea

- As with CP, patients with gait problems are at increased risk for dental trauma
Anesthesia issues in MD\textsuperscript{9,10,11}

- A number of reports have been published linking inhalation anesthetic agents & the neuromuscular blocker succinylcholine to hyperkalemia, cardiac arrest and rhabdomyolysis in individuals with Duchenne and Becker MD.
- Cardiac and respiratory complications during general anesthesia are considerations for individuals with all types of MD.
Treatment planning considerations for patients with neuromuscular impairment

• Carefully review the medical history; consult with physician if necessary
  – Are there any medical contraindications to proposed treatment?
  – Any medications that may impact oral health or interact with local anesthetics, antibiotics and/or pain medication?
• What is the patient’s ability to cooperate with dental treatment?
  – Patients with cognitive impairment may have difficulty cooperating – behavior management techniques?
  – Some CP patients may have involuntary movements that can interfere with treatment
• What is the patient’s ability to maintain oral health and/or any restorative treatment that may be planned?
  – Many of these individuals may be dependent on a caregiver to provide daily oral hygiene; does this person need instruction?
  – Some patients with cognitive impairment may be resistant to oral hygiene care
  – Does this patient have a history of repeated trauma to the dentition?
References (cerebral palsy)

References (cerebral palsy)

References (muscular dystrophy)

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Photo/illustration credits

Types of CP (head diagram): Eastern Central Alabama United Cerebral Palsy.  

Topography of CP: Japanese Society for Rehabilitation of Persons with Disabilities.  

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Photo/illustration credits

Pseudohypertrophy radiograph: University Hospital of Cleveland.  
www.uhrad.com/ray/peds2a.htm

Pseudohypertrophy photograph: Washington University, St. Louis.  
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Myotonia (hands): Netter, F.H. The CIBA Collection of Medical Illustration.  

Myotonia (Head & Neck): Netter, F.H. The CIBA Collection of Medical Illustration.  

Congenital MD: Washington University, St. Louis.  
www.neuro.wustl.edu/neuromuscular/syncm.html
THANK YOU

• Thank you to the Task Force on Special Dentistry Committee for their dedication to this project.

• Special thanks to the past and current Chair members of the Task Force on Special Dentistry:

  Dr. Alicia Bauman
  Dr. Craig Colas
  Dr. Nancy Dougherty
  Dr. Vincent Filanova

  Dr. Gary Goldstein
  Dr. Roderick MacRae
  Dr. Edward Riggins
  Dr. Maureen Romer
  Dr. Carl Tegtmeier