Congenital Muscular Torticollis:
Anatomic, Pathologic, and Management Considerations

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Objectives
- Describe the applied anatomy involved in CMT
- Review the proposed pathologic mechanisms involved in CMT
- Recognize the differential diagnosis of infantile torticollis
- List the essential components in the medical evaluation of CMT
- Explain the rationale for advanced conservative and surgical approaches in the treatment of CMT

Disclosure
- I have no actual or potential conflict of interest in relation to this program

Torticollis
- Definition:
  - Latin: Tortus – twisted
  - Cifnum – neck
  - Any asymmetric posturing of the head and neck
- Different types
  - Congenital muscular torticollis (infancy)
  - Benign paroxysmal torticollis (childhood)
  - Torticollis spasmodica (childhood to adulthood)
    - Cervical dystonia

Anatomy
Anatomy: Lateral Neck

- SCM
- Upper trapezius
- Scalenes
- Splenius capitis, cervicis

Action:
- Contralateral rotation
- Ipsilateral lateral flex/ext, shoulder elevation
- Ipsilateral lateral flexion
- Ipsilateral rotation
- Ipsilateral rotation
- Ipsilateral rotation/ext
- Ipsilateral rotation/ext
- Ipsilateral rotation/ext

Anatomy: Deep/posterior Neck

1. Semispinalis Capitis (head rotation/extension/backward)
2. Rectus Capitis (extends cervical vertebrae)
3. Longissimus Capitis (extends cervical vertebrae)
4. Longissimus Capitis (head rotation/extension/backward)
5. Longissimus Thoracis (extends/lateral flexion/rib rotation)
6. Iliocostalis (extends/lateral flexion/rib rotation)
7. Semispinalis Thoracis (extends/lateral flexion/rib rotation)

Anatomy: Suboccipital Neck

• Considerations
  - Primary muscles involved:
    • Contractures limiting PROM
      - SCM
      - Upper trapezius
      - Scalenes
      - Splenius capitis, cervicis
    - Secondary muscles involved (?):
      •  Secondary contractures (limited PROM
      - Longissimus
      - Iliocostalis
      - Suboccipital muscles

Anatomy: Functional Aspects

- Muscle
  - SCM
  - Upper trapezius
  - Scalenes
  - Splenius capitis, cervicis
  - Iliocostalis cervicis
  - Longissimus cervicis
  - Longissimus capitis
  - Obliquus capitis superior
  - Obliquus capitis inferior
  - Rectus capitis major

- Action
  - Contralateral rotation
  - Ipsilateral lateral flex/ext, shoulder elevation
  - Ipsilateral lateral flexion
  - Ipsilateral rotation
  - Ipsilateral rotation
  - Ipsilateral rotation/ext
  - Ipsilateral rotation/ext
  - Ipsilateral rotation/ext

Pathology and Etiology
Pathology

- **Pathology**
  - Unilateral fibrosis of the SCM leading to abnormal and symmetric head/neck posturing
  - Contralateral head rotation coupled with ipsilateral tilt
  - Secondary contractures of other musculature due to prolonged, shortened positions

Etiology

- **Etiology - ?**
  - Postulates:
    - Ischemic injury to SCM due to “kinking” of muscle during delivery (extreme neck flexion/lateral bending/rotation) (Davids, Wenger, Mubarek, 1993)
    - Compartment syndrome
      - Nerve and muscle damage
      - Swelling
      - Fibrosis
    - Abnormal vasculature -> ischemia/fibrosis/contracture
    - Direct muscle trauma/partial rupture -> fibrosis/contracture
    - Abnormal intrauterine posture -> contracture

Etiology/Pathology

- **Epidemiology**
  - Incidence: 0.04%-1.9%
  - Right>>Left
  - Male : Female - 1:1
  - Mean age at dx: 4 mos
  - Associated conditions:
    - Hip dysplasia: 2%-25% incidence
    - Plagiocephaly – 65% (Wei, et al. 2001)
    - Progressive facial asymmetry

Plagiocephaly

- **Causes**:
  - Craniosynostosis
    - Positional pressures (Torticollis)
  - Muscular forces / imbalances (Wolf’s Law) (Torticollis)

Differential Diagnosis
Differential Diagnosis: CMT

- Torticollis with unilateral SCM contracture and mass
  - Normal xray:
    - CMT

- Torticollis with unilateral SCM contracture, mass (-)
  - Normal xray:
    - CMT

- Torticollis without SCM contracture, mass (-)
  - Normal xray:
    - RP1
    - Congenital absence of a cervical muscle
    - Neurologic
      - CNS lesions: cystic formations, migrational defects
      - CNS tumors: brainstem, cerebellum/posterior fossa, spinal cord
    - Ocular abnormalities (rare)


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Differential Diagnosis: CMT

- Torticollis without SCM contractures, tumor (-)
  - Abnormal xray:
    - Vertebral anomalies
      - Hemivertebrae
    - Unsegmented bar vertebrae
    - Cervical rotatory subluxation (rare in CMT)
    - Klippel-Feil Syndrome
    - Congenital scoliosis


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Differential Diagnosis: Acquired Torticollis

- Rotatory subluxation, C1-C2
  - Inflammatory conditions
    - Rheumatoid arthritis
    - Osteomyelitis of c-spine
    - Cervical lymphadenitis
    - Pharyngitis
    - Tonsillitis
    - Mastoiditis
    - Cervical abscess

- Trauma
  - Fractures
  - Dislocations

Differential Diagnosis: Acquired Torticollis

- Non-traumatic, xray (-)
  - Ocular lesions
    - Ocular tumor
    - Diplopia
    - Extraocular paralysis
  - Sandifer Syndrome
  - Hiatal hernia + torticollis
  - Neurologic syndromes
    - Arnold Chiari Malformation
    - Dysautonomic syndromes
    - Postparaphrenic
    - Posterior fossa pathology
    - Spinal cord pathology - tumors, syrinx

Medical Evaluation

Physical Examination: Global Evaluation

- The physical examination necessarily consists of a broad assessment of the child, directed towards defining neurologic function, musculoskeletal development, and developmental status

  - Is there evidence of upper motor neuron/CNS pathology?
  - Is there evidence of developmental delays?
  - Is there evidence of other musculoskeletal abnormalities?
  - What is the degree of craniofacial abnormality?

  - KEY: Make sure there is nothing else going on

Physical Examination: Global Evaluation

- **Neurologic**
  - Tone: DTR’s, Spasticity/clonus
  - Coordination
  - Movement patterns
    - UE/LE
    - Facial
  - Ocular movements
    - Range?
    - Nystagmus?
  - Cranial nerves

- **Musculoskeletal**
  - Head shape
    - Plagiocephaly
    - Postural?
    - Structural?
  - Facial development
    - Asymmetries?
      - Cheek
      - Ocular
      - Mandibular
  - Spine
  - Cervical palpation
    - Masses?
    - Adenopathy?
    - Contractures?
  - Muscular development
  - Cervical ROM
    - Active
    - Passive
  - ROM, other
    - Hips, etc...

Physical Examination

- **Other**
  - Functional/developmental skills – age appropriate?
    - Crawling
    - Rolling
    - Transitions
    - Manual/fine motor skills
    - Cognitive/language

Medical Management

I'm on the pacifier path.
Conservative Treatment

- **Initial management**
  - Upon initial clinical diagnosis:
    - X-ray of cervical spine
      - Should be done before initiating PT
    - Initiation of outpatient PT / HEP
      - Begin as soon as possible
      - Before 3 mo age – 0 surgical need
      - After 3 mo age – 25% required surgery
    - With consistent, timely therapy 85-90% CMT should resolve within 4-5 months

To Radiate or Not to Radiate?

- 5/2 pts, 0-12 mos of age during a 10 yr period were studied
- All infants had plan radiographs for torticollis
- 10 had reported abnormal findings
  - 6 of the 10 had normal follow up studies
  - 4 of the 10 boney abnormality confirmed by additional studies
- 25 of the 492 with normal initial studies underwent further imaging due to continued concern for boney abnormality; 0 of the 25 patients had abnormalities
- Sensitivity 100%, Specificity 98% for detection of abnormality
- Prevalence of true c-spine abnormality ~ 0.8%
- Prevalence of clinically relevant abnormality ~ 0.2%
- No false negative from the initial radiograph were identified

Conservative Treatment

- **Refractory CMT**
  - Defined: Little to no improvement, or plateau in gains after 4-5 months of PT and/or 7-8 months of age
  - Concerns:
    - facial asymmetry
    - plagiocephaly
    - Irreversible contracture
    - Permanent alterations in cervical ROM
    - Other pathology
  - Approach:
    - Must consider/evaluate for other causes of torticollis
    - Modify treatment approach

Conservative Treatment: Refractory CMT

- **Evaluation**
  - 2 key steps:
    - Cervical spine X-ray (if not done)
    - Focused neurologic/developmental evaluation
  - IF:
    - X-ray abnormal / evaluation is normal:
      - Refrain from PROM, continue AROM and positioning
      - Refer to Orthopedic Surgery Clinic
      - Consider botulinum toxin injections

Conservative Treatment: Refractory CMT

- **Evaluation**
  - IF:
    - X-ray normal / evaluation abnormal (neurologic findings, developmental delays)
      - MRI scan of cervical spine, brainstem, brain
      - May continue PROM/AROM program
    - X-ray normal / evaluation normal
      - Botulinum toxin injections
      - Continue PROM/AROM program
      - Consider CT scan of cervical spine if no improvement after botulinum toxin injections

Refractory CMT: Conservative Treatment

- **Botulinum Toxin type A**
  - Evidence in the literature:
    - Collins A. et al. 2006
    - 2 of 7 patients (pediatric plus adult) with CMT responded
    - Oleszok JL et al. 2005
      - 20/27 patients (age 6-18 months) with improved cervical rotation
    - Injections to SCM and/or upper trapezius (30 units)
    - Adverse affects in 2/27 – mild dysphagia, neck weakness
    - Joyce MB. 2005
      - 14/15 children with improved cervical ROM
### Refractory CMT: Conservative Treatment

- **Botulinum Toxin type A**
  - CMH experience (anecdotal):
    - 2001-2008
    - Approximately 250-300 patients
    - 80-85% gain some benefit in PROM/AROM
  - Injection sites:
    - Rotational deficit:
      - SCM
    - Upper trapezias
    - Lateral flexion deficit:
      - Scalenes

### Refractory CMT: Surgical Treatment

- "a review of the literature on CMT shows that fewer than about 16% of children treated conservatively before one year of age will require surgery" – Karmel-Ross, 1997.
- Botulinum toxin will likely shift this dramatically
  - CMH: 40-50 new evaluations of refractory CMT per year
  - Approx. referrals to Plastic Surgery – 2-3/year (<5%)
- Surgical indications:
  - Refractory CMT, non-responsive to conservative treatment and botulinum toxin injections
  - Age > 18 months

Thank You!