Addressing a Moving Target: Treatment of Foot and Ankle Disorders in Patients with Charcot-Marie-Tooth Disease

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Disclosure of Relevant Financial Relationships

We have no financial relationships to disclose.

Disclosure of Off-Label and/or investigative uses:

We will not discuss off label use and/or investigational use in this presentation

Purpose

- Describe three typical gait patterns observed in patients with CMT and provide evidence to support non-operative and operative treatment options

Overview

- Utility of Gait Analysis
  - Understanding pathomechanics
  - Treatment decision-making
- Ankle motion – the basics
- CMT ankle
  - Gait characteristics (video, kinematics, kinetics)
  - Treatment options
- Discussion

Sources

- Literature
- Our experience of examining 68 patients with CMT with comprehensive motion analysis
- Õunpuu et al., Gait and Posture, 2013.


Gait & Posture

A comprehensive evaluation of the variation in ankle function during gait in children and youth with Charcot-Marie-Tooth disease

Background

- The optimal treatment of gait pathology requires a detailed understanding of the pathomechanics during gait
- Visual assessment is limited in providing a full understanding of movement pathology
  - It is just too complicated!

Gait Analysis Is…

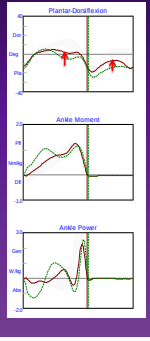
- The objective documentation of gait function in terms of the following:
  - Joint angles (joint kinematics) in 3D
  - Joint moments and powers (joint kinetics) in 3D
  - Muscle activity
- Includes integration of the above data with the impairments such as:
  - Weakness
  - Limited range of motion
  - Bony deformity

Gait Analysis Is…con't

- The development of a list of primary gait problems and their causes (that should be treated) and associated gait compensations (that should not be treated)
- The development of a list of proposed treatments including specific indications and the proposed outcomes

Gait Analysis Is…con't

- Objective evaluation of treatment outcomes using the same treatment decision-making methods

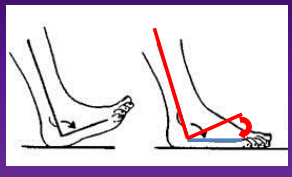


The First Step

- Know what you are talking about.
- Understand the gait analysis data.

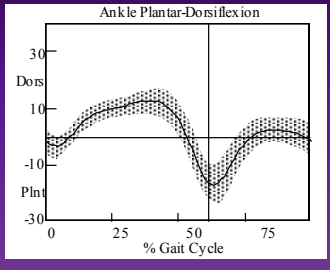
Angle Definition – Ankle Sagittal Plane

- The relative angle between perpendicular to the long axis of the shank and the plantar aspect of the foot
- As viewed by an observer looking along an axis perpendicular to the shank-foot plane



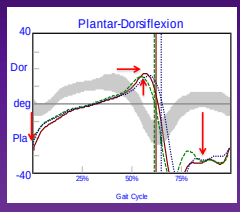
Please note: the ankle joint angle definition includes multiple joints (ankle and foot)

Ankle Sagittal Plane Motion



Pathological Ankle Motion

- Increased plantar flexion in swing and at initial contact
- Delayed and increased peak dorsiflexion in terminal stance



Charcot-Marie-Tooth (CMT)
(Hereditary Sensory and Motor Neuropathy)

- Most commonly inherited neurological disorder = de-myelination of large peripheral nerves
  - Myelin & axonal subtypes
- Characterized by:
  - distal muscle weakness and imbalance
  - foot and ankle deformities
  - associated gait implications
  - impairment progression at varying rates

Textbook Gait Description

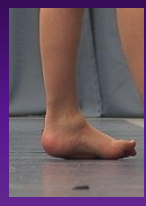
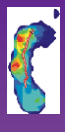
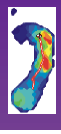
- Foot drop (excessive equinus) in swing
- Steppage (hyper-flexion of knee and hip in swing)
- Circumduction and pelvic hiking in swing

(Fenton, *JOPA* 1984)
(Morrisy, *Pediatric Orthopedics*)
(Vinci, *Archives of Physical Medicine & Rehab* 2002)

Textbook Clinical Description

- Forefoot equinus and adductus
- Hindfoot varus
- Pes cavus
- Toe deformities
  - claw toes

(Guyton; *Foot and Ankle* 2000)

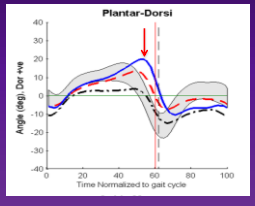





- Clinical experience:
  - Persons with CMT do not all have the same clinical presentation
  - Therefore, there are a variety of gait patterns and deformity…

Clinical experience shows variations in presentation...


- Peak ankle dorsiflexion in terminal stance = clinically relevant gait impact
- Three groups were defined:
  - greater than typical
  - within typical range
  - less than typical

Ankle Sagittal Plane Kinematics



- less than typical (dash-dot)
- typical (large dash)
- greater than typical (solid)

- Cavus deformity = clinically relevant gait impact
- Kinematically appears as “ankle plantar flexion” (even if due to foot)
- Prevalence:
  - Increased cavus (82% of feet – our experience)

Cavus

Framework for Treatment Decision-making

- Prerequisites of Typical Gait
  - Stance phase stability
  - Swing phase clearance
  - Appropriate prepositioning at initial contact
  - Adequate step length
  - Energy conservation

Perry J, Gait Analysis, 1992

The Flail Foot

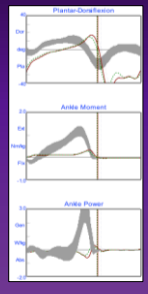
- Stance phase stability issues
- Swing phase clearance issues
- Inappropriate prepositioning at initial contact
- Reduced step lengths

The Flail Foot

- Clinical Examination Findings
  - Limited passive dorsiflexion range of motion
    - Knee flexed (1 ± 7 degrees)
    - Knee extended (8 ± 7 degrees)
  - Full plantar flexion and forefoot inversion/eversion
  - Strength: (median/maximum/minumum)
    - Plantar Flexors (2/5/2)
    - Dorsiflexors (4/5/0)
    - Forefoot Invertors (5/5/0)
    - Forefoot Evertors (4/5/2)

The Flail Foot

- Gait Characteristics
  - Increased and delayed peak dorsiflexion in terminal stance
  - Increased equinus in swing and at initial contact
  - Reduced peak plantar flexor moment and power generation in terminal stance



The Flail Foot

- Treatment Options
  - Brace
  - Surgery to maintain a "braceable position"

Flail Foot – TX

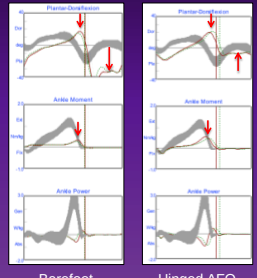
- Functional outcome of this ankle weakness includes instability in standing and during gait due to limited ability to bear weight over the forefoot
- Reduced base of support
- Excessive equinus in swing and associated clearance difficulties lead to tripping and falling

Ankle-foot Orthoses (AFO's)

- limit excessive dorsiflexion and allow weight bearing on the distal portion of the foot
- will provide more stability for the patient in standing and during gait
- limit excessive equinus and associated clearance problems in swing

Barefoot vs. Hinged AFO

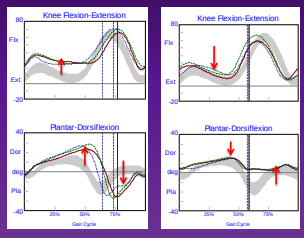
- Reduced excessive plantar flexion in swing
- No change in peak ankle dorsiflexion timing in terminal stance
- No improvement in peak ankle plantar flexor moment in terminal stance



Barefoot Hinged AFO

Barefoot vs. Solid AFO

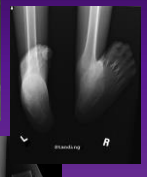
- Reduced excessive plantar flexion in swing
- Reduced excessive dorsiflexion in terminal stance
- Associated reduced excessive knee flexion in stance



Barefoot Solid AFO

Flail Foot TX: Surgery may be needed if foot "unbraceable"

Tendon transfers won't work if no strength



Flail Foot TX: Surgery

- Posteromedial release
  - Achilles Z lengthen
  - Posterior capsulotomies
  - Abductor Hallucis
  - FHL/FDL
  - TN capsulotomy
  - Plantar fascia release
- Closing cuboid osteotomy
  - (cuneiform too osteopenic to open)

The Cavovarus Foot

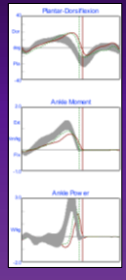
- Stance phase stability issues

The Cavovarus Foot

- Clinical Examination Findings
  - Limited passive dorsiflexion range of motion
    - Knee flexed (2 ± 6 degrees)
    - Knee extended (9 ± 7 degrees)
  - Full plantar flexion
  - Variable forefoot inversion/eversion
  - Strength: (median/maximum/minumum)
    - Plantar Flexors (4/5/2)
    - Dorsiflexors (5/5/4)
    - Forefoot Invertors (5/5/3)
    - Forefoot Evertors (5/5/3)

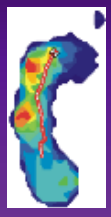
The Cavovarus Foot

- Gait Characteristics
  - Delayed peak dorsiflexion in terminal stance
  - Reduced peak plantar flexor moment and power in terminal stance



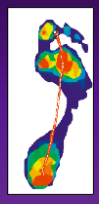
Foot Pressures

- Increased lateral weight bearing
- Reduced toe contact



Left

Right



Typically Developing

The Cavovarus Foot

- Treatment Options
- Consider presence of foot pain, shoe wear issues, stability in stance

Cavovarus Foot Treatment Considerations

- Cavus: Imbalance between peroneus longus (plantarflexes 1st ray) & anterior tibialis (dorsiflexes 1st ray)
- Varus: Imbalance between posterior tibialis (inverts hindfoot) & peroneus brevis (everts hindfoot)

Cavovarus Treatment: Non Op

- Plantar fascia stretch
- Strengthening exercises: dorsiflexors & evertors
- Bracing

Cavovarus Treatment: Surgical

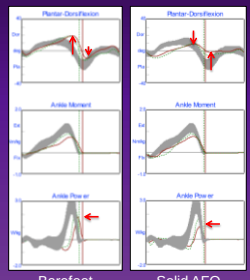
- Soft tissue release – if flexible
  - Plantar fascia
- Osteotomy – if fixed
  - Dorsiflexing 1st ray osteotomy
  - Calcaneal osteotomy
- Tendon transfers – to balance/delay recurrence
  - Peroneus longus to brevis
  - EHL to neck of 1st MT
  - (Anterior tibialis laterally)
- Arthrodesis – if severe/recurred

Cavus Component - TX

- Treatment of the cavus deformity may be a consideration depending on atypical foot pressures and associated pain
- The implications of plantar fascia release on "available" plantar flexor length in combination with weakness need to be considered to prevent excessive peak dorsiflexion post treatment

Barefoot vs. Solid AFO

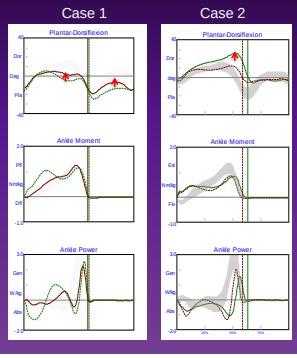
- Reduced excessive peak dorsiflexion in terminal stance
- Reduced plantar flexion range of motion
- Maintained peak plantar flexor moment in terminal stance
- Reduced peak power generation in terminal stance



Radiographic Findings

Treatment Outcomes Experience

- Through gait analysis we know that outcomes vary



Toe Walker

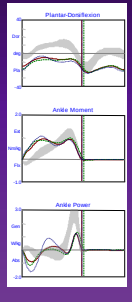
- Stance phase stability issues
- Inappropriate prepositioning at initial contact
- Swing phase clearance issues

Toe Walker

- Clinical Examination Findings
  - Limited passive dorsiflexion range of motion
    - Knee flexed (-2 ± 9 degrees)
    - Knee extended (-2 ± 13 degrees)
  - Full plantar flexion and forefoot inversion/eversion
  - Strength: (median/maximum/minumum)
    - plantar flexors (5/5/2)
    - Dorsiflexors (5/5/2)
    - Forefoot Invertors (5/5/3)
    - Forefoot Evertors (5/5/4)

Toe Walker

- Gait Characteristics
  - Increased equinus in stance and wing
  - Absence of dorsiflexior moment in loading
  - Reduced power generation in terminal stance



Toe Walker - TX

- implications of "toe walking" include instability in stance and standing due to the limited base of support under the foot

Toe Walker

- Treatment Options
  - Lengthening of plantar flexors or correction of cavus - never both
  - Cavus correction is adequate
  - Leave alone – increased body weight and weakness
- Clinicians must consider implications of
  - reducing plantar flexor contracture by lengthening a weak muscle which is likely to weaken more over time
  - reducing cavus deformity with implications on "available" plantar flexor length

Surgical Question?

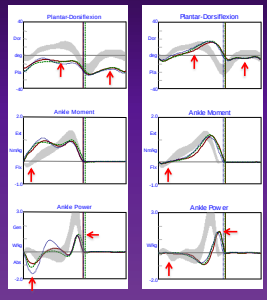
- What combination of options will be most appropriate to treat toe walking without creating excessive dorsiflexion?
  - Consider relationship between cavus and AVAILABLE plantar flexor length
  - Implications of plantar flexor weakness
  - Treatment effects go beyond target joint
- Can we predict treatment outcomes?

Evidence for Tx Recommendations

- Objective follow-up of surgical outcomes is needed to understand ultimately the indications and counter-indications for plantar flexor lengthenings

Gait Findings – Pre vs. Post

- Increased dorsiflexion in stance and swing
- Addition of dorsiflexor moment in loading response
- Maintained power generation terminal stance



RIGHT-Pre

Right-Post

Treatment - Beware

- Lengthening the plantar fascia
  - results in "lengthening" of the plantar flexors
- Dorsiflexing closing wedge osteotomy (first ray)
  - results in "lengthening" of the plantar flexors
  - IF YOU DORSIFLEX THE FIRST RAY W/O PLANTAR FASCIA, THE PLANTARFLEXORS MAY ACTUALLY TIGHTEN UP

HOW DOES THIS ALL INTERACT?

Treatment Summary

- Current Options:
  - Therapies
  - Bracing
  - Surgical Intervention
- Determine prerequisites of typical gait that are compromised
- Describe clinical and radiographic findings and associated gait issues
- Define treatment hypothesis

Principles

- Provide support when strength/stability issues are present
- Correct anatomical deformity to improve biomechanical function
- Consider treatment when pre-requisites of gait are compromised
- Progressive pathology – progression can be documented objectively using motion analysis techniques

Thank You