Answering Your Questions: Bracing and Charcot Marie Tooth

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CMT 1B

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Purpose of this Presentation .....

Point of view from Orthotist

- Description of CMT
- Some History
- Understand the disease process
  - Pathophysiology
  - Pathomechanics
  - Critical insight into best designs
- Patient Evaluation
- Orthotic Management Options

CMTA
Charcot-Marie-Tooth Association

History

1886

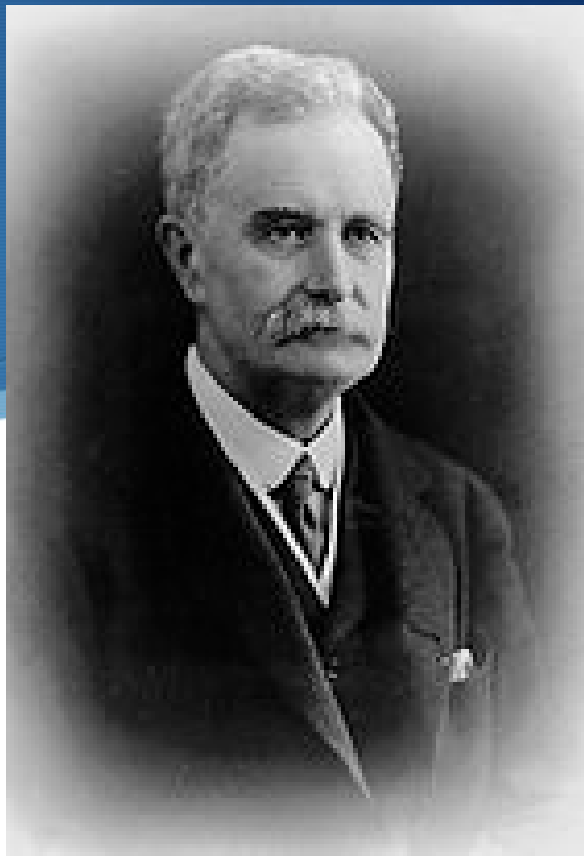
2 papers were submitted

Jean-Martin Charcot

61 y/o

Pierre Marie

33 y/o



Howard Henry Tooth

Cambridge Thesis:
“The Peroneal type of Progressive Muscular Atrophy”

29 y/o

Charcot Marie Tooth Disease Aka:

- Other names: Peroneal Muscular Atrophy, HMSN: Hereditary Motor Sensory Neuropathy, Charcot-Marie-Tooth-Hoffman, Tooth's Motor sensory neuropathy
- Description: A progressive inherited neuropathy that is characterized by motor and sensory loss, predominantly in the feet and legs but also in the hands and arms.

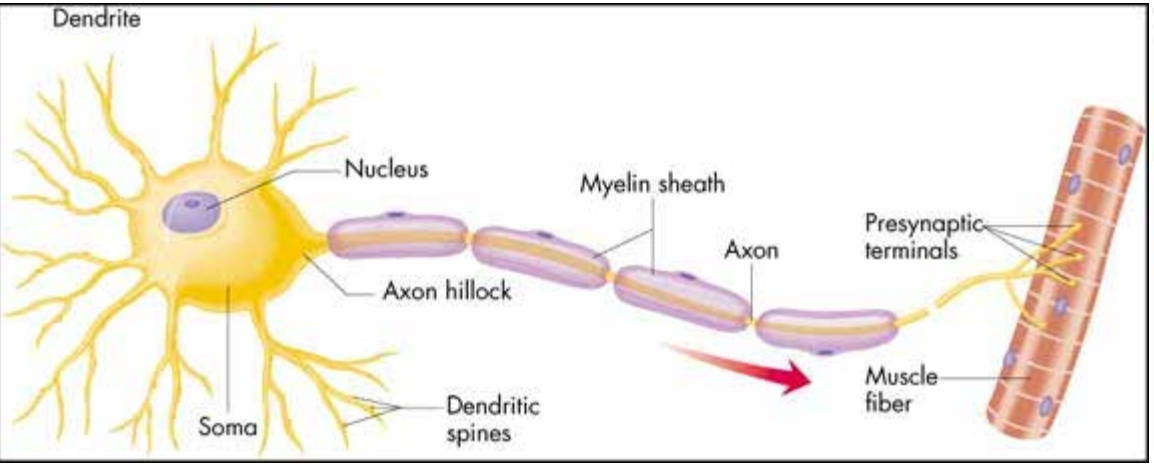
Proportion of CMT

- CMT1 Demyelination
- CMT2 Axonal degeneration
- Currently there are ~ 80 different kinds of CMT

EMG Studies

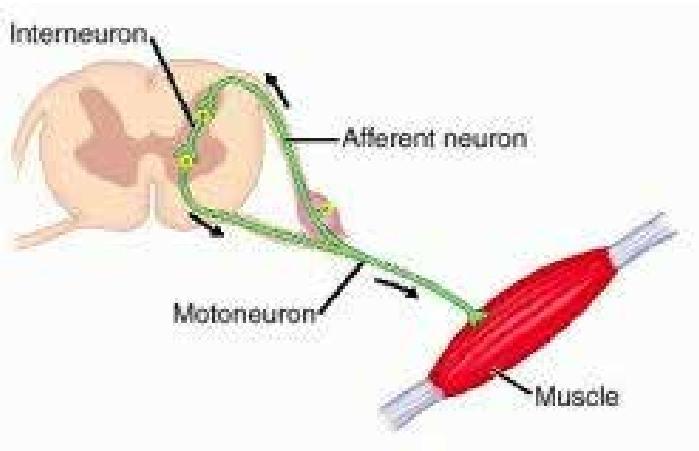
Demyelinating

- peripheral neuropathy characterized by:
- Slow nerve conduction velocity typically 5-30 meters per second;
- Normal CV
  - Tibial nerve 47.8 m/s
  - Peroneal nerve 47.1 m/s
- Hypertrophic peripheral nerves with onion bulbs biopsy
- Near normal amplitude



Axonal Degeneration

- Chronic denervation on EMG in distal muscles with
- Reduced compound motor action potentials
- Normal action potentials
  - Tibial nerve 8.8 mV
  - Peroneal nerve 6.0 mV
- Near-normal or normal motor conduction velocities
- 2B preserved sensory nerve action potentials, including the sural response



- Does not affect longevity
- Primarily affects below elbows and knees
- Can predict severity (?maybe?)
- Managed well

Complaints

- General foot weakness (foot drop, foot slap)
- Unsteady gait (poor balance)
- Chronic lateral ankle sprains
- Glove and stocking hypoesthesia
  - "numbness"
- Atrophy of hand muscles
  - "Hand weakness, dropping things"
  - Thumb opposition
- Atrophy of distal leg muscles
- Claw toes
- Painful calluses
  - Base and head of 5^{th} metatarsal
  - 1^{st} and 5^{th} met heads

Evaluation

- History
  - Occupation, present activities, desired activities.
  - Complaints (what is hard to do?, what limits you?)
    - Weakness, instability, balance, reduced activity levels

Evaluation

- Assessment
  - Sensory Testing
  - Range of Motion
  - Manual Muscle Test
  - Gait Deviations
  - Balance
  - Cadence (gait)

Evaluation

- Ask, Explain, Discuss, Agree
  - Get some specifics on you want
    - may not have even thought about options
  - Describe orthotic options
  - Agree on a treatment plan

Goals/ what can we achieve?

- Traditional orthotic goals:

  - Prevent deformity
  - Support and align skeletal structures
  - Limit or enhance motion about a specific joint

  - …….Balance, support, and reduce fatigue to muscles

Pathophysiology of CMT

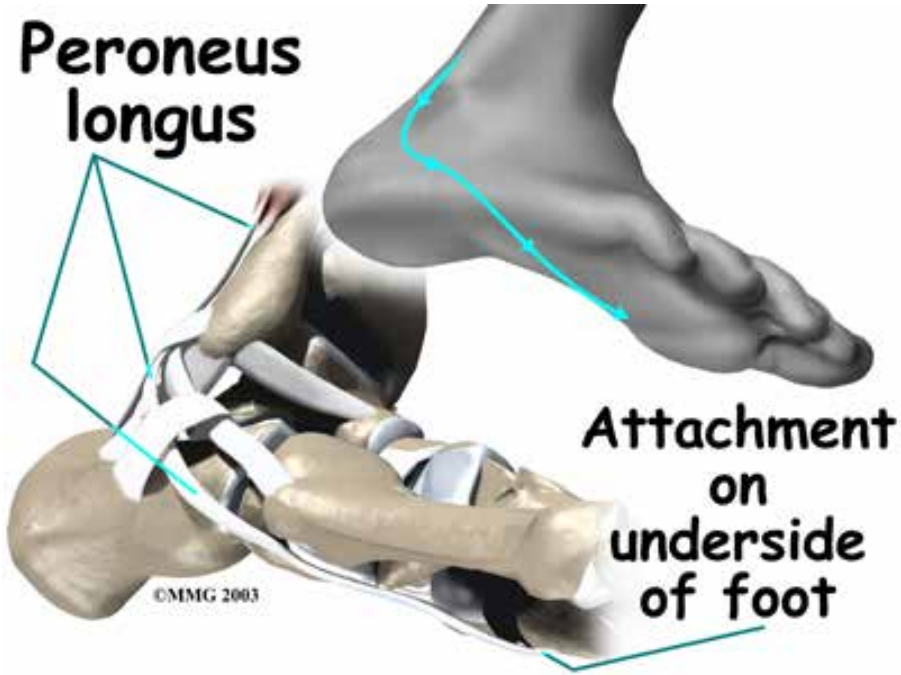
Lower Extremity insight into orthotic management

- A specific sequence of muscle loss results in muscle imbalances which over time develops into the classic CMT deformities of :
  - Intrinsic minus toes – claw toes
  - **Plantar flexed 1st ray**
  - Anterior cavus foot
  - Forefoot adduction at the mid tarsal joint
  - Inverted rear foot with lateral ankle instability
  - External rotation of the ankle and knee axes relative to the line of progression

Actual sequence of muscle loss

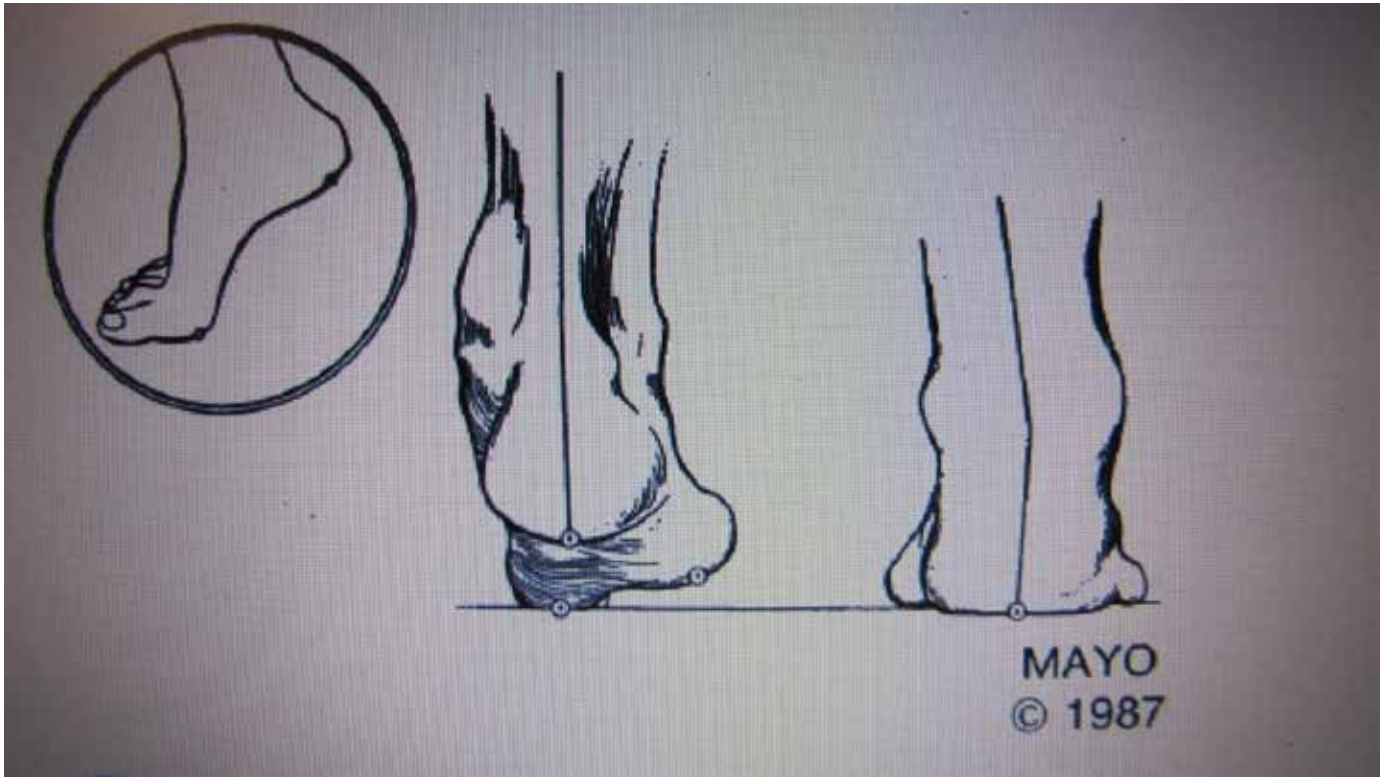
- Intrinsics are first to go leaving extrinsic long toe flexors unopposed to create claw toes
- Peroneus longus out lasts its antagonist anterior tibialis with resultant plantar flexion of the first ray
- A rigidly plantar flexed 1st ray is almost impossible to stretch out and weight bearing imparts an inversion twist to the rear foot creating the “tripod effect”.
- Posterior tibialis out lasts its antagonist the peroneus brevis with unopposed forefoot adduction.
- Long extrinsic toe flexors out last extensors creating anterior cavus

Peroneus Longus
Action: plantar-flex the 1st ray



Tripod Effect

In static weight bearing, a rigid plantar flexed 1st ray imparts an inversion twist to the hind foot with resultant calcaneal varus



Coleman (Lateral) Block Test

Pes Cavus!

Classic Characteristics of the deformity associated with CMT

What does your footprint say?

My footprint!!

- I find wearing my shoes and braces so empowering that I rarely go without them.
- I want you empowered at the end of this presentation.

Classic CMT foot deformity

ERD: External Rotary Deformity

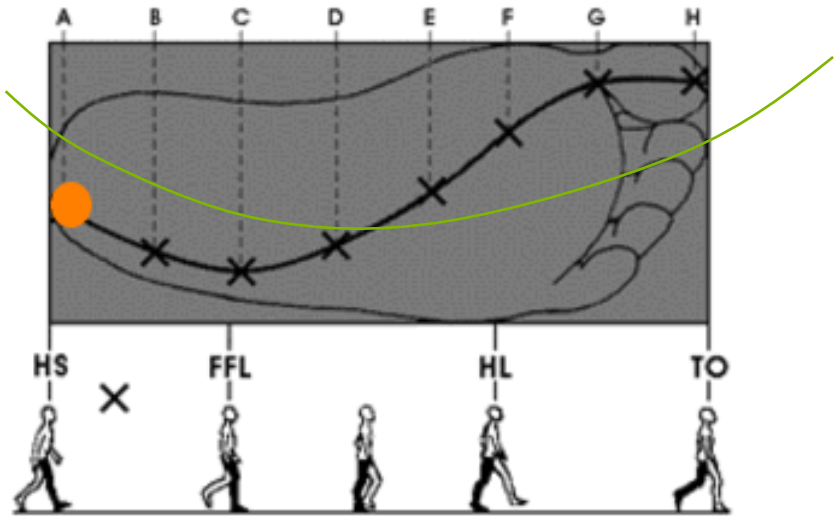
Classic CMT hand deformity

Atrophy of 1st dorsal interrosseus

Flat feet: pes planus IRD

- Less common
- Maybe created from balanced or equal weakness across joints.

Path of Pressure



- Normal Human Locomotion
- COG travels through a 2 inch square for maximum energy efficiency
- Normal Biomechanical Gait

Quality of the base

- An abducted or **adducted** forefoot will have an abnormal path of pressure
- Here as the COG passes over the base it will track lateral and off the base of the 5th metatarsal

Path of pressure.....evidence

Cast for custom AFO

Right foot

Left foot

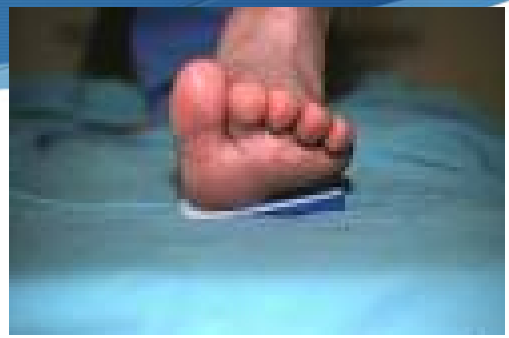
Treatment Options
Which Design is Right ?

- Determine Patient Goals
- device is like a tool

Prevention!!!!

- Simple FO's/SMO's
- Post to prevent lateral ankle instability
- Fore-foot varus post
- ST pad and 1st MP trap, and with weight bearing, prevent shortening of LA
- Stretching exercises to prevent tight achilles tendon, hamstrings

Accommodative/corrective FO's for fixed deformities



Forefoot valgus post for rigid plantar flexed 1st ray

Designs to prevent or slow the characteristics

- Inhibit lateral ankle instability with rear foot or fore foot posting
- Prevent forefoot adduction at mid-tarsal joint
- Prevent shortening of longitudinal arch:
  - trap 1st MP with intrinsic post
  - trap calcaneus with ST pad

Off the Shelf Carbon AFO ’s

Ground Reaction and dorsiflexion assist

Balance - Secondary to Sensory Deficits

- Peripheral neuropathy with proprioceptive deficits and poor balance especially with a history of falls
- Those who claim their balance is fine as long as:
  - They have a finger on a wall
  - Knee against a solid piece of furniture
  - Arm around someone
- Ground Reaction AFO's can dramatically improve balance by transferring forces to proximal segments where those forces are interpreted to let patient know where his center of gravity is over his base

Custom Energy Storing

Align Skeletal Segments Derotation

- ERD External Rotary Deformity
- Forefoot adduction at mid-tarsal joint
- Anterior cavus foot
- ER of ankle and knee axes relative to LOP
- COG not over base of support
- Extended medial foot plate
- Lateral mid-tarsal slot strap
- Lateral sub-talar slot strap
- Lateral calcaneal base modification
- ST pad and intrinsic FF valgus post
- Dorsi-assist ankle joints
- Extended medial proximal trim

samples

Silicone AFO ’s

Limit or enhance motion about a specific joint

- Sagittal plane:
  - Enhance Dorsiflexion
  - Limit Plantar Flexion
- Coronal plane:
  - Limit sub-talar varus
- Transverse plane:
  - Enhance forefoot abduction
  - Limit ER of ankle axis ankle
- Prevent dorsiflexion and provide a ground reaction for proprioceptive loss and improved balance
- Prevent shortening of longitudinal arch

Ground Reaction

"One good leg to stand on"

BALANCE

- Simple physics of balance
- the COG must lie over the base of support

What else?

Lateral Pre-tibial shell

Heel post to accommodate for PF contracture

Do Orthosis make the CMT worse?

No!!!!

Questions???

Thank you very much!!

Question 1

- I am an active person. However, in my current condition, if I walk for more than 5 minutes, my lower leg and thigh begin to really hurt. Will bracing make a difference for me, for example being able to walk for longer periods of time without experiencing pain?

Alignment?

Is it muscle fatigue? Potentially support muscles???

Diet related? Hydrated? Vitamins, electrolites (calcium, sodium, potasium)? Maybe get this checked by your doctor…..

Question 2

- Is there a special brand or make of KAFO for people with CMT? Is so, what should I look for? I have just recently joined a support group in my area and found that the AFO's I was first using were a lot different than the ones worn by others in my group now. They say that theirs is what is recommended for people with CMT. That is what lead to my question. I am on my second pair of KAFO's and they are somewhat different from each other. I have a very serious hyper-extension of the knees (they were measured to be at 20 degrees.) As it is right now I cannot stand in one spot without leaning on my crutches or something sturdy while wearing the KAFO'S.

- I try not recommend specific brands but rather concepts. Look for a **knowledgeable and interested orthotist.**

- Strong to control knee hyper-extension, lightweight (carbon), alignment for standing stability and balance.

- Stance control KAFO?

Question 3

- I'm not sure my braces are a good fit and serving the purpose for which I need, what should I do? I have a slight foot drop but my biggest issue is the rolling out of my right ankle. Even with my AFOs the ankle is still rolling inside the brace. Are silicone AFOs a good option for CMT patients? How can I still be trendy in shoe choices, as a young lady who AFOs.

- Talk to your orthotist. Look for a **knowledgeable and interested orthotist.** Open communication, listen. Establish goals together.

- Need to control pathomechanics.

- I wear the silicone AFO's, but I also wear carbon AFO's. I use custom FO's with each device. I consider a device like a tool……

Question 4

- I have bilateral foot drop as well as my ankles roll. Diagnosis with CMT at age 12 and now am 53. I have worn bilateral AFOs for the last 10 years. Can barely walk without them. My AFOs are custom fit and made of Kevlar. I'm thinking of carbon fiber bracing. What are the benefits and disadvantages? (Besides cost) Also, what do I look for in someone to give me a good fit with bracing? Have had problems with pain with custom bracing before fixing these. Thank you in advance.

- Custom fit vs custom made – discuss
- Kevlar flexible material like a strong canvas. Could be sewn or laminated into a device.
- Carbon fiber allows motion and returns energy. Springy. Can control - support, align, improve balance.
- Talk to your orthotist. Look for a **knowledgeable and interested orthotist.** Open communication, listen. Establish goals together. Take an active role in your treatment!!

Question 5 – comes from the CMTa online community

- I'm new to this site. (Diagnosed w/ CMT in April of 2012) I have good mobility with my CMT only effecting my lower body. At my latest appointment they suggested I get the Ottobock Walk-on Flex AFO's. (As well as in-sole orthotics which I will be getting fit for tomorrow) I tried them on and actually liked them a lot to my surprise. I was just curious if anyone else had any experience with these and if so, how they like them long-term and if they have helped in your everyday life or improvement in your walking?

- Yes – these work well for mild support. Otto Bock now has stronger model called the "Reaction". It offers better push off at toe off.

- Great that you are getting custom FO as well – This is key!!

- Other excellent models are Allard Toe-off & Blue-rocker, Kinetic Research – Noodle (& other variations), Bio-mechanical Composites: Custom Phat brace

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