CMT Guide

A Concept for the Orthotic Treatment of Patients with Charcot-Marie-Tooth

1st edition

Introduction

With the CMT Guide, we present a concept for the orthotic treatment of a peripheral nerve disease for the first time. Even though the indication Charcot-Marie-Tooth disease is not new, there are still issues with the supply with orthopaedic devices.

Choosing the right orthosis appears to be relatively simple: As most patients suffer from a drop foot due to a weakness of the shin muscles (dorsiflexors), they are usually treated with simple, conventional orthoses. If the calf muscles (plantar flexors) are also affected during the course of the disease, the minimal support provided by these devices is no longer sufficient.

Patients are often very dissatisfied, especially with preproduced devices. This supposedly simple approach not only leads to over- or undersupply, but also to fit problems due to foot deformities such as hollow feet, which can even lead to pain when wearing the orthosis.

This guide is intended to help you identify the special features of a weakness in these two muscle groups - the dorsiflexors and the plantar flexors - and to address them in a targeted manner with an individual orthosis. We also highlight the many possibilities of modern orthotics - from the Orthosis Configurator to plug + go modularity, with which you can adapt to many of the special particularities that occur during the course of the disease.

We hope that the CMT Guide will provide you with some guidance on the orthotic treatment of this condition and would be delighted if you could share your experiences with Charcot-Marie-Tooth with us.

Your FIOR & GENTZ team

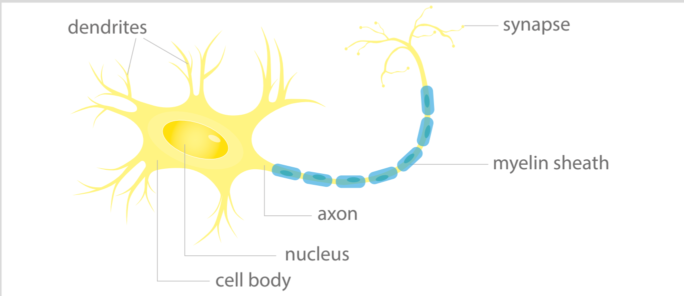
Content

Cover page: patient (CMT), treated with an AFO (NEURO SWING system ankle joint)

What is CMT?

Charcot-Marie-Tooth disease (CMT), also known as hereditary motor sensory neuropathy type I (HMSN I), is a group of hereditary diseases that affect the peripheral nerves and are characterised by a progressive loss of muscle tissue and touch sensation in various body parts.

The peripheral nerves are located outside the central nervous system (spinal cord and brain). These nerves control the muscles and transmit stimuli from the arms and legs to the brain to enable a person to perceive touch. A peripheral nerve consists, among other things, of the axon, which is the interior of the nerve conduction, and the myelin sheath, which forms a protective layer around the axon. CMT can affect either the axon of a peripheral nerve, its myelin sheath or both.



The disease is named after the three physicians who first described it: Jean-Martin Charcot (1825–1893), Pierre Marie (1853–1940) and Howard Henry Tooth (1856–1925). CMT is the most commonly inherited neurological disorder, affecting around one in 2500 people.

Causes

CMT is a genetic disorder caused by mutations in one or more genes. These mutations disrupt the structure and function of the axon and/or myelin sheath of the peripheral nerves, causing them to degenerate and impair the transmission of nerve signals between the brain and the extremities. The affected gene can be inherited from one or both parents. In rare cases, a person can be born with the disease spontaneously without inheriting it from their parents. Each of the five main types of CMT has different causes.

Classification of CMT

CMT1 is the most common form and accounts for about one third of all cases. It is caused by genetic defects that damage the protective myelin sheath and is commonly referred to as demyelinating CMT. Depending on which gene is affected, it is categorised into subtypes A to F:

- CMT1A, caused by a duplication of the PMP22 gene on chromosome 17 (the most common subtype of CMT1),
- CMT1B, caused by a mutation of the MPZ gene on chromosome 1 (the second most common subtype of CMT1),
- CMT1C, caused by a defect in the LITAF gene (rare),
- CMT1D, caused by a defect in the ERG2 gene (rare),
- CMT1E, also known as HNPP, caused by a defect in the PMP22 gene (rare), and
- CMT1F, caused by a defect in the NEFL gene.

CMT2 is caused by defects in a gene that plays an important role in the structure and function of the axon and is commonly referred to as axonal CMT. There are also subtypes of this disease: CMT2A is caused by a mutation in the MFN2 gene and is the most common axonal form of CMT, accounting for 30-40% of cases. Other subtypes are rare and include among others CMT2B, caused by defects in the RAB7 gene, CMT2C, caused by defects in the TRPV4 gene, and CMT2D, caused by defects in the GARS gene.

CMT3, also known as Dejerine-Sottas syndrome, is a rare form caused by defects in the P0 or PMP22 gene.

CMT4 is another rare type that affects the myelin sheath and is usually inherited in an autosomal recessive manner. It begins in early childhood and has several subtypes: CMT4A, caused by mutations in the GDAP1 gene, and CMT4B1, caused by a defect in the MTMR2 gene.

Charcot-Marie-Tooth Disease (CMT)

Symptoms

The first symptoms of CMT usually occur in childhood or early adulthood, in rare cases even later - in some patients as late as the age of 30 or 40. The symptoms start peripherally, i.e. in the distal body parts such as the hands and feet. Since it is a progressive disease, a worsening of the symptoms is to be expected. Proximal areas, i.e. those close to the torso, are also affected. Symptoms include:

- foot deformities such as hollow foot and hammer toes
- drop foot at the onset of the disease (initial symptom)
  - increased knee and hip flexion in the swing phase (stork walk)
  - set down with the toes at initial contact (foot drop)
- loss of muscle tissue and resulting reduction in the circumference of the thigh and lower leg
- muscle weakness in the legs and feet, later in the hands and forearms
  - quick muscle fatigue
  - difficulty standing and walking
  - frequent falling or stumbling
  - reduced ability to walk
- sensory disorders in the arms, legs and feet

Diagnosis

As part of a physical examination, the physician will check for signs of muscle weakness in the hands, arms and feet, foot deformities (e.g. hammer toes or hollow feet) and reduced reflexes. Other examinations include:

- electroneurography (ENG): measurement of nerve conduction velocity (speed and strength of the electrical signals transmitted by the nerves)
- electromyography (EMG): measurement of the electrical activity during muscle contraction
- nerve biopsy: removal and examination of a piece of peripheral nerve taken from the calf
- genetic tests based on blood samples: localisation of the defective gene or genes

Treatment Goal

Currently, there is no cure for the peripheral nerve damage caused by Charcot-Marie-Tooth disease (CMT). Therapy therefore focuses on restoring or maintaining bodily function by treating the symptoms. As the CMT symptoms are strongly centred on the lower extremities, CMT therapy is primarily focused on establishing a basis for painless, efficient and as physiological as possible stance and gait. The physiological gait shown below in its phases can be used as a reference for achieving this treatment goal. CMT therapy can include the following components:

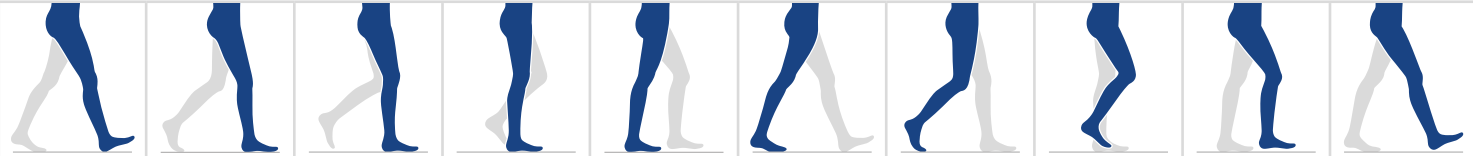
Analgesics: The muscle cramps or nerve damages are often accompanied by pain. Pain-relieving medication enables painless mobility without relieving postures or compensation mechanisms.

Physiotherapy/Occupational Therapy: Targeted and gentle exercises can help to strengthen and stretch the muscles and prevent muscle tension and progressive muscle atrophy.

Surgical Procedures: Severe foot deformities often develop as a result of the disease and lead to severe limitations. In certain cases, surgeries improve the biomechanical situation of the foot and prevent foot deformities from worsening.

Orthopaedic Devices: Medical devices such as orthoses are intended to provide stability when standing and walking. Many lightweight devices have actually been designed to keep a drop foot in a neutral position during the swing phase and thus enable the leg to swing freely.

Division of Physiological Gait into Different Phases According to Jacquelin Perry



Term (Abbreviation)									
initial contact (IC)	*loading response (LR)*	*early mid stance (MSt)*	*mid stance (MSt)*	*late mid stance (MSt)*	*terminal stance (TSt)*	*pre swing (PSw)*	*initial swing (ISw)*	*mid swing (MSw)*	*terminal swing (TSw)*
Percentage of Stride									
0%	0–12%	12–31%			31–50%	50–62%	62–75%	75–87%	87–100%
Hip Angle									
20° flexion	20° flexion	10° flexion	neutral position	5° extension	20° extension	10° extension	15° flexion	25° flexion	20° flexion
Knee Angle									
0-3° flexion	15° flexion	12° flexion	8° flexion	5° flexion	0-5° flexion	40° flexion	60° flexion	25° flexion	0-2° extension
Ankle Angle									
neutral position	5° plantar flexion	neutral position	5° dorsiflexion	8° dorsiflexion	10° dorsiflexion	15° plantar flexion	5° plantar flexion	neutral position	neutral position

Conventional Orthoses

Until now, the orthotic treatment has mainly been using lightweight medical devices such as insoles, bandages or mostly preproduced ankle-foot orthoses. Every device offers a certain benefit. However, the patient must also be aware of the disadvantages.
Bandages are provided to patients in various designs and materials, mainly as preproduced devices. The goal is to gently support the upper ankle joint to keep the foot in an approximately neutral position during the swing phase. However, in many cases the dorsiflexion-assist effect of these devices is insufficient. A stabilisation beyond this cannot be achieved without substantially restricting mobility in the ankle joint.

Bandages

Posterior-leaf-spring (PLS) AFOs are also made of either polypropylene or carbon fibre. In the Achilles tendon area, a narrow spring (made of polypropylene or carbon fibre) connects the foot piece and the posterior lower leg shell. This connection enables motion between the foot and lower leg. While lightweight and preproduced PLS AFOs made of polypropylene are used as simple foot lifter orthosis, PLS AFOs with strong carbon springs are used for patients with a weakness of the plantar flexors. All PLS AFOs allow a certain range of movement in the upper ankle joint. However, these are unphysiological motions, as there is no defined pivot point.

Posterior-leaf-spring AFO

Rigid AFOs are available with either an anterior or posterior shell. These are custom-made or preproduced AFOs made of either polypropylene or carbon fibre. Generally, rigid AFOs are used when the stabilisation of the knee and ankle joint in swing phase by lifting the foot is desired. Both foot lifting and joint stabilisation are achieved due to the rigid construction and the complete blocking of the range of motion in the upper ankle joint. However, this does not achieve a physiological gait.

SAFO FRAFO

Spiral orthoses made of carbon fibre are a new type of device. The spiral of this individually produced device can be adapted to the patient and lifts the foot in swing phase in the case of isolated peroneal palsy. Depending on the rigidity of the spiral, the alignment of the carbon fibres allows a range of motion in the direction of dorsiflexion and supports the swing phase initiation. However, a stabilisation of the knee and ankle joint cannot be achieved if the plantar flexors are weak. A spiral orthosis cannot be adjusted to the course of the disease at a later date.

Spiral Orthosis

Disadvantages of Conventional Orthoses

All currently available treatments can result in a successful therapy, but they can also have a negative effect, as each design not only has advantages, but also disadvantages. The characteristic symptoms of Charcot-Marie-Tooth (CMT) lead to recurring problems with conventional orthoses:

1. **Fit Problems**
   Since CMT is associated with foot deformities, many preproduced orthoses often have fit problems. Depending on how severe the foot deformities are, these issues can lead to uncomfortable wear, skin irritations or abrasions, or even pain. An orthosis must therefore always be custom-made for the patient based on the cast.
2. **Lack of Adjustment Options**
   CMT starts with problems due to the failure of the dorsiflexors. Many lightweight orthoses are therefore designed to compensate for a drop foot and allow the leg to swing freely. If the plantar flexors are also affected during the course of the disease, such devices do not provide enough stability to secure the knee and ankle joint. The lack of adjustment options means that it cannot be adapted. An orthosis must therefore always be adjustable.
3. **Lack of Range of Motion**
   If only the dorsiflexors are affected by the disease, the complete blocking of the range of motion in the upper ankle joint results in major restrictions when walking. This oversupply reduces the patient's acceptance of the device. If the plantar flexors are weak, stabilisation of the knee and ankle joint is often achieved by blocking the range of motion, which also prevents a physiological gait.

Requirements for an Orthosis

An unrestricted swing ensures motion without stumbling. Therefore, the most important requirement for an orthosis for CMT patients is that it keeps the foot in an approximately neutral position during swing phase. This position allows the affected leg to swing freely without the patient developing compensation mechanisms. Compensation mechanisms should be prevented, as they result in higher energy consumption when walking and put excessive strain on other body structures (see appendix). Beyond this minimum requirement, the following should be ensured:

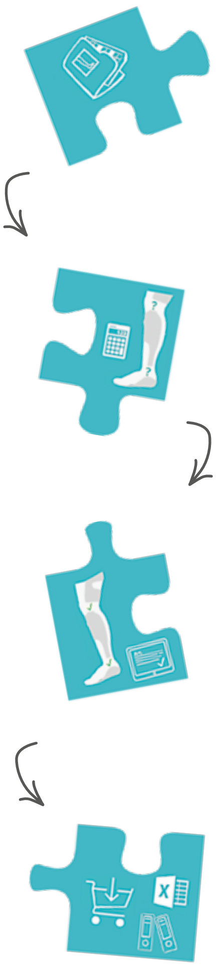
1. **Maintaining Mobility**
   As CMT is characterised by a loss of muscle tissue, additional atrophy caused by the complete immobilisation of the anatomical ankle joint in a rigid orthosis must be prevented. Orthoses must also support physiotherapy focused on muscle training and must not impede the success with such immobilisation. Orthoses must therefore allow range of motion in the upper ankle joint.
2. **Ensuring Stability**
   When standing and walking, the forefoot lever generated by the plantar flexors provides the necessary stability in the knee and ankle joint. If the disease affects not only the dorsiflexors, but also the plantar flexors, the forefoot lever is activated externally via a mechanical dorsiflexion stop. Orthoses must therefore provide sufficient resistance to the dorsiflexion when standing and walking.
3. **Enabling Adjustments**
   The symptoms of CMT progress. Even if only the dorsiflexors are initially affected, the muscle weakness can also extend to the plantar flexors at a later stage. Orthoses must be adaptable to the course of the disease to meet the changing support requirements.

To determine the level of support provided by a custom-made orthosis and to avoid under- or oversupply, the impaired muscle groups must be identified before planning the treatment. In CMT patients, the dorsiflexors often are affected first, which is characterised by an isolated foot drop and the compensation mechanisms developed in this context. A visual gait analysis, in which the patient's gait is examined for deviations from the physiological gait, can indicate whether the plantar flexors are also affected. You will find more information on the physiological stance and gait as well as deviations due to weakness of the dorsiflexors and plantar flexors in the following chapters. A muscle function test provides more detailed information about the condition of the muscle groups that may be affected. This is because the weakness of a muscle group does not always have a visible effect on standing and walking.

Orthosis Configurator

In addition to the muscle strength of the six major muscle groups in the leg, further patient data is collected as part of a comprehensive examination during the planning of the orthosis. This data is important for calculating the required functionality, the level of support and the expected load on the orthosis. The FIOR & GENTZ Orthosis Configurator does this calculation for you. During the configuration, you will receive recommendations on the orthosis type, construction, system joints, system width, the spring units to be used if applicable and many other orthosis data.

With the Orthosis Configurator, you can create a reproducible orthosis and save the orthosis data – an important element for your documented treatment. Use the completed orthotic treatment sheet and visit the Orthosis Configurator via our website or at www.orthosis-configurator.com. You will then be guided through the following steps:



1 Patient Data

In the first step, you enter all patient data that are relevant for planning your orthosis.

2 System Components

In this central step, you receive recommendations regarding the orthosis' design and choice of system components. The recommendations are functionally adjusted to the patient data and will withstand all expected loads.

3 Individual Adjustments

In the third step, you can adapt the shape and material of your system joints.

4 Result

In the last step, you can save or send your configuration result or print it for your treatment documentation. You can furthermore generate a calculation and order products directly from our webshop.

Orthosis Configurator

System Knee Joints with plug + go MODULARITY

Flexible Treatment now also at Knee Level

plug + go Modularity

As the disease progresses, the plantar flexors can also be affected in addition to the dorsiflexors. In these cases, CMT patients require a higher level of support to stabilise the knee and ankle joint. Such orthoses require functional elements that provide a dynamic resistance against the dorsiflexion. The plug + go modularity allows adaptation to changes caused by the disease without the need for a new orthosis. Thanks to the wide range of system ankle joints available from FIOR & GENTZ, the orthosis can be optimally adjusted to the patient's requirements.

In addition to the system joints listed below, other system ankle joints are also compatible with plug + go modularity. Under specific conditions, the NEURO CLASSIC-SPRING system ankle joint and the NEURO CLASSIC-SWING system ankle joint can also be used within the context of plug + go modularity.

System Ankle Joints with plug + go MODULARITY

The Dorsiflexors – Function and Pathology

Function of Healthy Dorsiflexors

The shin muscles are known as dorsiflexors because they cause dorsiflexion in the upper ankle joint. With a physiological gait, the dorsiflexors are active from pre swing to loading response. Over the duration of their activity, they perform different functions to stabilise the anatomical ankle joint, for foot lifting and for shock absorption during load bearing, thereby carrying out three different types of work.

Concentric Muscle Work of the Dorsiflexors

In pre swing, the dorsiflexors work against the activity of the plantar flexors through concentric muscle work and thus stabilise the upper ankle joint. From initial swing, the foot is brought into an approximately neutral position to prepare the leg for the swing phase.

Isometric Muscle Work of the Dorsiflexors

From mid swing to initial contact, the dorsiflexors keep the foot in an approximately neutral position through isometric muscle work. In swing phase, this position results in:

- a straight swinging of the leg
- a physiological knee joint angle of about 60°
- an upright body posture
- an initial contact with the heel

Eccentric Muscle Work of the Dorsiflexors

In loading response, the foot is lowered in a controlled manner in the direction of plantar flexion by eccentric muscle work of the dorsiflexors, stretching the muscle in a controlled manner. This mechanism significantly contributes to the body's shock absorption after initial contact and leads to:

- a controlled load bearing
- a flexion movement of the knee
- a physiological knee joint angle of about 15°

Effects of Weak Dorsiflexors

Problems during Push Off

Initial Swing
The weak dorsiflexors cannot bring the foot into a neutral position in initial swing to support the swing phase initiation. The push off is impaired. The toe can be lifted off the ground as follows:

- unphysiological knee joint angle of more than 60°
- excessive lifting of the pelvis on the affected side
- inclination of the upper body to the contralateral side

Drop Foot in Swing Phase

Mid Swing to Terminal Swing
No physiological foot lifting continues to occur after mid swing. The noticeably hanging foot leads to compensation mechanisms that are intended to achieve a swinging of the leg without stumbling (see appendix):

- increased hip and knee flexion (stork walk)
- lateral lifting of the pelvis (Hip Hiking)
- excessive hip abduction (circumduction)

In mid swing, a drop foot can lead to stumbling.

Incorrect Load Bearing (Activity Level 1 and 2)

Loading Response
Due to a drop foot, the forefoot touches the ground first in the case of a low activity level during initial contact, instead of the heel. This results in a resulting shortened stride length. The load bearing is reversed (foot drop):

- lowering of the heel and a dorsiflexion motion in the upper ankle joint
- extension movement and unphysiological knee extension
- insufficient shock absorption as a result

Uncontrolled Load Bearing (Activity Level 3 and 4)

Loading Response
In patients with a high activity level who take particularly large steps, initial contact is made with the heel despite the drop foot. However, the weak dorsiflexors cannot control the load bearing in loading response:

- too quick lowering of the foot
- audible slapping sound when the foot touches the ground
- unphysiological knee extension

Stance

Secure Stance due to Healthy Plantar Flexors

The calf muscles are known as plantar flexors because they cause plantar flexion in the upper ankle joint. The plantar flexors play an important role in the dynamic stance by activating the forefoot lever and thereby keeping the body's centre of gravity above the feet. The forefoot lever is the area between the ankle pivot point and the rolling-off line and forms the supportive area on the ground. The body's centre of gravity can be safely shifted forwards and backwards above the supportive area. The further the body is inclined forwards, the greater the torque in the ankle joint and therefore the greater the force to be applied by the plantar flexors. As long as the body's centre of gravity is above the supportive area, healthy plantar flexors keep the body in a stable balance. When balancing the body's centre of gravity over the supportive area, the physiological knee joint angle is approximately 0° to 5°.

Insecure Stance due to Weak Plantar Flexors

Due to weak plantar flexors, their muscle strength is reduced. The lower the muscle strength, the less the forefoot lever can be activated by the plantar flexors. The lower the muscle strength, the smaller the supportive area.

fig. 1

If the plantar flexors are completely paralysed, the forefoot lever also cannot be activated. As a result, there is no supportive area and the body weight cannot be shifted forwards. Consequently, the body can only be balanced in an unstable and unphysiological position exactly above the ankle pivot point. Shifting the body weight forwards would result in a fall. If only the plantar flexors of one leg are weakened, the patient can stand but tends to overextend the weaker leg (fig. 1). The resulting pathological knee joint angle temporarily improves stability, but permanently overstrains the ligaments in the knee as time goes on. This results in health problems.

Gait

Physiological Gait due to Healthy Plantar Flexors

During the stance phase of walking, from mid stance to pre swing, the plantar flexors are active and contribute to stabilising the knee and ankle joint as well as initiating the swing phase (push off). In mid stance, healthy plantar flexors stabilise the upper ankle joint by controlling the forward motion of the tibia and thus the dorsiflexion through eccentric muscle work. A physiological knee joint angle of 0° to 5° is maintained during this gait phase due to the knee-spanning m.gastrocnemius. The stabilisation of the knee joint continues in terminal stance. The plantar flexors activate the anatomical forefoot lever, which stabilises the upper ankle joint, thereby causing the heel to lift off the ground and lift the body weight. Lifting the body's centre of gravity significantly contributes to a fluid and energy-efficient gait. In pre swing, the plantar flexors initiate the swing phase (push off) through concentric muscle work and the resulting active plantar flexion (fig. 4).

Pathological Gait due to Weak Plantar Flexors

In stance phase, weak plantar flexors cannot stabilise the upper ankle joint. To compensate for the loss of stability, the knee is hyperextended from mid stance (fig. 2). During terminal stance, the forefoot lever cannot be activated to counteract the ground reaction force. As a result, the heel and therefore the body weight is not lifted (fig. 3), which greatly increases energy consumption during walking. The weak plantar flexors cannot perform active plantar flexion in pre swing, which prevents the physiological push off. In loading response, the knee shows excessive flexion on the contralateral side (fig. 4).

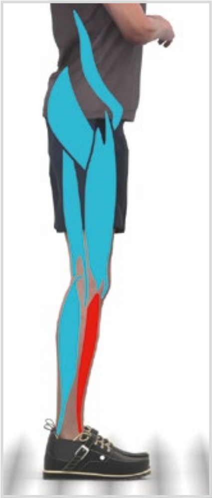
fig. 2

fig. 3

fig. 4

Appearance

Due to the damage to the peripheral nerves, CMT patients initially mainly suffer from a weakness of the dorsiflexors, which results in insufficient foot lifting. It appears as:



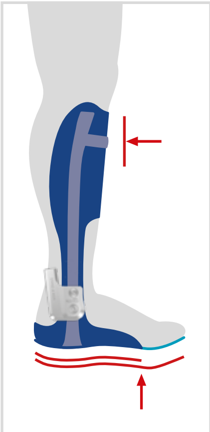
- problems during push off
- drop foot in swing phase
- compensation mechanisms

A muscle function test provides information about the severity of the muscle weakness. If only the dorsiflexors are affected by the disease, treatment with a foot lifter orthosis is sufficient. In order for this orthosis to allow the patient to walk without restrictions, it should:

- be custom-made
- allow range of motion in the upper ankle joint
- be adaptable to the course of the disease

Recommended Orthosis

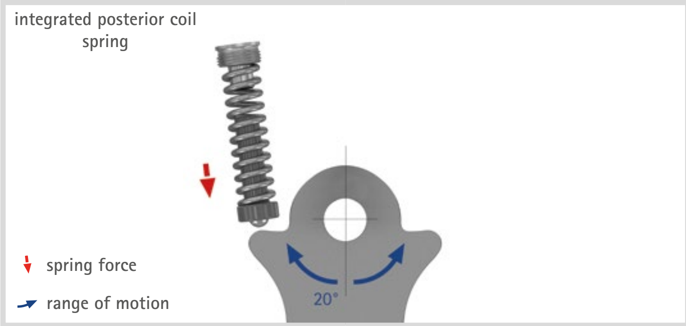
A dynamic AFO with high anterior shell, long and partially flexible foot piece (rigid sole with flexible toe area) and NEURO CLASSIC-SPRING system ankle joint is reccomended.



The NEURO CLASSIC-SPRING system ankle joint is equipped with an integrated coil spring with a normal spring force and 20° range of motion.

The NEURO CLASSIC-SPRING System Ankle Joint

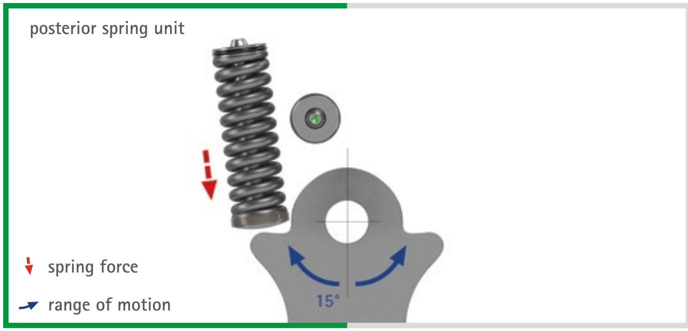
A NEURO CLASSIC-SPRING system ankle joint with plug + go modularity can be converted to any other system ankle joint with plug + go modularity by exchanging the functional unit.



Note: if the patient takes particularly large steps, initial contact with the heel can be achieved despite weak dorsiflexors (e.g. with an activity level of 3 or 4). In this case, an orthosis with a NEURO CLASSIC-SWING system ankle joint should be fitted.

The NEURO CLASSIC-SWING System Ankle Joint

The spring unit of the NEURO CLASSIC-SWING system ankle joint enables the foot to be lifted and lowered in a controlled manner in loading response. The support provided by the spring unit depends on the muscle strength of the dorsiflexors. Carry out a comprehensive muscle function test and enter the data in the Orthosis Configurator. The Orthosis Configurator provides a recommendation for the spring unit to be used.



Physiological Gait with an AFO

An AFO (ankle-foot orthosis) with a NEURO CLASSIC-SPRING system ankle joint is a common custom-made foot lifter orthosis. The mechanical pivot point of the otherwise rigid AFO is aligned with the pivot point of the anatomical ankle joint.

Initial Swing: The foot lifter orthosis already brings the foot into a neutral position during push-off and the leg can be accelerated into forward motion (fig. 1).

Mid Swing to Terminal Swing: An AFO with a NEURO CLASSIC-SPRING system ankle joint keeps the foot in a physiological foot lifting and leads to a physiological knee joint angle of 60°. The foot lift allows the leg to swing straight. Compensation mechanisms are thus avoided (fig. 2).

Initial Contact: The foot lifter function of an AFO with a NEURO CLASSIC-SPRING system ankle joint ensures that the heel touches the ground first during initial contact (fig. 3).

Loading Response: The correct mechanical pivot point of the orthosis and the physiological heel strike during initial contact ensure physiological, passive plantar flexion in the correct direction of ankle movement. This correct rotation supports the neurological reactivation of the dorsiflexors and leads to a physiological knee flexion angle of around 15° (fig. 4).

fig. 1

fig. 2

fig. 3

fig. 4

Information about the AFO

Activity Level 1 and 2: The coil spring of the NEURO CLASSIC-SPRING system ankle joint lifts a drop foot in swing phase, causing the heel to touch the ground first during initial contact. Due to the shortened stride length, the spring force of the coil spring of this system ankle joint with dorsiflexion assist is sufficient to enable a physiological load bearing in loading response.

Should the alignment of the finished orthosis need to be modified or adapted, a NEURO VARIO-SPRING 2 system ankle joint may be appropriate. In addition to the same functions as the NEURO CLASSIC-SPRING system ankle joint, it is equipped with an adjustable dorsiflexion stop.

Activity Level 3 and 4: With the NEURO CLASSIC-SWING system ankle joint, the resistance for controlled lowering of the foot can be adjusted using the exchangeable precompressed spring units. Controlling the passive plantar flexion has a positive effect on a physiological knee joint angle in loading response.

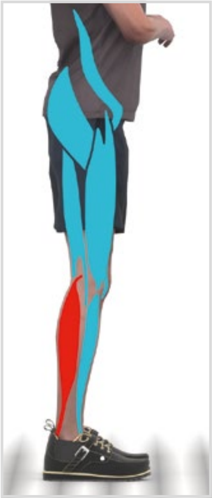
Should the alignment of the finished orthosis need to be modified or adapted, a NEURO VARIO-SWING system ankle joint may be appropriate. In addition to the same functions as the NEURO CLASSIC-SWING system ankle joint, it is equipped with an adjustable dorsiflexion stop.

Depending on the muscle strength and whether other muscle groups are affected by paralysis, a different type of orthosis with different system joints may be appropriate. Use the Orthosis Configurator from FIOR & GENTZ to configure the optimal orthosis with a system joint in the appropriate system width as well as all necessary components and materials.

Orthosis Configurator

Appearance

In severe cases, the damage to the peripheral nerves can also affect the plantar flexors in addition to the dorsiflexors. Due to the weak plantar flexors, the activation of the anatomical forefoot lever is impaired:



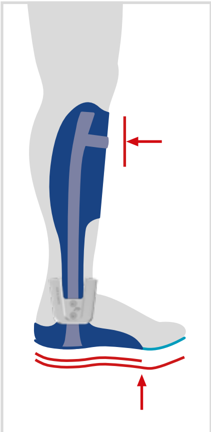
- missing stabilisation in mid stance
- no heel lift in terminal stance
- no lifting of the body weight

A muscle function test provides information about the severity of the muscle weakness. As the disease affects not only the dorsiflexors but also the plantar flexors, treatment with a foot lifter orthosis is no longer sufficient. An orthosis must contain functional elements that stabilise the patient's knee and ankle joint. Furthermore, it should:

- be custom-made
- allow range of motion in the upper ankle joint
- be adaptable to the course of the disease

Recommended Orthosis

A dynamic AFO with high anterior shell, long and partially flexible foot piece (rigid sole with flexible toe area) and NEURO SWING system ankle joint is recommended.



Spring units to be used:

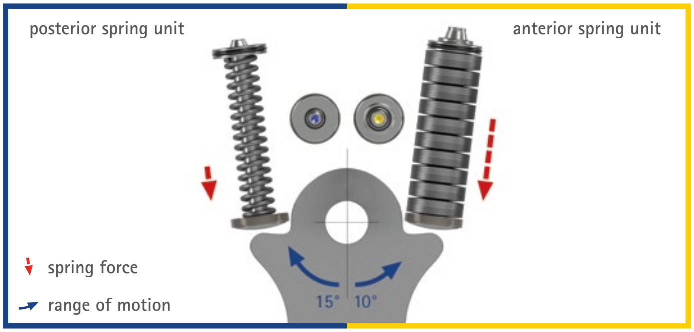
- posterior: blue marking (normal spring force, max. 15° range of motion)
- anterior: yellow marking (very strong spring force, max. 10° range of motion)

The NEURO SWING System Ankle Joint

Individual adjustment to the pathological gait by:

- exchangeable spring units;
- adjustable alignment;
- adjustable range of motion.

All three adjustment options can be chosen separately. They do not influence each other.



The recommended spring units represent a possible initial treatment. The support of the AFO can be adjusted via the spring forces of the spring units. It is important to avoid an oversupply and only select as much spring force as necessary.

The support provided by the spring units depends on the muscle strength of the dorsiflexors and plantar flexors. Carry out a comprehensive muscle function test and enter the data in the Orthosis Configurator. The Orthosis Configurator provides a recommendation for the spring units to be used.

Orthosis for Additional Weak Plantar Flexors

Physiological Gait with an AFO

A custom-made AFO with a NEURO SWING system ankle joint is equipped with a dynamic dorsiflexion stop via precompressed spring units. This AFO activates the forefoot lever and enables the patient to stand securely and restore a physiological knee joint angle of 0° to 5° (fig. 1).

Mid Stance: From late mid stance, the high resistance of the anterior spring unit of the NEURO SWING system ankle joint generates a knee extension moment, stabilising the knee and upper ankle joint. The patient can apply their body weight to the orthosis via the anterior lower leg shell, which prevents hyperextension of the knee (fig. 2).

Terminal Stance: The yellow spring unit of the NEURO SWING system ankle joint is strong enough to activate the forefoot lever externally and lift the heel (fig. 3). The heel lift raises the body's centre of gravity, which enables a physiological knee extension of the contralateral leg (fig. 4). This allows for a fluid gait with a reduced energy consumption.

Pre Swing: The energy brought into the anterior spring unit from late mid stance is then released until the basic adjustment is achieved, supporting push off (fig. 4).

fig. 1

fig. 2

fig. 3

fig. 4

Information about the AFO

For weak or completely paralysed plantar flexors, the optimal treatment consists of a custom-made AFO with a mechanical system ankle joint and dorsiflexion stop. The NEURO SWING system ankle joint with a dynamic dorsiflexion stop and precompressed spring units is the most suiteable. This AFO enables a safe stance. When walking, the range of motion in the upper ankle joint restores a physiological gait and the strength of the healthy muscles is maintained. At the same time, it prevents a hyperex-tension of the knee.

If the plantar flexors are weak, an AFO with a NEURO SWING system ankle joint can completely replace conventional walking aids, such as crutches and walkers, and also enable safe, free-handed standing and walking. If necessary, the NEURO SWING system ankle joint can be used to adjust the spring force, alignment and range of motion separately.

With a NEURO SWING FIT AFO test orthosis, you can check the extent to which your patient will benefit from a NEURO SWING system ankle joint without producing a custom-made AFO first. The NEURO SWING FIT AFO test orthosis with a pre-mounted NEURO SWING Carbon system ankle joint is a preproduced orthosis. It is used as a test orthosis prior to treatment with a custom-made AFO, in which a system ankle joint with dynamic stops (e.g. NEURO SWING) is mounted.

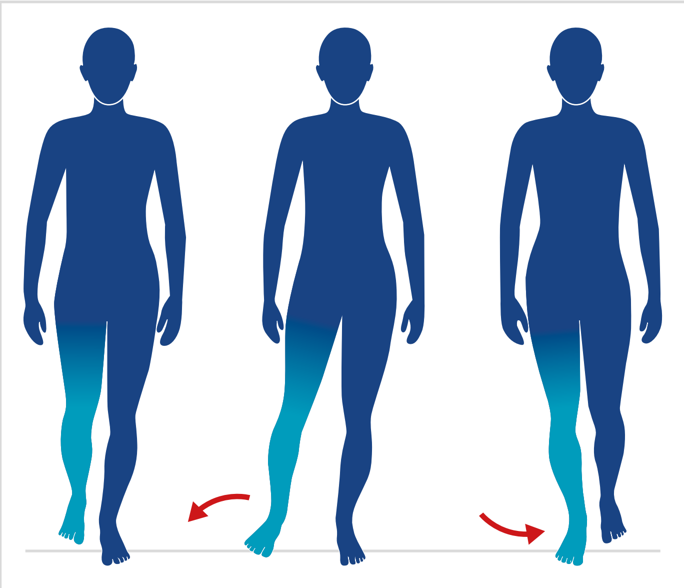
Depending on the muscle strength and whether other muscle groups are affected by paralysis, a different type of orthosis with different system joints may be appropriate. Use the Orthosis Configurator from FIOR & GENTZ to configure the optimal orthosis with a system joint in the appropriate system width as well as all necessary components and materials.

Orthosis Configurator

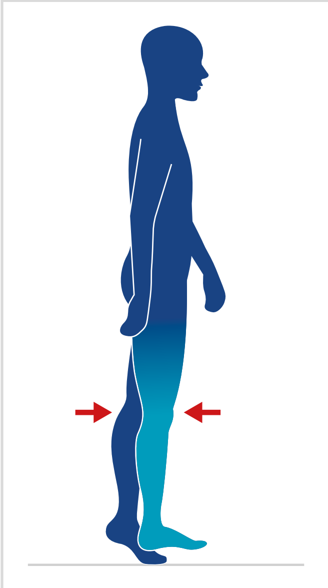
The swing leg must be effectively shortened to enable a forward movement without stumbling when walking normally. This requirement is met by a physiological hip and knee flexion as well as dorsiflexion during swing phase.

In certain gait disorders, this shortening of the swing leg is impaired, e.g. in the case of a failure of the hip or knee flexors. If the dorsiflexors fail, the swing leg is effectively extended due to an increased plantar flexion during swing phase. When wearing a locked KAFO, the permanent locking of the knee joint also prevents knee flexion.

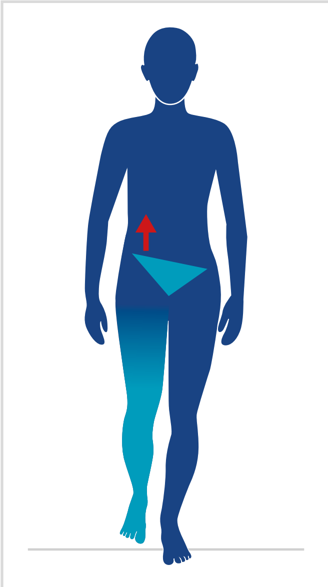
The body can compensate for this lack of functional shortening in swing phase in three different ways:



Circumduction
During swing phase, the leg is brought forward in a semicircular motion around the supporting leg. During this motion, external rotation occurs in the hip joint. In the long term, this motion can manifest itself and cause hip problems.



Vaulting
This compensation mechanism describes the contralateral plantar flexion. Since the affected leg is effectively extended or cannot be flexed, the contralateral supporting leg is extended to allow the swinging through.



Hip Hiking
Hip hiking refers to the excessive lifting of the pelvis on the swing leg side. This enables the extended swing leg the space to swing through without stumbling.

Abduction
(from Latin *abducere* = to withdraw, lead away): movement of a body part away from the centre of the body. The countermovement of adduction.

AFO
(ankle-foot orthosis): term for an orthosis encompassing the ankle joint and the foot

Autosomal Recessive
In autosomal recessive inheritance, unaffected parents pass on one mutated gene from the father and one from the mother to the patient. The disease only occurs if the same mutation is found in a specific gene on both chromosomes from 1-22.

Axon
(from Greek *axon* = axis): extension of a nerve cell. Transmits electrical impulses from the cell body to other nerve cells. The unit of axon and surrounding myelin sheath is called nerve fibre.

Carbon Spiral Orthosis
↑AFO made of carbon fibre that wraps around the lower leg in a spiral. The alignment of the fibres gives this orthosis particularly dynamic properties.

Charcot-Marie-Tooth
(abbr. CMT); also known as hereditary motor sensory neuropathy type I (HMSN I): disease of the peripheral nervous system with a variety of clinical and genetic causes and forms. CMT is a progressive disease with symptoms starting in the ↑distal body parts (hands and feet).

Chromosome
Structure that is located inside every cell and contains genes. Each body cell contains 46 chromosomes, arranged in 23 pairs. Half of the human chromosomes and therefore half of the genes come from one parent each.

Compensation Mechanism
(from Latin *compensare* = to compensate, to replace): compensation or replacement of a missing ↑physiological movement to achieve a certain goal. A deficient foot lift or knee flexion during the swing phase can be compensated by various mechanisms to achieve the goal (here: swing-through of the leg).

Concentric
(from Latin *con* = with; *centrum* = centre): moving towards a centre; having a common centre. In a mechanical context this means that the force is applied exactly in the centre. In a ↑physiological context, a muscle performs concentric work by shortening itself and thus causing a joint movement.

Contralateral
(from Latin *contra* = against; *latus* = side, flank): found on the opposite side of the body

Demyelinating
refers to the loss of the ↑myelin sheath.

Distal
(from Latin *distare* = to be distant): denoting a position away from the centre of the body. The opposite of distal is ↑proximal.

DNA
Deoxyribonucleic acid: a substance made up of different building blocks (deoxyribonucleotides) that contains the genetic information of living organisms and certain viruses

Dorsiflexion
Lifting of the foot or reduction of the angle between lower leg and foot. It is called dorsiflexion because of this movement (↑flexion). Functionally, however, it is a stretching movement in the sense of an ↑extension. The countermovement of ↑plantar flexion.

Dorsiflexion Stop
Constructional element of an orthosis that limits the degree of ↑dorsiflexion. The dorsiflexion stop activates the ↑forefoot lever, thereby creating an area of support. Furthermore, a dorsiflexion stop causes, together with the orthosis' foot piece, a knee extension moment and, starting at terminal stance, a heel lift.

Dorsiflexors
Colloquially known as the shin muscles. Muscles causing the lifting of the foot.

Drop Foot
Malfunction that prevents the foot from being actively extended or lifted and therefore the foot passively hangs down during swing phase. This malfunction is caused by peroneal palsy or a weakness of the dorsiflexors.

Dynamic
(from Greek *dynamikos* = active, strong): displaying movement, characterised by momentum and energy. Thus, a dynamic ↑AFO enables a movement in the anatomical ankle joint.

Eccentric
(from Latin *ex* = outside; *centro* = centre): located outside of a centre or away from a centre. In a mechanical context this means that the force is applied off-centre. In a ↑physiological context, a muscle performs eccentric work by actively extending itself and controlling a joint movement by decelerating it.

Extension
(from Latin *extendere* = to extend): active or passive straightening of a joint. Straightening is the countermovement of bending (↑flexion) and characteristically increases the joint angle.

Flexion
(from Latin *flectere* = to bend): active or passive bending of a joint. Bending is the countermovement of straightening (↑extension) and characteristically reduces the joint angle.

FRAFO
(floor reaction AFO): solid orthosis with an anterior shell which provides a knee or hip extension moment starting at terminal stance. FRAFOs can be made of either polypropylene or carbon fibre. They either have a rigid or a partially flexible foot piece. However, the name FRAFO is misleading since other ↑AFOs also interact with the ↑ground reaction force.

Forefoot Lever
anatomical lever arm running from the upper ankle joint to the metatarsophalangeal joints of the toes

Foot Drop
Pathologically altered manner of walking: By placing the tip of the foot on the ground, this gait looks like the movements of a stepper foot on a sewing machine.

Functional Element
Component of a system ankle joint that is responsible for the movement performed with the orthosis. For example, the orthosis allows, blocks or dynamically controls a movement.

Gene
A sequence of DNA that contains the blueprint for the production of proteins. The proteins created according to this blueprint are the basis for the functions of a living organism.

Ground Reaction Force
(abbr. GRF): force generated in the ground as a counterreaction to the body weight. The ground reaction force vector is a theoretical line representing the size, origin and direction of action of the ground reaction force.

Hammer Toes
deformity of the toes in which the metatarsophalangeal joint of a toe is hyperextended and the medial metatarsophalangeal joint is severely flexed, while the distal phalanx remains extended and points towards the ground

Hollow Foot (Foot Deformities)
Foot deformity in which the longitudinal arch of the foot is lifted upwards and the back of the foot is raised higher than normal. Often ↑hammer or claw toes develop simultaneously. Due to the hollow foot, the body weight is carried by a smaller proportion of the foot sole when standing and walking and the ball of the foot is therefore subjected to increased pressure.

Isometric
(from Greek *iso* = equal; *metros* = measurement): maintaining the same linear extension. Isometric muscle work is a type of muscle contraction without a change in length. No joint motion is caused.

KAFO
(knee-ankle-foot orthosis): term for an orthosis encompassing the knee, the ankle joint and the foot

Muscle Atrophy
(from Greek *atrophia* = depletion, emaciation): visible decrease in the circumference of a skeletal muscle due to reduced strain

Muscle Strength
Muscle strength is a parameter used to assess the force generated by a muscle group (e.g. knee flexors). This force is determined by the muscle function test (according to Janda), which tests each muscle group to assess the extent to which each respective movement can be performed. The muscle strength is classified on a six-level scale depending on whether or not the subject is able to overcome manually applied resistance or gravity:

0 (zero)	total paralysis, no evidence of contraction
1 (trace)	slight contraction, but no joint motion
2 (poor)	complete range of motion with gravity eliminated
3 (fair)	complete range of motion against gravity
4 (good)	complete range of motion against gravity with some resistance
5 (normal)	complete range of motion against gravity with full resistance

Mutation
(from Latin *mutare* = to change/alter, transform): a spontaneously occurring, permanent change in the genetic material (biology)

Myelin Sheath
(from Greek *myelos* = marrow): also called marrow sheath. A protective layer consisting of proteins and fats, which surrounds part of the nerve cell extensions (↑axons) of vertebrates in a spiral manner. This layer enables the nerve cells to transmit stimuli quickly.

Neurological
(from Greek *neuron* = nerve; *logos* = doctrine): concerning the nervous system

Pathological
(from Greek *pathos* = pain; disease): abnormally (changed)

Peripheral
(from Greek *peripherēs* = turning round): located in the outer zones of the body. The peripheral nervous system is the part of the nervous system that is not part of the brain and spinal cord.

Peroneal Palsy
damage to the peroneal nerve (calf bone nerve), which causes paralysis of the ↑dorsiflexors

Physiological
(from Greek *physis* = nature; *logos* = doctrine): concerning the natural life processes

Plantar Flexion
Lowering of the foot or increase in the angle between lower leg and foot. Countermovement of ↑dorsiflexion.

Plantar Flexors
Colloquially known as calf muscles. Muscles causing the lowering of the foot.

Posterior-leaf-spring AFO
(from Latin *posterior* = back): ankle-foot orthosis with leaf spring attached behind the Achilles tendon, mostly made of carbon fibre

Progressive
(from Latin *progredere* = to advance): advancement of a disease or manifestation of the ↑symptoms associated with a disease

Proximal
(from Latin *proximus* = the nearest): positioned towards the centre of the body. The opposite of proximal is ↑distal.

Push Off
Toe-off during pre swing. This accelerates the leg into a forward movement.

SAFO
(solid ankle-foot orthosis): rigid lower leg orthosis. The term SAFO is used internationally for rigid ↑AFOs made of polypropylene. The present use of this term is ambiguous since static AFOs are also rigid.

Skin Irritation or Abrasion
Skin irritation is a skin change caused by prolonged irritation, which manifests itself in redness, itching, burning, tightness or discomfort in the affected area. A skin abrasion refers to the graze or scraping away of one or more layers of skin in the affected area.

Spring Unit
precompressed coil springs or specifically layered disc springs intended for use in system ankle joints

Stork Walk
Pathologically altered manner of walking: This compensation mechanism is intended to enable swinging without stumbling in the case of a drop foot. The increased hip and knee flexion is reminiscent of a stork walk.

Subtypes
(from Latin *sub* = under, around, against, below, at; from Greek *týpos* = type, kind): sub category, variation

Symptoms
total of all signs detected by the patient or physician that occur in connection with a disease

Tibia
(lat. *tibia* = shinbone): the stronger of the two lower leg bones, which is part of both the knee and ankle joint

Abbr.	Source
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FIOR & GENTZ
ORTHOPÄDIETECHNIK MIT SYSTEM

Orthosis Configurator

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