2018년도 대한소아재활발달의학회 춘계연수강좌

Spasticity : Cause & Clinical presentation

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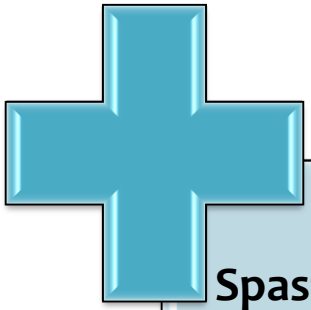
SPASTICITY : CAUSE

Definitions of spasticity

- A motor disorder characterized by **a velocity-dependent increase in tonic stretch reflexes ("muscle tone")** with **exaggerated tendon jerks**, resulting from **hyperexcitability of the stretch reflex**, as one component of the **upper motor neuron syndrome**

Lance JW et al. ,1980

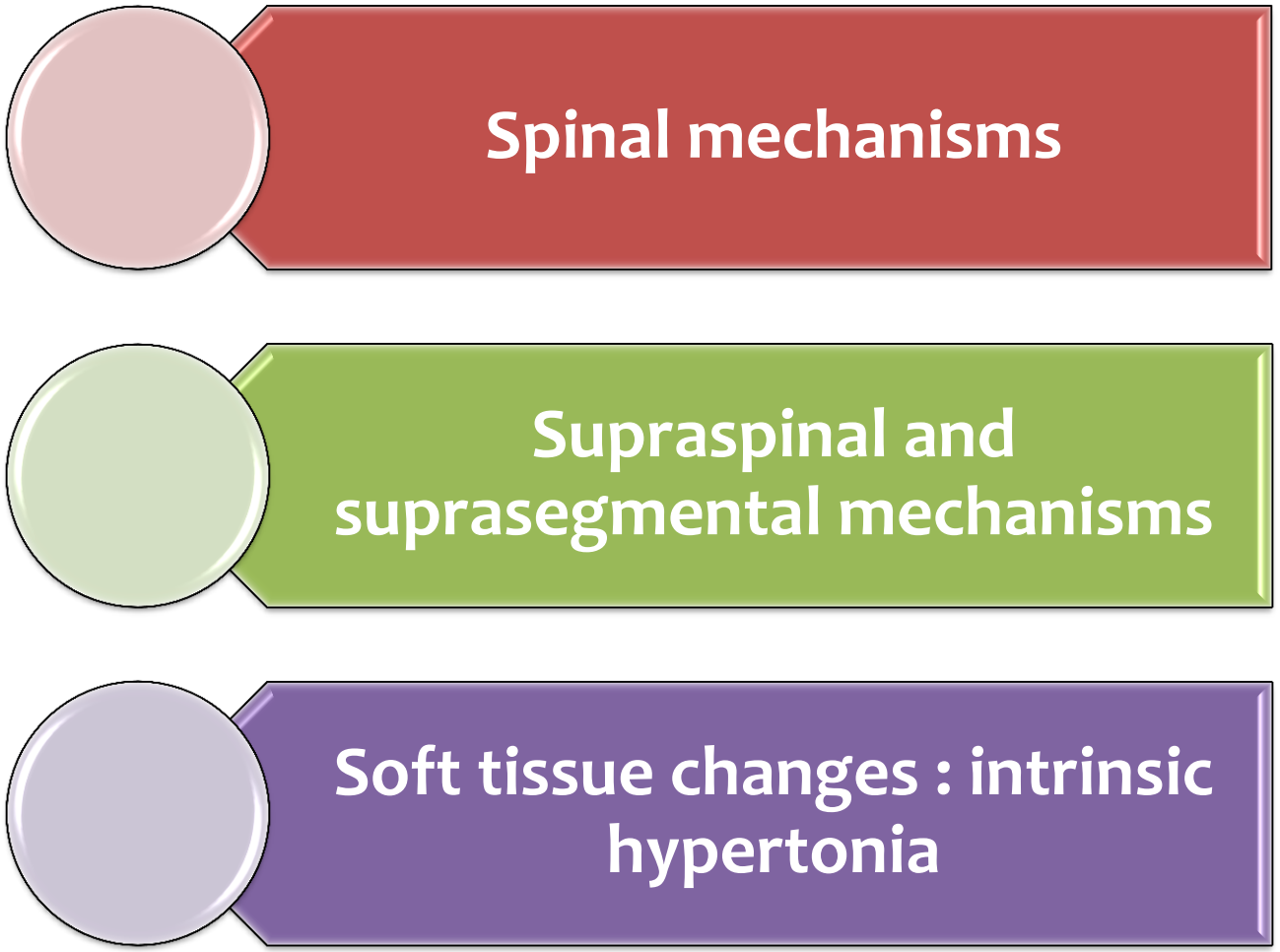
Upper motor neuron syndrome



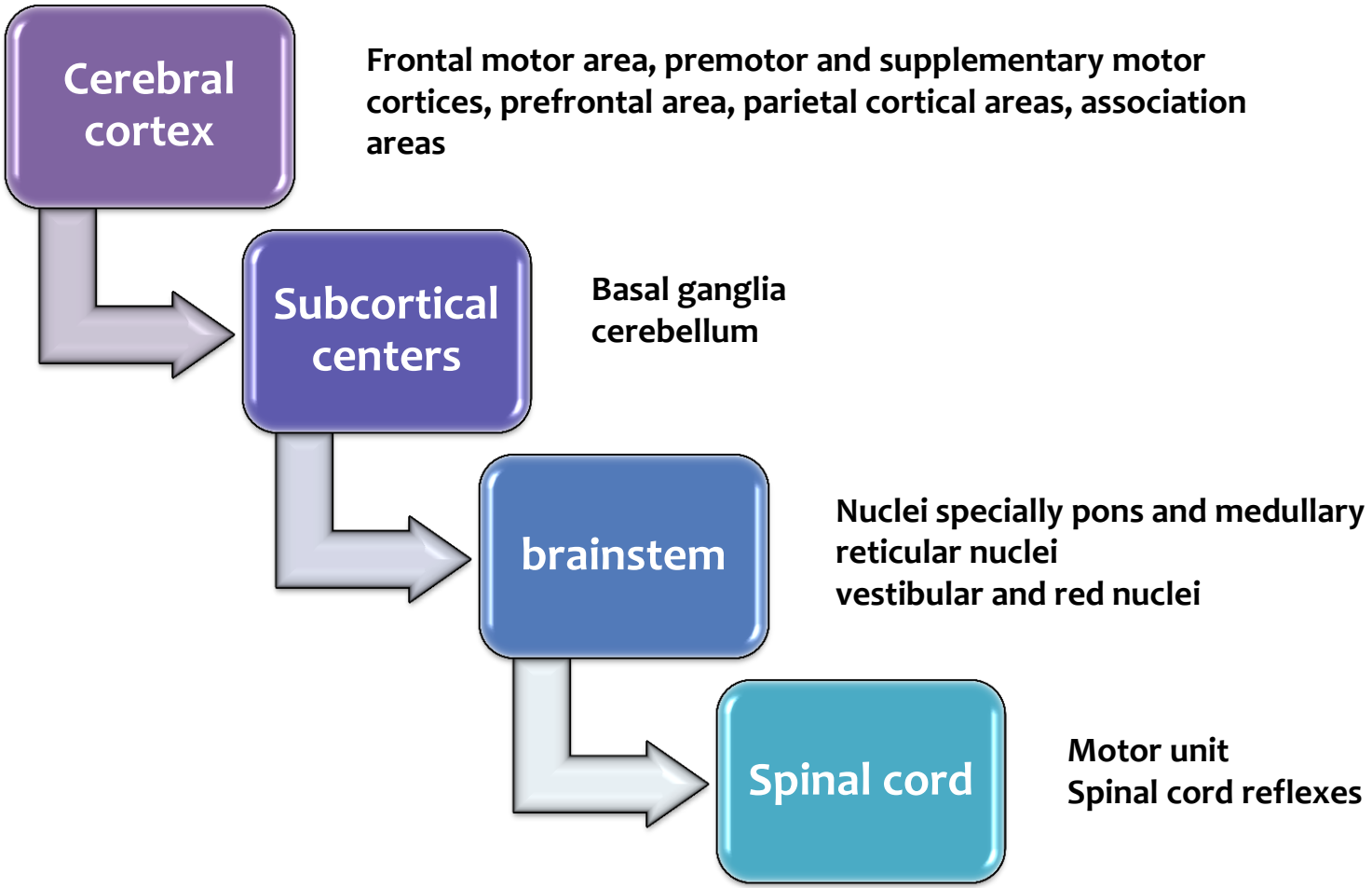
- Spasticity
- Exaggerated deep tendon reflexes
- Clonus
- Flexor and extensor spasms
- Extensor plantar responses
- Associated reactions
- Spastic dystonia
- Spastic co-contraction

- Weakness
- Loss of dexterity
- Fatigue

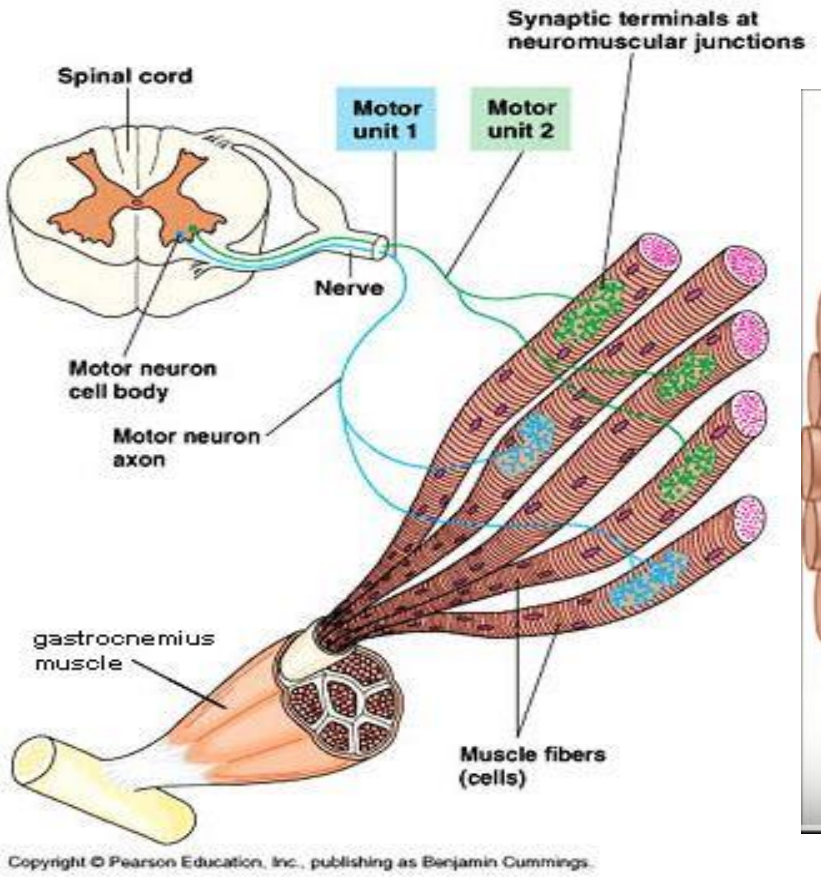
Pathophysiology of spasticity



Motor control system

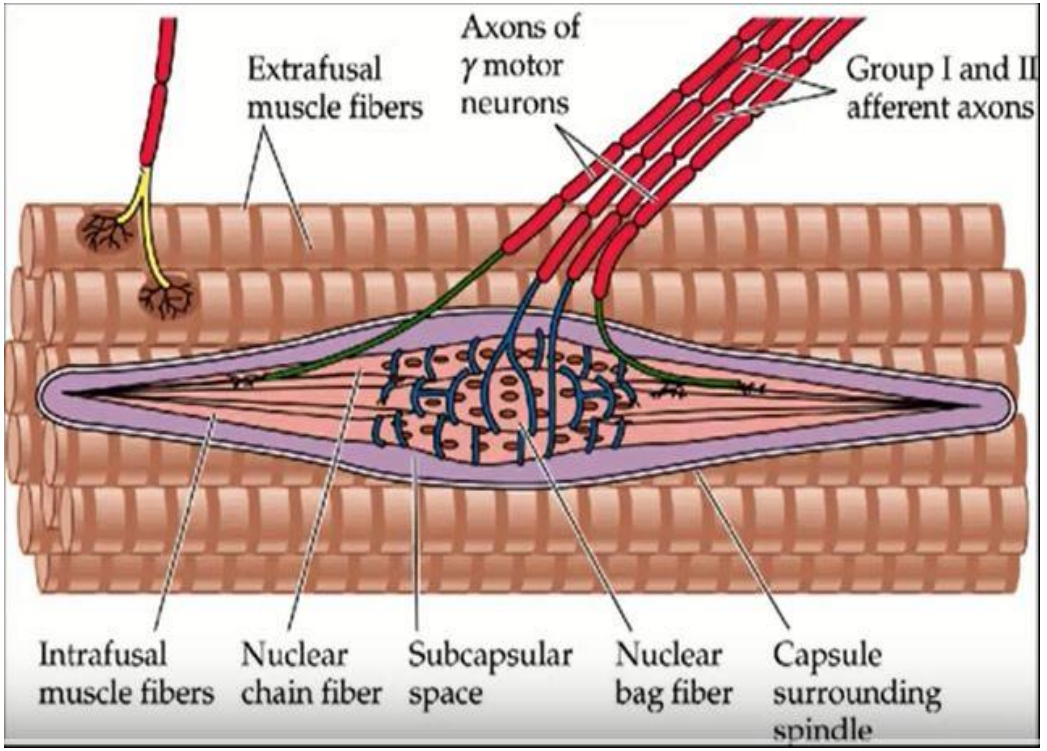


Motor functions of the spinal cord



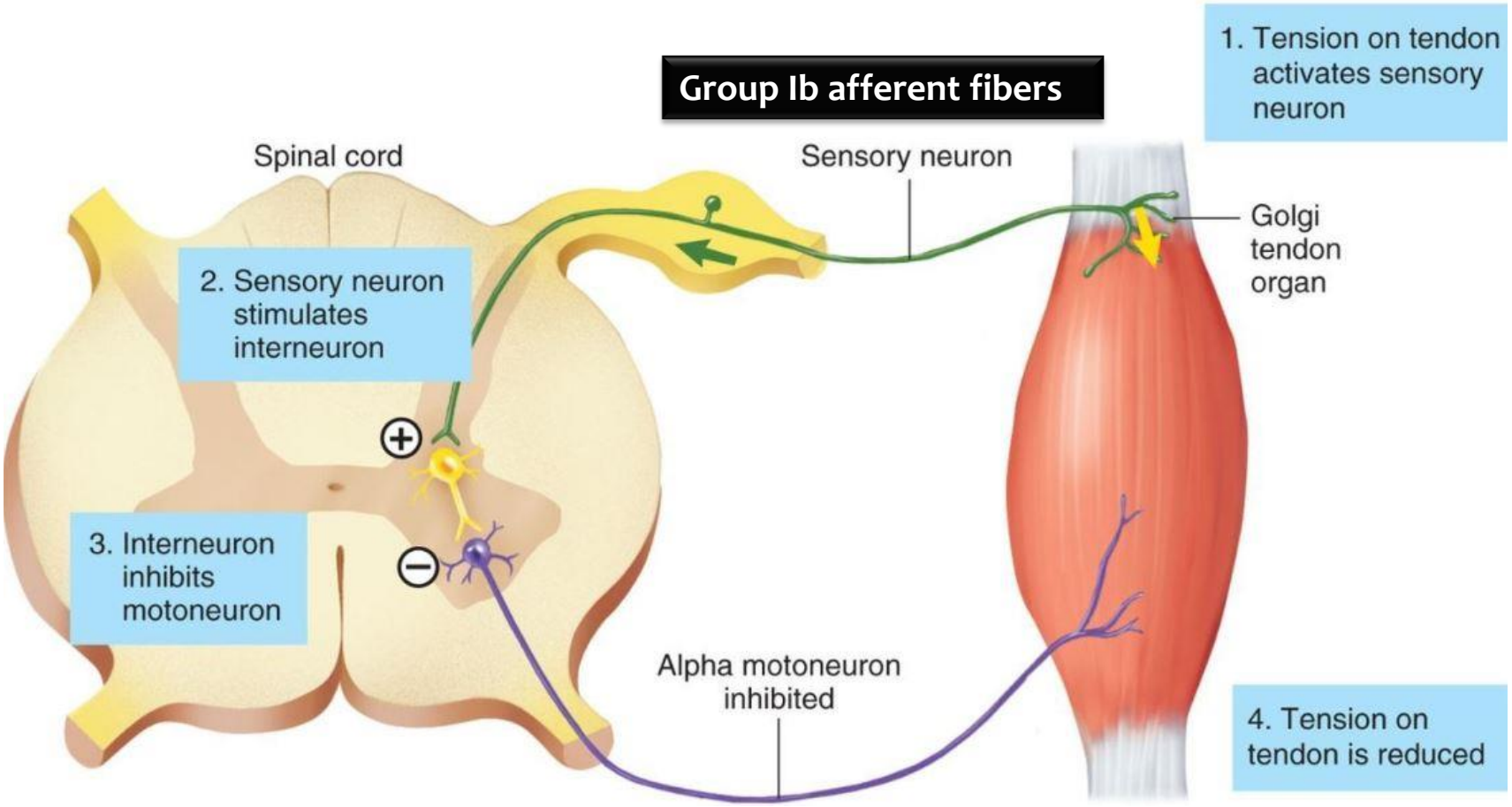
Motor unit

Group Ia & II afferent fibers



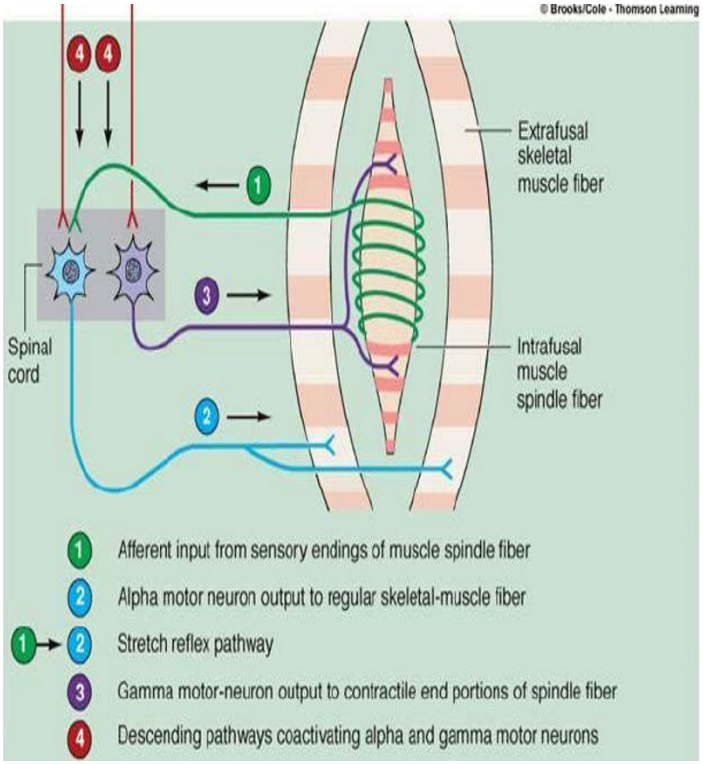
Muscle spindle

Motor functions of the spinal cord

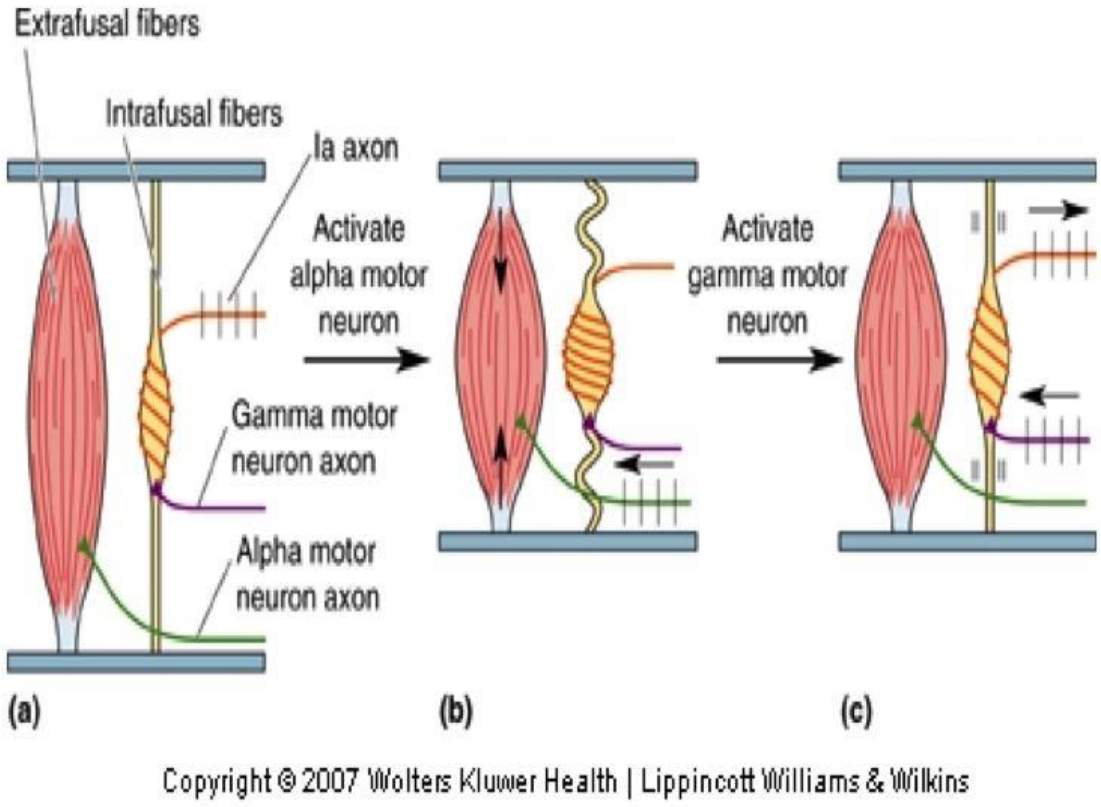


Golgi tendon organs

Motor functions of the spinal cord

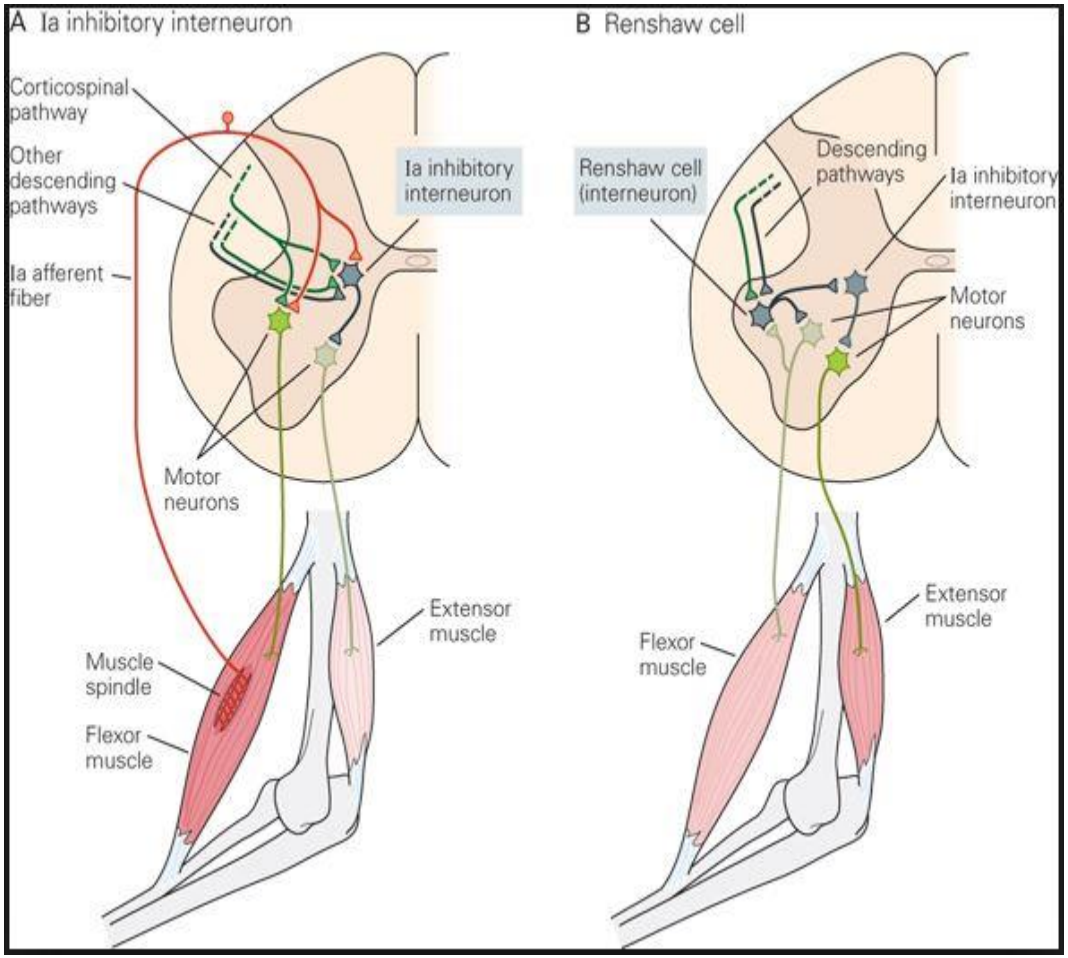


Stretch reflex



Alpha-gamma co-activation

Motor functions of the spinal cord



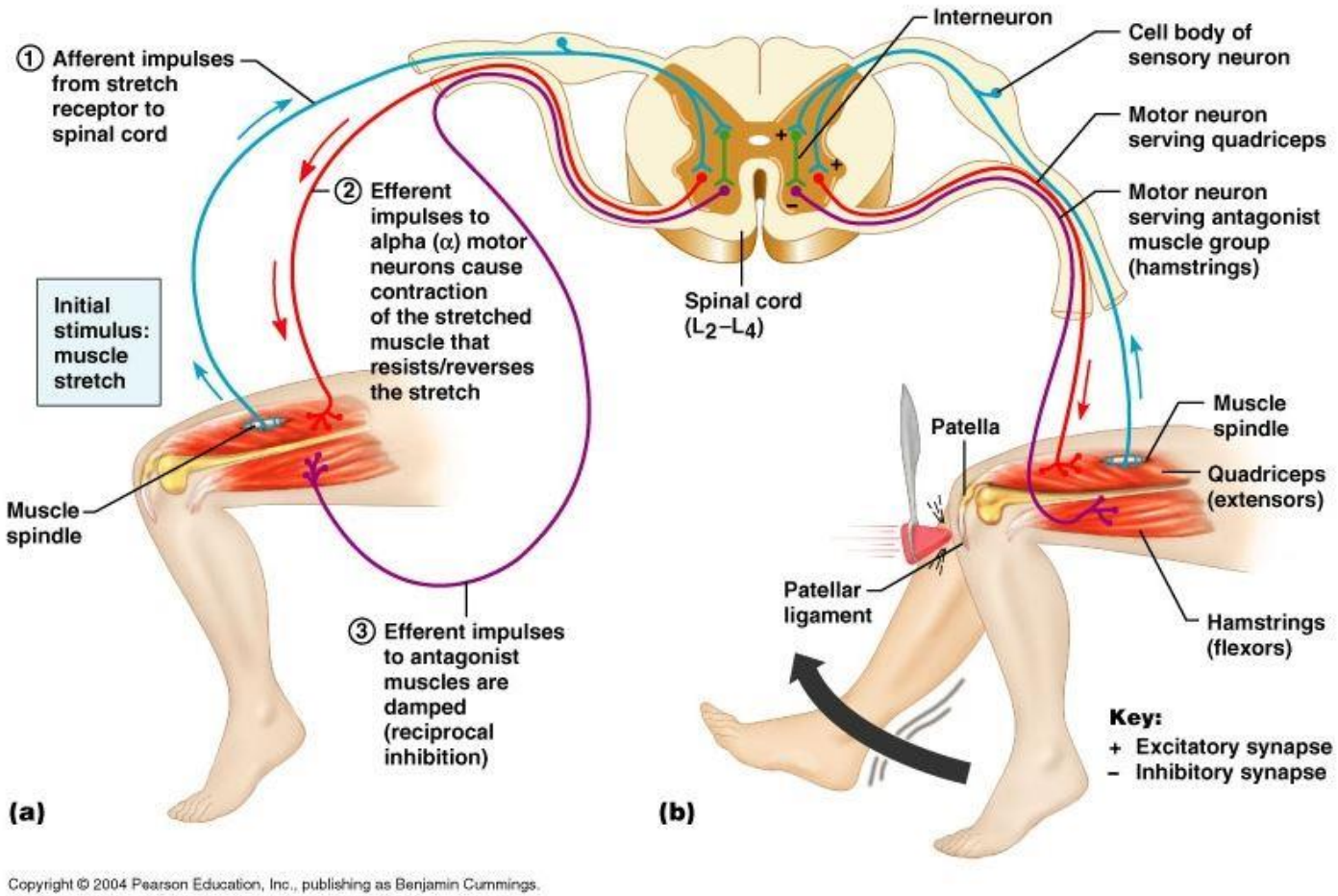
Spinal interneurons

- Renshaw cells and recurrent inhibition
- Reciprocal Ia inhibition
- Inhibitions from group II afferents
- Non-reciprocal Ib inhibition
- Presynaptic inhibition
- Flexor reflex afferents

Spinal interneurons

Mukherjee A & Chakravarty A, 2010

Motor functions of the spinal cord



Descending pathways

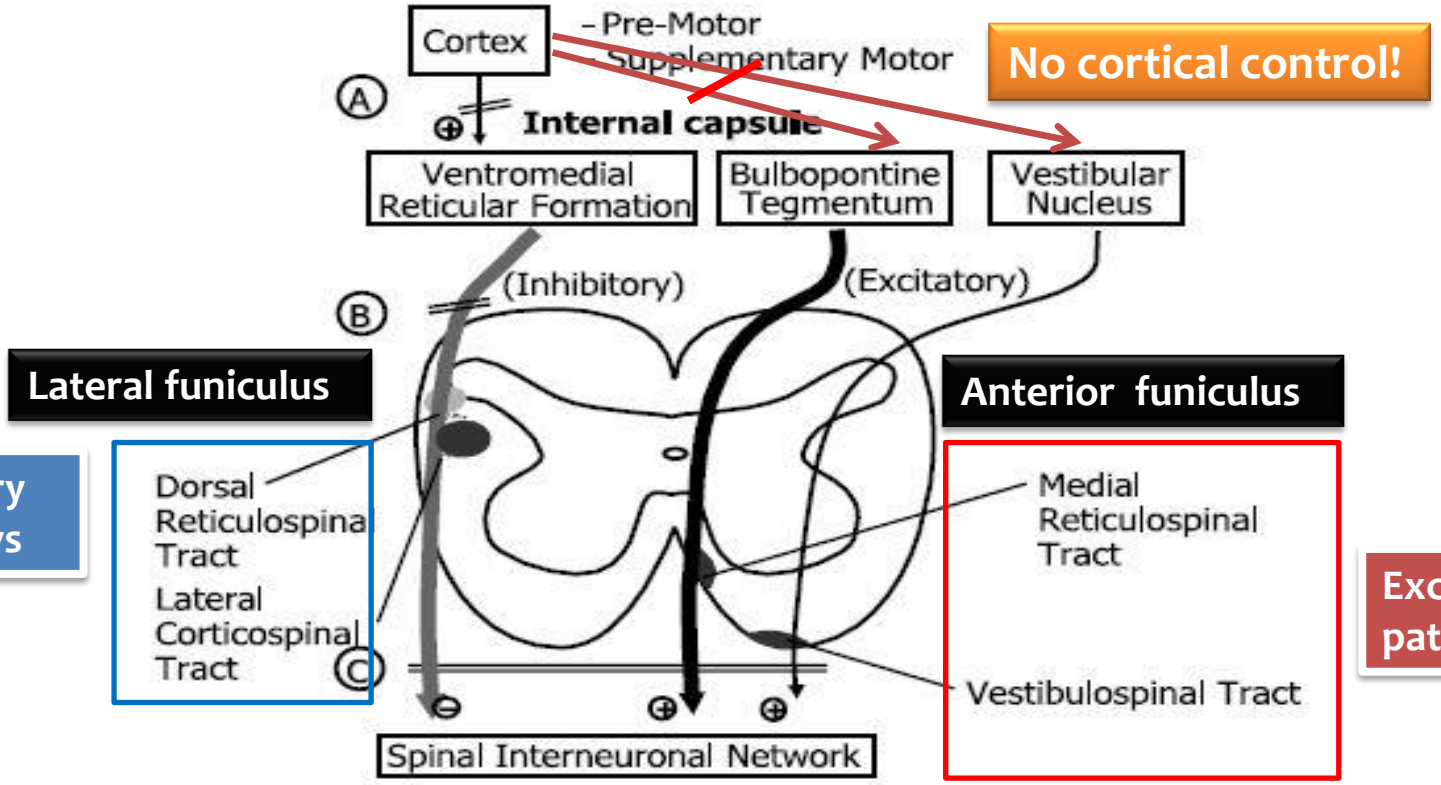


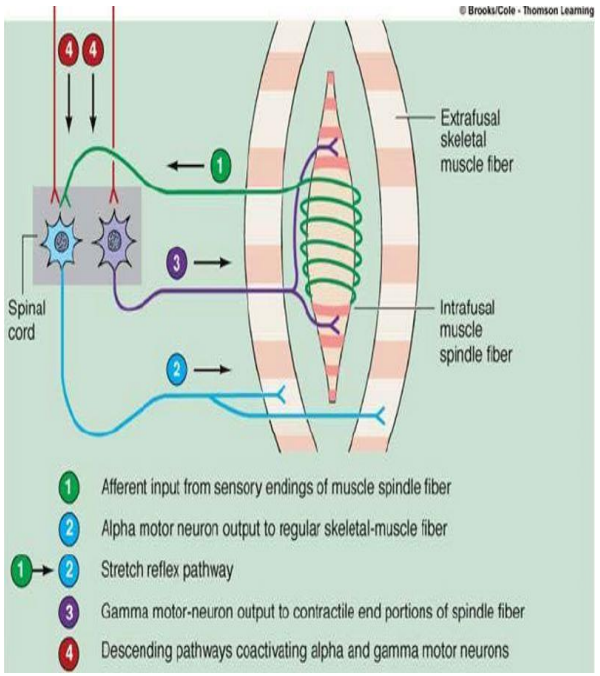
Figure 1 The major descending pathways controlling spinal reflex excitability. The inhibitory fibres are shown in grey.

Sheean G, 2002

Spinal excitatory mechanisms

Abnormal processing of sensory inputs in the spinal cord

-An excessive reflex activation of α-motoneurons

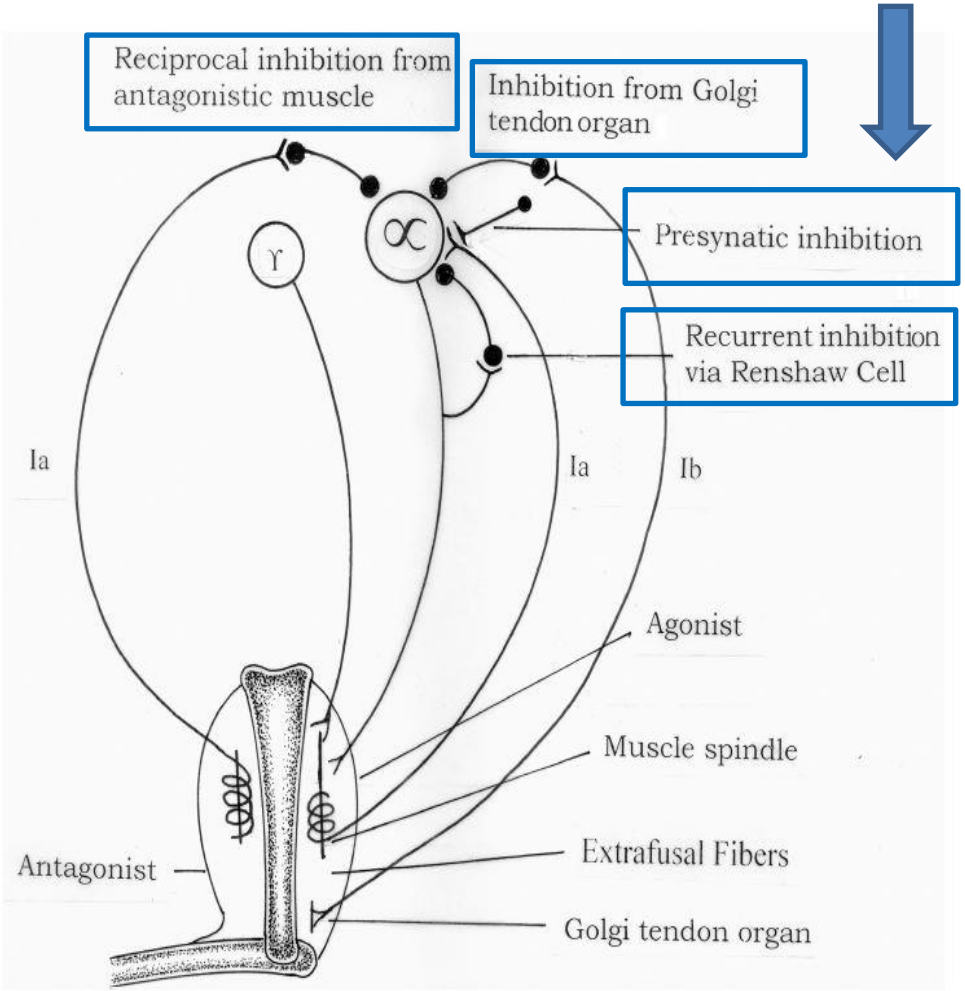


Increased excitability of muscle spindles

- Γ-motoneurons hyperactivity and subsequent muscle spindle hyperexcitability have a role in producing hypertonia

Trompetto C et al., 2014

Spinal inhibitory mechanisms



- Presynaptic inhibition of Ia afferent terminals
- Disynaptic reciprocal Ia inhibition
- Recurrent inhibition
- Non-reciprocal Ib inhibition

Spinal pathways which may be responsible for development of spasticity

Mukherjee A & Chakravarty A, 2010

Supraspinal and suprasegmental mechanisms

- **Muscle tone** is maintained by a **controlled balance** on stretch reflex arc by **inhibitory infleunce of CST and dorsal RST** and **facilitatory influence (on extensor tone) by medial RST and to a lessor extent in humans by VST**

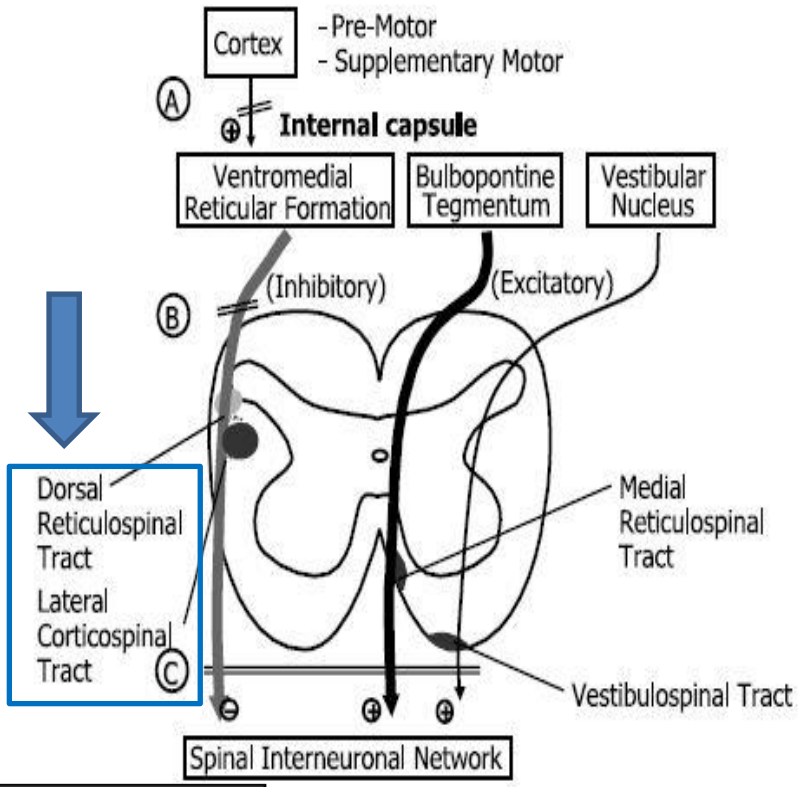
Mukherjee A & Chakravarty A, 2010

- **Imbalance of the descending inhibitory and facilitatory influences on muscle stretch reflexes** is thought to be the cause of **spasticity**

Lundberg, 1975

- In human spastic paretic syndrome, the three important pathways are –
  - **Corticospinal**
  - **Reticulospinal**
  - **Vestibulospinal**

Inhibitory supraspinal pathways



Lateral funiculus

Figure 1 The major descending pathways controlling spinal reflex excitability. The inhibitory fibres are shown in grey.

- Corticospinal pathway
  - Isolated pyramidal lesions : no spasticity < weakness, hypotonia, and hyporeflexia
  - Involvement of premotor and SMA : spasticity
- Corticoreticular pathways and dorsal reticulospinal tract (RST)
  - Medullary reticular formation : powerful inhibitory center to regulate muscle tone
  - Flexor spasm : release phenomenon of flexor reflexes due to damage to dorsal RST

Excitatory supraspinal pathways

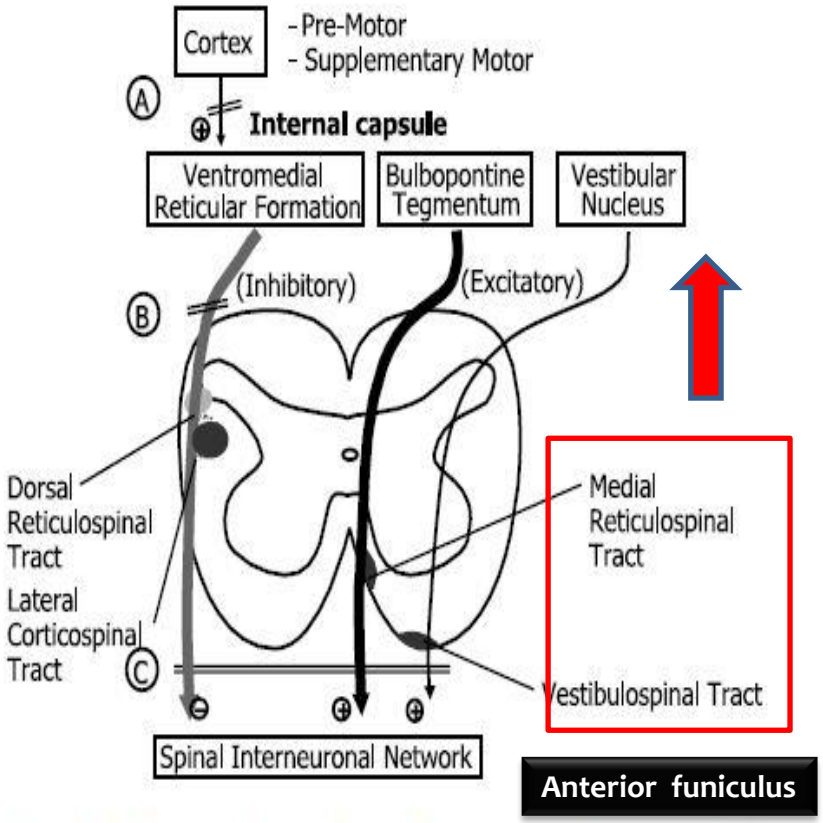


Figure 1 The major descending pathways controlling spinal reflex excitability. The inhibitory fibres are shown in grey.

- Vestibulospinal pathway (VST)
  - Maintain posture
  - Support against antigravity
  - Control extensors rather than flexors
- Medial reticulospinal tract (RST)
  - Mainly from pontine tegmentum
  - More important than VST in maintaining spastic extensor tone
- Cerebellum through its connections with the vestibular nuclei and reticular formation may indirectly modulate muscle stretch reflexes and tone

Soft tissue changes : intrinsic hypertonia

- Hypertonia in patients with UMNS can be divided into two components
  - **Hypertonia mediated by the stretch reflex, which corresponds to spasticity**
  - **Hypertonia due to muscle contracture, which is often referred as intrinsic hypertonia**
- The increased mechanical resistance may be caused by alterations in tendon compliance and physiological changes in muscle fibers
- Two components of hypertonia are likely to be **intimately connected**
  - **Reduced muscle extensibility due to muscle contracture → cause any pulling force to be transmitted more readily to the spindles → increasing spasticity**

Trompetto C et al., 2014

Muscle overactivity in the UMNS

- Spasticity
- Exaggerated deep tendon reflexes
- Clonus
- Flexor and extensor spasms
- Extensor plantar responses

Spinal reflexes

- Associated reactions
- Spastic dystonia

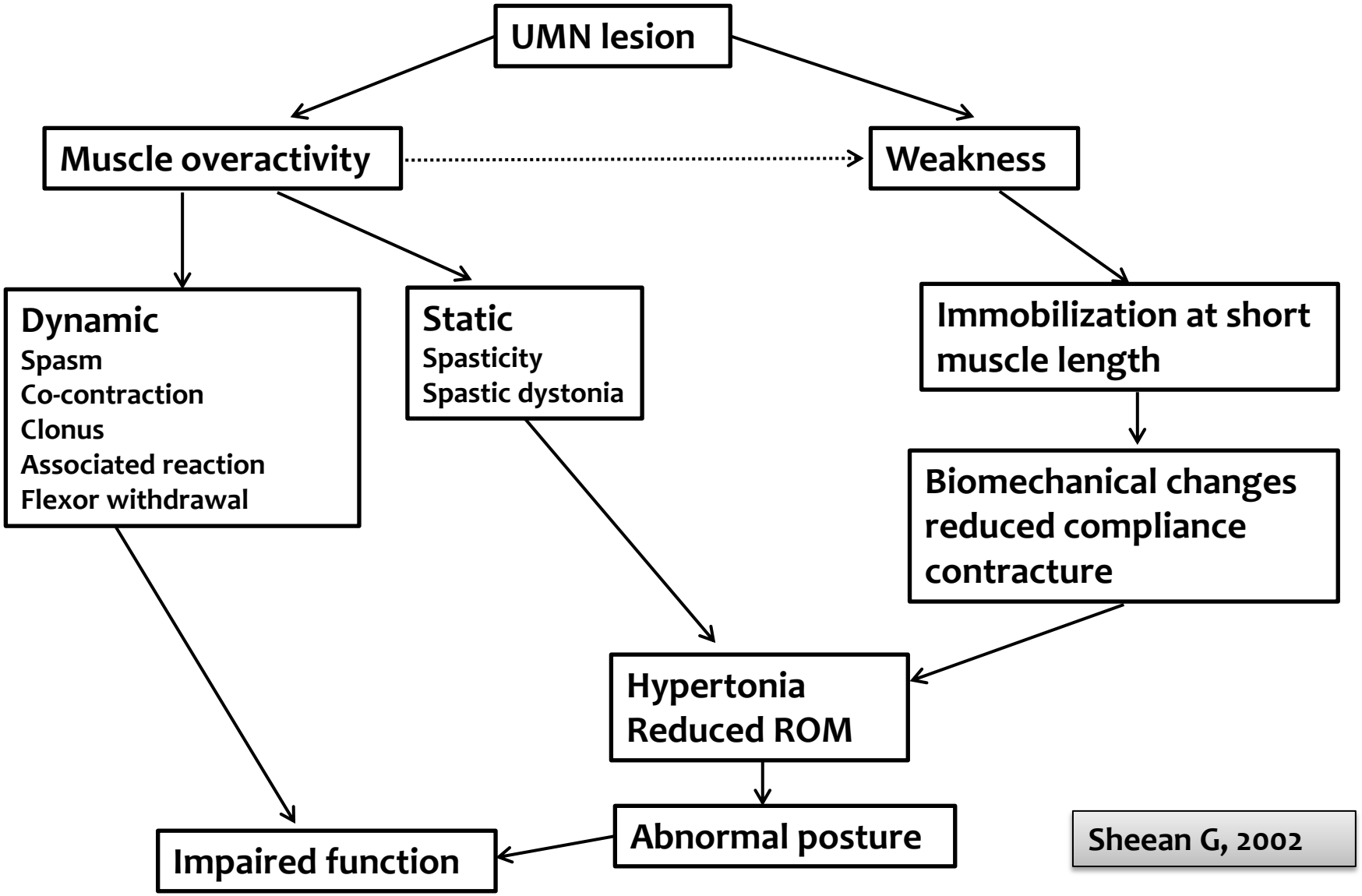
Efferent drive

- Spastic co-contraction

Disordered control of movement

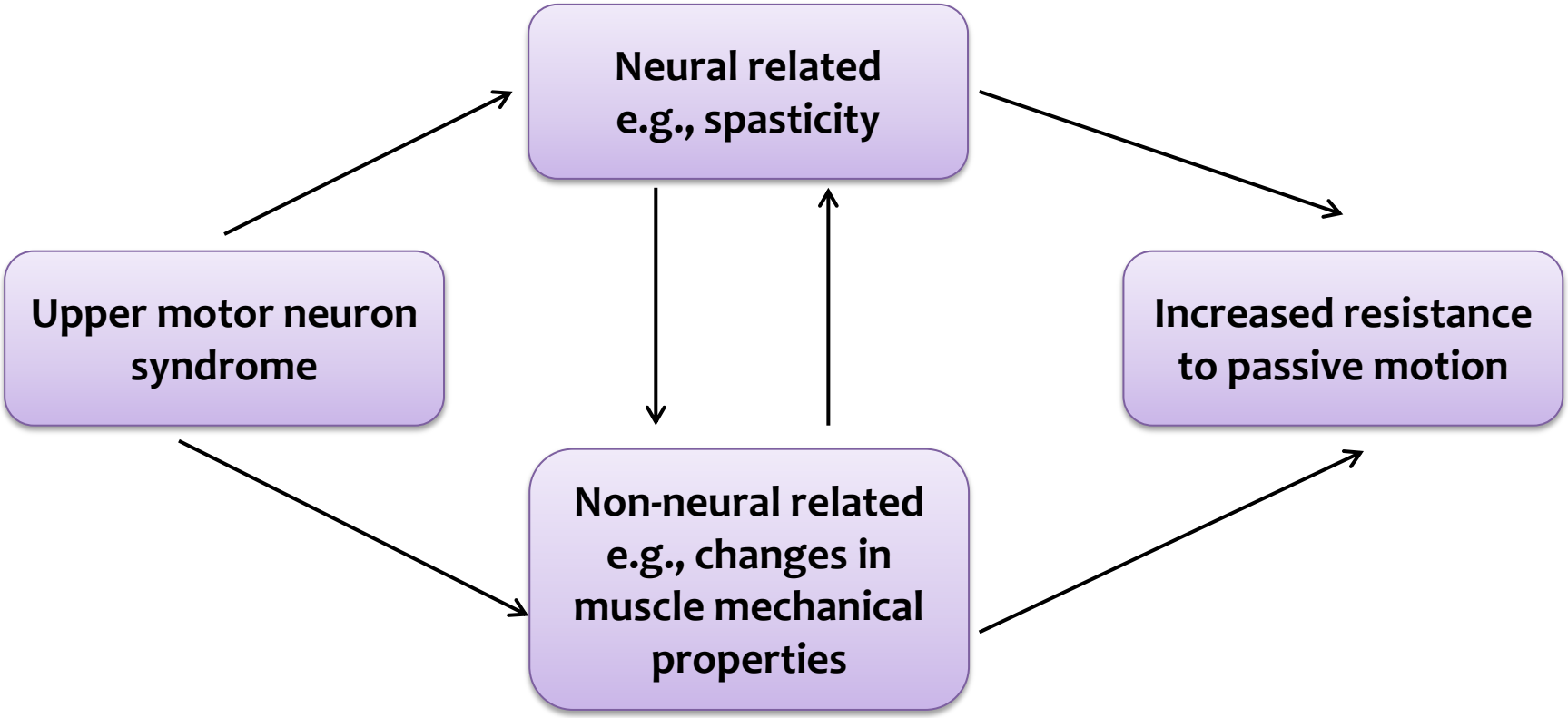
Sheean G, 2002

Overview of the features of the UMNS



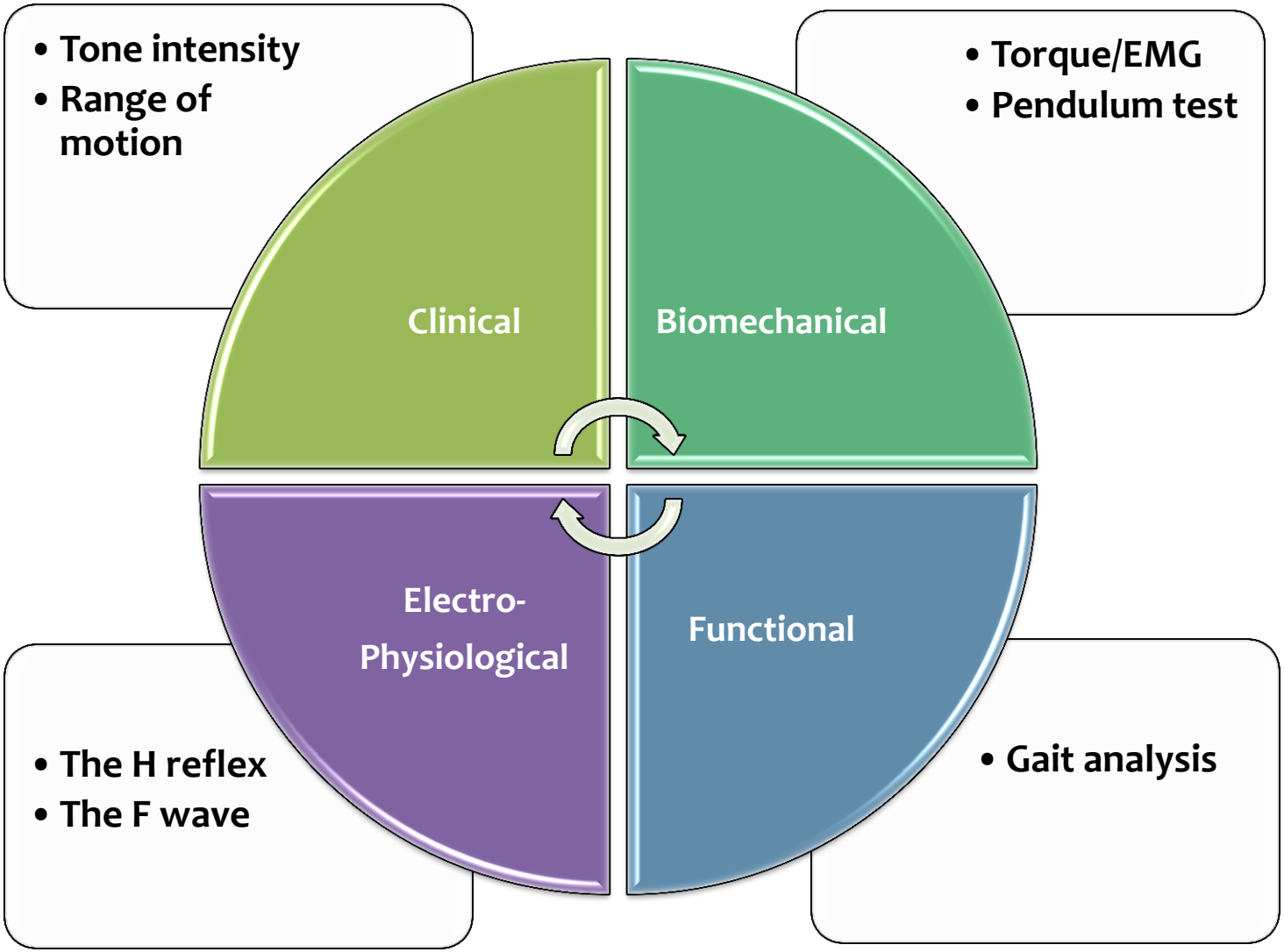
SPASTICITY : CLINICAL PRESENTATION

Neural vs non-neural components



Neural and non-neural mechanisms contributing to increased resistance to passive motion in an upper motor neuron syndrome

Measurements in spasticity



Ashworth scale

Score	Ashworth (Ashworth 1964)	Modified Ashworth (Bohannon and Smith 1987)
0	No increase in tone	No increase in tone
1	Slight increase in tone giving a catch when limb is moved in flexion /extension	Slight increase in tone giving a catch, release and by minimal resistance at the end of range of motion (ROM) when the limb is moved in flexion/extension
1+		Slight increase in tone giving a catch, release and minimal resistance throughout the remainder (less than half) of ROM
2	More marked increase in tone, but the limb is easily moved through its full ROM	More marked increase in tone through most of the ROM, but limb is easily moved
3	Considerable increase in tone – passive movement difficult and ROM decreased	Considerable increase in tone-passive movement difficult
4	Limb rigid in flexion and extension	Limb rigid in flexion and extension

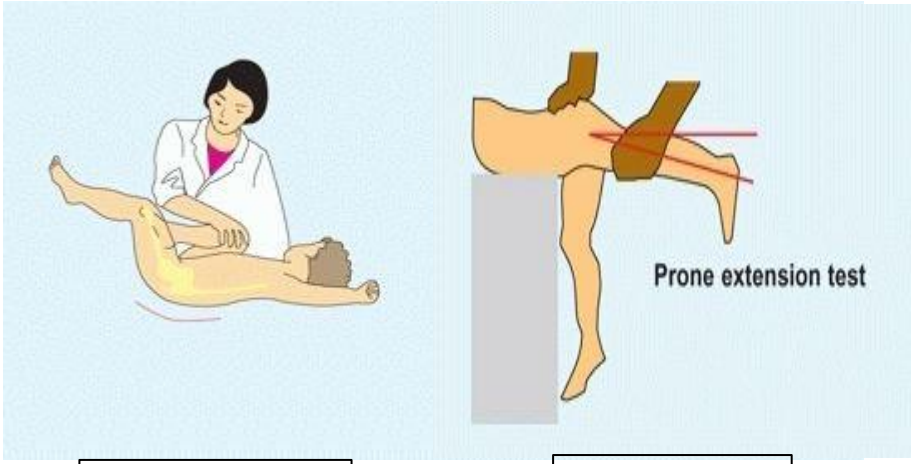
Tardieu scale

Y Angle (Dynamic Range of Motion)	
R1	Fast velocity: Movement through full range of motion
R2	Slow velocity: Passive joint range of motion or muscle length
Stretch Velocity	
V1	Slow as possible
V2	Speed of limb falling under gravity
V3	Fast as possible

R1	R2	R2-R1
Dynamic resistance	Static resistance	Used in effectiveness of chemoneurolysis in patients with cerebral palsy

Range of Motion

- **Hip joint**



Thomas test

Staheli test

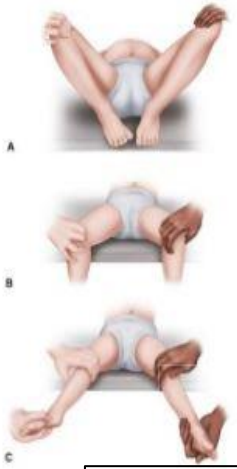


Figure 3

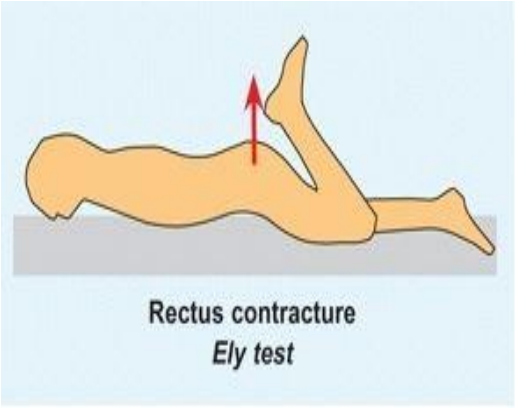
Testing for adductor and medial hamstring tightness

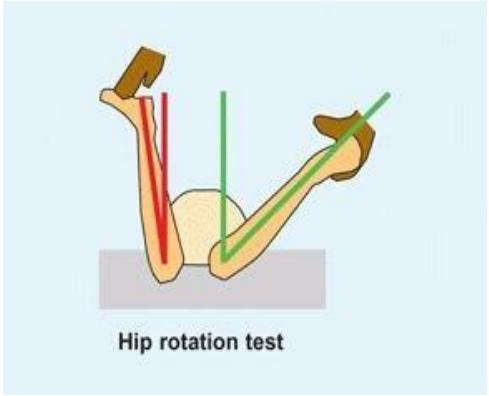
A, Thighs abduct well with hips and knees flexed, indicating no adductor contracture

B, With hips extended and knees flexed, hips abduct well

C, With hips extended, bringing knees into extension causes thighs to adduct, indicating medial hamstring spasticity.

Test for hip adductor tightness

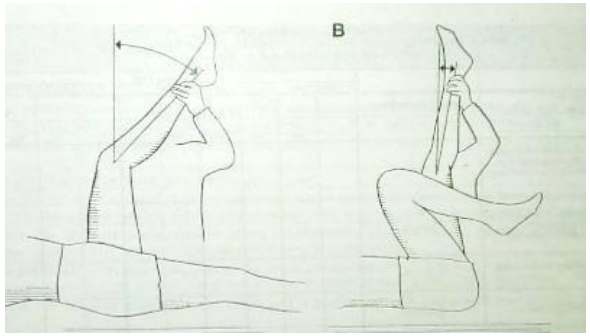




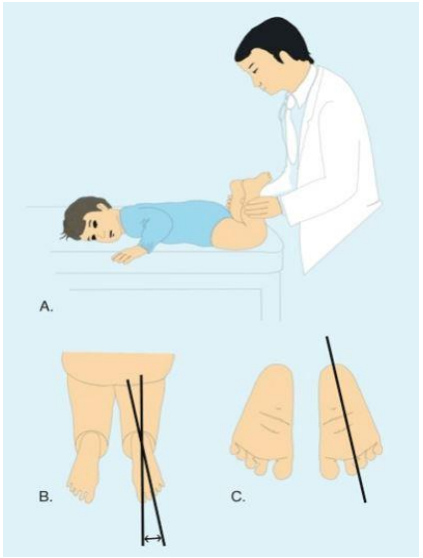
Gracilis	Adductor longus
2 joint muscle	1 joint muscle
Hip Ab with knee Ex→ Gr + AL	Hip Ab with knee Fl → Only AL

Range of Motion

- knee joint

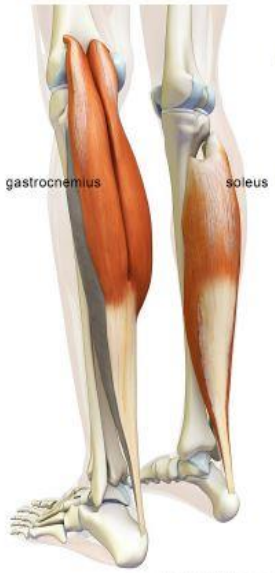


Unilateral and bilateral popliteal angle

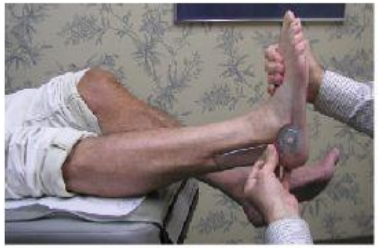


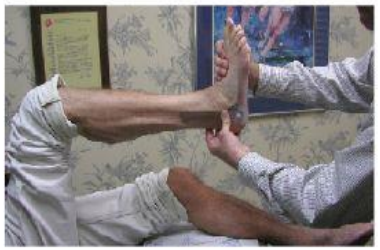
Thigh-foot angle

- Ankle joint



- Soleus : 1 joint muscle (ankle)
- Gastrocnemius : 2 joints muscle (knee & ankle)

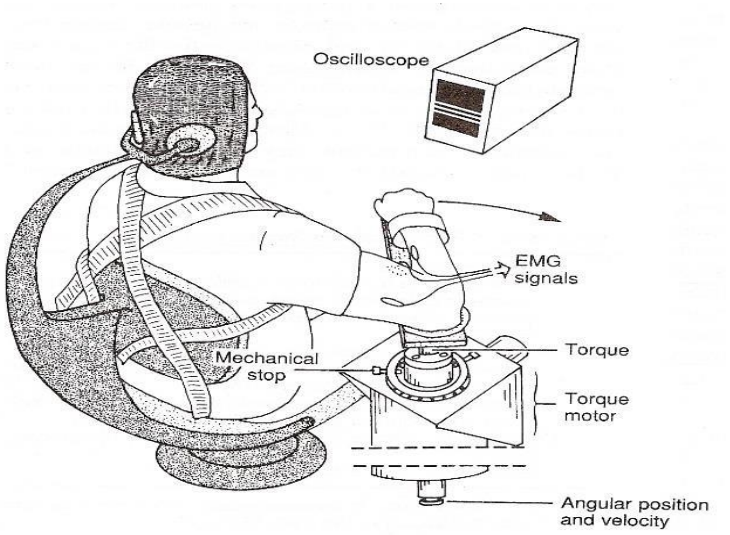


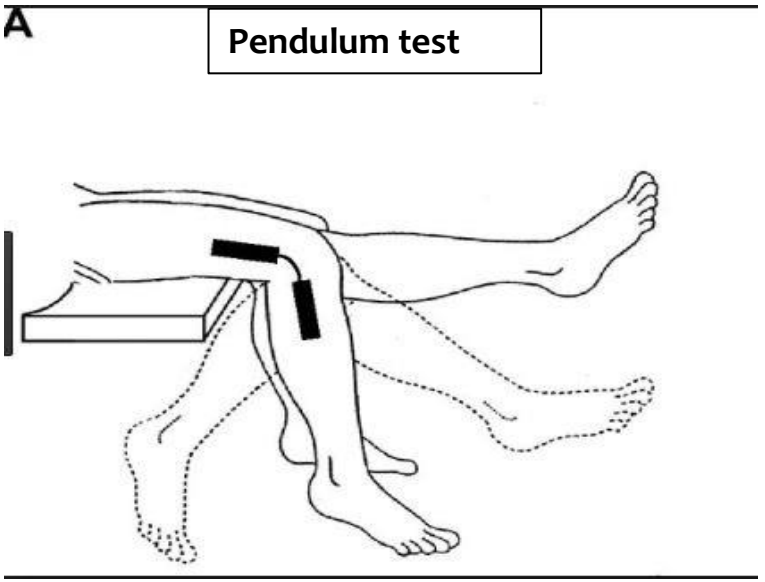


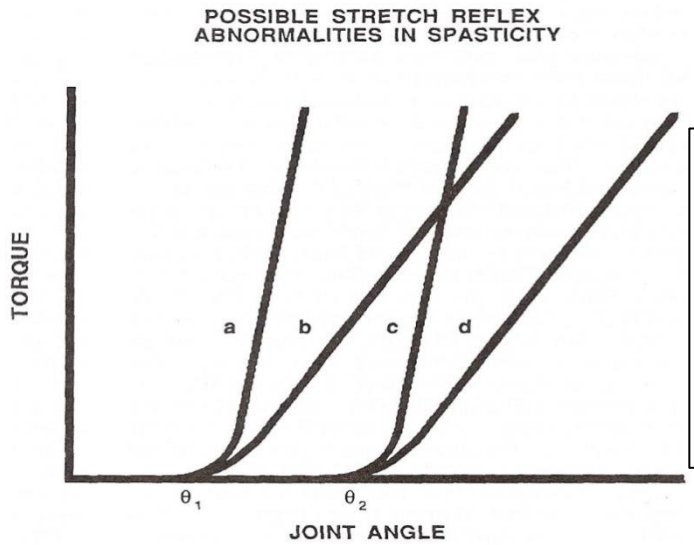
Silfverskiold test

Gastrocnemius	Soleus
2 joint muscle	1 joint muscle
Ankle DF with knee Ex → GCM + soleus	Ankle DF with knee Fl → Only soleus

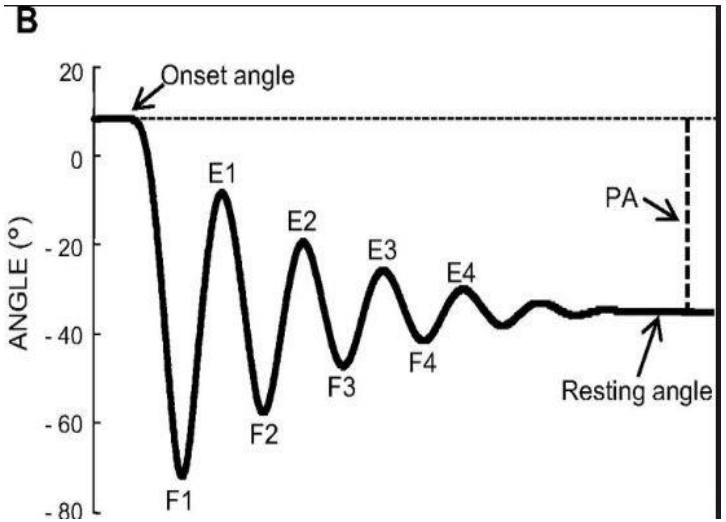
Biomechanical assessment



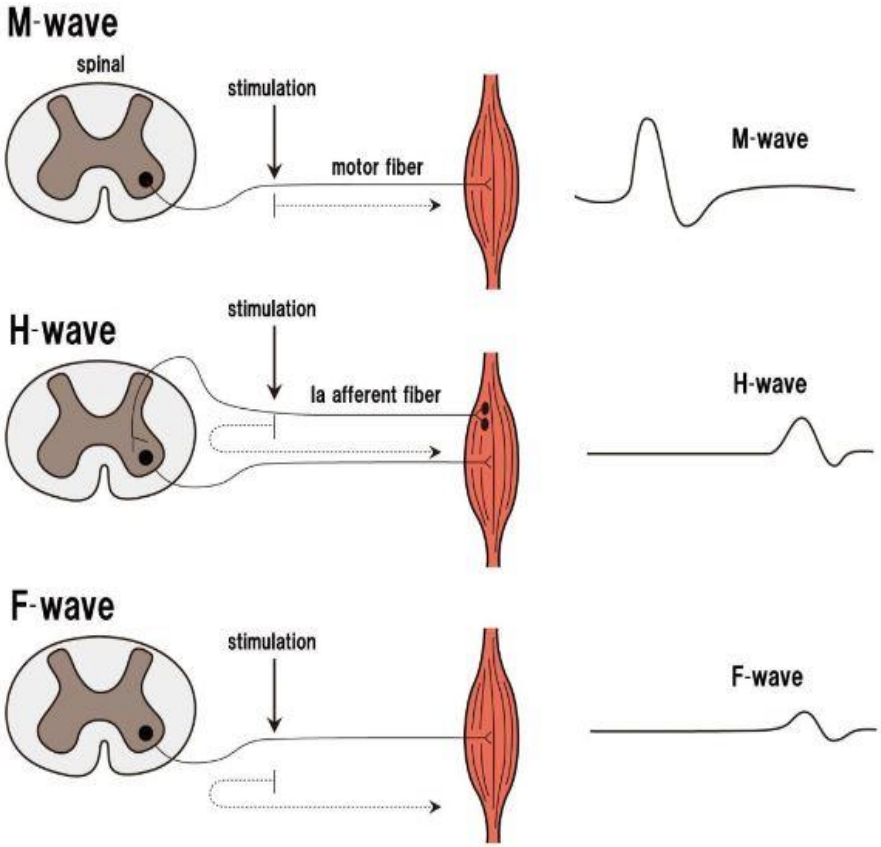




a; reduced threshold and increased stiffness
b; only reduced threshold
c; only increased stiffness
d; normal state of reflex threshold and stiffness



Electrophysiologic assessment



M-wave: Excitement is conducted to efferent. H-wave: Excitement is conducted via the monosynaptic reflex. F-wave: Excitement is conducted to antidromic.

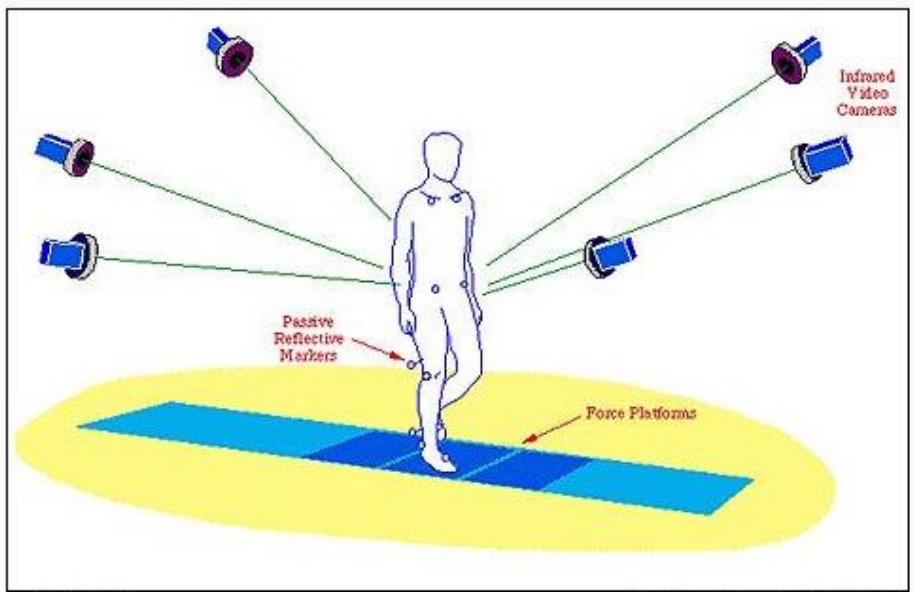
H reflex

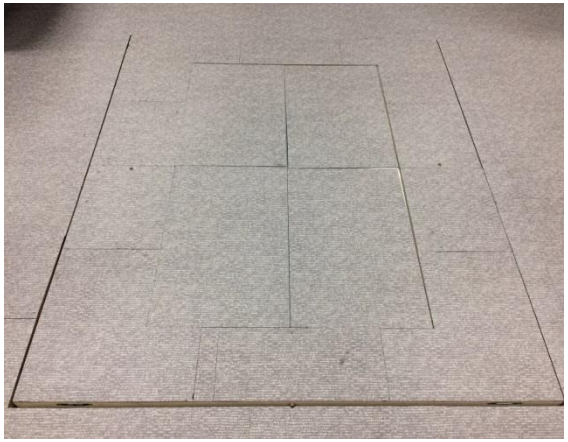
- sensory nerve -> Ia afferent sensory neuron -> alpha motor neuron -> motor nerve-> muscle
- H/M ratio

F wave

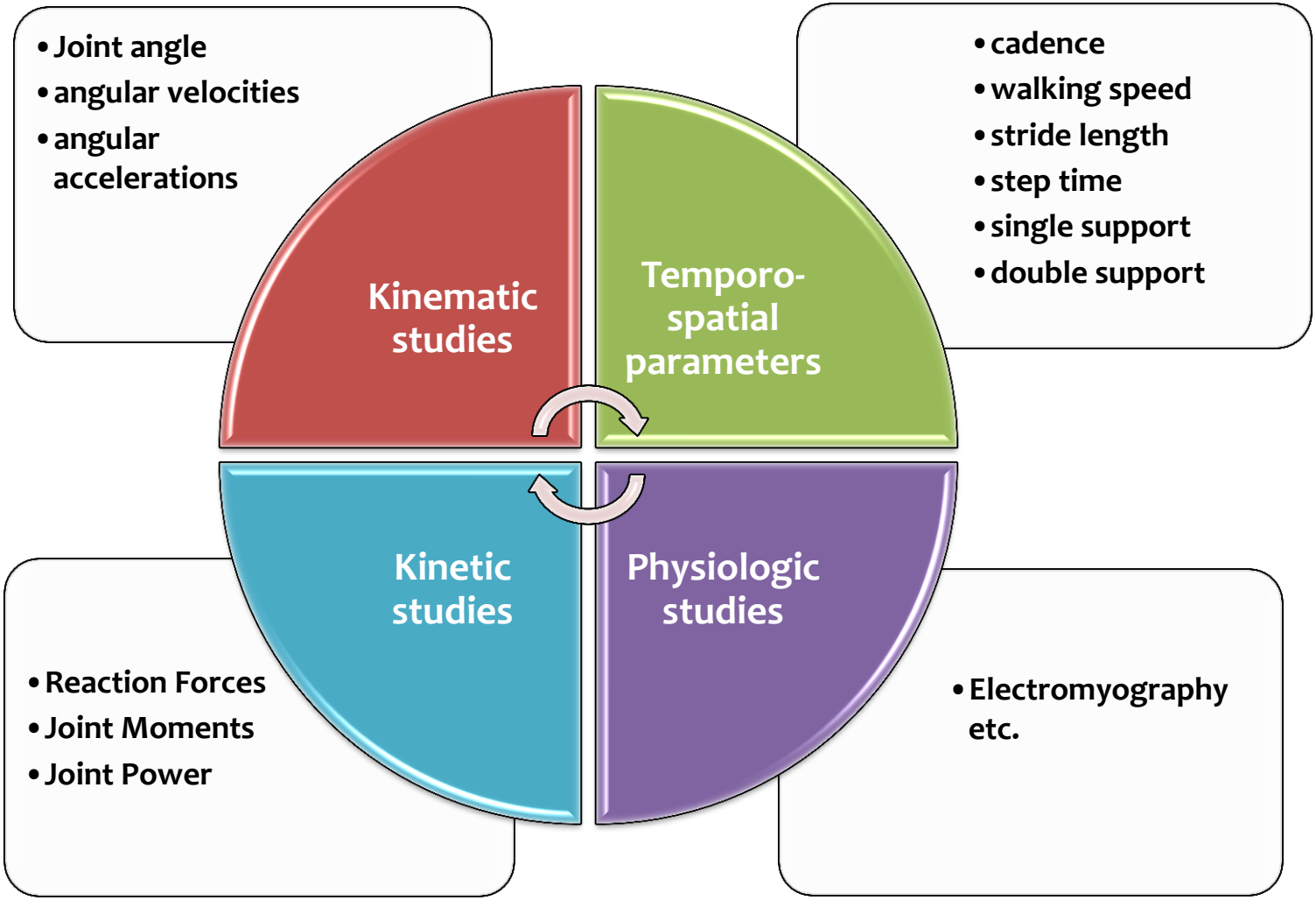
- motor nerve-> alpha motor neuron->motor nerve-> muscle
- F/M ratio

Gait analysis

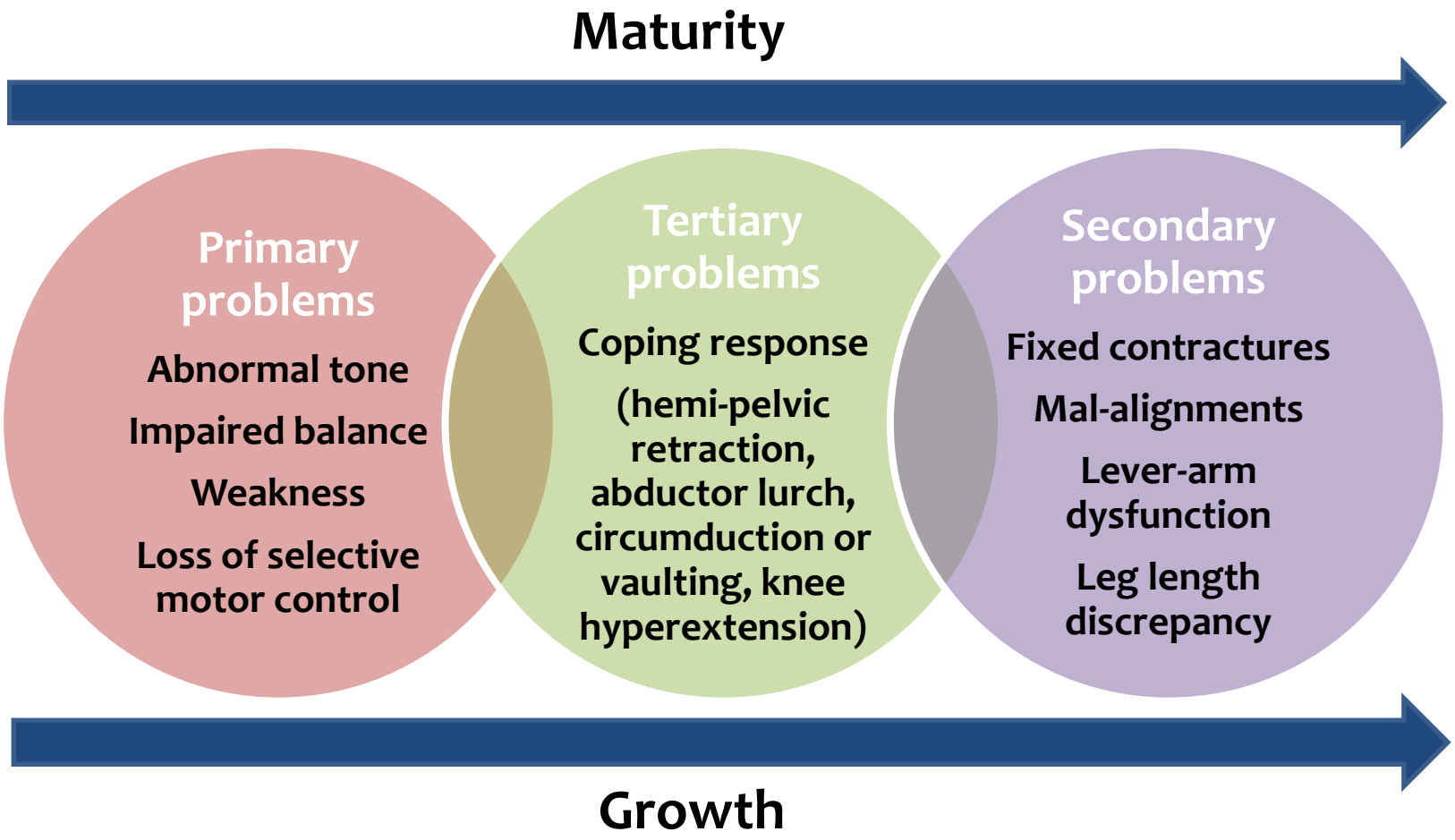




Gait analysis



Motor problems in CP



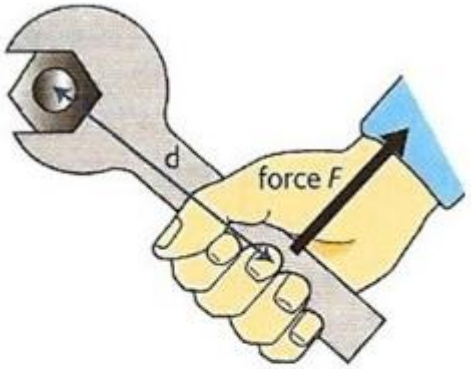
Motor problems in CP is a mixture of primary, secondary, and tertiary problems !

Molenaers G et al., 2010

Basic principles of gait biomechanics

- **Gait biomechanics**
  - **Force**
  - **Moment of force**
  - **Power**

- **Moment : turning effect of a force**
- The moment of a force about any point is defined
- **Force X perpendicular distance from the the line of action of the force**
- **Moment = F X d**
- Unit: **newton-metre (Nm)**
- Moments can be either **CLOCKWISE** or **ANTICLOCKWISE**



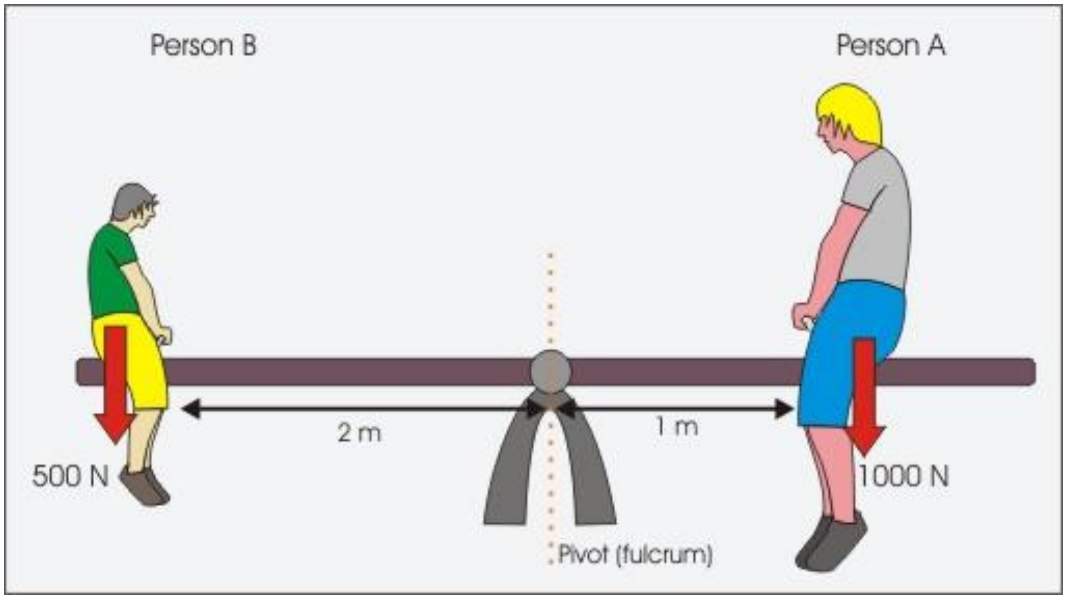
Force F exerting an ANTICLOCKWISE moment through the spanner on the nut

Basic principles of gait biomechanics

- In equilibrium

Total Anticlockwise Moment = Total Clockwise Moment

- **Person A' moment =**
  Force X perpendicular distance from fulcrum
  1000 X 1 = 1000Nm
- **Person B' moment =**
  Force X perpendicular distance from fulcrum
  500 X 2 = 1000Nm



- **Person A' moment = Person B' moment**
- **Anticlockwise moment = clockwise moment**
- Therefore seesaw is in equilibrium

Basic principles of gait biomechanics

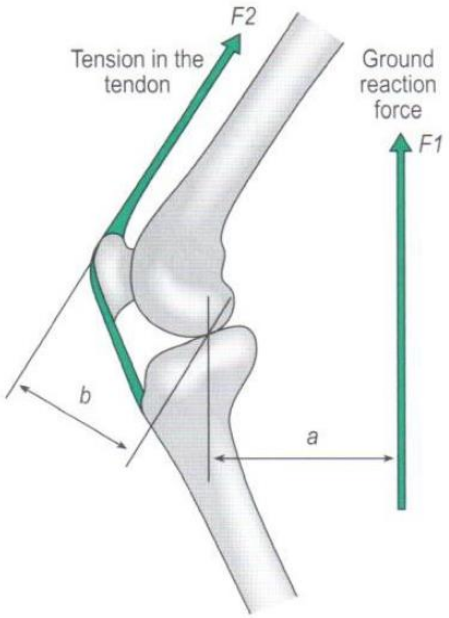


Fig. 1.22 • The moment of force due to ground reaction force, F_1 multiplied by a, is opposed by contraction of the quadriceps, producing a moment of force F_2 multiplied by b.

Force : muscle contraction
Perpendicular distance : lever arm
Fulcrum : joint
Internal force : muscle acting on the lever arm
External force : weight of limb or body or GRF

- The ground reaction force (**F1**) is acting a perpendicular distance **'a'** from the point of loadbearing
- Moment of force it provides the product of the tension in the tendon **(F2)** and the perpendicular distance **'b'**
- The presence of the patellar increases the value of b and reduces the muscle force needed to produce a given moment of force
- For equilibrium,

$F_1 \times a = F_2 \times b$

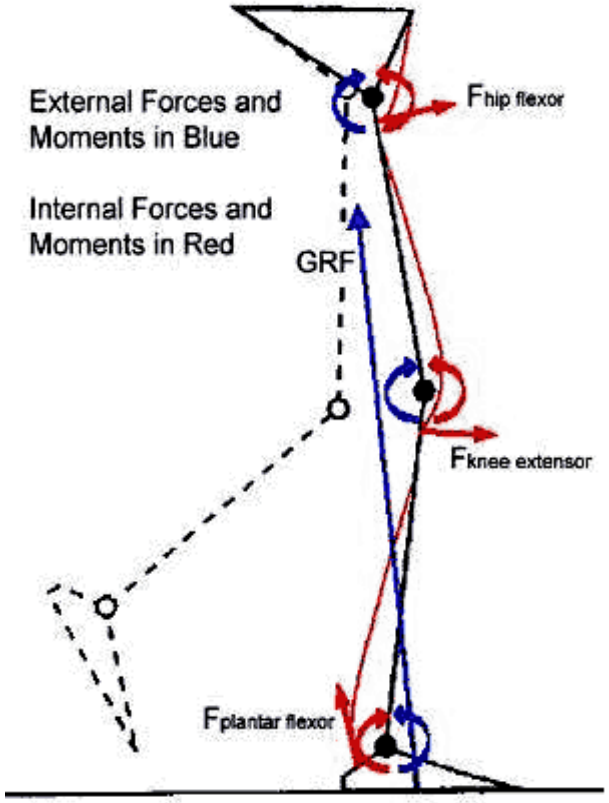
Basic principles of gait biomechanics

Internal moment

- **Active – muscular contraction (concentric, isometric or eccentric)**
- **Passive – bone-on-bone forces, tension in the soft tissues, esp. ligament**

External moment

- **Gravitational forces**



If an **external force** generates a moment at a particular joint, there must be a **corresponding internal moment**, generated within the joint, **to oppose it !**

Lever arm dysfunction

- The detrimental effect of secondary problems is mainly due to the **compromised function of the normal lever arm**

- Deformed bones, stiff joints and foot deformities all affect the ability of the skeleton to adequately respond to forces generated by muscle activity

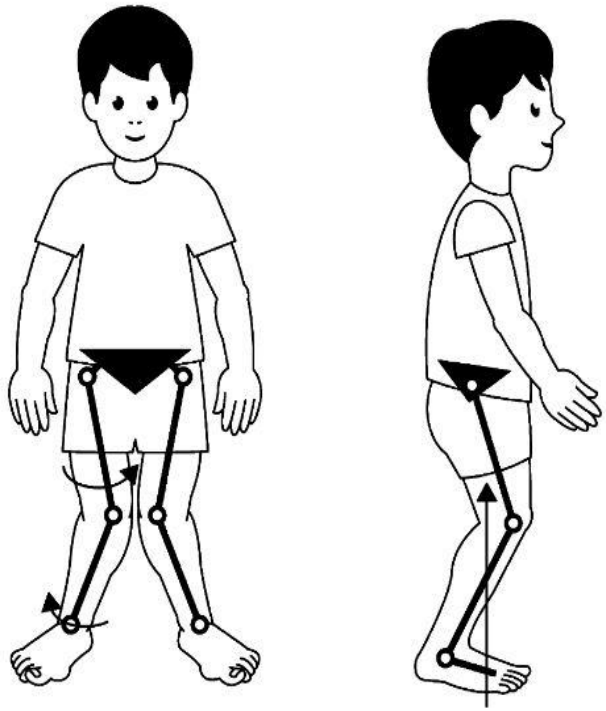
- The compromised lever arm prevents adequate transfer of forces and torques arising from the ground reaction force (GRF)

Lever arm dysfunction in CP

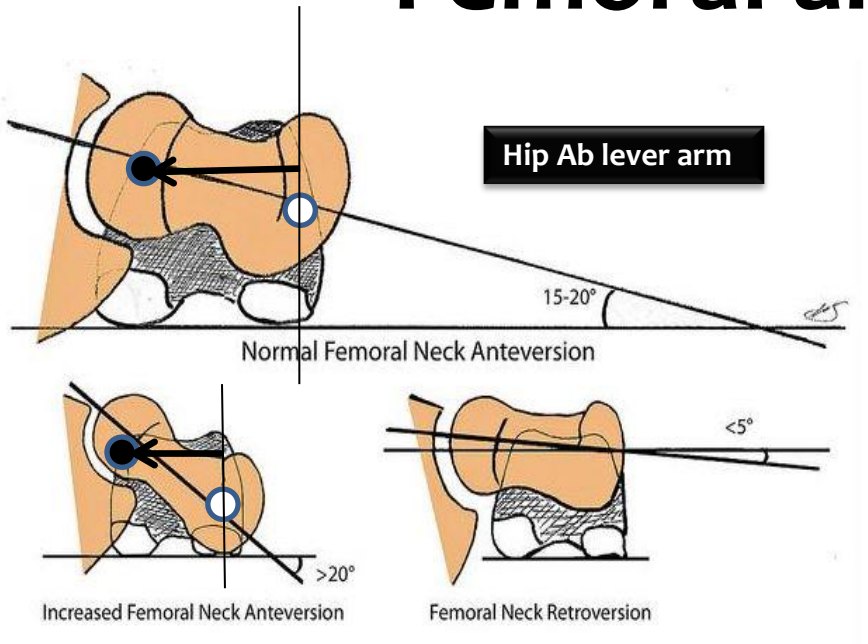
Torsional deformities of long bone
- Femoral anteversion
- External tibial torsion

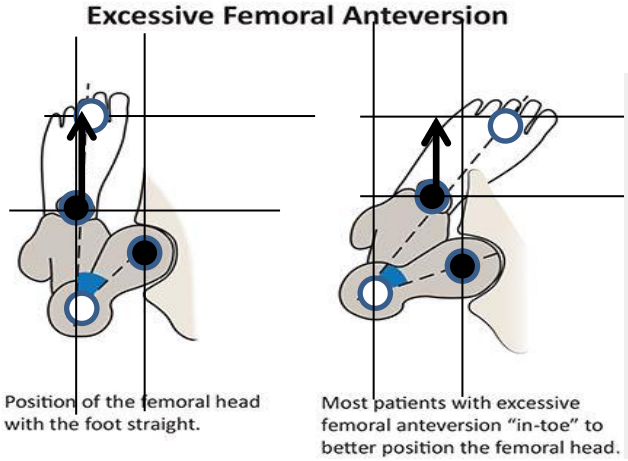
Positional anomalies
- Crouch gait

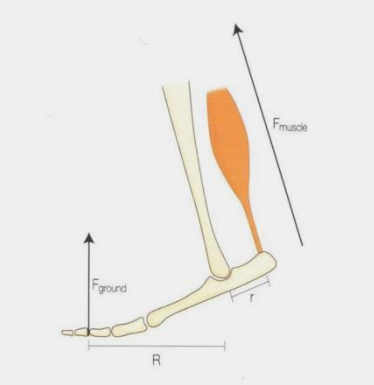
Foot deformities
-Equinus
-Equino-(plano)-valgus



Femoral anteversion

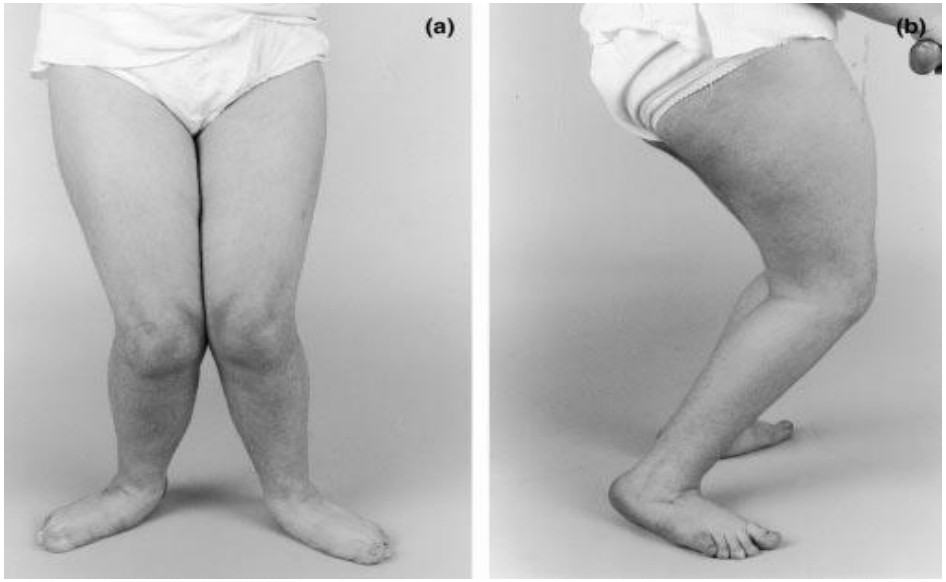


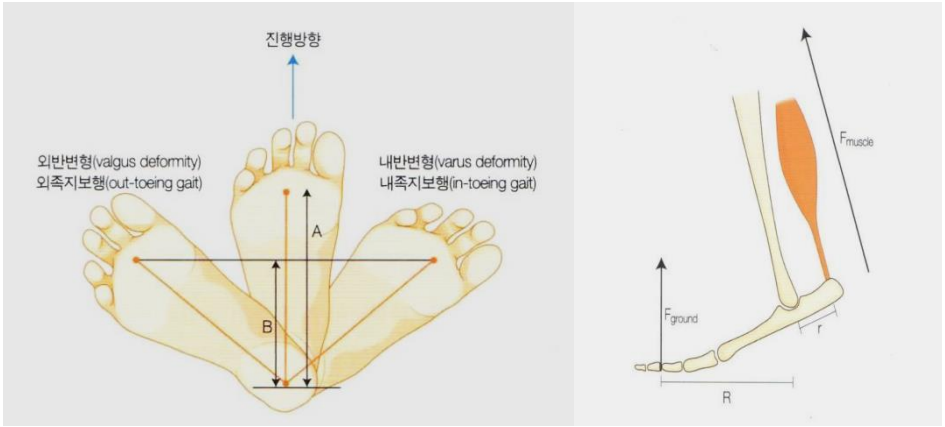




- Increased femoral anteversion compromises the lever arm of the hip abductors as the projection of the femoral neck length in the coronal plane is shortened
- Internal hip rotation may represent a compensation to bring the full femoral neck length to the coronal plane and increase the lever arm of the abductors
- Internal hip rotation during gait also affects the loading of knee and foot
- As the whole limb is internally rotated, the projection of the lever arm of forces acting in the sagittal plane is shortened
- For example, the internally rotated foot will represent a poor lever arm during push-off, as it projected length in the sagittal plane is shortened

External tibial torsion





- External tibial torsion is associated with equino-valgus foot deformity
- This compromises the alignment between the knee and the foot in the transverse plane
- The plantarflexor's moment is compromised as the projected length of the foot in the sagittal plane is shortened
- This further affects any pre-existing weakness of the plantarflexors
- The inadequate plantarflexor moment has a detrimental effect on knee extension
- The ineffective plantarflexors allow the tibia to progress forward during stance and allow the knee to flex excessively, leading to a crouched gait

Gage, 1991

The PF-KE couple

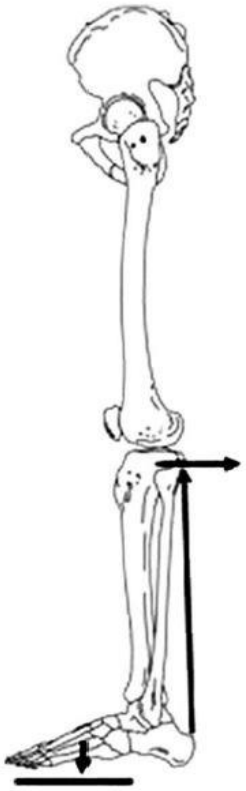
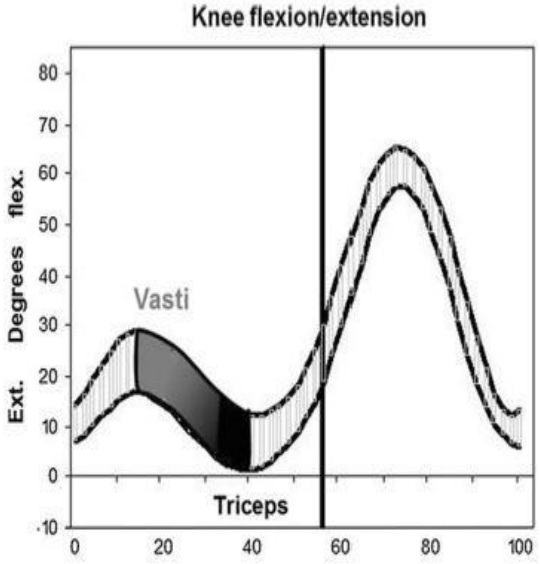
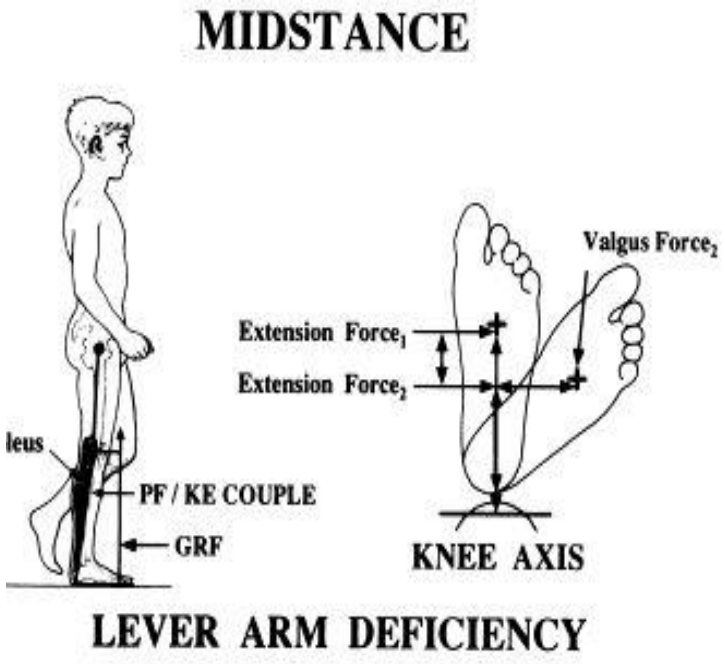


Fig. 1 Schematic drawing of the plantar flexion–knee extension couple (working only under load)

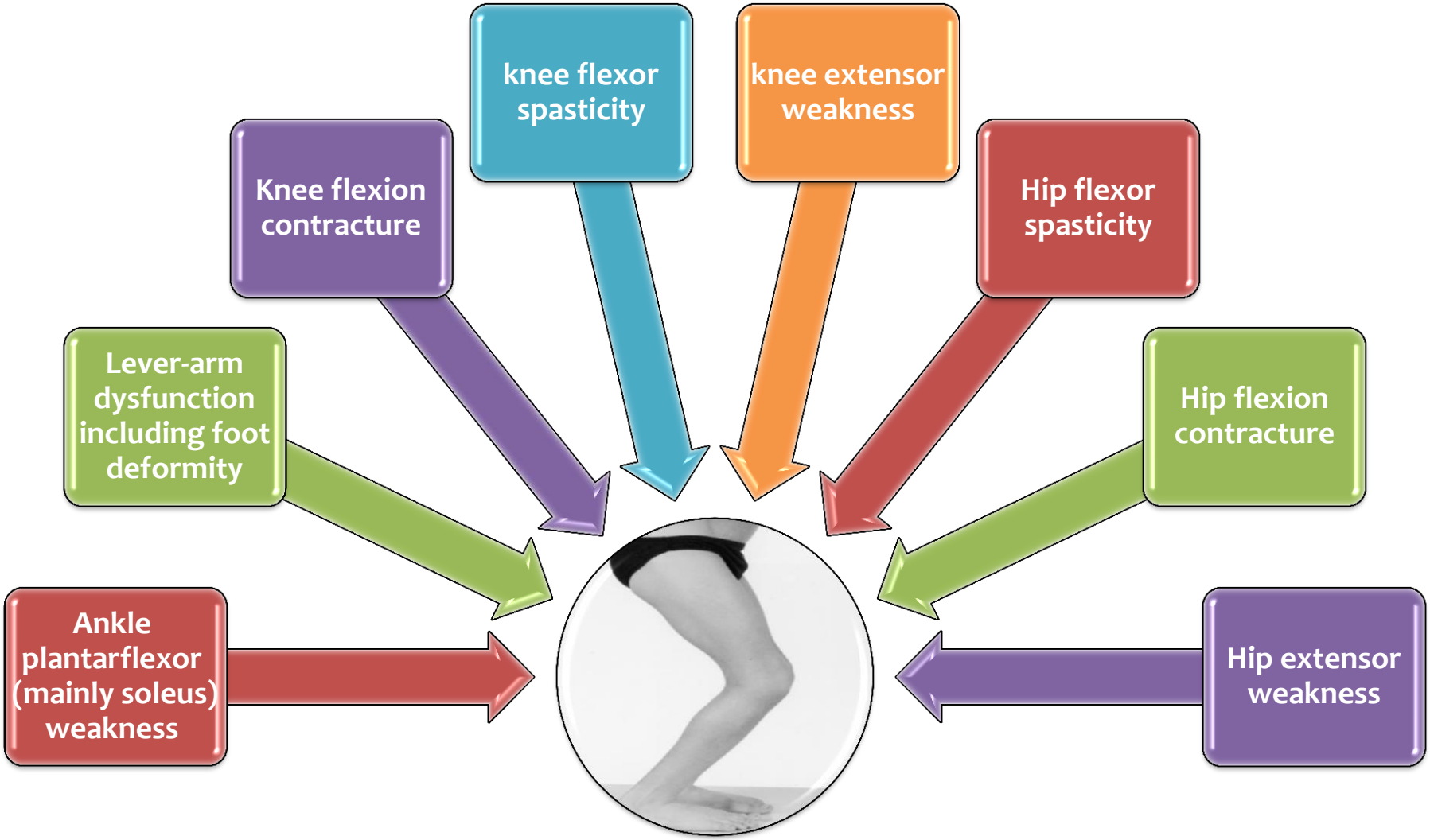




Critical biomechanical concept in understanding interrelationships between the foot-ankle and knee levels !

Brunner R et al., 2013

Crouch gait


knee flexor spasticity
knee extensor weakness
Knee flexion contracture
Hip flexor spasticity
Lever-arm dysfunction including foot deformity
Hip flexion contracture
Ankle plantarflexor (mainly soleus) weakness
Hip extensor weakness

Crouch gait

- GRF
  - **Person A : 500 N**
  - **Person B : 500 N**
- Perpendicular distance between the ground reaction force and knee joint
  - **Person A : 0.08m**
  - **Person B : 0.2m**
- External knee flexor moment
  - **Person A : 500 X 0.08 = 40 N·m**
  - **Person B : 500 X 0.2 = 100 N·m**
- **External knee flexor moment=Internal knee extensor moment**
  - **Person A : 500 X 0.08 = 40 N·m**
  - **Person B : 500 X 0.2 = 100 N·m**
- Perpendicular distance the quadriceps muscle acts away from the joint
  - **Person A : 0.06 m**
  - **Person B : 0.06 m**
- Force in quadriceps (M/0.06)
  - **Person A : 40 /0.06= 666.7 N**
  - **Person B : 100 /0.06= 1666.7 N**

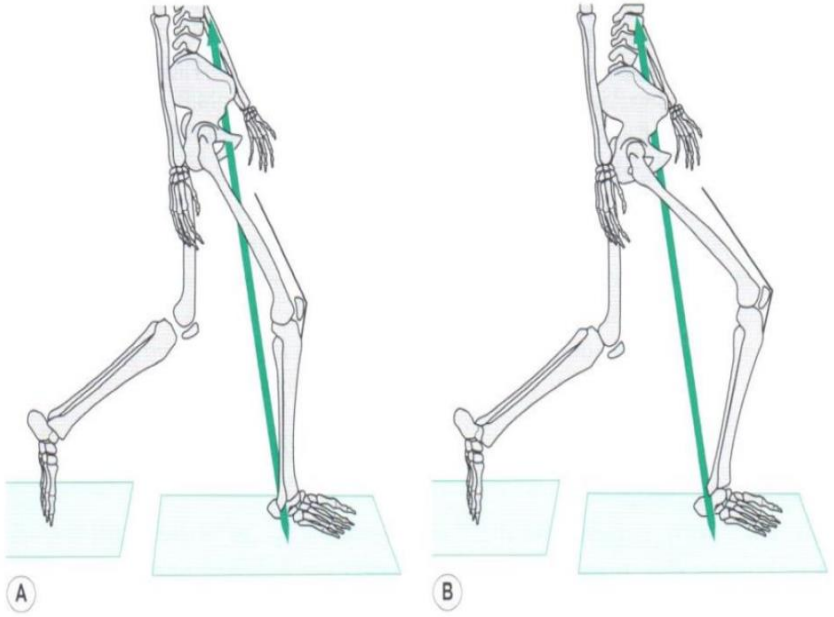


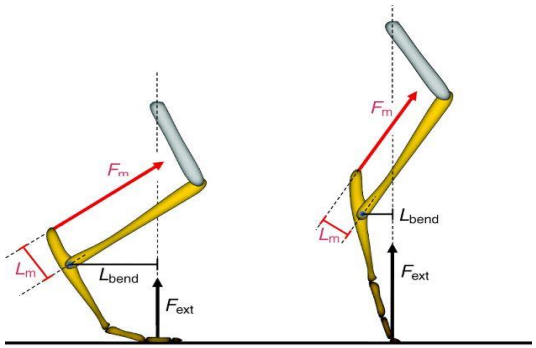
Fig. 1.23 • Ground reaction forces in (A) normal gait and (B) crouch gait.

Much greater external knee flexor moment
Much greater internal knee extensor (quadriceps) force demand

Much greater external hip flexor moment
Much greater internal hip extensor (gluteals & hamstring) force demand

Foot & ankle deformities in spastic CP

- Common among pediatric patients with CP at a reported rate of high as **93%**
- A spastic muscle group → overpower a normal or weak muscle group → abnormal foot position
- Flexible, dynamic deformity → soft tissue contracture → osseous deformity
- **Foot deformities**
  - **The position of ankle : equinus, calcaneus**
  - **The position of hindfoot : varus, valgus**
  - **The position of midfoot : cavus, planus**
  - **The position of foefoot : adductus, abductus**



- The most common foot deformities in children with CP are **equinus, planovalgus and equinovarus or equinocavovarus**

O'Connell PA et al., 1998; Kedem P et al., 2015

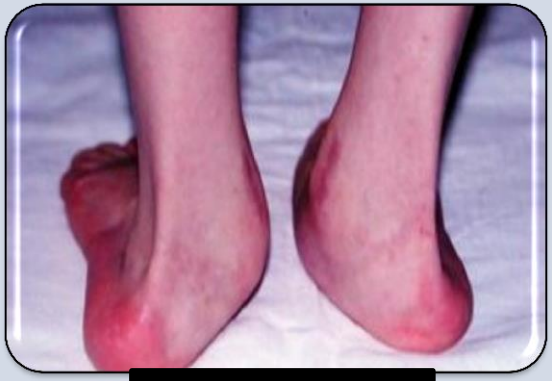
Foot & ankle deformities in spastic CP

equinus

The most common foot deformity among children with CP

Overactivation of the triceps surae complex compared with the ankle dorsiflexor

Midfoot and forefoot are in neutral



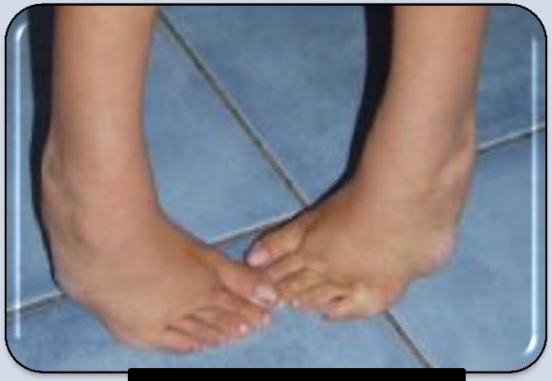
planovalgus

Commonly seen among diplegic and quadriplegic CP

Lateral column of foot : shorter than medial column

Accompanied and exacerbated by equinus

External tibial torsion frequently develops over time



equinovarus

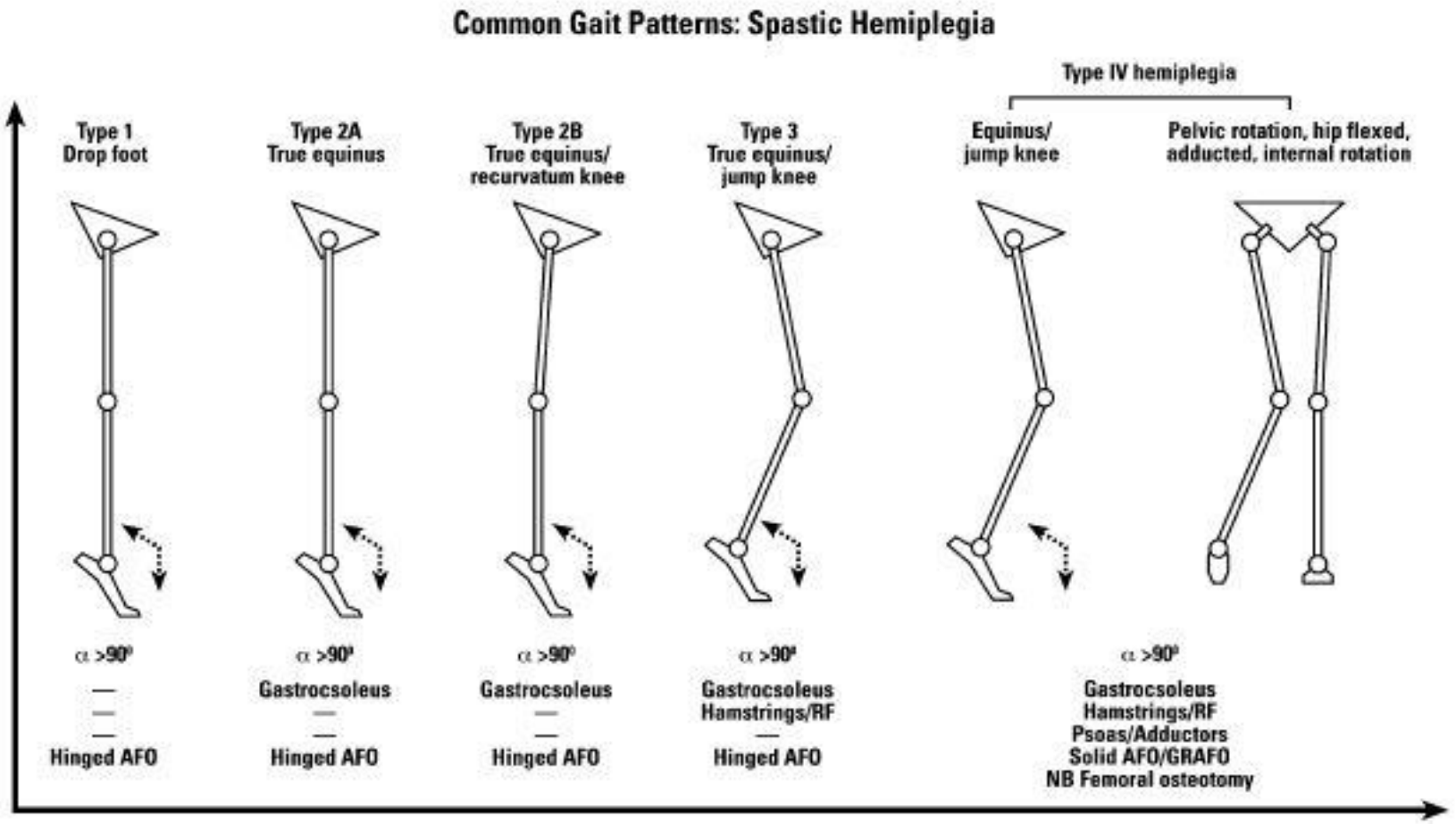
Observed mostly in hemiplegic patients

TP spasticity + TA overactivity

Cavovarus may be associated with equinus

The foot is typically stiff, with poor shock absorptioh & diminished WB area

Gait patterns in unilateral spastic CP

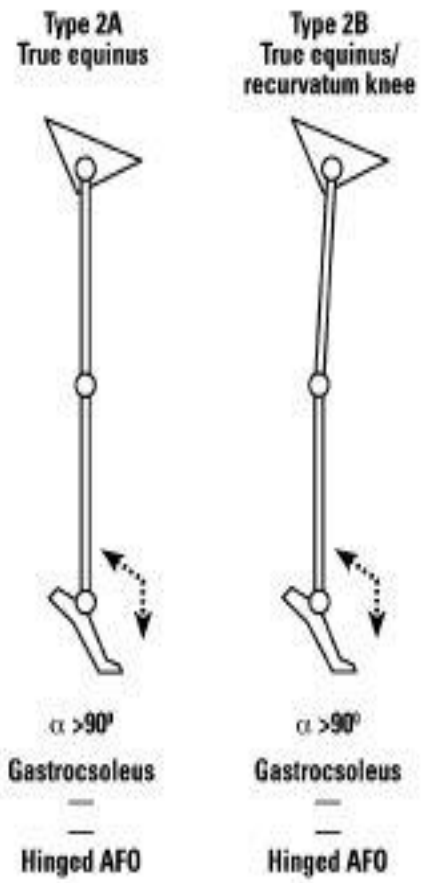


Rodda J et al., 2001

Type 1 drop foot

- Noted most clearly in the swing phase of gait due to inability to selectively control the ankle DF during this part of gait cycle
- Ankle DF during stance phase is normal due to no calf contracture
- This gait pattern is relatively rare
- Management
  - Spasticity management : not applicable
  - Contracture management : not applicable
  - Orthotic management : leaf spring or hinged AFO

Type 2 true equinus

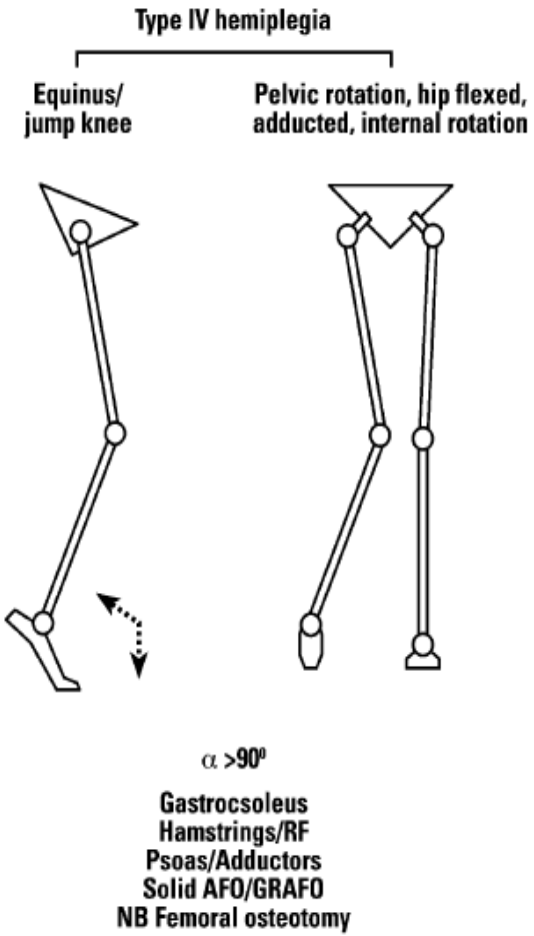


- The most common type in clinical practice
- True equinus is noted in the stance phase of gait because of spasticity and/or contracture of GCM-soleus
- The PF-KE couple is overactive and the knee may adopt a position of extension or recurvatum
- management
  - Spasticity management : BTX-A injection
  - Contracture management : Tendo Achilles lengthening, Strayer calf lengthening, if the contracture is confined to the GCM
  - Orthotic management : Hinged AFO or leaf spring AFO
  - Equinovarus deformity : additional injection of TP or by split transfer of TP
  - Equinovalgus deformity : calf injection + AFO
  - Older children with progressive valgus deformity : os calcis lengthening or subtalar fusion

Type 3 true equinus/jump knee

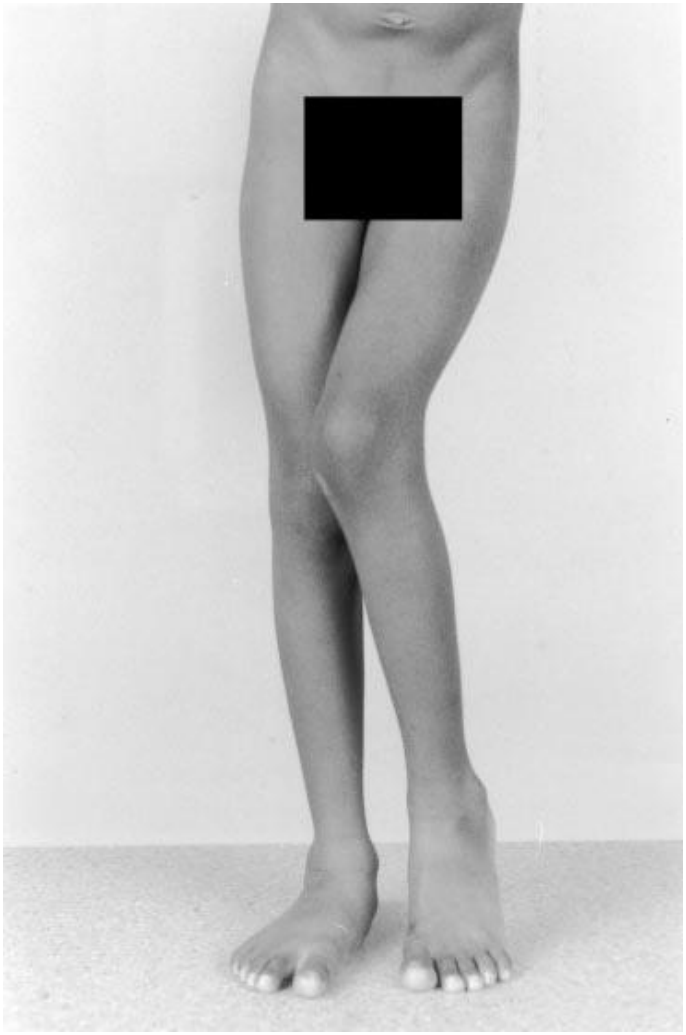
- GCM-soleus spasticity or contracture, impaired ankle DF in swing and a flexed, 'stiff knee gait' as the result of hamstring/quadriceps co-contraction
- Management
  - Spasticity management : BTX-A injections to the calf and hamstrings
  - Contracture management : tendo achillis lengthening combined with lengthening of the medial hamstrings and tranfer of the rectus femoris to the gracilis or semitendinosus
  - Orthotic management : solid or hinged AFO, according to the pre and post intervention integrity of the PF-KE couple

Type IV hemiplegia



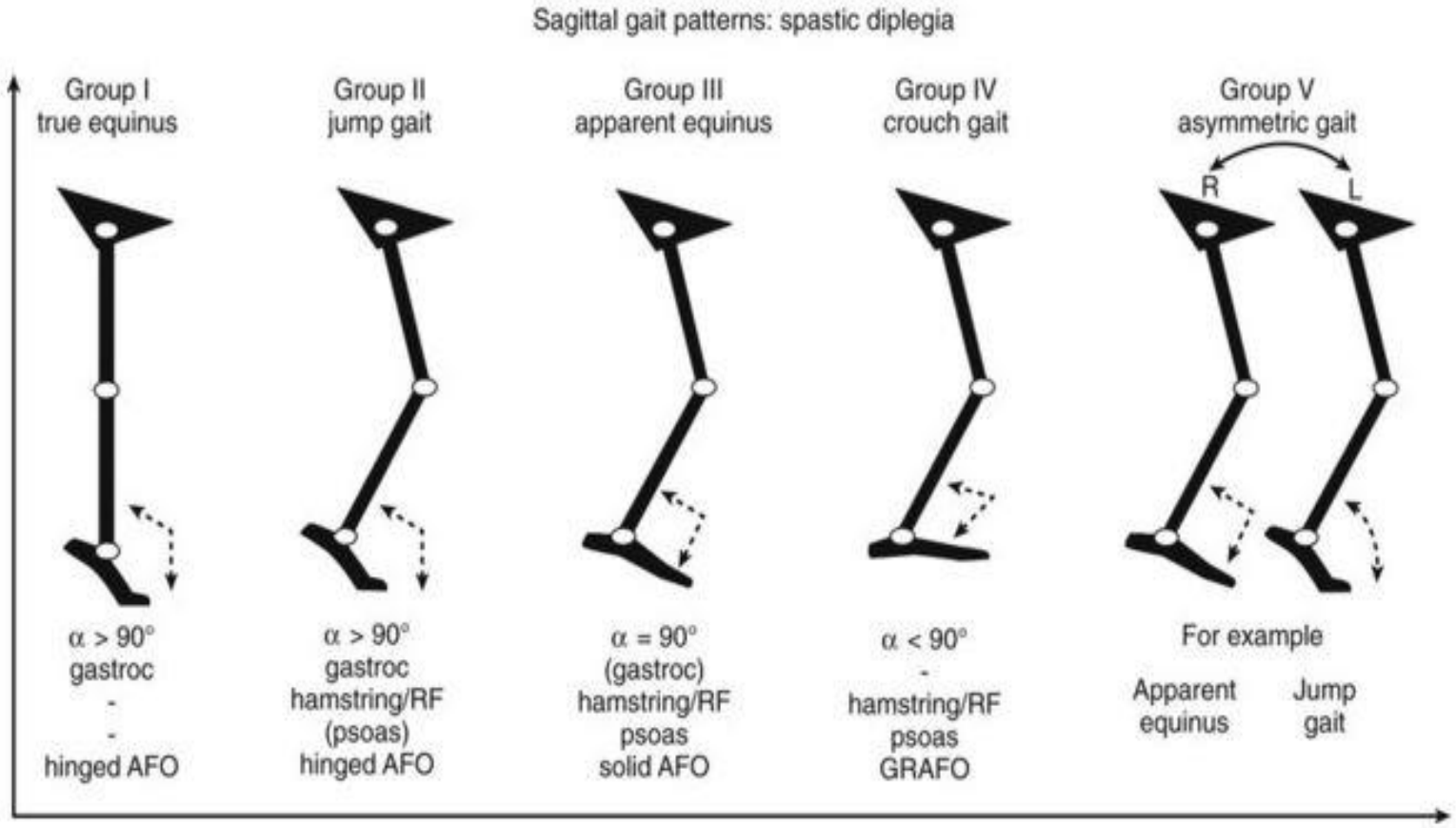
- Much more marked proximal involvement
- The pattern is similar to that seen in spastic diplegia
- Marked asymmetry including pelvic retraction due to unilateral involvement
- Failure to address the hip adduction and hip internal rotation → any distally focused intervention will fail and overall outcome will be poor
- Management
  - Spasticity management : multilevel injections of BTX-A including the calf and hamstrings, sometimes hip adductors and hip flexors
  - Contracture/deformity management : lengthening of the calf, the medial hamstrings (with rectus femoris transfer when indicated), hip adductors and iliopsoas. External rotation osteotomy of the femur
  - Orthotic management : ground reaction AFO, solid AFO or hinged AFO, according to the integrity of the PF-KE couple

Type IV hemiplegia



- Boy, severe left-sided spastic hemiplegia
- A fixed equinus at the ankle, a flexed knee, a flexed adducted and internally rotated hip
- Gait analysis showed all of these deformities had the expected effect on gait and that the knee was both flexed and very stiff throughout the gait cycle
- Management consisted of lengthening of the tendo Achillis, lengthening of the medial hamstrings and transfer of rectus femoris to semitendinosus, lengthening of adductor longus, lengthening of psoas at the brim of the pelvis and a left proximal femoral derotation osteotomy.
- Postoperatively he was managed in a solid AFO for the first 6 months and after his gastroc-soleus regained strength, a hinged AFO allowed a more normal gait pattern

Gait patterns in bilateral spastic CP



Rodda J et al., 2004

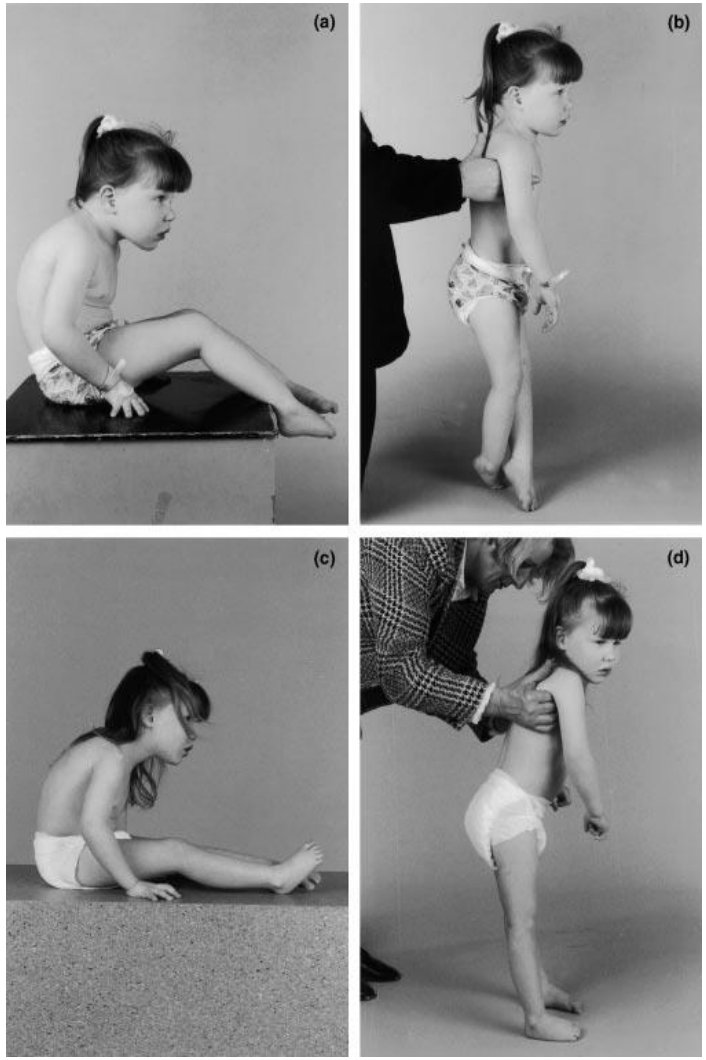
Group 1 true equinus

- When the younger child with diplegia begins to walk, calf spasticity is frequently dominant resulting in a 'true equinus gait'
- True equinus may be hidden by the development of recurvatum at the knee
- The persistence of this pattern is unusual and seen in only a small minority of children with diplegia
- management
  - Spasticity management : BTX-A injection to the calf
  - Contracture management : lengthening of the gastrocnemius
  - Orthotic management : solid or hinged AFO, according to the integrity of the PF-KE couple

Group 2 jump gait (with or without stiff knee)

- Very commonly seen in children with diplegia, who have more proximal involvement, with spasticity of the hamstrings and hip flexors in addition to calf spasticity
- The jump knee pattern and stiff knee pattern frequently coexist
- management
  - Spasticity management : in younger/less involved children BTX-A injections to the calf and hamstrings. Multilevel injections of BTX-A may be useful. SDR may be the optimum choice for small group of children
  - Contracture management : single event multilevel surgery, addressing all contractures and lever arm dysfunction
  - Orthotic management : ground reaction AFO, solid or hinged AFO, according to the integrity of the PF-KE couple

Group 2 jump gait



This 3-year-old-girl has severe spastic diplegia and is delayed in standing and walking.

(a) She has difficulty in long sitting
she sits on a posteriorly tilted pelvis with flexed knees.

(b) When supported in standing, she has a `jump gait' pattern with equinus at the ankle, flexion at the knee and flexion at the hip.
Clinical examination confirmed that her problems were mostly the result of spasticity in these muscle groups, not contracture.
She was managed by injection of botulinum toxin type A at a dose of 4 U/kg BW to the hamstrings bilaterally and to the gastroc- soleus bilaterally.
The total dose of botulinum toxin used was 16 U/kg BW

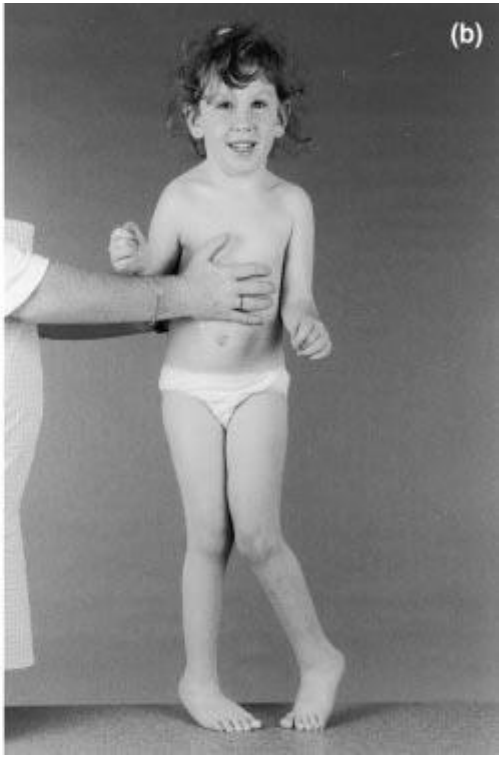
(c) It can be seen that she can now `long sit‘ more comfortably

(d) Her standing posture is improved.
Her feet are planti-grade, her knees extend fully but her hip flexion has increased as has her anterior pelvic tilt and lumbar lordosis.
She would have benefited from injections to the iliopsoas

Group 3 apparent equinus (with or without stiff knee)

- As the child gets older and heavier, equinus may gradually decrease as hip and knee flexion increase
- There is frequently a stage of 'apparent equinus'
- At this stage, weakening the gastrocnemius by injections of BTX-A or lengthening of the GCM will only provoke a crouch gait and result in impaired function
- management
  - Spasticity management : in younger/less involved children BTX-A injections to the hamstrings and iliopsoas.
  - Contracture management : single event multilevel surgery, addressing all contractures and lever arm dysfunction
  - Orthotic management : ground reaction AFO, solid or hinged AFO, according to the integrity of the PF-KE couple

Group 3 apparent equinus (with or without stiff knee)



(a) This 4-year-old girl stands on her toes with her heels on the ground.
There is also increased hip and knee flexion.
This is apparent equinus.
The foot is at 90° to the leg, the heel is off the ground because of the hip and knee flexion.

(b) In addition to the contractures of the iliopsoas and hamstrings, she has an intoed stance and gait because of bilateral medial femoral torsion, another type of lever arm dysfunction.

Group 4 crouch gait (with or without stiff knee)

- Defined as excessive dorsiflexion or calcaneus at the ankle in combination with excessive flexion at the knee and hip
- Natural history of the gait disorder in more severe diplegia and in the majority of children with spastic quadriplegia
- The commonest cause of crouch gait is isolated lengthening of the heel cord in the younger child
- Unattractive, energy expensive gait pattern, followed by anterior knee pain and patellar pathology in adolescence
- management
  - Spasticity management : in younger/less involved children BTX-A injections to the hamstrings and hip flexors
  - Contracture management : single event multilevel surgery, addressing all contractures and bony torsional abnormalities and joint instability
  - Orthotic management : long-term use of a ground reaction AFO until the integrity of the PF-KE couple is clearly re-established

Group 4 crouch gait (with or without stiff knee)

- This is the classic sagittal profile of crouch gait
- There is excessive ankle dorsiflexion, increased knee flexion and increased hip flexion
- The plantar flexion knee extension couple is incompetent with the ground reaction vector directed behind the knee
- This boy was managed by lengthening of the hamstrings and iliopsoas with ground reaction AFOs to redirect the ground reaction vector in front of the knee

Take home message

- **Imbalance of the inhibitory and excitatory influences on spinal and supraspinal pathways is thought to be the cause of spasticity**

- **Hypertonia in patients with UMNS can be divided into two components (spasticity and intrinsic hypertonia)**

- **If we try to understand the gait patterns in children with spastic CP according to growth, all motor problems including primary, secondary, and tertiary problems should be evaluated**

- **The gait patterns in children with spastic CP can be most accurately identified by using a combination of clinical examination (assessment of dynamic and fixed contracture), video recording of gait and instrumented gait analysis**

Thank you for your attention!