

Gait rehabilitation in people with hereditary spastic paraplegia

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Learning objectives

- To recognize the typical gait pattern in HSP
- To understand that management of gait impairments involves complementary non-pharmacological, pharmacological and surgical interventions

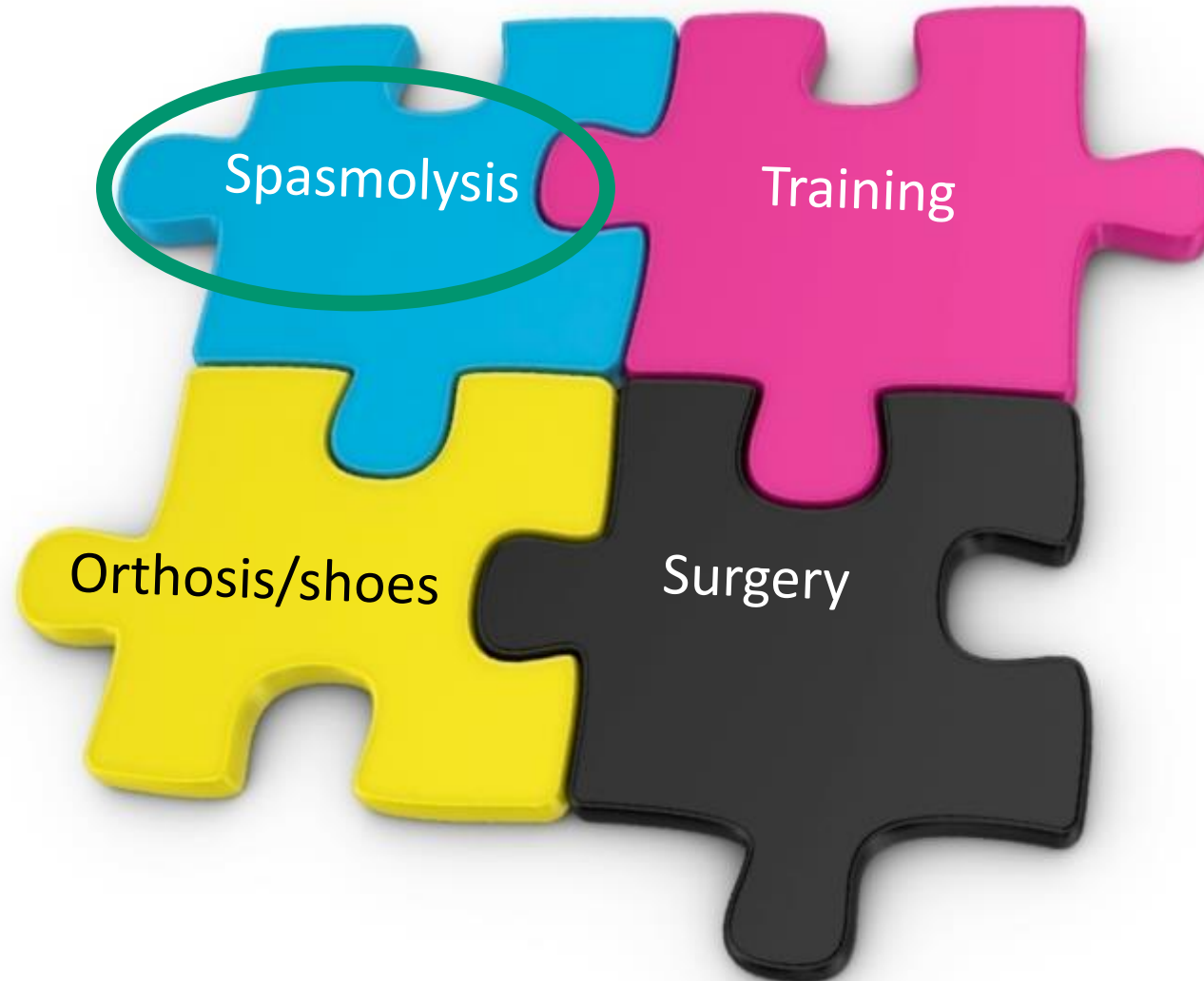
Gait pattern in hereditary spastic paraplegia



Why do gait impairments emerge?

- Spasticity
- Muscle weakness
- Reduced selectivity
- Difficulties to make rapid leg movements
- Impaired proprioception
- Contractures

Gait rehabilitation in hereditary spastic paraplegia



Focal spasms

Hereditary spastic paraplegia: from diagnosis to emerging therapeutic approaches

Samuel Shribman, Evan Reid, Andrew H Crosby, Henry Houlden, Thomas T Warner

Hereditary spastic paraplegia (HSP) describes a heterogeneous group of genetic neurodegenerative diseases characterised by progressive spasticity of the lower limbs. The pathogenic mechanism, associated clinical features, and imaging abnormalities vary substantially according to the affected gene and differentiating HSP from other genetic diseases associated with spasticity can be challenging. Next generation sequencing-based gene panels are now widely available but have limitations and a molecular diagnosis is not made in most suspected cases. Symptomatic management continues to evolve but with a greater understanding of the pathophysiological basis of individual HSP subtypes there are emerging opportunities to provide targeted molecular therapies and personalised medicine.

raisCS, is also valuable for mobility.

Oral antispasmodics, including baclofen and tizanidine, have an established role, although most patients do not gain significant clinical benefit. Gabapentin has been used for spasticity in multiple sclerosis and spinal cord injury, but a double blind crossover trial in a small cohort of SPG4 cases found no difference in effect from placebo.⁶⁹ An open-label trial of fampridine (4-aminopyridine) in a mixed cohort of HSP (SPG4, SPG7, SPG11) over 2 weeks suggested some benefit and deserves more rigorous testing.⁷⁰ With more severe spasticity intrathecal baclofen can be effective in reducing very high tone associated with pain and disability. It is often used for patients requiring wheelchairs, but also has been shown to improve gait if used earlier.⁷¹

Botulinum toxin injections can be helpful for targeting specific problematic muscle groups around the ankle, knee, and hip. One trial enrolled patients with SPG4, SPG3A, and SPG8 and studied the combination of

Instrumented gait analysis



Q1: I apply instrumented gait analysis before starting with focal spasmolysis

1. Yes
2. No

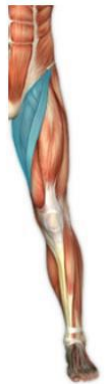
Focal spasmolysis

- Common target muscles:
 - Plantarflexors
 - Hip adductors

Feboch studies



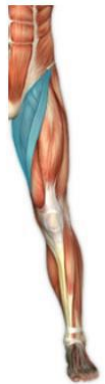
	Feboch I	Feboch II
Gait problem	toe walking	scissoring gait
Critical muscles	calves	hip adductors
Number of patients	15 (≥ 18 yrs)	25 (≥ 18 yrs)



Feboch studies



Treatment	Feboch I	Feboch II
<i>Medication</i>	Dysport®	Xeomin®
<i>Dose</i>	bilateral 500 U (MAS 1) or 750 U (MAS 2)	bilateral 150 U (MAS 1) or 200 U (MAS 2-4)
<i>Administration</i>	9 deposits per side (soleus and gastrocs)	7 deposits per side (add longus, add magnus and gracilis)
<i>Exercise</i>	2 x daily muscle stretching	3 x daily muscle stretching



Pre-post design

Treatment



Outcome measures

	Feboch I	Feboch II
Primary outcome	Comfortable gait speed	Gait width
Mod. Ashworth	+	+
MRC strength	+	+
Gait speed	maximal	comfortable + maximal

T0 (baseline) -T1 (4-6 weeks)

	Feboch I	Feboch II
Primary outcome	Comfortable gait speed (+9%)	Gait width (+12,6%)
Mod. Ashworth	↓	↓
MRC strength	↓	↓
Gait speed	Maximal =	Comfortable (+8.3%) Maximal =

T0 (baseline) -T2 (16-18 weeks)

	Feboch I	Feboch II
Primary outcome	Comfortable gait seepd (+12%)	Gait width (+9.7%)
Mod. Ashworth	↓	↓
MRC strength	=	=
Gait speed	Maximal =	Comfortable gait seepd (11,5%) Maximal =

Interim conclusions

Botulinum toxin in critical muscles in HSP results in:

- Reduction of muscle tone
- Slight, temporary loss of muscle force
- 8-12% increase in comfortable gait speed (both studies)
- 10-12% increase in gait width (FEBOCH II)

Randomized controlled trials are needed

Gait rehabilitation in hereditary spastic paraplegia



Training should be task specific



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Design MOVE-HSP

- Five-week, two-armed, open-label randomized RCT
- 36 people with pure HSP
- 1:1 randomized into intervention of control group
- Intervention: 10 one-hour sessions of gait adaptability training on C-mill
- Control group: usual care
- Primary outcome is the obstacle subtask of the Emory Functional Ambulation Profile
- Clinicaltrials.gov identifier: NCT04180098

Q2: Will levels of physical activity impact on gait impairments?

1. Yes, reduced physical activity will increase experienced gait impairments
2. Yes, reduced physical activity will reduced experienced gait impairments
3. No, physical activity will not impact on experienced gait impairments

Maintaining sufficient levels of physical activity is also important

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LETTER TO THE EDITORS



COVID-19 reveals influence of physical activity on symptom severity in hereditary spastic paraplegia

Lotte van de Venis¹ · Bart P. C. van de Warrenburg² · Vivian Weerdesteyn¹ · Bas J. H. van Lith¹ · Alexander C. H. Geurts^{1,3} · Jorik Nonnekes^{1,3}

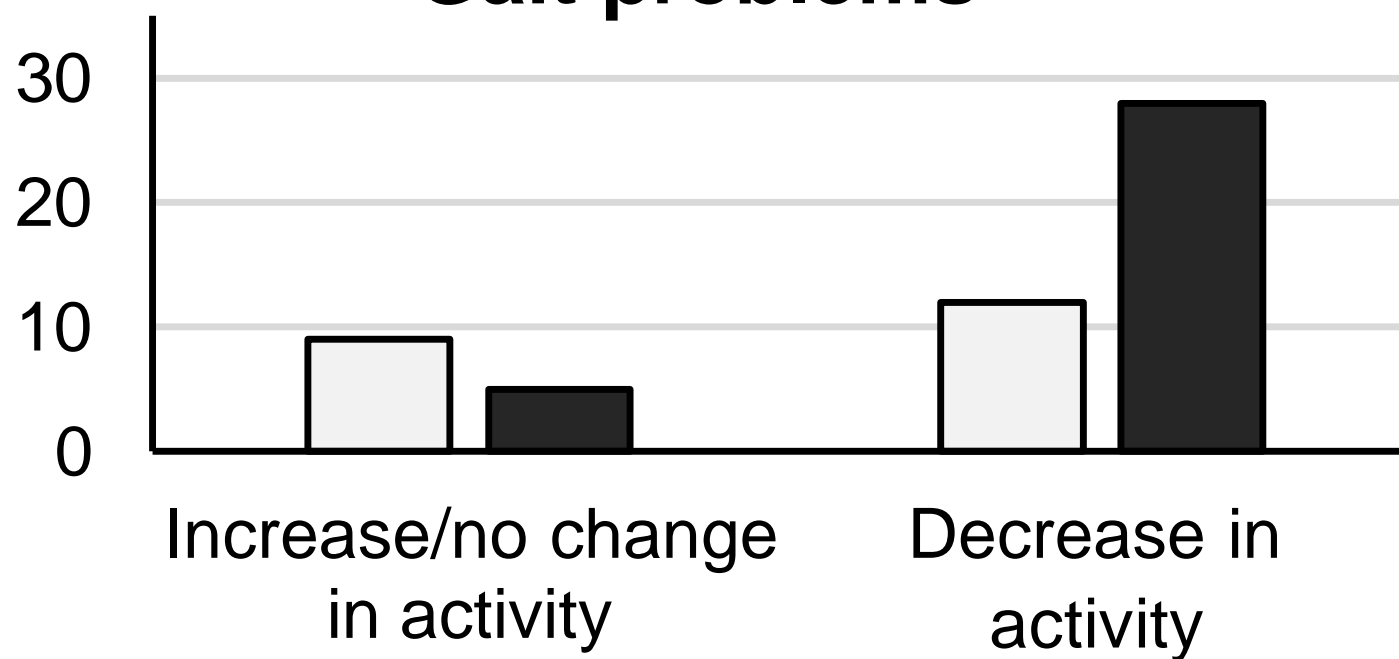
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Impact of Covid-19 lockdown on experienced symptom severity in HSP

- Web-based survey
- 109 people with pure HSP, 58 responders
- Reduction of physical activities by 74%
- Majority of patients also reported increase in symptom severity
 - 68% increase in muscle stiffness
 - 60% subjective increase in gait problems

Impact of physical activity on symptom severity

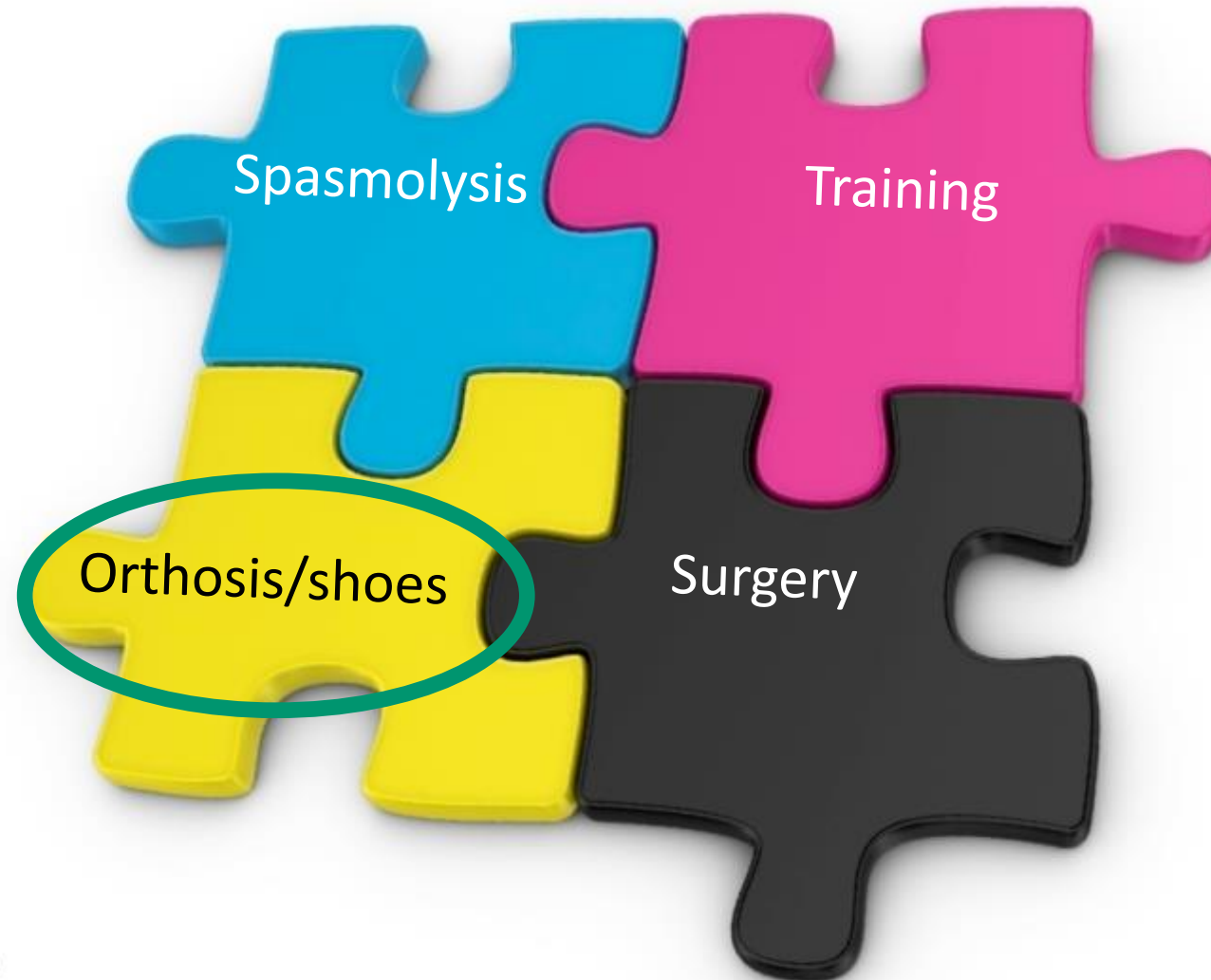
Gait problems



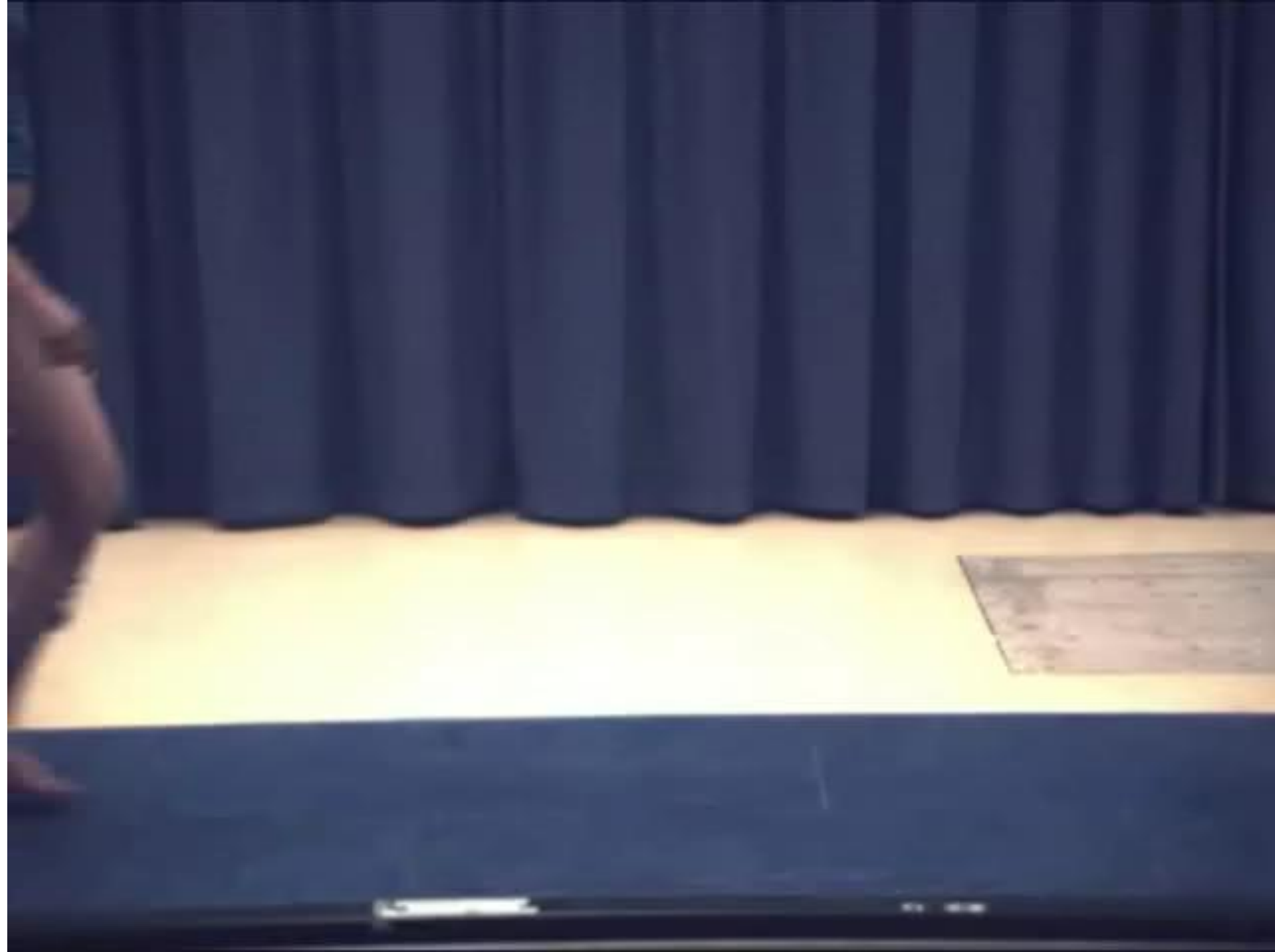
□ Decrease/no change in symptom severity

■ Increase in symptom severity

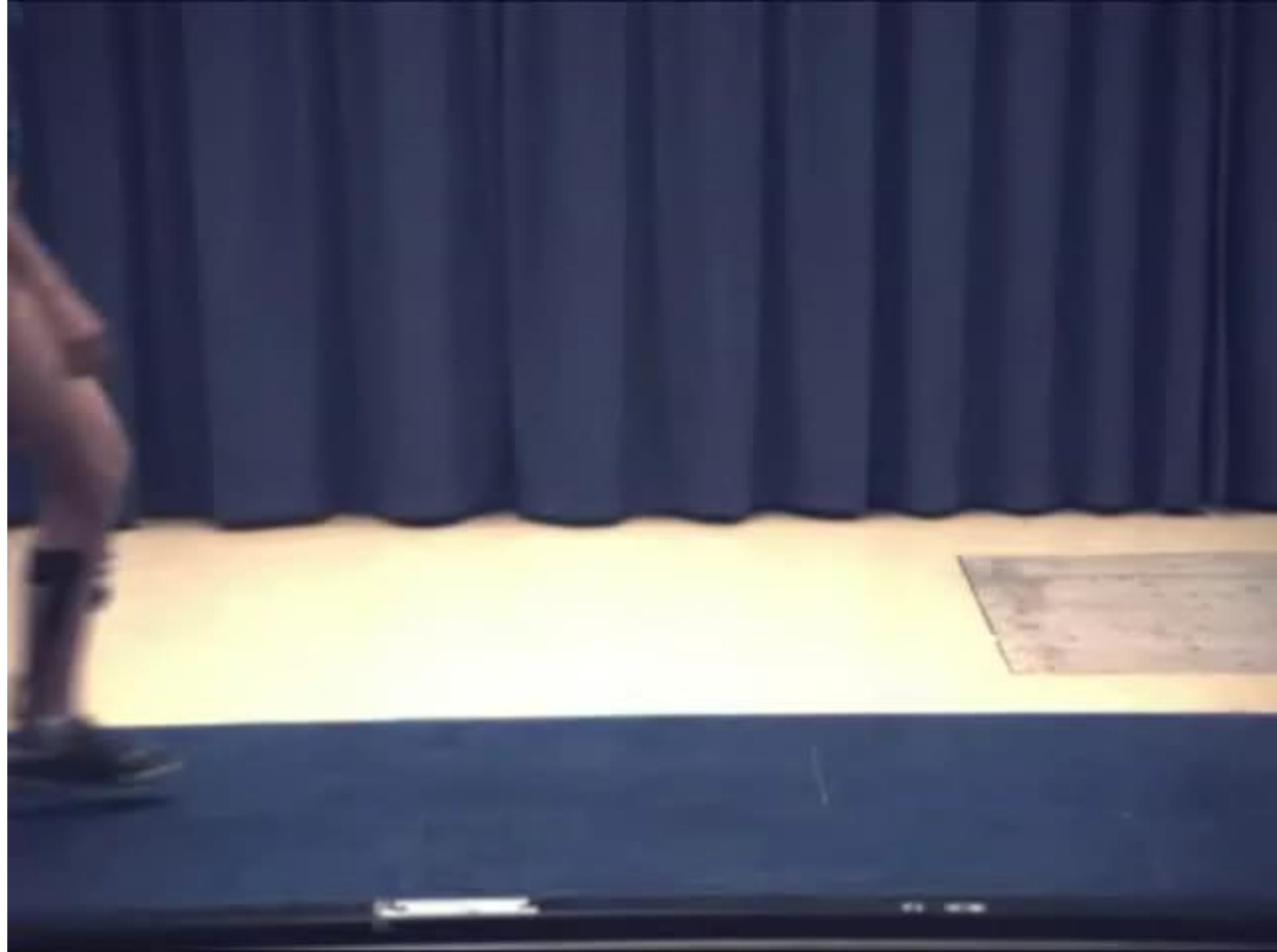
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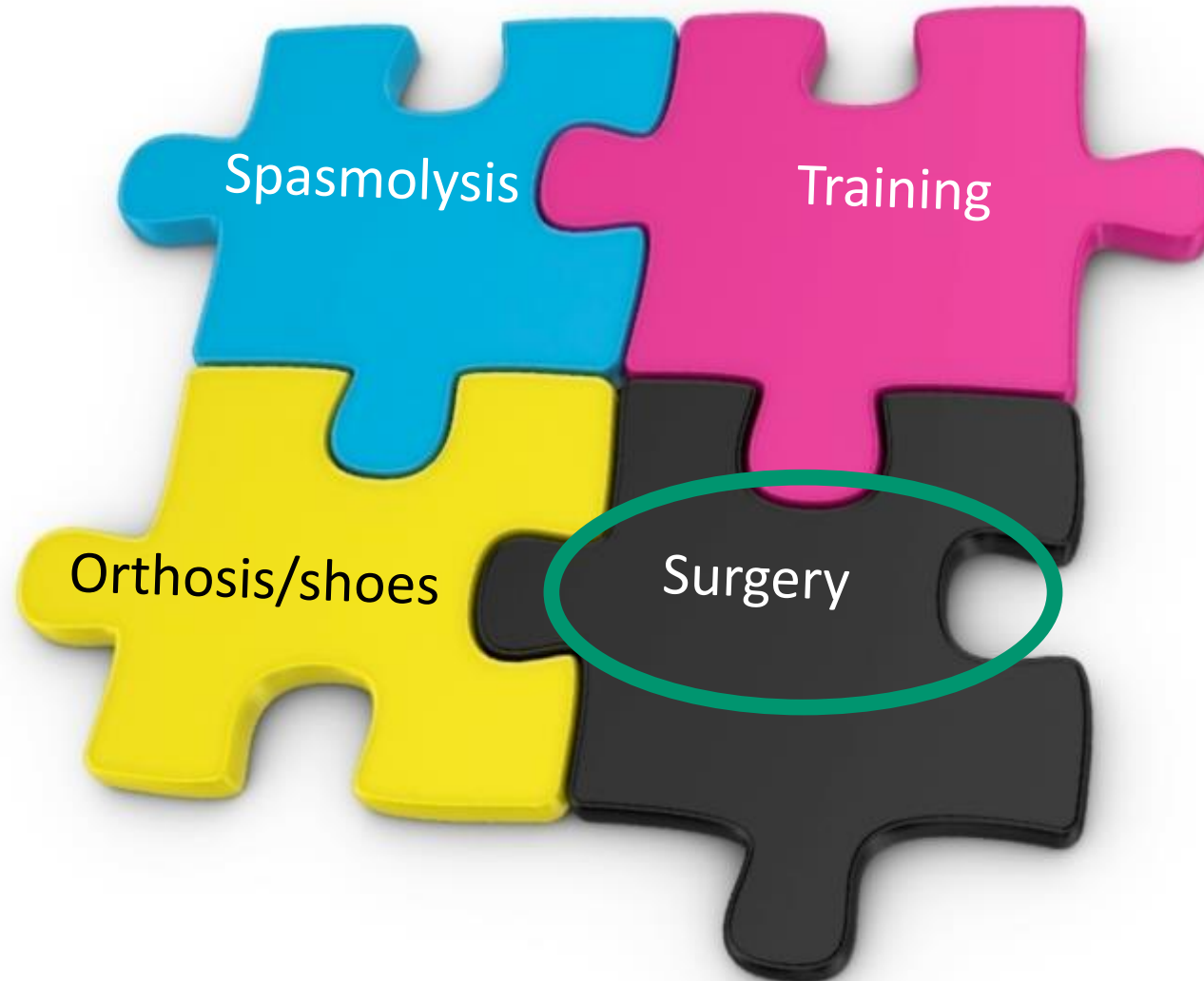








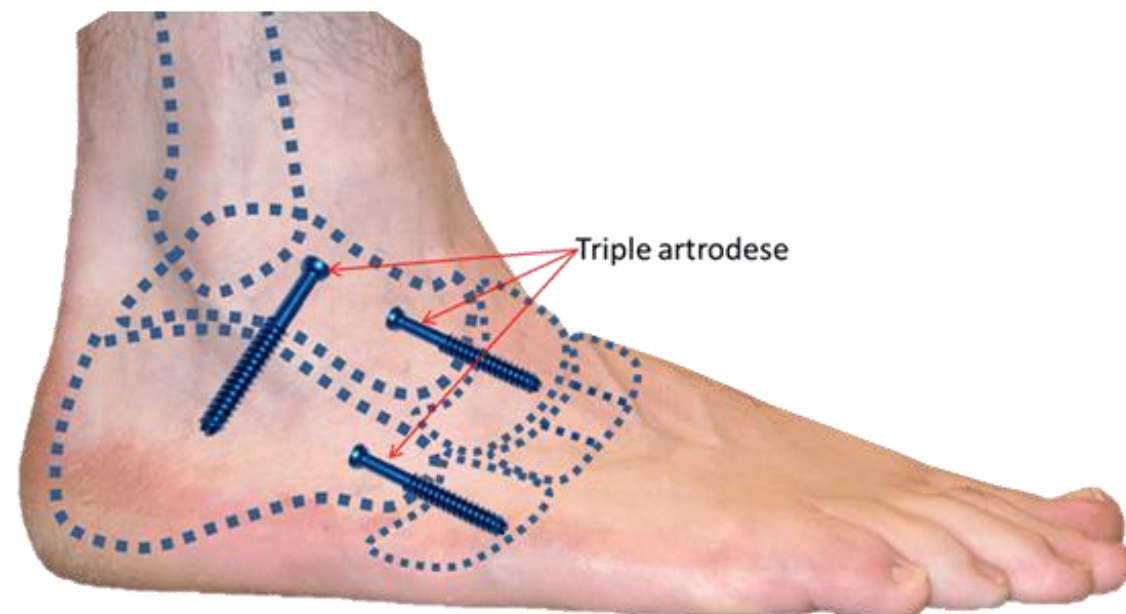
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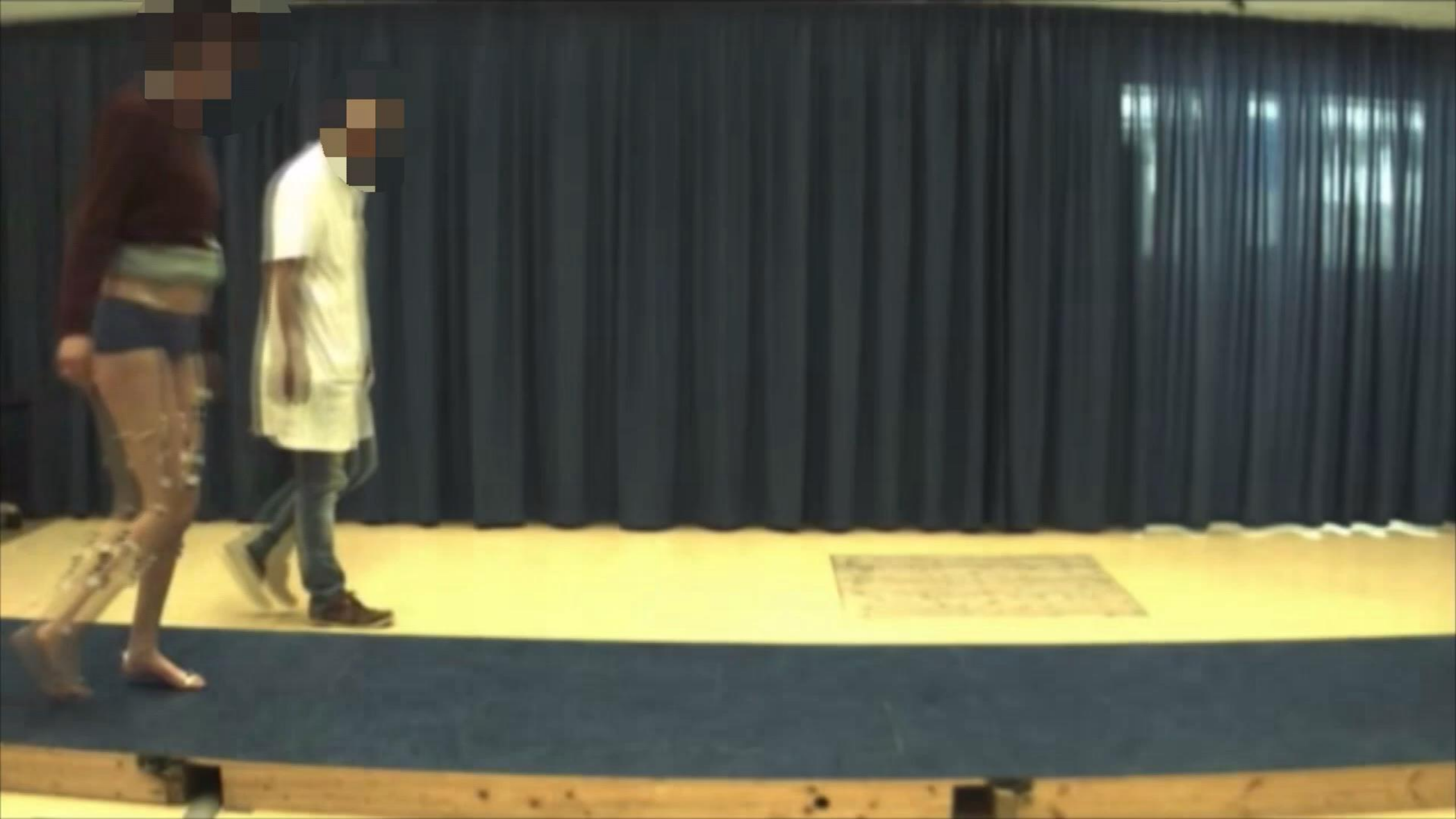


Q3: Do you consider surgical interventions in people with HSP?

- -Yes, I consider surgical interventions
- -No, I do not consider surgical interventions

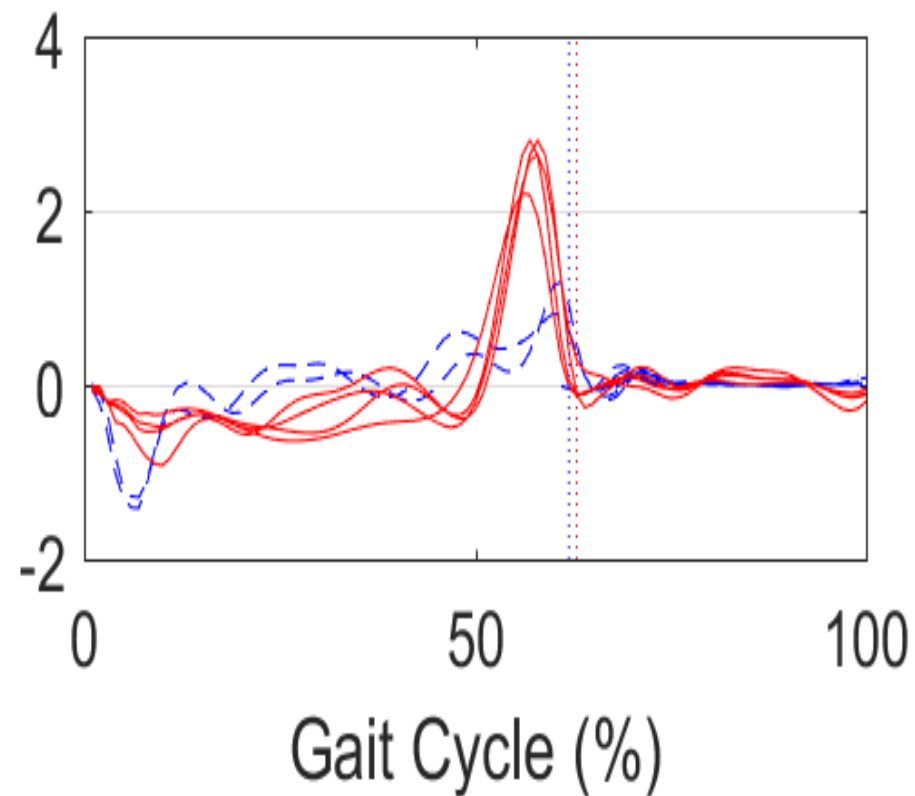
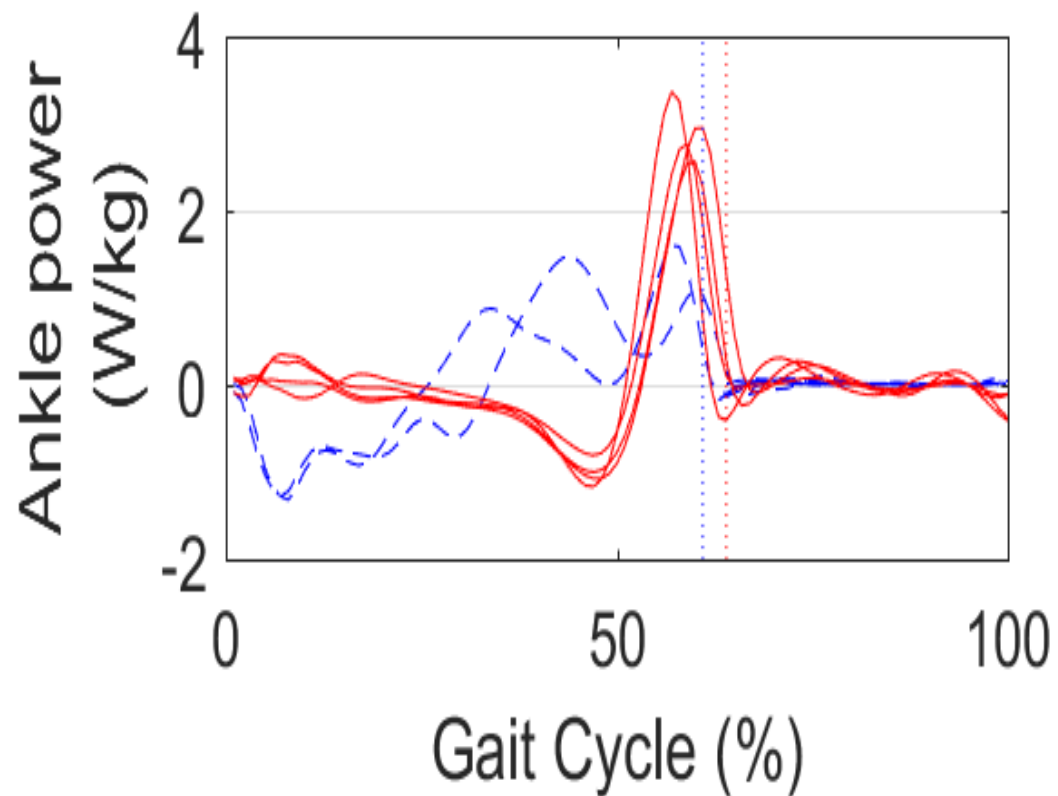
Surgery







Gain in ankle powers after surgery



Pre-surgery



Post-surgery

Key points

- Gait impairments are a key feature of hereditary spastic paraplegia
- Ideally, an instrument gait analysis is performed before starting treatment
- Treatment should be personalized and may involve complementary non-pharmacological, pharmacological and surgical interventions



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