

Neuromuscular Physiatry Sharing Session #2

Attendees:

Dr. Jessica Theriault – Presenter and Lead
Dr. Stephanie Plamondon
Dr. Xavier Rodrigue
Dr. Alasdair Rathbone
Dr. Geoffrey Frost
Dr. Beatrice Soucy
Dr. Lindsay Hubenig
Dr. Sandrine Boueilh

Summary of Case Study

Cases presentation

Case 1

An 84-year-old male presenting with an IBM-like pattern resides in a two-storey house. He has experienced an increased number of falls but declined home modifications. He has declined SC-KAFO and left knee orthosis because this prevents him from getting in the car.

Case 2

F 27 with SMA type 3 (4 copies SMN2), onset at 13 yo, on Nusinersen since 2019. Active lifestyle as she is the owner of an apartment rental company. She uses bilateral canes and she mostly avoid high-risk situations reporting no falls.

Case 3

M 75 with myofibrillar myopathy diagnosed in 2013, associated lumbar stenosis and chronic gluteal pain. He is currently walking with a 4WR and reporting 1 fall per month, indicating progressive decline in walking abilities. He has been refusing to wear his AFOs since 2020.

Summary of Selected Article

The article selected for this sharing session is "**Effect of specialist care lower limb orthoses on personal goal attainment and walking ability in adults with neuromuscular disorders**" by van Duijnhoven et al. 2023. In this study, the authors compared the impact of specialist care orthoses to that of UC orthoses on mobility-focused functional assessments.

Introduction

In current clinical practice, orthotic prescriptions for NMD vary widely, lacking strong evidence of their effectiveness. However, specialized orthotic care centers have reported success with custom ankle-foot orthoses (AFOs) and knee-ankle-foot orthoses (KAFOs), improving gait and reducing energy expenditure.

This cohort study aimed to assess the effectiveness of specialist care orthoses (including AFO, SC-KAFO, and locked KAFO) in individuals with NMD experiencing gait problems.

Methods

In this study, data collected between October 2011 and March 2021 at a university hospital's outpatient polio and orthosis expertise rehabilitation clinic in Amsterdam, the Netherlands, were analyzed. All participants were included in the analysis.

Eligibility Criteria:

- a minimum age of 18 years;
- weakness of the calf muscles;
- provision of their first specialist care orthosis;
- current use of a user of UC orthosis.

Outcome Measures:

- Leg muscle strength measured as the strength of the left and right hip flexors, extensors, abductors, and adductors, as well as knee extensors and flexors, ankle dorsiflexors, and plantar flexors, scored according to the MRC scale.
- Goal attainment scaling (GAS) Score, one or two personal goals selected with the patient based on their goals for the specialist care orthosis in daily living.
- Walking speed and net energy cost assessed with the 6-minute walk test measuring oxygen uptake and respiratory exchange ratios.
- Perceived Walking Ability assessed using an in-house questionnaire.
- Satisfaction with the Orthosis and Services Provided evaluated using the Quebec User Evaluation Satisfaction questionnaire.

Results and discussion

A total of 64 subjects with NMD using UC orthosis were included in the analysis. They were prescribed with specialist care orthoses as per the Dutch national guideline for lower limb orthoses in NMD, specifically 19 AFOs, 22 KAFOs, and 23 locked KAFOs. The custom-made specialist care lower limb orthoses did not objectively improve walking speed or net energy cost, only in patients wearing AFOs (9.5%), although subjects did report that walking required less effort. Sixty-one percent of subjects demonstrated a clinically relevant improvement in GAS score (78%), while all patients showed improvements in perceived safety, stability, intensity, fear of falling, and satisfaction while walking.

Prescription of specialist orthosis following dedicated guidelines could improve goal attainment and walking ability in adults with NMD.

Group Discussion

Guidelines/Criteria used when evaluating orthoses' efficacy

At select sites, a monthly interdisciplinary orthotics clinic, comprising physiotherapists, physiatrists, orthotists, and patients/families, is organized to address the needs of patients requiring orthotics.

Orthosis efficacy is not commonly and consistently evaluated across Canadian rehabilitation centers and there are not specific guidelines on OMs to evaluate orthoses' efficacy. Among the OMs used:

- 2 minutes walk test
- 25M walk test
- Timed up and go
- video gait analysis
- Video recording (eg of patient that received botulinum toxin treatment)

Recent experiences related to prescribing lower limb orthoses for neuromuscular patients

Successes

- Carbon fiber orthoses are lighter and slides better into the shoes.

- Availability of shoes design that facilitates wearing orthosis (Nike Go FlyEase, Billy Footwear, and customized shoes.)
- Turbo Med orthoses effective for CMT patients.
- Pediatric patients benefit from being followed up by a multidisciplinary team (rehabilitation – physiotherapy.)
- Similarly to what was reported in van Duijnhoven et al. 2023, patients that had poliomyelitis are 100% compliance.
- Feedback from an individual that run a 10 k with an exoskeleton (Keeogo): he believes that he can achieve this goal as he started wearing his Keeogo since early age.
- Access to standing frame to adults in special cases

Barriers

- Patients do not always believe the orthoses might help or is needed.
- Patients experience discomfort and they immediately give up on wearing orthoses.
- KAFOs are reported to be heavy and compromise patients' balance – not frequently used.
- For subset of NMD patient population (e.g. IBM), lower limb orthoses can be useful for a limited time until they manifest severe ankle weakness.
- Orthotists working in private clinics.
- If prescribed orthosis, patients might need bigger shoes that may make them feel clumsy.
- Lack of possibility to offer rehabilitation wearing orthoses.