Neuro-muscular diseases

Oral communications

CO019
The motor function measure (MFM) in the myotonic dystrophy type 1 population: Description and responsiveness
Capucine de Lattre 1,2, Pascal Rippert 2, Guillaume Bassee 2, Dalil Hamroun 3, Isabelle Poirot 1, Carole Vuillerot 5
1 Hospices civils de Lyon, l'escale MPR pédiatrique, Bron, France
2 Hospices civils de Lyon, Medical Information Department, Lyon, France
3 AP-HP, Neuromuscular Reference Center, GH Henri-Mondor, Creteil, France
5 CNRS UMR 5558, Biometric and Biology Evolutive Laboratory, Health Biostatistic group, Lyon, France
* Corresponding author.
E-mail address: capucine.de-lattre@chulyon.fr (C. de Lattre)

Objective To assess the applicability and the responsiveness of the motor function measure 1 (MFM) in the myotonic dystrophy type 1 (DM1) population.

Materials/patients and methods We conducted an observational, retrospective, multicenter cohort study using data from the MFM database (http://www.motor-function-measure.org/data-bank.aspx). Only DM1 patients with at least one MFM-32 score were included. The distributions of the MFM scores (total score and 3 subscores) were analyzed by age. MFM responsiveness was estimated in patients with at least two MFMs (at least 6 months between the two evaluations). Hypothetical sample sizes for specific effect sizes in clinical trial scenarios are given.

Results The descriptive study includes 618 patients from 29 physical medicine and rehabilitation or neurology department aged 6.2–80.4 years. 1038 MFM-32 for DM1 patients are registered in the MFM database and 228 patients have at least two evaluations. 930 MFM were realized in adults’ patients (> 18 years old). Mean age at the MFM execution was:

MFM D1 subscore (standing and transfers) is the more sensitive score to show deterioration (–2.32 points/year). MFM D2 (proximal and axial motricity) and D3 (distal motricity) subscores showed less changes over time (–1.43 + –4.25 points/year for D2, –0.53 + –4.18 points/year for D3). Significant responsiveness was obtained with the D1 subscore (standardized response mean [SRM] = 0.550).

Discussion/Conclusion MFM scale and particularly the D1 subscore is a reliable and valid outcome measure applicable in longitudinal follow-up and clinical trials in the DM1 population.

Keywords Motor Function Measure; Myotonic Dystrophy type 1; Motor evaluation; Clinical trial

Disclosure of interest The authors have not supplied their declaration of competing interest.

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CO020
Functional improvement with a rehabilitation programme in dermatomyositis an polymyositis: Results of a randomized controlled trial
Vincent Tiffreau 1,2, François Kopciuch 1, André Thevenon 1, François Rannou 3, Eric Hachulla 1, Philippe Thoumie 2
1 CHRU de Lille, MPR, Lille, France
2 AP–HP, hôpital Rothschild, MPR, Paris, France
3 AP–HP, hôpital Cochin, MPR, Paris, France
* Corresponding author.
E-mail address: vincent.tiffreau@chru-lille.fr (V. Tiffreau)

Objective To evaluate the medium-term functional impact and effect on quality of life of a standardized rehabilitation programme in patients with polymyositis and dermatomyositis.

Materials/patients and methods Design: a multicentre, randomized, controlled trial.
– Setting: four university hospitals in France.
– Participants: 20 patients suffering from polymyositis and dermatomyositis.
– Interventions: the intervention group participated in a four-week standardized, hospital-based rehabilitation programme and then a personalized, self-managed, home-based rehabilitation programme. The control group received Physiotherapy on an outpatient basis. The study participants were evaluated at inclusion, at the end of the rehabilitation programme (1 month) and then at 6 and 12 months.
– Main outcome measure(s): the primary efficacy criterion was the health assessment questionnaire disability index (HAQ-DI), and
the secondary criteria were quality of life (according to the SF36 questionnaire), muscle performance (isokinetic strength, the motor function measure, and the Kendall manual muscle test (MMT), gait, pain, fatigue and biomarkers of tolerance and disease activity.

Results At 12 months, the mean ± standard deviation HAQ-DI was significantly lower in the intervention group than in the control group (0.64 ± 0.53 vs. 1.36 ± 1.02, respectively; P = 0.026). The intervention group also had better scores than the control group for some quality of life dimensions (SF36 General Health: 53.44 ± 8.73 vs. 36.57 ± 22.10, respectively; P = 0.003; SF36 role physical (63.89 ± 43.50 vs. 17.86 ± 37.40, respectively; P = 0.023), the Kendall MMT score (85.89 ± 16.11 vs. 65.22 ± 31.50, respectively; P < 0.05) and pain levels (5.0 ± 10.61 vs. 33.38 ± 35.68, respectively; P = 0.04) at 12 months. Lastly, the programme was well tolerated by all the participants.

Discussion/Conclusion In patients suffering from polymyositis and dermatomyositis, the combination of a four-week standardized rehabilitation programme and a personalized, home-based, self-managed rehabilitation programme was well tolerated and had a positive medium-term functional impact.

Keywords Rehabilitation programme; Dermatomyositis; Polymyositis; Functional assessment; Quality of life.

Disclosure of interest The authors declare that they have no competing interest.

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CO021
2005–2015: Ten years clinical experience in treating DMD patients by corticosteroids in Lyon
Stéphanie Fontaine Carbonnel 1,*, Pascal Rippert, Isabelle Poirot, Dominique Gachet, Capucine de Lattre, Carole Vuillerot Hospices civils de Lyon, escale-rééducation pédia trique, Bron Cedex, France
* Corresponding author.
E-mail address: stephanie.fontaine01@chu-lyon.fr (S. Fontaine Carbonnel)

Objective Since 2005, in France, corticosteroid therapy is now widely used in Duchenne muscular dystrophy (DMD). This treatment has changed our practice of pediatric rehabilitation teams. We describe here our 10-year clinical experience in treating DMD patients by CS according to international guidelines i.e. prednisone 0.75 mg/kg/day started from the plateau of motor function.

Materials/patients and methods We conducted a retrospective observational cohort study of 130 neuromuscular adult patients having a positioning wheelchair consultation in Foundation of Garches. The assessment is done with the seated postural control measure for adults.

Results Most of the patients had severe intensity illness, only10% were walking and 29% were with tracheostomal ventilation. The reasons of consultation were: positioning, choice or change of wheelchair, pain and prevention (rarely). The own wheelchair’s patient was powered wheelchair with seat adapted to the person type 2 in only 91 cases. 109 patients (84%) experienced pain in their wheelchair. In Duchenne muscular dystrophy patients, 44 (88%) were painful in there wheelchair. The topography of pain is frequently ischiatic. 19 patients (14%) had a pressure ulcer. All the patients examined have deformities. In Duchenne muscular dystrophy, pelvic obliquity and trunk tilt are frequently observed; in FSHD pelvic anterior tilt is frequent, and in steinert myotonic posterior tilt is frequent.

Discussion/Conclusion In order to improve the quality of life of this population, a study about the posture in wheelchair is therefore essential, most precociously.

Keywords Neuromuscular disorder; Wheelchair; Seating postural control; Pain; Pressure ulcer

Disclosure of interest The authors have not supplied their declaration of competing interest.

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CO022
Seated postural in wheelchair in NMD
Nadine Pellegrini Centre hospitalier du vexin, SSR neurologie, Magny-en-Vexin, France
E-mail address: nadine.pellegrini@wanadoo.fr

Objective To study seated postural control in neuromuscular disorder.

Materials/patients and methods We conducted a retrospective observational cohort study of 130 neuromuscular adult patients having a positioning wheelchair consultation in Foundation of Garches. The assessment is done with the seated postural control measure for adults.

Results Most of the patients had severe intensity illness, only10% were walking and 29% were with tracheostomal ventilation. The reasons of consultation were: positioning, choice or change of wheelchair, pain and prevention (rarely). The own wheelchair’s patient was powered wheelchair with seat adapted to the person type 2 in only 91 cases. 109 patients (84%) experienced pain in their wheelchair. In Duchenne muscular dystrophy patients, 44 (88%) were painful in there wheelchair. The topography of pain is frequently ischiatic. 19 patients (14%) had a pressure ulcer. All the patients examined have deformities. In Duchenne muscular dystrophy, pelvic obliquity and trunk tilt are frequently observed; in FSHD pelvic anterior tilt is frequent, and in steinert myotonic posterior tilt is frequent.

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Keywords Neuromuscular disorder; Wheelchair; Seating postural control; Pain; Pressure ulcer

Disclosure of interest The author declares that she has no competing interest.

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CO023
Muscle activations during gait in children with Duchenne muscular dystrophy
Juliette Kopars 1,*, Mathieu Lempereur 2, Sylvain Brochard 1, Carole Vuillerot 1, Vincent Tiffreau 4, Jean-Marie Cuisset 2, Yann Péron 6, Fabien Leboeuf 7, Raphaël Gross 8, Ludovic Delporte 9, Yannick Delpiere 10
1 CHRU de Brest, pédiatrie, Brest, France
2 Laboratoire de traitement de l’information médicale LatTIM Inserm UMR 1101, Brest, France
3 Garches. The assessment is done with the seated postural control measure for adults.

Results Most of the patients had severe intensity illness, only10% were walking and 29% were with tracheostomal ventilation. The reasons of consultation were: positioning, choice or change of wheelchair, pain and prevention (rarely). The own wheelchair’s patient was powered wheelchair with seat adapted to the person type 2 in only 91 cases. 109 patients (84%) experienced pain in their wheelchair. In Duchenne muscular dystrophy patients, 44 (88%) were painful in there wheelchair. The topography of pain is frequently ischiatic. 19 patients (14%) had a pressure ulcer. All the patients examined have deformities. In Duchenne muscular dystrophy, pelvic obliquity and trunk tilt are frequently observed; in FSHD pelvic anterior tilt is frequent, and in steinert myotonic posterior tilt is frequent.

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