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Stéphanie Fontaine Carbonnel
by corticosteroids in Lyon
experience in treating DMD patients 2005–2015: Ten years clinical

Results

pain, fatigue and biomarkers of tolerance and disease activity.

function measure, and the Kendall manual muscle test (MMT), gait,

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

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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 prospective study. Information was given on the expected effects and side effects. A diet was prescribed. At baseline, a cardiac ultrasound, an orthopedic examination, a bone density test, a physical and biological examination to ensure the absence of against indication, check the anti-varicella immunization. The motor function measure were performed each 6 months with a medical examination and blood sample to verify the tolerance and adapt the treatment function to weight and side effects.

Results During this 10-year period, CS was proposed to 51 patients and 48 patients were finally treated up to 10.5 years. The age onset of CS varies from 6 years to 20 years between 2005 and 2010, from 5 to 8 years after 2010. Prematurity stops were observed for 14 patients: 9 in the first 2 years, 5 after 2 years for side effects or inefficacy. No acute adrenal insufficiency was observed. During the first 2 years of treatment, MFM D1 score (standing position and transfers) remained stable. Then a slow decrease was observed after 2years to be null at M72. MFM D3 score (distal motor function) is more preserved than D2 (axial and proximal motor function) and remained stable during the follow up for the great majority of patients. Eleven patients started corticosteroids after losing the ability to walk (mean age 9.1 ± 1.6 years), 7/11 had spinal fusion. Thirty-seven ambulant patients started, in this group, the mean age of losing ambulation was 11.3 ± 2.2 years and no patient had spinal surgery.

Discussion/Conclusion Increased appetite, irritability, cushingoid facies are common but well tolerated by children in view of efficacy. Dose adjustments were discussed during case if accelerated weight gain, sluggish growth in stature and behavioral disorders.

Keywords Duchenne muscular dystrophy: Corticosteroid; MFM; Prednisone

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CO022

Seated postural in wheelchair in NMD

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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 trunc 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

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CO023

Muscle activations during gait in children with Duchenne muscular dystrophy

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