

increases in length (up to ~600 mm): this differs markedly to most mononucleated cells, e.g. fibroblasts, that remain at a relatively small size (~10–20 µm diameter). In addition, myofibres are long-lived and probably persist throughout the life of an individual (in the absence of severe injury that results in myonecrosis). The consequences of a dynamic expanding sarcolemma during growth, compared with an adult myofibre of a fixed length, have many scientific and clinical implications.

<http://dx.doi:10.1016/j.nmd.2012.06.289>

G.P.118

Targeting sarcopenia: Molecular basis of denervation of aged muscles and benefits of life-long exercise

T. Shavlakadze¹, R.J. Chai¹, C. McMahon², M.D. Grounds¹

¹University of Western Australia, School of Anatomy, Physiology and Human Biology, Crawley, Australia; ²Agresearch Ltd., Developmental Biology Group, Hamilton, New Zealand

With ageing, the progressive loss of muscle mass and function (sarcopenia) results in frailty, loss of independence and is a major cause of increased falls and fractures. The precise reasons for sarcopenia are unclear. We have previously described the time course of sarcopenia in female C57Bl/6J mice with significant loss of muscle mass at 24 and 28 months (m) of age (Shavlakadze et al., 2010: Biogerontology). Muscle function and maintenance of muscle mass require electrical stimulation by a nerve, yet denervation as a primary cause for sarcopenia has barely been considered. We have described altered morphology and striking denervation of neuromuscular junctions (NMJs) for muscles in geriatric (28 m) compared to young (3 m) female C57Bl/6J mice, using immunostaining of whole intact fast extensor digitorum longus (EDL) muscle (Chai et al., 2011: PlosONE). Myofibre type changes indicative of muscle denervation and re-innervation were observed in all muscles examined. However, there was no loss of alpha-motoneuron cell bodies in the lumbar spinal cord of geriatric (28 m) mice. These data suggest that changes in the muscles per se are a primary cause of the age-related denervation. Establishing these baseline data for geriatric mice is essential in order to take full advantage of the wealth of genetically modified mice available to study sarcopenia, and for testing therapeutic interventions. Our molecular analyses of muscles from mice aged 3, 15, 24 and 28 months aim to identify the molecular basis for this age-related denervation. We have already demonstrated that life-long exercise significantly reduces sarcopenia (measured by loss of muscle mass) at 28 months of age (unpublished) and we are investigating the molecular basis for the benefits of this exercise on ageing muscles with a focus on NMJ innervation.

<http://dx.doi:10.1016/j.nmd.2012.06.290>

ASSESSING FUNCTIONAL OUTCOME 2 – POSTER PRESENTATIONS

S.P.16

Gen classification scale (Ek) for Duchenne Muscular Dystrophy and Spinal Muscular Atrophy: Validity and reliability study of the Turkish version

A. Karaduman¹, G.I. Yatar¹, I. Alemdaroglu¹, O. Yilmaz¹, H. Topaloglu²

¹Hacettepe University, Faculty of Health Sciences, Physiotherapy and Rehabilitation, Ankara, Turkey; ²Hacettepe University, Faculty of Medicine, Pediatric Neurology Unit, Ankara, Turkey

EK scale was developed to assess detailed functional ability of non-ambulatory Duchenne Muscular Dystrophy (DMD) patients who have

severe physical impairments by Steffensen et al. in 2002. EK total score was found to distinguish the different functional levels and show the prognosis of the nonambulatory DMD and Spinal Muscular Atrophy (SMA) patients. The aim of this study was to make the cultural adaptation and investigate the validity and intra/inter-tester reliability of Turkish Version of EK scale on non-ambulatory DMD and SMA patients. The original English version of EK scale was translated into Turkish and necessary arrangements were made after the consensus by the researchers. After the arrangements and cultural adaptation; the pre-test study was made on five patients, inter-tester and intra-tester reliability studies were made on totally 34 patients. Brooke Functional Classification Scale for Upper and Lower Extremities (BFC), Barthel's Index for Activities of Daily Living and Pulmonary Dysfunction Test (PDT) were used as gold standards to investigate the validity of the scale in Turkish DMD and SMA population. 11 SMA and 23 DMD patients (Mean age: 13.71 ± 4.25) with non-ambulatory functional status were included in the study. Intra-tester reliability of the EK scale was 0.979 (ICC) with % 95 Confidence Interval [0.930; 0.994] and inter-tester was 0.916 (ICC) with % 95 Confidence Interval [0.407; 0.990]. There were correlations between Turkish version of EK scale and Barthel Index ($r = -0.716$, $it\ p < 0.01$) and PDT ($it\ r = 0.541$, $it\ p < 0.05$) and BFC for Upper Extremities ($it\ r = 0.576$, $it\ p < 0.05$). The Turkish version of the EK scale has excellent inter and intra-tester reliability and validity on Turkish DMD and SMA patients. EK scale is an adequate and useful instrument to assess functional status and predict the prognosis of non-ambulatory DMD and SMA patients in Turkish population.

<http://dx.doi:10.1016/j.nmd.2012.06.291>

S.P.17

Standards of care: Ensuring physiotherapists are competent to treat babies and children with neuromuscular disorders

M. Main¹, A. Mayhew², L. Pallant³, R. Rabb⁴

¹Great Ormond Street Hospital for Children, Dubowitz Neuromuscular Service, London, United Kingdom; ²University of Newcastle, Neuromuscular Service, Newcastle, United Kingdom; ³Leeds Teaching Hospitals NHS Trust, Physiotherapy, Leeds, United Kingdom; ⁴Birmingham Children's Hospital, Physiotherapy Department, Birmingham, United Kingdom

There are 16 centres in the UK offering specialist neuromuscular services; some very large, with five full time neuromuscular physiotherapists and additional research physiotherapists. Others are small having one or two clinics per month. There are no specific qualifications in the UK that actually designate a paediatric physiotherapist, although there is the qualification of a masters degree in paediatric physiotherapy. This is an academic qualification and does not confer experience or expertise in any specific area. Many newly qualified therapists however are going straight to a Masters degree in the hope of securing future employment. So which physiotherapists are qualified to assess and treat children with neuromuscular disorders? Who teaches them and who teaches the teachers? The Association of Paediatric Physiotherapists (APCP) a clinical interest group and the official voice of paediatric physiotherapy in the UK has initiated a programme of published competencies for physiotherapists; the standards for physiotherapists working in neonatology having been completed. The standards for physiotherapists working in neuromuscular disorders are now being agreed by the relevant stakeholder physiotherapists in the field, based where possible, on published evidence. Understanding the underlying pathology, knowing the presenting features and progression of individual conditions; and managing the many aspects of treatment and rehabilitation are covered. The topics include assessment, respiratory treatment, mobility, management of contractures and deformity and appropriate exercise and activity. Once finalised, these topics will form