Upper limb evaluation in non-ambulatory patients with neuromuscular disorders


The aim of this study is to establish a reliable tool of reproducible assessment of muscle strength in children affected by Duchenne Muscular Dystrophy (DMD) which will be selected for mesoangioblasts transplantation. We have developed a potential treatment for DMD based on infusion of cells (mesoangioblasts) from a healthy donor capable. The results of the current functional study will hopefully establish reliable qualitative and quantitative tool to assess results of a future cell therapy clinical trial with mesoangioblasts. This is a single centre, prospective, non-randomised, study of validation of outcome measures on 30 ambulant patients aged 5–12 years old affected by DMD including a cohort of 15 healthy aged matched males. We perform 2 days evaluation each 3 month for 1 year. During each assessment the following outcome measures are applied to DMD subjects: North Star Scale and 6 min walking test during the first day; quantitative assessment using the Kin Com 125 machine during the second day. The controls subjects will perform quantitative assessment twice a year. Twice during this evaluation year patients perform spirometry, cardiac assessment and lower limb MRI. We divided the patients into three subgroups of age (5–7 years, 8–9 years, 10–12 years). The results of this preliminary part of the study show specific correlation between functional and quantitative tests in stronger children. Kin Com measurements correlate appropriately with functional tests for 10- to 12-years-old DMD boys, while show a major variability in muscle strength for 8- to 9-years-old DMD boys. The comparison with healthy subjects showed a difference of muscle strength that increases with age. This preliminary study demonstrates that our assessment may represent a useful tool to monitor the progress of DMD in ambulant children to determine the pre-transplantation story of the children who will be later treated with mesoangioblasts.

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Swallowing disorders in pediatric neuromuscular diseases: A pilot study

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Aim: The purpose of this study is to determine the swallowing disorders and the causes in neuromuscular diseases, to develop the appropriate management strategies in probable deficiencies. Methods: Average age of 7.21 ± 5.46, 14 patients which diagnosed by Medicine Faculty of Hacettepe University – Neurology Department included to this study. Patients have continuing or previously swallowing problems. Six patients (14.2%) were myotonic dystrophy, four patients (28.5%) were SMA, two patients (14.2%) fasciobulbar paralysis, two patients (14.2%) were DMD diagnosed. Subjects swallowing functions were evaluated by clinical swallowing test and videofluoroscopy. Peripheric oral functions oral and pharyngeal phases of swallowing were analysed in clinic test. In videofluoroscopy; lip closure, tongue-palate contact, tongue elevation, tongue base-pharyngeal wall contact, delay in swallowing reflex related to oral phase were evaluated. Airway closure, hyolaryngeal elevation, upper esophageal sphincter opening, penetration and aspiration related to pharyngeal phase were evaluated. Also residue and nasopharyngeal reflux were evaluated in oropharyngeal region. Results: In SMA and DMD groups; mainly oral phase, in fasciobulbar paralysis; deficiencies relating to pharyngeal phase occurs. Aspiration was seen in eight cases and penetration was seen in two cases. Five of aspiration was silent aspiration. Only at four of subjects aspiration sign was determined. Four of subjects were feeding non-oral. Discussion: In patients with neuromuscular disease, swallowing function is affected more often unlike known. Difficulties during oral intake could be one of the source of respiratory problems in the period ahead.


P4.39 Assessment of tongue pressure during swallowing in patients with muscular dystrophies
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In patients with muscular dystrophy, oro-dental abnormalities as well as weakness of oropharyngeal muscles can disturb swallowing function. To assess oral phase of swallowing function, we measured tongue pressure during swallowing in patients with muscular dystrophies by a newly developed sensor sheet. Participants were nine patients with myotonic dystrophy (DM), 11 patients with Duchenne muscular dystrophy (DMD) and 51 healthy controls. A T-shaped thin sensor sheet with five measuring points (three points in median line and two points on the posterior-lateral parts) was attached on the hard palate. Tongue pressure was recorded during swallowing of 10 ml water. In healthy controls, tongue pressure was generated firstly by close contact with anterior-median part, then the circumferential part and finally with the posterior-median part. Compared to healthy controls, the maximum tongue pressure was lower in both DM and DMD. In DM patients, the pressure on median line was particularly low. Some DM patients hardly contacted their tongue to the hard palate. In DMD patients, most parts of tongue simultaneously contacted to their hard plate and sequential pattern was disappeared. We assume that these results reflect functional and anatomical abnormality of these disorders; high arched palate and atrophic tongue in DM and low palate and macroglossia in DMD.