

Introduction: Several reports have shown the benefits of using steroids for Duchenne Muscular Dystrophy (DMD) patients. All agree the need of wheelchair can be postponed by using those drugs. What it is still controversial it is if we can get other good outcomes. We decided to analyze the pulmonary functions of DMD patients taking steroids. **Method:** We selected 35 DMD patients from different regions of Brazil seen in an outpatient clinic and checked who was taking steroids. Standardized parameters regarding pulmonary functions were analyzed. We also checked muscle strength, cardiac function, creatinokinase levels and quality of life. A table was prepared to compare the subgroups of patients using the drugs with those not using them. **Results:** All patients are between 20 and 25 years of age. Six patients were under that medication and 29 were not. Patients taking steroids used prednisone (0.5 - 1.0 mg per Kg per day). The age of onset, regarding the use of the drug varied from 3 to 14 years. The duration of use ranged between 2 and 7 years. Fifty per cent of patients taking steroids needed ventilatory assistance, between 2 and 10 hours per day while 75% of the patients not using the medication were in that condition (between 10 and 20 hours per day). **Conclusion:** Our results strongly suggest the importance to prescribe steroids for DMD patients, even after they lost ambulation. Keeping those patients in a good shape will give them a chance to be enrolled in a near future in clinical trial, for example, with exon-skipping or stem cell therapy.

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P4.16 **Effects of different exercises on hemodynamic responses in Duchenne muscular dystrophy**

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Aim: The aim of this study was to investigate the effects of different exercises on heart rate and oxygen saturation of children with Duchenne Muscular Dystrophy (DMD). **Subjects and method:** 30 subjects (mean age: 7.87 ± 1.45 years) with DMD in early stages (stage1/2) of disease according to the Brooke Functional Classification included in this study. 12 of 30 children were required to climb up and down the standardized 5 stairs during 3 minutes. 23 of 30 children did cycling during 40 minutes after a 5 minutes warming up period with stretching exercises. The day after cycling, the physical therapy exercises including specific stretching and aerobic exercises for both upper and lower extremities according to the functional status of children were applied by a physical therapist to 10 of 23 children during 40 minutes. The hemodynamic responses including heart rate and oxygen saturation of children were recorded before and just after the exercise applications. **Results:** The heart rate was elevated after climbing stairs and physical therapy exercises ($p < 0.05$), although didn't show a significant difference after cycling. Mean heart rate was % 58 of maximal heart rate as submaximal only just after physiotherapy exercises. There were no significant differences in oxygen saturation after 2 exercise applications ($p > 0.05$). **Discussion:** Heart rate is mostly affected by exercises including both upper and lower extremities than exercises restricted to lower extremities in DMD patients in early stages of disease. Submaximal heart rate was only achieved after physiotherapy exercises.

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P4.17

Effects of exercises on muscle strength and fatigue level in Duchenne muscular dystrophy

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Aim: The aim of this study was to investigate muscle strength responses and fatigue levels of children with Duchenne Muscular Dystrophy (DMD) after two different exercises. **Subjects and Method:** 23 children with DMD in early stages of disease (stage 1/2) according to the Brooke Functional Classification included in this study. All of them did cycling on a stable bicycle actively during 40 minutes after a 5 minutes passive warming up period with stretching exercises. 10 of 23 children was also applied physical therapy exercises by a physiotherapist including both of upper and lower extremities during 40 minutes the day after cycling. Lower extremity muscle strengths of children were measured with a hand-held dynamometer and fatigue levels were assessed with an effort rating scale called 'Pictorial Variant of the Children's Effort Rating Table' by self-report before and just after the exercises. **Results:** The fatigue levels were increased after two exercise applications ($p < 0.05$). The strengths of quadriceps and hamstrings and total lower extremity muscles decreased after cycling ($p < 0.05$), while didn't show a significant difference after physical therapy exercises. **Discussion:** The perceived exertion by children were similar after two exercises. It is thought that the decrease in muscle strength after cycling is due to the exhaustive effect of cycling specifically on quadriceps and hamstring muscles in supported sitting position. The muscle strength was maintained just after physical therapy exercises. The exercise programme which was planned and applied by a physiotherapist showed no risk of specific muscle fatigue.

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P4.18

Asphyxia in a Duchenne muscular dystrophy patient due to tracheal compression by brachiocephalic artery

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Background: Trachea and brachiocephalic artery are anatomically crossing. This anatomical location sometimes causes asphyxia and other respiratory problems due to tracheal compression by brachiocephalic artery (TCBA), as known as tracheomalacia, in patients with neuromuscular diseases. Scoliosis, opisthotonus and laryngo-tracheal separation are the risk factors of TCBA. Typical symptoms of TCBA are recurrent asphyxia and/or stridor. We present a Duchenne muscular dystrophy boy with TCBA and summarize the key points of the diagnosis and management of TCBA on our experiences with other patients with neuromuscular diseases with TCBA. **Case presentation:** A wheelchair-bound 13-year-old Duchenne muscular dystrophy boy was admitted to our hospital because of cyanosis, dyspnea, and difficulty in expectoration of sputum. His oxygen saturation was 85% and pCO₂ was 122 mmHg under 10 L/min oxygen mask. He was immediately intubated. Chest X-ray revealed no cause of his dyspnea. He was stable under mechanical ventilation but sometimes suddenly deteriorated and DOPE evaluation (tube displacement, tube obstruction, pneumothorax, equipment failure) detected no problems. He was extubated but re-intubated several hours later because of dyspnea. He received tracheotomy but dyspnea and asphyxia attack with no DOPE problem repeated. Further evaluation by bronchofiberscopy and contrast computed tomography

revealed tracheal compression by brachiocephalic artery. He became stable after his trachea was stented by T-shaped silicon tube. 5 months later, he was re-admitted to our hospital because of massive bleeding from trachea due to tracheal-brachiocephalic artery fistula. Emergency operation was performed and his life was saved. **Conclusion:** We propose that neuromuscular patients, with or without tracheotomy, should be evaluated for TCBA when following respiratory problems or symptoms occur: stridor, asphyxia, artificial ventilation failure, difficulty in sputum suction

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P4.19

Multiple aortic aneurysms in a patient with Becker muscular dystrophy

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Introduction: Cardio-vascular complications in patients with muscular dystrophies include congestive heart failure, dilated cardiomyopathy, arrhythmias, and heart blocks. Aortic aneurysms have been rarely reported. We now report a patient with Becker muscular dystrophy, who presented with multiple aortic aneurysm with fatal outcome with pathological findings. **Objectives** To report aortic aneurysm as a possible cause of death in patients of muscular dystrophy with autopsy findings and etiological considerations. **Methods:** A case report with genetic and autopsy pathological examinations. **Results:** A 60 year-old male patient developed muscle weakness and atrophy at the age of 16. His younger brother presented with similar symptoms and diagnosed as Becker Muscular dystrophy by genetic examination. The patient's muscle weakness and atrophy progressed, and heart failure ensued, which was controlled with ACE-I. He complained severe back pain at the age of 58. Enhanced CT study revealed multiple aortic aneurysms. Aortic aneurysms were observed with control of blood pressure. Patient's status excluded any surgical intervention including aortic reconstruction. After 2 years a patient died of rupture of abdominal aneurysm. Autopsy revealed multiple aortic aneurysms including thoracic and abdominal regions. **Conclusions:** Rupture of aortic aneurysm has been rarely reported during the course of muscular dystrophies. Radiology examinations in the presented case suggested paraspinal tumors as differential diagnosis. Autopsy examination revealed the final diagnosis. Sudden death cases of muscular dystrophies may include mortality with aortic aneurysm. Etiological correlation between aortic aneurysms and Becker muscular dystrophy is uncertain.

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P4.20

Telomere length in exercised wild type and mdx mice

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The length of DNA telomeres is an important parameter of the proliferative potential of skeletal muscles. A telomere shortening has been reported in athletes suggesting that chronic endurance exercise may be seen as a stressor to skeletal muscle. In Duchenne muscular dystrophy (DMD) muscle, telomere shortening, resulting from contin-

uous muscle degeneration-regeneration cycles, is thought to contribute to premature senescence of satellite cells and the following ultimate failure of regenerative activity. In *mdx* muscle telomere shortening occurs in tibialis anterior (TA) muscle only in 600-day old animals, whereas in diaphragm muscle in 100- 600-day old mice.

We presently investigated whether telomere length is affected in *mdx* mouse muscles, in relation to morphological pattern and physical activity. To this aim, diaphragm and tibialis anterior muscles from 84-112-day old *mdx* mice were compared with samples from age-matched wild type mice. Exercised animals underwent a protocol of chronic treadmill running (8-10 weeks).

Our results confirmed an effect of exercise in healthy muscle showing a significant telomere shortening in exercised WT mice compared to sedentary animals in both muscles. Sedentary *mdx* mice showed a telomere shortening only in diaphragm muscle compared to WT. We confirmed that exercised *mdx* mice had a more compromised muscle architecture in both diaphragm and TA muscles compared to sedentary *mdx*, but this paralleled a lower telomere length only in diaphragm muscle. These results reinforce the assumption that telomere shortening can be involved in replicative senescence of satellite cells. Therefore the modulation of these events by the available compounds targeting telomere maintenance related proteins might represent a novel strategy for treatment of DMD and other muscular dystrophies.

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MUSCLE IMAGING; POSTER PRESENTATIONS

P4.21

MR imaging and spectroscopy of the brain in DMD

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About one third of boys with DMD shows cognitive impairment. This may be caused by the lack of dystrophin, but the mechanism remains to be clarified. Following successful restoration of in vivo dystrophin expression in human skeletal muscle, therapeutic trials with exon skipping are being planned. Whether treatments that improve muscle strength will be effective in brain is unknown. An imaging parameter that correlates with cognitive functioning would be very helpful.

We will present pilot data of magnetic resonance imaging (MRI) and spectroscopy studies of the brain in DMD boys that focus on structural, functional and metabolic parameters. High field strength quantitative MRI (3 Tesla) and MRS(7 Tesla) will be performed. The results will be correlated to healthy boys as well as to cognitive and behavioral functioning.

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P4.22

Quantitative analysis of muscle wasting in Duchenne muscular dystrophy by a new computed tomography method

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