Bilag

Oversigt over undersøgelser, hvor man har undersøgt energiomsætningen i hvile hos patienter med muskelsvind

1. J Hum Nutr Diet. 2009 Oct;22(5):383-93.

A review of nutrition in Duchenne muscular dystrophy.

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Duchenne muscular dystrophy (DMD) is a recessive X linked genetic disorder characterised by progressive muscle weakness and reduced muscle tone. Affecting only boys, it limits life expectancy to approximately 20 years. A literature review was conducted using MEDLINE and the Cochrane Library, employing the term 'Duchenne muscular dystrophy'. A total of 1491 articles in English were recovered. These papers were searched thematically under the headings: body composition (n = 10), energy expenditure (n = 10), nutrition (n = 6), corticosteroid therapy (n = 55) and gene therapy (n = 199). Key dietetic practice points were identified relevant to nutritional management. Papers supporting these key themes were assigned a level of evidence and grade of recommendation. There is limited high-quality evidence to guide the nutritional management of boys with DMD. Currently, the majority of evidence is based on expert opinion and clinical expertise. Delayed growth, short stature, muscle wasting and increased fat mass are characteristics of DMD and impact on nutritional status and energy requirements. The early introduction of steroids has altered the natural history of the disease, but can exacerbate weight gain in a population already susceptible to obesity. Prior to commencing steroids, anticipatory guidance for weight management should be provided. Malnutrition is a feature of end stage disease requiring a multidisciplinary approach, such as texture modification and supplemental feeding. Micronutrient requirements are yet to be determined but, as a result of corticosteroid treatment, vitamin D and calcium should be supplemented. Some evidence exists supporting supplementation with creatine monohydrate to improve muscle strength. More research is needed to provide a higher quality of evidence for dietitians working within this area.

PMID: 19743977 [PubMed - indexed for MEDLINE]

2. Can J Diet Pract Res. 2008 Winter;69(4):208-12.

Body composition and resting energy expenditure of individuals with Duchenne and Becker muscular dystrophy.

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PURPOSE: The relationship between body composition and resting energy expenditure (REE) was investigated in two boys and two men with Duchenne muscular dystrophy (DMD) (ages 11 to 22.4 years) and two boys with Becker muscular dystrophy (BMD) (ages 7.75 and 13.75 years). METHODS: The REE was assessed by indirect calorimetry; body composition indices (weight, height, skinfolds, and mid-arm circumference) were measured using standardized techniques and compared with healthy reference data. RESULTS: Those with DMD had reduced corrected mid-upper-arm muscle area (C-MUMA) in comparison with healthy peers, and approximately twice the subcutaneous fat levels of subjects with BMD and healthy peers. Boys with BMD had remarkably lower muscle status than did boys with DMD and healthy peers. In both groups, REE was lower than in healthy peers; REE was associated with body weight (r=0.85), height (r=0.92), mid-upper arm fat area (MUFA) (r=0.97), and C-MUMA (r=0.65). CONCLUSIONS: Individuals with muscular dystrophy (MD) exhibit considerable disease-specific alterations in body composition. The REE had a stronger relationship with growth (weight and height) and subcutaneous body fat composition and a weaker association with C-MUMA. Understanding the effect of MD on body composition and REE will allow dietitians to individualize energy recommendations.

PMID: 19063812 [PubMed - indexed for MEDLINE]

3. Am J Phys Med Rehabil. 2008 Dec;87(12):1053.

A re-examination and confirmation of daily energy needs for people with severe muscular dystrophy.

Munn MW.

Comment on:
Am J Phys Med Rehabil. 2005 Aug;84(8):639-43.

PMID: 19033764 [PubMed - indexed for MEDLINE]

4. Eur Respir J. 2005 Apr;25(4):682-7.

Resting energy expenditure in Duchenne patients using home mechanical ventilation.

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Nutritional status is both important and difficult to assess in patients with Duchenne muscular dystrophy (DMD), particularly in those requiring mechanical ventilation (MV). The current authors evaluated body composition

(bio-impedancemetry), resting energy expenditure (REE; indirect calorimetry) and energy intake in 20 adult patients with DMD using home MV (nocturnal: n = 13; continuous: n = 7) and 12 age-matched healthy controls. The patients were smaller in height than the controls and had a lower body weight. Most of the reduction in body mass index was accounted for by a reduction in fat free mass (FFM). REE (kJ) was significantly reduced in the patients (4559+/-853 kJ x 24 h(-1) versus 7407+/-1312 kJ x 24 h(-1)), but the difference disappeared after correction for FFM. REE and FFM were correlated in both the controls and patients, but less strongly in the latter, the lower strength of the association being due to the patients using continuous MV (REE and FFM uncorrelated). The food intake of the patients was 1.2+/-0.4 greater than their REE. This study shows that patients with advanced forms of Duchenne muscular dystrophy have balanced energy intakes and resting energy expenditure.

PMID: 15802343 [PubMed - indexed for MEDLINE]

5. Muscle Nerve. 2005 Jun;31(6):713-8.

Nutritional inadequacy in adults with muscular dystrophy.

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Patients with muscular dystrophy may be prone to nutrient deficiency due to mobility limitations or oropharyngeal weakness. Patients with myotonic muscular dystrophy (DM1) may be particularly prone to nutritional deficiencies from associated dysmotility of the entire gastrointestinal tract. We prospectively evaluated nutritional intake, body composition, and muscle strength in adult patients with DM1 (n = 29) and other muscular dystrophies (n = 22) on two occasions separated by approximately 6 months. Handgrip was significantly lower and knee extension higher for DM1 compared to other dystrophies, with no between-group differences in nutritional intakes. Many patients in both groups demonstrated inadequate nutrient intake of protein, energy, vitamins (water and fat soluble), and minerals (calcium and magnesium). Significant correlations were found between measures of strength and certain individual nutrients (e.g., copper and water-soluble vitamins). These data indicate that a substantial number of adults with muscular dystrophy do not meet current dietary intake recommendations. The potential clinical implications of these findings are discussed.

PMID: 15786416 [PubMed - indexed for MEDLINE]

6. Am J Clin Nutr. 2004 Aug;80(2):357-64.

Postabsorptive and insulin-stimulated energy and protein metabolism in patients with myotonic dystrophy type 1.

Perseghin G, Comola M, Scifo P, Benedini S, De Cobelli F, Lanzi R, Costantino F, Lattuada G, Battezzati A, Del Maschio A, Luzi L.

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BACKGROUND: Exaggerated insulin resistance was described as the major metabolic abnormality in myotonic dystrophy type 1 (DM1). We reported recently that the severity of the impairment in insulin-stimulated glucose metabolism in these patients was overestimated. OBJECTIVE: The aim was to dissect out insulin action with respect to whole-body energy homeostasis and glucose, protein, and lipid metabolism in patients with DM1 to assess the relevance of insulin resistance to the heterogeneous clinical manifestations of this syndrome. DESIGN: Ten nondiabetic patients with DM1 and 10 matched healthy control subjects were studied by means of 1) dual-energy X-ray absorptiometry; 2) a euglycemic-hyperinsulinemic clamp (40 mU. m(-2). min(-1)) combined with a primed, continuous infusion of [6,6-d(2)]glucose and [1-(13)C]leucine; 3) indirect calorimetry; and 4) localized (1)H magnetic resonance spectroscopy of the calf muscles. RESULTS: Patients with DM1 had less lean body mass, greater fat mass, and greater intramyocellular lipid contents than did healthy control subjects. Energy expenditure and glucose and lipid metabolism did not differ significantly between the groups. In contrast, markers of proteolysis were higher in DM1 patients in the postabsorptive and insulin-stimulated conditions and were associated with lower plasma concentrations of insulin-like growth factor 1 ($P < 0.03$) and higher plasma concentrations of tumor necrosis factor alpha receptor 2 ($P = 0.04$). CONCLUSIONS: Despite greater body fat and intramyocellular lipid contents in patients with DM1, insulin sensitivity was not significantly different between patients and control subjects. In contrast, the loss of lean body mass in patients with DM1 was associated with abnormal postabsorptive and insulin-stimulated regulation of protein breakdown. Lower plasma insulin-like growth factor 1 concentrations and higher tumor necrosis factor system activity might be involved in the muscle wasting of DM1.

PMID: 15277156 [PubMed - indexed for MEDLINE]

7. Neuromuscul Disord. 2004 Feb;14(2):142-6.

Increased resting energy expenditure in subjects with Emery-Dreifuss muscular dystrophy.

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Comment in:
Neuromuscul Disord. 2005 Jul;15(7):515-6; author reply 516.

We have studied changes in energy expenditure and body composition in adult males

with Emery-Dreifuss muscular dystrophy, age-matched males with hyperCKemia and age-matched healthy controls. All participants were studied twice, 2-3 years apart. Resting energy expenditure was studied by indirect calorimetry, lean body mass and body fat by dual X-ray absorptiometry, and muscle mass was estimated based on 24-h urinary creatinine excretion. At baseline and 2-3 years later, body fat was significantly higher ($P < 0.011$ and $P < 0.003$, respectively) and lean body mass significantly lower ($P < 0.024$ and $P < 0.012$, respectively) in patients with Emery-Dreifuss muscular dystrophy as compared to subjects with hyperCKemia and healthy controls. Resting energy expenditure, over the study period, increased significantly in patients with Emery-Dreifuss muscular dystrophy ($P < 0.031$), but not in patients with hyperCKemia nor in healthy controls. Our study suggests that patients with Emery-Dreifuss muscular dystrophy may have increased energy expenditure relative to healthy subjects. If not met by increased caloric intake, this greater energy expenditure may partially contribute to a further deterioration in their muscle performance.

PMID: 14733961 [PubMed - indexed for MEDLINE]

8. Eur J Clin Nutr. 2003 Feb;57(2):273-8.

Body composition and energy expenditure in Duchenne muscular dystrophy.

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OBJECTIVE: To investigate the relationship between resting energy expenditure (REE) and body composition in Duchenne Muscular Dystrophy (DMD). DESIGN: An observational study. SETTING: University Research Centre. SUBJECTS: Nine Duchenne children (age range 6-12 y), mean relative weight 128%, agreed to undergo the investigation and all of them completed the study; INTERVENTIONS: Assessment of body composition (total body fat and skeletal muscle mass) by magnetic resonance imaging and resting energy expenditure by indirect calorimetry. MAIN OUTCOME MEASURES: Fat mass (FM; kg and percentage weight), fat-free mass (FFM; kg and percentage weight), muscle mass (kg and percentage weight), resting energy expenditure (kJ/kg body weight and kJ/kg fat-free mass). RESULTS:: In Duchenne children fat mass averages 32% and total skeletal muscle mass 20% of body weight. Resting energy expenditure per kg of body weight falls within the normal range for children of the same age range, while when expressed per kg of FFM is significantly higher than reference values. No relationship was found between REE and total skeletal muscle mass. CONCLUSIONS: Our results do not demonstrate a low REE in DMD boys; on the contrary REE per kg of FFM is higher than normal, probably due to the altered FFM composition. We suggest that the development of obesity in DMD children is not primarily due to a low REE but to other causes such as a reduction in physical activity and or overfeeding.

PMID: 12571659 [PubMed - indexed for MEDLINE]

9. Metabolism. 2001 Oct;50(10):1181-5.

Paradoxical weight loss with extra energy expenditure at brown adipose tissue in adolescent patients with Duchenne muscular dystrophy.

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We examined the energy expenditure in patients with Duchenne muscular dystrophy(DMD) to evaluate the cause of the paradoxical weight loss observed in large numbers of adolescent patients before any obvious impairment of their swallowing function. In the morning, resting energy expenditure (REE)/m(2) was almost the same as that in normal controls despite a reduction in fat-free mass (FFM); thus, REE/m(2)/FFM was significantly increased in patients (median, 21.2 kcal/m(2)/FFM kg; range, 17.7 to 44.2, P =.012). A thermographic examination in the morning showed an obvious elevation of the body surface temperature on the back. This phenomenon was consistent with a paradoxical fall in the low frequency (LF)/high frequency (HF) ratio at night analyzed using the inter-RR spectrum by 24-hour electrocardiogram, which indicated relative activation of the sympathetic nervous system. The urinary secretion of norepinephrine at night was also significantly greater in patients (median, 0.119 microg/kg/h; range, 0.061 to 0.219, P =.011). These results suggest that paradoxical activation of the sympathetic nervous system may accelerate the production of heat in brown adipose tissue (BAT) and increase the level of energy consumption in patients, and that adolescent DMD patients may require greater caloric intake than expected to maintain body weight, which is important to improve the prognosis of their respiratory function. Copyright 2001 by W.B. Saunders Company

PMID: 11586490 [PubMed - indexed for MEDLINE]

10. Phys Med Rehabil Clin N Am. 1998 Feb;9(1):127-43.

Nutritional aspects of neuromuscular diseases.

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Evidence suggests that individuals with DMD have reduced skeletal development, including decreased linear growth and bone mineral density, compared to normal subjects. Despite their reduced muscle mass, a high percentage of DMD patients are overweight. Body composition measurements can assist with monitoring changes in fat mass and skeletal muscle mass as the disease progresses. Weight management in overweight DMD patients is indicated because excess adiposity burdens mobility and breathing, but only one study in two DMD patients has documented that weight reduction can be done safely. In the latter stages of the disease most DMD subjects become underweight because of an acceleration in skeletal muscle protein degradation relative to its synthesis. Studies of energy, protein and branched

chain amino acid supplementation in DMD have yielded promising but inconclusive results, and more well-designed studies are needed in this area. Although there is currently no cure for DMD, studies on the role of nutritional therapy in increasing the quality of life in these patients are urgently needed. Studies in adults with various SP-NMDs indicate a reduction in fat-free mass and an increase in fat mass relative to controls. The newly developed method of air displacement plethysmography for measuring body composition is ideally suited for SP-NMD subjects because it requires very little effort and the measurement procedure is relatively fast. Dual energy x-ray absorptiometry technology has been proposed for distinguishing myogenic from neurogenic SP-NMDs from calculation of the fat-to-lean soft tissue ratio, which is higher in patients with myogenic muscular atrophy. Studies on the energy metabolism of ambulatory SP-NMD subjects indicate that their basal metabolic rate is either similar to or slightly lower than controls, but 24-hour energy expenditure is about 25% lower than controls. This reduction in 24-hour energy expenditure is due to a reduction in physical activity in SP-NMD. Studies examining the roles of energy expenditure, physical activity, and diet in the development of adiposity and risk for secondary chronic diseases in SP-NMD subjects are currently underway.

PMID: 9894137 [PubMed - indexed for MEDLINE]

11. Reprod Nutr Dev. 1998 Mar-Apr;38(2):181-6.

Duchenne muscular dystrophy: a model for studying the contribution of muscle to energy and protein metabolism.

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Duchenne muscular dystrophy (DMD) is associated with a dramatic muscle mass loss. We hypothesized that DMD would be associated with significant changes in both energy and protein metabolism. We studied the resting energy expenditure (REE) in DMD and control children using indirect calorimetry, and their protein metabolism using an intravenous infusion of leucine and glutamine labeled with stable isotopes. In spite of a 75% muscle mass loss in the DMD children, the REE only decreased by 10%. DMD was associated with increased leucine oxidation but neither protein degradation nor protein synthesis were different from that of the controls. In contrast, whole body turnover of glutamine, an amino acid mainly synthesized in the muscle, was significantly decreased. These studies emphasized the quantitatively poor contribution of muscle to energy and protein metabolism in children. The qualitative impact of muscle mass loss on amino acid metabolism (glutamine) offers a fascinating field of research for the next few years and has therapeutic potential.

PMID: 9638792 [PubMed - indexed for MEDLINE]

12. J Nutr Sci Vitaminol (Tokyo). 1992 Apr;38(2):155-61.

Predictions of energy intake and energy allowance of patients with Duchenne muscular dystrophy and their validity.

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Patients with Duchenne muscular dystrophy are so malnourished that energy supplementation is crucial. Their degree of energy deficiency was assessed as difference between their energy intake and their energy allowance, which were deduced from easily measured parameters. A significant, negative relationship was found between the basal metabolic rate (BMR) (Y, %, BMR/standard BMR) and body weight (X, %, body weight/standard body weight) in the patients, from which the formula for the BMR was deduced to be Y = -1.116X + 174.5 (n = 202, r = -0.72, p less than 0.001). Thus, it is possible to estimate the energy allowance for individual patients by a factorial procedure from the presumed BMR and a factor for physical activity. In addition, their energy intake was calculated from a constant protein-energy % (14.6%) in their diet and nitrogen intake which was deduced from a significant positive correlation between their nitrogen intake (Y, mg/kg/day) and their nitrogen excretion in 24 h urine samples (X, mg/kg/day). This correlation conformed to the equation Y = 1.053X + 32.4 (n = 267, r = +0.76, p less than 0.001). The validities of the above predictions for energy intake and allowance were examined by plotting the degree of energy deficiency (% ratio of presumed intake/presumed allowance) against the concentrations of retinal binding protein, prealbumin and transferrin in the serum, because rapid turnover proteins are sensitive to energy deficiency. Significant positive correlations were obtained with both variables, suggesting that these predictions were valid.

PMID: 1506921 [PubMed - indexed for MEDLINE]

13. J Nutr Sci Vitaminol (Tokyo). 1992 Apr;38(2):141-54.

Protein and energy metabolism in patients with progressive muscular dystrophy.

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Studies were made on whether body weight loss in patients with muscular dystrophy is due to reduced intake and/or abnormal expenditure of energy. For this, food intakes and various physiological variables were surveyed in totals of 310 patients with Duchenne muscular dystrophy (DMD) of 11 to 29 years old and 28 patients with limb-girdle muscular dystrophy (LGMD) of 30 to 47 years old. Energy and protein intakes, expressed on a unit body weight basis, in DMD patients were comparable to, or higher than the allowances for age-matched healthy controls, whereas those in LGMD patients were 92 and 94% respectively of these allowances. The basal metabolic rate (BMR), expressed as kcal/kg/day, of DMD patients of all ages was higher than that of controls, the difference increasing with age, and being about 20 to 30% higher than that of controls in older patients with DMD.

The BMR of LGMD patients was nearly normal. The maintenance requirements of conventional dietary protein in DMD and LGMD patients were 1.26 and 0.84 g/kg/day, respectively. These values were about 68 and 12% higher than the normal adult value (0.75 g/kg/day), indicating decreased protein utilization and increased protein catabolism. Daily excretion of urinary 3-methylhistidine (3MH) per unit muscle mass (micrograms/mg creatinine) by MD patients was significantly higher than that by controls, indicating increased degradation of muscle protein. The BMR, maintenance protein requirement and 3MH excretion of DMD patients suggest that DMD is a hypercatabolic disease. Comparison of the energy and protein intakes with the allowances estimated in consideration of increased requirements showed deficiencies of energy and protein in DMD patients. Thus, we conclude that the underweight of the DMD patients resulted from nutrient deficiencies due to hypercatabolism, despite their considerably high intakes of energy and protein, expressed as per kg body weight. These deficiencies were confirmed by demonstrating decreased concentrations of free essential amino acids, particularly branched chain amino acids, in their serum. The values of variables of LGMD patients were intermediate between those of DMD patients and control subjects.

PMID: 1506920 [PubMed - indexed for MEDLINE]