Nutrition-Related Problems of Pediatric Patients with Neuromuscular Disorders

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ABSTRACT. Children with neuromuscular disorders have special health needs that place them at risk of nutritional problems. These needs may have detrimental effects on their development, immune and respiratory system. Identified nutrition-related problems are malnutrition, obesity, food intolerance, food allergies, drug-nutrient interactions, constipation and reflux. In order to improve the nutritional status of these patients, a nutritional assessment is recommended which should include anthropometric measurements and laboratory tests. *Key words: Nutrition, Neuromuscular disorders.*

Nutritional deprivation is especially critical for children who are at-risk or disabled. Ten to 15% of all children in the United States and Puerto Rico have special health needs and it is estimated that at least 40% of them are at risk os nutritional related problems (1). The lack of appropriate nutrition results in a reduced immune response, attention span, circulation and motivation for play and rehabilitation activities in children with special health needs (2).

Patients with progressive neuromuscular disorders have difficulty meeting their nutritional requirements. Many nutritional concerns are common to all diseases causing progressive weakness, but others are unique to a particular disorder (3).

Nutrition-related problems often are associated with specific developmental disabilities. Most children with disabilities experience growth retardation, feeding and eating problems, food allergies, weight and drug-nutrient interactions. About 37% of these children require special diets while 32% need special feeding procedures (4). Children with mental retardation, cerebral palsy, and Down's syndrome often have growth retardation (5). Feeding and eating problems may be present in these children as well as in those with cleft lip and palate, visual impairments, and phenylketonuria (6). Food intolerance may also be associated in children with hyperactivity, learning disabilities, and mental retardation. Obesity often affects children with mental retardation, spina bifida, Prader-Willi syndrome, and heart disease. Drug-nutrient interaction problems are reported in children with mental retardation, cerebral palsy, autism, schizophrenia, and hyperactivity (7).

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Malnutrition

Starvation. Several studies have evaluated the effects of starvation and malnutrition on the neuromuscular system of animals and humans. Muscle energy utilization varies almost a hundred-fold from rest to maximum metabolic activity, whereas nerve cells continually have a high metabolic rate (8). As a result, early neuromuscular changes following malnutrition affect peripheral nerves more than skeletal muscle. The effects of malnutrition on peripheral nerves may be related to specific deficiencies of vitamins such as B_1, B_6, or B_{12}, as well as protein-calorie malnutrition (8). Because skeletal muscle is the largest and most important protein reservoir of the body and is the major site for various steps in the metabolism of amino acids, severe malnutrition eventually impairs muscle function (9).

To maintain proper nutrition, a person needs to have properly functioning mastication and swallowing mechanisms, esophageal motility, gastric emptying, intestinal motility and absorption and hormonal regulation during meals (i.e. insulin release). Several of these

functions may be impaired in neuromuscular diseases. The major cause of malnutrition is dysphagia, although defects in the other digestive steps may also be taking place (8).

Several factors predispose to fatigue of respiratory muscles. Because inspiratory muscles require energy to contract (expiration is passive), energy depletion of inspiratory muscles can result in respiratory failure in patients with neuromuscular disorders. Malnutrition and catabolic states, by depleting energy stores, further weaken diseased inspiratory muscles and play an important role in the pathophysiology of respiratory failure (10).

Simultaneous problems in the immune response as result of starvation may include lower cell-mediated responses, phagocytic activity, complement system, secretory antibodies, and antibody affinity. Such secondary immunodeficiency states may lead to a higher incidence of infections, which only further prevents recovery in the malnourished patient. Nevertheless, detailed effects of malnutrition on immune function in neuro-muscular disorders has not been studied. Since patients with neuromuscular disorders often develop intercurrent infections, usually pulmonary, immune dysfunction secondary to malnutrition may be contributing to their decline. Improvement in nutritional status, whether attributable to increasing strength or immunocompetence, can significantly and favorably alter the clinical course in selected patients (11).

Undernutrition. Thi is frequent problem in children with severe cerebral palsy (spastic quadriplegia) who often have significant impairment of their eating and swallowing mechanisms. During the past ten years, greater attention has been paid to the causes and management of undernutrition in chidren with neurodevelopmental disabilities (12).

The effect of undernutrition on physiologic, brain growth and development is well recognized (13). Under-nourished infants and children do not achieve normal height and weight for their ages. Also, their neurologic development is significantly impaired. A child who habitually chokes and is prone to aspirate food or liquids, reacts to feedings in a way that clearly signals discomfort (14).

Most children with neurodevelopmental disability and eating problems resulting in undernutrition are unable to meet their energy needs even with high-energy supplements (15). Underweight problems may be present in children with athetoid cerebral palsy, oral motor dysfunction, Rett syndrome, frequent infections, and poor interaction with their parents (16).

If there is no concern about food aspiration, the first step in the management of undernutrition is to increase the energy concentration of ingested food. Providing the child with supplemental beverages with an energy density of 4,200 to 6,300 kJ/L is helpful. In children who can eat purée solids, the energy density of these types of food can be increased by adding balanced fat (omega 6 and 3). An increase in the viscosity of liquids has resulted in a higher intake in some patients (17).

Obesity

Obesity is defined as an excessive ratio of fat to lean body mass and can be evaluated on the basis of weight-for-height and skin-fold thick-ness. Obesity can be a major problem for patients with neuromuscular disease, since excessive weight further impairs the ability to maintain independent ambulation and compromises respiration (3).

Since exercise in neuromuscular diseases is not easily accomplished and may be sometimes detrimental, an equally useful method of increasing the ratio of strength to body weight is by decreasing body weight (18). In such patients, relatively small gains in weight (3 to 5 Lb) can have a major impact on physical performance, such as climbing stairs or walking. An even more serious consequence of obesity in patients with neuromuscular diseases is respiratory failure (18). The added force of a distended abdomen predisposes to fatigue by increasing the work required for diaphragmatic motion. Loss of weight under these circumstances may significantly prolong the length and the quality of the patient's life (18).

Other Nutrition-related Problems

Constipation is a nutrition-related problem often found in children with spina bifida, hypertonia, and spastic cerebral palsy (19).

Pica (allotriophagia) is an eating/nutritional disorder frequently present in some individuals with mental retardation. These patients have an alteration in the taste sense with compulsive eating of edible and inedible objects. Pica often results in malabsorption of zinc and iron (16).

Pseudobulbar palsy, which occurs mainly in severe palsy, affects eating and swallowing. As a result, food intake is decreased and eating time increased. Thus, oral feeding may be of little success despite the efforts of caregivers.

Gastroesophageal reflux and aspiration of food often compound the problem celebral palsy (20). Many of this children have clinically important gastro-esophageal reflux regardless of whether they have been fed through a nasogastric tube. The cause of reflux in such patients is unclear; it is probably related to intestinal dysmotility and delayed gastric emptying (21-23). If reflux is significant

and frequent, it may increase the risk of food aspiration and aspiration pneumonia.

Nutritional Assessment of Patients With Neuromuscular Disorders

Clinical assessment of a patient's nutritional status is obtained by history and physical examination, anthropometric measurements, and specific laboratory tests. The Americal history should stress the presence or absence of recent weight changes, chronic conditions that may interfere with good nutritional status, physical handicaps that may result in inadequate food intake, and other predisposing conditions contributing to poor nutrition, such as alcoholism, chemotherapy, or drug use. Analysis of a dietary history through a daily caloric count may reveal evidence of vitamin, mineral, protein, or calorie deficiencies (3).

Anthropometric measurements provide quantitative data indicative of the nutritional status. The usual way to assess the nutritional status of a child is to measure weight and length (or height) and compare the results with growth charts. However, a child with quadriplegia has muskulosketal deformities that may make measurements of total length impossible (24). Height and weight should be measured and compared with ideal body weight. A decrease of greater than 10% of usual weight over a 6 month period should be considered a significant weight loss. Measurement of triceps skin fold is an indirect estimate of body fat or caloric stores and correlates well with other measures of body fat when done correctly. Measurement of mid upper arm circumference is an indicator of both caloric and proteic stores (25).

Krick and collaborators support the view that the energy needs of children with cerebral palsy cannot be determined from normal nutrition recommendations but should be calculated from the estimated basal metabolic rate. Unfortunately, the nomograms used to determine this rate are not always reliable in children with spastic quadriplegia (26).

The study by Fried and Pencharz in 1991 demonstrated that feeding disabled children a formula such as Pediasure® (Ross Products Division, Abbott Laboratories, Columbus OH, USA), which has a higher ratio of nutrients to energy, assures recommended nutrient intakes (15).

Laboratory tests commonly used to assess nutritional status include total lymphocyte count, serum albumin, serum transferrin, total iron-binding capacity, and serum vitamin levels, especially vitamins A and C (6). Bone mineralization in these children should be evaluated to determine whether present dietary levels of calcium and phosphorus intake are too low (14).

Intervention Strategies

There are many nutritional intervention possibilities for disabled children with special health care needs. The first type or intervention strategy emphasizes nutritionist and parenteral involvement in the care of these children (27).

A second approach that can be utilized emphasizes educational services for this child population. These intervention strategies include providing texture modifications, special eating devices, and feeding assistance for children with neuromuscular disorders. Educational strategies for children with special health care needs also include the development of a written nutritional plan for service providers as well as the inclusion of nutritional goals in their individual educational plans (4).

A third approach used as inter-vention strategy is the education of direct care providers of children with neuromuscular disorders. A recent training needs survey of direct care providers indicates great interest in liability issues associated with serving children with chronic health conditions; determining the nutrient value of menus; reducing fat, sodium, and cholesterol in diets; feeding techniques, procedures used in developing special diets, and understanding the physical and emotional needs of children with disabilities or special health care needs (27).

Resumen

Los niños con desordenes neuromusculares tienen necesidades especiales relacionadas a su condición que los coloca a riesgo de problemas nutricionales. Estas necesidades tienen efectos detrimentales en su desarrollo, así como en su sistema inmunológico y respiratorio. Los problemas nutricionales identificados son: malnutrición, obesidad, intolerancia y/o alergias a alimentos, interacciones de nutrientes y medicamentos, constipación, y reflujo gastroesofágico. Para poder mejorar el estado nutricional de estos pacientes se recomienda una evaluación nutricional que debe incluir medidas antropométricas y examenes de laboratorio.

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