Duchenne Muscular Dystrophy (DMD) is a genetic, progressive neuromuscular disorder affecting approximately one in 3,500 male infants across all populations. It is a recessive, single gene defect on the X chromosome of the mother. There are also cases of spontaneous mutation with no carrier identification identified in the mother. In 1987, the protein associated with this gene was identified and labeled dystrophin. In voluntary muscles, dystrophin is located under the cell membrane and attaches to a small group of other muscle proteins that, in turn, attach to the extracellular matrix outside the muscle fiber. Without dystrophin, muscle contractions can cause cracks in the cell membrane. As a result of the cracks in the cell membrane, muscle-digesting enzymes are activated to destroy the damaged cells. The destroyed muscle fibers are then replaced with fat and connective tissue.

With identification of the gene and concurrent research and therapy, boys with DMD are surviving longer, into their late 20’s and older. Medical management of DMD spans multiple sub-specialties and clinics including: Rehabilitation Medicine, Neurology, Orthopedics, Pulmonology and Cardiology. Nutrition care and management of DMD is an integral component of care. The following are some of the multiple issues associated with nutrition management in DMD:

- Accuracy and consistency of anthropometrics including weight and length.
- Determination of Ideal Body Weight
- Nutritional implication of corticosteroid therapy
- Osteoporosis and low-impact fractures
- Escalation of nutrition support with progression of muscle weakness
- Obesity with decreased activity
- Nutrition and emotional issues

Obtaining accurate weight and length measurements

A child who is able to stand and walk can be weighed and measured by the same means with the same tools as any pediatric patient. Infant scales and recumbent length boards can be used for infants. Regular scales and wall-mounted stadiometers are recommended for older children. However, when a boy with DMD is unable to stand, accurate weights and lengths are a challenge. Ideally, a wheelchair scale should be used. The child (in the wheelchair) is weighed, then a Hoyer lift is used to lift the child. The wheelchair is weighed alone, and its weight subtracted from the first weight. Growth assessment for older children can be challenging. Armspan has been used as an estimate of height. However, many patients with DMD have scoliosis, spinal fusion and reduced tubal growth, making this measurement inaccurate. One method is to use segmental lengths. There are also recumbent length measuring tools, which can be limited in accuracy secondary to contractures. What is important, however, is that no matter how cumbersome, weights and lengths need to be measured consistently by the same method at each clinic visit.

Estimation of Ideal Body Weight

Determination of ideal body weight using standard calculations becomes less relevant in children with DMD. Boys with DMD may lose up to 4% of their muscle mass per year. Specific guidelines for the actual calculation of ideal body weight, hydration needs and energy requirements are not currently published. One method of determining ideal body weight is to:

1) Determine the “height-age” (age at which the actual height is at the 50th percentile). 2) Determine IBW for the height-age. This is the 50th percentile weight that matches the height-age. 3) Calculate the percent IBW by dividing the actual weight by the IBW. Depending on the degree of progression of disease and deterioration of muscle mass, up to 20% less of the normal IBW can be factored into the equation.

Nutritional Implications in Corticosteroid Therapy

Of all the therapeutic drugs studied in the management of DMD, only prednisone seems to have the potential for providing interim, palliative, functional improvement for boys with DMD. Several studies describe prolonged function and ambulation in DMD with the use of corticosteroids, although there is no consensus recommending steroids as a standard therapy. However, the nutritional implications of steroid therapy include increased appetite, weight gain, linear growth, decreased absorption of dietary calcium and fluid retention. The side effects may necessitate some form of caloric restriction, weight monitoring, supplements of calcium and vitamin D and possibly some sodium restriction.

Osteoporosis

Studies conducted to determine the prevalence, circumstances and outcome of fractures in boys with DMD indicates that up to 20% have experienced fractures. Many of these (41%) have occurred in patients who were still walking. Bone densitometry studies in the DMD population show bone density in the proximal femur was profound-ly diminished even when gait was not affected and then progressed to four standard deviations below age-matched controls. Sixty-six percent of the fractures occurred in the lower extremities. Studies in osteoporosis in DMD (independent of steroid use) indicate that the occurrence of a fracture had a significant impact and possible loss of ambulation.

Escalation of Nutrition Support

Many boys with DMD require nutrition support at some point. The primary challenges in nutrition support are hydration (often affecting constipation and urinary tract infections), adequate fiber, adequate energy intake and adequate protein intake for prevention of skin breakdown. As the disease progresses, many require the use of oral nutrition supplements or gastrostomy tubes. Two calorie per cc tube feedings, if tolerated, provide the greatest number of calories in the fewest hours. The choice of tube feeding products is dependent on gastric motility, choice of feeding style (continuous drip vs. bolus), and the need for fiber to prevent constipation, fluid needs and the need for increased protein.

Obesity

For boys who are still ambulating, significant energy expenditure can occur as walking, climbing, playing and other childhood activities become more challenging. However, when a boy with DMD becomes wheelchair dependent and is no longer walking, energy requirements can decrease substantially. Unfortunately, by that time, most families have become accustomed to increasing energy intake to meet the needs of the previously ambulating child. Often, eating continues to be one of life’s pleasures as the child’s world becomes smaller. Creative ways to reward without the use of food, continued on page 6

PNPG Post

Practice Point

NUTRITIONAL ISSUES FOR DUCHENNE MUSCULAR DYSTROPHY

Susan C. Casey, RD, CD
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PNPG Post

SUMMER 2003 Page 5

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Susan C. Casey, RD, CD
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Duchenne Muscular Dystrophy (DMD) is a genetic, progressive neuromuscular disorder affecting approximately one in 3,500 male infants across all populations. It is a recessive, single gene defect on the X chromosome of the mother. There are also cases of spontaneous mutation with no carrier identification identified in the mother. In 1987, the protein associated with this gene was identified and labeled dystrophin. In voluntary muscles, dystrophin is located under the cell membrane and attaches to a small group of other muscle proteins that, in turn, attach to the extracellular matrix outside the muscle fiber. Without dystrophin, muscle contractions can cause cracks in the cell membrane. As a result of the cracks in the cell membrane, muscle-degrading enzymes are activated to destroy the damaged cells. The destroyed muscle fibers are then replaced with fat and connective tissue. 1,2

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NUTRITIONAL IMPLICATIONS IN CORTICOSTEROID THERAPY

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provide calorie-free or low calorie snacks and adequate after-school supervision are all integral to any successful management of rapid weight gain in a child who uses a wheelchair.

**Nutrition and Emotions**

In some cases, adolescents with DMD who require assistance with meals at school often stop eating at school. Many are reluctant to ask or rely on friends to help with the mechanics of feeding or are uncomfortable with the length of time a meal can take. Some teenagers may just adjust their mealtimes to a different time of day, however others are at risk for dehydration or inadequate intake. Some will take a high calorie beverage to school (commercial or homemade) so the child only has to have lunch through a straw. Unfortunately, some teenagers, without some thoughtful and creative planning, decide to no longer attend school, thereby isolating themselves from critically needed socialization.

While there are many review and research articles related to DMD, there does not appear to be a standard set of guidelines, care map or clinical pathway specifically for nutrition across the greater than 200 centers taking care of boys with DMD in the United States. To improve care, quality of life and survival, a collaborative process among dietitians to produce a set of nutrition standards for DMD or the use and benefit of all clinics is overdue. Any dietitian interested in a collaborative effort to create and publish nutrition guidelines for Muscular Dystrophy, contact the author at: susan.casey@seattlechildrens.org

**References**


**Journal Review**

**Kristin Ann Hillary  
Loyola Dietetic Intern**

According to Holly Renee Broadwater RN, BSN, MSN, FNP, “the number of overweight children has doubled within the last three decades”. In her article “Reshaping the Future for Overweight Kids”, in the November, 2002 issue of RN, Broadwater gives background information, the reasons behind the obesity epidemic, and the steps to take to both prevent the occurrence and improve the future for children who are already overweight.

Weight problems are no stranger to children; in fact 1-in-5 children could be considered overweight. While it is not accepted to say that a child is obese, a BMI in the 95th percentile has been shown to have the same effects as an adult in the same BMI percentile. High blood pressure, type 2 diabetes, low self-esteem, anxiety and feelings of rejection are all consequences of being overweight.

Broadwater states that, like adults, “children gain excess weight when there’s an imbalance between energy input and energy output”, and proposes that reasons may include unhealthy eating habits, a sedentary lifestyle, low socioeconomic status, obese parents or underlying disease such as hypothyroidism or Cushing Syndrome.

The first step in helping an overweight child is to set up a counseling session. It is important to counsel both the child and the child’s parents, according to Broadwater. Hypertension, diabetes, high cholesterol and psychological issues are all-important topics to discuss to help prevent an overweight child from becoming an overweight adult. Remember that “change needs to come in small steps” and should be a combination of nutrition, exercise and behavioral changes.

Broadwater suggests that healthy eating includes establishing a daily eating pattern, including less fried foods, teaching children about satiety, providing healthy snacks, minimizing empty calorie foods and involving the child with grocery shopping and meal preparation. The second piece of the puzzle is exercise. Broadwater states that there are several ways that a family can encourage physical activity including using parents as role models, finding an activity that the child enjoys, discouraging prolonged periods of sedentary activity and establishing daily routines. The third and final piece includes support and love from the child’s parents.

“Reshaping the Future for Overweight Kids” by Holly Renee Broadwater emphasizes the need for children to eat a healthy diet and get the right amount of exercise; starting when a child is young is crucial. With diet and exercise it is possible to help today’s overweight children become healthy, active adults tomorrow.

June first marked the beginning of a new year for PNPG. As your chair, I’m very excited about following up the activities completed this past year under the leadership of Robyn Wong. Some of the activities included our two publications: Building Block edited by Peggy Oomens and the PNPG Post, edited by Annie Frederik. Joan Walarius was our communications coordinator and did a spectacular job. Under Robyn’s leadership the web site, www.pediatricnutrition.org is now in place and is being used at a record pace. We are most indebted to Beth Ogata in developing what will be such a useful website. Look for the membership directory on this site along with announcements of continuing education events.

Our treasurer, Carol Parkman Williams, has overseen funding the many activities of the practice group for the past two years. Carol has done such a great job, monitoring what we spend, creating the budget following our program of work, and making sure that we follow it. Carol’s term ended May 31st and we welcome aboard, Patricia Edwards-Hare, clinical nutrition manager, All Children’s Hospital, St. Petersburg, Florida.

Many thanks to our outgoing committee chairs; Marion Taylor Baer, Grants and Awards; Susan Carlson, Scholarship; Terri Schindler, Member Support; Maria Puangco, Nominating; Kathleen Huntington, Legislative. Each of these committees is very important to this practice group.

Let me now introduce the new members of the executive board. Josephine Cialone is the chair-elect and will be planning the PNPG program for the annual meeting in 2004. Amy Brander, our secretary, continues for another year, Patricia Edwards-Hare is our treasurer, Aida Miles, is the professional support chair, and Debbie Hutslar is member support chair. Debbie will be supervising and coordinating the activities of the subspecialty units. Our three advisors, Barbara Emison Gaffield, Denise Sitka, and Faye Wong provide the executive board with much needed advice and consultation.

Last, but not least, is Robyn Wong who remains on the board as the past chair to provide guidance and look for funding. Other members of the leadership group are listed in the PNPG roster on our website at www.pediatricnutrition.org.

PNPG members are truly leaders in many of the activities of ADA. Beth Leonberg, our outgoing past chair, was elected director at large on the ADA Board of Directors. Susan Laraname, a past PNPG chair, is our new president-elect of ADA.

Mark your calendars for October and the FNCE meeting in San Antonio, PNPG will have a program on Monday afternoon on “Follow-up of the Very Low Birthweight Infant”. Our speakers are Ian Griffin, MD, Baylor Medical School; Nancy Nevlin-Folino, Dayton, Ohio; and Marsha Dunn Klein, OTR, Phoenix, Arizona. This year our breakfast and business meeting will be planned for Thursday and the reception for members Monday night.

In the next issue of the Post, I’ll describe some of the other activities of PNPG. We have many opportunities for your involvement in our practice group.

Harrriet Cloud, MS, RD, FADA

2002 Member Survey Results

The member directory was preferred as an electronic copy by 63% of respondents. The PNPG website has been visited by 42% of respondents less than once a month, 15% have visited more than once a month and 43% have never visited the website.

From additional comments, members were interested in pediatric seminars or CPEU opportunities and listed a variety of topics, PNPG involvement with childhood obesity support, leadership and programs was a top priority for many members.

The results were discussed in detail at the Spring board meeting and are being incorporated into PNPG’s action plan whenever possible. We appreciate your participation and input.