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Best Practices in Nutrition for Children With Myelomeningocele

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Abstract: *Children born with myelomeningocele (MMC), the most severe form of spina bifida, face multiple challenges throughout their life span. Neurogenic bowel, neurogenic bladder, Arnold-Chiari II malformation, and hydrocephalus are common complications that have implications for elimination, feeding, and learning. Nutrition education and intervention that use the concepts of family-centered care beginning in infancy and continuing throughout childhood, adolescence, and adulthood are important. These can help maximize wellness, fitness, and independence in children with MMC, as constipation, dysphagia, and obesity are common comorbidities seen in individuals with MMC. This article reviews the literature on nutrition assessment and intervention in this unique population to assist nutrition professionals in caring for children with MMC.*

Keywords: spina bifida; myelomeningocele; developmental disabilities; folic acid

Background

Myelomeningocele (MMC), also known as spina bifida, is one of the most common birth defects in the United States, affecting 1 in 2500 babies.[1] Advances in medicine have significantly increased the survival rate to 90% reaching adulthood.[2]

Myelomeningocele is a neural tube defect caused by incomplete closure of the spinal cord during early pregnancy. The neural tube typically closes by day 28 of pregnancy, usually before a woman even knows she is pregnant. When the neural tube fails to close, it leaves a sac or lesion along the midline of the spinal cord. The level and severity of the lesion determines the level of sensation and motor function in the individual. There are 4 types of spina bifida, as shown in Table 1. Table 2 shows some of the complex medical issues commonly found in MMC.[3]

Folic Acid Supplementation

In 1991, the Centers for Disease Control (CDC) recommended all high-risk women (ie, a diagnosis of spina bifida or a previous pregnancy/child with a diagnosis of spina bifida, anencephaly, or other neural tube defect) take 4 mg of folic acid daily (available through prescription) when they start trying to conceive and during the first trimester of pregnancy.[4] The US Public Health Service issued its recommendation in 1992 for all women of childbearing age to consume 400 mcg (micrograms) of folic acid daily to reduce the risk of MMC.[5] A few years later, the Food and Drug Administration required folic acid fortification of enriched grain products to begin January 1, 1998.[6] In addition to folic acid, genetics, maternal obesity, and environmental factors also play a role in the development of MMC. A recent study found a significant increase in a woman's risk of having a child with a neural tube defect with a serum B_{12} level ≤250 mcg/L at 15 weeks of gestation.[7]

Wellness

In 2001, the World Health Organization released the International Classification of Function, Disability and Health (ICF).[8] This is the first universal method to classify body structures, body functions, activities and participation, and environmental factors for individuals with disabilities. There has been a shift in focus to emphasize a person's function and health instead of his or her disability. Early emphasis on wellness and fitness at an early age fits into the ICF model and has been found to promote self-reliance, self-care, and lean body mass in children with MMC.[9-11] Interdisciplinary teams can use the ICF to address the multiple needs of a child with MMC during childhood and adolescence[8] as well as transition planning for adult care.[12]

Family-centered care is crucial when working with any child who has special health care needs, as family strengths and resources are important contributors to the success of the child's nutrition care plan.[13] Assessing a child's nutritional status and providing nutrition education on a regular basis in the context of family-centered care support the family in establishing good eating habits and managing some of the complications

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Table 1.

Types of Spina Bifida[2]

Type	Definition/Issues
Spina bifida occulta	A small gap or defect in some of the vertebrae that is not visible. There are usually no problems with the spinal cord or nerves. Most people do not know they have this type of spina bifida until they have an X-ray or magnetic resonance imaging for other medical reasons.
Meningocele	The meninges are exposed through the spine in a sac or cyst containing cerebrospinal fluid. Although there is typically no nerve damage, affected individuals may have some medical issues.
Occult spinal dysraphism	An infant presents with a dimple, red marks, or tufts of hair on the lower back. There may be medical issues, such as urologic, neurologic, or orthopedic issues, related to the spinal cord that require further evaluation.
Myelomeningocele	The most severe and complex type of spina bifida in which the meninges and spinal nerves are exposed through the spine at the level of the defect. This results in disabilities and nerve damage. Surgery to repair and close the defect is typically done within 24 to 72 hours after birth.

Table 2.

Common Medical Issues in Spina Bifida[3]

Endocrine	Growth hormone deficiency Precocious puberty
Neurologic	Hydrocephalus Arnold-Chiari II malformation Seizures Paralysis
Orthopedic	Scoliosis Lumbar lordosis Contractures of the hips/knees
Gastrointestinal	Dysphagia Neurogenic bowel
Other	Neurogenic bladder Hypertension Learning differences Insensate skin Pressure sores Latex allergy

related to MMC, such as constipation, osteopenia, and obesity.

The Ellyn Satter "division of responsibility" for feeding children is a useful tool to share with parents of children with MMC when establishing healthy eating habits. In this method, the child is responsible for how much and whether he or she eats, and the parent is responsible for the what, when, and where of feeding.[14] This approach can help families as they try to get their undernourished child diagnosed with failure to thrive to eat more or their "picky eater" to try more vegetables by clearly defining the feeding relationship. Parents of a child with special needs can feel particularly burdened with the multiple challenges related to feeding their child.

Anthropometrics

Consistently obtaining an accurate length/height measurement in children with MMC may be difficult secondary to scoliosis, contractures, and body structure differences. Measurements of length/height are typically obtained using length boards, wall-mounted stadiometers, or segmental length. The 2000 CDC growth charts[15] are used to plot and assess a child's growth trends. Linear growth usually slows down around 2 years of age, but weight gain may continue trending or increase at a faster rate, which reiterates the importance of accurate and consistent measurements beginning in infancy. MMC growth charts[16,17] may be used in conjunction with the CDC growth charts to evaluate growth velocity in a child with MMC. Recent charts include growth data according to lesion level and ambulatory status.[17] Arm span measurements are not typically used in children with MMC secondary to growth issues seen in older children with a higher level lesion, with the differences most notable in the lower extremities.[18] However, some studies have shown that the use of arm span measurements is accurate during growth hormone treatment.[19,20] Children with MMC have higher rates of precocious puberty and an earlier growth spurt[21]; however, their final adult height is shorter in relation to their peers,[10,21-24] even after

successful growth hormone treatment to increase linear growth.[20,25]

Overweight and obesity have long been recognized as comorbidity risks related to multiple factors, including ambulatory status, activity level, and hypotonicity in children with MMC[21,22,26] that continue through adulthood.[27] This can result in additional medical conditions associated with obesity, such as metabolic syndrome, hypertension, and diabetes. Nelson and colleagues[28] reported that one third of MMC patients met the criteria for metabolic syndrome. Contributing to these medical issues are the higher percentage of body fat[10,22,26,29-32] and lower amount of lean body mass[11,26,29,32,33] seen in children with MMC. Activity level plays a major role in body composition. Studies have shown that ambulatory children with MMC who have a higher level of function in daily living and physical activities have a higher level of fitness[27,31,34-36] that results in higher lean body mass[11] with reduced amounts of body fat.[24] However, the percentage of body fat between children who are ambulatory and those who are nonambulatory is similar.[24,34] In addition, children who have a higher MMC lesion typically have impaired linear growth and develop obesity.[18,30]

Body mass index (BMI) is widely accepted as a screening tool for obesity in children older than 2 years of age.[37] The difficulty in obtaining an accurate height measurement in children with MMC, along with their differences in body habitus, can make assessment of BMI inaccurate. Children with MMC have disproportionate body fat distribution in the trunk and upper limbs.[9,11] Skinfold measurements are useful and easy to use tools in a clinical setting that can be used longitudinally to assess trends in growth and body composition if used regularly by an experienced clinician. Humeral length, mid-arm circumference (MAC), and triceps skin fold (TSF) can be measured on a regular basis during clinic visits. Then MAC and TSF can be compared with reference percentiles.[38] Because MAC and TSF standards are based on data collected on children without disabilities, tracking serial measurements on individual children with MMC can serve as a method for assessing their growth trends over time in addition to growth charts.

Energy Needs

The aforementioned differences in body composition of children with MMC are reflected in their measured resting energy expenditure. Each child's energy needs should be determined on an individual basis, as typical predictive equations overestimate energy requirements in 1/5 of children with special needs.[26,39] A lower lean body mass and lower resting energy expenditure[10,11,29] that results in lower total energy expenditure[26,29,35] can make it a challenge for children with MMC to achieve and maintain a healthy weight. Children with MMC have been shown to maintain their weight with an energy intake meeting 80% of the Recommended Dietary Allowance (RDA).[23] In addition, children with MMC have been shown to expend 25% less energy than children of the same age not affected by MMC.[26] The RDA for energy is often used to calculate energy needs for infants, with adjustments made based on growth velocity. Grogan and Ekvall[11] published formulas in 1999 to calculate energy needs after infancy in children with MMC. Their recommendations are 50% of the RDA/age for weight maintenance after infancy, 9 to 11 kcal/cm for weight maintenance, and 7 kcal/cm for weight loss after age 6. Patt et al[40] describe an equation that uses a height measurement and injury level to determine energy needs in patients with spinal cord injury. This equation has been used in clinical practice to determine energy needs in children with MMC:

$$\text{Boys } (11.7 \times \text{height in cm}) + (30.7 \times \text{level}) - 931.23$$

$$\text{Girls } (12.4 \times \text{height in cm}) + (21.8 \times \text{level}) - 1036.42$$

where level is an injury number corresponding to the level of the affected vertebrae. The lesion level in children with MMC can be used as the injury number corresponding to the affected vertebrae.

Assessment of energy needs has to consider growth velocity, ambulatory status, MMC lesion level, and activity level. For further assessment of a child's resting energy expenditure, indirect calorimetry is considered the "gold standard," which can be helpful when the goal is to prevent over/underfeeding. Once resting energy expenditure has been calculated from indirect calorimetry measurements, a steady state should be verified to ensure accuracy of the measurement.[41] Then the addition of activity factors results in a value for total energy expenditure. The recently published Dietary Reference Intakes (DRI) has physical activity coefficients[42] that have been used as activity factors for indirect calorimetry in clinical practice.

Bone Health

Children with MMC have an increased risk of osteopenia and osteomalacia, as shown in Table 3. It is well established that ambulation significantly increases bone mineral density (BMD) in children with MMC[24,43,44]; however, this effect appears to dissipate in adulthood.[45] Children with a neurogenic bladder that has been augmented have shown lower BMD in previous studies.[46,47] Metabolic acidosis has not been determined as the cause of low BMD in children with MMC.[46,48] An earlier report by Quan et al[49] found increased urinary calcium excretion in nonambulatory children with lower BMD, and Boylu et al[47] theorized that the underlying neurologic processes related to a neurogenic bladder rather than bladder augmentation surgery resulted in a lower BMD.

There is a higher risk of osteomalacia and osteopenia/osteoporosis in children and adults with MMC related to lower BMD,[44,45,49] even though 1 study showed most children with MMC met the RDA for calcium.[44] Emerging research continues to expand the knowledge of the multiple factors involved in bone health in addition to dietary calcium intake and weight-bearing status and supports recommendations for a well-balanced diet with a variety of foods.

A nutrition assessment should include a child's typical dietary intake of calcium to ensure the DRI for age is met.[50] Clinical practice has suggested that calcium

Table 3.

Risk Factors for Osteopenia and Osteomalacia in Myelomeningocele

Ambulatory status
Anticonvulsant medications
Obesity
Lack of sensation
Limited sun exposure
Hypotonia
Malabsorption
Diet low in calcium and vitamin D

Table 4.

Risk Factors for Dysphagia in Arnold-Chiari II Malformation[53,54]

Problems sucking and swallowing
Difficulty positioning for feedings
Respiratory symptoms, stridor
Difficulty forming seal on nipple
Refusal of cup/sippie cup
Loss of food from mouth
Nasal regurgitation
Long feeding times
High number of formula changes to improve formula tolerance
Perceived lack of satiety
Refusal to advance textures/ increase variety
Delays in self-feeding
Choking with feedings
Poor salivary control
Aspiration
Weight loss/growth failure

supplementation is often necessary because of the individual food preferences of children with MMC.

In 2008, the American Academy of Pediatrics recommended increasing vitamin D intake of all infants, children, and adolescents to 400 IU daily.[51] Vitamin D status is assessed by checking levels of serum 25(OH)-D, with a deficiency diagnosed at a level <20 ng/mL. There is no established consensus or change in the DRI to recommend repletion doses in children, although maintenance doses in children have been suggested in the range of 400 to 1000 IU/d.[52] Once vitamin D supplementation has been started, levels of serum 25(OH)-D should be rechecked and assessed every 3 months to ensure levels return to normal.[51]

Common Challenges

Feeding Problems

Dysphagia

Arnold-Chiari II malformation is a hindbrain herniation of the brainstem and cerebellum present in most children (98%) with MMC. This malformation affects coordination of muscles involved in the suck, swallow, and breathe patterns in infants and can result in neurogenic dysphagia. Identifying dysphagia and providing intervention when dysphagia symptoms manifest (Table 4) is crucial in children with MMC. Mathisen and Shepherd[53] found that hypotonicity along with the Arnold-Chiari II malformation put infants with MMC at a higher risk for feeding issues. Feeding issues/delays resulted in low iron status, low protein levels, and energy intake that met 72% of recommended needs and ultimately impaired growth in this study. Severe cases of Arnold-Chiari II malformation can be treated with decompression surgery that may improve dysphagia symptoms; however, recurrence of dysphagia symptoms is common.[54,55]

Dysphagia can be evaluated and treated with a comprehensive, interdisciplinary feeding team. Difficulties in the oral phase, initiation of swallow, and/or pharyngeal phase of feeding characterize dysphagia. Feeding team members may include a physician, registered dietitian, speech-language pathologist, psychologist, occupational therapist, social worker, and nurse. A clinical feeding evaluation and a videofluoroscopic swallow study (VFSS) can provide valuable information to the feeding team. Observation of mealtime allows the team to evaluate many variables contributing to feeding, including the parent-child feeding relationship, the child's oral-motor skills, and typical positioning for feedings. Once this evaluation is completed, a VFSS or modified barium swallow study can be conducted to identify aspiration as well as abnormalities in oral-motor skills, swallow initiation, pharyngeal dysphagia, and slow esophageal motility. Silent aspiration can occur without obvious external symptoms. An interdisciplinary feeding team is valuable to work with families to implement team recommendations.[56,57] The National Dysphagia Diet[58] provides universal definitions and standards for thickened liquids and diet types for texture modifications.

Gastroesophageal Reflux Disease

A child with MMC who has difficulty with feeding and meeting his or her growth needs should be assessed for gastroesophageal reflux. Common symptoms reported by parents in clinical experience include difficult feedings, food refusal, emesis, stridor, grimacing, irritability, posturing, reactive airway disease, and difficulty gaining weight/weight loss. Although there is not a diagnostic test considered to be a "gold standard" by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN), there are several diagnostic tests commonly used in the diagnosis of gastroesophageal reflux disease (GERD). Although an upper gastrointestinal (GI) series is frequently used to diagnose GERD, it may not always be evident during the short testing period. This test is useful to identify structural abnormalities, such as

obstruction, malrotation, stenosis, esophageal strictures, or hiatal hernia in children with special needs and/or spinal abnormalities. Upper GI endoscopy can show reflux esophagitis, although additional studies may need to be performed to rule out other cause of esophagitis. A 24-hour pH probe can measure pH in the stomach over a 24-hour time period and can provide important diagnostic information when combined with caregiver or family report of GERD symptoms.[59]

Treatment of GERD in infants typically focuses on nonpharmacologic methods, such as thickened feedings, positioning changes, hydrolysate formula, and increasing caloric density of the diet.[59] The latest pediatric GERD Clinical Practice Guidelines published in 2009 suggested insufficient evidence for elimination of GERD-provoking foods in children and adolescents with GERD and recommended lifestyle changes, such as weight loss and positioning changes, along with drug therapy that may assist in GERD management. In severe cases of GERD, a fundoplication may be necessary.[59]

Bowel Management

Neurogenic bowel, dysmotility, and medications are common factors in children with MMC that predispose them to constipation that can result in multiple complications unless proactive steps are taken for bowel management. Constipation can affect gastric emptying, bladder health, and kidney health. The hypomotility noted in persons with MMC[60] increases colonic transit time and often necessitates a comprehensive program to address bowel continence. Bowel and bladder continence should be a goal for every child with MMC to reduce social stigma and the risk of skin breakdown.[61-63]

A nutritional assessment to evaluate the child's diet, with particular attention to fluid and fiber intake, is often the first step in bowel management.[61] Then recommendations can be made to increase fluid and fiber intake as needed. Fluid goals should primarily be based on body weight and adjusted as needed. If children with MMC have inadequate fluid intake, it may be helpful to send a water bottle and physician's orders to school to ensure the child is able to drink during the school day. Some families find it helpful to offer their child fluids after routine bladder catheterization.

Fiber goals are typically calculated using the "age + 5 = grams of fiber" rule of thumb.[64] The DRI for fiber intake in children is currently 19 to 38 grams daily[42] and is not realistic for most children with MMC because of a noted lack of high-fiber foods in their food preferences commonly seen in clinical experience. If dietary changes are not sufficient to increase fiber intake, then a fiber supplement may be needed in the regimen. Fiber intake should not be significantly increased unless fluid intake is sufficient, as this may worsen constipation. Medical management of constipation is often necessary to ensure regular evacuations and to preserve kidney, bladder, and bowel health. Leibold et al[61] have presented an excellent interdisciplinary model for formulating and monitoring a bowel management program in children with MMC that considers multiple variables, including the child's diet, learning issues, and family routines. This model reiterates the importance of adequate fluid and fiber intake, along with daily bowel movements, as a first step in achieving continence.[61] Starting a bowel continence program at an early age helps children with MMC maximize continence success.[61,62]

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Wounds

Children with MMC are at a high risk for skin breakdown and wounds related to multiple risk factors, primarily insensate skin and obesity. Children with MMC and their families should be educated on how to assess skin for breakdown on a routine basis.[65] There is a paucity of literature on wound healing in children. Most literature on wound healing in adults cites that adequate energy intake should be ensured to allow protein sparing for wound healing. Although obese adult patients have demonstrated slower wound healing,[66] weight loss is not recommended when trying to heal a wound. The focus should be strictly on providing sufficient nutrition to promote wound healing.

Adequate protein should be provided to ensure positive nitrogen balance during wound healing.[67,68] Clinical practice in children with MMC has shown good results with increasing protein intake to 2 to 3 g/kg/d with careful monitoring of serum urea nitrogen and creatinine levels. Pompeo[69] has suggested using a PUSH (pressure ulcer scale for healing) score to determine the amount of protein to provide for adult wound patients, which may be adapted for pediatric use, as shown in Table 5. Jaksic[70] has published recommendations for protein intake for critically ill children (included in Table 5) that can be used for wound healing in children. Ideal body weight should be used if the child is overweight or underweight, whereas actual weight should be used if the child is 90% to 125% ideal body weight.

All children with MMC who have a wound should be on a multivitamin with iron; other individual nutrients, such as zinc, should be supplemented when deficiency is evident.[66] Zinc has an established role in wound healing and immunity as a cofactor for multiple enzymes. Albumin is a zinc carrier through the body; therefore, when albumin levels are low, zinc absorption is reduced and zinc deficiency is probable. Plasma zinc levels should be evaluated as

Table 5.

Protein Recommendations for Wounds

Based on PUSH (Pressure Ulcer Scale for Healing) Score[69]	
PUSH Score	**Protein Needs, g/kg/d**
0-15	1.4-1.6
16-30	1.6-2.0
>30	2.0-2.4

Critically Injured Pediatric Patients[70]	
Age of Child	**Protein Needs, g/kg/d**
0-2 y	2.0-3.0
2-13 y	1.5-2.0
Adolescents	1.5

From Jaksic T. Effective and efficient nutritional support for the injured child. *Surg Clin North Am.* 2002;82:279-391. Reprinted with permission from Elsevier.

Table 6.

Foods With a High Food Allergy Risk for Children Who Are Latex Allergic/ Sensitive

Bananas
Kiwi
Figs
Plums
Avocados
Passion fruit
Peaches
Tomatoes
Chestnuts
Papaya
Nectarines
Celery

Table 7.

Learning Differences in Children With Myelomeningocele[78]

Perceptual motor problems
Comprehension
Attention
Sequencing
Hyperactivity/ impulsivity
Memory
Organization

part of a nutritional assessment, and supplementation should be initiated using the DRI as a reference.[71] When zinc levels normalize, supplementation should be discontinued, as supplemental zinc can impair copper and iron absorption.[66,72] Iron also plays an important role in collagen synthesis[66] and is routinely supplemented during wound healing when iron deficiency anemia is evident.

Vitamin A is involved in skin integrity and immunity. When vitamin A deficiency is evident, supplementation should be given according to the DRI for age.[73] Vitamin C, a water-soluble vitamin, acts as a cofactor with iron during collagen synthesis via proline and lysine hydroxylation. Supplementation to correct the deficiency should be the DRI for age,[74] although 100 to 500 mg/d is typically provided for 7 to 10 days with a documented vitamin C deficiency and then reduced to the DRI for age.[66,68]

Latex Allergy

Children with MMC have an increased risk of latex allergy, which appears to be associated with early and repeated exposures to latex.[75] Children with latex allergy/sensitivity are at risk of the following food allergies: banana, avocado, and kiwi (see Table 6 for a more comprehensive list of high-risk foods). When solid foods are introduced to the diet of an infant with MMC, parents should be reminded to monitor for signs and symptoms of food allergies, particularly if the child is latex allergic/sensitive. Children with MMC who are latex allergic/sensitive should avoid latex in the home and community.[76] However, they do not need to avoid foods containing some of the same proteins as latex unless they demonstrate allergy symptoms with these foods.

Learning Differences in Children With MMC

Children with MMC and shunted hydrocephalus often demonstrate learning differences that should be considered when providing nutrition education, as shown in Table 7.[77] Vachha and Adams[78] found that children with MMC have difficulty understanding abstract concepts (such as direction, position) and using appropriate language in social situations. They recommended using visual aids, short commands and sentences, and multiple opportunities for repetition and practice to enhance understanding in children

with MMC. For example, providing a handout with a pictorial rendering of the number of cups of fluid that need to be consumed daily helps a child keep track of his or her fluid intake by allowing him or her to check off each cup of fluid he or she drinks. Dietitians should consider learning differences when providing nutrition education to children with MMC to help them understand and maximize their success with recommended changes.

Conclusion

Children with MMC present a unique set of challenges throughout their life span to the dietetics practitioner. Early and frequent contact with the dietitian using a family-centered approach can help children with MMC and their families maximize wellness, fitness, and independence. Collaboration and consultation with interdisciplinary team members to consider the multiple issues that present in children with MMC, including medical and learning challenges, is critical when providing medical nutrition therapy. Recent medical advances in the care of MMC have helped these individuals grow up to become independent, self-sufficient, and successful.

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