

Spina Bifida Guideline

Nutrition, metabolic syndrome, and obesity: Guidelines for the care of people with spina bifida

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Abstract. Nutritional challenges and a lack of activity can lead to health problems across the lifespan for people with spina bifida. Children and adults with spina bifida are also at greater risk of being classified as overweight or obese compared to their peers without the condition. Therefore, early recognition of nutrition problems, weight management counseling, and timely referrals for evaluation and management of diet and activity can help those with spina bifida and their families achieve a healthy lifestyle. This article details the development of the Nutrition, Metabolic Syndrome and Obesity Guidelines, which are part of the 2018 Spina Bifida Association’s Fourth Edition of the Guidelines for the Care of People with Spina Bifida. It discusses the identification and management of poor nutrition and prevention of obesity for children, adolescents, and adults with spina bifida and highlights areas requiring further research.

Keywords: Spina bifida, nutrition, obesity, guidelines, myelomeningocele, neural tube defects

1. Introduction

Good nutrition is important for everyone to maximize health and wellness [1]. Healthcare providers are therefore often tasked with providing essential nutritional education to assist individuals incorporate healthful changes in eating patterns and behavior. Those with spina bifida often experience unique challenges related

to healthy dietary intake [2], which can affect many areas of health and wellness across their lifespan. People with spina bifida also have elevated rates of obesity [3] which can affect health, independence and quality of life [4]. This manuscript details the development and recommendations of the Nutrition, Metabolic Syndrome, and Obesity guidelines for individuals of all ages with spina bifida.

During infancy, some babies with spina bifida may experience slow weight gain and inadequate nutrition. Initial prolonged hospitalizations can negatively impact breast-feeding and bonding between infant and parents [5]. A complex interplay of medical and social

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factors can affect the nutritional intake of children as they grow, including brain stem dysfunction, shunt obstruction, silent aspiration, sleep apnea, recurrent infections, and altered feeding dynamics related to frequent hospitalization and caregiver stress [2].

As they grow older, children with spina bifida who have a latex allergy may limit their intake of fresh fruits and vegetables due to cross-reacting foods (see the Latex Guideline at <https://www.spinabifidaassociation.org/guidelines/>). Those with the Chiari II malformation may have an aversion to textured foods, which limits their dietary intake to highly processed food with low nutritional value [6]. Furthermore, many often have a neurogenic bowel [7]. Ensuring good bowel control may be more important to people when considering their dietary intake than the overall nutritional value of food [8,90]. Other psychosocial factors affecting nutritional intake include socioeconomic status. It is well documented that people with disabilities and their families frequently have lower incomes [10], which can result in food insecurity, a lack of access to sources of healthy food and inaccessible cooking environments [11]. When families are faced with these circumstances, the risks of poor nutrition affect the whole family, not just the individual with spina bifida.

The consequences of poor eating habits can be serious, including vitamin deficiencies, anemia, malnutrition, poor growth and/or excessive weight gain [2]. In turn, these can lead to constipation, skin breakdown, osteoporosis, hypertension, diabetes, cardiovascular disease and other preventable secondary conditions [12], which can require healthcare services including hospitalizations [13]. Previous studies estimated that 47% of hospital admissions for people with spina bifida were for potentially preventable secondary conditions [14], many of which may have been addressed with good nutrition. Therefore, it is critical that healthcare providers work with children, families and adults with spina bifida to emphasize the importance of healthy nutrition and a balanced diet on overall health and wellness [15].

Weight-management is an important component of delivering high-quality health care for people with spina bifida [3]. In a recent survey, 40% of parents of adult children with spina bifida and 44% of adults with spina bifida rated weight-management as one of their top five priority issues [16].

Most children with spina bifida follow typical growth patterns until they are around four years of age; after that time, increased fat mass (versus lean mass) has been found when compared with children without spina bifida [17]. The prevalence of obesity in children with

spina bifida has been calculated to be approximately twice that of their typically developing peers, with numbers cited up to 50% in children, and 64% in young adults [3,18,19]. A complex interaction of physiological, environmental, psychosocial and physical factors predisposes people with spina bifida to having obesity [3,90]. Mobility impairments, inaccessible health promoting spaces, reduced incomes, lower resting energy expenditure and social isolation can all contribute to weight gain [20,21]. In addition, girls with spina bifida diagnosed with premature puberty may experience weight gain [22]. Weight gain is also commonly seen in women with spina bifida and Polycystic Ovary Syndrome [23].

Obesity in children is usually defined as a certain Body Mass Index (BMI) above a given percentile for age and gender [24]. BMI in adults is calculated as a person's weight in kilograms divided by the square of height in meters, with obesity defined as a BMI > 30 [25]. However, calculation of BMI is challenging for clinicians as linear growth is affected in individuals with spina bifida and typically leads to short stature, which impacts a person's BMI [19]. Therefore, when BMI is calculated and plotted on norms based on typical developing children or the general adult population, classifications of weight will be skewed for people with spina bifida [26,27]. In all cases, BMI is only a proxy for body fat and does not consider muscle mass or other factors that are associated with a healthy weight [13,25].

Although contemporary weight science shows that higher weights do not automatically infer poor health [28], they can be associated with cardiovascular disease, diabetes, and musculoskeletal disorders [4,29]. For those with spina bifida, increasing body weight can affect mobility, leading to a sedentary lifestyle that can worsen the imbalance between caloric intake and expenditure [13]. In addition, higher weights in spina bifida may lead to increased pain, skin breakdown, sleep apnea and decreased independence [14,30]. It is also important to note that people with obesity, including those with spina bifida, often experience weight bias and discrimination; this initiates a cycle whereby the experience of weight stigma increases cortisol and metabolic abnormalities, contributing to higher weight and further increasing stigmatization [31]. It is critical, then, that healthcare providers do not stigmatize people with spina bifida who have higher weights, even unintentionally [32,33]. Focusing solely on weight loss can have the opposite effect and may encourage disordered eating in people with spina bifida [34]. Therefore, the guidelines presented here describe how

healthcare providers can take a collaborative, strengths-based approach to promote the benefits of healthy nutrition more broadly (see also the Health Promotion Guidelines and Physical Activity Guidelines at <https://www.spinabifidaassociation.org/guidelines/>).

Also, this approach aims to reduce the clustering of risk factors that can lead to metabolic syndrome, which includes three or more of the following risk factors: hyperglycemia, increased central adiposity, elevated triglycerides, decreased high-density lipoprotein cholesterol, and elevated blood pressure, although the exact diagnostic cut-offs vary between children and adults [35]. While data are lacking on metabolic syndrome in adults with spina bifida, some research has addressed risk factors in younger people. For example, one study identified metabolic syndrome in 32.4% of adolescents aged 11–20 years with spina bifida, with only six percent of the participants showing zero metabolic syndrome risk factors [36]. As a comparison, levels of metabolic syndrome in a general childhood/adolescent population have been reported to be approximately nine percent [35]. In a young adult population (16–30 years), 42% showed at least two risk factors for cardiovascular disease including total cholesterol, high density lipoprotein, systolic blood pressure and smoking [37]. Although some of these metabolic syndrome risk factors cannot be impacted by nutrition (e.g. smoking), it is important for healthcare providers to explain the link between a healthy dietary intake and reduced cardiovascular disease risk factors.

2. Summary and gaps

Given the benefits of healthy nutrition, the aim of these guidelines is to present the best available evidence for providing nutritional information to people with spina bifida across their lifespan. These guidelines also provide suggestions for how healthcare providers can approach weight-related conversations, as well as strategies for obesity prevention and management. Where relevant, recommendations for assessment of risk factors for metabolic syndrome have been provided.

3. Guidelines goals and objectives

The goals of the Nutrition, Metabolic Syndrome and Obesity Guidelines were both practical and aspirational. The objectives were to:

Primary objective

- Maximize and support wellness through the lifespan

Secondary objective

- Reduce and prevent secondary health conditions related to poor nutrition and overweight/obesity, including metabolic syndrome.

Tertiary objective

- Support the development of client/caregiver knowledge, self-management skills, and self-efficacy related to nutrition and dietary habits.

4. Methods

The Nutrition, Metabolic Syndrome and Obesity working group was convened in 2016 to create a new section that had not been included in the previous 2006 guidelines [38]. Initially, the Nutrition and Obesity guideline was distinct from the Metabolic Syndrome guideline. However, the two working groups identified that there was substantial overlap and thus they were combined with the approval of the Spina Bifida Association Steering Committee Chairs. The methodology used to create the guideline has been reported in detail elsewhere [39]. In brief, the working group members were identified by the Spina Bifida Association's Professional Advisory Council. One content expert was nominated to lead the work (ACM) with other members providing support in their area of expertise. A comprehensive literature review was conducted on the guideline's topic, supplemented by literature identified by the working group to ensure the content was informed by the best available evidence. The group created the objectives through consensus and the outline of the guideline followed a template provided by the Spina Bifida Association. This specified the sections to be included: Introduction, Outcomes, Clinical Questions, Guidelines and Research Gaps. Because Nutrition, Metabolic Syndrome and Obesity were not included in the 2006 guidelines, all material was newly created. Clinical questions that informed the guidelines can be found in Table 1.

5. Results

Not all the clinical questions could be answered using peer-reviewed literature. Where literature was lacking, the working group came to a consensus based on expertise and/or literature from other populations. A summary of the recommendations is provided here across broad age categories: infant (0–11 months), childhood (1–12 years), adolescence (13–17 years) and adulthood (18+ years). Many of the recommendations are similar

Table 1
Clinical questions that informed the nutrition, metabolic syndrome and obesity guidelines

Age group	Clinical questions
0–11 months	<ol style="list-style-type: none"> 1. What nutritional support should be provided when infants with spina bifida first go home from hospital? 2. What guidance on breastfeeding and/or use of breast milk should be given to parents of infants with spina bifida?
1–2 years 11 months	<ol style="list-style-type: none"> 1. What evidence-based information on nutrition, obesity prevention, and obesity management can be provided to parents? 2. What is the best way to manage constipation with diet for this age group? 3. How can healthcare providers communicate with parents about the benefits of a healthy diet in an understandable manner?
3–5 years 11 months	<ol style="list-style-type: none"> 1. What evidence-based information on nutrition, obesity prevention and obesity management can be provided to parents? 2. What is the best way to manage constipation with diet? 3. How can healthcare providers communicate with parents about the benefits of a healthy diet in an understandable manner?
6–12 years 11 months	<ol style="list-style-type: none"> 1. What information do schools and communities need to know about the special dietary and adapted equipment needs of students with spina bifida in order to help children eat and access food independently at school or in the community? 2. What parenting strategies can encourage a balanced and healthy diet for the whole family? 3. What is the best way to manage constipation with diet? 4. Are children with spina bifida and obesity at higher risk for metabolic syndrome? 5. Should screening for metabolic complications of obesity be performed in children aged 6–12 years with spina bifida? 6. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?
13–17 years 11 months	<ol style="list-style-type: none"> 1. What is the most effective protocol to approach diet and nutrition goals in annual spina bifida clinic visits? 2. What are the biggest barriers to healthy nutrition for children with spina bifida in this age group? 3. What self-management skills and resources related to healthy nutrition should be provided for children with spina bifida? 4. What is the best way to manage constipation with diet? 5. Are children with spina bifida and obesity at higher risk for metabolic syndrome? 6. Should screening for metabolic complications of obesity be performed in children aged 13–17 with Spina bifida? 7. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?
18+ years	<ol style="list-style-type: none"> 1. How do nutrition issues vary by different demographics (e.g., age, geography, level of lesion, economic status, race and ethnicity, gender, etc.) among adults with spina bifida? 2. What considerations should be given to nutritional intake when adults with spina bifida are taking medications to address other health concerns? 3. What is the best way to manage constipation with diet? 4. Are adults with spina bifida who have obesity at higher risk for metabolic syndrome? 5. Should screening for metabolic complications of obesity be performed in adults with spina bifida? 6. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?

across the age-ranges (e.g. exploring people's concerns about weight and nutrition) and will need tailoring to the age and ability of the person with spina bifida being seen. Table 2 provides detailed recommendations across more specific age-ranges.

5.1. Infancy (0–11 months)

The initial goal for infants is to ensure that their nutritional needs are met while they are undergoing surgery for neural defect repair, shunt placement or any other medical interventions. The mother should be

encouraged and supported to breastfeed if possible [40], which may include accessing breastmilk banks and/or using specialist pillows, nipples etc. [5]. A referral to a lactation consultant should be made if mothers continue to experience challenges, especially if the baby has severe Chiari malformation. Babies' weight should be carefully tracked to assess for failure to thrive.

5.2. Childhood (1–12 years)

Children should be monitored annually for weight and height or arm span [41]. While calculating BMI

Table 2
Guidelines for nutrition, metabolic syndrome and obesity

Age group	Guidelines	Evidence
0–11 months	<ol style="list-style-type: none"> 1. Assess weight, height, and occipital frontal head circumference at every clinical encounter. (see Supplemental File) 2. Ensure that the family's nutrition plan is followed closely by primary care provider(s): <ul style="list-style-type: none"> – Refer the family to community nursing and other support groups to ensure close monitoring of the child's growth and whether there are issues with feeding and elimination. – Connect the family with the spina bifida specialist clinic nearest them. 3. Provide parents and caregivers with pre- and post-natal guidance and support on breastfeeding. <ul style="list-style-type: none"> – Discuss with them that ideally, infants with spina bifida should breastfeed or be given breast milk exclusively for the first six months. Infants should continue to have breast milk for a year or more, as with all neonates. – Inform the mother that if the child's spinal surgery precludes immediate breast feeding, it will be necessary for her to pump breastmilk to feed the child until it is possible to transfer him or her to her breast. – Urge the mother to begin pumping breast milk within 6–12 hours of delivery. – Emphasize the need to pump frequently (8–10 pumping sessions per 24 hours for the first 7–10 days) to ensure enough will be available once the infant has surgery. – Advocate for babies to be hospitalized near their mothers to facilitate breastfeeding. – Provide mothers with information about accessing breast milk banks and planning for situations where she cannot provide it herself. – Encourage mothers to nurse their child in a flat position for five days following surgery to reduce pressure on the wound to avoid a cerebral spinal fluid leak. – Provide the mother with information about breastfeeding equipment options that can help meet the individual needs of the child with spina bifida (e.g., different types of propping pillows, nipples, bottles, pumps, latex-free equipment, and supplemental nursing systems). – Mention that severe Chiari malformation may affect successful latching and coordination of sucking, swallowing, and breathing. A referral to a lactation consultant should be made if mothers continue to experience challenges. – Provide support to mothers to thicken their breast milk to prevent aspiration. – Suggest breastfeeding or non-nutritive sucking (finger or pacifier) as ways to provide comfort to the infant and assist with pain management for acute procedures such as injections. – Highlight that the baby's transition from drinking breastmilk to eating solid food can cause constipation. – Refer infants with slow weight gain and failure to thrive for multi-disciplinary follow up. 	Clinical consensus as well as [5, 40, 41, 60–66]
1–2 years 11 months	<ol style="list-style-type: none"> 1. Assess weight, height at every clinical encounter (see Supplemental File) 2. Measure occipital frontal head circumference until two years of age (see Supplemental File)) 3. Support families to establish healthy food behaviors/relationships with their child. Tailor the advice to the family as appropriate. <ul style="list-style-type: none"> – Start introducing healthy foods as early as possible to get them integrated into food preferences. It may be harder to do so later when food preferences are more entrenched. – Recognize that children with Chiari malformation may have sensitivity to different food textures. – Consider that adults and families with lower incomes may experience food insecurity. – Caution the parents that habitually providing unhealthy foods can lead to a cycle of more requests and greater consumption of unhealthy foods Early healthy feeding practices can help avoid this. – Discuss with parents and caregivers that if they overly restrict food, especially energy-dense foods that are high in fat and have a low water content (e.g. cookies, chips, nuts), this can lead to over-eating of those foods when they become available. Therefore, balance is needed. – Caution parents against using food as a reward/ positive reinforcement, which can create an unhealthy relationship with food that is hard to break later and may lead to undesirable eating behaviors. Provide parents with other strategies for positive reinforcement rewards such as praise, stickers, and small toys. – Educate families on the importance of consuming a balanced diet and how it affects the whole body. – Discuss that some fluid and food choices used to help ensure hydration and bladder/bowel function are not necessarily the right choices for weight management (e.g. chocolate milk, juice, and sports beverages). Encourage them to hydrate by drinking non-caloric fluids (e.g. water, club soda, sugar free flavored drinks). 	Clinical consensus as well as [2, 3, 6, 8, 12, 15, 41–43, 60, 67–77]

Table 2, continued

Age group	Guidelines	Evidence
3–5 years 11 months	<p>4. Speak with parents about nutrition in terms of their child's health and growth.</p> <ul style="list-style-type: none"> – Provide regular opportunities for parents to discuss any concerns about their child's weight, growth and/or eating behaviors. A trusting therapeutic relationship can greatly facilitate an honest and open discussion. – Partner with parents to identify and address specific challenges that the family is facing. – Discuss that poor eating habits and reduced activity may lead to obesity, constipation, skin breakdown, osteoporosis, anemia, and other problems. Additionally, mention that children with spina bifida have a high risk of obesity because they have less calorie-burning tissue (lean body mass) and a lower rate of burning calories (metabolic rate). – Show parents the trajectory of a child's weight and height (or other measures of growth and adiposity), if appropriate. Use a growth chart as a visual aid, without referring to growth cut-offs developed for typically developing children. A steeply increasing trajectory would indicate that increased weight or obesity may be a concern and warrants proactive discussions of preventative strategies. – Highlight the importance of parents modeling healthy behaviors to their children from an early age. Encourage the whole family to get involved in healthy living activities, not just the child with spina bifida. – Discuss that children with spina bifida, especially those who are non-ambulatory, who undertake low levels of physical activity and have higher body fat levels or contractures, are at increased risk for bone fractures. Encourage physical activity and healthy lifestyles (See Physical Activity Guidelines) <p>5. Provide guidance on maintaining good bowel health.</p> <ul style="list-style-type: none"> – Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals. – Recommend the same guidelines for daily fiber intake that are recommended for all children: <ul style="list-style-type: none"> * 1–3 years: 19 g * 4–8 years: 25 g * 9–13 years: Female: 26 g; Male: 31 g * 14–18 years: Female: 26 g; Male: 38 g – Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. – Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the daily Maintenance Fluid Requirements (24 hour period) Calculation: <ul style="list-style-type: none"> * 100 mL/kg for the first 10 kg body weight * + 50 mL/kg for the next 10 kg body weight * + 20 mL for every kilogram of body weight over 20 kg – Further guidance can be found in the Bowel Function and Care Guidelines. <p>6. Screening for dyslipidemia (fasting lipid profile) is recommended every two years from 2 years of age if the child's BMI is above 95th percentile or if a family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives is present.</p> <p>1. Conduct annual assessments of weight, height, or arm span, and calculate BMI. (see Supplemental File)</p> <p>2. Discuss general weight-management principles with all families and highlight the importance of healthy behaviors for the entire household.</p> <ul style="list-style-type: none"> – Provide regular opportunities for parents to discuss concerns with their child's weight, growth and/or eating behaviors. A trusting therapeutic relationship can greatly facilitate an honest and open discussion. – Emphasize the broad benefits of healthy eating and physical activity, offering strategies to enable the child to incorporate healthy lifestyle behaviors appropriate to their abilities. – Consider that adults and families with lower incomes may experience food insecurity. – Highlight that early eating patterns and relationships with food are critical for ongoing good nutrition through the lifespan. – Discuss that some fluid and food choices to help ensure hydration and bladder/bowel function are not necessarily the right choices for weight management (e.g. chocolate milk, juice, and sports beverages). 	Clinical consensus as well as [2, 8, 9, 12, 17, 19, 42, 43, 46, 67, 68, 72, 76, 77]

Table 2, continued

Age group	Guidelines	Evidence
6–12 years 11 months	<ul style="list-style-type: none"> – Show parents the trajectory of a child’s weight and height (or other measures of growth and adiposity). Use a growth chart as a visual aid, without referring to growth cut-offs developed for typically developing children. A steeply-increasing trajectory would indicate that overweight or obesity may be a concern and warrant proactive discussions of preventative strategies. – Discuss with parents, if relevant, that BMI is an imperfect indicator of health in all young people and especially in children with spina bifida due to difficulties measuring height and body composition. – Consider monitoring other measures of adiposity, such as waist circumference. (see Supplemental File) – Explain that most children with spina bifida follow typical growth patterns until they are four years of age. After that, increased fat mass (versus lean mass) has been found when compared with children without spina bifida. – Discuss that linear growth or height will also be slower than peers without spina bifida due to paresis or paralysis of lower limbs, which also reduces calorie requirement. – Highlight that children with spina bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D. <p>3. Provide guidance on maintaining good bowel health.</p> <ul style="list-style-type: none"> – Explain that increased fiber in the child’s diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals. – Recommend the same guidelines for daily fiber intake that are recommended for all children: <ul style="list-style-type: none"> * 1–3 years: 19 g * 4–8 years: 25 g * 9–13 years: Female: 26 g; Male: 31 g * 14–18 years: Female: 26 g; Male: 38 g – Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. – Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the daily Maintenance Fluid Requirements (24 hour period) Calculation: <ul style="list-style-type: none"> * 100 mL/kg for the first 10 kg body weight * + 50 mL/kg for the next 10 kg body weight * + 20 mL for every kilogram of body weight over 20 kg – Further guidance can be found in the Bowel Function and Care Guidelines. <p>4. Screening for dyslipidemia (fasting lipid profile) is recommended every two years if the child’s BMI is above the 95th percentile or a family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives is present.</p> <ol style="list-style-type: none"> 1. Conduct annual assessment of weight, height or arm span, and BMI. (see Supplemental File) 2. Consider monitoring other measures of adiposity, such as waist circumference. (see Supplemental File) 3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. 4. Highlight dietary needs specific to living with spina bifida: <ul style="list-style-type: none"> – Discuss the importance of consuming fiber and water to manage bowel and bladder health. Sources of fiber include fruit, vegetables, whole wheat or whole grain bread and cereals. A mix of each along with regular fluids will help avoid constipation. – Advise limiting sugary drinks such as juice, chocolate milk, and sports beverages. – Discuss that children with Chiari malformation may have a sensitivity to different food textures. – Recommend that the child have access to food purchasing and preparation spaces. – Recommend a diet with adequate calcium and vitamin D for children with spina bifida, to avoid fractures due to osteoporosis. – Consider that adults and families with lower incomes may experience food insecurity. – Refer clients to the National Center on Health, Physical Activity, and Disability http://www.nchpad.org), which provides advice on nutrition and physical activity for persons with disabilities, including spina bifida. 	Clinical consensus as well as [2, 6, 8, 12, 44–46, 67–69, 73, 75, 76, 77–84]

Table 2, continued

Age group	Guidelines	Evidence
	<p>5. Provide families with nutritional information tailored to their circumstances.</p> <ul style="list-style-type: none"> – Consider a family’s geographical location, ethnicity, access to food, and other related factors when providing dietary education. – Consider that adults and families with lower incomes may experience food insecurity. – Encourage parents to include their children from an early age to participate in grocery shopping and food preparation, as appropriate to their age and ability. – Suggest parents let their children choose a new healthy food to try. Involving children in choices can lead them to increased independence and interest in their foods and learning about healthy choices. Repeating their exposure to healthy options can help them increase their acceptance and enjoyment of these healthy choices. – Involve children in discussions about healthy lifestyles in order to explore their understanding, perceptions, and priorities regarding nutrition. If appropriate, ask families to identify one or two small, healthy nutrition changes that they feel they could integrate into their daily life. – Consider referral to a “Healthy Lifestyle” program and/or use a smartphone app, while recognizing that few such programs are tailored to individuals with disabilities. – Celebrate any successes, such as drinking more water, introducing a new fruit or vegetable, cutting back on sugary drinks, and having regular meal- times. Focus upon the strengths of the family. – Highlight the importance of parents modeling healthy behaviors to their children from an early age. Encourage the whole family to get involved in healthy living activities, not just the child with spina bifida. – Understand that experiencing food insecurity may lead to a poor-quality diet and have developmental consequences. – Highlight that children with spina bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D. <p>6. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) every two years is recommended in children 10 years of age or older, or at the onset of puberty if it occurs at a younger age, with a BMI over the 85th percentile and have two or more additional risk factors including:</p> <ul style="list-style-type: none"> – family history of Type 2 Diabetes Mellitus (T2DM) in a first- or second-degree relative – high-risk ethnicity – acanthosis nigricans – hypertension – dyslipidemia – polycystic ovary syndrome (PCOS). <p>7. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children up to 8 years of age with a BMI over the 95th percentile or other risk factors for cardiovascular disease</p> <ul style="list-style-type: none"> – family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives – history of diabetes, hypertension, or smoking in childhood <p>8. Screening for dyslipidemia (fasting lipid profile) is recommended for all children aged 9–11 years.</p> <p>9. Provide guidance on maintaining good bowel health.</p> <ul style="list-style-type: none"> – Explain that increased fiber in the child’s diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and whole meal or whole grain bread and cereals. – Recommend the same guidelines for daily fiber intake that are recommended for all children: <ul style="list-style-type: none"> * 1–3 years: 19 g * 4–8 years: 25 g * 9–13 years: Female: 26 g; Male: 31 g * 14–18 years: Female: 26 g; Male: 38 g – Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. – Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the following daily Maintenance Fluid Requirements (24 hour period) Calculation: <ul style="list-style-type: none"> * 100 mL/kg for the first 10 kg body weight * + 50 mL/kg for the next 10 kg body weight * + 20 mL for every kilogram of body weight over 20 kg – Further guidance can be found in the Bowel Function and Care Guidelines. 	

Table 2, continued

Age group	Guidelines	Evidence
13–17 years 11 months	<ol style="list-style-type: none"> 1. Conduct annual assessments of weight, height or arm span, and BMI. (see Supplemental File) 2. Consider monitoring other measures of adiposity, such as waist circumference. (see Supplemental File) 3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. 4. Provide opportunities for teens and parents to talk about their priorities and concerns regarding nutrition and weight. <ul style="list-style-type: none"> – Discuss how nutrition can play an important role in helping individuals with spina bifida maintain a healthy weight, minimize skin breakdown, and increase activity and endurance. – Discuss that children with spina bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D. – Provide regular opportunities for teens to discuss any concerns with their weight, growth, and diet. A trusting therapeutic relationship can greatly facilitate an honest and open discussion. – Identify the teen’s priorities and negotiate goals that meet those priorities as well as the parents’ and clinician’s goals. – Use a strengths-based approach that highlights their nutritional achievements and celebrates successes. – Discuss with families, if relevant, that the BMI is an imperfect indicator of health in all young people and especially those with spina bifida due to difficulties measuring height and body composition. Instead, show the family the trajectory of the child’s weight and height (or other measures of growth and adiposity) on a growth chart as a visual aid. Do not refer to growth cut-offs developed for typically developing children. A steeply increasing trajectory would indicate that overweight or obesity may be a concern and warrants preventative strategies. – Avoid using scare tactics in older children with spina bifida. Instead, discuss the following potential negative consequences of having higher weights, as it relates to their individual circumstances: <ul style="list-style-type: none"> * Moving and transferring may become more difficult, which may also reduce independence and self-care activities. * Increased pressure on the skin when seated for long periods of time (such as when using a wheelchair) may result in skin breakdown * Weight gain alongside existing scoliosis or kyphosis may result in additional breathing problems. – Refer clients to the National Center on Health, Physical Activity, and Disability (http://www.nchpad.org), which provides advice on nutrition and physical activity for persons with disabilities, including spina bifida. – Consider referral to a “Healthy Lifestyle” program and/or use a smartphone app, while recognizing that few such programs are tailored to individuals with disabilities. 5. Consider the broader literature for all older children, given that there is little evidence specifically for teens with spina bifida. For instance: <ul style="list-style-type: none"> – Understand that eating habits generally worsen as children move into the teen years and become more autonomous. – Emphasize the positive health benefits of breakfast and eating fruits and vegetables. Skipping breakfast and low fruit and vegetable consumption is common in teens. – Consider that food insecurity and lower socioeconomic status can be related to poorer diets. – Emphasize to parents that the family setting remains important for teens. Parental modelling, dietary intake, and encouragement are all associated with fruit and vegetable consumption among teens. 6. Discuss opportunities for the teen to participate in nutrition-related activities, such as: <ul style="list-style-type: none"> – Identify the teen’s knowledge level about healthy eating habits. – Encourage the family to identify roles that the teen can play as part of daily life, such as in meal planning, shopping, and food preparation. – Encourage teens to select a new healthy food to try, which can encourage broader food preferences. – Identify the older child’s existing strengths and resources regarding nutrition and how they can be built upon to reach their goals. 7. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) every two years with a BMI \geq 85th percentile and have two or more additional risk factors including <ul style="list-style-type: none"> – family history of Type 2 Diabetes Mellitus (T2DM) in a first- or second-degree relative – high-risk ethnicity – acanthosis nigricans – hypertension – dyslipidemia – polycystic ovary syndrome 	Clinical consensus as well as [3, 8, 12, 42, 43, 46–48, 67, 68, 74, 76, 77, 78, 81, 85–9]

Table 2, continued

Age group	Guidelines	Evidence
18 + years	<p>8. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children with a BMI \geq 85th percentile or other risk factors for cardiovascular disease (family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives, history of diabetes, hypertension, or smoking in childhood).</p> <p>9. Provide guidance on maintaining good bowel health.</p> <ul style="list-style-type: none"> – Explain that increased fiber in the teens' diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and whole meal or whole grain bread and cereals. – Recommend the same guidelines for daily fiber intake that are recommended for all teens: <ul style="list-style-type: none"> * 1–3 years: 19 g * 4–8 years: 25 g * 9–13 years: Female: 26 g; Male: 31 g * 14–18 years: Female: 26 g; Male: 38 g – Recommend that if the teen is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. – Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the following daily Maintenance Fluid Requirements (24 hour period) Calculation: <ul style="list-style-type: none"> * 100 mL/kg for the first 10 kg body weight * + 50 mL/kg for the next 10 kg body weight * + 20 mL for every kilogram of body weight over 20 kg – Further guidance can be found in the Bowel Function and Care Guidelines https://www.spinabifidaassociation.org/guidelines/. <p>1. Conduct annual assessments of weight, height or arm span, and calculate BMI. (see Supplemental File)</p> <p>2. However, explain that BMI is not accurate for people with paralysis, who have lowered ratios of fat to lean muscle tissue and that looking at the trajectory over time may be more useful.</p> <p>3. Consider monitoring other measures of adiposity, such as waist circumference. (see Supplemental File)</p> <p>4. Conduct an annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension.</p> <p>5. Tailor the discussion around healthy nutrition to the adult's context. Consider that adults and families with lower incomes may experience food insecurity.</p> <ul style="list-style-type: none"> – Refer clients to the National Center on Health, Physical Activity, and Disability (http://www.nchpad.org), which provides advice on nutrition and physical activity for persons with disabilities, including spina bifida. – Identify who requires the information about healthy food (i.e. the adult with spina bifida, the caregiver, the attendant, the family member, or others). – Discuss the adult's access to cooking options and food preparation areas. – Involve a social worker or disability organization representative who can speak to adults about available local, state, and federal nutritional benefits such as the Supplemental Nutrition Assistance Program (SNAP), farmer's market vouchers or coupons, and other sorts of food vouchers that are available for eligible individuals. – Consider referral to a "Healthy Lifestyle" program and/or use a smartphone app, while recognizing that few such programs are tailored to individuals with disabilities. <p>6. Provide information about potential interactions between nutrition in foods and medications.</p> <ul style="list-style-type: none"> – Highlight that some medications, such as corticosteroids, have side-effects including weight gain, increased appetite, high blood pressure and a higher risk of developing osteoporosis or diabetes. – Provide information about specific foods and beverages that may interact with medications, such as anti-hypertensive, anticoagulant, or corticosteroid medications. – Encourage adults to disclose any prescribed, over-the-counter complementary and alternative medications they are taking to all of their healthcare providers, including pharmacists. – Emphasize the importance of reading medication labels to identify any dietary contraindications. If this is difficult, discuss other ways that the adults could find out about potential contraindications, such as making the medication labels available in a larger font or asking the pharmacists for assistance. 	Clinical consensus as well as [2, 46, 49–51, 81, 91–95]

Table 2, continued

Age group	Guidelines	Evidence
	7. Screening for abnormal blood glucose is indicated as part of assessing cardiovascular risk assessment in all adults aged 40 to 70 years who have a BMI > 25 kg/m ² . Persons who have a family history of diabetes, have a history of gestational diabetes or polycystic ovarian syndrome, or are members of high risk racial/ethnic groups may be at increased risk for diabetes at a younger age or at a lower BMI. Clinicians should consider screening earlier in persons with one or more of these characteristics.	
	8. Screening for dyslipidemia (fasting plasma profile) is recommended for all men ≥ 40 years of age, and women ≥ 50 years of age or postmenopausal. Adults with the following risk factors should be screened at any age: current cigarette smoking, diabetes, arterial hypertension, family history of premature coronary heart disease, family history of hyperlipidemia, high risk ethnicity (individuals of First Nations or of South Asian ancestry), or the presence of rheumatoid arthritis, systemic lupus erythematosus, psoriatic arthritis, ankylosing spondylitis, inflammatory bowel disease, chronic obstructive pulmonary disease, chronic HIV infection, chronic kidney disease, abdominal aneurysm, or erectile dysfunction.	

is included in the guidelines, it should be recognized that BMI is an imperfect indicator of body composition in children, especially those with spina bifida [42] (see more detail below).

The majority of recommendations across the childhood age-span focus on establishing and maintaining healthy feeding and nutrition practices. Parents should be provided with regular opportunities to discuss any concerns about their child's weight, growth and/or eating behaviors [43]. It is important to talk non-judgmentally with parents about nutrition in terms of their child's health and growth and how the early establishment of healthy eating habits can have long-term health benefits for the child and their family. This early guidance should highlight the balance between providing healthy options and not overly restricting certain foods (especially those thought of 'treats') [21]. As the child ages, parents can be encouraged to include them in food planning, shopping and preparation, as well as encouraging them to make choices, such as choosing a new healthy food [44]. Involving children in making choices can lead to increased independence and interest in their foods [45]. Appropriate fluid and fibre guidelines should also be provided according to the child's age (see Table 2).

However, while discussing ideal feeding practices, it should be recognized that food insecurity can greatly affect the family's access to nutritious food [2]. Furthermore, it should be acknowledged that children with Chiari malformation may have sensitivity to different food textures that limit their diet [6]. Developing a trusting therapeutic relationship can greatly facilitate honest and open discussions around these topics [8].

When plotting a child's weight and height on a growth chart, it is important that growth cut-offs developed for typically developing children are not used as comparators [2]. A steeply increasing trajectory in the child's growth may warrant proactive discussions of

possible obesity prevention strategies. Assessing height and/or length in children with spina bifida accurately can be challenging, which can bias BMI. Additional measures such as waist circumference should therefore be considered to augment other clinical information [46]. Recommendations for diabetes and dyslipidemia screening vary according to age, BMI percentile and presence of risk factors (see Table 2 for details).

5.3. Adolescence (13–17 years)

Healthcare providers should provide opportunities for teens and parents to discuss any concerns with nutrition, growth, and weight. The therapeutic relationship can be strengthened by identifying adolescents' priorities and goals and having strengths-based conversations that highlight their successes [8,47]. Adolescents should be encouraged to learn about nutrition and how to prepare healthy food. However, the impact of low income and food insecurity on nutrition should be recognized, especially if the adolescent is living independently [48].

In addition to regular growth monitoring, annual assessment of blood pressure is recommended to monitor for pre-hypertension or hypertension. Screening for dyslipidemia and diabetes should be conducted if the adolescent meets the criteria (see Table 2 for details). Fluid and fibre guidelines should continue to be provided.

5.4. Adulthood (18 years +)

Annual anthropometric monitoring continues to be important with the same caveat that BMI is largely inaccurate for people with paralysis [49]. Blood pressure should be assessed annually. In addition, screening for dyslipidemia should be conducted for all men ≥ 40 years of age, and women ≥ 50 years of age or postmenopausal [50]. Adults under this age with other risk

factors should also be screened (see Table 2 for specific criteria).

It is important to tailor conversations about nutrition and weight-management to the adult's context, which includes their access to healthy food, as well as food preparation and cooking facilities. Referral to a social worker or disability organizations may be appropriate to secure additional support. Also, possible interactions of certain food with medications should be discussed [51].

6. Discussion

The guidelines presented here on nutrition, metabolic syndrome and obesity were a new addition to the Spina Bifida Association's 2018 guidelines [7] as they were not previously part of the 2006 guidelines [38]. This guideline helps to address the nutritional needs of people with spina bifida to maximize wellness across their lifespan. Preventing conditions associated with poor nutrition –including overweight, obesity and metabolic syndrome– is also important to promote life-long health. However, through the process of creating these guidelines, we identified several gaps in the current evidence base that should be targeted in future research.

First, an evidence-based algorithm to determine the energy needs of people with spina bifida across their lifespan would be a critical tool to assist with nutritional intake and weight management. To be most useful in clinical practice, this algorithm would need to consider the heterogeneity of people with spina bifida, including their lesion level and mobility functioning.

Second, despite people with spina bifida being at a higher risk of developing obesity [3], there are currently no evidence-based guidelines for weight-management, obesity prevention, or obesity treatment specific to children and adults with spina bifida. This is a critical gap, because guidelines created for the general population [52,53] do not take into consideration factors that may present greater challenges for this population, such as altered body composition and metabolism, and sedentary lifestyles [19,26]. Other biopsychosocial factors can also uniquely impact nutritional intake, such as differences in tolerating certain tastes and textures, concerns around continence and unequal access to fresh food and preparatory methods [3,54].

However, focusing solely on weight-loss is not recommended, as it may encourage disordered eating [34]. This aligns with the guidelines' primary objective "to maximize and support wellness through the lifespan."

Therefore, it is recommended that healthcare providers focus on the broader benefits of good nutrition and physical activity in discussions with people with spina bifida. As always, these discussions should be tailored to the needs and circumstances of the individual and their families. Practical tools to enable supportive conversations have been requested by healthcare providers, patients and families alike [8]. In response to this, the 'Fostering Positive Weight-related Conversations Casebook' was recently developed that provides guidance specific to discussing weight-related issues in a spina bifida context (readers are directed to www.hollandbloorview.ca/conversationcasebook). However, further work is needed to identify the most effective strategies for implementing resources such as the Casebook into clinical practice [55,56].

Third, while metabolic syndrome is considered a concern in people with spina bifida [36,37], there are no comprehensive, high quality data to understand the risk profile of children and adults with spina bifida or guide clinical decision-making around screening protocols. Understanding the role of weight management interventions (if any) in preventing or managing metabolic syndrome is also needed.

Fourth, weight monitoring would be more informative if spina bifida-specific growth curves and weight classification cut-offs were developed. These would greatly assist in monitoring and managing children and adults' weight, rather than using norms developed for people without disabilities. In the absence of any spina bifida-specific growth norms, this guideline refers to BMI. However, healthcare providers are urged to remember that BMI is an inaccurate indicator of health generally [28] and especially in those with spina bifida [19]. Rather than focusing on a single measurement, monitoring trajectories over multiple time-points can provide more useful information about a person's growth and/or weight [2,28].

Finally, the supplemental file accompanying this article provides instruction on standardized assessments of BMI and body composition, such as waist circumference and skinfold thickness. Although these assessments have shown good correlations with fat mass measured by dual-energy X-ray absorptiometry [57,58], their accuracy is still dependent upon assessor skill [59]. Assessing body composition in an accurate and accessible manner is still extremely challenging, given that most clinical settings do not have access to specialized equipment.

7. Conclusion

The Nutrition, Metabolic Syndrome and Obesity Guidelines described here were created to maximize and support wellness for all people with spina bifida across their lifespan. While some recommendations are specific and technical, others are less exact due to a lack of evidence for this population. However, our objectives were deliberately aspirational. The working group believes that the recommendations here will help promote better health and wellness in people with spina bifida of all ages.

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The Spina Bifida Association has already embarked on a systematic process for reviewing and updating the guidelines. Future guidelines updates will be made available as they are completed.

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Conflict of interest

The authors have no conflicts to declare.

Supplementary data

The supplementary files are available to download from <http://dx.doi.org/10.3233/PRM-200753>.

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The hallmark clinical presentation is the inability to “voluntarily” control the defecation process or the inability to routinely perform elimination of stool/waste from the body. Patients with Spina Bifida may also experience:

- Constipation, very often severe
- Bowel obstructions, rectal impaction with sensory loss
- Hemorrhoids
- Nausea and vomiting
- Abdominal pain, bloating-distention, cramping, and lethargy – “sluggish feeling”
- Diet changes = decreased appetite - “grazing-snacking”
- Dehydration = electrolyte disturbances and increased UTI risk
- Soiling and unplanned evacuation of stool / social anxiety

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- ✓ Can assist in reducing time spent with patient for dressing/redressing due to episodes of incontinence or fecal discharge.²
- ✓ Can virtually eliminate episodes of incontinence.³
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