Chapter

# Impact of Nutrition in Spinocerebellar Ataxia

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## Abstract

This chapter explores the link between the health outcomes of spinocerebellar ataxia and diet and nutrition as well as overall quality of life and well-being that is achieved as a result of nutritional support and nutritional profile. Spinocerebellar ataxia is a hereditary condition characterized by degenerative changes to parts of the brain, extending to the spinal cord, that affects mobility and voluntary actions. Due to the deteriorating impact of this neurological disorder, the management of health and wellness of the individual is imperative in stemming physiological decline and morbidity. The connections between dietary intake, quality of life and well-being are important components of the health response in providing optimum health outcomes for clients diagnosed with spinocerebellar ataxia. Consequently, an examination of factors that impede, promote and generally affect dietary intake, nutritional status and profile is essential towards improving disease related quality of life and morbidity and mortality risk. The cyclical impact of the neurological condition on nutritional status and its corresponding impact on disease progression is an important exploratory point. Finally, recommendations and standardized guidance are crucial to expanding the health care approach and the overall wellness of individuals with spinocerebellar ataxia.

**Keywords:** nutritional support, quality of life, muscle strength, dysphagia, weight control and wellness



## 1. Introduction

### 1.1 Methodology and literature review

A content analysis of the literature, especially produced over the last decade, was carried out exploring the condition spinocerebellar ataxia and the possible impact that diet and nutrition may have on the outcome of the condition, utilizing the keywords as a guide. The paper was presented in sections concerning diet and wellness, analyzing each concept and the relationship between diet, nutrition, and spinocerebellar ataxia especially in light of quality of life and wellness.

## 2. Overview of spinocerebellar ataxia and diet and nutrition

Spinocerebellar Ataxia is a heterogeneous group of neurodegenerative ataxic disorders with autosomal dominant inheritance. It is an inherited progressive disorder

with clinical features including loss of balance and coordination accompanied by slurred speech. The clinical outcome of this disease is usually manifested in adulthood [1, 2]. This condition is similar to many non-communicable diseases insomuch as it is not transmissible from personal contact and has significant impact on quality of life and wellness. Considering the deteriorating neurological features of the condition, health and wellness maintenance is paramount. Diet and nutrition is featured heavily in its effect on clinical outcomes of this condition. Spinocerebellar Ataxia may have important health impact and nutritional risk profile effect due challenges with swallowing, dysphagia, dependence in meal preparations, muscle and coordination challenges. These physiological changes that are characteristic of spinocerebellar ataxia impact dietary intake and negatively affect lean body mass [3, 4]. Importantly weight loss is a predictor of morbidity risk where sepsis and concomitant illnesses were measured outcome criteria [5]. As a consequence of these physical changes and nutritional impact, the healthcare team must be responsive to the special needs of this cohort of individuals to stem possible negative outcomes. A focus on diet and wellness may accrue to significant health benefits in this population.

Diet and wellbeing are pervasive concepts impacting sociology, psychology, medicine, and human thought. Their relationship to health and happiness are significant to human development and livelihood. This chapter explores the connection between diet and wellbeing especially as they contribute to or impact health generally and particularly that of clients diagnosed with spinocerebellar ataxia. There are important factors that create the link between diet and wellness and include dietary intake, quality of life and wellbeing. The bridge between these concepts will contribute to a better understanding of the impact of diet and wellness and provide summative recommendations for the maintenance of health especially in individuals with neurological disorders such as spinocerebellar ataxia.

### 2.1 Dietary intake

Dietary intake is the food and nutrient consumed by an individual daily to maintain life, health, and functionality. Dietary intake is guided by set of recommendations/standards for the daily intake of nutrients and other food components based on the recommended daily allowances. These measurements are used to assess or track food, nutrient or any non-nutritional intake by individuals. The main purpose for assessing an individual's dietary intake is for nutritional screening and surveillance, which can be used to guide research. In individuals with neurological disorders, resting energy expenditure due to hypermetabolism is increased [6]. Resting energy expenditure is the caloric requirement of an individual needed to maintain life and the function of essential organs and systems at rest. It is an important feature of health management in people with neurological illness. Moreover, it is imperative that caregivers and the health care team manage caloric needs in this population, since under supply of energy increases health risks in this vulnerable group of individuals.

### 2.2 Quality of life

The World Health Organization defines "Quality of Life as an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" [7]. The quality of life can be affected by an individual's physical health, psychological state, their personal beliefs and social relationships in relation to significant features of their environment. The Centers for Disease Control and Prevention views quality of life as a concept that can be applied to various disciplines to evaluate important aspects of individuals' lives. The concept is usually defined differently for each

discipline. The concept of health-related quality of life has been explored since the 1900s. It evaluates a person's perception of their physical and mental health in relation to their health status and risks, social support and their socioeconomic status. Health- related quality of life also includes the community by identifying the resources, practices and policies of the community that may have an influence on the population's health perceptions and how well they function [8]. Importantly, in spinocerebellar ataxia many variables help to determine quality of life such as the capacity to carry out activities of living, the level of pain or discomfort and the independence to manage self care, Furthermore, the evidence considers disease related quality of life as an important element in neurological disorders and spinocerebellar ataxia in particular. This element of quality of life examines the progression of the disease relative the activities and competencies that the client retains [9]. The greater the physical independence, self care independence, the fewer the neurological deficit along with higher levels of comfort the higher the levels of general quality of life and disease related quality of life [9]. Nutrition and diet are inextricable to quality of life in these patients, so much as well balanced diets, lower health risks, meets energy needs and is associated with reductions in the progression of the disease. These concepts are explored in greater detail in the remainder of the chapter.

## 2.3 Wellbeing

The concept of well-being is usually viewed in a positive way for individuals as it indicates that they perceive that their lives are functioning well. An individual that has good living conditions such as proper housing and are employed is essential to their well-being [8]. Well-being is also recognized state where an individual experiences good health, happiness and prosperity, which includes their good mental health, having the ability to manage stress, satisfaction with their lives and the feeling that they have a meaning and purpose in life. Most people aim to achieve well-being because it represents positivity and general health [10].

The quality of life and wellness of an individual with a neurological disorder are affected by a cyclical relationship between nutrition and the disease state. Where poor nutrition is thought to exacerbate the clinical features of the disease and worsened diseases states negatively impact on nutritional risk profile. Specifically, chewing and swallowing difficulties affecting nutrient intake where suboptimal calorie intake is achieved. Furthermore, biochemical and physiology factors of neurological diseases in general and spinocerebellar ataxia in particular affect nutritional status through limited nutrient absorption and utilization, and physiological process affecting gross dietary intake including infection, depression, muscle atrophy, rigidity, tremor, dyskinesia, reluctance to feed, and dysphagia [6]. The combination of these factors may lead to undesired weight loss with worsened risk of infection, and sepsis. Furthermore, poorer nutritional states are associated with worsened ataxia, dyskinesia and tremors. Therefore, it is imperative to evaluate nutritional intake and specifically, caloric intake and expenditure of individuals properly in order to improve the quality of life and enhance health outcomes in clients with neurological diseases.

## 2.4 Factors that affect dietary intake

There are many factors that can have a positive or negative impact on an individual's dietary intake. The amount and quality of dietary intake is directly linked to nutritional profile of individuals. Considering the need for appropriate quantities of micronutrients and macronutrients to support nutritional wellbeing, the factors underpinning nutritional intake is essential for review. Some of the most

significant factors that impact dietary intake are appetite, importation, physical condition, built environment and health conditions. These factors will be explored in detail with a view to understanding how positive health outcomes can be accrued. Furthermore, these factors will be examined more closely in the context of neurological diseases and spinocerebellar ataxia.

The factors impacting dietary intake are expansive and are summarized in **Figure 1**.

In many countries, especially developing countries, meals are heavily dependent on importation. However, during a pandemic importation is unreliable and unstable. As a result, these countries are at a higher risk of food shortage [12], which will impact national dietary intake. This means that there may be acute or chronic impact on individual access to food primarily resulting in reduced dietary intake. A pandemic also impacts wages, whether through job loss, reduced working hours. The household purchasing power may be diminished creating impaired food access and decreased dietary intake based on the financial challenges brought on by the pandemic [12]. While importation is not controlled at the individual level, it has important personal impact. Spinocerebellar ataxia has been significantly associated with weight loss and BMI decline due to increases in metabolic demand. The risk of weight loss worsens with disease progression [13]. Importantly, weight loss and BMI decline are treated with regularly adequate intake of energy at the estimated levels to meet total daily energy expenditure. This means that efforts must be made by health care workers, families and government to facilitate safe, and adequate access to food so that energy supply may be appropriate in this population. Importation therefore has a tangential impact on the quality of life of individuals with spinocerebellar ataxia. This exists where importation affects food access, access affects dietary intake, which affects energy balance and body weight. If weight is suboptimal, individuals with ataxia are at greater health risk and the reverse is true when weight is ideal.

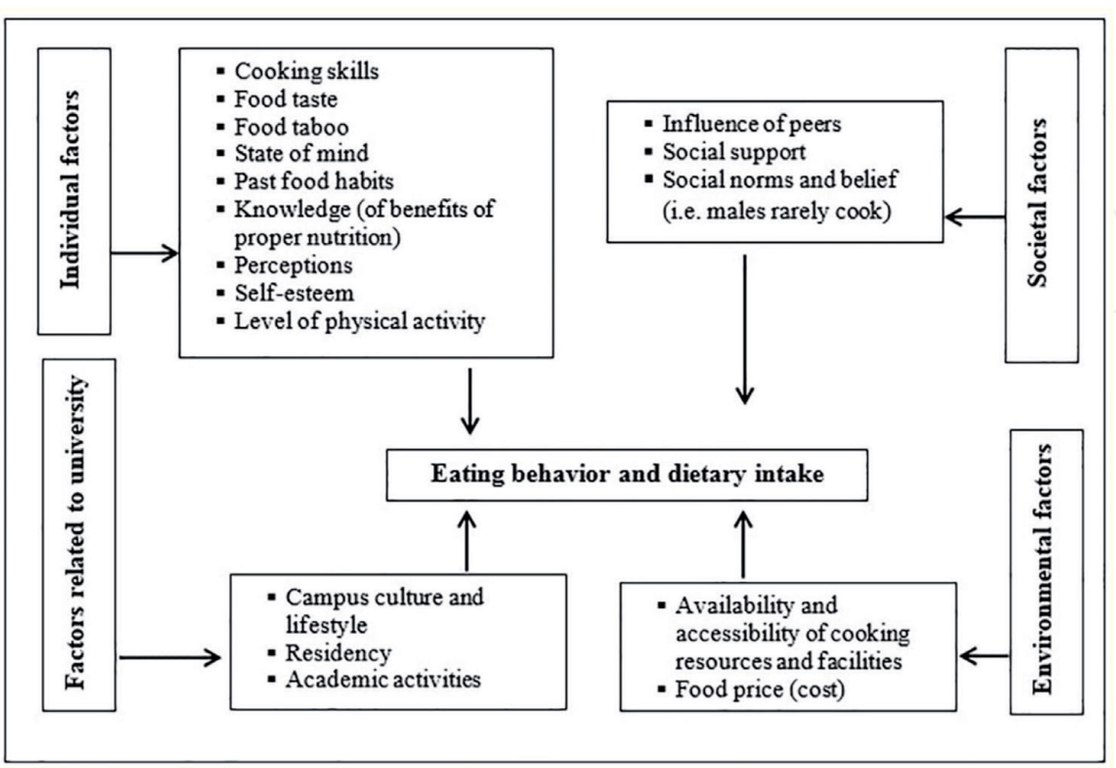


**Figure 1.**
*Factors affecting dietary intake [11], p. 6.*

There are some health conditions that affect an individual's dietary intake; that may range from challenges with digestion and absorption of nutrients or side effects from prescribed medications that interfere with absorption and utilization of nutrients and dietary adjustments may be needed in medical nutrition therapy for specified health conditions. As part of the treatment of some conditions, clients may be required to eat smaller portions, restrict some food or nutrients, or limit the amount they can tolerate. Clients with spinocerebellar ataxia are similarly afflicted. The condition is reported to impact swallowing capacity, digestion, and nutrient storage, especially fat ([6]; Ko, Qu, Black, & Tso, 2020). Some micronutrients including niacin, thiamine and tocopherol have high risk of deficiency in clients with spinocerebellar ataxia [14–16]. Consequently, nutritional support at times can be adjusted and specially formulated to meet their needs. In some instances where digestion is limited, elemental dietary formulations, texture modifications to support swallowing difficulties may be offered, or where gastrointestinal intake is severely restricted parenteral nutritional application may be necessary.

Nutrition and dietary intake have been established as major factors influencing the quality of life of patients diagnosed with spinocerebellar ataxia. Consequently, it is important to expand the dialog of nutrition in understanding how wellness is affected through dietary influence in this unique population. A major element of physical wellbeing is nutritional status which is directly proportional to the quality of dietary intake. Moreover, social, and emotional wellbeing are affected by dietary intake and nutritional status directly due to the components of food and indirectly because of the perceptions associated with the evaluation of personal nutritional status. Several factors affect dietary intake and include but are not limited to appetite, physiological development, health condition, the built environment, effects of a pandemic and social family settings. These factors will be explored in detail.

### 2.5 Appetite

Appetite is an individual's desire to eat food, the body's biological response to a lack of food is hunger. However, an individual's appetite can rise and fall due to various factors, sometimes causing people to eat less or more than their body needs [17]. Appetite can be affected by one's diet, mental health, pregnancy, medications or other health conditions. An individual experiencing a decrease in appetite may lead to a concordant decrease in the general desire to eat food and thus the person may consume less food and nutrients. Appetite is also an important factor impacting the nutritional status of patients with spinocerebellar ataxia. Appetite is primarily affected by the pharmacotherapy approaches used to manage/treat the condition. Drugs including Varenicline and Riluzole are used in the treatment of the neurological features of dystonia and ataxia [18]. The medications are reported to negatively impact on appetite and may potentiate weight loss. These concerns are necessary to be addressed systemically in view of the risks associated with weight loss in spinocerebellar ataxia. Corrective actions including eating by the clock, small frequent meals, colorful and attractive meals, and appetite stimulants may be important to address these appetite changes.

In the general population several personal and psychological factors were examined as factors that contributed to caloric intake. Hunger, appetite, and satiety were identified as important contributors to dietary intake among this population. Appetite was thought to relate to the psychological drive to eat [19]. There remains a concern regarding the factors that regulate appetite but dietary factors including protein and caloric load have been identified as features that influence satiety. At the alternate end of the spectrum, poor appetite is reported as a factor that affects the quality and quantity of food intake in older adults and influences health

outcome and morbidity risk [20]. Markedly appetite exerts an important influence across the lifespan and influences nutritional status and wellness. Appetite may also be affected by the capacity to coordinate movements and feed self especially among the psychological influences. In spinocerebellar ataxia, physiological outcomes including uncoordinated actions limit feeding independence may contribute to reductions in appetite [6, 18, 21]. As medications are introduced to correct physiological impact in these neurological conditions caution needs to be exercised in view of the physical impact that pharmacotherapy may have on appetite. Ultimately, a tight balance must be reached between the management of physical limitations to feeding and the impact on appetite, and pharmacological approaches to managing coordination and their impact on appetite. This must be done with careful observations of diet and appetite especially as the impact on weight balance and health related quality of life.

### 2.6 The built environment

Social changes that have led to structural advances have improved the quality of life of the global population. These changes had led to positive financial impact, changes in commerce, trading, and travel. Conversely, it has had instrumental impact on diet and nutritional intake. Furthermore, even the dynamic of the rural population has changed. Several scholars are identifying that caution should be introduced when these benefits are examined. As the referenced changes have also been recognized as significant drivers of changes in dietary quality, where wealth has led to increased dietary intake, particularly energy and serving sizes [22]. Urbanization has led to reduction in the value of home gardens, reduction in the reliance on agrarian subsistence with resulting declines in the consumption of complex carbohydrates [23]. There is also a notable increased in energy dense, nutrient poor and processed foods. Importantly as well, statisticians have associated the structural changes with dietary changes and with significant steep increases in non-communicable diseases, and sharp declines in quality of life. Moreover, there is a concern with changes in the built environment and green spaces for exercise and physical activity. With automation and innovations in construction, there has been a corresponding decline in physical activity with an increased risk of non-communicable diseases [24]. Physical activity is directly related to nutritional status as physical activity helps to create a balance between caloric intake and expenditure especially as weight maintenance in adulthood is desired. In clients with spinocerebellar ataxia muscle strength, disuse, and incoordination impact on the capacity to engage in the physical activity [6]. Notably, physical activity has been linked to improvement in the quality of life of patients diagnosed with spinocerebellar ataxia. It has been shown to improve physical capacity of the patient, slow the progression of the disease and limit the severity of the physical symptoms of the condition [25]. Furthermore, physical activity has been found to improve antioxidant capacity and reduction in prooxidant damage in patients with spinocerebellar ataxia [26]. It is therefore important, that green spaces are maintained, gyms and other facilities are created, and physical therapy is made available to this population so that the positive influence of physical activity can be accessed by this population.

### 2.7 Health condition

Non-communicable diseases account for more than half of the annual deaths in middle eastern countries [22]. The resultant public health advice has been targeted at reducing obesity through dietary and lifestyle changes to produce nutritional and general wellness. Despite the drive to improve dietary intake of

complex carbohydrates, fruits and vegetables and reduce simple sugars, saturated fats and total cholesterol the rates of overweight and obesity continue to rise with a concordant increase in the Disability adjusted life years (DALY) [23]. Neurological disorders categorized as other account for approximately 2.5% of all neurological disorders between 1990–2015 and remains a significant concern especially in developed countries (**Figure 2**). Nevertheless, health condition remains and important

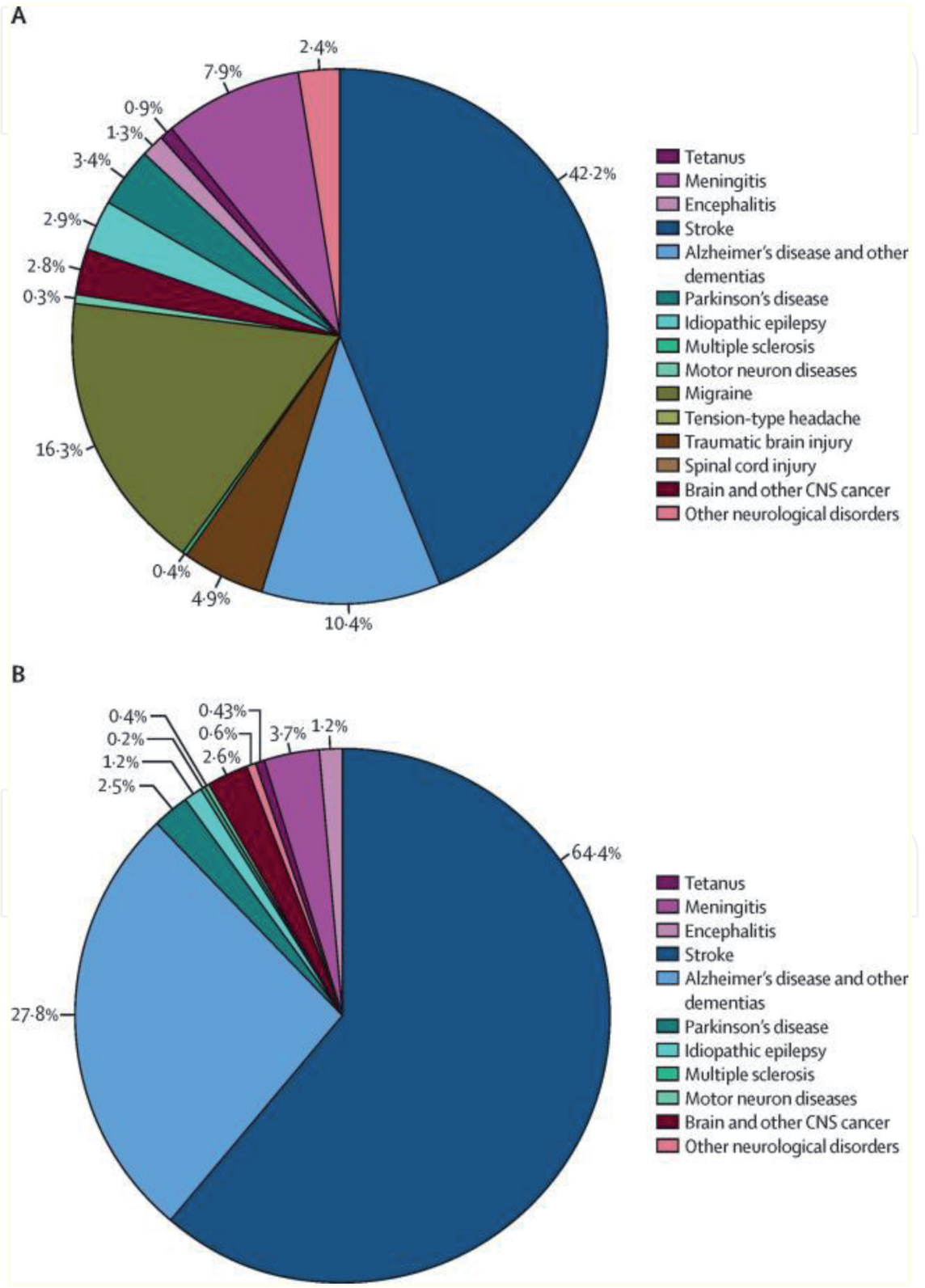


**Figure 2.**
*The global burden of neurological disorders ([27], p. 258).*

factor that influences dietary requirements and recommendations. In clients with neurological disorders especially spinocerebellar ataxia, there are several health conditions that impact on nutrition particularly requirements and dietary intake. Importantly the health conditions of these patients are also affected by nutritional status and intake. Individuals diagnosed with spinocerebellar ataxia experience, dysphagia, loss of lean body mass with weight loss, uncoordinated movement, micronutrient deficiencies including- niacin (Vitamin $B_3$), Tocopherol (Vitamin E) and thiamine (Vitamin $B_1$) as well as impaired fat metabolism and increased metabolism and prooxidant activity [3, 4, 14–16]. Dysphagia, uncoordinated movement, and loss of skeletal muscle mass may together create a challenge with feeding and limit dietary intake potentiating weight loss and reducing quality of life. Consequently, texture modification through thickening of food with caregiver support may help to treat swallowing difficulties and improve dietary intake. Limited muscle mass may be treated by introducing branched chain amino acids from supplements and complete proteins [28]. Micronutrient deficiencies constitute a health concern in spinocerebellar ataxia insomuch as they may influence negatively immune response, antioxidant capacity and energy metabolism ataxia [14–16]. It is therefore imperative that biochemical profile assessments are regularly conducted with a view to guide nutritional support. Increases in complete protein may supply adequate niacin levels, while thiamine levels can be improved from animal-based protein and tocopherol levels can be bolstered by consuming vegetable oils, green leafy vegetables and fortified cereals [29, 30]. Impaired fat metabolism is a critical concern in adults in general, and clients with spinocerebellar ataxia in particular, as this anomaly may lead to cardiovascular health risks including atherosclerosis and may also play a role in non-communicable disease development [6, 15]. To mitigate the negative health outcomes of impaired fat metabolism, caution needs to be taken with respect to meal preparation styles, limiting fry and trimming fats, and the selection of foods items [31]. Foods best suited in this case, should limit trans-fat, thereby reducing processed foods as well as increasing the intake of polyunsaturated fats from fish and nut oils while reducing saturated fats from red meats and animal fat. Finally, prooxidant activity in spinocerebellar ataxia can be managed through the reduction of processed foods, which are thought to have higher levels of prooxidant species and include foods with higher antioxidant capacity such as allium vegetables, fruits and a controlled amount of red wine [32]. Furthermore, when the immune status and antioxidant capacity of clients with spinocerebellar ataxia falls to suboptimal level, several nutritional approaches can be employed to improve these health statuses including supplementation and immunonutrition. Diet has been used to positively influence wellness through positive links with supplemental nutritional support, emotional wellness and immunonutrition. Supplemental nutrition includes the addition of a substance or product with the express goal of improving the intake of key micronutrients including vitamins and minerals [33]. This additional intake is aimed at improving nutritional status and wellness and has been a feature of the public health response in LMICs. It may also be employed in dietary options for clients who have low levels of micronutrients or those who are found to be clinically deficient, which is possible in clients with spinocerebellar ataxia. Supplemental nutrition may be formulated as tablets, pills, shakes and other products. Moreover, immunonutriton is a process or product of bolstering the immunity of individuals through the introduction of a targeted nutrients such as amino acids, especially essential and branched chain, essential poly unsaturated fatty acids, nucleotides, and antioxidants. It is particularly useful in reducing recovery time, improving quality of life and health of individuals who are immunosuppressed [21, 34]. This is another strategy that can be coupled with

biochemical analysis to improve the immune status of the client when per os dietary support has not significantly improved antioxidant capacity, immune status, or micronutrient profile.

## 3. Summary and recommendations

Diet is associated with Quality of Life and Wellness. Several factors impact on the quality of dietary intake including social, physiological, and psychological issues. The dietary quality has important health outcomes ranging from impacts on Disability Life Years to an increased impact on disease burden, risk of concomitant diseases and impact of disease progression. Nevertheless, diet has been shown to improve body weight, muscle strength, immune capacity, disease progression, life expectancy, recovery, and general wellness especially in clients with spinocerebellar ataxia. Given the significance of diet in influencing the illness wellness continuum, public health officials are constantly challenged to improve the population outcomes using diet. The benefits of diet particularly its impact on improving the quality of life of clients with spinocerebellar ataxia, can be best achieved if portion control, nutrient density and meal planning were to be engaged in this population. Portion control addresses the concerns of caloric intake, through a general reduction in the size and amount of a meal at any single sitting and matches caloric requirements with intake goals [35]. This is associated with a strong positive linear relationship with weight maintenance and disease related quality of life. As energy balance is reached, caloric intake equilibrium is achieved, and weight maintenance is maintained even in high metabolic states. This strategy is particularly important as weight loss is mitigated and weight maintenance is achieved. Weight stabilization is an important factor in spinocerebellar ataxia and is an important determinant of disease progression. Nutrient density is a concept of increasing the range of nutrients consumed, particularly micronutrients, for the smallest value of calories [36, 37]. This allows for the increased functional activity of antioxidants, immune supportive micronutrients, and biological supportive nutrients [38]. The concordant impact on recovery, immune support, and reduction in prooxidant activity is important in wellness especially in clients diagnosed with spinocerebellar ataxia in view of the morbidity risk. Meal planning includes multiple principles that benefit overall dietary pattern through the improvement with calorie intake, through energy control, improving diversity and variety and instituting moderation. These result in planned meals that are better aligned with nutritional guidelines and associated with better health outcomes. Generally, these three principles of portion control, nutrient density and meal planning improve the metabolic profile of the individuals who consistently institute these practices, with improved Body Mass Index (BMI) values, more ideal body weight, healthier body fat and lipid profile with lower chronic disease risk profile. The benefits to be had from diet are best achieved with consistency, moderation and diligence and provide significant sustainable advantages to quality of life and wellness. In clients with neurological disorders, coordinated nutritional support and control are positively associated with better disease related quality of life through weight control, healthy micronutrient status and calorie control. It is essential that these activities are guided by appropriate biochemical and clinical tests and involve personalized and individualized strategies for the most beneficial impact. When instituted in spinocerebellar ataxia, dietary programmes involving principles of meal planning, nutrient density and energy balance redound to significant health outcomes such as weight maintenance, with slowed disease

progression, improved immune status and healthy micronutrient. Therefore, a multi-team approach is recommended if the best health outcomes are to be achieved in spinocerebellar ataxia ensuring the involvement of the nutritionist/ dietician, the biochemist/ phlebotomist, physiotherapist, the neurologist and the general medical practitioner.





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