The Amino Acid Diet

The Amino Acid (AA) diet is one of many dietary approaches for people with Spinal Muscular Atrophy (SMA). Since every individual with SMA is unique, so too is their dietary management. There are many ways to provide an AA diet. Most people take this diet through a feeding tube, but others take it orally. The diet is individualized based on a person's calorie, protein, fat, carbohydrate, vitamin, mineral, and fluid needs. Other considerations include the person's respiratory status, SMA type, strength, lab values, family preference, diet tolerance, and insurance coverage of special formulas.

The AA diet is a moderate protein (7-10% of calories or 0.8-2 grams per kilogram of body weight), low fat (<30% of calories), high carbohydrate (60-75% of calories) diet that provides adequate nutrition for either growth (for children) or weight maintenance. Protein is supplied in the form of free amino acids in an elemental formula but some intact protein is given when vegetables and plant based milks are included. (For those who plan to administer the AA diet orally: elemental formulas do not taste very good. We recommend a trial of the formula before committing to this diet. Added juice can sometimes make the formula taste more acceptable.) An appropriate intake of protein, fat, and carbohydrate is needed to prevent deficiency or excess of these nutrients. A source of essential fatty acids is added to most AA diets to prevent essential fatty acid deficiency. The remainder of the calories comes from a variety of carbohydrates (juice, plant based milks, fruits, and vegetables). It is important to work with your dietitian to develop a formula recipe that meets you or your child's specific nutrient needs.

Elemental Formulas: For children older than 1 year of age, the diet typically starts with a low fat elemental formula such as Pediatric Vivonex and/or Tolerex, which both contain protein in its most broken down form of free amino acids. Pediatric Vivonex is 12% protein, 63% carbohydrate, and 25% fat. The fat comes from medium chain triglycerides (which are very easy to digest) and soybean oil (which contains both omega 6 and omega 3 essential fatty acids). Tolerex is 8% protein, 90% carbohydrate, and 2% fat. The small amount of fat comes from safflower oil (which contains mostly omega 6 essential fatty acids). Therefore, when using this formula as the only source of protein, oil that is rich in omega 3 essential fatty acids must be included as well as an additional source of omega 6. If additional protein is needed, then supplemental amino acid products are used such as Nutricia Complete Amino Acid powder or Essential Amino Acid powder.

Due to the role of fat in brain development, a low-fat AA diet is not typically recommended for infants less than 1 year of age. If an amino acid formula is desired in infancy, an elemental infant formula which contains adequate fat, such as Neocate Infant or Elecare is often used. In some cases, a low-fat elemental formula is used together with breast milk or higher fat elemental formula.

Breast milk is mentioned above for use in diets of infants less than 1 year. Some families of older children with SMA also incorporate breast milk into an AA diet and feel that breast milk is

well-tolerated. Proteins in breast milk are whey and casein, which are milk proteins and are not in the free amino acid form. Approximately 55-60% of the calories in breast milk are from fat.

Essential fatty acids: Omega 3 and omega 6 fats are considered essential fatty acids because they cannot be made by the body and must be obtained from the food we eat. If an individual is only getting Tolerex, an additional source of fat is needed to prevent essential fatty acid deficiency. This often comes from a combination of an omega 6 rich oil (like safflower or evening primrose oil) and an omega 3 rich oil (walnut oil, flaxseed oil), or even breast milk. A minimum of 3-5% of the calories in the diet must come from essential fatty acids to prevent deficiencies.

Fluid: Water and/or an electrolyte based drinks such as Pedialyte or Enfalyte along with some fruit juice and/or plant based milk are used in an AA diet. Fruit juices commonly used are apple, white grape, prune and pear. Plant based milks commonly used are almond, rice, and hemp. These fluids are added to provide 100-130% of a child's fluid needs since fluid needs are often increased in SMA due to increased respiratory losses and sweating and to thin out secretions.

Common Additions to AA diet: After adding the formulas and fluid, either pre-made baby fruits and vegetables or homemade pureed fruits and vegetables are often added. Add one fruit or vegetable at a time to check for tolerance. Commonly used fruits include apples, prunes, bananas, pears, and peaches. Commonly used vegetables are sweet potatoes, squash, green beans, and carrots. The amount of baby food added depends on calorie needs.

Vitamin/Mineral and Other Supplements: The last things added to an AA diet are vitamin, mineral, and other supplements. A separate vitamin/mineral supplement is often needed due to the low calorie needs of individuals with SMA. Even though amino acid based formulas contain vitamins and minerals, individuals with SMA have reduced calorie needs and do not receive a large enough amount of formula to meet basic vitamin/mineral requirements. Additionally, many people with SMA need extra vitamin D and calcium to prevent additional bone loss since many individuals with SMA are at increased risk for low bone density (primary to lack of weight bearing). Other commonly added supplements are vitamin C for immune health, vitamin K for bone health, and iron to prevent or treat iron deficiency anemia. Additional supplements may also be used such as carnitine to improve fat metabolism (or for carnitine deficiency), turmeric to decrease inflammation, a probiotic for gut health, elderberry for immune health, salt to ensure adequate sodium intake, and coenzyme Q10 to provide antioxidant benefits and to help with energy production. There are many other supplements that are often used based on individual needs and family preference. Megadoses of supplements are not recommended and can often do more harm than good. It is best to discuss vitamin, mineral, and other supplementation with your doctor and dietitian in order to meet specific needs and to avoid interactions or intakes that exceed recommendations.

Typical recipe for an AA diet:
Elemental formula (protein 0.8 to 2 grams protein/kg body wt and total fat <30% kilocalories)

Water (may have some portion as oral rehydration solution such as Pedialyte®)
Juice and/or dairy-free, soy-free beverage
Pureed fruit
Pureed vegetable
Oil (if needed to meet essential fatty acid needs)
Vitamins, minerals, and supplements as needed or per family preference
All ingredients are combined in a blender and refrigerated for use within 24 hours.

Signs of Intolerance: Once a recipe for the AA diet has been developed, no changes are made for several days to check for tolerance. For sensitive people with SMA, ingredients in the recipe are often added in stages: Formula and water/oral rehydration, juices, non-dairy/non-soy beverages, pureed fruits/vegetables (one kind at a time), added fats, vitamins, minerals and other supplements. Signs of intolerance might include increased heart rate, trouble breathing, increased secretions, and bloating/abdominal upset. Keep in mind, however, these signs may also indicate that a person with SMA is ill or that the disease is progressing and cannot always be attributed to diet.

Recommended nutrition labs for the AA diet: Labs are recommended at least annually for those individuals receiving an amino acid diet. Labs are used to optimize the diet and prevent deficiencies or excessive intakes. The most often recommended labs include: Essential fatty acid profile (recommended if on a very low-fat diet), 25 hydroxy vitamin D, comprehensive metabolic panel, complete blood count, protein status lab (such as prealbumin, plasma quantitative amino acid), and other labs as decided by doctor/dietitian based on dietary intake or signs/symptoms (i.e. iron studies, free and total carnitine, cholesterol panel, B vitamins, phosphorus, magnesium, zinc).

AA diets can be very complex and need to be individualized. We strongly recommend seeing a registered dietitian/registered dietitian nutritionist every 6-12 months to help optimize the diet.

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