

SMA Dietary Needs from a Metabolic Specialist

"The question you ask is a simple mathematical problem that, unfortunately, most physicians are not taught to solve. The average 23-month old, non-SMA girl is 34 inches long and weighs 11.6 kg. For such an "average" child, muscle constitutes 40% of body weight, whereas for the average SMA-I child, muscle is at most only about 10% of body weight, and often only 5%. Therefore, the ideal body weight of a 23-month-old with SMA-I should be 35% less than 11.3 kg, or about 8 kg (17.5 pounds). However, because an SMA infant needs some extra fat to provide "padding" that normally is provided by muscle (which helps prevent pressure sores and other related complications), we usually allow a somewhat greater weight than this calculation gives. Thus, all other things being equal, Elizabeth should weigh perhaps 18 or 18-1/2 pounds. Another way of expressing this is to say that her weight should be about 3 or 4 standard deviations (Z-score) below her length. If Elizabeth's length is only 32 inches, her ideal weight might be closer to 17 or 17.5 pounds.

Similarly, because about 40 to 50% of caloric expenditure is from muscle metabolism, a child with SMA needs far fewer calories, often only 60% of that recommended for age. For nutrition recommendations, physicians are taught to go by the book. However, unfortunately, there is no nutrition book written for SMA, and dietary recommendations made using standard scales are just not appropriate. For example, when a child is very small for age (below the third percentile), as some SMA children are, physicians are taught to use the "weight-for-height" chart to specify an ideal weight for a child's size rather than age. However, again, the weight-for-height charts were developed for children whose body composition is normal or at least potentially normal when better nourished, which never is the case for SMA. Thus, the published weight-for-height charts are not valid for SMA and should never be used.

Although what I have written here explains the basic principles behind the special weight and nutrition goals for SMA, in practice I usually look only at the length chart to make sure a child's linear growth has been steady. If so, then the rest of my recommendations are based on what a child looks and feels like, not a number that I calculate. However, for physicians who are not familiar with SMA and muscle disorders of similar severity, the calculations I have presented usually help them approach the problem correctly and avoid the almost universal problem of overfeeding in SMA."