basic understanding of the disorder and practical suggestions regarding food selection and preparation.* Meal planning is based on weighing the food on a gram scale; a less accurate method is the exchange list. As soon as children are old enough, usually by early preschool, they should be involved in the daily calculation, menu planning, and formula preparation. Using a computer, voice-activated calculator, cards, or colored beads can help children keep track of the daily allowance of phenylalanine foods. A system of goal setting, self-monitoring, contracts, and rewards can promote compliance in adolescents.

Preparation of the phenylalanine-free formula can present some challenges. The formula tends to be lumpy; mixing the powder with a small amount of water to make a paste and then adding the rest of the required liquid, helps alleviate this problem. A blender or mixer dissolves the powder more easily; a rechargeable hand mixer can be used when traveling. Although the taste is virtually impossible to camouflage, many new products are on the market today. Some of the complete formulas are chocolate, vanilla, strawberry, and orange flavored. Incomplete formulas are also available that do not contain the vitamins and minerals and are plain tasting; these can be added to cold foods instead of mixing them as a formula. Formula bars are convenient for active adolescents. Formula capsules are also available, but the patient would need to take 20 or more capsules per day.

Family Support†

In addition to the problem related to a child with a chronic disorder (see Chapter 17), the parents have the burden of knowing that they are carriers of the defect. Genetic counseling is especially important to inform the parents that prenatal testing is now available to detect the presence of the defective gene in heterozygotes. Counseling is also important for adults with PKU to inform them that all of their offspring will be carriers for PKU (see Genetic Evaluation and Counseling).

Galactosemia

Galactosemia is a rare autosomal recessive disorder that results from various gene mutations leading to three distinct enzymatic