

more important in the treatment of CF, but careful monitoring for adverse effects (gastrointestinal bleeding) is essential.

Management of Gastrointestinal Problems

The principal treatment for pancreatic insufficiency is replacement of pancreatic enzymes, which are administered with meals and snacks to ensure that digestive enzymes are mixed with food in the duodenum. Enteric-coated products prevent the neutralization of enzymes by gastric acids, thus allowing activation to occur in the alkaline environment of the small bowel. The amount of enzymes depends on the severity of the insufficiency, the child's response to enzyme replacement, and the practitioner's philosophy. Usually 1 to 5 capsules are administered with a meal, and a smaller amount is taken with snacks. Capsules can be swallowed whole or taken apart and the contents sprinkled on a small amount of food, such as cereal or fruit, to be taken at the beginning of the meal. The amount of enzyme is adjusted to achieve normal growth and a decrease in the number of stools to one or two per day. Pancreatic enzymes should be taken before a meal or snack or within 30 minutes of eating. The enteric-coated beads should not be chewed or crushed because destroying the enteric coating can lead to inactivation of the enzymes and excoriation of oral mucosa. The powder form is used with infants and young children but should be used cautiously because inhalation of the powder may precipitate acute bronchospasm and, if mixed with food, predigests the food, making it unpalatable. The mouth must be rinsed after enzymes are administered to avoid break down of the oral mucosa or a breast-feeding mother's nipples.

Children with CF require a well-balanced, high-protein, high-caloric diet (because of their impaired intestinal absorption). In fact, they often require up to 150% of the recommended daily allowances to meet their needs for growth. Breastfeeding with enzyme supplementation should be continued as long as possible and, when necessary, supplemented with a higher-calorie-per-ounce formula. For formula-fed infants, commercial cow's milk-based formulas are usually adequate, although frequently a partially hydrolysed formula with medium-chain triglycerides (e.g., Pregestimil, Alimentum) may be recommended. Because the uptake of fat-soluble vitamins is decreased, water-miscible forms of