

Fat-Soluble Vitamins (Vitamins A, D, E, and K)

Vitamins A, D, E, and K require fat to be absorbed and, since CF patients have trouble digesting and absorbing fat, the bloodstream levels of these vitamins are often low in patients with CF. As part of their daily medical therapy, CF patients are put on a "standard CF dose" of vitamins, based on age. This dose is higher than that recommended for people without CF. It is important that the vitamins prescribed are taken daily to prevent deficiency of those vitamins. To make vitamins easier to absorb they can be taken with meals when oral enzymes and food are supplied. A physical examination and blood tests can then tell if more vitamins are required.

Some people may want to take more vitamins and minerals than have been prescribed, thinking that more may be better. Actually the body uses only a certain amount of each vitamin and mineral and a large excess cannot be used. For some vitamins (water-soluble vitamins), the extra amount just ends up in the urine (some public health experts who laugh at Americans' overzealous use of vitamins say that "Americans have the most nutritious urine in the world!"). The fat-soluble vitamins are not excreted in the urine, and taking an excessive amount can be dangerous.

Vitamin A

Vitamin A is needed for fighting infections, preventing night blindness, and general growth and maintenance of the body. Retinol and carotene are two types of vitamin A. Vitamin A is found in animal products, particularly liver, egg yolks, and fortified milk. Carotene is found in dark green and deep yellow and orange vegetables and fruits, and is converted to active vitamin A in the body. Vitamin A

is normally absorbed from the intestine and then stored in the liver to be used when it is needed. Two proteins made by the liver—prealbumin and retinol-binding protein—are needed to extract the vitamin A from the liver. Blood levels of vitamin A have been found to be below normal in some people with CF, even when supplemental enzymes and vitamins are given, but actual symptoms of vitamin A deficiency are rare. The abnormal fat digestion is part of the problem with vitamin A in CF, but even when vitamin A is absorbed and stored in the liver, it will not be available to the body if there are low levels of prealbumin and retinol-binding protein (two forms of albumin). Zinc is a mineral that helps retinol-binding protein extract vitamin A from the liver, so it is important that patients receive zinc in their multivitamins.

Vitamin D

Vitamin D is needed for growth of strong bones and teeth, and for normal functioning of many other organs. It is important for the absorption of calcium and phosphorus from the diet into the bloodstream. Vitamin D deficiency causes a bone disease called *rickets*, in which the bones are abnormally soft. Vitamin D is found in fortified milk and dairy products. It is also made in the skin by the action of sunlight. Vitamin D from the diet or skin must be activated by the liver and kidneys, which means that people with liver disease or kidney disease are more susceptible to vitamin D deficiency. Rickets or other evidence of vitamin D deficiency is rare in CF, particularly when patients are given oral enzymes and a multivitamin with vitamin D. If deficiency does occur, it can be detected by blood tests and treated with special supplemental vitamin and mineral preparations. Plenty of sunshine is also helpful to keep an adequate amount of vitamin D in the body.

Vitamin E

Vitamin E is important for the functioning of a number of important body parts, especially nerves. Good sources of vitamin E are vegetable oil, wheat germ, and dried beans and peas. Symptoms of vitamin E deficiency may include unsteadiness while walking. Vitamin E deficiency also causes an abnormal knee-jerk reflex, which the doctor checks by hitting the knee with the rubber reflex hammer.

Vitamin E deficiency can be detected by blood tests. This is the most common fat-soluble vitamin to be low in CF patients, mainly because people do not eat a lot of foods that are rich in vitamin E, and amounts of vitamin E tend to be low in standard multivitamin preparations. It is important for CF patients to take extra vitamin E daily. If blood tests indicate a deficiency, a higher dose of vitamin E can be prescribed. Water-soluble forms of vitamin E are more easily absorbed than the more expensive health food store preparations of vitamin E.

Vitamin K

Vitamin K is needed by the liver for making some of the clotting factors that stop bleeding. Green leafy vegetables and cauliflower are good food sources of vitamin K, and it is also found in dairy products. The average diet contains plenty of vitamin K. In addition, bacteria that normally live in the intestines make vitamin K, so for most people no vitamin K supplements are needed. People with CF may need extra vitamin K, though, because both dietary and intestinal sources of bacteria may be less than normal: first, vitamin K may not be well absorbed by the patient with CF. A second problem that someone with CF might have is that antibiotics given to treat infection in the lungs may kill the bacteria in the intestines, cutting down on the number of intestinal bacteria available to make vitamin K. This means that some people with CF who take a lot of antibiotics may need additional vitamin K. Vitamin K deficiency more often occurs during infancy (before diagnosis) or with the onset of CF liver disease.

Low blood levels of vitamin K can lead to very serious bleeding. There are two blood tests for clotting factors to estimate vitamin K levels. These are the "PT" test (prothrombin time) and the "PTT" test (partial thromboplastin time). The PT becomes abnormal if levels of vitamin K are too low, and in more severe deficiencies, the PTT may also become abnormal. Vitamin K deficiency can be corrected by adding oral or injected vitamin K supplements. Since the liver is needed to make the clotting factors, even vitamin K given by injection may not provide enough if the liver is failing to work (as in severe cirrhosis, discussed in Chapter 4). In this case, the already-made clotting factors may be administered by giving a transfusion of "fresh-frozen plasma."