



Tax deductions for food expenses

A person with CF may consume 2-3 times the quantity of food that a person without CF would eat. Some may require special foods which when consumed in large quantities, can represent a significant expense.

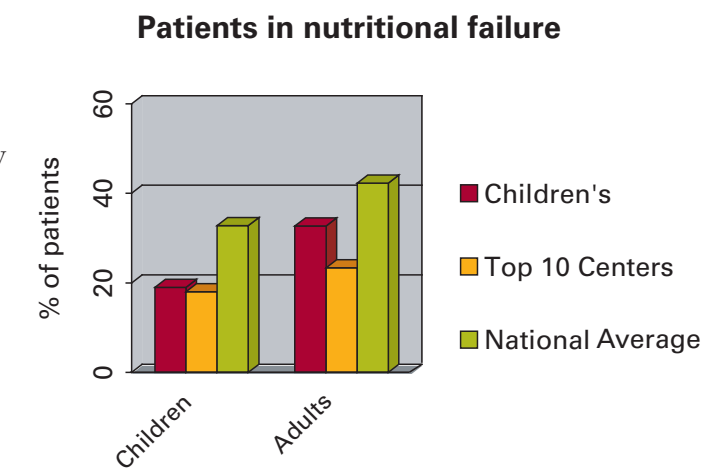
You may be able to claim as a medical deduction the amount of food that exceeds the costs for a person without CF. You will need to keep an accurate record of the amount, type and cost of food eaten. This will mean keeping store receipts.

For more information on how to save \$\$\$ on food expenses, log on to www.cff.org and type in tax tips and food expenses.

Our CF Center is making nutrition one of our top priorities. We are striving to improve how we look at nutritional issues in our clinic. As you know, nutrition plays a very important role in lung health.

Statistics from the 2003 CF Registry show that at our center 19% of children (< 18 years old) were in nutritional failure; 32% of adults are in nutritional failure. We can compare these numbers to the national average from all centers and from the top 10 centers in the country in each category.

As you can see in this graph, our CF Center Children's Hospital Boston is well below the national average for percent of patients in



nutritional failure in both age categories, but we feel that we have a lot of room for improvement. We ask that you partner with us to improve your nutritional status. Yearly appointments with the CF Center nutritionist, including diet recall to calculate the total number of calories consumed in a day, and having arm anthropometrics performed to determine fat stores, can help us tailor dietary interventions to help improve your weight and height.

We have covered a variety of topics in this newsletter to give you more information about nutrition and suggestions for improving your health. We hope you enjoy this issue of our CF News and learn something new.



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CF History

Thanks to a measure by the state legislature last year, children with cystic fibrosis will no longer stand out when it's time for lunch at school.

Led by State Representative Bob Coughlin of Dedham, Massachusetts state lawmakers approved a bill to allow students to self-administer the enzymes they need to properly digest food.

Coughlin, whose youngest son was diagnosed with CF in-utero, introduced the bill in January 2003 and it was passed into law in September 2004. The measure

means children with CF do not have to visit the nurse to take enzymes before snack or lunch times.

"It's important these kids have a chance to live as normal a life as possible," Coughlin said.

Enzyme supplements allow children with CF to absorb nourishment from food. Fourth, ninth and 10th graders testified in support of the bill at an Education Committee hearing in September 2003, saying the policy change would help them fit in with their peers.

The law is available online at:

www.mass.gov/legis/laws/seslaw04/sl040351.htm.

For more information contact:

Rep. Bob Coughlin at 617-722-2320.

Did you know that the goal of the CF Foundation is to have each child with CF achieve normal growth and development?

Amina Grunko, MS, RD, LDN, CF Dietitian

Good nutrition is essential for adequate growth and weight gain. People with cystic fibrosis (CF) should eat a diet high in calories and fat to help build strong lungs, preserve lung function and maintain breathing muscle strength. The body needs additional calories to fight off infection and also to maintain adequate growth. People with CF may need to eat 30-50% more calories than people without CF.

CF affects people differently. Approximately 85-90% of CF patients are in need of pancreatic enzymes. Enzymes are made by the pancreas to help digest food. In some people with CF, the ducts of the pancreas become clogged with thick, sticky mucus that blocks enzymes from reaching food. This lack of enzymes leads to poor digestion and absorption of nutrients, which in turn causes poor weight gain and growth.

Pancreatic enzymes are available to help people with CF digest the food that they eat. Each enzyme capsule contains small beads that dissolve in the small intestine and help break down food for absorption.

Enzymes should be taken just before eating. The number of capsules will likely increase over the years as the patient continues to grow. Several factors may affect the efficacy of enzymes including: the use of generic enzymes, crushing or chewing the beads, expired or bottles left opened, mixing of enzymes with non acidic foods such as milk or yogurt, and elapsed time between taking enzymes and eating. If you are unsure about any of your current practices involving provision of enzymes, ask your dietitian or health care provider. Patients in need of pancreatic

enzymes will need additional fat-soluble vitamins on a daily basis (vitamin A, E, D, and K), and it should be taken preferably with meals and enzymes.

If weight gain and adequate growth cannot be achieved despite eating a high calorie diet and pancreatic enzyme use, additional interventions may be needed. It is important for patients to follow-up with the CF dietitian to allow for early detection of other factors that may contribute to poor weight gain, including cystic fibrosis related diabetes.

Remember to visit the CF dietitian on a yearly basis, even if there are no problems with growth or weight gain. Other patients may need to see the dietitian more frequently for guidance and suggestions for high calorie meals.

Financial Assistance for Families

Paying for medications can be a major source of stress for many families. Programs do exist to help families pay for medications. Axcan Pharma™, the company that manufactures Ultrase® and ADEK®, offers two programs for families and patients with Cystic Fibrosis. Care First for CF, was developed for children less than two years old. This program provides free Ultrase enzymes and ADEK pediatric drops monthly, a free copy of Cystic Fibrosis: A Guide for Patient and Family educational book, and a free diaper bag. After registration in the program, a Pharmacy Benefit Card is sent out to the family. The card may be used at retail or mail order pharmacies and parents will not have to pay any out of pocket expenses for these medications. The second program offer is the Comprehensive Care Program for CF. Developed for patients over the age of two, families mail in proof-of-purchase of their monthly Ultrase® enzyme refill and then a 1-month supply of ADEK®, along with either 24 envelopes of Scandishake® or two 8-ounce canisters of Scandical® is shipped back to the family. Participants also receive a one-time certificate for a Flutter® mucus clearance device. The Comprehensive Care Program for CF does not have an upper age limit and patients may remain on this program for as long as they take Ultrase brand enzymes.

Solvay Pharmaceuticals, the makers of Creon®, offers the Wee Care™ program for children less than 2 years old. Once enrolled in the program, patients will receive free Creon® enzymes and Vitamax® pediatric drops through their second birthday through the CF Services Pharmacy. The Extra Helpings™ program offers a free month supply of Vitamax® when a proof of purchase of Creon® enzymes and a coupon for the program are mailed in.

Chiron Corporation sponsors two programs to help

patients and families increase access to Tobi®. The Tobi Access Hotline works with Priority Healthcare Corporation to review a patient's health insurance benefits and coverage verification. They also may assist with prior authorizations and denied claims. Patients or families that pay more than \$30/month for co-pays for Tobi® may find this program helpful. The Tobi Foundation is a non-profit organization that works with families who may be uninsured or underinsured to provide aid on a sliding scale based on eligibility.



The Genentech Endowment for Cystic Fibrosis was founded to increase access to Pulmozyme® for patients with cystic fibrosis. Patients and families may be eligible for one of the three programs that the Endowment offers: the Uninsured Patient Program, the Premium Assistance Program, and the Copayment Assistance Program.

Other drug companies offer programs to help families pay for medications. For more information you can look online at www.cfservicespharmacy.com and then click on Manufacturer Programs.

If you have any questions about these programs you may ask your health care provider in clinic or call the program hotlines directly.

Axcan Pharma (800) 472-2634.
Tobi Access Hotline (866) 598-8624
Tobi Foundation (877)-TOBI-4-CF.
Genentech Endowment for
Cystic Fibrosis (800) 297-5557
CF Services Pharmacy (800) 541-4959

Avocados

Did you know that avocados are a fruit and not a vegetable?

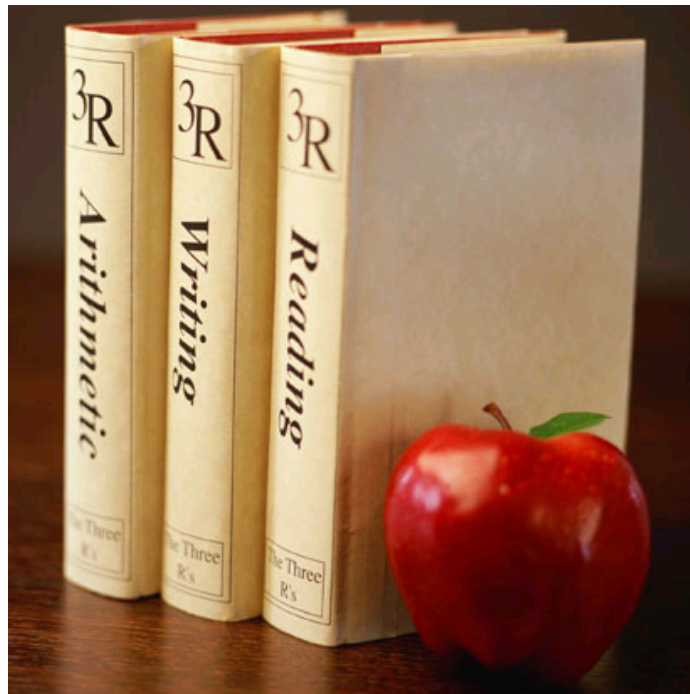
Avocados are cholesterol-free, sodium-free and low in saturated fat. They're a nutrient dense food that offers potassium, magnesium, folate, dietary fiber, riboflavin and vitamins C, E and B6.

One medium avocado contains approximately 300 calories and 30 grams of fat. Avocados are rich in omega-3-fatty acids, the heart healthy monosaturated fat which makes them a good food for growing children.

So if you have never tried an avocado, don't be afraid to do so. Buy the avocado when it is under-ripe, meaning it is firm, but not hard - squeezing it gently does not leave a dent. Store avocados at room temperature for three or four days until they are soft enough to dent on squeezing. To speed ripening, place the avocados in a paper bag and store at room temperature until they are ready to eat (one to three days). You can eat the avocado by itself or mash it up with fruits (bananas) to make them sweet or mix with tomatoes, garlic, onion, and lemon juice to make guacamole!

Preparing for Back-To-School

While now anticipating the lazy days of summer, we know that questions about planning for the upcoming school year will surface over the next few months. Several issues may arise as you plan for your own or your child's entry and/or return to school. Anticipating questions and concerns that you or your child may have about transitioning back to school and how these issues might be discussed and addressed by school staff can lead to an easier transition.



Communication between you and/or your family and the school is the critical first step. You may initially be unsure whether to tell school staff about your own or your child's condition or unsure about who should be informed (staff, other parents, classmates, etc). Parents can arrange to meet with their child's teacher and the school nurse before or at the start of school to talk about CF and their child's condition and what, if any, accommodations might need to be made during the school day (restroom privileges, permission to bring a water bottle, question of handling absences, etc). Parents have found it helpful to bring in written material for staff to learn more about CF. Parents also may consider having their child on an individualized education plan (IEP or 504 plan), which will spell out the

services and accommodations for that student. Specific accommodations may include establishing a plan to provide homework when the student is absent because of illness, allowing unlimited access to the bathroom, determining whether the student, teacher, or nurse holds enzymes, providing for students to eat snacks during the school day, etc. Parents often question when the right time to set up this plan is. Many parents decide that it is best to be prepared rather than to wait until their child is ill or needs to be in the hospital. Written material is available in clinic and Judy Bond LICSW and Lynne Helfand LICSW, the CF Center social workers, can assist you with this process.

Planning for college likewise may entail considering whether accommodations might be indicated and whether

contacting the college/university office for students with disabilities or health needs to assist in developing this plan might be helpful. Possible accommodations might include providing a specific type of dorm room, providing parking on campus, and planning to have notes taken for the student who is absent because of illness. For additional tips to consider, please read Joan Finnegan Brooks' article titled "Helping the Student with CF Prepare for College Life"

in the 2004 edition of *Homeline* (available online at www.CFServicesPharmacy.com).

Anticipating and planning with school staff around your or your child's health needs can ensure that the transition back to school will be as smooth as possible.

Amazing Gains

Ellen Schwanke

I was taken totally by surprise recently when my eleven-year-old daughter showed a significant weight gain after only one quarter.

Like many other CF patients, my daughter is a very picky eater. She was diagnosed at one year. The main reason her CF was not picked up until one year was she had good weight gain as a baby. While she does suffer from pancreatic insufficiency, she continues to this day to maintain her weight consistently at the 50th percentile.

Neither her pediatrician nor her CF doctor had ever raised a concern about her need to gain more weight. After all, with the rising population of obese children, the pediatricians these days are delighted to see a child hovering consistently at the 50% mark.

Earlier this year, our new CF doctor suggested we schedule a visit with the nutritionist. I must admit that, although we are diligent about PFT's, x-rays, blood work and the related important CF data gathering, nutrition was not a priority – we thought we were doing just fine. We also didn't think there were any achievable improvements. After all, our daughter was able to consistently maintain her weight with no problem, even when she was sick.

We agreed to meet with the nutritionist, and it was an interesting meeting. She commended me on my efforts to offer a wide variety to a picky eater who would sooner skip a meal than try something new. Truthfully, I thought – 'that was that'. But then she went on to explain that we could do even better. I was skeptical at first. Well, now I'm writing to share with other CF parents our success story. I would

never have believed we could achieve the results we have – and in such a short time. I owe it ALL to the visit with the nutritionist that day.

For our little girl, the answer is all in the milkshake. I was already serving up lots of high calorie ice cream anyway. What I learned that day has helped us to add another few hundred calories to my daughter's daily intake – effortlessly. By adding heavy cream to her milkshakes, and offering them up more than once a day, we can sneak in another few hundred calories without any fuss. And there were no meat or vegetables called into question! The results have astounded me and made me a believer in the importance of meeting with the nutritionist annually.

In short, I hope this quick story will convince all CF parents and patient to take the time to make that extra appointment. Good nutrition is a critical piece of the CF care pie. I was lulled into complacency at the 50th percentile for ten years and was convinced we couldn't do any better. I guess it's true what they say, "if you always do what you always did, you'll always get what you always got". Our gains have come only weeks after the appointment, my daughter bumped right up to the 75 percentile within the time span of one quarter.

It takes so much to provide all the care a CF patient needs; my hope is that you too may find some easy ways to enhance each meal as part of your care giving efforts.



Here's the recipe for the Schwanke Shake:

(Measurements are approximate)

1 cup whole milk ice cream

$\frac{3}{4}$ cup whole milk

$\frac{1}{4}$ cup heavy cream

3 tablespoons Hershey's syrup

$\frac{1}{2}$ packet Carnation instant breakfast powder

Mix it all together and blend to perfection.

The Use of Growth Hormone Therapy In Pre-Pubescent, Growth Restricted Children with Cystic Fibrosis

Tremendous progress has been made in the search for the ways to undo or get around the basic defect in CF. Thus, it is a very exciting time in CF research because nearly every month an important piece of the puzzle is discovered and new experimental treatments come to light. The prospects for even better treatment in the upcoming months and years are very bright. Having said this, I would like to take a few moments to talk about a large, multicenter study that involves treating children with CF who are growth restricted (slow growing) with Nutropin AQ, a recombinant human growth hormone.

Cystic fibrosis affects the digestion and absorption of nutrients because abnormal mucus blocks the ducts of the pancreas causing maldigestion (when the food which is eaten is not digested), and malabsorption (food is not very efficiently absorbed). This can cause many problems including: malnutrition, poor growth, and loss of fat and protein in the stool (steatorrhea and azotorrhea respectively). Consequently, to stave off malnutrition, an increased intake of calories, protein, fat, vitamins, and minerals is necessary along with pancreatic enzyme replacement.

Despite the emphasis on adequate nutritional intake and ad-

vances in the treatment of infections, malnutrition is a common occurrence. In adults, malnutrition first shows up as weight loss. In children, who should grow and gain weight steadily, a slowing down of the normal height and weight gain may be the first sign of malnutrition. Consequently, children with CF frequently remain short and underweight compared to their age-mates without CF. Studies have shown that poor linear growth is characteristic of CF, and is associated with a decline in lung function. Additionally, recent trials have demonstrated that nutrition alone may not be adequate to ensure maximal linear growth in children with CF.

Human growth hormone has been available for use in humans for more than 30 years. The use of growth hormone for the treatment of growth restriction has been met with great success in children with chronic diseases like growth hormone deficiency, Turner Syndrome, and chronic kidney disease. In fact, for these conditions it is now a standard of care. It was only natural then, that doctors would want to see if growth hormone could help short children with CF to grow. To date only a few studies involving a small number of children with CF have been conducted. This early research has demonstrated that giving hu-

man growth hormone, Nutropin AQ, to slow-growing children with CF can and does result in improvement in height and weight. One study also showed that those children treated with growth hormone had a significant improvement in their forced vital capacity (FVC, a measure of lung function) compared to their baseline, and the number of hospitalizations and the use of outpatient antibiotics also decreased. With these encouraging results, we are proceeding to a larger clinical trial to see if the results of these smaller studies hold true.

Currently we are participating in a Phase II trial of Nutropin AQ in prepubertal children with CF between the ages of 5 and 13. Approximately 100 children will be enrolled at 20 CF clinical sites around the country. Each subject will be randomized (assigned a group by chance, like flipping a coin) to receive either Nutropin AQ or no treatment (control group) for 12 months. The entire study lasts 18 months.

Please feel free to speak to your CF doctor or Dana Dorman, CF Research Coordinator at (617) 355-3197, if you have questions.

Database Studies	Cystic Fibrosis Foundation Registry- Open to all patients ESCF- Open to all patients Gene Modifier- Ages 8 and older Tissue Bank- Open to all patients Twin/Sibling-Twins and/or siblings with CF
Drug Studies	AI-005 (Aztreonam)- Ages 10 and older Inspire- Ages 8 and older Epic- Under age 13
Procedure Studies	AIM- Ages 10 and older Infant PFTs- Infants and toddlers
Nutrition Studies	DHA- Ages less than 8 weeks Growth Hormone- Ages 5-13
Upcoming Studies	TIP-003

• **Johns Hopkins University is conducting the CF Twin/Sibling Study.** The study will look at the clinical symptoms and medical history of twins and siblings and compare these results to a blood test. Parents as well as patients will be asked to give a blood sample. The study hopes to identify additional genes that affect the severity of CF. For more information, please contact Meredith Little at (617) 355-6665.

• **The Inspire study** is a follow up study to the previous Inspire study that our center participated in. The objectives of the study are to evaluate the safety, tolerability and efficacy of 28 days of treatment with INS37217 solution compared to placebo in patients with CF. This drug hopes to improve mucociliary clearance. Please contact Camille Puronen for more information at (617) 355-2093. A BIG THANKS to everyone who participated in the prior Inspire trial. Thanks to your help, researchers gained valuable information about this promising drug!

• **The Family Based Gene Modifier study** is designed to determine how genes other than the cystic fibrosis gene (CFTR) contribute to the severity or mildness of lung disease. This study uses a novel design that incorporates genetic analysis of parents and children. Patients must agree to provide a blood sample. Parents will also be asked to give a blood sample, but this is not necessary for patient participation. For more information contact Ian Huntington at (617) 355-1910.

• **Recently, a new technique, Infant Pulmonary Function Testing,** has been developed to measure pulmonary function in infants and young children. The purpose of this research study is to see how well this technique detects abnormalities in lung function in children with cystic fibrosis. 15 infants/toddlers will be enrolled in this study. Please contact Jennifer Treseler, RN at (617) 355-6077 for more information.

• **The AI-005 study** is a Phase III study assessing the safety and efficacy of an inhaled form of aztreonam lysinate for the treatment of Pseudomonas aeruginosa. Patients will be randomized to receive either 28 days of aztreonam for inhalation or placebo, following a course

of TOBI. Please contact Summer Adams at (617)-355-2446 for more information.

• **The AIM study** will look at using induced sputum as a tool to evaluate anti-inflammatory markers in patients with CF. Patients will take either ibuprofen or no treatment and perform two induced sputum procedures. Please contact Meredith Little at (617) 355-6665 for more information.

• **The DHA study** will compare the growth and nutrition of infants with cystic fibrosis on standard Enfamil formula vs. formula fortified with DHA (Docosahexaenoic Acid). Patients will be randomly assigned to a formula group and will receive free formula for the first year of life. Please contact Summer Adams at (617) 355-2446 for more information.

• **The Tissue Bank for Pulmonary Diseases** is designed to collect discarded clinical specimens (sputum, throat cultures, blood, etc.) for the various research projects of Children’s Hospital, Boston investigators. Please contact Meredith Little for more information at (617) 355-6665.

• **The Growth Hormone study** is a phase II study of the safety and efficacy of daily administration of growth hormone injections for treating slowed growth, improving growth and size (lean body mass) in children, and improving lung function in children with CF. Patients will be randomized to receive either growth hormone, or no treatment. For more information, please contact Dana Dorman at (617) 355-3197.

• **The EPIC clinical trial** is designed to find out which antibiotic regimens are most effective at eradicating new Pseudomonas aeruginosa infections. Patients will be randomized to one of four arms comparing different courses of TOBI and TOBI in conjunction with Cipro. Please contact Camille Puronen at (617) 355-2093.

• **Upcoming Research Update:** TIP-003 This study will look at effectiveness and distribution of a new powdered formulation of TOBI. We hope to begin enrolling for this study in the next year.