

Spring

PFIC.org Newsletter

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New Web-site for Liver Families

- [Http://www.Liverfamilies.net](http://www.Liverfamilies.net)

Offers Support, Knowledge and Hope to an international online community of families whose lives have been touched by pediatric liver disease and transplant.

Shadow Buddies

My daughter, Anna, was recently given a liver transplant shadow buddy doll as a gift. The doll was darling with blond curly hair and heart shaped eyes, but most importantly a scar that matched Anna's transplant scar. Anna named the doll Annie and while she plays with Annie, the doll's most important role seems to be when Anna is sick. Anna will show us what is wrong using her doll. I thought the dolls were such a good idea that I went on the internet to learn more. Here is the Shadow Buddy story.

Miles Postlethwait has been in and out of the hospital since he was born with multiple congenital defects. During one hospital stay, the Olathe boy told his parents he wished he had a friend "just like me," someone who had similar defects and, more importantly, someone who could share his feelings of loneliness, fear and frustration. Miles' mother, Marty Postlethwait, decided to find that little friend for her son and other children who spend a lot of time in the hospital. She did research and later developed Shadow Buddies, a line of dolls that are customized to have the same illness as that of the child who receives it.



method of educating children and their families about a particular disease, or medical condition and its treatment. Shadow Buddies are a unique form of medical play therapy. The Buddies remain with the child throughout their treatment, providing long-term psychological support. Each Buddy has a smile that children can always see when they need one and view themselves in a positive manner. We strive to provide as many smiles as possible.

Some hospitals give out the Shadow Buddies to their kids or the buddies may be purchased from the Shadow Buddies website. They are unique in that you can choose the skin, hair, eye color and medical condition that matches your child's. The picture to the left shows the Liver Transplant Shadow Buddies and to the right are Ostomy Bag Shadow Buddies. Each Buddy is 12" in length, weighs approximately 10 oz. and is very huggable! All of the Buddies have heart-shaped eyes for love, and wear a big smile -- belly buttons are included, and each Buddy arrives with a brightly colored hospital gown.

For more information visit: www.Shadowbuddies.com

Shadow Buddies are condition-specific dolls designed to be a friend, "just like me." These buddies provide physicians a hands-on

Tax Deductions for Medical and Dental Expenses By Gene Gregorio CPA

General Information

If you itemize your deductions on Form 1040 Schedule A, you may be able to deduct expenses you paid that year for medical care (including dental) for yourself, your spouse, and your dependents. A deduction is allowed only for expenses paid for the prevention or alleviation of a physical or mental defect or illness. Medical care expenses include payments for the diagnosis, cure, mitigation, treatment, or prevention of disease, or treatment affecting any structure or function of the body. The cost of drugs is deductible only for drugs that require a prescription, except for insulin.

Deductible Fees

Medical expenses include fees paid to doctors, dentists, surgeons, chiropractors, psychiatrists, psychologists, and Christian Science practitioners. Also included are payments for hospital services, qualified long-term care services, nursing services, and laboratory fees. Payments for acupuncture treatments or inpatient treatment at a center for alcohol or drug addiction are also deductible medical expenses. You may include amounts you paid for participating in a smoking-cessation program and for drugs prescribed to alleviate nicotine withdrawal.

Deductible Items

You may deduct the cost of:

- participation in a weight-loss program for a specific disease or diseases, including obesity, diagnosed by a physician.
- false teeth
- prescription eyeglasses or contact lenses
- laser eye surgery
- hearing aids
- crutches
- wheelchairs
- guide dogs for the blind or deaf
- admission and transportation to a medical conference relating to the chronic disease of either yourself, your spouse, or your dependent (if the costs are primarily for and essential to the medical care)

Non-Deductible Items

You may not deduct:

- the cost of purchasing diet food items.
- the costs for meals and lodging while attending the above mentioned medical conference.
- amounts paid for nicotine gum and nicotine patches, which do not require a prescription.
- funeral or burial expenses
- health club dues
- over-the-counter medicines, toothpaste, toiletries, cosmetics, a trip or program for the general improvement of your health, or most cosmetic surgery.

Transportation Expenses

Transportation costs primarily for and essential to medical care qualify as medical expenses, the actual fare for a taxi, bus, train, or ambulance is deductible. If you use your car for medical transportation, you can deduct actual out-of-pocket expenses such as gas and oil, or you can deduct the standard mileage rate for medical expenses. With either method you may include tolls and parking fees.

Lodging and Meals

You may include in medical expenses the incidental cost of meals and lodging charged by the hospital or similar institution if your main reason for being there is to receive medical care.

Qualifying Expenses

You can only include the medical expenses you paid during the year, regardless of when the services were provided. Your total medical expenses for the year must be reduced by any reimbursement. It makes no difference if you receive the reimbursement or if it is paid directly to the doctor or hospital.

You may include qualified medical expenses you pay for yourself, your spouse, and your dependents, including a person you claim as a dependent under a multiple support agreement. If either parent claims a child as a dependent under the rules for divorced or separated parents, each parent may deduct the medical expenses he or she actually pays for the child. You can also deduct medical expenses you paid for someone who would have qualified as your dependent except that the person didn't meet the gross income or joint return test.

Calculating Medical and Dental Expenses

You may deduct only the amount by which your total medical care expenses for the year exceed 7.5% of your adjusted gross income. You do this calculation on Form 1040 Schedule A in computing the amount deductible.

Insurance Premiums Considered as Medical Expenses Include:

- insurance premiums paid for accident and health
- qualified long-term care insurance.

You may **not deduct** insurance premiums for life insurance, for policies providing for loss of wages because of illness or injury, or policies that pay you a guaranteed amount each week for a sickness. In addition, the deduction for a qualified long-term care insurance policy's premium is limited. You may not deduct insurance premiums paid by an employer-sponsored health insurance plan (cafeteria plan) unless the premiums are included in Box 1 of your Form W-2.

If you are self-employed and have a net profit for the year, or if you are a partner in a partnership or a shareholder in an S corporation, you may be able to deduct, as an adjustment to income, 100% of the amount you pay for medical insurance for yourself and your spouse and dependents. You can include the remaining premiums with your other medical expenses as an itemized deduction. You cannot take the special 100% deduction for any month in which you are eligible to participate in any subsidized health plan maintained by your employer or your spouse's employer.

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Happy Birthday Cade Charles and Anna



Please help us celebrate Cade Charles and Anna's birthdays. Cade Charles birthday is April 15. He will be 2 years old. Anna's birthday is May 24. She will be 4 years old. In her picture is Dr Saul Karpen, Texas Children's Hospital.



Surgical Treatment of PFIC

By Benjamin Shneider, MD
Professor of Pediatrics
Chief, Division of Pediatric Hepatology
Mount Sinai School of Medicine

One of the most apparent and debilitating problems associated with progressive familial intrahepatic cholestasis (PFIC) is pruritus (itching). This relatively constant problem has tremendously deleterious effects on children and their families. Child behavior and family dynamics are understandably and often adversely affected. Traditional remedies for itching associated with other diseases are typically ineffective. For instance, anti-histamines that are utilized and effective for allergy-related itching are rarely useful in the long-term for PFIC. Anti-histamines often sedate children and may facilitate going to sleep, although the effect may be transient. A variety of other medications has been tried as treatment for pruritus in PFIC, although most are not effective in the long run. The mechanisms that underlie the pruritus in PFIC and other forms of cholestatic liver disease remain unclear. Pruritus is often seen in individuals with high levels of bile acids in their blood, leading some to believe that bile acids are part of the pathogenesis of the pruritus.

Bile acids are chemicals that are made by the liver from cholesterol. The bile acids are secreted by the liver and stored in the gallbladder. After a meal, the gallbladder contracts and the bile acids are expelled into the first portion of the small intestine. In the small intestine they mix with juices from the pancreas and facilitate the absorption of fats and fat-soluble vitamins (vitamins A, D, E and K). Most (95%) of the bile acids that are secreted by the liver are reabsorbed in the last portion of the small intestine (terminal ileum). The bile acids that are reabsorbed cycle back to the liver through the bloodstream so that the bile acids can be conserved and reutilized. The cycle of bile acids going from the liver to the terminal ileum and back to the liver is referred to as the enterohepatic circulation of bile acids.

Interrupting the enterohepatic circulation of bile acids has the potential to reduce the total amount of bile acids in an individual's body and could be useful as a treatment for the pruritus associated with certain types of liver disease. At present, there are primarily two surgical means by which interruption of the enterohepatic circulation of bile acid can be achieved – namely partial external biliary diversion and partial ileal exclusion.

Partial external biliary diversion (PEBD) is the surgical approach most commonly utilized for PFIC and another cholestatic liver disease called Alagille syndrome. This technique has been used successfully for over 15 years. The results can be quite dramatic and rewarding. In children who respond well to this procedure, pruritus diminishes markedly and often disappears completely. Blood tests of liver injury often improve and liver histology (examination of liver tissue under a microscope) stabilizes or may improve. PEBD involves sewing a small piece of intestine between the gallbladder and the abdominal wall. A small hole is made in the gallbladder and one end of the intestine is attached at that opening. The other end of the intestine is attached to a hole in the abdominal wall, called an ostomy. This ostomy allows contents within the segment of intestine to drain externally, typically into a small bag that is secured to the abdominal wall. Therefore after completion of the external biliary diversion, bile in the gallbladder has two routes of excretion. Approximately 30 to 50% of the bile goes into the diversion and drains out the ostomy. This bile is discarded each day. The remainder of the bile is excreted into the intestine following normal pathway through the bile ducts. This surgical procedure is well tolerated and relatively safe in the hands of a surgeon experienced with these kinds of procedures.

An alternative surgical approach is ileal exclusion. There is a limited experience with the use of this procedure for children with PFIC. Not all clinicians are convinced that it is as effective as PEBD, although success with this procedure has been reported at some centers including our own. This procedure has been performed successfully in over 400 adults as a means of treating elevated cholesterol levels. The procedure involves bypassing the last 15% of the small intestine. The first step of this procedure involves cutting the intestine at a point that divides the intestine into the proximal (upper) 85% and distal (lower) 15%. The proximal intestine is sewn to the colon and the distal intestine is closed off. Thus the distal 15% of the intestine is no longer in contact with the flow of intestinal contents and is bypassed. Bypassing the distal 15% of the small intestine reduces the ability of the intestine to reabsorb bile salts. Those bile salts pass into the colon and are excreted with stool. The distal intestine remains in the abdominal cavity and can be reutilized in the future if necessary. The major advantage of this procedure is that it does not require an ostomy. The success of the procedure may be dependent on the amount of intestine that is excluded. Excluding too much may lead to significant diarrhea, while excluding too little may not be effective.

Surgical Treatment of PFIC- continued

At present, it is uncertain as to whether these procedures are equally effective for PFIC due to FIC1 disease (Byler's disease = PFIC1) or due to BSEP disease (PFIC2). My own personal bias is that they are more effective for PFIC1 versus PFIC2, although milder PFIC2 disease may be very responsive to these surgical interventions. On-going investigations are attempting to answer this important question. Continued medical follow-up in children who undergo either of these surgical procedures is important even if there is complete resolution of the pruritus. Liver disease can still progress and nutritional problems related to vitamin deficiencies are still possible.

An alternative surgical procedure for PFIC is liver transplantation. A full discussion of liver transplantation is beyond the scope of this review. Suffice it to say, liver transplantation is associated with significantly greater potential morbidity and mortality than either PEBD or ileal exclusion. Both PEBD and ileal exclusion can be reversed if liver transplantation is required. The surgical procedures described above may be difficult and ill-advised for a child with cirrhosis, and liver transplantation may be essential in these circumstances. PFIC2 is a disease that appears to specifically affect the liver and liver transplantation corrects all of the problems associated with PFIC2. In contrast, PFIC1 is potentially a more systemic disease. A variety of medical problems may arise after liver transplantation for children with PFIC1. Therefore, in our program, we strongly encourage the use of either PEBD or ileal exclusion in children who we suspect have PFIC1. Decision-making for presumed PFIC2 may be more complicated, although a trial of PEBD may be very reasonable. Recent studies have revealed that PFIC2 may be associated with an increased risk of liver tumors. Therefore, screening for liver tumors needs to be conducted in children with PFIC2 who have had either successful PEBD or ileal exclusion.

Programs for Buying Peptamen Jr

Peptamen Junior is a peptide-based elemental diet for the nutritional needs for GI-compromised children. Sometimes doctors recommend this product, but it is very expensive and not always covered by insurance. Recently I found two programs that may help families buy this product.

Nestle Homelink, 1-800-776-5446, sells **Peptamen Jr** directly to you and offers free home delivery with a 3-to-5 day delivery window.

Nestle Nutrition products has also established a financial assistance program for individuals based on medical and financial need. For information on that program you can contact them through the web-site

www.nestleclinicalnutrition.com

or write:

Nestle Nutrition Products
Financial Assistance Coordinator
Nestle Nutrition
3 Parkway North, Suite 500
Deerfield, IL 60015

Kids Indoor Fun

Here are some fun and free webpages full of games and stories for those days spent inside.

See the animals live at the National Zoo's web-cameras.
<http://nationalzoo.si.edu/Animals/WebCams/>

Play games with Arthur, Dragon Tails, Barney at PBS Kids.
<http://pbskids.org>

Visit Sesame Street Workshop.
<http://www.sesameworkshop.org/sesamestreet/>

Play games with your favorite Disney character at Disney's Playhouse.
<http://disney.go.com/home/today/index.html>

Play animal games with The Little People of Fisher Price On-line.
http://www.fisher-price.com/fp.aspx?st=10&e=gamesLanding&mcat=game_infant,game_toddler,game_preschool&site=us

Visit Seussville with Cat in the Hat.
<http://www.seussville.com/catinthehat/>

Do jigsaw puzzles with Winnie the Pooh.
<http://www.dltk-kids.com/crafts/cartoons/winniethepooh.html>

Play with everyone's favorite monkey, Curious George.
<http://www.houghtonmifflinbooks.com/features/cgsite/>

Play the vegetable game with Peter Rabbit.
<http://www.peterrabbit.co.uk/>

Save the Popcorn Factory at Tonka Trucks.
<http://www.hasbro.com/tonka/pl/page.da/dn/default.cfm>

Play games with Mr. Potatohead, My Little Pony, and Candyland at Hasbro.
<http://www.hasbro.com/monkeybar/>

Make music and games with Noggin.
<http://www.noggin.com/>



Sharing Talent

Our web-site is simple and so is our newsletter. I am not talented when it comes to graphics layout. If you have a talent you would like to share, in web design, with the newsletter design, or in any other area please e-mail me.

Robin

PFIC LIVE CHAT

I thought we might have our first online Chat, April 3rd Monday at 7:00pm Central time zone. Come share your concerns, inspirations, and stories. Hope to see you there.

Below Cade Charles gives us a two binkie solute.

