

# *Cystinuria Support Network News*

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The *Cystinuria Support Network*, a Non-Profit Mutual-Aid Support Network, has been developed to provide a resource for putting individuals in touch with each other for support and practical advice. Since no one can understand the issues like those who are sharing the same experience, this Network will allow us to come together with our strengths, hopes and concerns to offer support and understanding to each other.

The Cystinuria Support Network in no way endorses any drug or treatment that is reported herein and should not be regarded as a substitute for personal, professional medical advice. It is our wish only to provide information and encourage you to always check any treatment with your physician.

## **From the Editor**

This is the third newsletter to be published by the Cystinuria Support Network. Since the last publication we have started work on a UK based support network and we have gone on-line with the Cystinuria Listserv (see the article Cystinuria in Cyberspace for more details)

As always we couldn't have put this newsletter together without the input and help of the doctors and many thanks to David Goldfarb who has contributed two articles this time.

We hope you are all well and keeping up with the fluids. May more of us be stone free.

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# Stone Soup

A column containing bits and pieces of information, questions, anecdotes and ideas for discussion.



## If it wasn't for cystinuria!

Two years ago, after I graduated from graduate school, I decided to go to Israel for a year to live.

I had to delay my trip for a month in order to have a perc. and remove 5 stones from my right kidney. A week after the surgery, thinking I was "stone free" for the first time in my life, I headed off to Israel feeling great. Less than a week later, I developed another stone, this time in my left kidney. I was caught completely off guard. It was so demoralizing, so frustrating.

I stayed in Israel a few months, but when the pain got unbearable, I returned to the states (to my parents house in Maryland) for treatment. My doctor at the time told me I had developed around 9 stones in my right kidney in addition to the one still in my left, and he strongly advised me to stay in the country.

I had no job, no apartment, no plans, and no idea what I was going to do with myself. It was probably the most depressing time of my entire life.

I decided to go back to New York to visit friends and figure out my life. My first night back in New York, I went out to dinner with some friends, and sitting across the table from me was the nicest, sweetest, cutest most unbelievable guy I had ever met.

Because of my stones, I had all the time in the world to get to know this person and develop a relationship. Five weeks from now, we will be married!

Had it not been for cystinuria, I would have been half-way across the world and I never would have met Neil. For once I am grateful.

Marni

## Use the BRAN Principle

During yet another visit to the hospital for renal colic, a doctor asked whether he could do a rectal examination. Apart from not relishing the idea of having a finger put where it shouldn't, the thought of lying still was impossible. Fortunately my sister was there and asked whether this examination was vital to his diagnosis. The intern explained that although it wasn't necessary, it was something he 'liked' to do.

He was told in no uncertain terms that if it wasn't necessary it wasn't going to be done. We have now adopted the BRAN principle to all procedures we are asked to undergo.

- |             |  |
|-------------|--|
| B Benefits  | What are the benefits of doing this procedure? |
| R Risks     | What are the risks involved?                   |
| A Actions   | What do they have to do?                       |
| N Necessity | What would happen if it wasn't done?           |

If we are convinced that all this information makes the procedure worthwhile we will allow it to be done.

We believe this cuts down, not only the amount of defensive medicine practiced on us, but also procedures where the humiliation etc outweighs the benefits

Sue

## Ask Another Cystinuric

**Has anyone got any ideas for making stones pass more quickly. I have two stones on each side and the usual stuff doesn't seem to be working. I really don't want to go into hospital!**

Laura

Drink a pot of Oolong Tea. You will have never peed so much in your life. Keep drinking till you pass something. When ever I drink Oolong tea I always seem to pass something. Plus it's that really yummy kind of tea you get at chinese restaurants. Probably doesn't help that I add a pound of sugar :)

Yara



## Ya Gotta Laugh

I was once admitted into hospital with really bad renal colic.

The next day, after the pain had subsided an X-ray was taken. A few hours later I saw two young doctors looking at my X-rays. They beckoned me over, and both with very grave looks on their faces told me that they had some very bad news! They said that I had a 2 inch staghorn calculi in my left kidney.

I couldn't believe it as I had only had an ultrasound a month before which declared me clear of stones.

I asked to see the X-ray. I couldn't see any stones! I asked them to point the stone out. They pointed to a light band sloping across the kidney. 'Surely they didn't mean that ???' I thought to myself.

'Are you sure?' I said. One nodded and looked at me with great sympathy!! I looked at the X-Ray again. That did not look like a stone, it looked more like a rib! No, it was a rib!

I said 'Are you sure it's not a rib?' They looked at each other - then realisation struck!

This taught me one important lesson.

**NEVER ACCEPT ANYTHING ANYONE SAYS ABOUT YOUR HEALTH WITHOUT CHECKING IT OUT FOR YOURSELF !**

Pete

When I was about 8 years old I unknowingly had a stone the size of a golfball. My mother took me into the ER kicking and screaming. After puking all over the lobby they took me into a private room.

The doctor asked what it felt like. I told him it was a like a herd of Elephants stampeding across my kidney.

The doctor tried so hard not to laugh. He just couldn't take it and started laughing. Everyone else in the room had a good laugh too.

At the time it wasn't very funny to me but they sure thought it was. Now I think it was hysterically funny. You sure get some strange responses out of children.

Yara

## Urinary Alkalization

By David S. Goldfarb, M.D.  
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Center

Alkalinization of the urine is important in cystinuria because it increases the solubility of cystine, meaning that more cystine can be dissolved in a given amount of urine. Alkalinization means neutralizing the acid in the urine by adding base. When acid is neutralized there are fewer H<sup>+</sup> molecules (also called protons) and the pH rises. pH is a measure of the amount of acid in the urine. Human urine can have pH ranging from about 4 (acid) to about 8 (alkaline). When urine pH rises above 7, cystine becomes much more soluble, so achieving a urine pH of 7.5-8 for a good part of the day is desirable. Measuring and recording your urine pH at various times of the day is very helpful to you and your doctor to show whether you are getting to the desired range.

You can alkalinize your urine by decreasing the amount of acid you take in. You can lower the amount of acid you eat (and therefore the amount of acid your kidneys have to get rid of) by eating less animal protein. Protein is what muscle is made of, and includes fish, beef, chicken and pork. These products also contain some cystine, so limiting your intake of these has 2 benefits.

You can also take in more base to alkalinize your urine. If you eat more fruits and vegetables when you reduce your protein intake, you will take in more base. Base comes in the form of molecules called "organic anions", such as citrate and malate. They are converted to bicarbonate by the liver. Bicarbonate is the blood's form of base. One citrate is converted to 3 bicarbonates. So taking citrate and bicarbonate are equivalent. Some of the citrate also is found in the urine where it helps prevent calcium stone formation in non-cystinuric people with the more commonly found calcium oxalate stones. Citrus fruits like oranges and lemons and all fruits and vegetables contain these organic anions.

For most people adequate alkalinization does not occur without taking in extra base. It comes in many preparations. Potassium (K) citrate is preferable to sodium citrate preparations because sodium may increase cystine excretion. This is also why I don't usually prescribe baking soda, which is sodium bicarbonate. But the alkalinizing effect, if it works, could override the increase in cystine excretion. If you are doing well with sodium preparations I would not change your prescription.

The major reason why I sometimes prescribe sodium citrate instead of potassium citrate is if there's too much potassium in the blood, which is rarely a problem in young people with normal overall levels of kidney function. Another reason to use sodium citrate is taste. Some people prefer it. A third reason is gastrointestinal

tolerance. Some people find that potassium citrate causes heartburn, or diarrhea, or abdominal cramps. These are not usually serious side effects but can be avoided by changing preparations.

Sodium bicarbonate comes as baking soda and in pill form. Sodium citrate can be taken as Bicitra, Shoal's solution. Polycitra (NOT the same thing as Polycitra-K!) has both sodium citrate and potassium citrate in it. All three contain sodium citrate and citric acid. Why is it OK to take citric acid if you are trying to avoid acid? Because the citric acid provides both base (citrate) AND acid, which neutralize each other. It has no net effect on urine pH, unlike the citrate in food which has only the base part, not the proton (H<sup>+</sup>) part. Why is it there then? To help dissolve the sodium citrate.

Potassium citrate comes in various preparations. Polycitra-K comes as a liquid and in crystals (packets) that you mix in water. It comes in several flavors which are worth trying. In either case they can be sufficiently diluted or mixed into other juices to minimize the taste. Another option is K-Lyte which comes as an effervescent tablet that dissolves in water, like an Alka-Seltzer. It also comes in different flavors worth trying on your kids. It's a combination of potassium citrate and potassium bicarbonate; that's OK because citrate and bicarbonate are equivalent. It also comes as "DS" or double strength. (You DON'T want K-Lyte/Cl which is potassium chloride and has no alkalinizing property). Another popular form of potassium citrate is Urocit-K, a pill form. They are actually in a wax matrix from which the drug dissolves. People often see the unabsorbed, undissolved wax in their bowel movements; this does not mean the mineral is not being absorbed.

Compare doses of these preparations in milliequivalents (mEq) of bicarbonate equivalents; ignore the number of milligrams. Most people need anywhere from 20 to 120 mEq per day, but measuring the urine pH is the way to determine how much you need. Bicitra and Shoal's solution are 15 mEq per tablespoon (1 tbsp=15 cc, cubic centimeters), or 1 mEq per cc. Polycitra liquid has 2 mEq per cc (half as sodium, half as potassium). Polycitra-K liquid is 2 mEq per cc, all potassium. Polycitra-K crystals come as 30 mEq per packet. Urocit-K comes in 5 and 10 mEq tablets. K-Lyte comes as 25 mEq per tab, and 50 mEq for the "double-strength" DS. The standard generic sodium bicarbonate tab (325 milligrams, like an adult aspirin) is about 4 mEq.

**I know that people often hesitate when a doctor reaches for a prescription pad. I tell my patients that potassium citrate is more like a vitamin, not a drug. Potassium and citrate are in all of your cells, and all the fruits and vegetables you eat. Both are normally found in urine in significant amounts because we take in more than we need. You can't be allergic to these minerals, though rarely people are allergic to dyes in the preparations. If your blood potassium is in the normal range you should not have a problem: the extra potassium is excreted by the kidneys. The occasional heartburn or other GI symptoms can usually be overcome by taking them with meals, which doesn't diminish their absorption or effect on the urine. Sodium**

**citrate or bicarbonate may be a problem for people with decreased heart function, kidney function, or high blood pressure, and can increase urinary cystine levels, but like eating salty pretzels should not cause problems for most otherwise healthy people. I wouldn't be concerned about taking these "supplements" or about giving them to children. I view these medications as safe and effective, though inexplicably expensive.**

## Cystinuria in Cyberspace

by Philip M

If you're reading this, you already know (or are about to discover) what a great resource the Cystinuria Support Network can be.

Now there's a great way for you to be in touch with other members of the network every day: the cystinuria listserv.

Back in early 1987, support network member Michelle Culbertson had the bright idea of setting up an e-mail loop as a way to stay in touch. It was primitive, but effective: members sent e-mail to each other by using the "Reply All" function — so that each time a message was sent, it went to everybody who was part of the loop.

Then, in spring 1998, Keith Swenson, whose wife has cystinuria, set up an honest-to-goodness listserv. The way it works is simple: you sign onto the list (see details below); a message posted to the list goes to a computer which then immediately distributes it to everyone subscribed to the list. The list differs from the loop (though some people still call it "the loop!") in that replying to a message simply involves replying to the list address. And you can easily sign on or sign off anytime you want.

Since so many of us who have cystinuria (or live with cystinuric family members) rarely if ever meet others with the disease, the list is a superb way to keep up-to-date, find support, and pick the brains of people who are knowledgeable about cystinuria (including four doctors who are experts at treating cystinuric patients).

Recent topics of discussion include the odds of developing cystinuria; dealing with very real pain when tests turn up nothing; the challenges faced in school by cystinuric children; the pros and cons of various types of pain medication; vegetarian recipes; and tips for passing stones and dealing with the pain they cause.

Keith, who is apparently shouldering the cost of running the list himself (thanks Keith!), has also set up an archive that anyone subscribed to the list can access. The archive includes every message ever sent to the list, and it's a great resource. If you need to find out more about polycitrac, and you vaguely remember

a discussion about it on the list, you can head over to the archives and pull up the whole thing.

To get onto the cystinuria list, send e-mail with subscribe cystinuria in the body text to: listserv@home.ease.lsoft.com. Once you've signed on, you'll get a welcome message that should answer any questions you may have about using the list — including how to sign yourself off.

To use the archives, go to:

<http://home.ease.lsoft.com/archives/cystinuria>

log in, and conduct your search! Please note that you must be a member of the list in order to search the archives.

With 75 members, the list has become a friendly cyber-place, packed with information and support. It provides important information, helpful messages to members who are suffering, and provides a potent reminder to keep drinking!

Edit 2006: In 2005 the CSN Support Group moved from ListServ to Yahoo Groups. We have over 350 members, still going strong.

## Cystinuria and Transplantation

By David S Goldfarb, M.D.

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There is a web-site journal called HDCN you probably know: hypertension, dialysis, clinical nephrology, run by John Daugirdas, a very prominent dialysis expert. Most of the content I believe is limited to health professionals. I'm on the editorial board, and review stone disease especially for John.

There's an "ask the professor" feature, questions from other nephrologists, and recently a transplant surgeon asked whether cystinuria recurs after transplantation. I took the occasion to discuss some aspects of the disorder, then answered the question. The response is below.

### Cystinuria

The question asked is whether cystinuria can recur after renal transplantation. I'll review the basic pathophysiology with an update on recent investigations of the defective cystine transporter, and then examine this issue.

Cystinuria is an autosomal recessive disorder of transepithelial transport of cystine and other dibasic amino acids. Cystine is relatively insoluble, and its presence in the tubular lumen at concentrations of

more than 250 mg/l is associated with precipitation and stone formation. Treatment requires increasing urinary volumes to keep cystine concentrations at or below 250 mg/l, urinary alkalization with potassium citrate, restriction of dietary sodium, and reduction of cystine to the soluble cysteine with penicillamine, alpha-mercaptopropionylglycine.

The abnormal gene was mapped via linkage studies to human chromosome 2p (1). At the same time, the mutated gene was demonstrated to be rBAT (basic amino acid transporter) (2), a cystine transporter previously identified in proximal tubular membrane vesicles from humans, rats and rabbits. This transporter mediates sodium-independent, electrogenic, apical membrane uptake of cystine into the cells of the proximal straight tubule (S3). It is also present in the apical brush border membranes of the jejunum where it mediates absorption of cystine.

When expressed in *Xenopus* oocytes, this transporter also mediates transport of dibasic amino-acids (lysine, ornithine, arginine) and some neutral amino-acids as well (11). This high-affinity process is augmented by a low-affinity process in the proximal convoluted tubule (S1), the mediator of which is not yet identified. After uptake across the apical membrane, cystine, essentially a cysteine dimer, is reduced intracellularly to cysteine which exits the cell across the basolateral membrane.

Studies by Harris (3) and Rosenberg (4) suggested that there were 3 phenotypes of cystinuria.

Type I, the most severe form, appears to be caused most frequently by a mutation of rBAT residue 467 from methionine to threonine. This mutation, called M467T, accounted for 40% of the abnormal chromosomes in the Spanish cohort studied by Calonge et al (2), and 30% of the abnormal chromosomes studied in the entire group (n=36) from Spain and Italy. Heterozygotes for Type I have normal urinary levels of cystine and other amino-acids.

Type II patients have impaired in-vitro intestinal transport of lysine, but cystine transport is present in the homozygote. Type II heterozygotes have increased urinary levels of cystine, ornithine, arginine, and lysine.

Type III patients have some intestinal cystine absorption, and can partially absorb an oral load. Type III heterozygotes also have moderately increased urinary amino-acid excretion.

The molecular correlates for Type II and Type III have not yet been described but presumably represent the manifestations of other mutations in the rBAT gene, or in some cases, combinations of M467T with other abnormal alleles.

In answer to the question about renal transplantation, one would not expect cystinuria to recur after cadaveric renal transplantation since the renal transport of cystine in the graft would be expected to be normal. Intestinal absorption of cystine would be absent in Type I patients and impaired in most Type II and III patients, so cystinuria would not occur. There are no demonstrations of other metabolic abnormalities of

proven clinical significance associated with failure of cystine transport.

Deficiency of other amino-acids, like lysine, are not limiting, as their absorption as constituents of oligo-peptides is not impaired (5). One letter describes a patient who received a living-related transplant, though it fails to describe the relationship of the donor to the recipient (6). More than 3 years later, urinary amino-acid levels were normal, with cystine excretion of 37æmol/24 hours, and no recurrence of nephrolithiasis. This letter cites an article purporting to have 3 cases of renal transplantation; in fact, my review of this article finds no mention of transplantation in it (7)!

Another report of stones in renal transplant recipients notes no cases of cystine stones in 88 cases (8). However, cystine stones account for (only) up to 3% of stones in the general population, so not finding a case is not very surprising. One might expect increased urinary cystine levels in recipients of living-related grafts obtained from heterozygotes with Type II- and Type III phenotypes. Since I can find no heterozygotes reported with active cystine stone disease, the clinical significance of the finding would appear to be nil, assuming of course that the prospective donor has no history of nephrolithiasis. In Rosenberg's reports, levels of cystinuria that occurred in Type II and III heterozygotes were below 250 mg/gm creatinine, levels unlikely to cause nephrolithiasis. The utility therefore of measuring cystine levels would be negligible. The cyanide-nitroprusside test can be used to screen qualitatively for cystinuria with excellent sensitivity (5).

An additional issue is the relative frequency with which patients with cystinuria, and perhaps their relatives, develop calcium or urate stones. Other metabolic abnormalities accounting for this have been described. Sakhaee (9) found that 5 of 27 cystinurics had hypercalciuria, 6 had hyperuricosuria, and 12 had hypocitraturia. These abnormalities may or may not be resolved by renal transplantation and recipients may then be at risk for recurrent stone disease after transplantation. Of course it is also possible that these abnormalities are intrinsic to the native kidneys, or the result of recurrent stone disease (like renal tubular acidosis with hypocitraturia). Morin (10) described a family in which several heterozygotes for cystinuria had hypercalciuria and/or hyperuricosuria.

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## A Stone Owners Manual

by The Cystinuria Listserv Group

### What Is Cystinuria?

Cystinuria is an inherited metabolic disorder characterized by the abnormal movement (transport) in the intestines and kidneys, of certain organic chemical compounds (amino acids). These include cystine, lysine, arginine, and ornithine. Excessive amounts of undissolved cystine in the urine (cystinuria) cause the formation of stones (calculi) in the kidney, bladder, and/or ureter.

One of the peculiar things about cystinuria is that the amount of cystine excreted by the kidneys is not always related to the number and size of the stones formed. Some cystinurics with very high cystine levels form very few, if any stones. Other cystinurics with comparatively low cystine levels are prolific stone formers.

Diagnostic tests for cystinuria include a 24 hour urine collection to determine the levels of cystine in the urine and a one off urine sample to detect higher than normal levels of cystine in the urine.

Cystinuria has been linked to the chromosome 2P. A mutation in the gene causes cystinuria. It is hoped that in the future, gene therapy may be a cure for the condition.

There are three types of cystinuria, I, II and III.

In Type I Cystinuria, there is a defect in the active transport of cystine and the amino acids (dibasic) lysine, arginine, and ornithine in the kidneys and small intestine. People who are carriers of the gene for this type of the disorder generally have no symptoms.

In Type II Cystinuria, cystine and lysine transport is severely impaired in the kidneys and only somewhat impaired in the intestines.

In Type III Cystinuria, kidney transport of cystine and lysine is defective; intestinal transport is normal. People who are carriers of the gene for this variety of the disease typically have slightly elevated levels of cystine and lysine in the urine.

In Hypercystinuria, there is generally a moderate elevation of cystine in the urine; intestinal absorption of cystine and the dibasic amino acids is normal.

Cystinuria can also be classified by the age at which symptoms first appear. I.e. infantile, juvenile and adolescent.

Regardless of which type you have all cystinurics can look forward to a lifetime of kidney stones. There are various ways of lowering the formation of stones, however as yet there is no cure.

Other problems, apart from kidney stones, cystinurics may encounter are:

- Knowing they have an incurable disease, which can remain silent and is like living with a time bomb
- Knowing the disease has no respect for the cystinurics personal circumstances and can present when the cystinurics least wishes it to.
- Knowing that at any time they can experience severe pain without warning when a stone is on the move.
- Knowing there will be times when they will require hospitalization for treatments, sometimes long term.

Other symptoms not generally associated with stone disease but which are commonly talked about are low grade chronic pain, tiredness, depression, unquenchable thirst, irritability, mood swings and generally being a pain in the neck to friends and families.

This can lead to a certain amount of depression and hopelessness in the cystinuric. This can be overcome with the support of other cystinurics and a very supportive and loving family.

However, conversely, many cystinurics find that when they are well, they have less to complain about than other people and tend to enjoy life more when the disease gives them a break. They tend to have a higher pain threshold for other illnesses and a lower sick rate for minor ailments than the general population.

The most inconvenience some cystinurics have is that they know the location of every toilet in their home

town and are accused of drinking too much! Others find that they spend the majority of their time and money in a continual round of consultants, x-ray departments and treatments. Cystinuria is no respecter of wealth or convenience.

## What Are The Odds?

This is the technical bit. The odds of becoming a cystinuric are dependent on your parents. As cystinuria is a recessive gene it is one of those conditions that should eventually die out in your blood line.

The odds are as follows:

For children of parents who are both full cystinurics the chances of the children becoming cystinurics are 100%. That's the bad news.

For children of parents of whom one is a full cystinuric and the other a carrier the chances of each child becoming a full cystinuric is 50% or 1 in 2. The chances are the same of becoming a carrier.

For children of parents who are both carriers the chances of each child becoming a cystinuric is 25% or 1 in 4.

For children of parents of whom one is a cystinuric and the other is neither a cystinuric or carrier, the chances of each child becoming a cystinuric is nil. However the child will be a carrier. If that child has a child to someone who is neither a cystinuric nor a carrier then that child has a 50% chance of becoming a carrier.

This is all good news for the cystinurics and not good news for cystinuria. The most conservative ratio of a person being a carrier is 1 in 150. So you have a 149 chances out of 150 of having a child with someone who isn't a carrier.

## Symptoms of Cystinuria

### Renal And Ureteric Colic

Renal colic is often the first symptom cystinurics get which gives the doctors a clue to diagnosing cystinuria. Renal colic in young children and adults in the general population is rare. Kidney stones normally take a lifetime to grow. Only in cystinurics and patients with other stone forming disorders are stones present at a young age.

Renal colic is described as a moderate to severe sharp spasmodic pain in the back, side and groin area, often travelling from front to back. Over time can be tracked downwards. Sometimes described as the worst pain a patient has ever had. Can be accompanied by cystitis and bladder spasms, especially when the stone has travelled that far down. Some cystinurics talk of transferred pain to the centre of their back and in their bladder. Cystinurics tend to describe this pain as 'excruciating, stabbing, feels like a knife twisting, doubles me over', 'it feels like someone sticking a knife into your side and slowly turning it', 'sharp, stabbing pain'.

For most people the pain is a lot worse at night. This may be due to the fact that at night it is quieter, with less going on and very few distractions from the pain.

If the pain persists for longer than usual, some investigations are usually necessary to ensure that an obstruction is not the cause for the pain. An obstruction may be signaled by low grade pain in the area of the kidney following a bout of renal colic, patient sensitive to their kidneys being poked (often striking the offender), low or high grade pyrexia (temperature). Care should be taken with cystinurics with an obstruction as a renal infection may develop leading to septicemia.

Investigations would normally include an ultrasound to either find the obstruction and to measure the size of the kidney and an IVP/IVU to find the obstruction.

If these investigations appear to have a negative result, the patient may still have renal colic. Even very small stones can be extremely painful, especially stones which have been previously broken up by lithotripsy or laser treatments and so have sharp jagged edges. These smaller stones do not always show up on investigations.

### **Chronic pain**

Some cystinurics suffer from chronic kidney pain most of the time. This is often described as 'hard, achy feel to my kidneys most of the time', 'squeezing my kidneys in a vice, as hard as they can', 'dull throb which wears me down'. This continual pain can cause a cystinuric to become 'worn down', snappy and difficult to live with. Apart from pain medications there seems to be little that can be done for this pain.

### **Depression**

There has been discussion amongst cystinurics about depression. People were concerned that the essential amino acids that are not metabolized may lead to a deficiency in certain minerals, namely zinc. There is a well known correlation to depression and zinc deficiency. However, it appears that this is not the case. Many cystinurics suffer from a form of depression. This may be due to the nature of the disease. It is difficult to live with a condition which is not visible, extremely painful at times, and does not ever go away. Most of the procedures associated with cystinuria are painful, humiliating or both. For some people the disease can mean many in-patient stays in hospital, disrupting a normal lifestyle. These stays can also be without warning leading the cystinuric to feel permanently off balance and unable to plan. For cystinurics living in the United States and other countries without a national health plan, this can also lead to crippling medical bills which their private insurance just doesn't cover. A cystinuric may find it difficult to find a permanent job, due to the amount of time spent in hospital.

So if you can imagine intermittent severe pain, humiliating procedures, unexpected hospital stays away from the family, huge medical bills and no job leading to the feeling of isolation. If you add to this the rarity of the disease, the overall lack of understanding/interest by the medical profession, having constantly to explain the condition over and over to different doctors, chronic kidney pain, no cure and having to plan around the nearest toilet/drinking place is it any wonder some cystinurics are depressed.

There are treatments for depression. The most common of these are anti-depressants. However, time is the best healer and the support of a loving family and network of friends. Being able to talk out the problems with another cystinuric is also a good treatment which is why organizations like the CSN and the online group are so valuable.

### **Hydronephrosis**

Hydronephrosis is where the kidney is enlarged due to an obstruction. It is a very serious condition. If left untreated Hydronephrosis causes the fluid in the kidney to become stagnant leading to kidney infection, possible septicemia and further complications.

Treatment is not pleasant but very necessary.

### **Tiredness**

This is a big problem for many stone patients. It is characterized by that 4pm feeling when sleep is so attractive. It may be a symptom of underlying depression or could be due to nature of the disease, for example during a stone episode cystinurics may have difficulty in sleeping due to the pain and worry.

## **Prevention Of Stones**

### **Drink gallons and gallons of water.**

This is perhaps the most common prevention method used. Nearly every cystinuric you come into contact with will tell you to drink, drink, drink.

To find out how much you need to drink you need to get a 24hr collection done. When you get your 24hr cystine level, check to see if you have the total amount in mg and the total literage. By dividing the total amount of mg by the litres you get your untreated concentration. You need to get this rate down 200-250mg to prevent stones forming.

For example:

1500mg in 24 hrs with 3 litres of fluids = 500mg/litre. You would need to double your fluid intake to get to your optimal dilution i.e. drink 6 litres of fluid in 24hrs.

At least a third of this should be drunk at night. However this is not always possible so the aim should be to drink as much as possible during the night and the rest during the day.

Or another way to look at it is that the conversion factor for mg to millimole is 240 mg of cystine = 1 millimole, or 1mg = 0.04 millimoles. Thus, production of 1000 mg of cystine per 24 hours is almost exactly 4 millimoles. The rule of thumb is to drink one litre of liquids for every millimole of cystine to keep the average concentration below 1 millimole. If the 24 hour cystine were 1500 mg (about 6 millimoles), one would need at least 6 liters of water.

With regards to what you should drink, there are many theories. Some cystinurics swear by water and water alone. Others say that it doesn't matter what you drink, providing there is plenty of it. Others try to avoid

liquids such as tea, coffee, soda and alcohol. Anything, in fact which has a dehydrating effect or would make the urine more acid.

Medical opinion is also divided. However the most sensible advice would be anything in moderation, providing it is in liquid form and the majority of which is a fluid such as water, which doesn't dehydrate.

Some cystinurics, however, cannot face alcohol at all, others feel that their kidneys have suffered enough, others that it is vital in keeping stones moving. It is one of those things that is personal to each cystinuric.

Some cystinurics, especially those who suffer a lot with stones, have a PICC line inserted. This is a semi-permanent IV line where the cystinuric can hook up fluids which can run through the night, increasing fluid intake at a time when the kidneys are most sensitive to producing stones. This is not a permanent line, is visible and definitely not for most people. Certainly a cystinuric who is controlling their hydration, pH levels and medications would not normally consider this option. It is a very drastic measure but one which has been successful for some people.

#### **Follow a methionine free diet.**

Methionine is the amino acid from which we get cystine. To reduce the amount of methionine ingested would also mean a reduction in the cystine produced. Methionine is found in animal protein.

The diet is simple. Exclude animal protein by not eating any meat, limited milk, yoghurt, cheese and other dairy, no eggs and no fish. Stock up on green vegetables and other forms of vegetable protein such as nuts, beans and pulses.

It is a very hard diet to follow strictly, however, there are a few cystinurics who do it successfully. The value of the diet has not been proven scientifically, however it makes logical sense to reduce the amount of cystine we eat to reduce the amount of cystine available to form into stones.

Many cystinurics follow a modified form of the diet and just do the best they can not to go overboard on animal products.

Some cystinurics have been advised that eating fish at all is worse than eating some meat.

#### **Alkalization**

Raising your pH to a level of 7.5 - 8.0. Having a high pH means that the environment in the kidneys is not conducive to forming stones.

Alkalize your urine by using an alkali such as Urocit-K, Polycitra, sodium citrate, potassium citrate or sodium bicarbonate. To work out how much alkali is necessary just take 24 hour urine profiles to find your low spots and alkalize accordingly.

A person's pH is dependent on what they eat. This is why some cystinurics follow a low meat, high green vegetable diet, even if they are not following the low methionine diet. Green vegetables and lemon juice naturally raises the pH of the urine. Meat, fish, sodas and alcohol naturally lower it

Remember that a pH of 8.0 and higher is likely to encourage calcium stones to develop.

You could also use lemon juice in water to a dilution which is drinkable. Although lemon juice is acidic it acts on the body like an alkaline and raises pH.

If you are using a sodium based alkali, you may be advised to reduce your salt intake. You may choose to reduce your added salt, avoid processed foods and foods which have a high salt content.

#### **Medication**

##### Tiopronin (Thiola)

This drug prevents the formation of kidney stones when there is too much cystine in the urine. It is the medication of choice for many people. It is not available yet in the UK., however it is available in the US and Ireland. However it can have some severe side effects such as skin rashes, itching skin, mouth sores, mouth ulcers and a condition which decreases the elasticity of the skin. It can also cause abdominal pain, gaseousness, diarrhea, nausea and vomiting. It has certain unwanted effects on an unborn child and should not be taken until the mother has discontinued breastfeeding. The good news is that hydration increases the effects of Thiola and it is often effective in controlling stone formation.

##### d-Penicillamine (Depen, Cupramine, Distamine)

This drug combines with cystine to prevent cystine stones. It helps the solubility of cystine. This is the drug that is widely available in the UK. Although it is slightly less effective as Thiola, it is effective enough to stop stones from forming. It is not commonly used in the US as Thiola is found to be more effective with fewer side effects.

The user may have side effects, the most common of which are a rash, itchy skin, swollen lymph glands, appetite loss, nausea, diarrhea, vomiting and impaired taste. It can also cause a sore throat, fever, unusual bruising, swollen feet or legs, bloody or cloudy urine, weight gain, fatigue, weakness and joint pain. It can also cause double or blurred vision, pain, ringing in the ears, ulcers, sores, white spots in the mouth, difficult breathing, coughing up blood, jaundice, abdominal pains, skin blister and peeling skin. Not a pretty list!

The cystinuric must be monitored with regular blood tests to ensure compatibility, for example white blood count, platelet count, red blood count, hemoglobin and hematocrit, kidney function and liver function. When a cystinuric first starts to take Penicillamine, it is generally started slowly and gradually increased to minimize the side effects. The side effects are the main reason that this is not the drug of choice for many people. In addition to all this, it may be advisable to increase your B6 vitamin intake by taking a supplement as a B6 deficiency is common with this drug.

It is not advised that a patient gets pregnant whilst taking this drug and the drug carries over into the breastmilk.

However for some it is the only choice and is used effectively by many people.

## Captopril (Capoten)

This is the drug of choice for those who are intolerant to Thiola and d-Penicillamine. Although it is not as effective as the drugs above, it can be useful in severe cases where the drugs above are no longer tolerated. It is the drug of choice for cystinurics with poor kidney function, for whom the other two drugs are not suitable.

It is more commonly used as a treatment for high blood pressure and for patients with congestive heart disease.

Its side effects are fewer than with the other two drugs. For example a user may find they have a rash and loss of taste. It can, however, cause a severe anaphylactic reaction for those who are intolerant. Other side effects are swelling of the mouth, face hands and feet, dizziness, fainting, chest pain, fast or irregular heartbeat, coughing, confusion and nervousness. Also diarrhea, headache tiredness, sore throat, cloudy urine, fever, chills, nausea, vomiting, indigestion and abdominal pain. It must be stressed, however, that generally this is a safe drug with few side effects presenting themselves.

You should not get pregnant on Captopril and it passes over to breastmilk.

Prolonged use may cause a decrease in white cells and proteinuria (protein in the urine). Regular blood tests should be taken to confirm continued compatibility and the drugs should not be discontinued abruptly.

## **Diagnostic Devices For Stones**

### **KUB**

KUB stands for flat x-ray of the kidney, ureters and bladder. This is the standard form of x-ray. Cystine stones do not show up on these x-ray at all unless they are mixed with another ingredient such as calcium or oxalate or are very big. Their usefulness is doubtful however if it keeps the doctors happy...

### **Ultrasound**

This is where a patient lies on their back, jelly is applied to their belly (often freezing cold) and a device similar to a computer mouse is run over the renal area. A patient may be asked to roll on their side and both belly and back are scanned. The operator may also try to scan the ureters and bladder for stones/obstructions. The kidney can be measured using this device. Stones show up as bright stars in the kidneys. Bright stars without shadows may not be stones but collections of crystals. It takes a very skilled operator or a cystinuric to spot the stones.

### **IVP/IVU**

This is an x-ray which involves having a line inserted into a vein, dye is injected into the line and the cystinuric undergoes a series of x-rays. A patient may feel a warm sensation in various parts of their body. Sometimes a belt is used to slow down the flow. These x-rays can take anything from one to six hours. Cystinurics are often left in the x-ray room inbetween times. They are very boring. Sometimes this is the only way to see cystine stones.

## **Nephrostogram**

This is an x-ray of the kidney and ureters where contrast is introduced through the nephrostomy tube, already in situ. If the kidney is obstructed this may be painful as the kidney is filled with contrast to its full capacity and causing dilation.

### **VCUG (Voiding cystourethrogram)**

This is a particularly humiliating x-ray examination of the urethra and bladder. A patient is catheterized, contrast is introduced into the bladder and the catheter is clamped. A number of films are taken in various positions. The catheter is removed and the patient is encouraged to urinate on the table. A film is also taken whilst voiding. This is a particularly distressing examination and mild sedation is sometimes used.

### **Cat Scan/Spiral Cat Scan**

Cat (Computer applied tomography or CT, computerized tomography) scans provide a three dimensional picture of the abdomen, kidneys, ureter, bladder, and surrounding tissues. It yields definitive information about tumors, cysts, masses, or obstructions. They do use a bowel prep the night before and no food after midnight.

It is performed in radiology and a contrast media is injected in the vein or given in the form of a fizzy drink to help visualize the images, then pictures are taken. Not all procedures use a contrast medium. Some cystinurics now insist that the CT scan is performed without contrast as they have found the contrast covers up the cystine stones.

This procedure is similar to an IVP except for the fact that the cystinuric is asked to lay on a moving table which moves them inside the machine. Some people find this uncomfortable or claustrophobic.

The cystinuric may be asked to remove their clothes and put on a gown. This can be avoided by wearing soft, loose clothing with no metal fastenings, no jewellery or bra. Then they lay down on the little table that moves them into the machine. It is open ended so it isn't usually a problem for claustrophobics. They hold their arms above their head, take in a DEEP breath and hold it for about 45 seconds. If they can't hold their breath that long they can VERY slowly let it out. The technicians are trying to avoid their organs moving to obscure the picture.

Some cystinurics say it feels like a line of static electricity running over their body, but it is not at all uncomfortable.

The scan can diagnose stones which show up "bright shiny" spots in the kidneys and other areas. It also will show other anatomy of the kidney. It can also show up "brush borders" which are areas of the kidney which have been hardened by the cystine.

# Treatment Of Stones

## Invasive Procedures

### Open Surgery (Extended Pyelolithotomy)

A patient is given a general anesthetic. Surgeons physically remove the whole stone by cutting into the kidney and removing the stone. Recovery time 6 weeks. Hospital stay 10-14 days. Complications such as severe pain on recovery from the anesthetic, initial mobility problems as associated with any major surgery, need for recovery time and time off work, reduced kidney function. The kidney will only tolerate this procedure a few times. Benefits include quick fix and possibly only solution for very large stones. The procedure leaves a 10 to 12 inch scar.

### Percutaneous Surgery (keyhole)

A patient is given a general anesthetic. The surgeon will make three or four small one inch incisions in the back. A camera is passed through one of the incisions. A laser or lithoclast is inserted into the kidney and the stone is broken up. Some of the stone can be removed through the incisions. Others are left to pass naturally. Hospital stay 4-7 days. Recovery time 2 weeks depending on the patient. A nephrostomy tube may be left in temporarily to aid drainage and to allow the kidney to heal. A Stent tube is sometimes inserted, especially if there is a narrowing of the ureters.

### Minimally Invasive Surgical Procedures

The following procedures are minimally invasive and require less recovery time than standard surgical procedures. They can be used following Percutaneous surgery or ESWL to remove fragments.

### Cystoscopy (Retrograde Cystoscopy)

This is a procedure which is used in conjunction with various probes and lasers. It can either be used as a diagnostic test, in the case of pain without a stone, or as a way of inserting a probe to break up stones and a dornier basket to remove stones.

Either under a local or general anesthetic a long flexible tube containing the camera is inserted into the urethra. It then passes into the bladder and, depending on what it required, is passed through into the ureter. Once in position, various other instruments are inserted through the tube and treatment is carried out.

Following the procedure you may feel discomfort in the bladder. However it may be very difficult to pass water the first few times. It feels like a severe case of cystitis. It may be worth holding on before going to the toilet. The cystoscope will have emptied the bladder during the procedure, but the trauma to the bladder makes it want to expel almost immediately afterwards. Once enough fluids have been drunk to necessitate voiding, it is best to relax as much as possible. The urethra may still be in spasm and so does not want to release as well as normal. It can be incredibly painful the first time, but after 1 – 2 hours the bladder and urethra returns to normal.

Depending on the procedure carried out, you may pass a little blood and some gravel which should be handled as you would normally.

### Dornier Basket Removal via Cystoscopy

This is a procedure where, under a general anesthetic, a camera is inserted through the urethra into the bladder. It is then moved into the ureter until it reaches the stone. A basket type attachment (like a grabber in the funfair side shows) is inserted alongside the camera and passed along to the stone where the basket is opened put around the stone and closed. The basket is then removed from the ureters, bladder and urethra, with the stone inside.

This is normally a straight forward procedure and is useful for small stones which are not too far up the ureters. The effectiveness of the dornier basket is dependent on whether the operator is able to get the equipment to the stone and the size of the stone.

### Holmium Laser Therapy

This is a very powerful laser used to break up stones in situ. It is inserted in the same way as the dornier basket and can also be used in conjunction with percutaneous surgery. The laser is fired at the stone, cutting it up. Only a very skilled operator can use this equipment as the laser is extremely powerful.

### Other Laser Therapy

There are other lasers which are used to cut up stones in situ. However their effectiveness is dependent on the density, hardness and size of the stone as they are less powerful.

### Direct Lithoclast

A Lithoclast is a type of lithotripter probe, also used in conjunction with cystoscopies and Percutaneous surgery. The probe is placed directly on the stone and the shockwave is fired, hopefully fracturing the stone. Again, effectiveness is dependent on hardness, density and size of stone.

### Nephrostomy

This is where a tube is inserted through an 1 inch incision in the back, directly into the kidney. Local anesthetic is given to the area of the skin where the incision is made. Some form of tranquilizer is also given to calm the patient. The time taken for this procedure depends on the state of the patient. It is sometimes seen as a barbaric procedure, however it is sometimes necessary to reduce Hydronephrosis due to an obstruction. An obstructed kidney which is not drained may become septic, damaged and possibly lead to septicemia. Once inserted nephrostomies (although unsightly and embarrassing) are not usually painful.

Nephrostomies are safe to leave in for up to six weeks. They are a source of infection, however and care must be taken to keep them as clean as possible.

## Caring for a nephrostomy tube

So you are going home with a nephrostomy tube and legbag. Now what?

With a bit of care, you can make this experience as comfortable as possible. You will need: tape, non-stick gauze pads, a type of combine dressing or padded dressing, and replacement leg bags. Do not use the type of gauze pad with a slit for the tube as this tends to stick to the tube and can be painful to remove. Tape the nephrostomy tube, about 4-6 inches down the tube, on a downward angle so that it ends up coming down your hip and the outside of your leg. Taping the tube in place helps relieve any pressure or pulling on the tube caused by the legbag. Place the bag on the outside of your leg at a level that you find comfortable. You do not want the tube placed on your buttocks so that you will sit on it. Sitting on the tube can cause punctures and leaks. Place the non-stick pad on the combine or padded dressing so that it faces the nephrostomy tube entry point. Tape in place taking care not to tape the bandage to the tube. I personally prefer using a 5in x 9in (12.7cm x 22.9cm) combine dressing folded in half for additional padding, using a minimum of tape to hold the bandage in place. Tape that goes on - must come off - ouch!!

Since baths are generally frowned on while wearing a tube, showers are great. A handheld shower head allows you to aim the water directly at the tube insertion point washing away any discharge. A wet or dry cloth used gently on the site will also help remove any irritating dry discharge. Take care that you do not pull on the tube or the stitch that holds it in place.

Legbags come in different sizes. A smaller person would use a smaller bag. The best type is the kind that has a nozzle or flip top on the port. The types with rubber caps can easily be dislodged by pants and result in leakage. Fold the port under the bag, facing up your leg so that it is protected by the bag. This helps minimize any embarrassing accidents. Cloth/Velcro or elastic legbag bands are much more comfortable than latex bands which can irritate the skin. Change your bag every week to avoid unpleasant odors.

## **Stent Tubes**

This is another way in which kidney are allowed to drain. Stent tubes are often used after surgery to allow the kidney to heal and to dilate the ureters. This is useful if the ureter has narrowed due to an obstruction. They are sometimes used in conjunction with a nephrostomy. They are inserted under a general anesthetic but often removed under a local.

As the bladder is a smooth muscle, Stent tubes can cause severe spasmodic pain for a period after they have been inserted. Various drugs can be used to relieve this spasm pain. Cystinurics can also find relief in using a hot water bottle on the bladder. If the stent lies on a certain place in the bladder they can also cause nerve pain. They often also cause the urethra to spasm which is very uncomfortable. The constant discomfort of a Stent tube can cause some cystinurics to become tired and run down. They can also cause

depression in cystinurics who have to have them for an extended amount of time.

Stent tubes are safe to leave in for up to six months. They are often removed in a day unit without anesthetic with a Cystoscopy. Some cystinurics find that crystals form around the Stent tube which can make removal uncomfortable. Other cystinurics do not have any problem with stents tubes and prefer them to the nephrostomies.

## **Non Invasive Procedures**

### **Extra Corporeal Shock Wave Lithotripsy (ESWL)**

This is a procedure where the sound shock waves are used to break up stones without the need for surgery. A lithotripter comes in two forms. The patient either lies in a bath of warm water, under general anesthetic, and the shock waves are fired through this water onto the kidney. This is an older form of lithotripter which is not always effective. The newer forms of lithotripter use a table with a hole in it. A patient lies on the table and the dome which includes the shock wave mechanism is raised to meet the cystinurics back. There is often a rubber or silicone sheet filled with jelly or warm water between the patient and the mechanism. The dome is pressed into the patients back and the shock wave is fired.

The shock waves are timed to the cystinurics heartbeat. It can take anything from 500 to 2000 shock waves to fracture the stone depending on the size and density of the stone. Monitoring of the progress is done by either ultrasound or image intensifiers. Careful placing of the sound shock waves is necessary to ensure accuracy. The patient must lie completely still during the process.

The process is not always painful although some cystinurics find the pain is either intense kidney pain, or irritating on the lines of water torture. Various drugs are used to control this pain.

Usually lithotripsy is performed as a day case and a hospital stay is not necessary. Stone fragments are usually passed within 1-2 days but further treatment may be required.

Please note not all makes of lithotriptors are effective in breaking up cystine stones.

## **Pain Relief Medications**

Our list of favourite and well used medications is as follows:

Morphine Sulfate (MST)  
Hydromorphone Hydrochloride  
Meperidine Hydrochloride (Demerol)  
Oxycodone Hydrochloride (Percocet)  
Oxycodone Terephthalate (Percodan)  
Diclofenac Sodium (Voltaren, Volterol)  
Diclofenac Potassium (Cataflam)  
Buprenorphine Hydrochloride (Temgesic)  
Pethidine Hydrochloride (Pethidine)  
Meptazinol (Meptid)  
Hydrocodone Bitartrate (Vicodin, Lorcet, Lortab)  
Propoxyphene Napsylate (Darvocet)  
Propoxyphene Hydrochloride (Darvon)  
Promethazine Hydrochloride (Phenergan)

Fentanyl (Duragesic)  
Dihydrocodone Bitartrate (DF 118)  
Gas & Air (Entonox)  
Codeine Phosphate with Acetaminophen Tylenol &  
Codeine (Furinol)  
Acetaminophen (Tylenol)  
Methadone Tablet  
Stadol Nasal Spray  
Ibuprofen Tablets (Brufen, Buprofen, Advil)  
Hypnoval (for nephrostomies)  
Valium Tablet (for sedation)  
Oxybutinin Tablet (for spasm)  
Stemetil Tablet (for sickness)

### To keep with you just in case.

Some cystinurics carry with them drugs such as Vicoden or Ibuprofen with Codeine just in case they get a bout without warning. There is a lot to be said for the feeling of security this gives you.

### Self Help Pain Relief

- Soak in a hot bath, use a hot water bottle or heat pad. The heat is a good form of pain relief.
- Keep busy, talk to people, use diversionary tactics. This may sound easier than it is to do but for long term or persistent pain this is a good habit to get into.
- Drink plenty. This is a recurring theme. Although it may increase the pain in the short term, it will make the pain go quicker.
- Go for a run, walk round the block or go swimming. Again, although you may not feel like doing this, if you are able it helps to relax the muscles and reduce the muscle spasms.

### Some Suggestions For Moving Stones Quickly

With all stones it is important, before you make the decision to encourage the stone to move, to ensure that the stone is small enough to pass through your tubes and not to cause an obstruction. For stones that are too large to be passed naturally other forms of stone removal are probably best. Take advice from your doctor before trying any of these methods.

- Get and use pain relief promptly. Don't wait until it is really bad. The quicker you take the medication the more use it will be.
- Stand on your head to move the stones from the bottom parts of your kidney (this may sound strange but this has been suggested to several people)
- Drink pints and pints of water. This may be the last thing you want to do with renal colic, but it is the only way to keep those stones rolling.
- Go for a run or walk round the block. Again, probably the last thing you want to do but it gets the muscles in that area moving.

- Have a beer. Be careful that the stone isn't too large but this one way many cystinurics expel the stones.
- Drink Oolong Tea, the chinese tea you get in restaurants. It helps to clear out the system.

### Useful Contacts:

Cystinuria Support Network  
C/o Jann Ledbetter  
21001 NE 36<sup>th</sup> Street  
Redmond WA 98053  
[Cystinuria@aol.com](mailto:Cystinuria@aol.com)

Cystinuria Support Network UK  
C/o Sue Holden  
93 Dragon Parade  
Harrogate  
North Yorkshire  
England  
[sue@cystinuria.com](mailto:sue@cystinuria.com)

## A Glossary of Terms

**Acute Pain** = Sudden or Severe pain. Symptoms appear, change or worsen rapidly. Severe pain, as may follow surgery or trauma.

**Acid** = A sour substance. Reacts to a base to form a salt.

**Amino Acids** = A group of chemical compounds that forms the basic structural units of all proteins. There are 20 different amino acids that make up all the proteins in humans. Of these, 12 can be made by the body, because they do not need to be obtained from the diet. The other eight, the essential amino acids, cannot be made by the body and must be obtained from the diet. The 20 amino acids that make up proteins also occur free within cells and in body fluids. In addition, there are more than 200 other amino acids that are not found in proteins but which play an important part in chemical reactions within cells.

**Analgesic** = A drug that relieves pain.

**Asymptomatic** = Absence of symptoms. An illness or condition may be present without recognizable symptoms.

**Calculus** = A stone that has formed or is present in the kidneys, ureters, or bladder, caused by precipitation from a solution of substances in urine.

**Catheter** = A flexible tube used either for draining fluid from or injecting fluid into the kidney.

**Cat scan** = A diagnostic technique in which the combined use of a computer and x-rays passed through the body at different angles produces clear cross sectional images of the tissue being examined. Cat scanning provides clearer and more detailed information than x-rays used by themselves.

**Chemical Analysis** = Check ketones, sugar protein and blood.

**Chronic Pain** = Pain that continues or recurs over a prolonged period, caused by various diseases or abnormal conditions.

**Creatinine** = A substance formed from the metabolism of creatine, commonly found in blood, urine, and muscle tissue.

**Cystine** = A non essential amino acid found in many proteins in the body, including keratin and insulin. Cystine is a product of the oxidation of two cysteine molecules.

**Cystine Stones** = Stones formed from the amino acid cysteine. Cystine stones are

**Cystinuria** = An inherited metabolic kidney disorder characterized by stones in the kidney, ureter, and bladder; caused by excessive excretion of certain amino acids (protein building blocks) because of genetic abnormality. The Kidneys do not adequately reabsorb certain amino acids during the filtering process, resulting in excess excretion of these amino acids.

**Cystitis** = Inflammation of the inner lining of the bladder, caused by an infection that is usually due to bacteria.

**Cystoscopy** = The examination of the urethra and bladder cavity using a cystoscope (viewing tube inserted up the urethra).<BR>

**Cystourethrogram, (Voiding) (VCUG)** An x-ray procedure for studying a person's bladder while he or she is urinating. A catheter is inserted into the urethra and passed through the bladder. A contrast is injected through the catheter.

**Dehydration** = Excessive loss of water from the body tissues. Dehydration is accompanied by a disturbance in the balance of essential electrolytes, particularly sodium, potassium and chloride. Dehydration may follow prolonged fever, diarrhea, vomiting, acidosis, any condition in which there is rapid depletion of body fluids.

**Diagnosis** = Identification of a disease or condition.

**Diagnostic Radiology** = Medical imaging using external sources of radiation.

**Endoscope** = An illuminated optic instrument for the visualization of the interior of a body cavity or organ.

**Endoscopy** = Examination of a body cavity by means of an endoscope, a tube like instrument with lenses and a light source attached.

**Extracorporeal Shockwave Lithotripsy (ESWL)** = A procedure which shatters kidney stones into fragments so small they can pass through the ureter and the bladder where they finally leave the body via the urethra when a person urinates.

**Foley Catheter** = The most common type of catheter which is used to drain urine from the bladder.

**Genetics** = The study of inheritance. How the characteristics of living organisms are passed from one generation to another, the chemical basis by which such characteristics are determined, and the cause of the similarities and differences among individuals. Genetics includes the study of deoxyribonucleic acid (DNA), the substance in cells that determines the characteristics of an organism, and of genes, which are units of inheritance corresponding to specific bits of DNA.

**Hematoma** = A localized collection of blood (usually clotted) caused by bleeding from a ruptured blood vessel. A hematoma may occur almost anywhere in the body and, depending on the site and amount of accumulated blood, may vary in seriousness from a minor to a potentially fatal disorder.

**Hematuria** = Abnormal presence of blood in the urine.

**Hydronephrosis** = Distention of the pelvis and calyces of the kidney by urine that cannot flow past an obstruction in a ureter.

**Hypertension** = A common, often asymptomatic disorder characterized by elevated blood pressure exceeding 140/90. A person with hypertension has a high blood pressure at rest.

**Idiopathic** = Without a known cause.

**Intravenous** = (IV) A term meaning within a vein.

**Invasive** = An invasive medical procedure is one in which body tissues are penetrated by an instrument.

**Intra Venous Pyelogram (IVP)** = A procedure for obtaining x-ray pictures of the urinary system. The technique involves the introduction of a radiopaque, iodine-based dye into the kidneys, ureters, and bladder so that they show up well on x-rays. Pyelography is performed to help diagnose disorders of the urinary system.

**Kidney** = The organ responsible for filtering the blood and excreting waste products and excess water in the form of urine.

**Laser** = A device that produces a concentrated beam of light radiation; laser is an abbreviation for light amplification by stimulated emission of radiation.

**Laser Treatment** = The use of laser beam in a variety of medical procedures. Treatment with low-intensity beams stimulates tissue healing and reduces pain, inflammation, and swelling. High-intensity treatment destroys cells directly under the beam while leaving adjacent cells undamaged making it useful in the treatment of some tumors. The beam cuts through tissue and, simultaneously, causes blood clotting, making it a useful surgical tool. Laser is also used to disintegrate bladder and kidney stones.

**Leukocytes** = Any type of white blood cell. Excessive leukocytes suggest a bacterial or other infection.

**Lithotripsy** = The process of using shock waves or ultrasonic waves to break up calculi (stones) for excretion. ESWL is used to break up smaller stones. This technique uses a machine called a lithotripter to produce external shock waves to break up the stones. X-ray imaging systems are used to show the position of the stone and to monitor its destruction into a fine sand which is passed out urine over the following few weeks.

**Lithotripter** = The machine used in ESWL to disintegrate small stones.

**Metabolic** = The aggregate of all chemical processes that take place in living organisms, resulting in growth, generation of energy, elimination of wastes and other bodily functions as they relate to the disturbances of nutrients in the blood after digestion.

**Methionine** = An essential amino acid needed for proper growth in infants and for maintenance of nitrogen balance in adults.

**Narcotic Drugs** = A type of analgesic (painkiller) used in the treatment of moderate and severe pain. Abuse of narcotic drugs for their purpose often causes tolerance (the need for greater amounts to have the same effects) and physical and psychological drug dependence. Non narcotic drugs are useful in the treatment of mild or moderate pain.

**Nephrectomy** = Surgical removal of one or both of the kidneys.

**Nephritis** = Inflammation of one or both kidneys.

**Nephrolithomy** = The surgical removal of calculus from the kidney by cutting through the body of the kidney. Can be done by abdominal incision or through puncture incision made through the skin in the back and directly into the kidney.

**Nephrologist** = A specialist in the diagnosis and treatment of kidney disease.

**Nephrostomy** = The introduction of a small tube into the kidney to drain urine to the abdominal surface, thus bypassing the ureter. Nephrostomy is sometimes performed after an operation (to remove calculus) on the ureter or kidney to allow healing to take place.<BR>

**Non-invasive** = Any medical procedure that does not involve penetration of the skin or entry into the body through any of the natural openings.

**Percutaneous** = A medical term meaning performed through the skin. Percutaneous procedures include the injection of drugs into veins, muscles, or other body tissues and biopsies in which tissue or fluid is removed with a needle.

**Percutaneous Lithotripsy** = A nephroscope (type of endoscope) is inserted into the kidney via a small flank incision. An ultrasonic probe is directed through the nephroscope to break up the stone; fragments are removed through the nephroscope.

**PH** = A measure of the acidity or alkalinity of a solution. The PH scale ranges from 0 to 14. 7.0 is neutral, below 7.0 is acidic and above 7.0 is alkaline. The PH of body fluids must be maintained very near 7.4 for the body's metabolic reactions to proceed properly.

**Proteins** = Large molecules that consist of hundreds of thousands of amino acids linked to form long chains, which are often folded in various ways. Protein is the major source of building material for muscles, blood, skin, hair, nails and the internal organs. It is necessary for the formation of hormones, enzymes, and antibodies and as a source of heat and energy and it functions as an essential element in proper elimination of waste materials. Rich dietary sources are meat, poultry, fish, eggs, milk and cheese, which are classified as complete proteins because they contain the eight essential amino acids. Protein deficiency causes abnormal growth and tissue development in children, whereas in adults it results in lack of vigor and stamina, weakness, mental depression, poor resistance to infection, impaired healing of wounds and slow recovery from disease. Excessive intake of protein may in some conditions result in fluid imbalance.

**Proteinuria** = The presence in the urine of abnormally large quantities of protein, usually albumin. Proteinuria rarely causes any symptoms, although the urine may appear frothy.

**Pyelogram** = An X-Ray picture of the kidney.

**Pyelonephritis** = Inflammation of the kidney, usually caused by a bacterial infection.

**Pyuria** = The presence of white blood cells in the urine, usually a sign of an infection of the urinary tract.

**Referred pain** = A referred pain is one felt in a site other than an injured or diseased part. Sensory nerves from certain body areas converge before they enter the brain, causing confusion about the source of pain signals.

**Reflux** = An abnormal backward or return flow of fluid in a body passage due to failure of the passage's muscle to close fully. One type is the back flow of urine from the bladder into one or both ureters. Persistent urinary reflux may lead to kidney damage.

**Renal** = The medical term for anything related to the kidneys.

**Renal biopsy** = A procedure in which a small portion of kidney tissue is removed and examined under microscope.

**Renal colic** = Sharp, severe pain (intermittent spasms) in the lower back over the kidney's radiating forward into the groin. Renal colic usually accompanies forcible dilation of a ureter followed by spasm as a stone is lodged or passed through it.

**Renal Failure** = Reduction in the ability of the kidneys to filter waste products from the blood and excrete them in the urine, to control the body's water and salt balance, and to regulate the blood pressure. Renal Failure can be acute or Chronic. In acute renal failure, kidney function usually returns to normal once the underlying cause has been discovered and treated. In chronic renal failure, function is usually irreversibly lost.

**Spasms** = An abrupt and forceful contraction of a muscle, usually maintained for several minutes or hours and frequently associated with marked pain.

**Specific Gravity** = Also called relative density, the ratio of the density of a substance to that of water. Materials with a relative density of less than 1 are less dense (lighter) than water. Those with a relative density of more than 1 are denser (heavier) than water. The specific gravity of urine shows if it has a large amount of material dissolved in it (near 1.030) or if it is almost water (near 1.010).

**Staghorn Calculus** = Stones that form within and fill the pelvis of the kidney.

**Stent** = A thin silastic tubing running from the kidney through the bladder to allow safe passage of the shattered stone fragments.

**Streptococcal** = An infection caused by bacteria of the streptococcus family. Almost any organ of the body may be involved. The infections occur in many forms including urinary tract infection.

**Struvite** = Infection stones. Struvite stones form because certain bacteria that grows in the urine produce ammonia, thus making the urine alkaline.

**Symptomatic** = Showing symptoms or concerning a symptom.

**Ultrasound Scanning** = A diagnostic technique in which very high frequency sound waves are passed into the body; the reflected echo's are detected and analyzed to build a picture of the internal organs.

**Ureter** = Either of two tubes that carry urine from the kidneys to the bladder. Each is about 10 to 12 inches (25 to 30 cm) long. Urine flows down the ureters partly by gravity but mainly by peristalsis, a pumping action as waves of contraction pass several times per minute through the muscle layers

**Ureteritis** = An inflammatory condition of the ureter that may be caused by blockage with a stone or by infection spreading up from the bladder.

**Ureterolithotomy** = The surgical removal of a urinary tract calculus.

**Urethra** = The tube by which urine is excreted from the bladder.

**Urinalysis** = A physical, microscopic or chemical examination of urine. The specimen is physically examined for color, turbidity, specific gravity and pH. Microscopically for blood cells, casts, crystals pus and bacteria.

**Urinary tract** = The part of the body concerned with the formation and excretion of urine. The urinary tract consists of the kidneys, the renal pelvis, the ureters, bladder and urethra.

**Urinary Tract Infection (UTI)** = An infection anywhere in the urinary tract. UTI is usually characterized by urinary frequency, burning, pain with voiding, and, if the infection is severe, visible blood and pus in the urine. UTI include cystitis, pyelonephritis, and urethritis.

**Urine** = The fluid secreted by the kidney's transported by the ureters, stored in the bladder and voided by the urethra.

**Urine Retention** = Inability to empty the bladder and difficulty in doing so.

**Urography**= Any of a group of x-ray techniques used to examine the urinary system. A radiopaque substance is injected, and x-ray films are taken as the substance is passed through or excreted from the part of the system being studied. Some kinds of urography are cystoscopic, urography, intravenous pyelography and retrograde pyelography.

**Urologist** = A physician who specializes in the practice of urology.

## Ask the Doctor

### 1. Are we OK to eat tomatoes? I understand they are quite acidic.

Tomatoes are OK. Fruits and vegetables lead to urinary alkalization. Yes there is citric acid in tomatoes, which has NO NET EFFECT on urinary pH. The citrate part of citric acid is a base, and neutralizes the effect of the acid. That's why there is no problem with Bicitra, as one example, containing citric acid even if the preparation is used for urinary alkalization. Who could give up tomatoes?

### 2. Does a low methionine diet really help?

Protein ingestion leads to more urinary acid to excrete. Fruits and vegetables lead to alkalization. This may be a more important reason to restrict protein intake than limiting methionine intake. I agree that the benefit of diet in the long run is not well studied, in children or adults and that protein intake could be overcome by more water drinking and effective alkalization. But I've noted that some of you out there claim to have benefited from a more vegetarian diet and I would bet that the effect on urine pH is more important than the effect on cystine excretion.

### 3. Isn't Captopril a diuretic and therefore something we should avoid?

Captopril (brand name Capoten) is not really a diuretic, though it may increase urine output in some patients, especially with heart disease, and can potentiate the effect of some diuretics. It lowers blood pressure and has beneficial effects on the kidneys for people with decreased kidney function and a tendency to progressively lose kidney function (like diabetes), which most people with cystinuria don't have. It also happens to have a sulfhydryl group, which combines with the sulfhydryl groups in cystine to help solubilize the latter. The literature on its benefit is very spotty, with claims both ways. It's reasonable to use it though

its effect is probably small and my own feeling is that urinary cystine levels should be measured to show the benefit or its not worth continuing. But if you think its helping continue it anyway! It is absolutely contraindicated in women who might conceive while taking it. It won't affect your ability to conceive later, it just causes significant birth defects if you conceive while on it. Cough is a common side effect.

### 4. Should we avoid diuretics which could lead to damage to the kidneys?

Diuretics are not drugs that to my mind "damage" the kidneys with time. They can be used safely for long term treatment of high blood pressure. They have some mild side effects, but thiazides, as an example, are a class of drugs that have been used safely by millions of people for high blood pressure and they are currently in the first line of recommended drugs for high blood pressure according to the authoritative Joint National Committee (JNC) VI.

### 5. I am a little concerned that these amino acids we don't absorb are 'essential' amino acids. What are they 'essential' for?

The diminished absorption of various amino acids in people with cystinuria does not lead to nutritional deficiencies. This is because the ability to absorb small peptides (small chains of 2 or 3 amino acids) is NOT impaired. Only single amino acids. Therefore someone with this defect in intestinal absorption will still absorb enough cystine, lysine, etc., in a different form, that then can be broken down to the single amino acids.

Similarly, the loss of cystine and the other amino acids in the urine will not lead to deficiencies of them; it's just not enough to lead to "negative" balance of them. There is no evidence that I know of that there are nutritional defects of any amino acids.

### 6. If we are not able to utilise certain minerals because we don't absorb the amino acids, is there any point in taking supplements?

Minerals, like zinc, are not amino acids, and absorption or utilization of them as far as I know, is not affected by cystinuria.

### 7. Is there any research into this subject?

Sure. I've given you what I think is the best current information.

### 8. Could my slowness to conceive be anything to do with cystinuria and/or mineral deficiencies?

I can't think of any reason why. I also don't think that 12 months is really "slowness" yet. But that's outside of my professional expertise. Though within my personal expertise!

Answers supplied by Dr David S Goldfarb, M.D.

### 12. There are many different makes and types of Lithotripsy machine ESWL, and I was wondering if there is a difference in the effectiveness of the machines.

The Dornier is the most powerful lithotripter but uses old technology x-ray imaging which makes it difficult to see cystine stones. We use the Storz SLX which uses ultrasound for localization and this is a plus for cystine. It is also very powerful - but as you all know cystine is tough to break in general.

Answer supplied by Dr Mike Grasso, M.D.

## Thank You!!!

Thank you to the following people who have made contributions to The Cystinuria Support Network since the last newsletter was printed:

All donated funds are used to offset the costs of mailing, printing, phone calls, on-line services, and to pay annual membership dues for CSN in organizations such as National Organization of Rare Disorders and the Alliance of Genetic Support Groups.

Thank you to all of you who took the time to write articles or share your stories for this newsletter.

*Send me an article about yourself and your experience with Cystinuria so that other participants can get to know you.*

*Send me a concern you are struggling with that someone else may have some insight into for the "Mailbag" column. This could be a question and answer type column with medical opinions if appropriate.*

*Send me ideas you have for articles for future newsletters and I will try to locate someone who is willing to address the issue.*

### **This newsletter requires your input.**

*Send your ideas, questions, articles etc. to:*

Jann Ledbetter  
Cystinuria Support Network  
21001 NE 36th Street  
Redmond WA 98053  
206-868-2996  
email: [cystinuria@aol.com](mailto:cystinuria@aol.com)

# The Cystinuria Questionnaire

Compiled by Tim Gode

My name is Tim Gode. I was diagnosed with cystinuria about 12 years ago and have been a member of the cystinuria support group for about 3 years. Recently, I decided to conduct a study on ourselves that would answer questions that we, as a group, have about our condition. Many of us were sharing our experiences and providing alternatives to situations that we ran across. This is great but we can do more. A lot of questions have been put out to the group that no single one of us could answer.

For instance, someone once posed the question that there might be a relationship between how we sleep and which kidney our stones form in most often. Many of us responded with our own experience. I form stones only in the right kidney and I toss and turn at night. Other people sleep on one side and wrote that they form stones in both kidneys. So what's the answer? The only way to find out is to gather these facts on everyone and then see if the relationship exists.

Someone else wondered if cystinurics might have a weight problem. The only way to truly answer this is to get everyone's weight, height, and age on a list and see if the data shows our group to be higher than the average for the rest of the population.

Another area we've discussed at length is side effects with our drugs. Many of us have experienced side effects with either the cystine binding drugs or painkillers we use during stone attacks. How does our experience with side effects compare to the medical literature? When the drug pamphlet says there is a 1 in 50 chance of a certain side effect occurring, is that what we've experienced?

Since this information either does not exist or is not available to us, an organized study of ourselves is the only way we'll get answers. A questionnaire was developed using input from all the on-line cystinuria support group members. The result was a form that probes into the lives, habits, and medical histories of ourselves. Over a period of about 2 months, the content of this questionnaire was refined. Since August 5, 1998, gathering information on ourselves is exactly what we've been doing.

I sincerely hope everyone participates. Our information gets better with each additional questionnaire.

What follows is a questionnaire that probes into the lives, habits, and medical histories of the cystinuric. The content of this questionnaire was formulated by the cystinuria support group and is intended to provide information about us so that we may answer questions about ourselves that any single one of us could not.

Please know up front that many of the questions are very personal in nature. If you participate, you will be submitting your answers to a fellow cystinuric who is NOT a physician. It is for this reason that participation is completely optional.

I sincerely hope everyone participates. Our information gets better with each additional questionnaire.

As the conductor of this study and as a fellow cystinuric, my pledge to you is this:

- 1) I will do everything in my power to protect your privacy.
- 2) The information gathered and the results obtained will never be released to insurance companies.
- 3) The results will only be used for the good of cystinurics.

Below is the final version of the Cystinuria Support Network Questionnaire. Revision 7 is the official final version. Make sure you have the correct one before answering.

#### INSTRUCTIONS:

- Do not put your name anywhere on the questionnaire. You will be anonymously and uniquely identified with a number.
- The questionnaire is not as long as it looks. Many sections appear long but require just a few checks.
- Most questions can be answered by placing an 'x' on the blank line provided. Try to use as few "other" options as possible.
- Each questionnaire is intended to be filled out from the point of view of the cystinuric.
- For children too young to answer for themselves, parents should answer for them from the child's perspective.

Mail the completed questionnaire to:  
CSN Study  
c/o Tim Gode  
218 Albania Dr.  
Luling, LA 70070

Thanks to everyone that helped develop the questions.

Tim

## Revision 7 July 31, 1998

THE INFORMATION GIVEN IS STRICTLY CONFIDENTIAL AND IS ONLY TO BE USED IN THIS STUDY.

#### BASIC INFORMATION

Cystinuric Unique ID Number: \_\_\_\_\_ (Don't fill in, this will be assigned.)

Type of Cystinuria:  I  II  III  Don't know

City: \_\_\_\_\_ State: \_\_\_\_\_

ZIP code: \_\_\_\_\_ Country: \_\_\_\_\_

Sex:  M  F

DOB: \_\_\_/\_\_\_/\_\_\_ (m/d/y)

Weight: \_\_\_\_\_ lbs OR \_\_\_\_\_ kilos

Height: \_\_\_\_\_ ft \_\_\_ ins OR \_\_\_ mtrs \_\_\_ cms

Marital Status:  Married  Single

Other: \_\_\_\_\_

Highest education level completed:

- Elementary School
- High School
- Technical School
- College/University - B.A., B.S.
- College/University - MA, MS
- College/University - PhD

Occupation: \_\_\_\_\_

Hobbies: \_\_\_\_\_

#### UROLOGICAL HISTORY

Do you have to urinate frequently?  
 Yes  No  Sometimes

Do you get up at night more than once to urinate?  
 Yes  No  Sometimes

Do you have difficulty in passing your urine?  
 Yes  No  Sometimes

Has your urine stream slowed down?  
 Yes  No  Sometimes

Do you have pain or burning during urination?  
 Yes  No  Sometimes

Do you ever lose urine involuntarily when you cough, sneeze, laugh, getting up from bed or while in a sitting position?  
 Yes  No  Sometimes

Do you ever have the sudden urge to urinate and cannot hold your urine?  Yes  No

Have you ever passed blood in your urine?  
 Yes  No

Have you ever been told you have blood in your urine in a lab test?  Yes  No

#### PAST MEDICAL HISTORY – ADULT AND CHILDHOOD

Have you had: Check the appropriate space(s)

- |  |  |
|--|--|
| <input type="checkbox"/> Heart Disease or Murmur     | <input type="checkbox"/> German Measles            |
| <input type="checkbox"/> High Blood Pressure         | <input type="checkbox"/> Mumps                     |
| <input type="checkbox"/> Diabetes                    | <input type="checkbox"/> Rheumatic Fever           |
| <input type="checkbox"/> Stroke                      | <input type="checkbox"/> Chicken Pox               |
| <input type="checkbox"/> Cancer: _____               | <input type="checkbox"/> Tuberculosis or +TB Test  |
| <input type="checkbox"/> HIV or AIDS                 | <input type="checkbox"/> Pneumonia/ Pleurisy       |
| <input type="checkbox"/> Syphilis                    | <input type="checkbox"/> Arthritis                 |
| <input type="checkbox"/> Gonorrhea                   | <input type="checkbox"/> Asthma                    |
| <input type="checkbox"/> Chlamydia                   | <input type="checkbox"/> Kidney Disease            |
| <input type="checkbox"/> Skin Disorder               | <input type="checkbox"/> Kidney/Bladder Infection  |
| <input type="checkbox"/> Liver Problems              | <input type="checkbox"/> Kidney Stone              |
| <input type="checkbox"/> Hemorrhoids                 | <input type="checkbox"/> Gout                      |
| <input type="checkbox"/> Intestinal Disorders        | <input type="checkbox"/> Blood transfusion         |
| <input type="checkbox"/> Back Trouble                | <input type="checkbox"/> Hay Fever                 |
| <input type="checkbox"/> Ruptured Disc/Sciatica      | <input type="checkbox"/> Chronic Bronchial Trouble |
| <input type="checkbox"/> Vein Trouble                | <input type="checkbox"/> Jaundice or Hepatitis     |
| <input type="checkbox"/> Blood Disease or Anaemia    | <input type="checkbox"/> Gallbladder Disease       |
| <input type="checkbox"/> Bleeding Tendency           | <input type="checkbox"/> Goiter or Thyroid Trouble |
| <input type="checkbox"/> Stomach or Duodenal Ulcer   | <input type="checkbox"/> Nervous Disorder          |
| <input type="checkbox"/> Concussion/Head Injury      | <input type="checkbox"/> Irritable Bowel Syndrome  |
| <input type="checkbox"/> Serious accident or injury? |  |

If so, how and when? \_\_\_\_\_

Broken bones \_\_\_\_\_

(Which?) \_\_\_\_\_

Other illness or disease not listed: \_\_\_\_\_

### SOCIAL HISTORY

Have you ever smoked?  Yes  No

Currently If yes, how long? \_\_\_\_\_ Packs/day

Alcohol:

Never  Rare  Occasional

Moderate  Often

### IMMEDIATE FAMILY HISTORY

Does/did your mother have cystinuria?

Yes  No  Don't know

Mothers date of birth: \_\_\_\_/\_\_\_\_/\_\_\_\_ MM/DD/YY

Does/did your father have cystinuria?

Yes  No  Don't know

Father's date of birth: \_\_\_\_/\_\_\_\_/\_\_\_\_ MM/DD/YY

Do you have children?  Yes  No

If yes, how many do you have? # of girls: \_\_\_\_ # of boys: \_\_\_\_

How many of these are cystinuric?# of girls: \_\_\_\_ # of boys: \_\_\_\_

Do you have any siblings?  Yes  No

# of brothers w/cystinuria \_\_\_\_

# of brothers w/o cystinuria \_\_\_\_

# of sisters w/cystinuria \_\_\_\_

# of sisters w/o cystinuria \_\_\_\_

Do you have any other relatives you know have cystinuria?

Yes  No  Don't know

If yes, please list relationship to you, approximate age, and sex:

Relationship	Age	Gender
_____	_____	<input type="checkbox"/> M <input type="checkbox"/> F
_____	_____	<input type="checkbox"/> M <input type="checkbox"/> F
_____	_____	<input type="checkbox"/> M <input type="checkbox"/> F

Do any of your relatives form non-cystine stones?

Yes  No  Don't know

If yes, list relationship, approximate age, sex, and type of stones:

Relationship	Age	Gender	Type of stones
_____	_____	_____	_____

\_\_\_\_\_  M  F  
 \_\_\_\_\_  M  F  
 \_\_\_\_\_  M  F  
 \_\_\_\_\_

### ETHNIC BACKGROUND

Check all that apply.

North American/Caribbean

<input type="checkbox"/> Canadian	<input type="checkbox"/> French Canadian
<input type="checkbox"/> American	<input type="checkbox"/> Black American
<input type="checkbox"/> Mormon	<input type="checkbox"/> Native American
<input type="checkbox"/> Mexican	<input type="checkbox"/> Nahua
<input type="checkbox"/> West Indian	<input type="checkbox"/> Cuban
<input type="checkbox"/> Haitian	<input type="checkbox"/> Puerto Rican
<input type="checkbox"/> Other: _____	

Central/South America

Please specify: \_\_\_\_\_

Europe

<input type="checkbox"/> Celt	<input type="checkbox"/> Irish	<input type="checkbox"/> Scottish
<input type="checkbox"/> Welsh	<input type="checkbox"/> English	<input type="checkbox"/> Norwegian
<input type="checkbox"/> Swedish	<input type="checkbox"/> Finnish	<input type="checkbox"/> Danish
<input type="checkbox"/> French	<input type="checkbox"/> Belgian	<input type="checkbox"/> Dutch
<input type="checkbox"/> German	<input type="checkbox"/> Swiss	<input type="checkbox"/> Austrian
<input type="checkbox"/> Slav	<input type="checkbox"/> Polish	<input type="checkbox"/> Czechoslovak
<input type="checkbox"/> Croatian	<input type="checkbox"/> Slovakian	<input type="checkbox"/> Hungarian
<input type="checkbox"/> Romanian	<input type="checkbox"/> Bulgarian	<input type="checkbox"/> Albanian
<input type="checkbox"/> Gypsy	<input type="checkbox"/> Portuguese	<input type="checkbox"/>
<input type="checkbox"/> Spaniard		
<input type="checkbox"/> Galician	<input type="checkbox"/> Basque	<input type="checkbox"/> Catalan
<input type="checkbox"/> Andalusian	<input type="checkbox"/> Sard	<input type="checkbox"/> Italian
<input type="checkbox"/> Sicilian	<input type="checkbox"/> Macedonian	<input type="checkbox"/> Greek
<input type="checkbox"/> Other: _____		

Middle East/North Africa

Please specify: \_\_\_\_\_

Africa

Please specify: \_\_\_\_\_

Central/Northern Asia

<input type="checkbox"/> Baltic People	<input type="checkbox"/> Russian	<input type="checkbox"/>
<input type="checkbox"/> Ukranian		
<input type="checkbox"/> Tartar	<input type="checkbox"/> Bashkir	<input type="checkbox"/> Daghestani
<input type="checkbox"/> Georgian	<input type="checkbox"/> Armenian	<input type="checkbox"/> Turkmen
<input type="checkbox"/> Uzbek	<input type="checkbox"/> Kazakh	<input type="checkbox"/> Tadjik
<input type="checkbox"/> Kirghiz	<input type="checkbox"/> Mongolian	
<input type="checkbox"/> Other: _____		

India/South Central Asia

Please specify: \_\_\_\_\_

China/East and South-East Asia

Please specify: \_\_\_\_\_

Australasia/Pacific

<input type="checkbox"/> Australian	<input type="checkbox"/> New Zealander	<input type="checkbox"/> Papua New Guinea
<input type="checkbox"/> Melanesian		
<input type="checkbox"/> Other: _____		

Other: \_\_\_\_\_

Don't know

## DIAGNOSIS

Age diagnosed with cystinuria: \_\_\_\_\_  
years

Number of times mis-diagnosed.  
\_\_\_ None \_\_\_1 \_\_\_2 \_\_\_3 \_\_\_4 More: \_\_\_

Age of first stone attack:  
\_\_\_\_\_ years \_\_\_ Haven't had one yet

## KIDNEY STATUS

Number of kidneys remaining:

- I still have both
- I have my right kidney only (lost left to cystinuria)
- I have my left kidney only (lost right to cystinuria)
- I was born with only a right kidney
- I was born with only a left kidney
- Right kidney normal, left kidney horseshoe (or other abnormality)
- Left kidney normal, right kidney horseshoe (or other abnormality)
- Both kidneys horseshoe (or other abnormality)
- I lost both to cystinuria
- I lost one or both to other causes

Explain: \_\_\_\_\_

Kidney function:

Right: \_\_\_\_\_% \_\_\_ Don't know  
Left: \_\_\_\_\_% \_\_\_ Don't know

Typical cystine output: \_\_\_\_\_ mg/liter

Note: We are looking for an untreated (not even hydration) cystine output per 24 hours. This would have to be measured right after diagnosis OR if you have a recent 24 hour test result, it can be approximated by dividing the 24 hour output by what a non-cystinuric person would put out (about 1.5 litres)

Approximate number of stones formed per year:

\_\_\_\_\_/year \_\_\_ Don't know

Typical size of stones:

- I get a little gravel
- I get lots of gravel
- I get gravel and large stones
- I get a few large stones
- I get lots of large stones

Largest stone ever formed: \_\_\_\_\_ (cm) or \_\_\_\_\_ (mm)

What is the longest period (years, months) that you have been stone free since you were diagnosed?

\_\_\_\_\_ years \_\_\_\_\_ months

Typical stone composition:

- Pure cystine
- Cystine and Calcium Oxalate % cystine \_\_\_\_\_  
%calcium \_\_\_\_\_
- Cystine and Uric Acid % cystine \_\_\_\_\_ % uric acid
- Other \_\_\_\_\_
- \_\_\_\_\_
- Don't know

Stone location: My stones form...

- always in the right kidney
- mostly in the right kidney
- in both kidneys equally
- mostly in the left kidney

\_\_\_ always in the left kidney

## ROUTINE MONITORING

I visit my doctor regularly. \_\_\_Yes \_\_\_No  
If yes, how frequently? \_\_\_\_\_

While at the doctor I get (check all that apply):

- Ultrasound \_\_\_\_\_ Always \_\_\_\_\_
- Sometimes
- KUB X-ray \_\_\_\_\_ Always \_\_\_\_\_
- Sometimes
- IVP X-ray \_\_\_\_\_ Always \_\_\_\_\_
- Sometimes
- Blood drawn for tests \_\_\_\_\_ Always \_\_\_\_\_ Sometimes
- Urine Ph test \_\_\_\_\_ Always \_\_\_\_\_
- Sometimes
- Urine tested for blood \_\_\_\_\_ Always \_\_\_\_\_
- Sometimes
- Other: \_\_\_\_\_

I routinely monitor at home:

- Urine for Ph
- Urine for blood
- Leukocytes
- Other: \_\_\_\_\_

## SLEEPING PATTERNS

I sleep:

- only on my back \_\_\_\_\_ mostly on my back
- only on my right side \_\_\_\_\_ mostly on my right side
- mostly on my left side \_\_\_\_\_ only on my left side
- mostly on my stomach \_\_\_\_\_ only on my stomach
- I toss and turn \_\_\_\_\_ I don't know how I sleep

## CURRENT TREATMENT

Check all that apply.

- High fluids
- Diet
  - Vegetarian
  - Low methianine
  - Low sodium
  - Other \_\_\_\_\_
- Cystine binding drugs
  - Tiopronin(Thiola) Dosage/day: \_\_\_\_\_ mg
  - Penicillamine(Depen, Cuprimine) Dosage/day: \_\_\_\_\_ mg
  - Other \_\_\_\_\_ Dosage/day: \_\_\_\_\_units \_\_\_\_\_
- Alkalyzers
  - Sodium Bicarb Dosage/day: \_\_\_\_\_ mg
  - Potassium Citrate
    - Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_
    - Urocit-K Dosage/day: \_\_\_\_\_ units \_\_\_\_\_
    - Polycitra-K Dosage/day: \_\_\_\_\_ units \_\_\_\_\_
  - Sodium Citrate & Citric Acid
    - Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_
    - Bicitra Dosage/day: \_\_\_\_\_ units \_\_\_\_\_
    - Polycitra Dosage/day: \_\_\_\_\_ units \_\_\_\_\_

Citrolith Dosage/day: \_\_\_\_\_  
 units \_\_\_\_\_  
 Tricitrates Dosage/day: \_\_\_\_\_  
 units \_\_\_\_\_  
 Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Acetyl Sulfoxasole  
 Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Gantrisin Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 units \_\_\_\_\_  
 Blood pressure drugs(also used as diuretics)  
 Captopril  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Capoten Dosage/day: \_\_\_\_\_ mg  
 Capozide Dosage/day: \_\_\_\_\_ mg  
 Clonidine  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Catapres-TTS Dosage/day: \_\_\_\_\_ mg  
 Propranolol Hydrochloride  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Inderal Dosage/day: \_\_\_\_\_ mg  
 Inderide Dosage/day: \_\_\_\_\_ mg  
 Metoprolol Tartrate  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Lopressor Dosage/day: \_\_\_\_\_ mg  
 Furosemide  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Lasix Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ mg  
 Heartburn drugs  
 Propulsid  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Cisapride Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ mg  
 Vitamins  
 B-6 Dosage/day: \_\_\_\_\_ mg  
 C Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ mg  
 Bladder instability control  
 Oxybutynin Chloride  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Ditropan Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ units \_\_\_\_\_

**HISTORY WITH TREATMENTS:**

Treatments tried in the past: Check all that apply.

High fluids  
 Diet  
 Vegetarian  
 Low methianine  
 Low sodium  
 Other  
 Cystine binding drugs  
 Tiopronin(Thiola) Dosage/day: \_\_\_\_\_ mg  
 Penicillamine(Depen, Cuprimine) Dosage/day: \_\_\_\_\_ mg  
 mg  
 Other \_\_\_\_\_ Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Alkalyzers  
 Sodium Bicarb Dosage/day: \_\_\_\_\_ mg  
 Potassium Citrate

Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Urocit-K Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Polycitra-K Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Sodium Citrate & Citric Acid  
 Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Bicitra Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Polycitra Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Citrolith Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Tricitrates Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Acetyl Sulfoxasole  
 Generic Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Gantrisin Dosage/day: \_\_\_\_\_ units \_\_\_\_\_  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 units \_\_\_\_\_  
 Blood pressure drugs(also used as diuretics)  
 Captopril  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Capoten Dosage/day: \_\_\_\_\_ mg  
 Capozide Dosage/day: \_\_\_\_\_ mg  
 Clonidine  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Catapres-TTS Dosage/day: \_\_\_\_\_ mg  
 Propranolol Hydrochloride  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Inderal Dosage/day: \_\_\_\_\_ mg  
 Inderide Dosage/day: \_\_\_\_\_ mg  
 Metoprolol Tartrate  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Lopressor Dosage/day: \_\_\_\_\_ mg  
 Furosemide  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Lasix Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 mg  
 Heartburn drugs  
 Propulsid  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Cisapride Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 mg  
 Vitamins  
 B-6 Dosage/day: \_\_\_\_\_ mg  
 C Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 mg  
 Bladder instability control  
 Oxybutynin Chloride  
 Generic Dosage/day: \_\_\_\_\_ mg  
 Ditropan Dosage/day: \_\_\_\_\_ mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_  
 mg  
 Other: \_\_\_\_\_ Dosage/day: \_\_\_\_\_ units \_\_\_\_\_

**WATER**

How much do you actually drink per 24 hours?  
 \_\_\_\_\_ Liters \_\_\_ Don't know  
 How much do you think you should drink per day?  
 \_\_\_\_\_ Liters \_\_\_ Don't know  
 How much did your doctor say you should drink per day?  
 \_\_\_\_\_ Liters \_\_\_ Didn't advise  
 Do you drink something other than water?  
 Yes  No  
 If yes, specify: \_\_\_\_\_

Do you regularly drink water at night?  Yes  No  
If yes, how many times/night? \_\_\_\_\_ how much?  
\_\_\_\_\_ oz

### DIET

If vegetarian, what type?

- Vegan-Strict vegetarians who eat only foods from plant sources
- Lactovegetarian-Eat plant foods plus dairy products
- Ovovegetarian-Eat plant foods plus eggs
- Lacto-ovovegetarian-Eat both dairy products and eggs
- Fruitarian-Eat fruits, nuts, olive oil and honey
- Semivegetarian-Eat chicken and fish but no red meat
- Pescovegetarian-Eat fish but no other meat products
- Pollovegetarian-Eat poultry but no red meat
- Other(please explain): \_\_\_\_\_

How successful do feel you your diet has been in controlling stone formation?

- Very successful
- Pretty good
- Average
- Not so good
- Unsuccessful
- Don't know

Have you seen a measurable reduction in stone formation due to diet therapy?

Yes  No  Don't know

If low sodium, what is your daily limit?

- \_\_\_\_\_ mg
- I just try to minimize by not adding salt to anything
- I make sure everything I eat is as sodium free as possible

### THIOLA

(also known as Tiopronin, Mercaptopropionyl)

Tried it?  Yes  No  
If yes, date \_\_\_\_\_ started? \_\_\_\_\_ ended?  
 Currently use

Had a reaction?  Yes  No  Don't know

If yes, what type? check all that apply

- Gastrointestinal-nausea, emesis(throwing up), diarrhea, anorexia, abdominal pain, bloating, flatus(gas)
- Impaired taste and smell
- Dermatologic complications-pharyngitis(sore throat), oral ulcers, rash, ecchymosis(bruising), prurites(itching), urticaria(skin disorder marked by raised, swollen patches of skin with intense itching), warts, skin wrinkling, pemphigus(large blisters on skin or

mucous

membranes with itching or burning), elastosis perforans serpiginosa

Hypersensitivity reactions-laryngeal edema(swollen larynx),

dyspnea(difficult or labored breathing), respiratory distress, fever, chills, arthralgia(pain in joints), weakness, fatigue, myalgia(muscle pain),

adenopathy

Hematologic abnormalities-increased bleeding, anemia, leukopenia(low blood leukocytes), thrombocytopenia(persistent decrease in blood platelets), eosinophilia

Renal complications-proteinuria, nephrotic syndrome, hematuria

Pulmonary manifestations-bronchiolitis, hemoptysis, pulmonary infiltrates, dyspnea(difficult or labored breathing)

Neurologic complications-myasthenic syndrome

Dizziness

Other: Please describe:  
\_\_\_\_\_

If you had a reaction, were you able to reduce dosage and gradually build up again?  Yes  No  Didn't try

If you had a reaction to Thiola, did you switch to D-pen?  
 Yes  No

If you switched, was it successful?  
 Yes  No

### D-PENICILLAMINE:

(also known as Cuprimine, Depen, Penacillamine)

Tried it?  Yes  No  
If yes, date \_\_\_\_\_ started? \_\_\_\_\_ ended?

Currently use

Had a reaction?  Yes  No  Don't know

If yes, what type? check all that apply

- Gastrointestinal-nausea, emesis(throwing up), diarrhea, anorexia, abdominal pain, bloating, flatus(gas) Impaired taste and smell
- Dermatologic complications-pharyngitis(sore throat), oral ulcers, rash, ecchymosis(bruising), prurites(itching), urticaria(skin disorder marked by raised, swollen patches of skin with intense itching), warts, skin wrinkling, pemphigus(large blisters on skin or

mucous

membranes with itching or burning), elastosis perforans serpiginosa

Hypersensitivity reactions-laryngeal edema(swollen larynx),

dyspnea(difficult or labored breathing), respiratory distress, fever, chills, arthralgia(pain in joints), weakness, fatigue, myalgia(muscle pain),

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Hematologic abnormalities-increased bleeding, anemia, leukopenia(low blood leukocytes), thrombocytopenia(persistent decrease in blood platelets), eosinophilia

Renal complications-proteinuria, nephrotic syndrome, hematuria

Pulmonary manifestations-bronchiolitis, hemoptysis, pulmonary infiltrates, dyspnea(difficult or labored breathing)

Neurologic complications-myasthenic syndrome

Dizziness

Other: Please describe:  
\_\_\_\_\_

If you had a reaction, were you able to reduce dosage and gradually build up again?  Yes  No  Didn't try

If you had a reaction to D-pen, did you switch to Thiola?  
 Yes  No

If you switched, was it successful?  
 Yes  No

### ALKALIZERS

(such as Urocit-K, Polycitra, Polycitra-K, Sodium Bicarb, etc.)

What Ph do you shoot for? \_\_\_\_\_

### SURGICAL HISTORY

Check all that you've had.

	No. treatments	% successful
<input type="checkbox"/> Ultrasonic Lithotripsy	_____	_____
<input type="checkbox"/> ESW Lithotripsy	_____	_____
<input type="checkbox"/> EHL Lithotripsy	_____	_____
<input type="checkbox"/> Percutaneous Lithotripsy	_____	_____
_____		
<input type="checkbox"/> Holmium Laser	_____	_____
<input type="checkbox"/> Cystoscopy with basket removal	_____	_____
<input type="checkbox"/> Nephrectomy	_____	_____
<input type="checkbox"/> Nephrostomy	_____	_____
<input type="checkbox"/> Pyelolithotomy	_____	_____
<input type="checkbox"/> Stent	_____	_____
<input type="checkbox"/> Cystolithopyxy	_____	_____
<input type="checkbox"/> Prostate Biopsy & Surgery	_____	_____
_____		
<input type="checkbox"/> Incontinence Surgery	_____	_____
<input type="checkbox"/> Other	_____	_____

Surgery: \_\_\_\_\_

**PAIN CONTROL**

Check all that you've had at home and in hospital.

These are listed as: generic name (brand names)

<input type="checkbox"/> Morphine Sulfate		
<input type="checkbox"/> Generic		<input type="checkbox"/> Kadian
<input type="checkbox"/> MS Contin		<input type="checkbox"/> MSIR Oral
<input type="checkbox"/> Oramorph SR		<input type="checkbox"/> RMS
Suppositories	<input type="checkbox"/> MS/S Suppositories)	<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Hydromorphone Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Dilaudid
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Meperidine Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Demero
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Hydrocodone Bitartrate		
<input type="checkbox"/> Generic		<input type="checkbox"/> Hycodan
<input type="checkbox"/> Hycomine		<input type="checkbox"/> Hydrocet
<input type="checkbox"/> Cortab		<input type="checkbox"/> Norco
<input type="checkbox"/> Tussend		<input type="checkbox"/> Vicodin
<input type="checkbox"/> Vicodin-ES		<input type="checkbox"/> Vicodin-HP
<input type="checkbox"/> Vicoprofen		<input type="checkbox"/> Zydone
<input type="checkbox"/> Ceta Plus		<input type="checkbox"/> Co-
Gesic		
<input type="checkbox"/> Endal-HD		<input type="checkbox"/> Lorcet
<input type="checkbox"/> Lorcet-HD		<input type="checkbox"/> Lorcet Plus
<input type="checkbox"/> Lortab 2.5/500		<input type="checkbox"/> Lortab 5/500
<input type="checkbox"/> Lortab 7.5/500		<input type="checkbox"/> Lortab 10/500
<input type="checkbox"/> Lortab-ASA		<input type="checkbox"/> Tussafed-HC
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Oxycodone Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Percocet
<input type="checkbox"/> Oxycontin		<input type="checkbox"/> Oxy IR
<input type="checkbox"/> Roxicodone		<input type="checkbox"/> Tylox
<input type="checkbox"/> Endocet		<input type="checkbox"/>
Endodan		
<input type="checkbox"/> Roxicet		<input type="checkbox"/> Roxilox
<input type="checkbox"/> Roxiprin		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Oxycodone Terephthalate		
<input type="checkbox"/> Generic		<input type="checkbox"/> Percodan
<input type="checkbox"/> Percodan-Demi		<input type="checkbox"/> Endodan
<input type="checkbox"/> Roxiprin		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Buprenorphine Hydrochloride		

<input type="checkbox"/> Generic		<input type="checkbox"/> Temgesic
<input type="checkbox"/> Buprenex		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Pethidine Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Pethidine
<input type="checkbox"/> Pamergan		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Meptazinol		
<input type="checkbox"/> Generic		<input type="checkbox"/> Meptid
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Propoxyphene Napsylate		
<input type="checkbox"/> Generic		<input type="checkbox"/> Darvocet N-50
<input type="checkbox"/> Darvocet N-100		<input type="checkbox"/> Darvon-N
<input type="checkbox"/> Propacet 100		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Propoxyphene Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Darvon
<input type="checkbox"/> Wygesic		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Diclofenac Sodium		
<input type="checkbox"/> Generic		<input type="checkbox"/> Voltaren
<input type="checkbox"/> Voltaren-XR		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Diclofenac Potassium		
<input type="checkbox"/> Generic		<input type="checkbox"/> Cataflam
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Codeine Phosphate		
<input type="checkbox"/> Generic		<input type="checkbox"/> Brontex
<input type="checkbox"/> Fioricet w/codeine		<input type="checkbox"/> Fiorinal w/codeine
<input type="checkbox"/> Nucofed		<input type="checkbox"/> Soma
Compound		<input type="checkbox"/> w/codeine
<input type="checkbox"/> Tylenol w/codeine		<input type="checkbox"/> Butalbital
<input type="checkbox"/> Fiorital w/codeine		<input type="checkbox"/> Phenaphen
		<input type="checkbox"/> w/codeine
<input type="checkbox"/> Paracodal		<input type="checkbox"/>
CoProxamol		
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Acetaminophen (ex: Tylenol)		
<input type="checkbox"/> Promethazine Hydrochloride		
<input type="checkbox"/> Generic		<input type="checkbox"/> Phenergan
<input type="checkbox"/> Phenergan Suppositories		<input type="checkbox"/> Other: _____
_____		
<input type="checkbox"/> Dihydrocodeine Bitartrate		
<input type="checkbox"/> Generic		<input type="checkbox"/> DHC plus
<input type="checkbox"/> Synalgos-DC		<input type="checkbox"/> DF 118
<input type="checkbox"/> Other: _____		
_____		
<input type="checkbox"/> Gas & Air		
<input type="checkbox"/> Entonox		<input type="checkbox"/>
Other: _____		
_____		
<input type="checkbox"/> Fentanyl		
<input type="checkbox"/> Generic		<input type="checkbox"/> Duragesic
<input type="checkbox"/> Other: _____		
_____		
Other: _____		
_____		
Which pain control did you like best? _____		
Why? _____		
_____		
Which pain control did you like the least? _____		
_____		

Why?

\_\_\_\_\_

Side effects experienced with pain killers:

- Nausea
- Insomnia
- Rash
- Drowsiness
- Allergic reaction
- Constipation
- Nervousness
- Other:

\_\_\_\_\_

**FOR WOMEN ONLY**

Medical History:

Female

Operations: \_\_\_\_\_

Pelvic Inflammatory Disease

Other: \_\_\_\_\_

Are you still having menstrual periods?  Yes

No

Have you found your menstrual cycles coincide with forming/passing stones?  Yes  No  Don't know

Are you taking birth control medication?  Yes  No

If yes, specify

type: \_\_\_\_\_

This section is only for women with cystinuria who have been pregnant.

How long did it take to conceive?

First Pregnancy \_\_\_\_\_ years \_\_\_\_\_ months  Don't know

Second Pregnancy \_\_\_\_\_ years \_\_\_\_\_ months

Don't know

Third Pregnancy \_\_\_\_\_ years \_\_\_\_\_ months  Don't know

Fourth Pregnancy \_\_\_\_\_ years \_\_\_\_\_ months  Don't know

Fifth Pregnancy \_\_\_\_\_ years \_\_\_\_\_ months  Don't know

Generally, did you find pregnancy:

Reduced stone formation.

Increased stoned formation.

No change

Don't know

Generally, did you find pregnancy:

Reduced passing frequency

Increased passing frequency

No change

Don't know

Generally, did you find pregnancy:

Reduced kidney function

Increased kidney function

No change

Don't know

Did you have complications during pregnancy?

Yes  No

If yes, please list:

Urinary Tract Infection  Passed stones

Hypertension  Proteinuria

Ureteric Colic

Other: \_\_\_\_\_

Did you have any miscarriages?

Yes  No  Don't know

If yes, how many? \_\_\_\_\_

Did you breastfeed?

Yes  No

If yes, did breastfeeding affect your cystinuria?

Yes  No

If yes, in what way? \_\_\_\_\_

**FOR MEN ONLY**

Medical History:

Vasectomy  Trauma to Groin/Testicles  Prostate Trouble

Have you ever had difficulty in obtaining or maintaining an erection?  Yes  No

Have you ever had painful ejaculations?

Yes  No

Have you ever seen blood in your sperm?

Yes  No

Have you ever fathered children?

Yes  No

Have you ever had a discharge from the penis?

Yes  No

Thank you for taking the time to complete the questionnaire.

# Membership Form

If you are interested in participating in the Cystinuria Support Network and have not already done so, please fill out the following form and send it to:

CSN c/o Jann Ledbetter  
21001 NE 36th Street  
Redmond WA 98053

Name \_\_\_\_\_

Address \_\_\_\_\_

City, State, Zip \_\_\_\_\_

Phone \_\_\_\_\_ Best time \_\_\_\_\_

Additional Patient Information:

Sex \_\_\_\_\_ Date of Birth \_\_\_\_\_

Number of Years since diagnosed \_\_\_\_\_

Current Medications \_\_\_\_\_

Surgeries and other procedures you've experienced \_\_\_\_\_

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

Other information you would like to share \_\_\_\_\_

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

How did you hear about CSN? \_\_\_\_\_