

# **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 advise 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 persons 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 heamocrit, kidney function and liver function. When a cystinuric first starts to take Penicillamine, it is generally started slowly and gradually increased to minimise 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 drugs 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 is 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:**

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