

What can you do?

- **Drink LOTS of water.**

It is recommended that you drink at least 3 litres or more per day. It is important that your urine does not become concentrated at night. You will have to drink plenty before bed and when you get up to urinate.

- **Follow a methionine-free diet.**

Methionine is the amino acid from which cystine is formed and is found in animal protein. Therefore try to reduce the amount of animal protein in your diet. This includes meat, fish, eggs and dairy products but it is important that you replace these with plenty of green vegetables, nuts, beans and pulses. (The hospital can organize dietary advice.)

- **Stones dissolve in alkaline urine -** you can take potassium citrate and monitor the pH (acidity) of your urine with dipsticks. The diet above also helps achieve this.

- **Lower your salt intake.**

Contact information

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International Cystinuria Foundation

www.cystinuria.org

Your local Pharmacist

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The Trust provides free monthly health talks on a variety of medical conditions and treatments. For more information visit www.uhb.nhs.uk or call 0121 627 7803

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A large green rectangular graphic with a white circular logo at the top left. The logo consists of three overlapping white circles. Below the logo, the word 'Cystinuria' is written in large white font. Underneath, in smaller white font, is the text: 'A leaflet for patients with Cystinuria; cause, diagnosis, treatments and self-help measures.'

Cystinuria

A leaflet for patients with Cystinuria; cause, diagnosis, treatments and self-help measures.

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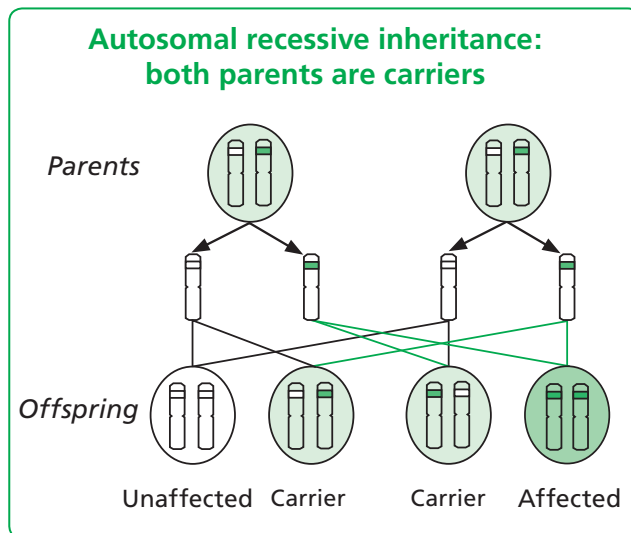
What is Cystinuria?

Cystinuria is a rare condition affecting the kidney. It is characterized by the excess collection of amino acids in the urine (most notably cystine) which are insoluble and therefore form hard, crystallised stones in the urine.

It affects approximately 1:7,000 and is usually diagnosed between 10 and 30 years-of-age.

How is it caused?

Cystinuria is a genetic condition. It has a recessive inheritance pattern-needing a copy of the altered gene to be inherited from both parents. This leads to the condition tending to 'appear' without other members of the family having the same condition.



Symptoms

Renal colic is often the first sign (moderate to severe sharp, spasmodic pain in the back, side and groin area.) This pain is caused by a kidney stone falling into the ureter (tube that joins kidney to bladder). It can sometimes be linked with urinary infection (cystitis) and bladder problems.

Patients can also report feeling a constant ache in the kidney area.

Tiredness and depression can be associated with Cystinuria.

Stones can cause obstruction and pain should be investigated as blockage can cause problems with infection and kidney damage. Blockage can also cause the kidneys to become enlarged-hydronephrosis.

How we diagnose Cystinuria

- 24 hour urine collection to test levels of cystine
- Spot urine for initial detection of cystine
- Genetic tests can be undertaken

Finding stones

Stones can be detected using the following:

- KUB (X-ray of the kidneys, ureter and bladder)
- Ultrasound

- CT (body scan) scan
- Nephrostogram (X-ray with introduction of dye into the kidney via a nephrostomy tube where a patient already has one inserted)

Medical treatment

D-Penicillamine – This drug combines with cystine to prevent stones forming.

Side effects include taste disturbance, rash and nausea. Other risks are rarer and you would be monitored closely.

Captopril – Used if the patient is intolerant to the above again with the aim of reducing stone formation this can also cause taste disturbance, postural hypotension and cough.

Tipronin (Thiola) – Unfortunately this drug is not yet available in the UK.

Surgical treatment for stones

- Open or keyhole surgery. Open surgery requires a general anaesthetic and a 10-14 day hospital stay
- Cystoscopy
- Laser Therapy of several types

Risks of surgery should be discussed with your surgeon.