

What I tell my patients about haemolytic uraemic syndrome

Haemolytic uraemic syndrome (HUS) is a rare condition, which can affect people of all ages, but usually affects babies and young children. There are different kinds of HUS, but the most common kind happens following diarrhoea caused by a bacterium called *Escherichia coli* (*E coli*). This bacterium releases a harmful substance, called verotoxin, into the bloodstream. It is verotoxin that causes the collection of problems which we call haemolytic uraemic syndrome.

Haemolytic

This refers to the abnormal breakdown of red blood cells. Your child may appear very pale because the red blood cells are being broken down and this will cause anaemia. Blood transfusions may be required.

The number of cells that help the blood to clot (platelets) are also reduced during the illness. This may result in easy bruising.

Uraemic

The verotoxin also attacks the kidneys, preventing them from carrying out their normal function of getting rid of waste products and fluid. One of these waste products is called urea – the term uraemic means too much urea in the blood. If urea and other waste products build up in the bloodstream because the kidneys are unable to remove them, your child may feel listless, sleepy and sick. Other waste products include potassium and phosphate. The kidneys also remove excess water from the urine. If they are not working properly, the amount of urine produced may be vastly reduced and your child may start to look puffy, often in the face and legs. When the kidneys are unable to get rid of enough waste products and water, this is called kidney failure.

How is it treated?

It is difficult to predict the progression of your child's HUS. In some children it is mild and in others severe. If it is severe and your child is not passing urine then dialysis may be required. You should expect your child to be in hospital for two to three weeks if dialysis is needed but some children will get better more quickly than others, while some will take much longer. The doctors and nursing staff looking after your child should

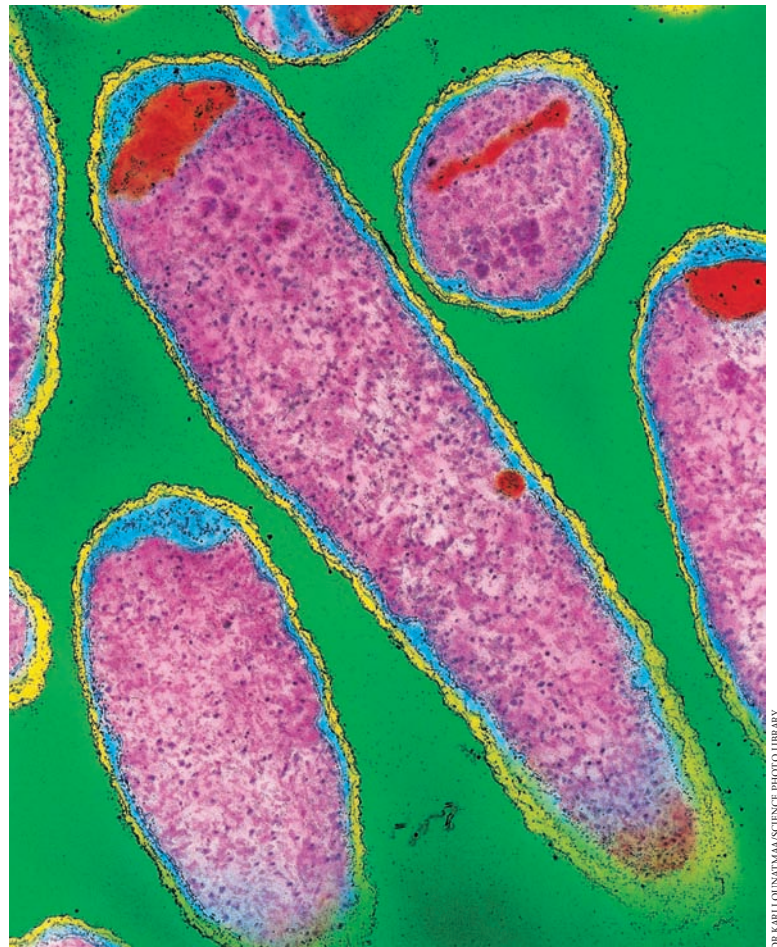
keep you informed about your child's progress day-by-day. Most children with HUS will need blood transfusions to treat the anaemia. However, the low platelet numbers do not usually need treating.

Because the kidneys are not able to remove potassium and phosphate, your child may be referred to a dietitian who will give you advice about foods containing a lot of potassium or phosphate. Your child will need to avoid these foods while the kidneys are not working properly. The dietitian may also prescribe a high calorie drink or feed for your child. Sometimes, if a child has a very poor appetite during the illness, a small tube (a nasogastric tube) is placed into the stomach through the nose. The high calorie feed can then be put through this.

Another main function of the kidneys is to remove excess water. While they are not working

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*Haemolytic uraemic syndrome is caused by the bacterium *Escherichia coli**





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ROBERT HARDING

Peritoneal dialysis is usually carried out by placing a soft plastic tube into the stomach

properly, the amount of fluid your child is allowed to drink may be limited.

Dialysis may be required if levels of waste products in the blood become too high, or if the kidneys cannot get rid of enough water. Dialysis is usually carried out by placing a soft plastic tube into the stomach while your child is sedated. Fluid is then run in and out, using a dialysis machine. This is called peritoneal dialysis. Sometimes haemodialysis is used. This involves placing a plastic tube into a large blood vessel and using a different type of dialysis machine to clean the blood. These procedures will be explained in more detail if your child needs dialysis.

Unfortunately, the only way we can tell how well the kidneys are working and what type of dialysis is needed is by carrying out blood tests. Usually these have to be taken once or twice a

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day at the beginning of the illness in order to monitor your child's condition and to help to make decisions about what treatment is needed. As your child's condition improves, the blood tests are needed less often.

How did it happen?

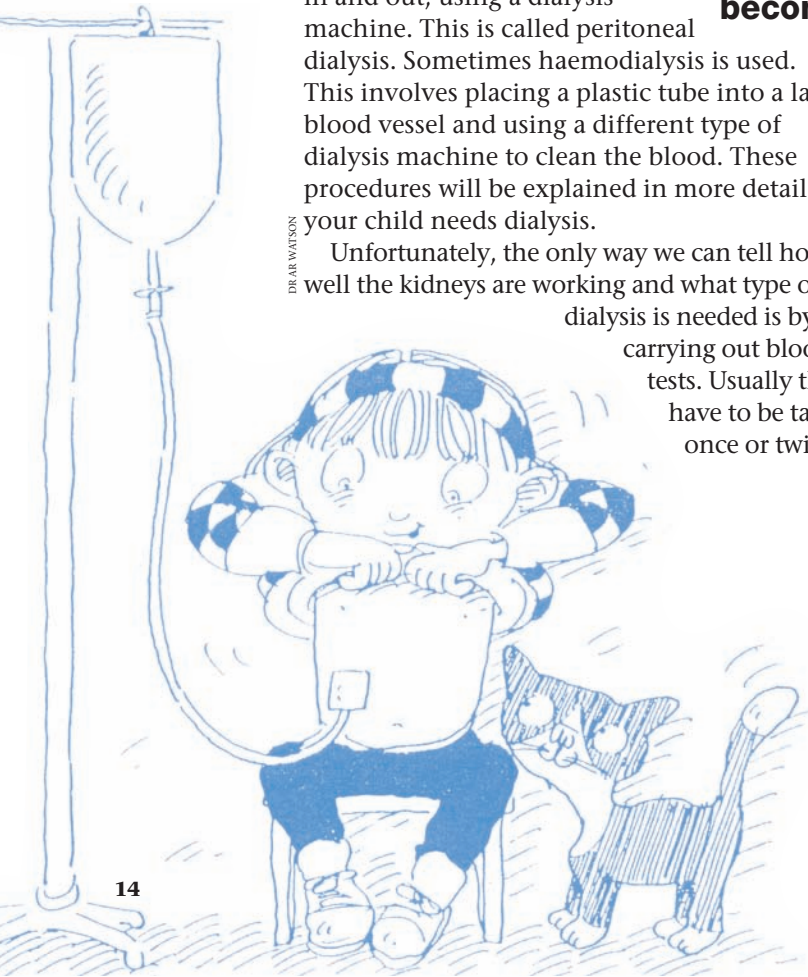
The strains of *E coli* that produce verotoxin generally infect cattle, so uncooked beefburgers and unpasteurised milk are common sources of the infection – although other foods can be contaminated. However, HUS has been associated with all kinds of food, including other meats, fresh seafood, and even vegetables. As the disease may be spread by food, if your job involves the preparation or serving of food, please inform the doctor on the ward and ask for his or her advice.

It has also been shown that the *E coli* can be picked up from contaminated water in swimming pools and the sea.

Very occasionally, other children in the same area or playgroup are affected. If other children in the family, or others with whom your child has been playing, become unwell then they should be seen by a doctor, or the hospital staff informed.

What's the long-term outlook?

Most children seem to recover completely from HUS. However, the particular form of HUS caused by *E coli* has only been recognised for the past 20 years or so and we do not know exactly how well the kidneys will be working in 40 to 50 years' time. Until we have a clearer picture, we are





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CHRISTIA STADLER/PHOTOFUSION

continuing to monitor all children who have had HUS, even if dialysis was not required, unless the disease was particularly mild. The majority of children do very well and we can confirm that the kidneys are continuing to work normally.

In a few cases, however, the kidney function gets worse over time and treatment or more frequent follow-ups may be needed. The normal follow-up is individualised for each paediatric nephrology unit in the country.

In Nottingham, we recommend a basic kidney assessment once a year after the illness. At these visits, your child will have their blood pressure measured and a sample of urine checked to see if extra protein is leaking out. A blood test will only be required will if there is a problem with one of these basic checks. The blood test may be done at the hospital outpatient department or by your GP.

We recommend a more detailed kidney test at one year after the illness and then once every five years. This involves a glomerular filtration rate (GFR) test to measure exactly how well the kidneys are working, an accurate measurement of the amount of protein in the urine, and a normal blood pressure check. The GFR test takes several hours, but your child will not need to stay overnight.

Getting information

We appreciate that you may be upset and confused by your child's sudden illness. This information leaflet is just intended to give you some simple facts about HUS. The doctors and nurses aim to keep you well informed about your child's condition so please feel free to ask

should you have any questions during the course of your child's illness or if you require more information ■

Further reading

Fitzpatrick M. Haemolytic uraemic syndrome and *E coli* 0157. *BMJ* 1999; **318**: 684-685.

Fitzpatrick MM, Dillon MJ. Current views on aetiology and management of haemolytic uraemic syndrome. *Postgrad Med J* 1991; **67**: 707-709.

Fitzpatrick MM, Shah V, Trompeter RS, Dillon MJ, Barratt TM. Long term renal outcome of childhood haemolytic uraemic syndrome. *BMJ* 1991; **303**: 489-492.

Karmali MA, Petric M, Lim C *et al*. The association between idiopathic hemolytic uraemic syndrome and infection by verotoxin-producing *Escherichia coli*. *J Infect Dis* 1985; **151**: 775-782.

Japanese Pediatric Nephrology Association. Diagnosis and therapeutic guidelines for hemolytic uraemic syndrome associated with enterohemorrhagic *Escherichia coli* infection. *Pediatr Int* 1999; **41**: 449-451.

Key points

- **HUS is rare, but it can be a potentially serious disease.**
- **HUS is usually caused by infection with a bacterium called *E coli*, which releases a harmful substance called verotoxin into the bloodstream.**
- **The main features of the syndrome are anaemia, low platelet count and poor kidney function.**
- **Treatment may include blood transfusions and dialysis.**
- **Most children will not have major kidney problems in the future, but long-term follow-up of kidney function after HUS is recommended at present.**



HUS support groups



HUSH aims to increase public awareness of E coli 0157 and produces a number of information leaflets for this purpose

There are a number of charitable organisations that have been set up to provide support for anyone who is affected by HUS. These groups also aim to create a means of communication and increase public awareness of this and other related syndromes.

Haemolytic Uraemic Syndrome Help

HUSH is a charitable organisation set up to:

- Create a means of communication between families affected by *Escherichia coli* 0157 to prevent a feeling of isolation and, in particular, to put recent sufferers in contact with long-standing sufferers who have previously experienced this illness
- Increase public awareness of *E coli* 0157 and disseminate information as to the steps that can be taken to avoid infection
- Increase knowledge/awareness of GPs and Accident and Emergency Departments in order to speed up diagnosis
- Encourage the government to legislate effectively in order to decrease *E coli* 0157 poisoning cases
- Collate scientific research that is being undertaken in various areas, such as vaccination/prevention, diagnostic methods, treatment and knowledge of long-term effects and possible treatment with antitoxins or toxin receptor analogues.

HUSH consists of:

- Families affected by the *E coli* 0157 bacterium

- Legal, medical, microbiological, public health and environmental health experts who wish to reduce illness caused by this bacterium
- Approximately 120 members.

HUSH activities include:

- Liaison with medical experts and public health departments
- Research into the condition
- Compilation of information
- Quarterly meetings
- Fundraising
- Production of a quarterly newsletter and information leaflets.

The Heather Preen Trust

The Heather Preen Trust was set up in memory of Heather Preen, an eight-year-old girl who died after developing HUS while on holiday in Devon.

The Trust aims to:

- Increase usage of the all-encompassing term verotoxigenic *E coli* (VTEC) as opposed to the more common terms *E coli* 0157 and HUS
- Increase professional awareness of the vital need for stool tests
- Educate and inform about the preventive hygiene methods which can help to minimise infection.

Further information is available from:



**HUSH
PO Box 1303
Loxley
Sheffield S6 6YL.**

**The Heather Preen Trust
Chiltern House
Leys Road
Brierley Hill
West Midlands DY5 5UP.**

In August this year, the Trust organised a consensus meeting of experts where key messages for the general public and relevant health professionals were agreed. This resulted in 'Heather's checklist for *E Coli* (VTEC)' which provides advice for the public.

The Trust has produced information leaflets for both the general public and for professionals. These leaflets will be available from mid October by sending a stamped addressed envelope to the Heather Preen Trust ■

Heather's checklist

If you can answer 'yes' to any of the following questions for someone who is suffering from the symptoms described you should take them to see a doctor today:

- ✓ He/she has had diarrhoea for more than 48 hours
- ✓ The diarrhoea has been bloody
- ✓ He/she has or has had diarrhoea and has now stopped passing urine, or is passing less urine than normal
- ✓ It is possible that the person may or may not have suffered from stomach cramps.

If any of the above happens, or has happened in the last few days, and the person is now also listless, disinterested or paler than normal, they should be taken to their local hospital, or the emergency services should be contacted immediately ■