



Haemolytic uraemic syndrome (HUS)

This leaflet explains about haemolytic uraemic syndrome (HUS), what causes it and how it can be treated. It also describes what you can expect when your child comes to hospital for treatment.

What is haemolytic uraemic syndrome?

Haemolytic uraemic syndrome (HUS) is the name given to a group of illnesses that in some cases can lead to acute kidney failure in children. The name consists of medical terms describing the symptoms:

- Haemolytic = destruction of red blood cells
- Uraemic = build up of waste substances in the blood
- Syndrome = collection of symptoms seen together

What causes HUS?

There are three types of HUS, the two most common of which follow an infection, either after a tummy bug (gastroenteritis) or a Pneumococcal infection (chest infection). However, only a small number of tummy bugs and chest infections lead to HUS. The tummy bug most commonly linked with HUS (although not the only one) is a special type of the Escherichia coli (E. coli) bacterium. This can be eaten in undercooked meat, rarely dairy produce and on several occasions in the UK has been contracted through contact with infected farm animals. In most people, the bacteria pass through their system in a few days leaving no lasting effects. In some people, particularly children, the poisons (toxins) that the bacteria release cause HUS. The second type follows an infection with the Pneumococcus bug, which can give quite a severe illness usually associated with a chest infection. The bug changes the proteins present on the surface of the red blood cells so that the body does not recognise them any more. The body then antibodies that attack the red blood cells making them break down. HUS due to these two causes is very unlikely to come back after successful treatment. HUS can develop without a tummy bug or chest infection (atypical HUS) but this is much more rare. It can be due to genetic causes. This atypical type of HUS tends to come and go after treatment, so very careful follow-up is necessary. In all three types of HUS the reason for the symptoms is the same. The red cells are broken into fragments. The body then tries to repair the problem by attracting platelets, the tiny circulating cells that form clots. This means that the number of red cells and platelets fall in the blood so that the child becomes anaemic (we say that the haemoglobin is low) and their blood does not clot so well. Children may need to have a blood transfusion if the haemoglobin falls too low. One of the first signs of recovery is that the number of platelets in the blood starts to improve again and the haemoglobin stops falling.

The little clots can clog up blood vessels, and are particularly likely to do so in the kidney. For this reason, the kidney can shut down and either produce less urine or stop producing any urine. The kidneys usually recover over time ranging from a few days to weeks. The most important sign of recovery is the return of urine output. Very rarely, the small clots affect other parts of the body. We check the blood sugar regularly in case the pancreas is affected. Very rarely the brain may be involved so we keep a regular check of your child's ability to communicate and respond to us.

What are the symptoms of HUS?

With E.Coli HUS, your child will have had a preceding episode of diarrhoea, which is usually bloody. For this reason this type of HUS is sometimes call Diarrhoea positive (D+) HUS. It is also sometimes called 'typical' HUS, as in the past it was always the most common. In the Pneumococcal type, your child may have had a chest infection. The 'atypical' type may be set off by any infection in a child who is susceptible, or sometimes no reason for it starting can be found. Your child may feel weak, tired and look pale. He or she may also have small bruises or nosebleeds. This is due to the red blood cells and platelets being destroyed. Red blood cells carry oxygen around the body so fewer of them in circulation means that your child will feel weak, tired and pale (anaemic). The bruises and nosebleeds happen because the platelets are being used up as well as red blood cells. Platelets help our blood to clot so fewer of them in circulation leads to bruising and bleeding. Your child may also seem to pass less urine (pee) less than usual, and his or her urine may be a pink or brown colour due to blood cells. The destroyed red blood cells that are stuck in the tiny blood vessels in the kidneys reduce the amount of urine they can produce. This means that the kidneys are unable to remove excess salt and water from the body, which can lead to swollen face, tummy, hands or feet (oedema), and raised blood pressure (hypertension).

How is HUS diagnosed?

Your doctor will ask you lots of questions about how your child became ill and examine your child. Various tests will be needed to confirm or rule out HUS:

- blood tests to check how many red blood cells are present, to look for red cell fragments and to check the number of platelets
- urine tests to check for blood and protein in the urine
- stool (poo) and blood samples to try to find signs of E.coli or Pneumococcus Other tests may also be needed if the results of these tests are not completely clear.

How common is HUS?

HUS is quite a rare condition. One study estimated that less than one child in 100,000 under-sixteen develops HUS each year in the UK. However, when looking at just under-fives, the figure rises to 1.54 in every 100,000 children in the UK each year. Our experience is that it is much more common in young children than teenagers and young people. HUS that develops after a chest infection is becoming more common than the tummy bug type, probably due to increased publicity around food hygiene. A vaccine against Pneumococcal infections is now part of the regular childhood immunisation schedule so the numbers may drop in future. Many studies have reported that children seem to develop HUS after a tummy bug more often during the summer months. It is likely that this is linked to people serving undercooked meat at barbeques. The atypical type of HUS is very rare. All three types of HUS seem to affect boys and girls in equal numbers, and no one race is more likely to develop HUS than another.

How is HUS treated?

HUS usually requires a stay in hospital for close monitoring and treatment. The most immediate treatment your child will need is fluids and minerals to stop any dehydration. Dehydration is common after severe vomiting and diarrhoea and can be life threatening if not treated quickly. The fluids and minerals will be given through an intravenous drip (IV) into a vein in your child's hand or arm. The amount we give is checked very carefully by measuring the amount going in as fluids and the amount coming out as diarrhoea, vomiting and urine, and by regularly checking the temperature of the fingers and toes and the blood pressure. He or she will also need regular blood tests, to check the levels of both red blood cells and platelets.

Your child may need a transfusion of red blood cells, if the blood tests confirm that your child's red blood cell count is very low. The blood tests continue throughout treatment. As platelets are often destroyed as well as the red blood cells, a rise in platelet levels shows that the breakdown of red blood cells is slowing.

In severe cases your child will need kidney dialysis if his or her kidneys show evidence of shutting down by producing reduced amounts of urine. This is usually done by inserting a soft catheter into the tummy cavity under general anaesthetic so that it can be used to flush fluids in and out (using a machine). The fluid can take away salt, water and the impurities the kidneys are unable to remove. This may only be for a short while, to take over the work that the kidneys are unable to do. However, the average time would be around 7 days.

The amount of urine that your child passes will be monitored closely, as this tends to improve slowly as your child is recovering. This allows us to reduce the amount of dialysis your child has, until we can stop it when his or her urine output is satisfactory.

Sometimes children are left with kidneys that do not recover completely. This is called chronic renal failure. In this case they will need very careful follow-up. Very, very rarely, prolonged dialysis may be needed. Some children may benefit from plasma exchange, but this is used extremely rarely and selectively in special situations.

What is the outlook for children with HUS?

Most children recover well. Only very rarely does HUS lead to the death of a child; this is more likely to happen when body systems as well as the kidneys have been affected. In most children, kidney function recovers, but your child will need long term follow up to make sure that they do not show any signs of developing kidney problems in the future. His or her blood pressure will be checked regularly at first then at least every year. The level of protein in the urine will also be checked, as this can be a sign that the kidneys are not working as well as they should. This just involves sending a urine sample (the first in the morning if possible) off to the laboratory. If your child's urine protein levels do not decrease, he or she may need to take a medicine called an ACE inhibitor. These aim to control the levels of protein in the urine. More information about these medicines is in the leaflet *ACE inhibitors*.

Long-term follow up is essential to be sure that your child's kidneys do not show any deterioration in their ability to work as time goes by. This may be done with a test called a glomerular filtration rate (GFR) test, firstly one year after the illness and then sometimes five years afterwards. For more information about the GFR test, please see our leaflet. If the GFR test results show that your child's kidney function is not recovering as expected, he or she may need more frequent follow up checks. A small number of children develop kidney damage and kidney failure, so require long-term dialysis and transplantation. This is more common with the 'atypical' type of HUS

Can HUS be prevented?

It is impossible to say whether HUS can ever be prevented totally, but there are several steps that reduce the risk of it developing.

HUS following a tummy bug

- Always cook meat products, such as hamburgers and kebabs, right through so there are no pink bits and the juices run clear when pressed. If you are barbequing, cook the meat in the oven or on the hob first, and then transfer to the barbeque to warm through rather than cook.
- Use separate chopping boards and kitchen utensils for raw and cooked meats to avoid contamination. Raw meat should always be stored at the bottom of the fridge to avoid juices dripping onto foods below.
- Avoid unpasteurised milk and cheeses when your child is growing up. Although the evidence linking HUS and unpasteurised food is less clear, these products can contain bugs that cause other illnesses as well as HUS.
- If your child develops a tummy bug and you think it may have been caused by something he or she ate, you should take your child to your family doctor (GP) to have this confirmed. Certain illnesses, particularly those caused by food, have to be reported officially in case there is an outbreak.
- Teach your child good hygiene rules, such as hand washing before and after eating and going to the toilet. If your child has been in contact with animals, hand washing is especially important. Lack of hygiene is not usually implicated in the spread of HUS but is a good rule to follow to prevent spread of other illnesses, such as tummy bugs, coughs and colds.

HUS following a chest infection

- The vaccine against Pneumococcal infections is the best hope for reducing the number of children developing HUS following a chest infection. This is now part of the regular immunisation schedule that should be followed by each child in the UK. However, the vaccine does not work against all strains of Pneumococcus, so it is unlikely to rule out HUS after chest infections completely.

Atypical HUS

- The only way of preventing atypical HUS coming back is to monitor children at risk of developing this closely to ensure they do not have further relapses. Regular blood pressure monitoring is essential as a rise in blood pressure can be an early sign that HUS is coming back.

Is there a support group for children with HUS?

Yes, the following organisation should be able to offer you support and advice:

HUSH (Haemolytic Uraemic Syndrome Help) UK PO Box 59 Hayes UB4 8XE Tel: 0800 7 4679

Website: www.ecoli-uk.com

Notes

If you have any questions, please call