

Hemolytic Uremic Syndrome

What is HUS?

HUS is a syndrome or disorder that has two major components:

Hemolytic: There is a process going on in which little clots form in the smallest blood vessels of the body. These clots use up platelets and break up red blood cells, the oxygen carrying units of the blood. Doctors call this a microangiopathic process. The low platelet counts can cause bruising and bleeding. Low red blood cell counts can cause fatigue. When

the blood breaks down, the skin and eyes can develop a yellow, jaundiced color.

Uremic: One of the major places that clots form in HUS is the kidney. This process can cause mild problems with kidney function detectable only in a laboratory, full-blown kidney failure, or anything in between.

Clots can form in other organs and cause malfunction and damage. This process may affect all pancreas, liver, lung, and brain.

Types of HUS

Diarrhea + (typical): The most common form of HUS in children follows an illness with diarrhea, usually with blood in the stool. A number of germs can cause this, but they all produce a toxin that can enter the blood and attack the kidneys and other

organs. The most common cause is a form of E. coli (O157:H7).

Atypical: This type of HUS does not follow another illness. It may be associated with inherited problems of the immune system or the blood clotting system.

How do you diagnose HUS?

HUS is usually diagnosed by finding typical laboratory abnormalities. There is no test that is specific for HUS. The germs that cause the diarrhea that brings it on can be grown by special stool

cultures. New lab studies can identify the DNA for the toxin in the stool as well. However, by the time HUS develops, the diarrhea may have cleared and none of these tests may be positive.

How do you treat HUS?

There is no treatment specifically for HUS. Careful attention must be directed to the child's fluid balance and nutrition. Transfusions can be given to treat severe anemia and bleeding due to low platelet counts. If kidney failure occurs, dialysis may be necessary. Drugs may be needed to treat high blood pressure.

Typical HUS may follow a bacterial infection, but antibiotics play no role in its treatment. Atypical HUS may sometimes be treated with plasma exchange or plasmapheresis, a special type of blood therapy.

In typical HUS, 95% of children survive. Of these children, 95% recover sufficient renal function that they do not need dialysis. The other 5% go on to long-term dialysis and kidney transplantation.

Typical HUS almost never occurs again in the same child. Long-term follow-up is essential, since some children develop kidney problems 5 or 10 years later. These include protein in the urine, high blood pressure, or loss of renal function. At this time, we cannot tell who will develop later problems, so we watch everybody.

Atypical HUS has a worse prognosis. This type tends to occur over and over again in the same patient, and it often leads to permanent kidney failure. Even with kidney transplantation, this type of HUS may happen again in the new kidney.