Hemolytic Uremic Syndrome

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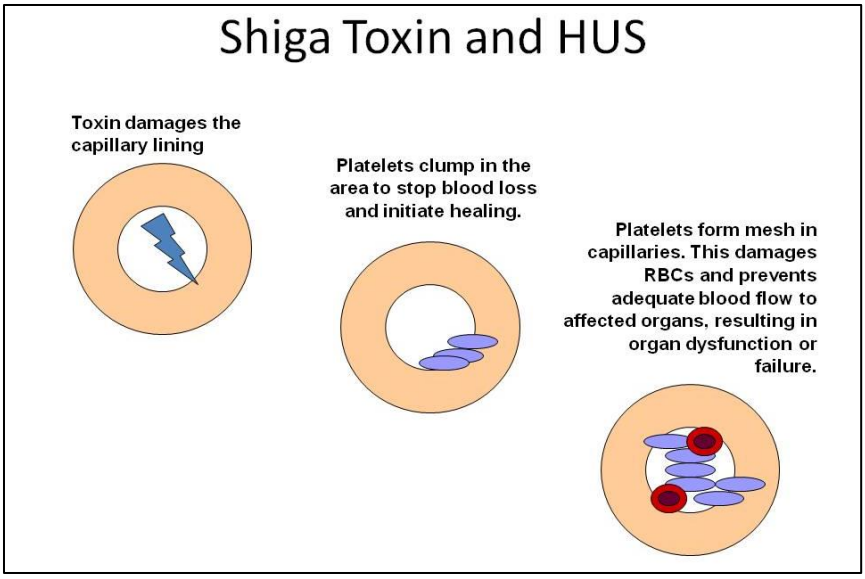
What is HUS?

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 organs, but especially pancreas, liver, lung, and brain.

How do you diagnose 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 treat HUS?

There is no treatment specifically for typical 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 can be treated with eculizumab (Soliris™), an antibody that stabilizes the complement immune system.

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 can be a life-long illness that returns in the kidney transplant. Eculizumab appears to control this disorder and prevent progressive kidney failure.