Hepatocerebral degeneration

Hepatocerebral degeneration is a brain disorder that occurs in people with liver damage.

Causes

This condition may occur in any case of acquired liver failure, including severe hepatitis.

Liver damage can lead to the buildup of ammonia and other toxic materials in the body. This happens when the liver doesn't work properly. It does not break down and eliminate these chemicals. The toxic materials can build up in the brain and damage brain tissue. However, ammonia may not be the cause of hepatocellular degeneration.

Specific areas of the brain, such as the basal ganglia, are more likely to be injured from liver failure. The basal ganglia help control bodily movement. This condition is the "non-Wilsonian" type. This means that the liver damage is not caused by Wilson disease, which leads to copper deposits in the liver. Damage to the basal ganglia can cause symptoms similar to Parkinson disease.

A condition called hepatic encephalopathy can also cause similar symptoms. It is more frequent than hepatocerebral degeneration. It also can occur with hepatocerebral degeneration.

Symptoms

Symptoms may include:

- Difficulty walking
- Impaired intellectual function
- Jaundice
- Muscle spasm (myoclonus)
- Rigidity
- Shaking of arms, head (tremor)
- Twitching
- Uncontrolled body movements (chorea)
- Tremor of the hand when the wrist is extended (also called a flapping tremor or asterixis)
- Unsteady walking (ataxia)

Exams and Tests

A physical exam may reveal signs of cirrhosis of the liver or liver failure, including:

- Fluid in the abdomen that causes swelling (ascites)
- Gastrointestinal bleeding from enlarged veins in the food pipe (esophageal varices)
- Jaundice (yellowing of the skin)

A nervous system (neurological) exam may show signs of:

- Coma
- Confusion
- Dementia
- Involuntary movements
- Walking instability (ataxia)

Laboratory tests will show advanced liver disease or liver failure. Lab tests can also check for other causes of the symptoms.

Other tests may include:

- MRI of the head
- EEG (may show general slowing of brain waves)
- CT scan of the head

Treatment

The treatment includes trying medicines that are used for hepatic encephalopathy. This includes antibiotics (such as rifaximin) or a medicine such as lactulose, which lowers the level of ammonia in the blood.

Trying to correct the “shunt” (blood flow changes) from the liver disease may help. This may be done by interventional radiology or surgery.

There is no specific treatment for the neurologic syndrome, because it is caused by irreversible liver damage. A liver transplant may cure the liver disease. However, this operation may help, but not reverse the symptoms of brain damage.

Outlook (Prognosis)

This is a long-term (chronic) condition that may lead to permanent nervous system (neurological) symptoms.

The person may continue to get worse and die without a liver transplant. If a transplant is done early, the neurological syndrome may improve.

Possible Complications

Complications include:

- Hepatic coma
- Severe brain damage with a wide range of possible symptoms

When to Contact a Medical Professional

Contact your health care provider if you have any symptoms of liver disease.

Prevention

It is not possible to prevent all forms of liver disease. However, alcohol-related, metabolic-related, and many types of viral hepatitis may be prevented.

To reduce your risk of getting alcohol-related or viral hepatitis:

- Avoid risky behaviors, such as IV drug use or unprotected sex.
- Don't drink, or drink only in moderation.

Alternative Names

Chronic acquired (Non-Wilsonian) hepatocerebral degeneration; Hepatic encephalopathy; Portosystemic encephalopathy

References

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Updated by: Jenifer K. Lehrer, MD, Gastroenterologist, Philadelphia, PA. Review provided by VeriMed Healthcare Network. Also reviewed by David C. Dugdale, MD, Medical Director, Brenda Conaway, Editorial Director, and the A.D.A.M. Editorial team.

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