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Hepatic encephalopathy

Hepatic encephalopathy is a worsening of brain function that occurs when the liver is no longer able to remove toxic substances in the blood.

Causes

Hepatic encephalopathy is caused by disorders that affect the liver. These include disorders that reduce liver function (such as cirrhosis or hepatitis) and conditions in which blood circulation does not enter the liver. The exact cause of hepatic encephalopathy is unknown.

An important job of the liver is to change toxic substances that are either made by the body or taken into the body (such as medicines) and make them harmless. However, when the liver is damaged, these "poisons" may build up in the bloodstream.

Ammonia, which is produced by the body when proteins are digested, is one of the harmful substances that is normally made harmless by the liver. Many other substances may also build up in the body if the liver is not working well. They can cause damage to the nervous system.

Hepatic encephalopathy may occur suddenly in people who previously had no liver problems when damage occurs to the liver. More often, the condition is seen in people with chronic liver disease.

Hepatic encephalopathy may be triggered by:

- Dehydration
- Eating too much protein
- Electrolyte abnormalities (especially a decrease in potassium) from vomiting, or from treatments such as paracentesis or taking diuretics ("water pills")
- Bleeding from the intestines, stomach, or esophagus
- Infections
- Kidney problems
- Low oxygen levels in the body
- Shunt placement or complications (See: Transjugular intrahepatic portosystemic shunt)
- Surgery
- Use of medications that suppress the central nervous system (such as barbiturates or benzodiazepine tranquilizers)

Disorders that can mimic or mask symptoms of hepatic encephalopathy include:

- Alcohol intoxication
- Complicated alcohol withdrawal
- Meningitis
- Metabolic abnormalities such as low blood glucose
- Sedative overdose
- Subdural hematoma (bleeding under the skull)

- Wernicke-Korsakoff syndrome

Hepatic encephalopathy may occur as an acute, potentially reversible disorder. Or it may occur as a chronic, progressive disorder that is associated with chronic liver disease.

Symptoms

Symptoms many begin slowly and gradually worsen, or they may begin suddenly and be severe from the start.

Symptoms may be mild at first. Family members or caregivers may notice that the patient has:

- Breath with a musty or sweet odor
- Change in sleep patterns
- Changes in thinking
- Confusion that is mild
- Forgetfulness
- Mental fogginess
- Personality or mood changes
- Poor concentration
- Poor judgment
- Worsening of handwriting or loss of other small hand movements

More severe symptoms may include:

- Abnormal movements or shaking of hands or arms
- Agitation, excitement, or seizures (occur rarely)
- Disorientation
- Drowsiness or confusion
- Inappropriate behavior or severe personality changes
- Slurred speech
- Slowed or sluggish movement

Patients with hepatic encephalopathy can become unconscious, unresponsive, and possibly enter a coma.

Patients with hepatic encephalopathy are often not able to care for themselves because of these symptoms.

Exams and Tests

Nervous system signs may change. Signs include:

- Coarse, "flapping" shaking of the hands when attempting to hold the arms out in front of the body and lift the hands
- Abnormal mental status, particularly cognitive (thinking) tasks such as connecting numbers with lines
- Signs of liver disease, such as yellow skin and eyes (jaundice) and fluid collection in the abdomen (ascites), and occasionally a musty odor to the breath and urine

Tests may include:

- Complete blood count or hematocrit to check for anemia
- CT scan of the head or MRI
- EEG
- Liver function tests

- Prothrombin time
- Serum ammonia levels
- Sodium level in the blood
- Potassium level in the blood
- BUN and creatinine to see how the kidneys are working

Treatment

Hepatic encephalopathy may become a medical emergency. Hospitalization is required.

The first step is to identify and treat any factors that may have caused hepatic encephalopathy.

Gastrointestinal bleeding must be stopped. The intestines must be emptied of blood. Infections, kidney failure, and electrolyte abnormalities (especially potassium) need to be treated.

Life support may be necessary to help with breathing or blood circulation, particularly if the person is in a coma. The brain may swell, which can be life-threatening.

Patients with severe, repeated cases of encephalopathy may be told to reduce protein in the diet to lower ammonia production. However, dietary counseling is important, because too little protein in the diet may cause malnutrition. Critically ill patients may need specially formulated intravenous or tube feedings.

Lactulose may be given to prevent intestinal bacteria from creating ammonia, and as a laxative to remove blood from the intestines. Neomycin may also be used to reduce ammonia production by intestinal bacteria. Rifaximin, a new antibiotic, is also effective in hepatic encephalopathy.

Sedatives, tranquilizers, and any other medications that are broken down by the liver should be avoided if possible. Medications containing ammonium (including certain antacids) should also be avoided. Other medications and treatments may be recommended. They may have varying results.

Outlook (Prognosis)

Acute hepatic encephalopathy may be treatable. Chronic forms of the disorder often keep getting worse or continue to come back.

Both forms may result in irreversible coma and death. Approximately 80% (8 out of 10 patients) die if they go into a coma. Recovery and the risk of the condition returning vary from patient to patient.

Possible Complications

- Brain herniation
- Brain swelling
- Increased risk of:
    - Cardiovascular collapse
    - Kidney failure
    - Respiratory failure
    - Sepsis
- Permanent nervous system damage (to movement, sensation, or mental state)
- Progressive, irreversible coma
- Side effects of medications

When to Contact a Medical Professional

Call your health care provider if any change in mental state or other nervous system problem occurs, particularly if there is a known or suspected liver disorder. Hepatic encephalopathy can rapidly get worse and become an emergency condition.

Prevention

Treating liver disorders may prevent some cases of hepatic encephalopathy. Avoiding heavy drinking and intravenous drug use can prevent many liver disorders.

If there are any nervous system symptoms in a person with known or suspected liver disease, call for immediate medical attention.

Alternative Names

Hepatic coma; Encephalopathy - hepatic

References

Garcia-Tsao G. Cirrhosis and its sequelae. In: Goldman L, Ausiello D, eds. *Cecil Medicine*. 24th ed. Philadelphia, Pa: Saunders Elsevier; 2011:chap 156.

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