

Metabolic errors stall recovery

RARE In genetic disorder, body cannot convert food into energy

HT Correspondent

htreporters@hindustantimes.com

NEW DELHI: Almost two in three — 61% — patients admitted to Delhi's Indraprastha Apollo Hospital are malnourished on admission, which influences how quickly they bounce back after surgery.

The problem, say clinical nutritionists, is not always imbalanced diets but also "Inborn Errors of Metabolism," another name for a group of rare genetic disorders in which the body cannot properly turn food into energy.

The disorders are usually caused by defects in specific proteins (essential enzymes) that help break down food.

"These disorders are present from birth but often remain undiagnosed even though they manifest early in life. Unmetabolised food products can build up in the body and cause developmental delays, mental retardation, kidney and/or liver failure," says Dr Anupam Sibal, group medical director and paediatrician, Apollo Hospitals.

Early diagnosis is essential as simple diet changes can help control the disorder.

The treatment — type and extent of the changes — depends on the specific metabolic error.

For example, a baby with galactosemia should avoid milk and milk products, including yoghurt, cheese, and ice-cream.

"A baby with galactosemia is unable to metabolise galactose, which reaches high levels in the body, causing damage to the liver, central nervous system and various other body systems.

TYPES AND SYMPTOMS

- Disorders that cause toxic accumulation
- Disorders of protein metabolism
- Disorders of carbohydrate intolerance
- Disorders of energy production, utilisation
- Symptoms may begin abruptly or be chronic and progressive
- Some symptoms include:
Poor feeding, vomiting;
Unexplained neonatal or sudden infant deaths in siblings;
Recurrent episodes of vomiting, seizures, lethargy, coma

An infant with galactosemia may develop jaundice, vomiting, lethargy and convulsions. But galactose and lactose-free milk substitutes such as soy milk can prevent the problem," says Sibal.

"Simple blood and urine spot screening at birth can detect many Inborn Errors of Metabolism such as phenylketonuria (PKU), galactosemia and fructose intolerance right after birth. In undiagnosed cases, the disorder may manifest as an emergency, causing seizures and coma when the child is under stress," said Dr Suresh Vijay, consultant in paediatric-inherited metabolic disease at Birmingham Children's Hospital, UK.

The tests are inexpensive and done routinely in developed countries.

In countries with limited funds, doctors should recommend screening if there has been infant death in the family within the first few weeks or months of birth.

SUNDAY hindustan



New Delhi / METRO

Vol. X No. 31

₹ 5.00 / *Price along with Hindustan ₹ 8.50

hindustantimes.com
times
JULY 31, 2011

smartcheck 16 |

hindustantimes

HEPATITIS DECODED

Keeping your liver safe
from viral attack

Dr Anupam Sibal

One in every twelfth person in the world has viral Hepatitis B or Hepatitis C, making the dismore common than cancers and AIDS, says the World Health Organisation.

Types: Hepatitis A and E spread through contaminated water and food and cause jaundice outbreaks. Hepatitis B and C are blood-borne viruses that spread through unsafe blood transfusion, unsafe sex, use of unsterilised medical equipment, shared razors, tattooing or body piercing tools and haemodialysis in kidney-failure people.

Vaccines: Hepatitis B vaccine should be given on day one of life, as the risk of virus transmitting to a child from the mother is high. The virus can lie silent for years and suddenly become active.

Three doses of Hepatitis B vaccine costs ₹100 and is given every three months. Hepatitis A vaccine costs ₹600 and is given in two doses at the age of one and two.

Prevention: Don't share needles. Cover open cuts or wounds. Use condoms, when getting pierced or tattooed, insist on a new needle and ink pot.

Symptoms of Hepatitis A and E (that cause seasonal outbreaks): Fever, stomach ache, appetite loss, jaundice, clay-coloured stools, dark yellow urine. Hepatitis B and C are silent viruses that do not cause symptoms till 60% of the liver is damaged. After that, the symptoms are the same.

Tests: Liver function tests and ultrasound detect the virus in the liver and deformation in the organ surface. ELISA-based blood tests are then done for confirmation.

Dr Sibal is a liver specialist and group medical director, Apollo Hospitals



think!

SUNDAY HINDUSTAN TIMES, NEW DELHI
JULY 31, 2011

think!