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METABOLIC AND DEGENERATIVE DISORDERS

HEREDITARY FRUCTOSE INTOLERANCE

Symptoms of neurological impairment in five children with hereditary fructose intolerance are described from the Service de Pediatrie, Hopital Antoine Beclere, Clamart, France. The diagnosis was proved by the deficiency of fructose-1-phosphate aldolase hepatic activity. Neurological symptoms during or after the acute phase of fructose intoxication included seizures, intracranial hypertension, tetraplegia, mental retardation, and deafness. Roentgenographic examination showed hydrocephalus, intraparenchymatous hemorrhage, cortical atrophy with ventricular dilatation, and ischemic or hypoxic cerebral lesions. In three patients cerebral impairment was secondary to cardiovascular collapse, prolonged hypoglycemia, or hemorrhagic diathesis related to liver insufficiency. Improvement followed treatment with a fructose-free diet but seizures necessitating anticonvulsant treatment persisted in three patients. (Labrune P et al. Unusual cerebral manifestations in hereditary fructose intolerance. Arch Neurol Nov 1990; 47:1243-1244).

> COMMENT. Hereditary fructose intolerance is a metabolic disease of autosomal recessive inheritance that is due to a deficiency of aldolase B, the enzyme which catalyzes the catabolism of fructose-1-phosphate. The main symptoms are abdominal pain, vomiting, hypoglycemia, and liver dysfunction following the ingestion of fructose. Central nervous system involvement is unusual and serious sequelae may develop in cases that present with hemorrhage or hypoglycemia.

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