

A 10 year-old boy is brought to the physician by his mother because of a 1-day history of severe vomiting and mission. She says that he had he had he symptoms I'verent ago, but his condition improved with bed rest and exprin therapy. He is lethergic. Abdominal examination shows moterate hepatomegally Serum studies after

Glucose 55 mg/st. (3.1 mmol/L)

Ammonia 210 µg/dL (12) µm/dL) (N+29-70 µg/dL 0 17-41 µm/dL)

AST TOUL ALT 65 UL

Despite appropriate treatment, he dies 1 day later. Gross and microscopic appearances of his liver that were obtained at autupay are shown. The most fixely underlying cause of this patient's disorder involves which of the following?













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