CASE 32

ID/CC A 6-year-old boy is brought by his parents to the emergency room in a **comatose state**.

HPI The child had been suffering from **chickenpox** and had been given **aspirin** by the family physician for fever.

PE VS: **fever**. PE: comatose child with **papulovesicular rash** all over body; fundus shows **marked papilledema**; no icterus; moderate hepatomegaly; asterixis.

Labs **Marked hypoglycemia; increased blood ammonia concentration; elevated AST and ALT; prolonged PT**; serum bilirubin normal. LP (done after lowering raised intracranial pressure): normal CSF.

Imaging CT: findings suggestive of **generalized cerebral edema**.

Gross Pathology Severe cerebral edema; acute hepatic necrosis.

Micro Pathology Liver biopsy reveals microvesicular steatosis with little or no inflammation; electron microscopy shows marked mitochondrial abnormalities.

Treatment Specific therapy not available. Supportive measures include lactulose to control hyperammonemia, fresh frozen plasma to replenish clotting factors, mannitol or dexamethasone to lower increased intracranial pressure, and mechanical ventilation. Exchange transfusion; dialysis.

Discussion Although the cause of the highly lethal Reye's syndrome (hepatoencephalopathy) is unknown, epidemiologic evidence strongly links this disorder with outbreaks of viral disease, especially influenza B and chickenpox. Epidemiologic evidence has also prompted the Surgeon General and the American Academy of Pediatrics Committee on Infectious Diseases to recommend that **salicylates not be given to children with chickenpox or influenza B**.