Background

Reye’s syndrome is a childhood disorder which was first recognized and described by Dr. Douglas Reye in Australia in 1963. The disease is characterized by several clinical features, manifested progressively in a quick succession. The outstanding clinical features are profoundly disturbed consciousness, fever, convulsions, vomiting, disturbed respiratory rhythm, altered muscle tone and altered reflexes. It is usually preceded by a prodromal viral infection. The disease usually progresses over hours or a few days to coma and death. At necropsy remarkably uniform pathological changes are found. They are severe fatty degeneration of liver and viscera and brain edema.

The mortality is high varying from 20 % to 100 % in some series. Thus, this disease constitutes a major health problem and posses a great risk to the younger generation.
REYE'S SYNDROME โรคบาดทะพางเกิน
คำว่า เฮียนดีรี่

Family or village very rare occurrence
Seasonal distribution - rainy and early winter
Signs and symptoms - characteristic
toms
Mortality 60 – 70 %

Symptoms
Prodome present in 80 % of cases fever, vomiting: diarrhea and URI
Onset very sudden convulsions followed by sustained coma usually in the morning.

Signs
Temp. normal to moderate elevation
Pulse Tachycardia
Resp. irregular or hyperpnea
B.P. normal
Nutritional status normal
Jaundice not present
Liver usually not enlarged on admission
Neck supple
Pupils dilated, sluggishly reactive
Fundus normal
Muscular tone usually increased, sometimes flaccid
Convulsions generalized

Laboratory findings
Leukocytosis c neutrophilia
Hypoglycemia
Elevated transaminases
Prolonged prothrombin time
Elevation of the blood ammonia
Hyperkalemia.
Abnormal increase and decrease in serum amino acids levels
Metabolic acidosis, respiratory alkalosis, or mixed.
C.S.F. usually no cell, glucose decreased, glutamine increased

Pathological findings
Liver Increased weight, uniformed of patchy yellow microscopic severe fatty metamorphosis
Brain Edematous, increased weight, microscopic edema, no inflammation, fatty degeneration in wall of blood vessels
Kidney Increased weight, pale cortex fatty dege

Differential Diagnosis
1. Acute gastroenteritis c febrile convulsion
2. Pneumonia c febrile convulsion
3. Encephalitis, meningitis, brain abscess
4. Fulminant hepatitis
5. Salicylate intoxication

Treatment Symptomatic
1. Correction of hepatic dysfunction of tubular epithelium - 50% Glucose IV. Push 10 – 20 c.c.
- Maintain fluid 10% Glucose in 1/3 NSS.
Amount 2/3 of maintenance need
- NB Tube lavage, Neomycin 50 - 100 mg/kg/day in divided doses
- Vit. K 5 mg I.M.
- Insulin 1 u Per 5 gm Glucose
- Cxchange transfusion

2. Control ICP
- Manitol 1 gm/kg/dose IV q 4 - 6 hrs.
- Dexamethasone 1 mg/kg/day in divided doses
- Fluid restriction
- Phenobarbital IM. Valium IV

3. Correction of electrolytes & Acid - base imbalances
- Na. K
- HCO₃ not usually given

4. Supportive treatment for coma
- Intubation, maintain adequate air way
- Positioning
- Urinary catheterization

Unfavorable signs
Hyperkalemia
Elevated blood ammonia
Elevated transaminases
Abnormal respiration
Abnormal pupils
Severe delirium, deep coma

Etiology
Not definitely Known
Possible etiologies include
- Viruses
- Drugs, chemical, toxins
- Viral – toxin interaction
- Metabolic defects.

Interested Observations
- Regularity of the disease
- Racial predilection
- Time of onset
- No recurrence

A case demonstration
A five years old Thai female was not ill until the day of admission. The night before admission, she went to bed, appeared to be well. At 5 a.m. work up and asked for water. Convulsions started abruptly at 7 a.m. followed by deep and sustained coma. She was seen at the hospital about 10.30 a.m.

P.E. T 37.4 c, P. 124/min R. 32 B.P. 100/70 mmHg
A well developed, well nourished Thai girl was in deep coma. Spastic decorticated posture was observed. Rales were heard bilaterally. Pupils were partially dilated, Pupils were partially dilated, sluggishly reacted to light. D.T.R was hyperactive. Babinski sign was positive.

Laboratory findings
Hgb. 11.4 gm % W.B.C. 7000 n. 83 % L 16 %, M 1 %
Blood sugar less than 30 mg. % SGOT 220 SGPT 310
CSF clear, 8 lymphocytes, sugar less than 100 mg. % glutamine 10 mg. %

Hospital course
50 % glucose 20 c.c was given. I.V. fluid was maintained. Ornietil 10 gm. Was added into the I.V. fluid. High cleansing enema with 5 % dextrose in ½ N.S.S. was performed. The patient show Immediate response but remained comatose. She expired 3q hours after admission.