

## Reye's syndrome

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### Abstract

Five and half years male child with one day history of pain abdomen and vomiting who was on aspirin for suspected rheumatoid arthritis presented initially with acute gastritis. Next day, however he developed the signs of encephalopathy with altered liver function.

**Key words:** Rheumatoid Arthritis, Aspirin, Hepatic Encephalopathy, Reye's syndrome.

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### Case summary

Five and half years male child was admitted in paediatric ward of Kathmandu Medical College Teaching Hospital, Sinamangal on 05/09/18 with one day history of excessive vomiting and abdominal pain over the right hypochondriac region without fever.

During the past two years, the child was suffering from high-grade fever of 103-104°F and joint pain on and off involving multiple small and big joints. He had also been treated as septicemia and tubercular pericarditis in India. Later on he was diagnosed to have juvenile rheumatoid arthritis from Kanti Children's Hospital and was put on aspirin, which he took for about 1 week with the relief of the symptoms.

On admission, the child was conscious and cooperative, but showed some pallor. He was moderately dehydrated and agitated but was not icteric. The temperature was 97°F and pulse 100/min, blood pressure 90/60 mm/Hg, respiratory rate of 26 per minute. Systemic examinations were within normal limits except for mild tenderness over the epigastrium and right hypochondriac region.

The child was rehydrated with Ringer's lactate solution and treated for gastritis with ranitidine injection, domperidone suspension and antacids orally.

Early in the morning the next day, the child complained of severe pain in the abdomen and vomiting, but there was no fever and the motion was normal. He was very much irritable, semiconscious with Glasgow Coma Scale (GCS)

of 9/15, pulse 68/min, BP systolic 100 mmHg RR 40 per minute, pupil reacting sluggish to light, later on fundoscopy was done which revealed bilateral papilloedema. The neurological examination showed exaggerated deep tendon reflexes, equivocal Babinsky reflex with persistent ankle clonus. There was no sign of meningism.

The child was immediately shifted to High Care Unit (HCU) and was treated with intravenous Mannitol, Dexamethasone, Vitamin K, Ceftriaxone, Metronidazole and Lactulose via nasogastric tube. But the condition of the patient further deteriorated with worsening of the consciousness.

The results of investigations sent from the HCU were as follows: Haemoglobin 9.9gm%, total leucocyte count 13,700/cumm predominantly polymorph 64%, random blood sugar was 44mg/dl, serum bilirubin 2.1mg/dl, SGPT-1230U/L, prothrombin time - 28 secs (control-11 secs). Blood urea, serum creatinine, electrolytes, blood uric acid, serum amylase were within normal limit. The serum ammonia could not be estimated due to lack of facility. On the third day the child remained critical with the deterioration of the general condition, GCS remaining low - 3-4/15. The same treatments were continued for another day. On the fourth day the child showed gradual improvement particularly of the conscious level, GCS became 15/15 late in the

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evening on the fourth day and vital signs normalized with pulse 80/min blood pressure 110/80 mmHg respiratory rate 24-28 per minute and temperature 98°F. Repeat random blood sugar was 108mg/dl and prothrombin time decreased to normal (12 secs) SGPT was not repeated immediately as normalization is less likely in two days. Mannitol and vitamin K was stopped but other treatment continued for one week. The child improved well and was discharged with full recovery on 7th day with the final diagnosis of Reye's syndrome. SGPT done in the follow up after one week became normal.

### **Introduction**

Reye's Syndrome was first recognized by R. Douglas Kenneth Reye an Australian Pathologist in 1963<sup>1</sup>. The symptom of the disease is believed to be triggered by the ingestion of the salicylate containing drugs such as aspirin in a patient who is exposed to the viral illnesses like influenza, cold and chicken pox that affect all the system of the body mostly affecting liver and brain lethally.

### **What is Reye's Syndrome?**

Reye's syndrome is a serious disease manifested as acute encephalopathy with hepatic dysfunction. It is considered to be a two phase illness because it is almost always associated with a previous viral infection such as influenza, cold or chickenpox in a child who is on salicylate treatment. It is not contagious and the cause is unknown. It occurs during the recovery period following a viral infection. Symptoms are persistent vomiting, loss of consciousness, convulsions, delirium and personality changes. If not treated in time, it can lead to hepatic as well as cerebral dysfunction. The serum level of ammonia rises while the blood sugar level drops and hepatic function alters. At the same time the liver may swell up and develop fat deposits. Swelling can also develop in the brain and cause such emergency symptoms such as seizures or convulsions and eventually lead to coma and brain death<sup>2</sup>. Reye's Syndrome is so rare and if it is usually diagnosed in time it prevents severe damage to the body.

### **There are five grades identified with Reye's syndrome<sup>3</sup>**

**Grade I** usually quite, lethargic, with vomiting and laboratory evidence of hepatic dysfunction.

**Grade II:** Deep lethargy, confusion, hyper reflexic

**Grade III:** Obtunded, light coma, decorticate rigidity, intact pupillary reaction.

**Grade IV:** Seizure, deepening coma, decerebrate rigidity, loss of oculocephalic reflexes, fixed pupil

**Grade V:** Coma, loss of deep tendon reflexes, respiratory arrest, fixed dilated pupil, decerebrate rigidity

### **What causes Reyes Syndrome?**

At present, no one knows precisely what causes Reye's Syndrome to develop in some children and not in others. It often follows a viral infection such as chicken pox or influenza. However, given the high incidence of these conditions in children, only very few children actually go on to develop Reye's Syndrome<sup>4</sup>.

Researchers are therefore speculating that the children who suffer may have some kind of genetic susceptibility to the disease. Aspirin may exacerbate this susceptibility<sup>4</sup>.

### **Aspirin and Reye's Syndrome**

Many clinical studies have indicated a link between Reye's Syndrome and aspirin, where aspirin was used in children during the viral stage of the illness. There is no conclusive proof for this, but the Committee for the Safety of Medicines (CSM) in the UK regarded this as enough of a risk for them to warn against the use of aspirin in children under the age of 12 years in 1986<sup>1</sup>.

In 1980 the number of cases peaked in the United States at 555. After that the figure began a sharp decline<sup>4</sup>. In 1984-94 there were no more than 36 cases of Reye's syndrome per year, and since then only a few cases have been reported each year<sup>5</sup>. In Nepal there are no data of the cases of Reye's syndrome as it is so rare disease.

### **Conclusion**

This child was suffering from fever and joint pain of long duration suggestive of juvenile rheumatic arthritis. He had received aspirin for one week but he developed signs and symptom strongly suggestive of Reye's syndrome within a week. With supportive therapy he improved well with in next few days of admission.

Rapid deterioration leading to hepatic encephalopathy and full recovery with in few days was probably due to the use of aspirin in this child. Therefore Aspirin under the age of 12 years is not recommended due to its serious side

effects including gastric irritation and hepatic encephalopathy. Its use in children under special circumstances has to be closely supervised so as to prevent serious complications.

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