

REYE SYNDROME – AN OVERVIEW

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ABSTRACT

Acute non-inflammatory encephalopathy with fatty liver failure is the definition of Reye syndrome, a rare and potentially fatal juvenile condition. This syndrome was first outlined by Australian pathologist R.D.K. Reye in 1963. Early in the 1970s, it became the focus of national surveillance in the United States, which resulted in strong recommendations against giving children aspirin. It often starts with vomiting and bewilderment before quickly progressing to a coma and death. This exercise discusses the etiology, pathophysiology, and clinical manifestations of Reye syndrome and emphasizes the value of an interdisciplinary approach to treatment.

Key words: Reye syndrome, Treatment

1. INTRODUCTION

In youngsters, syndrome often starts with vomiting and bewilderment before quickly progressing to a coma and death. This syndrome frequently manifests in the days following recovery from a viral illness treated with aspirin. Toxins, drug interactions, and inborn metabolic errors—particularly those involving the metabolism of fatty acids—may also predispose to or contribute to the development of Reye syndrome.

Etiology

The most frequent causes of Reye syndrome are viral diseases like varicella and influenza A and B. According to surveillance data collected by the Centers for Disease Control and Prevention (CDC) between 1980 and 1997, influenza infection occurred in 73%, varicella infection in 21%, and gastrointestinal infections in 14% of instances of Reye syndrome. In 82% of instances, serum salicylate concentrations could be found. Coxsackie, parainfluenza, Epstein-Barr (EBV), cytomegalovirus (CMV), adenovirus, and hepatitis are less frequently related viral connections. It has also been linked to bacterial diseases such Chlamydia, Bordetella pertussis, Mycoplasma, and Shigella. Studies on the epidemiology of disease have linked salicylate consumption to the emergence of Reye syndrome. While fewer than 0.1% of youngsters who used aspirin experienced More than 80% of kids with Reye syndrome had taken aspirin in the three weeks prior to their diagnosis. As a result of these findings, aspirin usage in children was discouraged in 1980. Following the widespread recommendations against giving aspirin to young children, the number of Reye syndrome cases that have been reported has substantially decreased. ⁽³⁾⁽⁴⁾

Pathophysiology

Reye syndrome's precise etiology is unknown, however it appears to include mitochondrial damage in the context of a viral infection. Aspirin may inflict or maintain damage to cellular mitochondria, which inhibits the metabolism of fatty acids. Neurologic symptoms are most likely caused by hepatic mitochondrial failure, which raises ammonia levels. Hyperammonemia may cause widespread cerebral edoema and subsequently increased intracranial pressure by inducing astrocyte edoema. Pathology studies have identified astrocyte edoema, neuronal loss, fatty kidney degeneration, and enlarged and fewer mitochondria.

History and Physical examination

Making this uncommon diagnosis calls for a high degree of suspicion based on the patient's medical history, clinical symptoms, and lab results. Reye syndrome symptoms and signs often appear between 12 hours and 3 weeks after a viral illness, such as gastroenteritis or an upper respiratory tract infection, has subsided. Vomiting usually starts 3 to 6 days following a viral illness, on average. Clinical development is divided into 5 separate stages, according to the CDC:

Stage 1

Persistent, copious vomiting, Lethargy, nightmares, increased somnolence and Confusion

Stage 2

Stupor, disorientation, combativeness, delirium, Hyperreflexia, positive Babinski sign, lack of appropriate response to noxious stimuli, dilated and sluggish pupils, Hyperventilation and tachycardia

Stage 3

Obtunded, comatose and decorticate rigidity

Stage 4

Pupil dilation with minimal response to light or fixed and dilated pupils, deconjugate gaze with caloric stimuli and Deep coma with decerebrate rigidity

Stage 5

Seizures, Flaccid paralysis, absent deep tendon reflexes, no pupillary response, Respiratory arrest

And Deaths ^(5,6)

Diagnostic Evaluation

The following criteria have been used by the CDC to define Reye syndrome

"Acute non inflammatory encephalopathy is defined clinically as cerebral edema without perivascular or meningeal inflammation, as well as a change in consciousness and, if available, a record of cerebrospinal fluid (CSF) containing fewer than or equal to 8 leukocytes/cu.mm.

liver biopsy or autopsy that is deemed to be diagnostic of Reye syndrome, or b) a threefold or more increase in the levels of serum glutamic-oxaloacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), or serum ammonia, are both regarded to be signs of hepatopathy.

Raised liver function tests (ALT, AST, bilirubin), hyperammonemia, abnormal coagulation studies, elevated amylase and lipase, decreased serum bicarbonate, and lab findings consistent with dehydration are among the anomalies in the body's chemical make-up that are linked to Reye syndrome. The most typical test result is an early spike in ammonia that occurs within 1 to 2 days of changes in mental status. The leukocyte count must be less than 8 in a lumbar puncture to meet diagnostic requirements. Opening pressure may increase in the later clinical stages of the disease, although it may also stay normal in the beginning ^{[8] [9] [10]}

2. MANAGEMENT / TREATMENT

To preserve hemodynamic stability and appropriate respiratory function in Reye syndrome, which is a rapidly advancing condition, early invasive operations may be necessary. A Foley catheter may be used to track urine output, central venous access insertion, and airway intubation, among other procedures. It may also be necessary to perform further specialized tests like liver biopsy and intracranial pressure monitoring.

Dextrose-containing fluids (D50, D10, D5, etc.) can be used to treat hypoglycemia with a blood glucose target of 100–120 mg/dl. Treatment options for acidosis include breathing control and sodium bicarbonate (care must be taken not to overcorrect or correct too rapidly). Treatment options for hyperammonemia include phenyl acetate-sodium benzoate (Immunol) and sodium polystyrene sulphate (Kayexalate), however hemodialysis may be necessary if the level is higher than 500mcg/dl., fresh frozen plasma (FFP), or vitamin K may be used to treat coagulopathy (particularly before invasive operations or with clinically substantial bleeding).

Treatment strategies for elevated ICP may include the following:

Raising the bed's head by 30 degrees, Control of temperature to avoid rigors and increased cerebral metabolism, ICP observing the use of Lasix, careful fluid management to avoid becoming over hydrated. Anesthesia and analgesia, Hypertonic saline or mannitol and seizures and subsequent prevention ⁽¹¹⁾

3. CONCLUSION

This child had a fever and chronic joint pain that was indicative of juvenile rheumatic arthritis. He had been taking aspirin for a week, but within that week, he began to exhibit symptoms and signs that were highly suggestive of Reye's syndrome. With supportive care, he made good progress in the days after his admission. Aspirin consumption in this youngster was likely the cause of the rapid deterioration that resulted in hepatic encephalopathy and full recovery within a few days. Since aspirin has major adverse effects, it is not advised for children under the age of 12.

4. REFERENCE

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