



Pathology Biology Section – 2006

G101 Hyperglycemic Hyperosmolar Nonketotic Syndrome in a Sixteen-Month Old Child With Rotaviral Diarrhea

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The goal of this case study is to present a child death due to severe dehydration from hyperglycemic, hyperosmolar, nonketotic syndrome (HHNS), with concomitant rotavirus diarrhea.

This presentation will impact the forensic community and/or humanity by reviewing the rare entity of HHNS in children; and discussing the differentiation between stress-induced hyperglycemia, diabetic ketoacidosis (DKA) and HHNS.

A sixteen-month-old male child, born at twentyfive weeks and five days gestation, had several congenital anomalies including serious mental and neurological deficiencies.

The initial five months of his life were spent in the hospital during which time he never demonstrated hyperglycemia or showed signs of being diabetic.

Three days prior to his death he developed episodic, profuse watery diarrhea. He was fed a banana, rice, applesauce, toast, (BRAT) diet, without toast. He was also treated with acetaminophen and bismuth subsalicylate. There were no visits to a physician.

On the third day of his illness, he was fussing at 0430. His mother tried to give him cough medicine but he refused. She changed his diaper, laid him prone, and rubbed his back. He was notably limp. When she checked on him approximately two hours later, he was unresponsive. Paramedics were called, but death was obvious and no resuscitation was attempted.

Postmortem examination revealed a slightly dysmorphic male, weighing 18 pounds and having a crown-heel length of 29.5 inches. The occipital frontal circumference was 16.8 inches. The eyes appeared sunken. The organs had dull surfaces and were tacky to touch. There was no physical or historical evidence of previous or recent abuse.

The stomach contained 300cc of light pink fluid without any food fragments. The intestinal contents were of a similar consistency to that of the stomach and light tan in color. The brain had markedly small cerebellar hemispheres and atrophic optic nerves. The hippocampi were atrophic and the lateral and fourth ventricles were mildly dilated. Microscopic sections from the pancreas, heart, lungs, liver, kidney, adrenals, thymus, and trachea were normal. Vitreous electrolytes included a glucose of 598mg/dL, vitreous osmolality 430mOsm (285-305mOsm), sodium-158mEq/L, chloride-140 mEq/L, urea nitrogen-37mg/dL, and creatinine-1.0 mg/dL. Acetone was negative. Stool culture was positive for rotavirus. Toxicology was positive for liver acetaminophen with 25mcg/gm, and liver salicylate of 3.5mg/100gm.

Diarrhea illness, world wide, represents a leading or second cause of death for children less than five years old. In the United States only about 300 child deaths per year are due to diarrhea.

HHNS is almost always a disorder of Type II diabetes mellitus in elderly, neglected, or debilitated adults. In HHNS, glucose levels are elevated, often as high as 1000 mg/dL or more. Ketones are negative because lipolysis is inhibited. Serum osmolality is high, with the measured level being higher than the calculated level. Acidosis may occur, and is usually due to lactate from hypoperfusion.

Fewer than 30 cases of HHNS have been reported in children since 1960. In most of these, the children are less than two years old and/or neurologically impaired. Mortality is as high as seventy five percent, and occurs from dehydration or from cerebral edema if rehydration occurs too rapidly.

HHNS in children represents either the initial presentation of diabetes mellitus, or it is associated with gastroenteritis, usually rotavirus, as was the case with this child. When HHNS is associated with gastroenteritis it is a transient condition and, if the child survives, they have no greater risk of developing diabetes mellitus than the rest of the population. The mean glucose level (634 mg/dL) is lower, and the sodium is higher (mean 135 mEq/L), when gastroenteritis is the cause of HHNS.

Suggested laboratory studies needed to make the diagnosis of HHNS, and to exclude diabetes mellitus, include glucose, osmolality, HgA1C, plasma insulin levels, and islet cell antibodies.

Although samples to exclude diabetes mellitus did not remain in this case, this child, who had a severe neurologic disorder and no prior history of a hyperglycemic event, and who was hyperosmolar with rotavirus diarrhea, represents a rare case of HHNS.

Hyperglycemic, Hyperosmolar, Nonketotic