

SEQUELAE IN SURVIVORS OF REYE'S SYNDROME

William B. Svoboda, MD

Pediatric Neurology & Epileptology Division

1035 N. Emporia

Suite 270

Wichita, Kansas 67214

In the 1960's, the question was "What is Reye's Syndrome?" The question of the 1970's was one of survival. The question of the 1980's focuses on the quality of survival, for Reye's Syndrome has come to be viewed as a disorder leading either to death or, more commonly, to full recovery. The mortality rate is now less than 20% to 30%. Still, some individuals suffer sequelae. When survivors are studied, significant psychologic deficits may occur in up to 64%, significant neurologic deficits in up to 54%; and major handicaps in as many as 42%. Significant neurologic impairments include blindness, spasticity, tics and, occasionally, seizures.

The range of residual problems extend from mild (such as a learning disability, incoordination or unclear speech) to severe (as with mental retardation, spasticity or disturbed speech). Such sequelae occur infrequently, often are transient, and, if lasting, often are subtle. Those working with children surviving Reye's Syndrome need to know (1) the predictor risks, (2) the potential problems of language, learning and behavior, and (3) both the prognosis and problem management so that they can offer early support and help.

PROBLEM PREDICTORS & RISK FACTORS

The risks for sequelae are significant if (a) the attack is severe, (b) the coma is prolonged or (c) if the child is young. The quality of the outcome does not correlate with the ammonia level, the SGOT increase or the treatment.

THE PROBLEMS FOLLOWING REYE'S SYNDROME

Three of every five children following Reye's experience speech problems and a similar percentage experience emotional problems. Three of every ten experience difficulties in learning.

The most common speech problems are those of distortions of pronunciation as well as difficulties in the motor act of speaking. Nearly 30% experience difficulties in expressive language, but this is seldom a lasting problem. The abnormal speech is commonly described as soft, harsh, hoarse, choppy, low-pitched, monotone, shallow or snorting. This does not relate to the use of intubation during the time when the child is on a respirator. Swallowing and drooling problems may occur. A few may retain a slurred speech pattern with a soft, breathy quality. Some may speak slowly, whereas others may rush what they say.

LEARNING PROBLEMS:

Three types of learning problems may be seen; namely, mental retardation (in 10% to 40%), learning disabilities (in 30%), and a temporary drop in performance (in 20%). About half of the survivors of Reye's Syndrome may show such difficulties.

Soon after the child's return to school, the teachers and parents complain that the child can do the work but works more slowly. Of more concern is that the child's performance in school may deteriorate. Grades are poor. The child has problems in reading, writing, spelling and math. Attention problems are prominent. The child is described as more distractable, inattentive and forgetful.

In some children, these may all be transient problems. Some children (about 20%) initially show a declining IQ. These children often have frank problems in getting along with others. This may be temporary. Some children start to recover within months. A falling intelligence is more often seen in teenagers.

Retardation is more to be seen in younger children, especially following a severe attack of Reye's Syndrome. Other neurologic handicaps often are present.

Learning disabilities may be seen following an attack of Reye's Syndrome. This is one of a group of medically related learning disabilities. Certain medical conditions have been found to be associated with specific patterns of learning disabilities. Those conditions that interfere with blood flow to the brain may result in disturbances of the visual learning channel, especially of perception. Those chemical insults in the body may impair the auditory learning channel, especially memory.

The process involved in Reye's Syndrome can affect the brain in multiple ways, principally (1) the metabolic disturbances, (2)

the pressure interference with blood flow to the brain and (3) pressure herniation of portions of the brain. The metabolic disturbance, theoretically, may impair the auditory learning channel and result especially in memory problems. The increased intracranial pressure may interfere with blood flow to the brain, resulting in impairments of the visual learning channel, affecting, especially, perception. If the pressure is too great and part of the brain herniates, further damage may occur to those areas of the brain related to immediate memory skills and attention. Thus, one might predict problems of memory, perception, language and attention in those who survive Reye's Syndrome. Such problems have been found.

The younger child, especially one recovering from a severe attack, may be left with retardation or at least a learning disability. Visual motor learning problems and difficulties in expressing one's thoughts are most commonly seen, especially in those with lowered intelligence. Such children have problems in relating items together and getting items in the right order. Problems with memory (both short-term and long-term memory), order and difficulties in developing thought concepts have been noted. Memory problems, especially auditory, tend to be an early difficulty from which the child may recover.

School grades often drop. This falling performance may be temporary, lasting about 6 months. The child may perform normally in intelligence and academic tests but not do well in the classroom. Math is especially a problem for teenagers. After a year or two, the child may show a remarkable recovery.

EMOTIONAL PROBLEMS:

Three types of problems result in 50% to 70% of children surviving Reye's Syndrome. Transient emotional upsets may be seen. Lasting irritability is reported. The family must undergo a series of adjustments.

Immediately after the attack, there is anxiety and apprehension that the attack may recur. The child and the family share a common fear of bodily harm and potential death. Early emotional problems include anxiety, acting-out behaviors, attention seeking, attention deficit symptoms (such as inattention, distractibility, excessive activity and agitation), bodily complaints (such as headaches), demanding behaviors, easy frustration, immaturity, infantile behavior, irritability, manipulateness and excessive whining. Yet, several years later, only the irritability persists.

The family is filled with concerns. There may be uncertainty as to how to treat the child. These include difficulties in limit

setting. The child is often looked at as vulnerable. Initially, attitudes may be positive; but after a year or two, interactions tend to turn negative.

Vulnerability is a common sequel to Reye's Syndrome (and to most other major medical conditions). There is the feared, if not actual, loss of an ability or of life itself. The parents tend to react with overprotection, overindulgence, overrestriction, yet underdiscipline. The child may become spoiled, immature and a frequent visitor to the doctor.

The brothers and sisters tend to be more aggressive, yet, underachieve in school. They also often complain of bodily aches and ills.

PROGNOSIS & PROBLEM MANAGEMENT

The outlook in Reye's Syndrome relates to the severity of the attack, the duration of the coma and the age of onset with younger children being more susceptible. The outcome is not predictable. The recovery period may be stormy at first, but a remarkable recovery often occurs later. When sequelae are seen, they occur infrequently, may be transient and are often subtle, if persistent.

The child and the family should be screened in follow-up. If problems are suspected, intervention should ensue. Early speech therapy may aid in recovery. Occupational, as well as physical therapy, may help with motor and coordination problems. Special educational supports may help both with transient school problems as well as any lasting learning difficulties. Counselling for the child and the family may begin in the hospital during the recovery period and be continued thereafter as needed. If problems occur, early intervention can help both child and family.

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