Treating acute bilirubin encephalopathy--before it's too late: early signs of bilirubin-induced neurologic damage in healthy term and near-term newborns are often vague. A comprehensive approach to management helps prevent rapid progression and irreversible consequences.

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Authors: Vinod K. Bhutani, Lois H. Johnson and Ron Keren
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Main content

A late-night telephone call from a parent of a recently discharged newborn reporting that the baby has, or may have, jaundice heralds a potential medical emergency--especially when jaundice is coupled with concerns about poor feeding, excessive sleepiness, irritability, or lethargy. This constellation of complaints requires detailed questioning about specific signs and symptoms, a review of the birthing and postnatal history and predischarge data, and, possibly, an emergency neurologic evaluation of muscle tone, alertness, and cry pattern for evidence of bilirubin-induced neurologic dysfunction (BIND).

Although the clinical signs described by the parent sound vague, and may simply reflect a new parent's anxiety or inexperience, they also could be early, nonspecific--but nevertheless sentinel--signs of acute bilirubin encephalopathy (ABE). ABE refers to the acute, and often progressive, manifestations of bilirubin toxicity that are seen in the first weeks after birth. When unmonitored or untreated, they may progress rapidly to advanced manifestations such as opisthotonus and seizures. If intervention to reduce the bilirubin load rapidly is neither timely nor efficient, chronic, permanent clinical sequelae of bilirubin toxicity--referred to as kernicterus--result and become increasingly apparent during infancy.

Newborn jaundice generally has a reassuringly benign outcome. It is kernicterus following an acute, brief, preventable exposure to extreme hyperbilirubinemia that can have lifelong deleterious effects. This article discusses the clinical signs, symptoms, and known causes of kernicterus and presents a systems-based strategy for preventing it. The strategy is based on predischarge screening and a bilirubin measurement plotted on the hour-specific bilirubin nomogram to identify newborns at risk of severe hyperbilirubinemia and to target follow-up to prevent irreversible damage.

The broad spectrum of BIND

Bilirubin-induced neurologic dysfunction refers to a wide spectrum of disorders, including kernicterus, that are caused by increasingly severe hyperbilirubinemia. The common insult in all cases of BIND results from a total serum bilirubin (TSB) level that exceeds the infant's neuroprotective defenses and leads to neuronal injury, primarily to the basal ganglia, central and peripheral pathways, hippocampus, brain stem nuclei for oculomotor function, and cerebellum. (1-5)

The damage ranges from minimal to severe with signs of ABE: kernicteric sequelae, isolated auditory neuropathy (a form of sensorineural hearing loss), extrapyramidal movement disorders, or a combination of neuromotor, sensorineural hearing disability, and visual disability. Some experts believe that BIND may have milder, subler neurologic manifestations, but this theory is unproven. (6-11)

The actual incidence of ABE is unknown for several reasons: No longitudinal surveillance studies of the condition have been performed; awareness and recognition of the diagnosis in healthy babies is limited; and the diagnosis usually is not coded on discharge summaries. Recent case reports and registries, however, suggest that kernicterus has reemerged as a public health problem after years of near extinction (see "Kernicterus makes a comeback," page 74). (5, 11, 12)

Clinical signs of ABE

The classic signs of ABE in the severely hyperbilirubinemic term infant (Figure 1) include increasing hypertonia--especially of extensor muscles and accompanied by retrocollis (spasmodic torticollis in which the head is pulled straight backward) and opisthotonus--along with varying degrees of drowsiness,...
The modified bilirubin induced neurologic dysfunction score assigned by residents was compared with the clinical diagnosis of acute bilirubin encephalopathy by expert consultants. Demographic information was obtained. Known risk factors were also evaluated among infants with and without acute bilirubin encephalopathy in addition to exploratory analyses. Data were analyzed by Statistical Analysis System; statistical significance was set at p < 0.05. Three hundred and thirty three paired modified bilirubin induced neurologic dysfunction scores (333) were analyzed and showed excellent agreement. Jaundice, or high bilirubin, is common in newborns. However, if improperly treated, it can result in a serious form of brain damage called kernicterus. What causes high bilirubin in newborns? Symptoms, diagnosis, and treatment of elevated bilirubin. What kinds of brain damage can jaundice cause? Can conjugated bilirubin cause harm? Legal help for infant jaundice, kernicterus | birth injury attorneys. Bilirubin encephalopathy is a rare neurological condition that occurs in some newborns with severe jaundice. Infants with the first signs of jaundice have bilirubin level measured within 24 hours. If the level is high, the infant should be screened for diseases that involve the destruction of red blood cells (hemolysis). All newborns have a follow-up appointment within 2 to 3 days after leaving the hospital. This is very important for late preterm or early term babies (born more than 2 to 3 weeks before their due date). Alternative Names. Bilirubin-induced neurologic dysfunction (BIND); Kernicterus. Patient Instructions. Newborn jaundice - discharge. Does bilirubin damage the brain of healthy infants? It has been claimed that “most studies have failed to substantiate significant association between a specific level of total serum bilirubin (TSB) during ‘non hemolytic’ hyperbilirubinemia in term newborns and subsequent IQ or serious neurologic abnormality (including hearing deficits)” [1]. Report and analysis of cases on healthy term and near-term infants who sustained kernicteric damage (contributed by colleagues and parents from a broad geographic area) during the period in question have been presented and recorded [9, 12, 13].