

**Kernicterus: a Preventable Disorder on the Rise due to Early Hospital Discharge**  
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**Introduction**

Two young parents come in with their first child. Her diagnosis includes cerebral palsy. The child is now 15 months old and had her first MRI at six months. The parents are puzzled because the pregnancy was uneventful, there was no fetal distress and the Apgars, whose import they now know, were 8 and 9. Although the baby came a few weeks early (35 ½ weeks), she weighed over 6 lbs. The mother breast fed the baby to give her the best possible start; it was difficult waking her to feed. After a discharge at 48 hours, the infant was readmitted five days after birth with severe jaundice. Following a period of phototherapy, the doctors sent the baby home. Her developmental milestones have been slow. Yet the child has never had seizures. Nor does she have the characteristic spasticity of most people with cerebral palsy. She seems "floppy," and the term athetotic cerebral palsy has been used by the medical personnel. The doctors are worried about her loss of upward gaze and seem concerned about her hearing as well. When she got her first teeth, they appeared yellow and weakened and broke off.

The family's expenses for special equipment have been mounting: a wheelchair, special collars and neck supports; there is talk of a feeding tube if nutrition cannot be maintained. The child does not crawl or walk or sit unaided. She isn't talking yet. But her intelligence seems normal, superior even. She is alert and interested in everything; her eyes sparkle. She works incredibly hard during her therapies. She wants to learn. She is a busy baby.

This child may or may not be seeing the same pediatrician who treated her at birth.

She may or may not have been diagnosed with kernicterus. The parents may or may not know that kernicterus was virtually eradicated decades ago with the advent of phototherapy and exchange transfusion. They probably do not know that slightly premature breast fed babies are most at risk for developing kernicterus.<sup>1</sup> Unless the child has been diagnosed and the parents are activists, they almost certainly do not know that: early discharge (48 hrs for a pre-term baby)<sup>2</sup> has given rise to unprecedented numbers of children with kernicterus; a Kernicterus Registry is maintained at the University of Pennsylvania School of Medicine by Dr. Lois Johnson and colleagues; a parents' group has formed called Parents of Infants and Children with Kernicterus (PICK); and the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) has issued a *SENTINEL EVENT ALERT*<sup>3</sup> regarding kernicterus.

### The Syndrome

Kernicterus is a preventable life-long neurologic syndrome caused by severe and untreated hyperbilirubinemia during the neonatal period. High levels of bilirubin are toxic to the developing newborn. In full-term infants, hyperbilirubinemia symptoms include

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<sup>1</sup> Ryan AS. The resurgence of breastfeeding in the United States. *Pediatrics* 1997. Available at <http://www.pediatrics.org> Accessed June 2001.

<sup>2</sup> Brown AK, Johnson L. Loss of concern about jaundice and the reemergence of kernicterus in full-term infants in the era of managed care. In: Fanaroff AA, Klaus MH, eds. *Yearbook of neonatal and perinatal medicine*. St. Louis, Missouri: Mosby Yearbook;1996:xvii--xxviii. Liu LL, Clemens CJ, Shay DK, Davis RL, Novack AH. The safety of newborn early discharge: the Washington state experience. *JAMA* 1997;278:293--8.

<sup>3</sup> Joint Commission on Accreditation of Healthcare Organizations. Sentinel event alert issue 18: kernicterus threatens healthy newborns; May 2, 2001. Available at <http://www.jcaho.org>. Accessed June 2001.

severe jaundice, lethargy, and poor feeding. There is hardly a parent or grandparent who has not seen or given birth to a baby who became jaundiced.<sup>4</sup> The characteristic color of the condition gives rise to the term "yellow jaundice." In a term, healthy baby bilirubin is transported into the bile and excreted in the stool. The jaundice does not become severe, nor is it usually dangerous to the baby. However, in pre-term and some term infants, bilirubin is not as successfully eliminated. The levels of bilirubin become elevated. This bilirubin can cross the blood/brain barrier and cause brain damage. The constellation of symptoms that result from this neurotoxicity is called kernicterus. Features of kernicterus may include choreoathetoid cerebral palsy, sensorineural hearing loss, dental dysplasia and gaze paresis. They may or may not include mental retardation. Many children with kernicterus have intelligence that is normal or above. Kernicterus is not a reportable condition in the United States, and its true prevalence is unknown.<sup>5</sup> However, prior to managed care and the early discharge of mothers and babies, it was as rare as polio. As one author has noted, "The problem was solved a generation ago."<sup>6</sup> Tragically, this is no longer the case.

As the medical literature makes clear, the key to the prevention of kernicterus is to control bilirubin levels. The key is, therefore, adequate perinatal care.

### The Problem

Managed care is implicated because jaundice peaks in the term infant between 48-

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<sup>4</sup> Each year approximately 60% of the 4 million newborns in the United States become clinically jaundiced.

<sup>5</sup> CDC June 15, 2001/50(23) 491-4.

<sup>6</sup> Valaes, Pediatrics, 1992.

72 hours, in the pre-term infant even later (4<sup>th</sup> -5<sup>th</sup> day). A baby who has been discharged early under an aggressive managed care cost containment policy will not be where its bilirubin can be measured or treated. The opportunities for negligent care increase exponentially once the baby who is at risk is discharged prematurely.<sup>7</sup> Bilirubin is measured by a heel stick (non-invasive methods are being developed as well). It cannot be measured at home. Neither the parents' nor the doctor's visual assessment of jaundice is likely to be accurate. Nor do all pediatricians have the capability to measure bilirubin in their offices. Weight loss and the need for supplemental feedings--which aid the metabolism of bilirubin--are less likely to be timely noted after discharge. New parents do not know if a newborn is feeding well; they cannot assess stool color or temperature instability like a nurse can. Readmission is necessary for intense photo therapy and exchange transfusion if levels are too high or if photo therapy fails. Since the advent of early discharge, the most frequent reason for an infant to be admitted to the hospital in the first week of life is the presence of significant jaundice.<sup>8</sup>

### **The Allegations**

**Thus, the first allegation of negligence may be early discharge of an at risk baby**

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- <sup>7</sup> **Major Risk Factors for Hyperbilirubinemia in Full-Term Newborns :**  
**Jaundice within first 24 hours after birth.**  
**A sibling who was jaundiced as a neonate.**  
**Unrecognized hemolysis such as ABO blood type incompatibility or Rh incompatibility.**  
**Nonoptimal sucking/nursing.**  
**Deficiency in glucose-6-phosphate dehydrogenase, a genetic disorder.**  
**Infection.**  
**Cephalohematomas/bruising.**  
**East Asian or Mediterranean descent.**

<sup>8</sup> **Seidman D, Stevenson DK, Ergaz Z, Gale R. Hospital readmission due to neonatal hyperbilirubinemia. Pediatrics 1995;96:727--9.**

with or without a bilirubin level at discharge. If the baby is preterm, that in itself is sufficient risk. Discovery will show the role of the health insurance plan versus the role of the discharging pediatrician. The second allegation is likely to be failure to control the bilirubin level in a timely manner.

### **The Standard of Care**

Hyperbilirubinemia is a medical condition for which algorithms abound. The standard of care for management of hyperbilirubinemia in the healthy term newborns is set out in the American Academy of Pediatrics (AAP) Practice Guidelines (October 1994).<sup>9</sup> In general, the guidelines describe when to administer photo therapy or exchange transfusion depending upon the age of the newborn and the bilirubin level. An example of the specificity of the guidelines will illustrate the difficulty a physician will have in defending a deviation. For example, for the treatment of the 25- to 48-hour-old infant, photo therapy may be considered when the Total Serum Bilirubin (TSB) level is >12 mg/. Photo therapy should be implemented when the TSB level is >15 mg/dL. If intensive photo therapy fails to lower a TSB level of >20 mg/dL, exchange transfusion is recommended. If the TSB level is >25 mg/dL when the infant is first seen, intensive photo therapy is recommended while preparations are made for an exchange transfusion. If intensive photo therapy fails to lower the TSB level, exchange transfusion is recommended.

### **The Experts**

It is important to note that these guidelines are for treatment of jaundice in the

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<sup>9</sup> American Academy of Pediatrics. Management of hyperbilirubinemia in the healthy term newborn. Pediatrics 1994;94:558--65. Available at [www.aap.org](http://www.aap.org)

healthy term newborn. Your client is likely to be sick or pre-term. Your expert will be able to extrapolate from these guidelines for the baby even more at risk. Evaluation of your case ideally should include a consult with one of the authors of the AAP's Practice Guidelines. The doctors who devised the above guidelines are listed by the AAP as the Subcommittee on Hyperbilirubinemia.<sup>10</sup> Since these world class experts have probably testified more often for the defense, having one on board makes a powerful statement about the strength of your case. Given the cost of these cases (between \$100,000-\$200,000), contacting a top medical expert in neonatal jaundice only makes sense. It is the most reliable way to determine for yourself and your client whether the case should be brought and can be won.

#### **The Defenses**

The first standard-of-care defense is that there is/was controversy about the dangers of hyperbilirubinemia. This defense is given credence by some of the language in the AAP's Practice Guidelines. For example, the defense will cite the statement, "Few issues in neonatal medicine have generated such long-standing controversy as the possible adverse consequences of neonatal jaundice and when to begin treatment. Questions regarding potentially detrimental neurologic effects from elevated serum bilirubin levels prompt continuing concern and debate. . . ."<sup>11</sup> The other principle defenses, as in birth injury

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<sup>10</sup> William O. Robertson, MD; Jon R. Almquist, MD; Irwin J. Light, MD; Jeffrey Maisels, MD; Thomas B. Newman, MD, MPH; Ronald W. Poland, MD; Gerald B. Merenstein, MD; James A. Lemons, MD; Lawrence M. Gartner, MD; Lois Johnson, MD; Audrey K. Brown, MD; David K. Stevenson, MD; Avroy Fanaroff, MD; James Cooley, MD.

<sup>11</sup> AAP Practice Guideline, Pediatrics (October 1994). See also Newman TB, Maisels MJ. Less aggressive treatment of neonatal jaundice and reports of kernicterus:

cases-- which this is not--are the causation defenses. They include, but are not limited to:

a) The statistical defense. Since most cerebral palsy springs from unknown causes; the cause here also is likely to be unknown.

b) The MRI shows periventricular leukomalacia (PVL), not kernicterus (injury to the globus pallidus). (A good pediatric neuroradiologist is necessary to test the validity of this defense. PVL can be distinguished from Kernicterus by MRI and also by the stormy newborn course, very early gestation, and severe respiratory distress requiring resuscitation--not the usual presentation of Hyperbilirubinemia.)

c) The infection defense. There either was, or was not, an adequate work-up for sepsis. An intrauterine infection is always a defense and sepsis should have been considered by the pediatrician and/or neonatologist who is/are your defendant(s). There are some experts who are always named in an infection defense whose identity is well-known among section listmates. In fact, however, infection *increases* the likelihood of brain damage from bilirubin, so its presence can be integrated into your case. In addition, the characteristics of a viral encephalopathy are different from those of kernicterus. There you would expect hydrocephalus or microcephalus, calcifications on the MRI, heart defects, blindness, spastic cerebral palsy, global brain damage and/or multiple small hemorrhages throughout the brain.

d) The brain damage is from hypoxia (though not during labor and delivery, which would give rise to accountability). Your neonatologist can distinguish hypoxia from bilirubin encephalopathy. The defense may be valid if in your case there are low Apgar scores,

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lessons about practice guidelines. Pediatrics 2000;105:242--5.

seizures, low Ph, apnea, coma, multiple organ failure, increased intra cranial pressure, intrauterine growth retardation, fetal distress, and/or an abnormal placenta.

e) The placenta is abnormal, which shows an earlier injury than one due to jaundice.

There are myriads of defenses related to the placenta that will need to be considered by your geneticist, neonatologist, placental pathologist, and other experts. In a pure kernicterus case, the placenta is likely to have been normal and not sent to pathology.

f) The underlying cause is genetic. For this defense it is important to remember that a genetic predisposition to jaundice is not a defense; the treatment options are the same, whatever the cause of the elevated jaundice is.

These defenses, in the face of elevated bilirubin levels and a diagnosis of kernicterus, tend to be at once both predictable and creative. You must know the medical literature related to these and other defenses to properly cross-examine the defense experts on them at deposition and trial.

### **Final Thoughts**

The best plaintiff's cases have a diagnosis of kernicterus by a treating neonatologist or developmental pediatrician who was aware of the prenatal, labor and delivery, and neonatal history when he or she made the diagnosis. If the child has no diagnosis at the time of the initial client consult, helping to find the most highly qualified subsequent treating physician is crucial. If the child is still seeing the original doctors who failed to adequately prevent or treat the severe jaundice, the diagnosis of kernicterus may not have been made or may not have been communicated to the parents.

For various reasons, you may wish to file a Motion to prevent subsequent treating physicians from being asked standard of care questions by the defense. Other Motions to

**file include protective orders to prevent production of sibling medical records and protective orders to prevent defense medical exams (DME). The Motions and supporting briefs are on file with ATLA.**

**Because the damages are so astronomical, the cases may be fought via the scorched earth policy. If you and your client's physicians are satisfied that the child has kernicterus, however, there is not more satisfying case to work up and try. Hopefully, good verdicts and settlements along with the JCAHO Sentinel Alert will raise the level awareness of insurance and their doctors to again make kernicterus a thing of the past.**

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