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KERNICTERUS RISK IN 2010

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WHAT IS KERNICTERUS?

Kernicterus is a very rare condition afflicting full-term and pre-term newborns. Caused by severe neonatal hyperbilirubinemia (jaundice), it often has tragic consequences: death or profound neurological impairment.[I] The precise incidence of kernicterus is not known, but studies show that, at a minimum, 10% of the neonates (a newborn in the first 28-30 days of life) with this rare diagnosis will die and 70% of those surviving will have lifelong impairment. Treatment is either phototherapy (so-called "bili lights") or, for severe cases, exchange transfusions.[II]

Trends in early post-partum hospital discharges beginning in the 1970s raised concerns in the pediatric community about the identification and treatment of elevated bilirubin levels. These concerns caused the American Academy of Pediatrics (AAP) to promulgate treatment guidelines in 1994 and again in 2004. The National Quality Forum (NQF) deemed kernicterus a "never event." The Joint Commission on Accreditation of Healthcare Organizations (JCAHO) has issued Sentinel Event Alerts on kernicterus twice in the last decade (2001 and 2004). Despite this increased attention to the problem and the attempts to address it, kernicterus cases still occur.[III] Recent articles in the pediatric literature suggest that kernicterus can occur even when appropriate care is provided.

This article will focus on liability trends and risk management implications for kernicterus.

PEDIATRIC LIABILITY TRENDS

The national medical malpractice environment remains stable in 2010. The industry combined ratio indicates profitability, that claim frequency has dropped markedly and claim severity has moderated. The analysis of national medical malpractice claim

trends reveal two types of claims as problematic for severity: those arising in either obstetric or pediatric settings.

There are three reasons for this. First, while enactment of damage caps in many states since 2000 has helped improve the national malpractice environment, non-economic damage caps do not help defendants very much in any case involving a severely injured child. Second, long-term survival of these children has improved over the last 20 years due to medical advances and therefore the life-care plans are expensive, especially as health care costs have risen dramatically over the decades. Finally, the sympathy factor makes these cases difficult to defend, especially at trial.

Four types of recurring pediatric cases in the category of high frequency/low severity are

particularly problematic: 1) meningitis, 2) malrotation of the bowel/volvulus, 3) retinopathy of prematurity (ROP) and 4) kernicterus. Verdicts and settlements in these cases are often in multiple millions of dollars. The trial bar seeks out these high-damage cases through diverse advertising strategies, especially through the internet.[IV]

Meningitis and malrotation of the bowel cases frequently involve misdiagnosis and/or delayed treatment. However, retinopathy of prematurity cases and kernicterus cases often involve system breakdowns resulting in neonates not receiving requisite initial eye examinations or subsequent follow-up examinations (ROP); or initial and follow-up testing and/or treatment for jaundice/highly elevated bilirubin levels in the blood (hyperbilirubinemia) resulting in kernicterus. ROP occurs only in low birth-weight neonates treated in Level II or Level III Neonatal Intensive Care Units (NICUs). Kernicterus cases can arise from neonates treated in any newborn nursery, not just NICUs. All neonates, even those full-term and seemingly healthy at birth, are at risk for developing elevated bilirubin levels, which undetected and/or untreated, can become extreme hyperbilirubinemia and result in kernicterus, a "rare but highly preventable condition," that manifests itself in profound permanent neurological impairment.[V]

Despite increased attention from many national organizations focused on patient safety over the last 10 years, particularly the JCAHO, the NQF and the AAP, kernicterus cases still occur.[VI] Malpractice verdicts and settlements in kernicterus cases, including those in recent years, are very large, almost always in multiple millions of dollars.

A CLINICAL OVERVIEW

Jaundice is not uncommon in many newborns; in most cases it is benign. However, due to the potential neuro-toxicity of bilirubin, newborns must be monitored to identify those at risk for severe hyperbilirubinemia and those relatively few cases of newborns at risk for acute bilirubin encephalopathy, or kernicterus.[VII] Kernicterus is defined as a rare neurologic syndrome that results in severe brain damage or death of the affected infant caused by exposure to hyperbilirubinemia (highly elevated levels of bilirubin). Highly elevated levels of bilirubin are neuro-toxic in a developing neonate, as for some full-term infants and especially for some pre-term infants, the bilirubin crosses the blood brain barrier and can result in permanent brain damage. The AAP recommended that the term "kernicterus" be used solely for infants that exhibit the signs of "chronic and permanent clinical sequelae of bilirubin toxicity."[VIII]

POSTPARTUM CARE

The incidence of kernicterus prior to 1990 was very low, especially in full-term infants. Possibly, the now greater risk of exposure to high levels of hyperbilirubinemia in full-term and near-term infants especially, is due to the greater number of breast-feeding mothers with early post-partum hospital discharges (termed "drive-thru deliveries"), inconsistent post-discharge care and follow-up, and a lack of concern about the danger of high bilirubin levels among providers.[IX]

This paradigm shift in postpartum care and the increased incidence of breastfeeding created health care delivery system challenges that were not immediately recognized and addressed by providers. It is very important for health care professionals to observe the newborn for evidence of hyperbilirubinemia during the period that the infant is at risk, as set forth in AAP or American Academy of Family Practice (AAFP) clinical practice guidelines.

ADDRESSING THE PROBLEM

1994	**AAP ISSUES A CLINICAL PRACTICE GUIDELINE FOR THE MANAGEMENT OF HYPERBILIRUBINEMIA IN HEALTHY, FULL-TERM NEWBORNS, RAISING KERNICTERUS AS A PEDIATRIC TREATMENT PROBLEM.**[X] (The guidelines did not necessarily change clinical practice immediately. However, an article by Burke et al. in *Pediatrics* in February 2009 covered their analysis of Healthcare Cost and Utilization (HCUP) data from 1988-2005, concluding that there was "...a substantial reduction in hospitalizations with a diagnosis of kernicterus" in that time frame, although admittedly difficult to prove that the drop was due merely to the issuance of the guidelines.)[XI]
1990s	**CONCERTED EFFORTS TO ADDRESS THE PROBLEM OF KERNICTERUS BEGIN WITH THE MAJIC PROJECT, A CONSORTIUM TO IMPROVE CARE FOR NEWBORNS WITH HYPERBILIRUBINEMIA FUNDED BY THE AGENCY FOR HEALTHCARE RESEARCH AND QUALITY AND INCLUDING THE AAP AND OTHERS.**[XII]
2000	**OCTOBER - PARENTS OF INFANTS AND CHILDREN WITH KERNICTERUS (PICK) IS FOUNDED.** (One of their primary goals is working with health care delivery systems to help protect newborns and prevent future cases of kernicterus, employing system-based approaches that include such measures as universal pre-discharge bilirubin screenings.)[XIII]
2001	**APRIL - THE JCAHO ISSUES ITS FIRST SENTINEL EVENT ALERT (SEA) ON PREVENTION OF KERNICTERUS: "KERNICTERUS THREATENS HEALTHY NEWBORNS."**[XIV]
2002	**THE NQF DEEMS KERNICTERUS A "NEVER EVENT" IN ISSUING ITS FIRST LISTING OF THESE INCIDENTS.** The term "Never Event" referred to particularly egregious medical errors (such as wrong-site surgery) that should never occur. Over time, the list of "Never Events" has been expanded by NQF to signify adverse events that are unambiguous (clearly identifiable and measurable), serious (resulting in death or significant disability), and usually preventable. The NQF initially defined 27 such events in 2002, including "death or disability (kernicterus) associated with failure to identify and treat hyperbilirubinemia in neonates."[XV]
2004	**JULY - AAP ISSUES UPDATED CLINICAL PRACTICE GUIDELINE: "MANAGEMENT OF HYPERBILIRUBINEMIA IN THE NEWBORN INFANT 35 OR MORE WEEKS OF GESTATION."**
2004	**AUGUST - JCAHO ISSUES ANOTHER SENTINEL EVENT ALERT PROVIDING REVISIONS FOR THEIR 2001 ALERT.**[XVI] The JCAHO's revisions are based on the AAP July updated guidelines.

AAP GUIDELINES

The AAP guidelines suggest that if their recommendations are followed by health care professionals, kernicterus would be largely prevented. The guidelines also emphasize the importance of universal, systematic assessment for the risk of severe hyperbilirubinemia, close follow-up and prompt intervention when indicated. The recommendations apply to the care of infants at 35 or more weeks of gestation and seek to further the aims defined by the Institute of Medicine as appropriate for health care: safety, effectiveness, efficiency, timeliness, patient-centeredness and equity. They specifically emphasize the principles of patient safety and the key role of timeliness of interventions to prevent adverse outcomes resulting from neonatal hyperbilirubinemia.[XVII]

FOLLOWING ARE KEY ELEMENTS OF THIS GUIDELINE. CLINICIANS SHOULD:

1. Promote and support successful breastfeeding
2. Establish nursery protocols for the identification and evaluation of hyperbilirubinemia
3. Measure the total serum bilirubin (TSB) or transcutaneous bilirubin (TcB) level on infants jaundiced in the first 24 hours
4. Recognize that visual estimation of the degree of jaundice can lead to errors, particularly in darkly pigmented infants
5. Interpret all bilirubin levels according to the infant's age in hours

MALPRACTICE CLAIM TRENDS

Drs. Newman and Maisels, in a joint letter to the editor of the journal *Pediatrics,* summarized the most common issues in kernicterus malpractice cases:

> "Although quantitative literature on this point is admittedly sparse, the causal relationship between hyperbilirubinemia and kernicterus is generally not disputed. In our experience, however, most malpractice cases involving kernicterus do not revolve around the question of whether a specific bilirubin level produced brain damage or whether an intervention at a lower bilirubin level would have prevented the damage. More commonly, cases involve infants admitted to the hospital with bilirubin levels of >35 mg/dL who manifest the classical signs of acute bilirubin encephalopathy. In these cases...it can be asserted that "with reasonable medical probability" that if this infant had been seen and treated earlier, the bilirubin would not have risen to dangerous levels and the kernicterus could have been prevented. Whether someone is to blame for this sequence of events, however, is often the most contentious issue, and specifically whether there was a breech in the standard of care."[XVIII]

The first decade of the 21st century has seen malpractice verdicts involving kernicterus with very large settlements. The table below contains selected settlements we are aware of. It is based on our own informal research and does not represent all court verdicts.

KERNICTERUS

MEDICAL MALPRACTICE VERDICTS/SETTLEMENTS

DATE	LOCATION	AMOUNT (in millions)
12/09	California	$5
7/07	California	$15.4
4/06	Boston	$5
4/04	Chicago	$30
Unknown	Minnesota	$3.08

CASE IN POINT

Allegation: The plaintiff alleged that several defendant physicians failed to diagnose and treat jaundice in a newborn infant in the first weeks of life, resulting in severe, lifelong brain damage.

In this case, a newborn healthy baby was discharged from the hospital without any instructions to the mother about the signs and symptoms of jaundice, such as abnormal coloring and difficulty feeding. Evidence discovered by counsel showed that, at the time the infant was discharged from the hospital the day after birth, photographs taken by the family showed differences in skin color, which should have led the physicians to recognize the development of jaundice. The baby's jaundice went unnoticed and untreated by her doctors.

When the infant was five days old, the pediatrician, who was scheduled to see the patient at two weeks of age, was informed of feeding problems and skin and eye discoloration. The mother was instructed to take the infant to the hospital the following morning for bilirubin testing for jaundice. However, the urgent need for testing was not communicated to the parents. Several crucial hours passed before the child was brought in for testing An exchange transfusion was ordered but was not carried out for seven hours.

SETTLEMENT: $5 MILLION

The theories driving the verdict of $5 million dollars in the case were:

- Failure to instruct the mother at the time of discharge on the signs of neonatal jaundice
- Failure of the pediatrician to stress the need for immediate testing to the parents
- Failure of the hospital to perform the exchange transfusion in a timely manner[XIX]

RISK MANAGEMENT TECHNIQUES

- Techniques for preventing kernicterus revolve around proper patient instructions, early diagnosis and prompt intervention. The **AAP has toolkits** available for hospitals and physicians' offices that include discharge readiness checklists, follow-up letters, inventories, assessments and documentation tools and parent handouts.[XX]

- Discharge assessment for risk factors for developing hyperbilirubinemia should be done and documented.
- Discharge instruction for kernicterus must include documentation that the parents understand the signs and symptoms of jaundice as well as the emergency nature of this condition.
- Parents need to be instructed on hyperbilirubinemia and when to call the physician.
- Testing must be done in a timely manner and treatment instituted immediately.
- Compliance with the AAP Guideline (most current version as of this publication is the 2004 Guideline) is essential to establish practice within the current standard of care, although recent findings as set forth below suggest that not all cases can be prevented. Providers can help promote patient safety by being current with the latest medical literature findings.

RECENT DEVELOPMENTS

Now, in 2010, there is some question as to whether kernicterus should have been deemed a "never event." The implication by the NQF was that if elevated bilirubin levels were monitored appropriately and there was timely intervention (primarily phototherapy and, for severe cases, exchange transfusions), kernicterus could be eliminated. Because of recent articles, the current state of clinical knowledge regarding the kernicterus problem is more muddled.

Over the last year, authors such as Maisels et al. state that while elimination of all cases of kernicterus is a worthy goal, certain clinical presentations, such as a particular glucose deficiency (called G6PD), a genetic predisposition, sepsis and "other unknown stressors" may cause severe hyperbilirubinemia and "can produce brain damage despite appropriate monitoring and intervention."[XXI]

In that same October 2009 issue of *Pediatrics*, Trikalonis et al. suggested that "although screening can predict hyperbilirubinemia, there is no robust evidence to suggest that screening is associated with favorable clinical outcomes."[XXII] The U.S. Preventive Services Task Force went even further in a Recommendation Statement issued in October 2009 concluding "...the evidence is insufficient to recommend screening infants for hyperbilirubinemia to prevent chronic bilirubin encephalopathy (kernicterus)." They go on to state that:

> "Early treatment can decrease the number of infants with elevated serum bilirubin levels. However, the USPSTF found inadequate evidence that treating elevated bilirubin levels in term or near-term infants to prevent severe hyperbilirubinemia resulted in the prevention of chronic bilirubin encephalopathy."[XXIII]

The USPSTF also challenges the clinical connection between hyperbilirubinemia and kernicterus "in every case,"[XIV] and states that "...not all children with chronic bilirubin encephalopathy have a history of hyperbilirubinemia."[XV]

Maisels et al. recommend in their article that that at present, and until there is more clinical evidence, all newborns receive pre-discharge screening and the follow-up management as recommended by the AAP "in the absence of better evidence."[XVI]

They state:

> "...it is our opinion that universal screening, when combined with the clinical risk factors (of which gestational age and exclusive breastfeeding are most important) and targeted follow-up, is a systems approach that is easy to implement and understand, and it provides a method of identifying infants who are at high or low risk for the development of severe hyperbilirubinemia." [XVII]

Debate continues in the pediatric community on the approach to preventing kernicterus. The current use of TBS screening was challenged in the same October 2009 issue of *Pediatrics* by Fay et al., suggesting that the TBS test is flawed due to sample, subject follow up, and the fact that positive hyperbilirubinemia test results did not always correlate with kernicterus.

CONCLUSION

Kernicterus is a devastating disorder that can result in lifelong disability or death of a child. The magnitude of this problem has been recognized by both the AAP and the Joint Commission on Accreditation of Hospitals. JCAHO issued Sentinel Event Alerts after guidelines had been created by AAP. Kernicterus cases are an area of specialty for many plaintiffs' lawyers due to the severe nature of the disorder and potentially expensive life-care plans. But recent findings suggest that at least some of these cases may be successfully defended on a causation basis if there is compliance with current screening and treatment guidelines.

Effective assessment of the newborn for risk factors, timely testing and thorough discharge planning are essential in preventing many cases of kernicterus, but recent articles in the pediatric literature suggest that not all cases can be prevented, even with optimal screening and follow-up. Compliance with the AAP guidelines is essential to kernicterus prevention and creating a defensible case at present, although advancements in the understanding and treatment of the effects of hyperbilirubinemia on kernicterus will alter these guidelines in the future.

[I] Ip,S, Chung M.,Kulig, J, O'Brien, R. et al., "An evidence-based review of important issues concerning neonatal hyperbilirubinemia," *Pediatrics,* Vol 114, No.1, 2004, (abstract) pp. 130, July 2004.

[II] US Preventive Services Task Force, "Screening of Infants for Hyperbilirubinemia to Prevent Chronic Bilirubin Encephalopathy: US Preventive Services Task Force Recommendation Statement," *Pediatrics,* Vol.124, No.4, October 2009, pp. 1172-1177, at 1175.

[III] Fay, David L. M.D. et al., "Bilirubin Screening for Normal Newborns: A Critique of the Hour-Specific Bilirubin Nomogram," *Pediatrics,* Vol. 124, No.4, October 2009, pp. 1203-1205, at 1203.

[IV] Author's note: Using Google or other search engines to search on the key words "kernicterus" and "malpractice" will result in a lengthy listing of a number of plaintiff's law firm sites, such as "New Jersey Jaundice Lawyers."

[V] Sentinel Event Alert, "Revised Guidance to Help Prevent Kernicterus," Issue 31, August 31, 2004, JCAHO.

[VI] Fay et al., Ibid, p. 1203.

[VII] AAP, Clinical Practice Guideline, "Management of Hyperbilirubinemia in the Newborn Infant 35 or More Weeks of Gestation," *Pediatrics,* Vol.114, No.1, July 2004, pp. 297-316, and www.pediatrics.aappublications.org/cgi/content/full/114/1/297, pp. 1-45 at p. 1.

[VIII] Ibid., p.3

[IX] Harris, Mary Catherine M.D. et al., "Developmental Follow-Up of Breastfed Term and Near-Term Infants with Marked Hyperbilirubinemia," *Pediatrics,* Vol.107, No.5, May 2001, pp. 1075-1080, at 1078.

[X] Ibid.

[XI] Burke, Bryan M.D. et al., "Trends in Hospitalizations for Neonatal Jaundice and Kernicterus in the United States, 1988-2005," *Pediatrics,* Vol.123, No.2, February 2009, pp. 524-532, at 531.

[XII] Ibid.

[XIII] Sheridan, Susan, "Parents of Infants and Children with Kernicterus," Journal of Perinatology, Vol.25, No.4 2005, pp. 227-228, at 227.

[XIV] Sentinel Event Alert, JCAHO, Issue 18, April 2001.

[XV] AHRQ, "Never Events," www.psnet.ahrq.gov.primer.aspx?primerID=3.

[XVI] Sentinel Event Alert, JCAHO, Issue 31, August 2004.

[XVII] The Joint Commission, "Revised Guidance to Help Prevent Kernicterus" www.jojintcommission.org/SentinelEvents/SentinelEventAlert/sea_31.htm.

[XVIII] Maisels, M. Jeffrey M.D. and Newman, Thomas B. M.D., "Kernicterus, the Daubert Decision, and Evidence-Based Medicine," Letter to the Editor, *Pediatrics,* Vol.119, 2007, at p.1038,available at www.pediatrics.org.

[XIX] Verdicts and Settlements http://www.kernicteruslaw.com/verdicts/#5mil.

[XX] American Academy of Pediatrics, http://practice.aap.org/content/aspx?=2577.

[XXI] Maisels, M. Jeffrey et al., "Hyperbilirubinemia in the Newborn Infant >35 Weeks Gestation: An Update with Clarifications," *Pediatrics,* Vol.24, No. 4, October 2009, pp. 1193-1198, at 1193.

[XXII] Trikalonis, Thomas A. M.D. et al., "Systematic Review of Screening for Bilirubin Encephalopathy in Neonates," *Pediatrics,* Vol124, No.4, October 2009, pp. 1162-1170, at 1162.

[XXIII] US Preventive Services Task Force, Screening of Infants for Hyperbilirubinemia to Prevent Chronic Bilirubin Encephalopathy: USPSTF Task Force Recommendation Statement, *Pediatrics,* Vol.124, No.4, October 2009, pp. 1172-1177, at 1174.

[XXIV] Ibid at 1175.

[XXV] Ibid at 1173.

[XXVI] Maisels et al. at 1197.

[XXVII] Ibid.

[XXVIII] David L. Fay, M.D., Kenneth G. Schelhase, M.D., MPH, and Gautham K. Suresh, "Bilirubin Screening for Normal Newborns: A critique of the hour-Specific Bilirubin Nomogram, *Pediatrics,* Vol 124, Number 4, October 2009.

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