Neonatal Jaundice, Hyperbilirubinemia, and Kernicterus for the Medical Negligence Litigator

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Anyone who has had a child or spent any time around newborn babies probably has at least some informal experience with neonatal jaundice (icterus), caused by elevated levels of bilirubin. For litigators handling birth trauma cases, however, superficial familiarity is not enough. Although jaundice in the newborn is common and usually benign, it nevertheless remains an issue of concern in the medical community, because of one significant risk associated with elevated bilirubin levels: kernicterus.[1]

The term kernicterus was first used in 1904 by Christian Georg Schmorl to describe yellow staining in the basal ganglia of infants who died with severe jaundice.[2] Since that time researchers and physicians have used the term to describe: (1) an acute illness associated with jaundice, lethargy, poor feeding, changes in muscle tone, a characteristic type of posturing called opisthotonus, fever, and a high-pitched cry; (2) the neuropathological findings (yellow staining of brain tissue) on autopsy of infants who died during or after the acute illness; or (3) a chronic condition seen in those who survive the acute illness, characterized by athetoid cerebral palsy.[3]

Kernicterus is a rare but proven cause of cerebral palsy.[4] It is rare because it can be, and usually is, prevented by the exercise of good medical care. Despite the fact that it is now considered a "never event" by medical professionals,[5,6] and although it is extremely rare, kernicterus is not extinct and cases still occur in the United States. Experts agree, however, that kernicterus is *the most easily preventable cause of neonatal mortality and brain damage in the United States*.[7,8,9] As a preventable "never event" associated with a catastrophic signature injury, every case of kernicterus is also a potential claim for medical malpractice. A true kernicterus case may cross your path but once in your entire legal career—what follows is the information you need to recognize and pursue it.

Understanding bilirubin metabolism: the basics

To understand and manage the issues pertinent to a kernicterus claim, one must first have an understanding of what bilirubin is, and how it is metabolized in the body. Bilirubin has traditionally been discussed in the literature as a 'waste product' created by the metabolism of red blood cells, although it has been shown to have some protective, antioxidant effects as well.[10] Make no mistake, however—bilirubin, at the high levels discussed in this article, is a neurotoxin.

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[1] Maisels, MJ. Jaundice in a Newborn: Answers to questions about a common clinical problem. First of Two Parts. *Contemporary Pediatrics*, May 1, 2005.

[2] Schmorl, G. Zur Kennntnis des Ikterus Neonatorum, *Verh. Dtsch. Pathol. Ges.* 1904; 6:109-115.

[3] Maisels MJ and Watchko JF, Eds. *Monographs in Clinical Pediatrics: Neonatal Jaundice. Chapter 7: The Pathophysiology of Bilirubin Toxicity*. 2000. Harwood Academic Publishers, The Netherlands.

[4] Maisels, MJ. 2005, Supra, Note 1.

[5] Bhutani, VK. Kernicterus as a 'Never Event': A Newborn Safety Standard? *Indian J. Pediatr* 2005; 72 (1): 53-56;.

[6] Bhutani, VK, Johnson L. Kernicterus: A Preventable Neonatal Brain Injury. *J. Arab Neonatal Forum* 2005; 2:12-24.

[7] Bhutani VK 2005, Supra, Note 5.

[8] Johnson LH, Brown AK, Bhutani VK. System-based Approach to management of neonatal jaundice and prevention of Kernicterus. *J. Pediatr* 2002; 140:397-86.

[9] Bhutani VK, Johnson LH, Maisels JM, Newman TB, Phibbs C, Start AR, et al. Kernicterus: epidemiological strategies for its prevention through systems-based approached. *J. Perinatol* 2004Oct: 24(10):650-662.

[10] See, e.g., Neužil J and Stocker R: Free and albumin-bound bilirubin are efficient co-antioxidants for ά-tocopherol, inhibiting plasma and low density lipoprotein lipid peroxidation. *J. Bio. Chem.* 1994; 269(24):16712-16719; and Stocker R, Glazer, AN, Ames BN: Antioxidant activity of albumin-bound bilirubin. *Proc. Natl. Acad. Sci.* 1987; 84:5918-5922.

The creation of bilirubin typically begins with the death of red blood cells (erythrocytes). Red blood cells have a lifespan of approximately 45-50 days in newborns.[11] They can, of course, die sooner as a result of trauma, hemolytic disease, and a number of other causes. When red blood cells die, the body's normal physiologic processes break them down and eliminate the resulting by-products.

Most people are aware that red blood cells are the main source of the iron-containing heme molecule, which facilitates the transport of oxygen and carbon dioxide in the bloodstream in the form of hemoglobin. Heme, therefore, is one of the key components of red cells that must be metabolized and eliminated when these cells die. Heme from a few other sources also contributes to bilirubin production, but the vast majority of bilirubin formed in the body is produced by the normal metabolic breakdown of red blood cells.[12]

Bilirubin has a yellow pigment, and is eliminated from the body in urine, stool, bile, and other secretions. Bilirubin is what lends a yellow/brown hue to normal urine, bile, and stool. Unless or until it is modified by the body, bilirubin is not soluble in water (hydrophobic). Blood plasma, however, is 91% water[13], which makes it a less-than-hospitable transport medium for bilirubin. Therefore, hydrophobic bilirubin travels through the bloodstream bound to the blood protein albumin.

Because bilirubin is naturally insoluble in water, it cannot be eliminated from the body efficiently[14] unless or until it is converted into water-soluble form. This conversion process, an enzyme-mediated chemical reaction known as "conjugation," takes place in the liver. Once it arrives, the albumin releases it and the liver takes over. Thus, bilirubin comes in two varieties: conjugated (a.k.a. "direct" or "direct-reacting") and unconjugated (a.k.a. "indirect" or "indirect-reacting").

Simply stated, the normal processing system for bilirubin involves moving it into the circulation, where it is then bound to albumin, transported to the liver, unbound from albumin, conjugated, and ultimately, excreted in the bile, urine, and feces. Any condition that disrupts this process, at any stage in the process, can result in the accumulation of excess bilirubin in the body. Accumulated bilirubin may be bound or unbound, and conjugated or unconjugated, depending on the mechanism and timing of the interruption to the process.

For instance, a shortage of the enzymes needed by the liver for the conjugation reaction (not an unusual occurrence in newborns, whose livers are immature) is a common cause of elevated unconjugated bilirubin levels. Similarly, if there is a shortage of albumin, bilirubin may linger unbound, and therefore unable to travel to the liver for processing and elimination. Generally speaking, any process working to break down red blood cells (creating more bilirubin), such as traumatic delivery resulting in extravasation of blood (hematomas or bruising), hemolytic disease, coagulopathies, maternal-fetal blood incompatibilities, sepsis, polycythemia, and/or macrosomia in the infant of a diabetic mother, will work to increase the blood concentration of bilirubin.

Likewise, any process that serves to decrease bilirubin clearance, such as prematurity, glucose-6-phosphate dehydrogenase ("G6PD") deficiency, genetic disorders such as Gilbert's Syndrome or Crigler-Najjar Syndrome, and metabolic disorders such as hypothyroidism or hypopituitarism, will work to increase serum concentrations as well.

Serum bilirubin concentration is measured clinically in milligrams per deciliter of blood (mg/dL). Total Serum Bilirubin ("TSB") concentration is the sum of the conjugated and unconjugated bilirubin present in the blood serum. The lab reports in your client's medical records may report the TSB level only, or may break that level down into its component parts, which is obviously

[11]Maisels MJ and Watchko JF, Eds. 2000, Supra, Note 3, *Chapter 1: Fetal and Neonatal Bilirubin Metabolism*, p.3.

[12]Small amounts of bilirubin also derive from ineffective erythropoiesis in the bone marrow and/or the breakdown of tissue heme and heme proteins in the liver.

[13]Patton, KT. *Survival Guide for Anatomy and Physiology.* Elsevier Mosby, 2006, St. Louis, MO., at p. 225.

[14]Phototherapy and exchange transfusion can assist in eliminating unconjugated bilirubin from the body. More information on the use of medical therapies to eliminate bilirubin follows.

more useful. The differentiation between conjugated and unconjugated bilirubin is important because kernicterus is only caused by *unconjugated hyperbilirubinemia.*[15]

Assessing jaundice and measuring bilirubin levels

Jaundice is a clinical manifestation of hyperbilirubinemia, characterized by a yellow discoloration, visible to the naked eye, of the whites of the eyes (sclera), skin, and secretions. It progresses from the head downward, showing up first in the eyes and face and moving downward from there.[16] Like most phenomena in medicine, jaundice manifests along a continuum; some patients have an obvious, dark discoloration while color change in others is more subtle.

Importantly, the degree of discoloration does not necessarily correlate to the actual TSB level. In other words, some babies will be severely discolored with only a mild elevation in their TSB concentration, while others can have significantly elevated TSB levels with little or no noticeable change in color. Additionally, race can play a role in confounding a clinician's ability to identify jaundice, which can be both more prevalent[17] and less noticeable[18] in darker-skinned infants. While it is generally true that visible jaundice confined to the face represents a lower TSB concentration than jaundice throughout the whole body, this is most useful when comparing levels taken from the same patient, as a means of watching progression.[19] **It is not possible to accurately and reliably estimate the TSB levels by visual assessment alone**. Therefore, it is not sufficient to visually estimate the *bilirubin levels* by simply looking at the baby and assessing the degree of jaundice present.[20]

Current technology allows for transcutaneous measurement of the bilirubin level (TcB), obviating the need for a blood draw in some cases. The TcB and TSB tests are not interchangeable, however, and the TSB is more accurate.[21] Generally TcB measurements are used for screening purposes, where a result over a certain level (determined by the institution's internal protocols) will prompt an actual TSB measurement and closer evaluation. Given the unreliability of visual assessment of bilirubin levels based on the observable degree of jaundice, coupled with the ability to obtain pain-free, low-cost, non-invasive measurements of TcB levels, there is no legitimate justification in modern medical facilities for failing to measure the bilirubin levels in a jaundiced infant. Indeed, even the procedure required for a TSB measurement is relatively low-cost and involves only a blood draw, which, while associated with some discomfort for the patient, is poses little risk, cost, or disadvantage.

Physiologic versus pathologic jaundice

Not all jaundice is cause for alarm. In fact, most neonatal jaundice is easily and routinely managed and therefore perfectly harmless. Although up to 60% of full-term newborns have clinical jaundice in the first week of life, few have significant underlying disease.[22] Most neonatal jaundice is the result of normal physiologic processes that are self-limiting in nature, and need not be associated with poor outcomes or a danger to the baby.

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[15] Hyper-bilirubin-emia: from hyper- (too much); bilirubin; -emia (in the blood)

[16] Maisels, MJ. 2005, Supra, Note 1.

[17] See, e.g., Huang MJ, Kua KE, *et. al.*, Risk Factors for Severe Hyperbilirubinemia in Neonates. *Pediatric Research* 2004; 56(5):682-689; and Halamek LP, Stevenson D. Diseases of the fetus and infants, in Fanaroff AA, Martin RJ, eds. *Neonatal-Perinatal Medicine.* 1997 Mosby, St. Louis, MO. pp.1345-1389.; and Watchko, JF. Genetics and the Risk of Neonatal Hyperbilirubinemia. *Pediatric Research* 2004; 56(5): 677-678.

[18] Maisels, MJ. 2005, Supra, Note 1.

[19] Maisels MJ and Watchko JF, Eds. 2000, Supra, Note 3, *Chapter 10: The Clinical Approach to the Jaundiced Newborn*, p. 144.

[20] Kramer LI: Advancement of dermal icterus in the jaundiced newborn. *Am. J. Dis. Child.*1969; 118:454; and Bhutani VK, Meloy LD, Poland RL, *et. al*. Correlation of clinical assessment of jaundice, transcutaneous and total serum bilirubin levels in health term and near-term infants. *Pediatr. Res.* 2004; 55:591A; and Moyer, VA, Ahn C, Sneed S: Accuracy of clinical judgment in neonatal jaundice. *Arch. Pediatr. Adolesc. Med.* 2000; 113:1628.

[21] Maisels, MJ. 2005, Supra, Note 1.

[22] Porter, ML, Dennis BL. Hyperbilirubinemia in the Term Newborn. *American Family Physician* 2002; 65(4):599-606, 599.

Often, neonatal jaundice is simply the result of the baby's new liver requiring time to attain full functional capacity. During pregnancy, the placenta does the work of clearing the baby's system of bilirubin, but after birth, that role is assumed by the liver.[23] This transition of responsibility for bilirubin management from the placenta to the liver is often accompanied by a transient rise in serum concentrations of bilirubin, and therefore, predictably, some jaundice. In such cases, when the cause of jaundice is unrelated to disease, and providing that the levels do not rise high enough to be considered a risk to the patient, it is referred to as *physiologic jaundice*. On the other hand, when the cause of the jaundice is disease, or the severity of the hyperbilirubinemia presents a risk to the patient, it is referred to as *pathologic jaundice.* Newborn babies typically have physiologic, not pathologic, jaundice.[24]

Physiologic jaundice in the newborn is generally predictable and follows a typical pattern. The average TSB level usually peaks at 5-6mg/dL on the third or fourth day of life, and then declines over the first week after birth.[25] Serum concentrations in most cases of physiologic jaundice do not exceed 12mg/dL when their levels peak, but occasionally when multiple risk factors are present, cases of physiologic jaundice can include peak levels as high as 17mg/dL. After the first week of life, the liver will usually have reached a level of function sufficient to maintain the bilirubin levels within normal limits.[26]

By contrast, there are a number of risk factors[27] for hyperbilirubinemia in the full-term newborn, including:

- Jaundice within the first 24 hours of life
- A sibling who was jaundiced as a neonate and/or required phototherapy
- Hemolysis (Rh incompatibility, ABO incompatibility)
- Oxytocin use during labor
- Gestation 35-38 weeks
- Male sex
- Short hospital stay
- Impaired sucking/nursing
- G6PD deficiency
- Infection
- Bruising, hematomas
- East Asian or Mediterranean descent
- Breastfeeding

When treating babies at high risk for developing severe hyperbilirubinemia, TSB levels that may not cause alarm in low-risk babies should be considered pathologic and treated aggressively.[28]

Signs that jaundice may be pathologic in origin include appearance of jaundice within 24 hours after birth, a TSB concentration that rises by 5mg/dL or more per day, and/or a TSB level higher than 17 mg/dL in a full-term newborn. Other non-reassuring signs include (1) prolonged jaundice that is unresponsive or poorly responsive to therapy; (2) jaundice that is relieved by therapy but rebounds again when therapy is discontinued, to the same or a higher level than that obtained before therapy was instituted; (3) other evidence of underlying disease; or (4) serum conjugated bilirubin levels greater than 2mg/dL or more than 20% of the TSB level (indicating that, although the bilirubin has been

[23] Jackson, C and Gower, L, eds. *Maternal, fetal, and neonatal physiology: a clinical perspective. Chapter 18: Bilirubin Metabolism.* St. Louis: Saunders Elsevier 2007.

[24] Practice Parameter: management of hyperbilirubinemia in the healthy term newborn. Pediatrics. 1994; 94(4 pt 1): 558-562; and Behrman RE, Kliegman RM, Jenson HB, eds. *Nelson Textbook of Pediatrics.* 16th Ed. Philadelphia: Saunders, 2000: 511-28; and Porter, ML 2002, Supra, Note 23, at p. 599.

[25] Maisels MJ and Watchko JF, Eds. 2000. *Monographs in Clinical Pediatrics: Neonatal Jaundice*. Harwood Academic Publishers, The Netherlands; *Chapter 10: The Clinical Approach to the Jaundiced Newborn,* p.139.

[26] Maisels, MJ. 2005, Supra, Note 1.

[27] Maisels MJ and Watchko JF, Eds. 2000. Supra, Note 25, at p.154; and Porter, ML 2002, Supra, Note 23.

[28] This was part of an expert opinion rendered by Jeffrey Maisels in a case handled by our office, and is consistent with Dr. Maisels' book chapter in Maisels MJ and Watchko JF, Eds. 2000, Supra, Note 25.

converted to water-soluble form, the body is still not able to eliminate it).

Serum bilirubin levels: monitoring and interpretation

The serum concentrations mentioned thus far seem to be very high numbers—especially when considered in comparison to the "normal" level of bilirubin in an adult, which is 0.3-1.0 mg/dL. How much bilirubin is too much in an infant?

The TSB level in a newborn is constantly changing in the hours and days after birth. Normal bilirubin values vary according to the infant's age, *in hours*. TSB levels also vary considerably depending on a variety of patient-specific factors including race, breastfeeding, and other epidemiologic and genetic factors, thus the exact parameters of what constitutes a "normal" TSB level are somewhat inexact and amorphous. As such, TSB levels are interpreted in terms of the infant's age in hours, with the use of an hour-specific nomogram, as shown in Figure 1. Indeed, M. Jeffrey Maisels, M.D., a leading expert on neonatal jaundice, has even written that "It makes no sense to attempt to interpret bilirubin levels without considering the infant's precise age in hours."[29]

The nomogram is essentially a graph that shows trending in the patient's TSB levels over time, and quantifies the patient's risk for clinically significant hyperbilirubinemia according to established patterns. As you can see from Figure 1, newborn infants can have TSB levels as high as 12 mg/dL and still be in the low risk zone, depending on their age in hours. That same value of 12 mg/dL could, alternatively, be in the highest risk zone in a younger infant. This variability helps to illustrate the usefulness of the nomogram in interpreting the TSB levels.

An 'abnormal' TSB level, therefore, is most easily understood in relation to the hour-specific nomogram: it is one that is either (a) above the 95th percentile, or (b) crossing from a lower percentile into a higher percentile.[30]

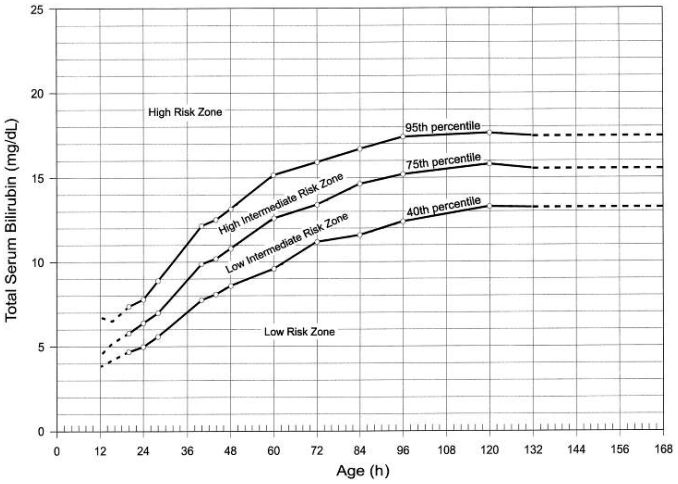

Figure 1: Hour-Specific Nomogram for TSB Interpretation[31]

The 95th percentile on the nomogram represents a rate of rise in TSB level of 0.2 mg/dL/hour. The American Academy of Pediatrics recommends that clinicians serially measure the TSB level and plot the values on the nomogram to determine whether the TSB is crossing percentiles and to quantify the infant's risk for developing hyperbilirubinemia.[32]

Medical intervention

When the level of bilirubin presents a risk to the patient, it can be treated using ultraviolet light waves, which break apart the bilirubin molecule into component parts that can be more easily eliminated from the body. This process is called phototherapy. Usually, phototherapy is sufficient to bring the bilirubin levels back into a safe range.

[29] Maisels MJ and Watchko JF, Eds. 2000, Supra, Note 25, at p.149.

[30] Bhutani VK, Johnson LH, Sivieri EM: Predictive ability of a pre-discharge hour-specific serum bilirubin for subsequent significant hyperbilirubinemia in healthy term and near-term newborns. *Pediatrics* 1999; 103:6.

[31] This widely used nomogram was first published in *Pediatrics*, 1999; 103(1): 6-14 by Drs. Vinod Bhutani, Lois Johnson, and Emedio Sivieri. It was reprinted in "Management of hyperbilirubinemia in the newborn infant 35 or more weeks of gestation", the current guideline for the management endorsed by the American Academy of Pediatrics, in *Pediatrics*, 2004; 114(1): 297-316.

[32] Maisels MJ, Baltz RD, Bhutani VK, et al. Management of hyperbilirubinemia in the newborn infant 35 or more weeks of gestation. *Pediatrics* 2004; 114:297.

Exact parameters can vary slightly, but as a general guideline, phototherapy is indicated when, within the first 48 hours of life, the TSB concentration rises above 15mg/dL; above 18mg/dL in hours 48-72; and above 20mg/dL in hour 72 and beyond.[33] Obviously phototherapy can be initiated earlier, at lower TSB levels if, in the physician's clinical judgment, there is an increased level of concern. Early use of phototherapy constitutes better care, and would not likely provide the basis for any negligence claim.

In cases where the TSB levels are not responsive to phototherapy, or when the levels are so high that phototherapy alone cannot lower them into a safe range quickly enough to ensure protection from brain injury, exchange transfusion can be performed. Exchange transfusion is a procedure in which the baby's blood serum, containing high levels of bilirubin, is filtered out and replaced with 'clean' blood serum. This is accomplished using a machine similar to that used for hemodialysis in renal patients. Because the procedure is invasive and risky, it is used only in cases of dire emergency after phototherapy has failed or is found to be insufficient.

Untreated hyperbilirubinemia

When elevated levels of unconjugated bilirubin persist too long, kernicterus can develop. Kernicterus, as earlier noted, is a specific type of brain damage caused by deposits of unconjugated bilirubin in certain parts of the brain. This yellow staining can be seen on autopsy, as further discussed below, but can also be seen on MRI and through clinical examination when the diagnosis is made in a living patient.

The precise pathological mechanism underlying the neurotoxicity of bilirubin in the central nervous system is quite complex and still not entirely clear. Postmortem studies of the brains of kernicterus-afflicted babies demonstrate that bilirubin deposits its yellow pigment in the brain tissue, leaving a characteristic discoloration as evidence of the injury. Postmortem findings of kernicterus are characterized by yellow staining of an intense, canary yellow to yellow-orange color, which remains brightly colored even after fixation of the brain in formalin, never oxidizing or fading, even in brains that have been immersed in formalin for many years.[34] The areas most often affected are the globus pallidus, subthalamic nucleus, metabolic sector of the hippocampus, and a few others.

The exact mechanism by which bilirubin crosses the blood-brain barrier is not well understood. Some theorize that bilirubin is unable to cross the blood-brain barrier when bound to albumin because the size of the albumin molecule prevents it from passing through blood vessel walls and cell membranes.[35] Conversely, unbound bilirubin that is circulating freely is able to pass through the blood vessel wall and cell membranes, into the brain tissue, causing injury. Immaturity of or damage to the blood-brain barrier can increase its permeability, as can several other factors including acidosis, increased carbon dioxide levels (hypercarbia), irradiation, asphyxia, and others.[36]

When reviewing medical records for a client with a diagnosis of kernicterus, it is occasionally possible to pinpoint the timing of the injury by looking for one key piece of information: an exceptionally high level of unconjugated bilirubin that decreased in a relatively short period of time *in the absence of any medical intervention*.

In a recent case handled by our office, the client's records demonstrated a bilirubin level in the high thirties, which dropped precipitously in a short period of time, without any treatment. Although somewhat counterintuitive, this finding, if present, is an ominous sign. Recall the earlier discussion regarding the hydrophobic properties of

[33] Beers, MH. et al., eds., The Merck Manual, 18th Edition., at 2267, 2278.

[34] *See, e.g.,* Ahdab-Barmada M, Moossy, J. The neuropathology of kernicterus in the premature neonate: Diagnostic problems. *J. Neuropath. Exp. Neurol.* 1984; 43:45-56; and Larroche JC. Kernicterus. In: Handbook of Clinical Neurology. Volume 6. Vinken, PJ and Bruyn GW, eds. North Holland Publishing Company, Amsterdam 1968, pp.491-516; and Blanc WA and Johnson L, Studies on kernicterus. *J. Neuropathol. Exp. Neurol.* 1959; 18:165-189.

[35] Maisels MJ and Watchko JF, Eds. 2000, Supra, Note 3.

[36] *Id.*

unconjugated bilirubin, and how this hinders the body's ability to excrete bilirubin until it is conjugated by the liver. In this context, it follows that an abrupt drop in serum concentrations of bilirubin *in the absence of therapy* may reveal a critical point of no return for your injured client. If it cannot be excreted from the body but is disappearing from the blood that means only one thing: it is being moved from the blood to another location. A precipitous decrease in the serum concentration occurs in the absence of medical therapy *because the bilirubin is moving from the blood into the brain*. If you see this, you have a good idea of when your client's injury occurred.

Recognizing acute bilirubin toxicity and the late sequelae of Kernicterus

Early clinical indications that an infant is suffering from acute bilirubin toxicity can include hypotonia (a floppy baby), a poor sucking reflex and difficulties in feeding. After the first several days, the presentation often changes and the baby becomes very stiff (hypertonia) and may exhibit a type of posturing associated with brain injury called opisthotonus (Figure 2), fever, a high-pitched cry, and, in the most severe cases, seizures, coma, and death.[37]

You need to ask your clients what they recall about their baby's presentation during the time in question, and look for indications in the medical records that any of these findings were reported by the parents or noted by the healthcare providers.

Figure 2: Opisthotonus posturing

If the child survives the acute phase of the illness, later manifestations of kernicterus are described as "Perlstein's Tetrad"[38] and include extrapyramidal abnormalities (such as facial grimacing, drooling, dysarthria, and athetosis), hearing loss due to injury to the cochlear nuclei in the brain stem, limitation of upward gaze, and dental dysplasia. Not every child will exhibit all these conditions, nor are they all required to confirm a diagnosis.

In terms of a definitive diagnosis, some will argue that the only means of confirmation is by examining the brain on autopsy. This is not necessary for litigation to be successful. MRI findings and one or more of the clinical findings discussed above will suffice for your experts to make this diagnosis. You will need to causally link the result back to an elevated bilirubin level, of course, but interestingly, the level of hyperbilirubinemia does not necessarily correlate to the severity of injury. In other words, a baby who suffered kernicterus with a TSB of 38 mg/dL will not necessarily be more severely injured than one who developed kernicterus at a TSB level of 26 mg/dL. There are no studies that demonstrate a specific bilirubin level that will cause kernicterus, or a level below which kernicterus will not occur.[39]

Mistakes in Medical Care—Liability and Causation

Errors and omissions giving rise to liability in a claim for kernicterus can include:

Making treatment decisions with the assumption that there is a single 'normal' bilirubin level. Remember the nomogram. The 'normal' values change according to the baby's age *in hours.* A value that is within normal limits for a 72-hour old baby can be dangerously elevated in a younger infant.

Failing to screen and monitor babies to ensure that serum concentrations of bilirubin do not reach dangerous levels. Prevention is the best medicine here. Remember, kernicterus is always preventable. It is easy to obtain a TcB level or TSB level, and these can be compared over time to

[37] Bhutani VK 2005, Supra, Note 6, at p.15.

[38] Perlstein, MA. The Late Clinical Syndrome of Posticteric Encephalopathy, *Ped Clin N Am* (1960) 7:665-687.

[39] Bhutani VK 2005, Supra, Note 6, at p.16.

demonstrate a trend. There is no excuse for not tracking bilirubin levels in a jaundiced baby.

Failing to examine a baby for signs of acute bilirubin encephalopathy. This is especially true when the parents report problems that might be consistent with hyperbilirubinemia, such as difficulty feeding, a stiff baby, a floppy baby, high-pitched cry, etc.

Disregarding a lab result in the erroneous belief that the level is so high that it must be a laboratory error. Our defendant told his patient's parents that the TSB concentration reported by the lab of 37.2mg/dL was a lab error and that they should not be concerned. Several days passed before any intervention was attempted, and by then it was already too late. Treatment should never be delayed in the face of dangerously high bilirubin levels.

Turning off the phototherapy lights to perform for diagnostic testing or procedures. If medical treatments are needed while a baby is undergoing phototherapy, they should be done under the lights. If that is not possible, i.e., the baby needs an MRI or X-ray, the lights should be left on as long as possible and treatment resumed at the very first opportunity as soon as the test or procedure is concluded. If there is some reason for the baby to leave the unit, the lights should be left on and taken with the child. This therapy should not be interrupted unnecessarily.

Operating under the erroneous belief that since unconjugated bilirubin is the form that causes kernicterus, it is the only value that needs to be monitored. This is not the standard of care. Treatment decisions need to be made based on the TSB level, not the unconjugated bilirubin level alone.

These mistakes are the most common, and can be used to establish liability. Causation is an easier burden because kernicterus is a signature injury—unlike CP in general, which has numerous causes and sometimes occurs where no cause can be identified, only unconjugated hyperbilirubinemia causes kernicterus. If you can establish a breach of the standard of care, the causation opinions will follow.

Damages

Children who survive the acute phase of bilirubin toxicity and go on to live with kernicterus and the resulting cerebral palsy face a life of difficulty and catastrophic medical needs. Kernicterus by itself, once the acute phase is over, does not necessarily limit the child's life expectancy, however, and it is not uncommon for these children to live out a normal lifespan. The severity of their CP will dictate the costs associated with caring for them, for the duration of their life expectancy. These children require home care, multiple modalities of therapy, assistive technology, mobility adjuncts such as specialized wheelchairs, accommodations to their living space, work space, and vehicles, surgeries to manage muscle and tendon contractures, medications to manage spasticity, and a myriad of other interventions that increase the cost of the life care plan.

One critical fact of which you should be aware when handling one of these cases, not only from a case management perspective but also as you begin to think about damages, is that the cerebral cortex is largely spared with this injury. As such, intelligence, even in the most severely injured kernicterus patient, is normal or very close to normal. The heartbreaking result is a child who is catastrophically injured, but simultaneously *aware* of their predicament.[40] The questions these children ask and the comments they make, whether through a dysarthric speech impediment or by the use of assistive computer technology that speaks for them, clearly indicate that these children, while severely injured and brain damaged, are *smart*. Their athetoid cerebral palsy serves to trap them in a body that does not work, but does so without hindering their ability to think, feel, and understand the world around them. Be sensitive to this when talking to the children and their parents, and remember as you talk about the injured child that he or she can probably understand everything you say.

[40] See, e.g., Bhutani VK 2005, Supra, Note 6.

Resources for further reading

Materials on kernicterus are abundant. The sources cited throughout this article are a good start and will lead you to multiple other resources as well.

A valuable online resource can be found at http://www.kernicterus.org/, a website maintained by the Virginia Commonwealth University and Dr. Steven Shapiro. Dr. Shapiro's work in this area is highly regarded and he is an excellent resource.

For general information on this injury, look at the website for Parents of Infants and Children with Kernicterus, located at: http://www.pickonline.org/, and the Centers for Disease Control, at: http://www.cdc.gov/ncbddd/dd/kernichome.htm.

Litigators evaluating a kernicterus claim are invited to contact the author for more information or assistance in screening and pursuing this type of claim.

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Jennifer Keel is admitted to practice in Colorado, Maryland, Minnesota, District of Columbia, and California. Her practice focuses exclusively on medical malpractice litigation, to which she is uniquely well suited given her extensive medical background. In addition to her undergraduate education, which was heavily weighted towards pre-medical studies, Jennifer also completed the rigorous didactic and clinical education program for New York State's highest level of prehospital care providers, and became a paramedic in 1995. She worked as a full-time Paramedic for six years in the City of Rochester, New York, during which time she treated thousands of patients in need of emergent medical and surgical intervention in the prehospital setting. She also spent two years as a Tissue Procurement Specialist and Procurement Team Leader for the Rochester Eye and Human Parts Bank, where she performed sterile surgical tissue procurement procedures to recover transplantable joints and tissues for recipients across the country. Jennifer has also worked in the operating room as an anesthesia technician and scrub-assist technician, where she had the opportunity to scrub in and observe a wide variety of surgeries firsthand.