Prevention of kernicterus

New guidelines and the critical role of family physicians

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In the 1940s and the 1950s, severe neonatal hyperbilirubinemia and kernicterus were most often encountered with hemolytic disease of newborn (HDN), which occurs most often as a result of the incompatibilities of the Rh and ABO blood groups. With the advent of prenatal testing, maternal Rh°(D) immunoglobulin, phototherapy, and exchange transfusion, the incidence of severe hyperbilirubinemia drastically decreased to the point that most physicians practising today have never encountered a bilirubin-induced neurologic disorder.

Children affected with complications of hyperbilirubinemia can present with choreoathetoid cerebral palsy, dystonia, sensorineural hearing loss, paralysis of upward gaze, and dental enamel dysplasia. Unfortunately, severe hyperbilirubinemia continues to be the most common cause of neonatal readmission to hospital in North America, and kernicterus continues to occur in infants without risk factors or evidence of HDN.

In Canada, a recent 2-year Canadian Paediatric Surveillance Program study of severe hyperbilirubinemia reported 258 cases of term infants, 60 days of age or younger, with either exchange transfusion or an unconjugated bilirubin level of 425 µmol/L or greater. Most of these infants (72%) were readmitted to hospital at a median age of 5 days. More important, 81% of infants were exclusively breastfed, and 11% of confirmed cases had a documented 10% to 15% weight loss. Of those with available data, only 36% had a cause identified; the most common cause was ABO blood group incompatibility and glucose-6–phosphate dehydrogenase (G6PD) deficiency.[1] Regardless of the rarity of kernicterus even with bilirubin levels of 425 µmol/L, readmission creates potentially unnecessary distress and disruption for these families and can be prevented.

Prevention measures

Given that most of the infants in the Canadian Paediatric Surveillance Program study lacked traditional risk factors, these guidelines highlight more aggressive prevention measures:

Documentation of the mother's blood group. For mothers whose blood group is O or has not been identified, perform targeted cord blood screening for ABO and Coombs testing, as infants born to mothers with O blood group are at increased risk of developing HDN due to ABO incompatibility.

Jaundice risk evaluation. Assess all infants and determine their risk for severe jaundice based on history, including family history, physical examinations, and laboratory investigations.

Transcutaneous bilirubin (TcB) or serum bilirubin measurement before discharge. Documentation in all newborns of either a TcB or serum bilirubin measurement before discharge, between 24 and 72 hours of life,[2,3] is a favoured approach because visual inspection is inaccurate for determining both the presence and severity of jaundice, particularly in darkly pigmented infants.[4]

The Canadian Paediatric Society has recently published new guidelines for the detection, management, and prevention of hyperbilirubinemia in term and late preterm newborn infants.[2] Risk factors for severe hyperbilirubinemia include the following:

- hemolysis due to ABO or other incompatibility,
- cephalhematoma or substantial bruising,
- excessive weight loss (≥ 10%), particularly in breastfeeding infants,
- East Asian ancestry,
- gestational age of 35 to 36 weeks,
- history of a sibling receiving phototherapy, and
- jaundice observed in the first 24 hours.

Any infant who requires resuscitation at birth or treatment for sepsis is also at increased risk.[2]

Noninvasive TcB measuring devices, such as BiliCheck and ColorMate III, have been shown to provide valid estimates of bilirubin serum levels in term newborns and can be used as screening tools.[5]

Transcription of the bilirubin result. Transcribing the bilirubin measurement result on the predictive nomogram based on age in hours and gestational age allows for categorization of infants as low, medium, or high risk.[6] The nomogram should be given to parents when the newborn is discharged. Clinicians can also make use of an on-line tool (**www.bilitool.org**) to determine infants' risk of developing severe neonatal jaundice, or the need for phototherapy.

Confirmation of serum bilirubin level. Serum bilirubin levels should be confirmed for all infants who are clinically jaundiced within the first 24 hours or for those with a high TcB measurement.

Screening for G6PD deficiency. Screen for G6PD deficiency in selected at-risk non-white infants who develop jaundice, such as those of African, Asian, Mediterranean, or Middle Eastern descent, and in infants with severe hyperbilirubinemia.

Community programs. Community programs where breastfeeding is encouraged and supported should be established.

Ongoing assessment. Mother and baby should be continually assessed after discharge, especially when discharge occurs earlier than 48 hours after birth.[7,8]

The recommendation for measuring TcB or serum bilirubin levels for all infants before discharge or within 72 hours will have an effect on the provision of both hospital and community newborn care services, as many infants are discharged before 72 hours. Family physicians play a pivotal role in the 48-hour post-discharge follow-up—as recommended by both the Canadian Paediatric Society[7] and the American Academy of Pediatrics.[8] These visits have the potential to identify infants with feeding difficulties and weight loss, assess infants with persistent or worsening jaundice, and ensure priority access for repeat TcB or serum bilirubin measurements.

Conclusion

Although there is no reliable strategy to identify all infants who will develop serious hyperbilirubinemia, nor any one bilirubin level that predicts the development of neurologic damage, these new guidelines emphasize heightened awareness of risk factors, a low threshold for measuring serum bilirubin while in hospital, and rigorous attention to close follow-up. Renewed efforts on the part of primary care providers have the potential to reduce both readmission rates and bilirubin-induced neurologic disorders.

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Competing interests

None declared

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