NEWBORN SCREENING
ONTARIO
DÉPISTAGE DES NOUVEAU-NÉS
DE L'ONTARIO

BULLETIN #67

June 10, 2013

Updates from Newborn Screening Ontario (NSO): newborn screening for SCID, prematurity/low birth weight recommendations, and reminder of telemedicine

Please distribute in relevant areas at your institution/practice:

1. **Newborn screening for Severe Combined Immune Deficiency (SCID):**
   Due to unexpected delays related to the revision and approval of provincial regulations surrounding newborn screening required to start SCID screening, the start date has been postponed. Our understanding is that this process is nearing completion and a revised start date will be announced as soon as Cabinet approval has been officially obtained. We anticipate delaying the start by as much as a month in order to allow time for communications designed to ensure that the public and prospective parents are aware of this change; this has not been possible during the regulatory change process.

   Thank you for your patience and understanding. Please do not hesitate to contact NSO if you have any questions.

2. **Recommendations for significantly premature or very low birth weight (LBW) infants who require a packed red blood cell transfusion (PRBC):**
   NSO has received a number of questions for clarification of the recommended process for significantly premature (<33+0 GA) or LBW (<1500g at birth) babies who require a PRBC transfusion. To help clarify the recommendations for all, NSO has created a flow diagram to accompany the recommendations. Please see the recommendations + diagram attached to this bulletin.

3. **Reminder: Telemedicine, Thursday, June 13, 2013 |12:00 PM – 1:00 PM**
   Topic: Newborn Screening Ontario's (NSO) Submitter Survey Results
   Presenter: Lauren Higgins, MSc, CCGC

   This event is scheduled through the Ontario Telemedicine Network (OTN). OTN Telemedicine contacts who have access to Ncompass can register their site by clicking on the link. Otherwise, OTN partners wishing to attend can email their site and system info to OTN Customer Service customerservice@otn.ca

   URL: https://schedule.otn.ca/tsm/portal/nonclinical/details.do?request.requestId=27870623

These bulletins are circulated to keep you abreast of changes to Newborn Screening Ontario. Please share this information with any relevant personnel in your hospital / practice. If there is someone who would like to be added to this list, contact NSO at: 1-877-627-8330, option #0 or at NewbornScreening@cheo.on.ca.

Recommendations for Premature or Low Birth Weight Infants

Premature or low birth weight babies may have a delayed rise in TSH even if they have Congenital Hypothyroidism (CH). As Newborn Screening Ontario (NSO) uses elevation of TSH as the screening marker for CH, there is an increased risk of a false negative result (missed case) if these babies are screened only once in the early neonatal period. As well, premature newborns are known to have a lower number of T-cell receptor excision circles (TRECs) and are more likely to falsely screen positive for Severe Combined Immune Deficiencies (SCID). Therefore, the first follow up investigation for screen positive premature infants is to retest a second newborn screening sample taken at a later age. It is therefore important that screening samples are taken as outlined below.

Premature (<33^0 WGA) or very low birth weight (<1500g) babies should have:

1. A first Newborn Screening specimen collected between 24 and 72 hours of age.

2. A second specimen collected at 3 weeks of age or when the baby is being discharged home from the hospital, whichever comes first.

   2.1. If the baby is discharged home prior to 3 weeks of age from a hospital with a robust tracking system to ensure follow-up, an outpatient appointment between 3 and 4 weeks of age can be arranged without the need for a blood draw prior to discharge.

   2.2. If the baby is discharged home with the second specimen collected before 3 weeks of age consideration should be given to having a third specimen arranged as an outpatient between 3 and 4 weeks of age.

3. If the baby is being transferred to another hospital after 3 weeks of age, the hospital receiving the baby should confirm that the second sample was taken prior to transfer. If it was not taken, the receiving hospital should take the second sample as soon as possible after the baby arrives. The receiving hospital can also call NSO to inquire whether a second sample was received.

The following exceptions to the above policy apply:

1. A specimen should be collected prior to the baby receiving a blood transfusion, even if this is prior to 24 hours of age. This will ensure a satisfactory screen for:
   a. Galactosemia since the test relies on measurement of the Galactose-1-Phosphate Uridyltransferase enzyme activity in red blood cells.
   b. Hemoglobinopathies since the adult hemoglobin from the transfused blood may mask an abnormal hemoglobin pattern.
   c. If the first sample is taken at less than 24 hours of age, a second sample should be taken between 24 hours and 7 days of age.

2. Total Parenteral Nutrition (TPN). Amino acid solutions administered as part of TPN are a common reason for false positive screening results for amino acid diseases in premature / LBW babies. Ideally, a specimen should be collected at a time when the baby is receiving lower amounts of TPN. For example, if the baby is receiving 1.5 g/kg/d of amino acids at 24 hours of age, it is preferable to take the screening sample before this amount is increased. Please note that there is no increased risk of a false negative result if a sample is taken when the baby is receiving higher amounts of amino acid solutions; screening should not be delayed beyond 72 hours of age for this reason.

Premature infants ≥33^0 WGA and >1500g should be screened no differently than term babies.

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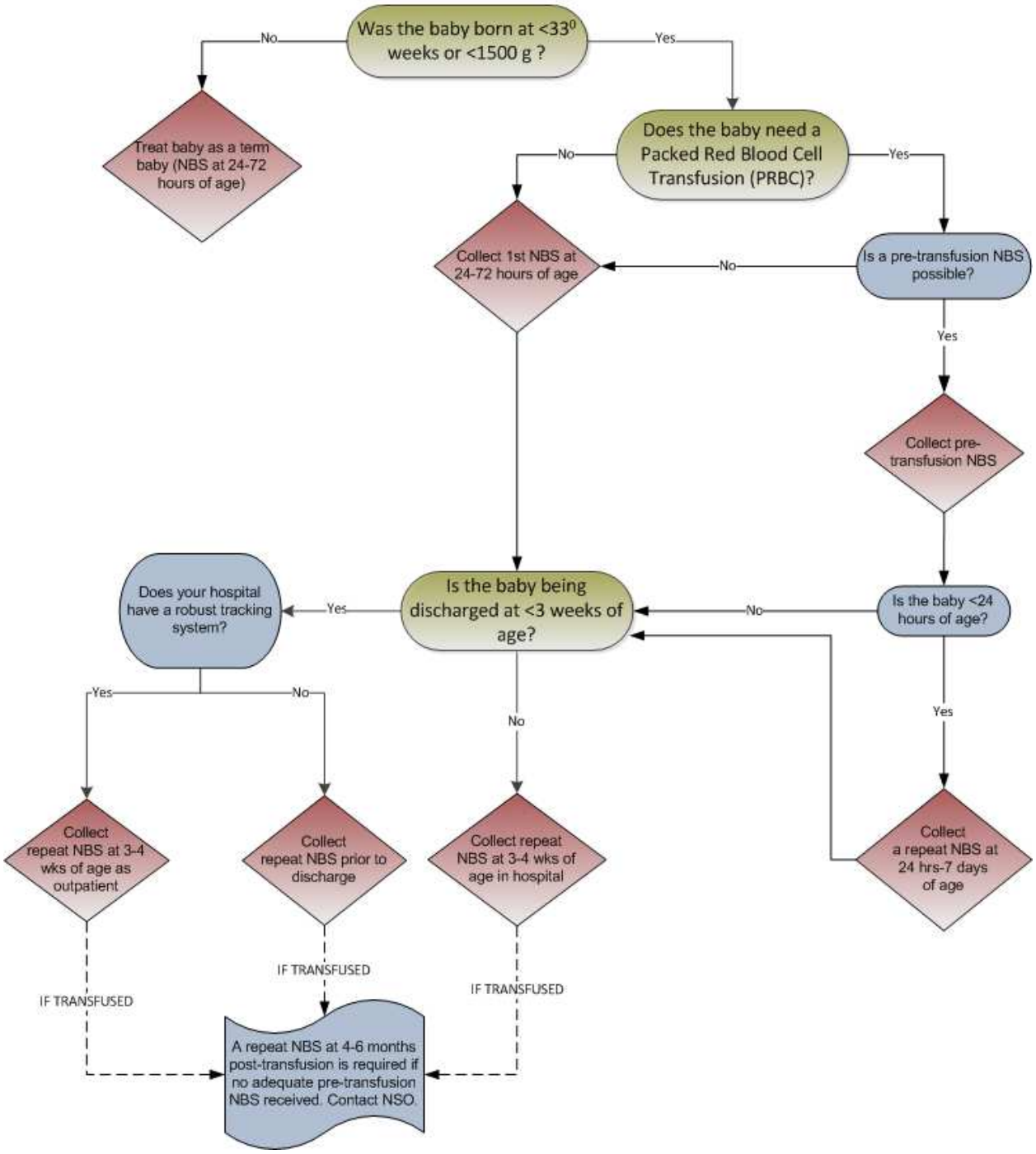
Premature or Very Low Birth Weight Babies +/- PRBC Transfusions



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