



Biliary Atresia Screening in Ontario

Information for Front Line Health Professionals



NEWBORN SCREENING ONTARIO
DÉPISTAGE NÉONATAL ONTARIO



What is Newborn Screening Ontario (NSO)?

- Ontario's provincial newborn screening program
 - Screening babies born anywhere in ON, NU, NL
- >30 diseases on the Ontario screening panel:
 - Dried bloodspot
 - Point-of-care CCHD screening
 - Risk Factor Hearing Screening (cCMV)
 - **At-home screening for biliary atresia – NEW!**
- Laboratory based out of CHEO in Ottawa, ON





Overview for Presentation

What is BA?

Screening for BA in Ontario

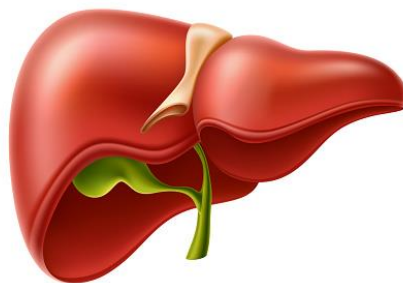
How do I help families screen?

Supportive resources

Questions?



Biliary Atresia (BA)



A pediatric liver disease

Caused by:

obliteration of the bile duct that drains bile from the liver to the intestines

Leading to:

bile accumulation in the liver causing liver damage

Resulting in:

most frequent cause of pediatric liver transplant and liver related death

1 in 16,667 babies in

Ontario

1 in 19,05 babies in
Canada



Symptoms

Babies with BA usually appear healthy at birth

Symptoms appear around two weeks of life

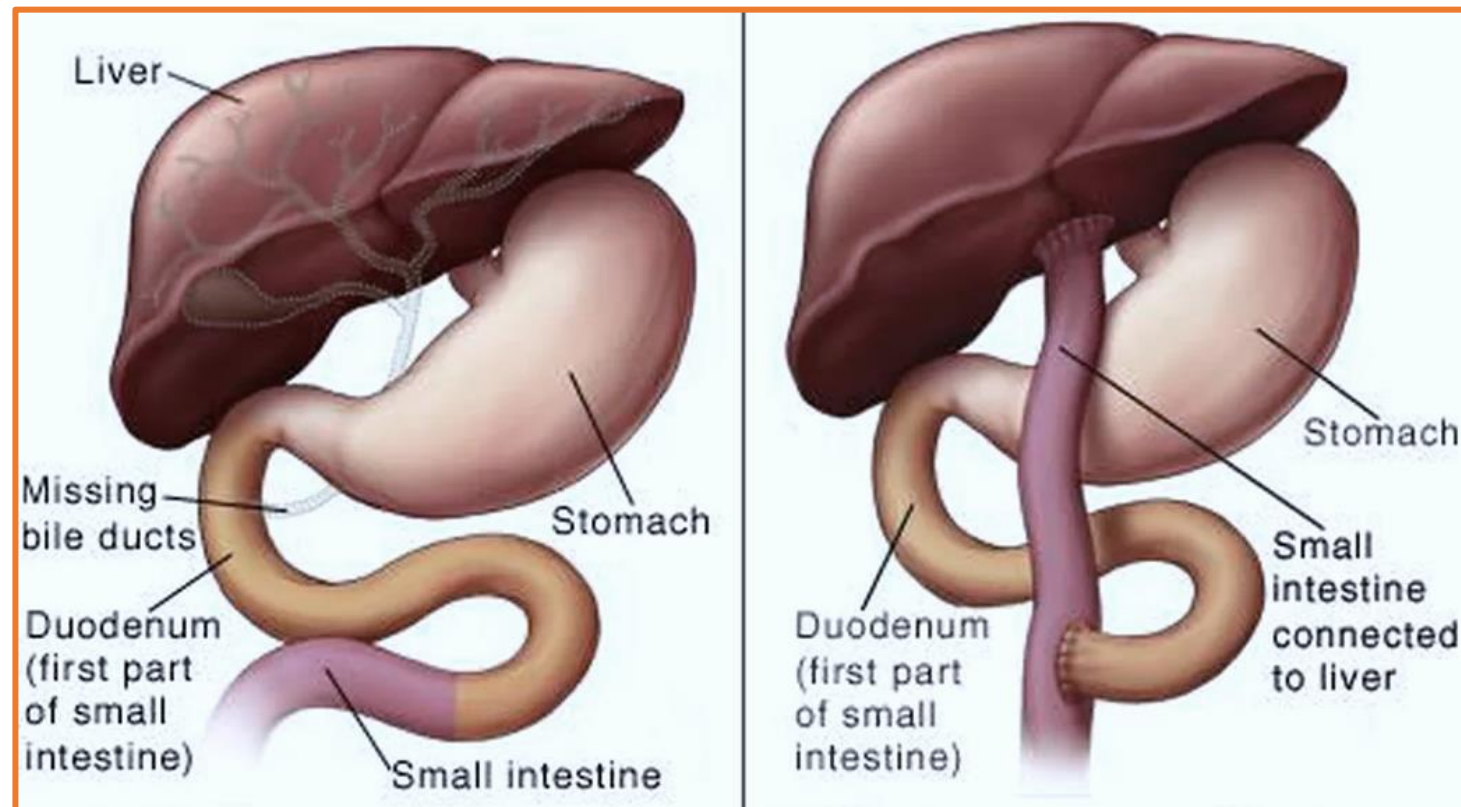
Early: persistent jaundice, dark urine, **pale stool**

Late: distended abdomen, weight loss, GI bleeding



Treatment

- The **Kasai Procedure (KP)** (hepatoportoenterostomy)
- Surgical procedure to establish bile flow from the liver into the intestine





Treatment timing is key!

Without KP intervention, children with BA rarely survive beyond 2 years of age without liver transplantation.

Without surgical repair or a liver transplant, liver failure and mortality is certain.

Performing the KP early is essential. Best outcomes are achieved if it is done <30 days.



Current BA diagnosis and treatment timing

(ICES 2002-2017)

	Median age at diagnosis	Median age of KP	KP >90d	KP in target timeframe
Ontario	62 days	66 days	17%	25% (<45d old)
Canada	55 days	65 days	20%	8% (<30d old)



Current system challenges

Jaundice is common in newborn infants.

There is a lack of awareness among some parents and HCPs that **jaundice > 2 weeks of age, and/or pale stool** can indicate liver disease.

Most well babies do not have a scheduled visit with their HCP between 2 weeks and 2 months of age.



Newborn Screening Ontario is proud to implement screening for **Biliary Atresia**

Welcoming families/caregivers into the screener role using an **Infant Stool Colour Card (ISCC)**

Simple, non-invasive and cost effective

Monitor babies daily for pale stools at home for the first month of life

Leads to an earlier diagnosis and earlier treatment, resulting in **better outcomes** for Ontario babies



Screening for BA using an ISCC works!

Japan since 1994, Taiwan since 2002


In Canada, British Columbia started provincial screening in 2014

Switzerland (2009), France (2014), Germany (2016), and Portugal, Egypt, Brazil have screening programs, education and awareness campaigns


Taiwan: no BA cases with KP performed >90 days of age, and 5 year post-Kasai liver survival rates increased from 37% to 64%

Infant Stool Colour Card (ISCC)

- Visual reference tool
- Low cost, easy to use
- Early detection of abnormally pale stool and connection with specialized help



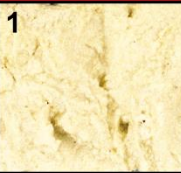

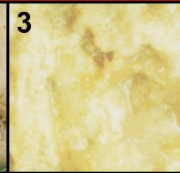



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BILIARY ATRESIA
ATRÉSIE DES VOIES BILIAIRES


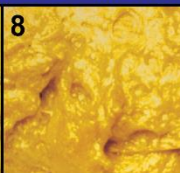
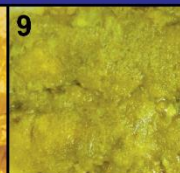
Infant Stool Colour Card Carte colorimétrique des selles du bébé

ABNORMAL Stool Colours
Couleurs des selles ANORMALES

Consult the physical version of this card for greater colour accuracy.
Consultez la carte en papier pour une plus grande précision des couleurs.

NORMAL Stool Colours
Couleurs des selles NORMALES

		
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INSTRUCTIONS

Check your baby's stool (poop) colour every day for the first month after birth to screen for biliary atresia.

If your baby has an abnormal stool colour (see above colours 1,2,3,4,5,6), please contact Newborn Screening Ontario by phone or email or by scanning the QR code below.

INSTRUCTIONS


Vérifiez la couleur des selles (cacas) de votre bébé chaque jour durant le premier mois de vie pour dépister l'atrésie des voies biliaires.


Si les selles de votre bébé ont une couleur anormale (voir couleurs ci-dessus 1,2,3,4,5,6), veuillez contacter Dépistage Néonatal Ontario par téléphone, par courriel, ou en scannant le code QR ci-dessous.

Phone / Téléphone : **1-833-POOP-CHK**
(1-833-766-7245)


Online / En ligne : www.newbornscreening.on.ca/poop

Email / Courriel : NSOBA@cheo.on.ca





NEWBORN SCREENING ONTARIO
DÉPISTAGE NÉONATAL ONTARIO



BILIARY ATRESIA
ATRÉSIE DES VOIES BILIAIRES

When identification and treatment start early, babies with biliary atresia can have a better chance to grow and develop normally.

What is biliary atresia?
Biliary atresia (BA) is a rare but serious disease where bile cannot get from the liver to the stool (poop). Babies with BA usually seem well but they can develop **pale yellow or whitish coloured stools** in the first month of life. Along with the pale poop, these babies can have yellow skin and eyes (**jaundice**) **lasting longer than the first two weeks of life**. Without treatment, babies with BA will develop life-threatening liver damage. Treatment is most effective when babies are identified before they are one month old.

How can you screen for biliary atresia?
You can screen for BA by comparing your baby's stool (poop) colour to the numbered pictures on the front of this card, for the first month after birth. Screening for abnormal stool colour can help to identify problems with the liver earlier. Keep this card close by when you change your baby's diaper.

Who do I contact and when?
If at any time during the screening you notice that your baby has an abnormal stool colour (number 1,2,3,4,5, or 6), please contact Newborn Screening Ontario using the information on the front of this card. Please have available the baby's name, birth date, and health card number. If you also choose to see your doctor, please take this card with you.

What happens next?
First, don't be alarmed. This is a screening tool and not a diagnostic test. A clinical team member from Newborn Screening Ontario will return your call by the next business day to discuss your concern and begin next steps. Your privacy and health information is protected. Learn more: www.newbornscreening.on.ca/privacy

If you have concerns about your baby **other than stool colour**, please contact your health care provider. For more information about biliary atresia, visit our website or scan the QR code on the front of the card.

Information is available in other languages. / Ces informations sont aussi disponibles dans d'autres langues.
www.newbornscreening.on.ca/poopcard

Le dépistage précoce et le traitement sans délai chez les bébés atteints de l'atrésie des voies biliaires peuvent les aider à grandir et se développer normalement.

Qu'est-ce que l'atrésie des voies biliaires?
L'atrésie des voies biliaires (AVB) est une maladie rare et grave dans laquelle la bile ne passe pas du foie aux selles (cacas). Les bébés atteints de l'AVB semblent être en bonne santé, mais leur selles peuvent être de couleur **jaune pâle ou blanchâtre** au cours du premier mois de leur vie. En plus d'avoir des selles pâles, les bébés peuvent avoir une coloration jaune de la peau et des yeux (**jaunisse**) **qui dure plus longtemps que les deux premiers semaines de vie**. Sans traitement, les bébés atteints de l'AVB développeront de sérieux dommages au foie, potentiellement mortels. Le traitement est plus efficace quand les bébés sont identifiés avant l'âge d'un mois.

Comment puis-je dépister l'atrésie des voies biliaires?
Pour dépister l'AVB, il suffit de comparer la couleur des selles (cacas) de votre bébé aux images numérotées au recto de cette carte pendant le premier mois suivant la naissance. Le dépistage d'une couleur anormale des selles peut aider à identifier plus tôt les problèmes liés au foie. Gardez cette carte près de l'endroit où vous changez la couche de votre bébé.

Qui devrais-je contacter, et quand?
Si, à tout moment pendant le dépistage, vous remarquez que les selles de votre bébé ont une couleur anormale (numéro 1, 2, 3, 4, 5 ou 6), veuillez communiquer avec le Dépistage Néonatal Ontario en utilisant les renseignements au recto de la carte.

Veuillez avoir à portée de main le nom du bébé, sa date de naissance et son numéro de carte santé. Si vous souhaitez également de consulter votre médecin, veuillez apporter cette carte avec vous.

Que se passe-t-il ensuite?
D'abord, ne vous inquiétez pas. Il s'agit d'un outil de dépistage et non d'un test diagnostique. Un membre de l'équipe clinique de Dépistage Néonatal Ontario vous rappellera d'ici le prochain jour ouvrable pour discuter de vos préoccupations et amorcer les prochaines étapes. Votre vie privée est protégée et vos renseignements médicaux sont confidentiels. Pour en savoir plus : www.newbornscreening.on.ca/fr/vie-privee

Si vous avez des préoccupations concernant votre bébé **autres que la couleur des selles**, veuillez communiquer avec votre fournisseur de soins de santé. Pour en savoir plus sur l'atrésie des voies biliaires, consultez notre site web ou scannez le code QR au recto de la carte.

We thank / Nous remercions
Health Promotion Administration, Ministry of Health and Welfare, Taiwan, and Professor Mei-Hwei Chang, National Taiwan University, College of Medicine
for authorizing the use of stool colour photos. / pour avoir autorisé l'utilisation des photos de couleur des selles.
For permission to use any content on this card, please contact NSO. Pour obtenir l'autorisation d'utiliser le contenu de cette carte, veuillez contacter NSO.

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How to Screen

Compare baby's stool colour for the first month of life to numbered images on the ISCC.

If an abnormal coloured stool is noticed, information on the card directs families to contact Newborn Screening Ontario.

IS THAT

POOP???



Contact NSO



Phone: 1-833-POOP-CHK



Email: NSOBA@cheo.on.ca



Website: www.newbornscreening.on.ca/poop or QR Code

- NSO website portal for entering contact info and stool photos
- Leverage CHEO's VOYCE translation service
- NSO will contact the family within 1 business day and perform a telephone assessment, request stool photos, arrange next steps



Screen Positive = Acholic Stool

IDENTIFICATION – **parental report** to NSO, plus **telephone triage assessment** by NSO health care provider incorporating information from submitted stool photos

CONFIRMATION – **referral by NSO to Gastroenterologist/Hepatologist** at the nearest Pediatric Academic Health Sciences Centre to arrange for outpatient **urgent bilirubin** (direct/conjugated)



Diagnostic Workup

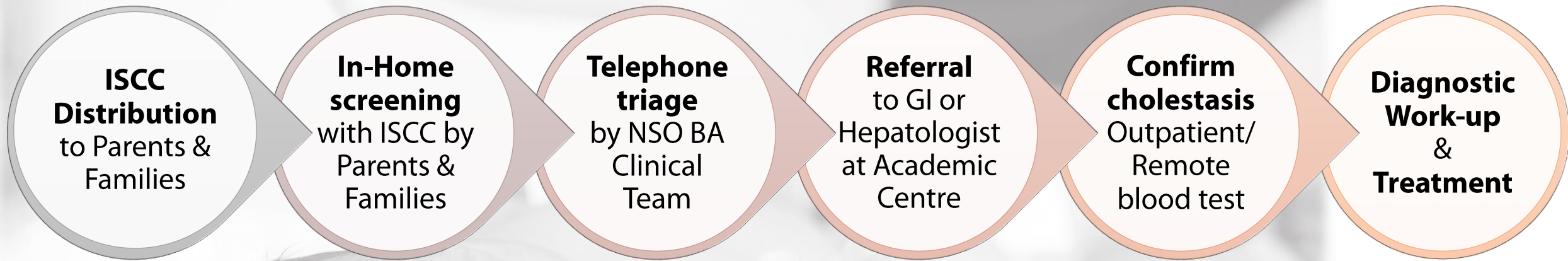
CHOLESTASIS CONFIRMATION – Elevated **direct/conjugated bilirubin** levels indicate the **presence of cholestasis** (blockage of bile flow) but not the cause.

DIAGNOSIS – **Further diagnostic tests** (other blood tests, liver imaging, etc.) determine the **cause of cholestasis**. BA is the leading cause of cholestasis in newborns; there are many others.

- Other treatable causes: sepsis, some metabolic disorders, pituitary problems, gallstones, and bile acid synthesis problems



BA Screening Workflow Summary





Newborns Admitted to Hospital

Screening for biliary atresia applies to **all newborns**... even those who don't spend their first month at home!

- ✓ Keep ISCC at bedside of all admitted babies in their first month of life.
- ✓ Notify NSO if an admitted baby has pale/acholic stools.

Premature babies are at **higher** risk for BA and can also present with symptoms later.

- Babies born **less than 37 weeks gestation**, screen the stool (or instruct discharged families to screen) for four weeks/one month past the baby's due date.
- Ensure families have an ISCC to continue screening at home if discharged during this screening window.



If families present to their PCP...

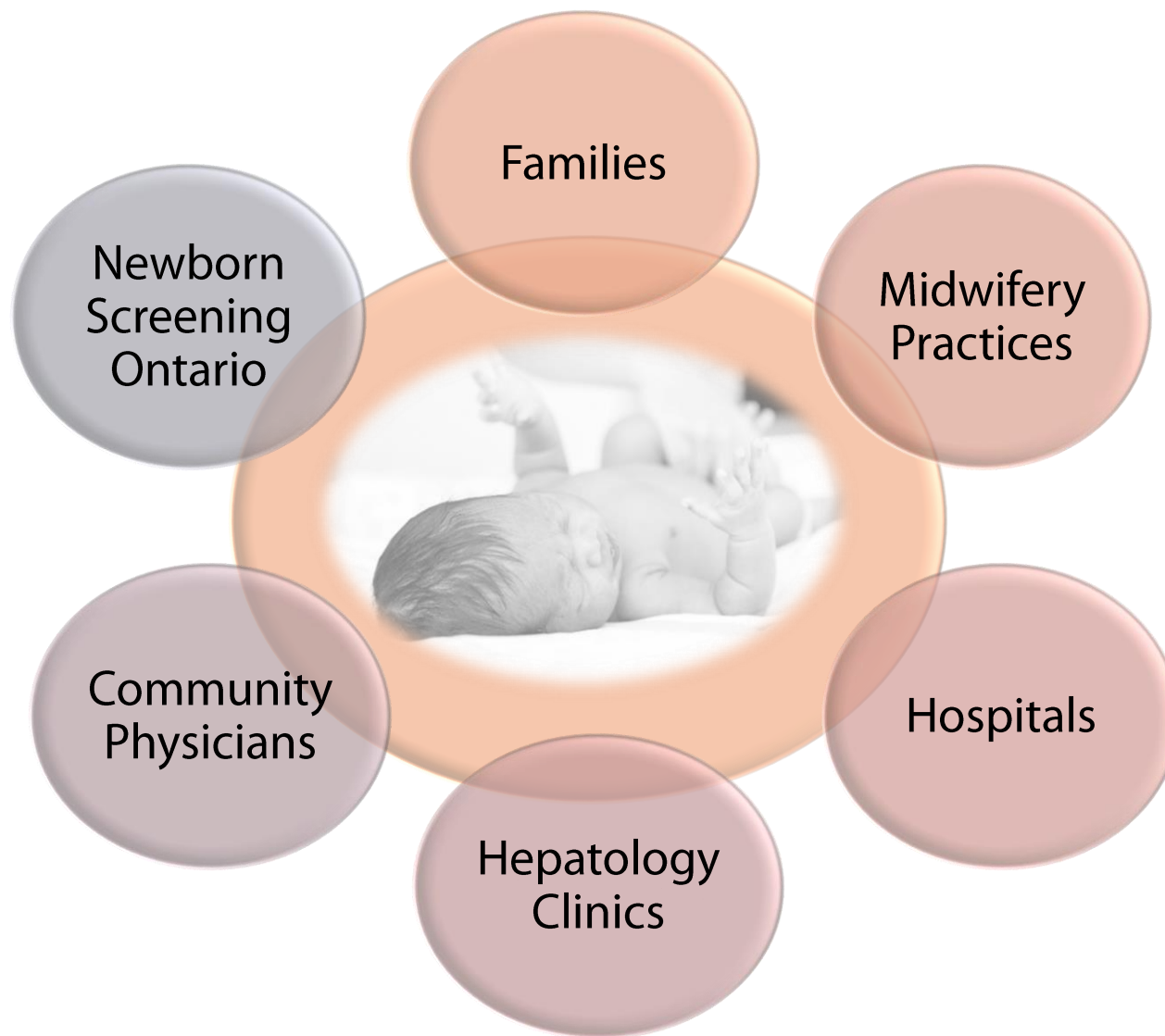
Families might choose to go directly to their primary care provider (or the ED, community GI clinic...)

- We ask them to bring the card with them to the appointment
- We ask the primary care provider to notify NSO if they also believe the stools are pale

Notifying NSO will expedite referral to GI for bilirubin measurement, interpretation, and will help NSO track screen positive cases and monitor the effectiveness of the program.

A large, light orange starburst graphic with several radiating lines, positioned behind the main title.

Collaboration means Success



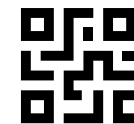


How do I help families screen?

Ensure families have an **ISCC** to use at home or when discharged, and explain how to screen, as well as the importance of screening.

Refer to the *Front-Line Script for BA Screening*.

Instruct families that if an abnormal stool colour is noticed during the screening period, **contact Newborn Screening Ontario** using one of the methods on the ISCC.





Supporting Resources

All materials for families available in French and English.

- Translations of ISCC in many languages will be available to print on our website

Biliary Atresia Screening Portal:

- More information on BA
- How to screen videos + tutorials
- FAQs for families and HCPs
- Materials for HCPs
- Materials for parents
- Contact form for parents for submission of stool colour concerns + images



How to order ISCCs

An initial starter supply of Infant Stool Colour Cards are provided directly to you from NSO in your launch kit. (approx. 3-6 month supply)

Like the dried bloodspot cards:

- Free of charge
- 25 ISCCs per package
- Same distributor (VWR International)
- Same size: 5.5" x 11"

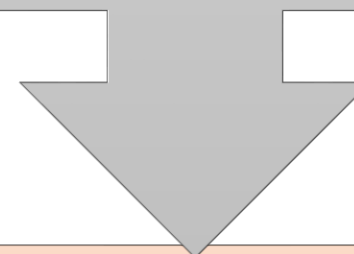
Important:

- ISCCs are a different product and must be added separately on the bloodspot card order
- ISCCs can be ordered alone if you don't require the dried blood spot cards



KEY MESSAGE

Screen baby's stool for 1
month after birth using the
ISCC



Call NSO with any concerns
regarding stool colour



For more information on
Newborn Screening Ontario and biliary atresia, please visit

www.newbornscreening.on.ca

Email: nsoba@cheo.on.ca

“

When identification and treatment start early, babies with biliary atresia can have a better chance to grow and develop normally.

”





*Thank you for your front-line
contribution to
newborn screening in
Ontario*

QUESTIONS...??





Questions

Who is responsible for organizing the referral to the GI clinic after a parent contacts NSO and a referral is deemed necessary after the telephone assessment?

- NSO is responsible for organizing this referral.

Will NSO inform the infant's primary care provider (if applicable) to keep them up to date if a family contacts NSO with concerns about stool colour?

- Yes, we will send a notification letter to the child's primary care provider (specified during the clinical assessment) about any contact that NSO has with a family about their infant's stool colour.



Questions

Can we order infant stool colour cards in a specific language? My clinic targets a population that is not French or English-speaking.

- No. We have translated the infant stool colour card in 30 languages. These translations will be available to download and print on our website. We recommend printing translated copies in advance and attaching them to the card for non French or English-speaking families. It is very important to rely only on the colours of the original card and not those of the translated copy (on paper or viewed on a display) since these colours may appear different.

Do we need to order our initial shipment of cards?

- No. NSO will take care of sending a biliary atresia screening launch kit to your organization which will include the right number of cards corresponding to a 3-6 month birth volume. Please let us know if you would like this shipment addressed to someone specific at your organization.

Are the launch kits specific to each organization? My hospital would hand out 25 infant stool colour cards in approximately 2-3 days.

- Yes! We will send you the right number of cards to get you started for the first 3-6 months. We will also include instructions for you to reorder more cards as you use up your initial supply.



Questions

Will the infant stool colour cards be replaced with a mobile app in the future to allow families to log in and upload pictures?

- This is something we are working on for a future improvement to the biliary atresia screening program! We will be happy to share more details in the future!

Do we order the infant stool colour cards on the NSO website after we receive the initial kit?

- No. The infant stool colour cards will need to be order via VWR International. This is the same distributor that is used for newborn screening dried bloodspot collection cards. Exact instructions for how to order more cards will be included in your launch kit and made available on our website. You will be able to order dried bloodspot collection cards and infant stool colour cards at the same time!

Should we start using/distributing the infant stool colour cards as soon as we receive them, or should we wait until a specified date?

- Yes, by all means! By the time the first organization receives their launch kit in Ontario, NSO's systems and processes will be ready. We encourage you to start distributing infant stool colour cards to families as soon as possible, though we understand it may take some time to implement at your organization.



Questions

What is the process if families do not have a newborn care provider or family doctor?

- We built the referral process with this in mind! NSO makes referrals directly to one of five paediatric academic health sciences centre gastrointestinal clinics. The clinic will order the required bloodwork remotely and begin workup without needing to involve a care provider. If a care provider is available for the newborn, they will be notified of the referral.

Will there be a time where the infant stool colour card will be attached to the newborn screening package similar to the CCHD screening result card?

- At this time, no. This is something that we may consider for the future, however.

Can we use this presentation in our education for staff?

- Absolutely! Please share with staff and any newborn-related areas at your organization.

For infants that are born at less than 37 weeks gestational age, are instructions for screening longer than 1 month shown on the infant stool colour card?

- No. With very limited space on the card, these specific instructions are not included. By providing you with a specific education script for admitted babies/premature babies, we are hoping that you can educate these families and specify exactly how long their screening window will be.



References

- <https://www.cincinnatichildrens.org/health/b/biliary>
- <https://www.liver.ca/patients-caregivers/liver-diseases/biliary-atresia/>
- <https://medizzy.com/feed/23396658> (KP image)
- Fawaz R, Baumann U, Ekong U, Fischler B, Hadzic N, Mack CL, et al. Guideline for the Evaluation of Cholestatic Jaundice in Infants: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition. *J Pediatr Gastroenterol Nutr* 2017 Jan;64(1):154-168.
- Chen SM, Chang MH, Du JC, Lin CC, Chen AC, Lee HC, et al. Screening for biliary atresia by infant stool color card in Taiwan. *Pediatrics* 2006 Apr;117(4):1147-1154.
- Newborn Screening Ontario, (Oct 2020) *Proposal for Biliary Atresia (BA) Screening in Ontario*