Screening for Biliary Atresia Using Infant Stool Colour Cards: Improving The Health of British Columbian Newborns

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OBJECTIVES

• Biliary Atresia 101

• The Canadian experience

• CIHR sponsored BA screening program: results from Phase 1 at Women’s Hospital

• Provincial Program Implementation by Perinatal Services BC
Biliary atresia (BA) is a rare condition (1:19,000 births) but it is the most important chronic pediatric liver disease.

- Most common cause of cirrhosis and liver disease related death in children.
- Leading indication for liver transplantation in the pediatric population accounting for more than 60% of all pediatric liver transplants.
In all patients there is progressive complete obliteration of the bile duct (The pipe draining fluid from the liver to the intestine) that begins in the perinatal period.

The condition manifests in the first 2 weeks of life with jaundice in association with pale chalk white or acholic stools.
BILIARY ATRESIA: Treatment is surgical

KASAI OPERATION = KP

IF SUCCESSFUL
RE-ESTABLISHES BILE FLOW,
JAUNDICE CLEARS
STOOLS BECOME COLOURED

• PROLONGS PATIENT SURVIVAL WITH THEIR OWN NATIVE LIVER WITHOUT NEEDING A LIVER TRANSPLANT

• IF KP FAILS
LIVER TRANSPLANT IS NEEDED OR INFANT DIES BY 2 YEARS
The most important prognostic factor influencing post KP survival with your own liver

**THE BABY AGE AT SURGERY:**

- **BEST OUTCOME IF KP DONE <30 Days old (50% 10 yr with own liver)**
- **WORST OUTCOME IF KP DONE >90 Days old (13% 10 yr with own liver)**

*In CANADA*

- 8% of kids have the KP at < 30 days old
- 20% of Kasai done at >90 days old
AGE OF KASAI AND OUTCOME

CASE 1
BA DELAYED DIAGNOSIS:
WHAT’S THE PROBLEM*

- **BA is a rare condition:**
  - In Canada 1/19,000 births
  - In BC 1/10,000 births (3-5 cases/yr)
  - Few health care providers will see any BA cases in their entire career

- **Finding the yellow needle in the haystack**
  - Neonatal Jaundice is the most common clinical problem among newborns (66% of newborns)
  - but rarely is it due to BA (1:10,000).
  - Even the vast majority with prolonged jaundice beyond 2 weeks are healthy with “breast milk jaundice”

BILIARY ATRESIA: DELAYED DIAGNOSIS

- **STOOL COLOUR**

There is a lack of awareness in Canada (and elsewhere) among parents and health care providers for the need to look at stool colour as a disease indicator and persistent jaundice is often not considered “abnormal” in these infants.

- **The Routine Infant Health Care Visit schedule:**

In Canada by 2 weeks and then at 8 weeks of age... missing the ‘window of opportunity’ for early diagnosis and treatment intervention (< 45 days)
BILIARY ATRESIA:
THE PROBLEM OF DELAYED DIAGNOSIS

- Lack of clinical awareness and assessment
- Late referral and delayed management
- Negative overall disease prognosis:
  lower rate of native liver survival
  greater need for liver transplant
- Inefficient use of health care dollars
BILIARY ATRESIA:
Newborn screening using an infant stool colour card

• First developed in the 1990s in Japan

• Taiwan introduced a national program in 2004

• No case of KP operation after 90 days of age

• 5 year post KP native liver survival increased from 37 to 64%
Why Screen for Biliary atresia in Canada?

- Improved outcome
- Better native liver survival
- Less demand for transplant
- Lower health care costs

SHIFT Distribution to earlier detection

Timely referral and diagnosis

Optimal age at Kasai

20% KP at >90 days

8% KP at ≤ 30 days old

IN CANADA:

Current Distribution

8% 72% 20%

Age after birth (weeks)
Home Based Screening for Biliary Atresia Using Infant Stool Colour Cards: A Large Scale Feasibility Study at Women’s Hospital

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PHASE 1 STUDY AT BC WOMEN’S MATERNITY UNIT

STUDY AIM

TO EVALUATE THE OPTIMAL WAY OF INTRODUCING AN INFANT STOOL COLOUR CARD SCREENING PROGRAM FOR BILIARY ATRESIA IN CANADA

TO ASSESS COST EFFECTIVENESS OF THIS PROGRAM IN CANADA
METHODS

INFANT STOOL COLOUR CARDS are issued to families at discharge from maternity

WARD NURSES INSTRUCT PARENTS TO:
Monitor their infant’s stool colour daily for the first month of baby’s life.

Complete, tear off and return the card by mail when their infant is one month old, or earlier should an abnormal stool colour be recognized.
A SERIES OF INCREMENTAL INTERVENTION STRATEGIES TO INCREASE USE AND RETURN OF CARDS

| Step 1 | Simple Distribution at maternity |

- Phone Surveys to assess card utilization
- Conducted Cost Effectiveness Analysis
RESULTS
Study at BC Women’s Hospital
Vancouver, BC Dec 1 2010- Dec 31 2011.

• 6,843 births
• 6,187 families enrolled into the study
• A 90% enrolment rate!!
• > 90% utilization of the infant stool colour card!!
• Highly cost effective
Universal screening using a passive card distribution to families at maternity is best - the most parsimonious cost effective strategy.

The screening strategy is unique:

- A family centered care model
- No blood tests
- No samples to collect
Biliary Atresia
HOME SCREENING PROGRAM
Rationale

- The current BC Newborn screening program identifies several rare neonatal diseases, including those having a similar incidence to Biliary Atresia (ie Phenylketonuria; 1:15,000).

- No convenient and accurate laboratory test for early Biliary Atresia detection that can be incorporated into routine neonatal screening regimens.

- An infant stool colour card has proven efficacy to instigate earlier referral and more timely intervention for infant affected by biliary atresia with improvement in outcome and health care cost saving.
BILIARY ATRESIA HOME SCREENING PROGRAM: IMPLEMENTATION

- Launched summer 2013

- Rolled out by health authority: VCH; FHA; Island…

- All maternity ward nurses given in-servicing and provided with scripted information about the screening process that they relay to the families at discharge

- Letters mailed to health care providers and midwives

- Public health nurses to remind families about the program

- Website with parent information about biliary atresia and the screening program
• Check your baby’s stool colour every day for the first month after birth

• If you notice that your baby has an abnormal stool colour of pale yellow, chalk white, or clay coloured stool (#1, #2, or #3 on the card), promptly contact PSBC

  Phone: 1-877-583-7842 (1-877-5-test-4-BA)
  Email: psbc@phsa.ca
  Fax: 604-875-3244

SIGN UP FOR WEEKLY TEXT OR EMAIL REMINDERS
FOREIGN LANGUAGES ONLINE
Maternity Ward Nurse Script

BC INFANT STOOL COLOUR CARD®
SCREENING PROGRAM FOR BILIARY ATRESIA

NURSE SCRIPT FOR PARENTS PRIOR TO DISCHARGE:

1. Show parents the Biliary Atresia INFANT STOOL COLOUR CARD (turn page over to show example of the card)
2. The INFANT STOOL COLOUR CARD, located in your discharge package, is for you to screen for Biliary Atresia at home. This is an official provincial screening program for newborns.
3. Biliary Atresia is rare but serious liver disease that begins to affect newborns in the first month of life.
4. It is important to check the colour of the baby’s stool (poop) EVERY DAY for the first month after birth.
5. These are the normal infant stool colours: Numbers 4, 5, or 6 (turn page over, show colours to parents: mustard seed yellow, orange, green)
6. These are the abnormal infant stool colours: Number 1, 2, or 3 (turn page over, show colours to parents: pale, chalk white, clay coloured)
   If at any time you notice the baby has an abnormal coloured stool, you should promptly contact Perinatal Services BC by:
   - Phone Toll Free Number: 1-877-583-7842 (1-877-5-TEST-4-BA)
   - Email: psbc@phsa.ca
   - Fax: 604-875-3244
7. If parents have a smart phone, they can scan the QR code to receive weekly reminders to check their baby’s stool colour for 4 weeks.

For more information: www.perinatalservicesbc.ca

AFTER YOU DISCUSSED THE INFANT STOOL COLOUR CARD WITH FAMILY, PLEASE INITIAL THE BC NEWBORN CLINICAL PATH FORM.
Click on screening programs tab; select biliary atresia

Infant Stool Colour Card available in multiple languages

Can re-order ward supply of cards on line
Improving the outcomes of BC Children
YOU CAN MAKE A DIFFERENCE!!