Pulse Oximetry Newborn Screening for CCHD: Indiana's Story

Bob Bowman, MS
Director of Genomics & Newborn Screening
Indiana State Department of Health

Indiana State Department of Health

Background

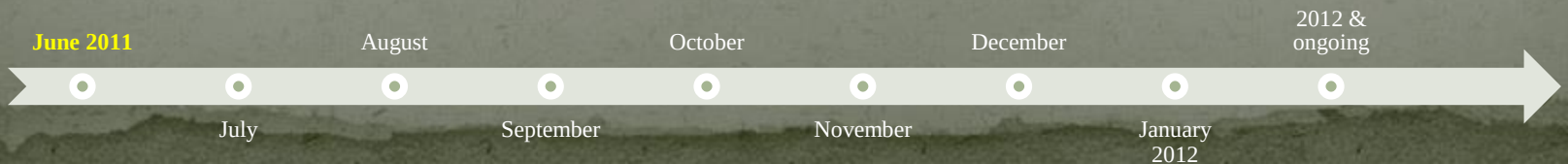
Mission of the ISDH Genomics & Newborn Screening Program

- Responsible for the following:
  - Ensuring that all infants born in Indiana receive state-mandated screening for all designated conditions;
  - Maintaining a centralized program to follow up & ensure that infants who test positive for screened condition(s) receive appropriate diagnosis & treatment & to ensure that parents/families receive genetic counseling; &
  - Promoting genetic services, public awareness, & education concerning genetic conditions

Legislative Action

- Indiana General Assembly amended state newborn screening law (IC 16-41-17) to include pulse oximetry screening for critical congenital heart disease (CCHD)
  - Implementation date: January 1, 2012

- What did this mean for Indiana?
  - Pulse oximetry screening for CCHD now included as part of state-mandated NBS panel
  - Every baby born in Indiana is required to receive CCHD
    - Only legal reason parents/guardians may refuse: based on parents'/guardians' religious beliefs
      - Refusal must be documented in writing with signatures of parent/guardian & witness at birthing facility

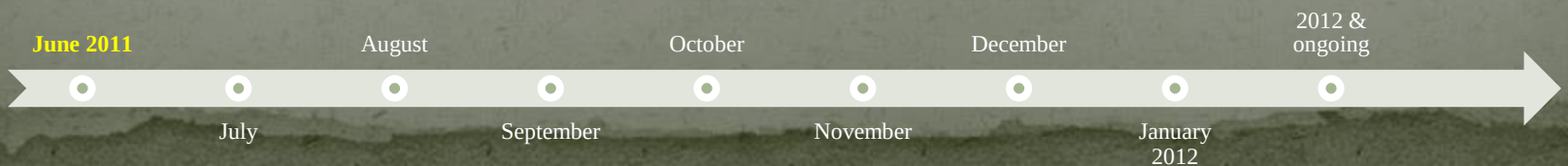


Initial Steps Toward Implementation of CCHD Screening

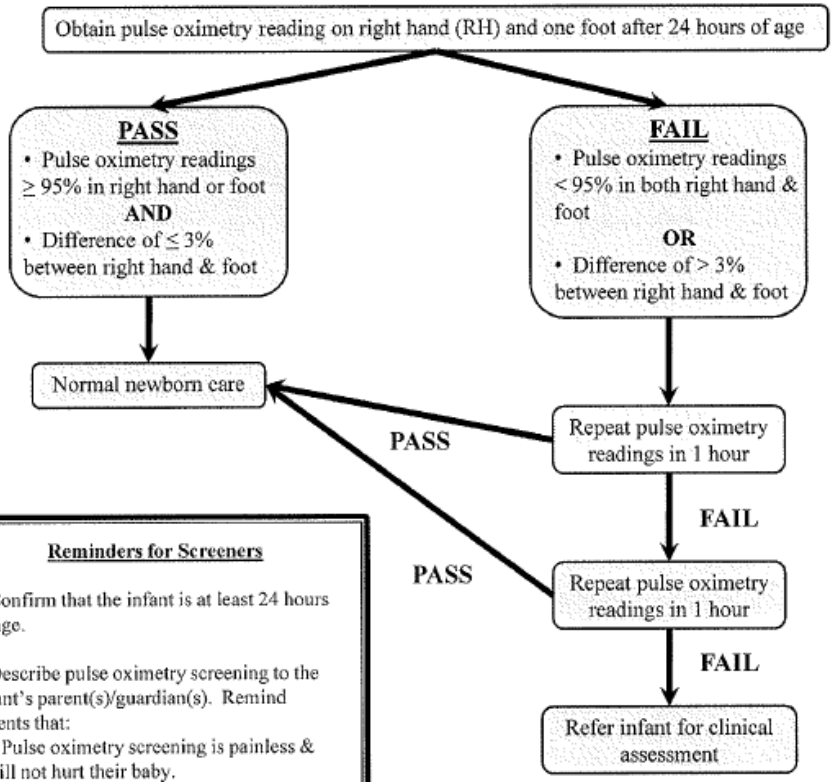
Implementing CCHD Screening

- ISDH contacted representatives listed below for assistance in finalizing Indiana's CCHD screening protocols
  - Neonatologists
  - Nurses
  - Pediatric cardiologists
  - Birthing facilities

- Ultimately, Indiana decided to utilize the screening protocols previously approved by SACHDNC
  - <u>One modification:</u> Instead of waiting 1 hour to rescreen an infant who does not pass, Indiana facilities will only wait 15 minutes
    - Based on recommendations made by Indiana physicians



Pulse Oximetry Newborn Screening Protocols for Healthy Infants (Born at 35+ Weeks Gestation)



Reminders for Screeners

- Confirm that the infant is at least 24 hours of age.
- Describe pulse oximetry screening to the infant's parent(s)/guardian(s). Remind parents that:
  - Pulse oximetry screening is painless & will not hurt their baby.
  - It is possible for a child to have a normal pulse oximetry result & still have a congenital heart defect (CHD).
- Perform the screening in a quiet, peaceful environment. Keep the infant warm & calm.
- Select screening sites that are clean & dry.

Clinical Assessment

- ***Babies with saturation < 90% in right hand or foot should be immediately referred for clinical assessment.***
- ***Babies with 3 failed readings*** (pulse oximetry < 95% in right hand & foot **OR** > 3% difference between right hand & foot) should receive:

1. Clinical assessment (infectious & pulmonary pathology should be excluded)
2. Complete echocardiogram
3. Referral to Pediatric Cardiology
   - Immediately if symptomatic
   - In a timely manner if asymptomatic

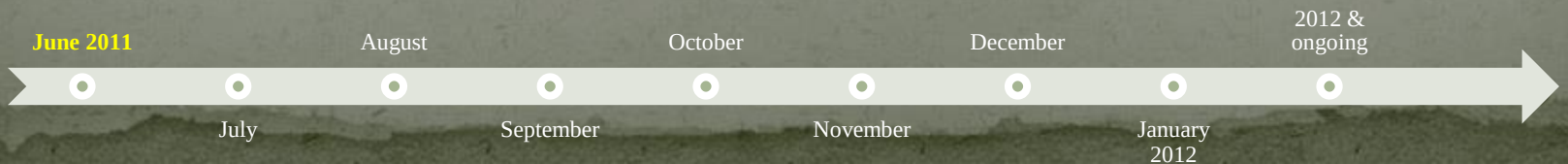
Indiana State Department of Health

Available on ISDH Genomics & NBS Program Professionals website (http://www.in.gov/isdh/25250.htm)

Implementing CCHD Screening (cont.)

- ISDH initially announced the addition of CCHD screening via e-blast to:
  - Primary care physicians
  - Midwives
  - OB/Labor & Delivery/Nursery department manager at each birthing facility

- Collaborated with Indiana Perinatal Network (IPN) to distribute all CCHD-related messages to OB/L&D/Nursery managers
  - IPN had current list of contact information & offered to assist ISDH in reaching these individuals



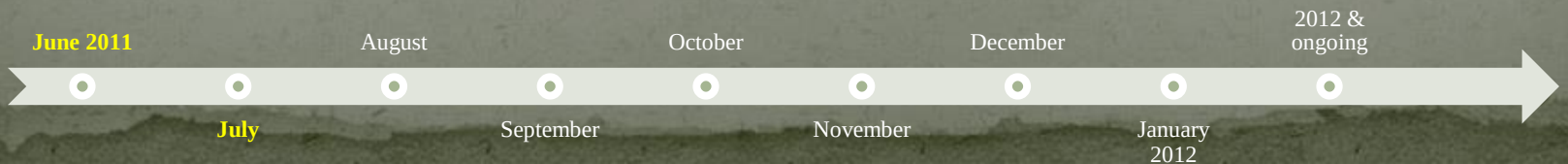
Birthing Facility Survey #1

- Asked OB/L&D/Nursery managers to participate in survey to obtain an overall picture of the state's ability to implement CCHD screening
  - Created through SurveyMonkey
  - Invitation included in initial CCHD announcement

- <u>Questions on this survey:</u>
  - Does your facility already perform pulse oximetry screening on all infants as part of routine newborn care?
  - Does your facility currently have the capacity (equipment, staff, etc.) to implement routine CCHD screening?
  - How many pulse oximeters with pediatric probes are available?
  - Has your entire OB/L&D/Nursery staff been trained to perform pulse oximetry on infants?
  - Where will infants who do not pass the pulse oximetry screen receive follow-up care (defined as pediatric cardiology services, including pediatric echocardiogram)?

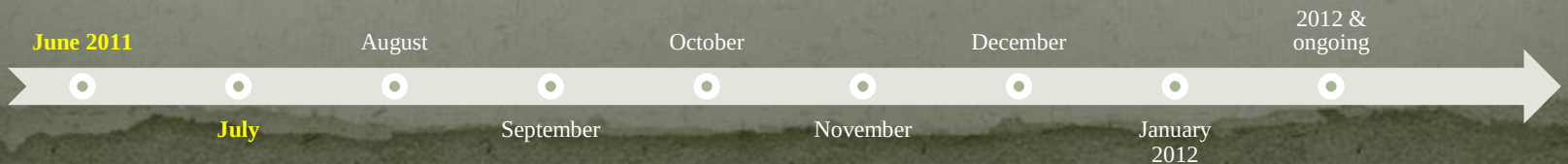
Results of Birthing Facility Survey #1

- <u>49 responses were received</u>
  - 83.7% (41/49) stated their facility did not currently perform routine pulse oximetry screening on newborns
  - 83.7% (41/49) stated their facility already had the capacity to implement routine pulse oximetry screening
  - Facilities reported anywhere from 1 - 10 pediatric pulse oximeters were available for use
  - 71% (34/49) stated that OB/L&D/Nursery staff had already been trained to perform pediatric pulse oximetry screening



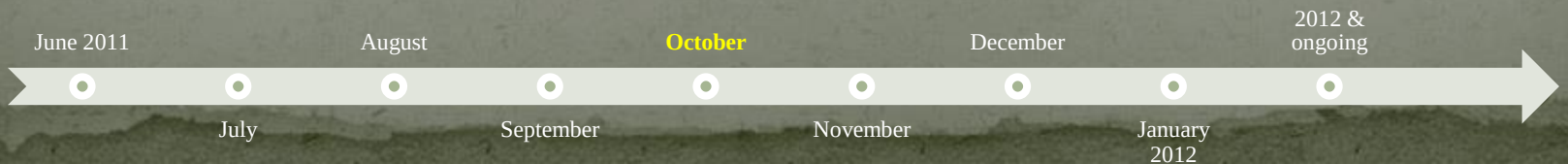
Results of Birthing Facility Survey #1 (cont.)

- 58.3% stated that pediatric echocardiograms would be performed & interpreted at their own facility, negating need to transfer infant to another facility
  - Most of these facilities have
    - Pediatric cardiologists who provide routine outpatient services on-site and/or
    - Can have echos interpreted by pediatric cardiologists via internet
  - <u>If transfer did occur, respondents indicated the following facilities:</u>
    - Riley Hospital for Children, Indianapolis (29.2% or 14/49)
    - St. Vincent hospitals, Indianapolis & statewide (16.7% or 8/49)
    - Out-of-state facilities (Louisville, KY; Cincinnati, OH; Chicago, IL)



Implementing CCHD Screening (cont.)

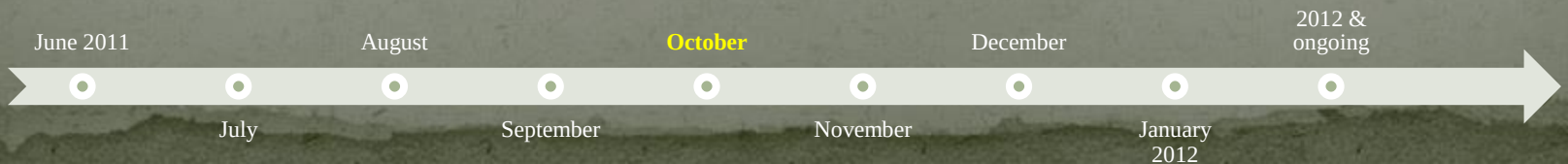
- <u>October 2011 - 2nd e-blast announcement sent to:</u>
  - Primary care physicians
  - Midwives
  - OB/Labor & Delivery/Nursery department manager at each birthing facility
  - Genetic counselors
  - Geneticists
  - Pediatric cardiologists
  - Newborn screening coordinator at each birthing facility



Implementing CCHD Screening (cont.)

- 2nd e-blast included:
  - Updated letter from ISDH NBS Program
  - Finalized screening protocols
  - Link to ISDH NBS Program's Professionals' website
  - FAQ sheet for CCHD

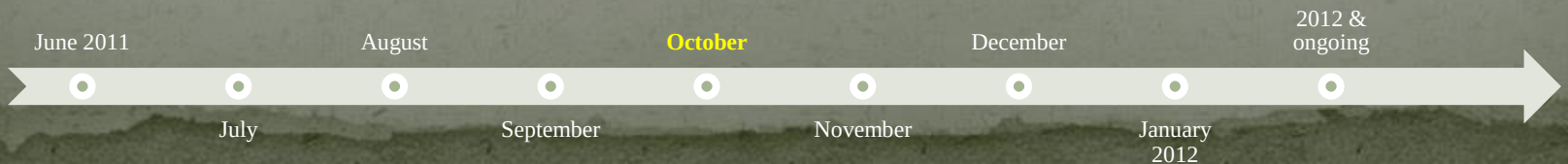
- All birthing facilities were required to submit:
  - Name & contact information of person(s) who would be completing CCHD Monthly Summary Report (MSR)
  - Answers to 2nd birthing facility survey



Birthing Facility Survey #2

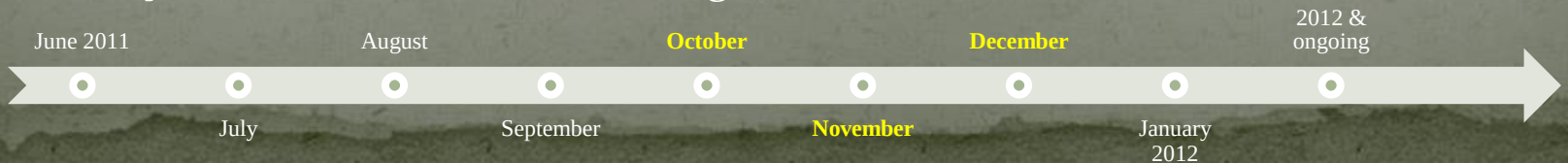
- OB/L&D/Nursery managers received link to 2nd CCHD survey
  - Created through SurveyMonkey

- Questions on this survey:
  - Which birthing facility do you represent?
    - Respondents selected facility name from drop down menu
  - Where will a child born at your facility receive follow-up care (defined as pediatric cardiology services, including echocardiogram) if he/she does not pass the pulse oximetry newborn screen?
    - Respondents could select multiple facility names via checkbox list and/or select "Other" to write in additional facility name
  - What questions do you have for the ISDH Newborn Screening Program?
    - Free text box for respondents to write in questions
  - Contact information
    - Completed if respondents wished to be contacted by ISDH with answers to questions



Results of Birthing Facility Survey #2

- <u>94/101 facilities responded (as of 12/27/2011)</u>
  - 46.8% (44/94) reported that pediatric echos would be performed in-house
    - Echos would be interpreted by in house pediatric cardiologist or by pediatric cardiologist performing outreach services at that facility
  - 29.8% (28/94) would refer child to Riley Hospital for Children at Indiana University Health or St. Vincent facilities
    - 2 largest hospitals/hospital systems with pediatric cardiology services in Indiana
  - 4.3% (4/94) would transfer child to an out-of-state hospital for pediatric cardiology follow-up
    - All of these facilities are located near IN's borders & are typically 2.5 hours' driving distance from an IN hospital offering pediatric cardiology services
  - Remaining facilities responded by indicating another hospital/network within IN's major metropolitan areas that provide in-house pediatric cardiology services
- ISDH NBS Program currently contacting facilities who did not respond to survey invitation to obtain missing information



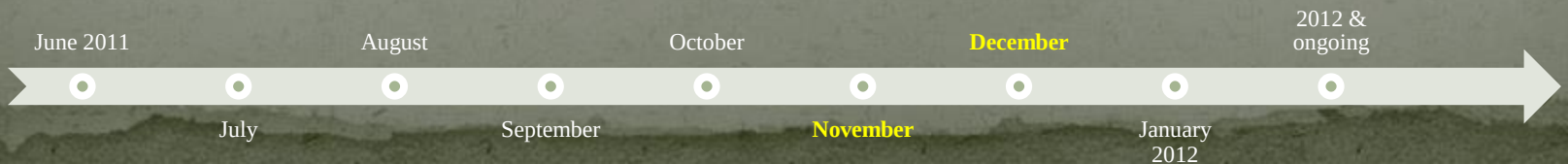
Questions Received - Birthing Facility Survey #2

- Questions submitted by respondents in this survey fell into the following general categories:
  - Detailed questions related to CCHD screening protocols
    - *"[Ar her r comm ndations for] positioning he oxim r prob ?"*
    - *"Is ther a r comm nded leng h of tim w sho ld monitor ach baby on a pulse oximeter?"*
    - Questions related to screening infants born < 35 weeks gestation and/or infants in the NICU
  - Acceptability of screening infants who are discharged prior to 24 hours of age
  - Concerns from midwiferies about feasibility of performing CCHD screening at 24 hours of age
    - *"Our babies ar hom within ho rs of birth. This nec ssita s hom visits, which is an added cost. Do s he AAP support [CCHD scr ning]?"*
    - *"Sinc o r babies go hom a 4 6 hours, can we do the [pulse oximetry screen] at the home visit when we collec he m abolic scr n?"*
  - Availability of religious refusal of CCHD screening
  - Questions related to recommendations for follow-up care (e.g., when should an echo be performed?)
  - Concerns about the length of time required for asymptomatic infants to be seen by Pediatric Cardiology
    - *"I alr ady ak s 2 3 weeks to refer an infant for possible cardiac problems. With the increase in referrals, how long will it ak for asymptomatic babies o b seen? Do sn't his put mor liability on s during he wait [for an infant o b seen for an chocardiogram]?"*
  - Billing for CCHD screening
  - Types of pulse oximeters that are acceptable for CCHD screening
  - How pulse oximetry screen results will be reported to ISDH NBS Program

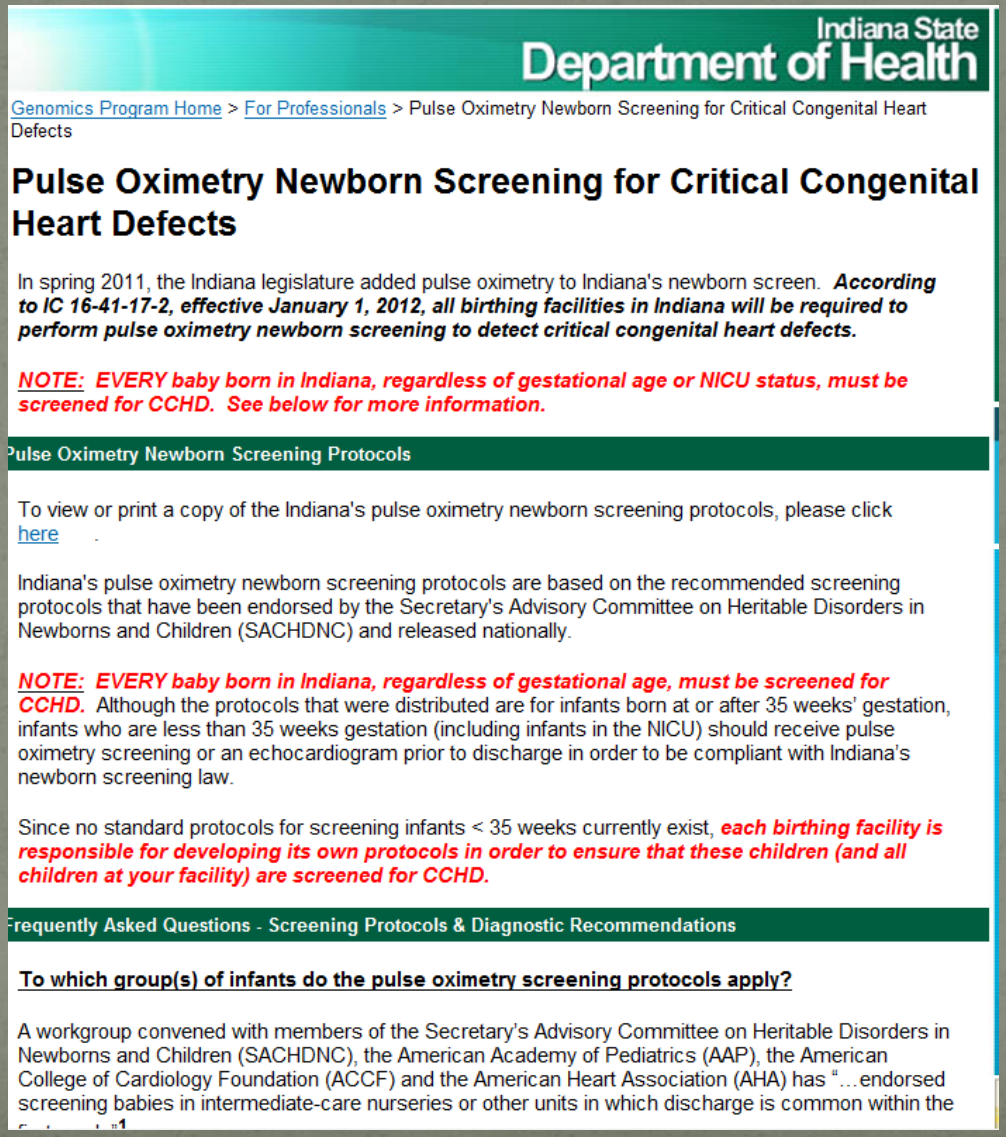
Putting the Pieces Together

Implementing CCHD Screening (cont.)

- Based on results & feedback from birthing facility surveys, ISDH NBS Program:
  - Updated state Religious Waiver form to include refusal of pulse oximetry screening
    - Indiana's NBS law states that the ***only*** legal reason parents/guardians can refuse any portion of NBS is if they object to NBS due to their religious beliefs
  - Updated NBS Professionals' website to include current information about CCHD screening & answers to FAQs
  - Created parent education sheet for CCHD screening in Indiana
    - General information sheet for all parents



ISDH NBS Professionals' Website for CCHD Screening



Indiana State Department of Health

Genomics Program Home > For Professionals > Pulse Oximetry Newborn Screening for Critical Congenital Heart Defects

Pulse Oximetry Newborn Screening for Critical Congenital Heart Defects

In spring 2011, the Indiana legislature added pulse oximetry to Indiana's newborn screen. ***According to IC 16-41-17-2, effective January 1, 2012, all birthing facilities in Indiana will be required to perform pulse oximetry newborn screening to detect critical congenital heart defects.***

NOTE: EVERY baby born in Indiana, regardless of gestational age or NICU status, must be screened for CCHD. See below for more information.

Pulse Oximetry Newborn Screening Protocols

To view or print a copy of the Indiana's pulse oximetry newborn screening protocols, please click here .

Indiana's pulse oximetry newborn screening protocols are based on the recommended screening protocols that have been endorsed by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) and released nationally.

NOTE: EVERY baby born in Indiana, regardless of gestational age, must be screened for CCHD. Although the protocols that were distributed are for infants born at or after 35 weeks' gestation, infants who are less than 35 weeks gestation (including infants in the NICU) should receive pulse oximetry screening or an echocardiogram prior to discharge in order to be compliant with Indiana's newborn screening law.

Since no standard protocols for screening infants < 35 weeks currently exist, ***each birthing facility is responsible for developing its own protocols in order to ensure that these children (and all children at your facility) are screened for CCHD.***

Frequently Asked Questions - Screening Protocols & Diagnostic Recommendations

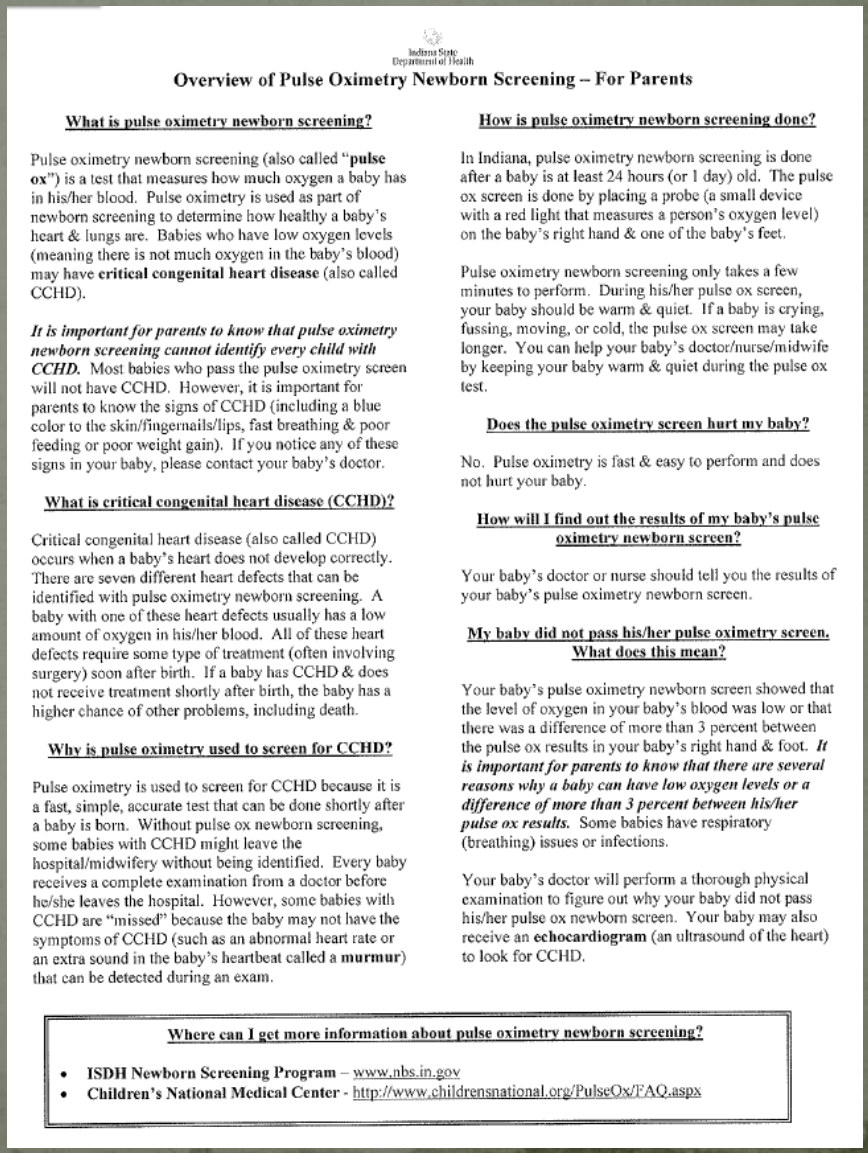
To which group(s) of infants do the pulse oximetry screening protocols apply?

A workgroup convened with members of the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC), the American Academy of Pediatrics (AAP), the American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) has "…endorsed screening babies in intermediate-care nurseries or other units in which discharge is common within the

http://www.in.gov/isdh/25250.htm

Parent CCHD Education Sheet

- Developed/written by ISDH NBS Program
- Approved by ISDH Office of Public Affairs (OPA)
- Reviewed for parent-appropriate content & literacy level by mother of Indiana child whose daughter had one of the heart defects detectable by CCHD screening



Indiana State Department of Health

Overview of Pulse Oximetry Newborn Screening – For Parents

What is pulse oximetry newborn screening?

Pulse oximetry newborn screening (also called "**pulse ox**") is a test that measures how much oxygen a baby has in his/her blood. Pulse oximetry is used as part of newborn screening to determine how healthy a baby's heart & lungs are. Babies who have low oxygen levels (meaning there is not much oxygen in the baby's blood) may have **critical congenital heart disease** (also called CCHD).

It is important for parents to know that pulse oximetry newborn screening cannot identify every child with CCHD. Most babies who pass the pulse oximetry screen will not have CCHD. However, it is important for parents to know the signs of CCHD (including a blue color to the skin/fingernails/lips, fast breathing & poor feeding or poor weight gain). If you notice any of these signs in your baby, please contact your baby's doctor.

What is critical congenital heart disease (CCHD)?

Critical congenital heart disease (also called CCHD) occurs when a baby's heart does not develop correctly. There are seven different heart defects that can be identified with pulse oximetry newborn screening. A baby with one of these heart defects usually has a low amount of oxygen in his/her blood. All of these heart defects require some type of treatment (often involving surgery) soon after birth. If a baby has CCHD & does not receive treatment shortly after birth, the baby has a higher chance of other problems, including death.

Why is pulse oximetry used to screen for CCHD?

Pulse oximetry is used to screen for CCHD because it is a fast, simple, accurate test that can be done shortly after a baby is born. Without pulse ox newborn screening, some babies with CCHD might leave the hospital/midwifery without being identified. Every baby receives a complete examination from a doctor before he/she leaves the hospital. However, some babies with CCHD are "missed" because the baby may not have the symptoms of CCHD (such as an abnormal heart rate or an extra sound in the baby's heartbeat called a **murmur**) that can be detected during an exam.

How is pulse oximetry newborn screening done?

In Indiana, pulse oximetry newborn screening is done after a baby is at least 24 hours (or 1 day) old. The pulse ox screen is done by placing a probe (a small device with a red light that measures a person's oxygen level) on the baby's right hand & one of the baby's feet.

Pulse oximetry newborn screening only takes a few minutes to perform. During his/her pulse ox screen, your baby should be warm & quiet. If a baby is crying, fussing, moving, or cold, the pulse ox screen may take longer. You can help your baby's doctor/nurse/midwife by keeping your baby warm & quiet during the pulse ox test.

Does the pulse oximetry screen hurt my baby?

No. Pulse oximetry is fast & easy to perform and does not hurt your baby.

How will I find out the results of my baby's pulse oximetry newborn screen?

Your baby's doctor or nurse should tell you the results of your baby's pulse oximetry newborn screen.

My baby did not pass his/her pulse oximetry screen. What does this mean?

Your baby's pulse oximetry newborn screen showed that the level of oxygen in your baby's blood was low or that there was a difference of more than 3 percent between the pulse ox results in your baby's right hand & foot. ***It is important for parents to know that there are several reasons why a baby can have low oxygen levels or a difference of more than 3 percent between his/her pulse ox results.*** Some babies have respiratory (breathing) issues or infections.

Your baby's doctor will perform a thorough physical examination to figure out why your baby did not pass his/her pulse ox newborn screen. Your baby may also receive an **echocardiogram** (an ultrasound of the heart) to look for CCHD.

Where can I get more information about pulse oximetry newborn screening?

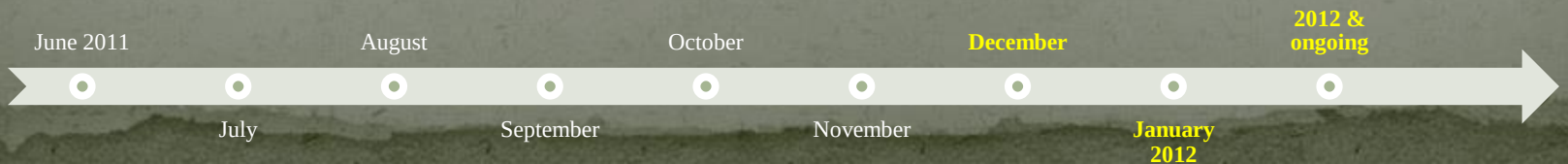
- **ISDH Newborn Screening Program** – www.nbs.in.gov
- **Children's National Medical Center** - http://www.childrensnational.org/PulseOx/FAQ.aspx

Data Collection

CCHD Screening Data Collection

- ISDH NBS Program is currently evaluating available methods of collecting CCHD screening data for all children
  - Including demographic data & pulse oximetry screen results

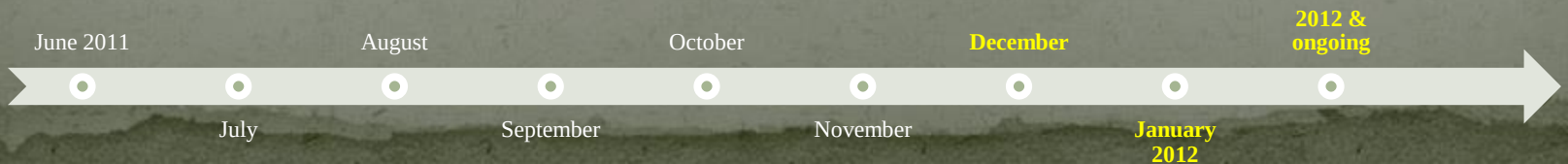
- <u>Possible methods of data collection include:</u>
  - Manual entry of pulse oximetry data onto heelstick card at birthing facility
    - Also involves manual entry of this information into database at state-contracted NBS Laboratory
    - Not very reliable
      - Previous experience with manual entry of hearing screening data has shown that manual data entry leads to high level of incomplete and/or inaccurate data
  - Direct electronic transmission of birth data & pulse oximetry data from birthing facilities to ISDH Repository
    - Utilizing third-party vendor software



INSTEP - CCHD Monthly Summary Report (MSR)

- Indiana Newborn Screening Tracking & Education Program (INSTEP)'s web-based application is already used by all birthing facilities to submit NBS monthly reports for heelstick data

- ISDH NBS Program added a new Monthly Summary Report (MSR) for CCHD screening
  - All birthing facilities will be required to submit CCHD MSR beginning in 2012
    - CCHD MSR users who already use INSTEP (for heelstick MSRs) will submit CCHD MSRs through INSTEP beginning in January
    - New CCHD MSR users will need to attend INSTEP training in 2012
      - Will submit CCHD MSRs using paper forms until trained to use INSTEP

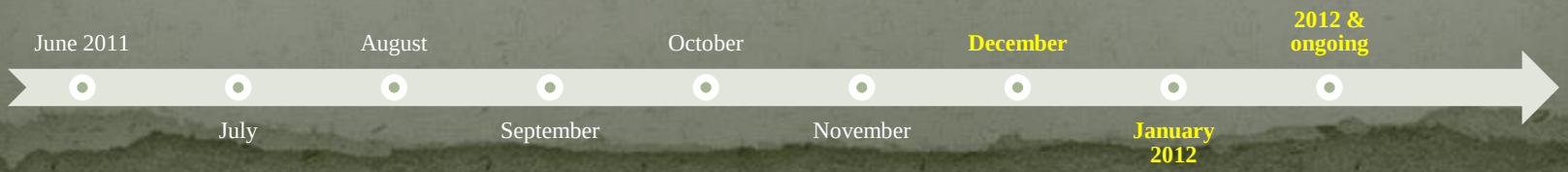


INSTEP - CCHD Monthly Summary Report (MSR)

- Includes detailed information on children born in Indiana who either:
  - Did not pass CCHD screening
    - Facilities are required to report where child will receive his/her follow-up care
  - Did not receive a valid CCHD screen
    - Facilities are required to report the reason ("exception") why a child did not receive a valid CCHD screen
      - Transferred to other birthing facility before 24 hours of age
      - NICU
      - Deceased before 24 hours of age
      - Prenatally/postnatally diagnosed with CCHD
      - Parents/guardians signed Religious Waiver refusing CCHD screen
      - Discharged home without receiving valid CCHD screen (typically due to unauthorized refusal of CCHD screen by parent/guardian)

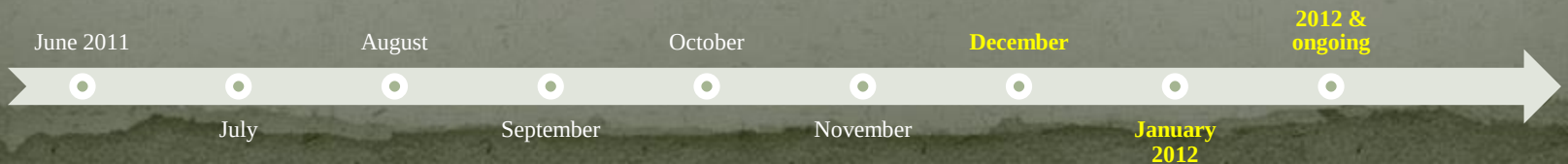
INSTEP & IBDPR

- In order to obtain follow-up information (including health outcomes) on children who do not pass CCHD screening:
  - INSTEP will partner with Indiana Birth Defects & Problems Registry (IBDPR)
    - Another web-based application developed, maintained, & used by Genomics/NBS Program
  - IBDPR collects information on birth defects for all children born in Indiana from birth up to age 3 (age 5 for FAS & autism)
    - Data obtained from direct physician reporting & hospital discharge data (HDD)
      - IBDPR staff performs medical record audits on all children reported to IBDPR through HDD



INSTEP & IBDPR (cont.)

- <u>ISDH NBS Program will utilize data contained in IBDPR in order to:</u>
  - Ensure that all children who did not pass CCHD screen received timely & appropriate follow-up care (including echocardiogram)
  - Evaluate health-related outcomes for children who are diagnosed with at least one of the critical congenital heart defects detectable by CCHD screening
    - Regardless of child's CCHD screen results (passed/did not pass)
  - Evaluate & potentially modify current standards of care for Indiana children with CCHD



Conclusion

Future Activities

- CCHD screening in Indiana will be a "work in progress" for the next few months!

- <u>Future activities include:</u>
  - Implementation of CCHD screening data collection
    - Direct electronic transmission from oximeters to ISDH and/or manual entry on heelstick card
  - INSTEP trainings for MSR contacts who are new users
  - Development of additional parent educational materials and parent website