Newborn Screening Program Report
CY2018 – CY2019 Comparison

Submitted by collaborated team of:

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Introduction

Newborn screening for inborn conditions has been mandatory in Arkansas since Act 192 of 1967 stipulated screening of all newborns for phenylketonuria. Since that time, the number of conditions screened for has grown substantially. The program oversees follow-up on 29 genetic disorders screened using the blood spot card in addition to two point of care tests, hearing screen and critical congenital heart disease, for a total of 31 core disorders. In 2018 and 2019, Ninety-eight point five percent (98.5%) of the approximately 36,000 babies born in Arkansas each year are screened for genetic disorders.

Reports

Several reports are prepared throughout the year and shared with stakeholders.

Newborn Screening Quality Improvement (QI) Graph

Three key data points are monitored monthly: time of birth to time of collection, time of collection to time of receipt in the lab and time of receipt to time of reporting results. Data is provided to ADH Health Statistics representative for compilation. The combined goal for all three data points is less than 168 hours.

NBS QI project:

NBS continues to monitor timeliness data monthly at three points during the process: date of birth to specimen collection, collection to receipt in the lab and receipt to reporting of test results. Time is measured in hours and a goal of less than 168 hours (7 days) from birth to reporting of test results was set.

In 2018, we saw a high of 181.1 hours in November, likely due to three holidays and the HPLC machine was out of service and a low of 141.6 hours in June. ***The 2018 report indicated the average was 149 hours = 6.2 days.***

In 2019, every month was below the goal of <168 hours. January was the highest at 163.7 hours and March was lowest at 136.8 hours. ***The 2019 report indicated the average was 144 hours = 6 days.***

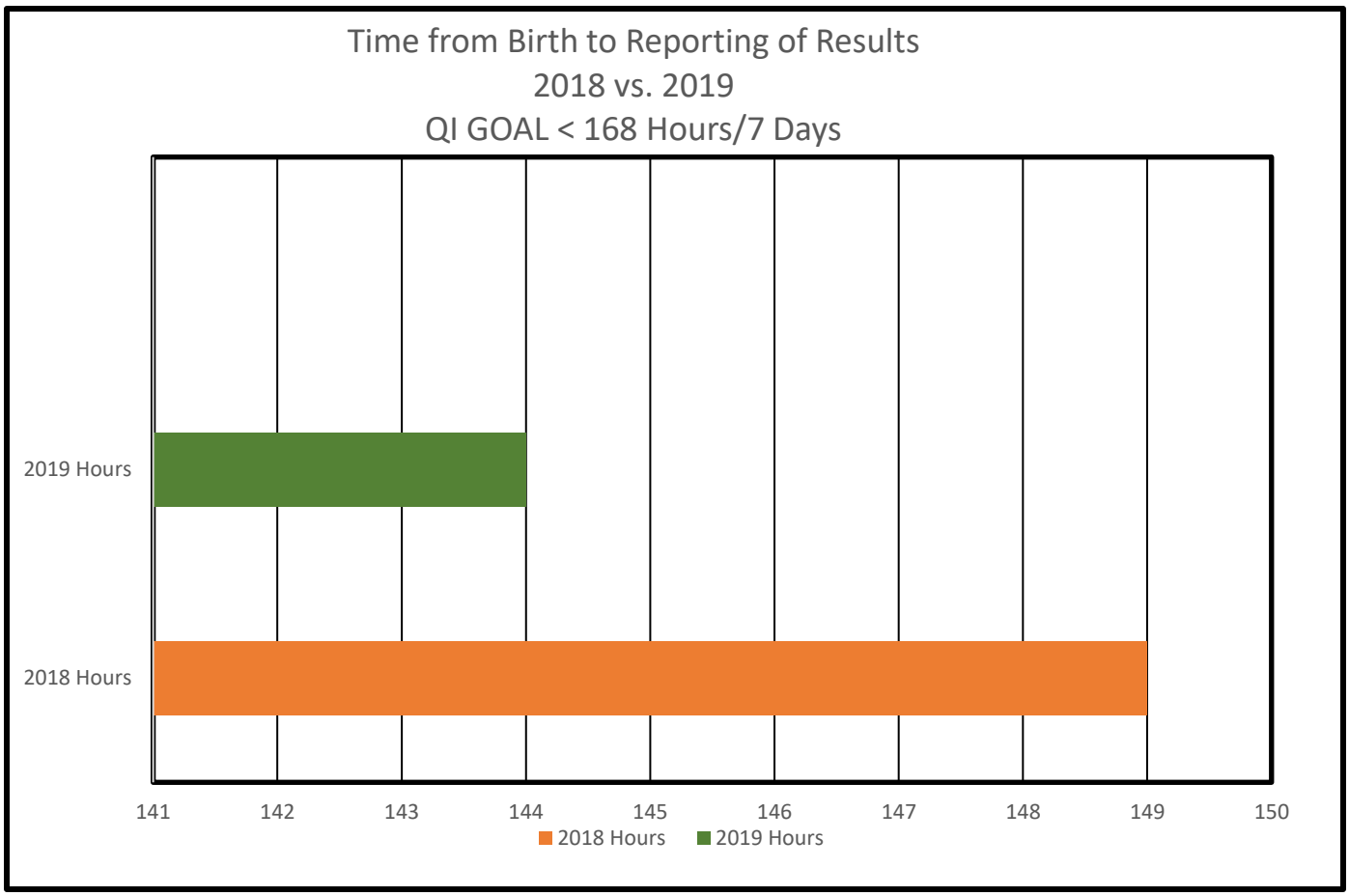


Figure 1

Hospital Timeliness Report

As illustrated in Figure 1: Timeliness reports are distributed quarterly to thirty-nine birthing hospitals and Arkansas Children's Hospital. Reports are sent to three key personnel at each facility: Hospital Administrator, Lab Manager and Nursery/NICU Manager. Data presented include the number of specimens submitted by the facility and a bar graph showing the percentage of those specimens that were received by the lab within 48 hours of collection.

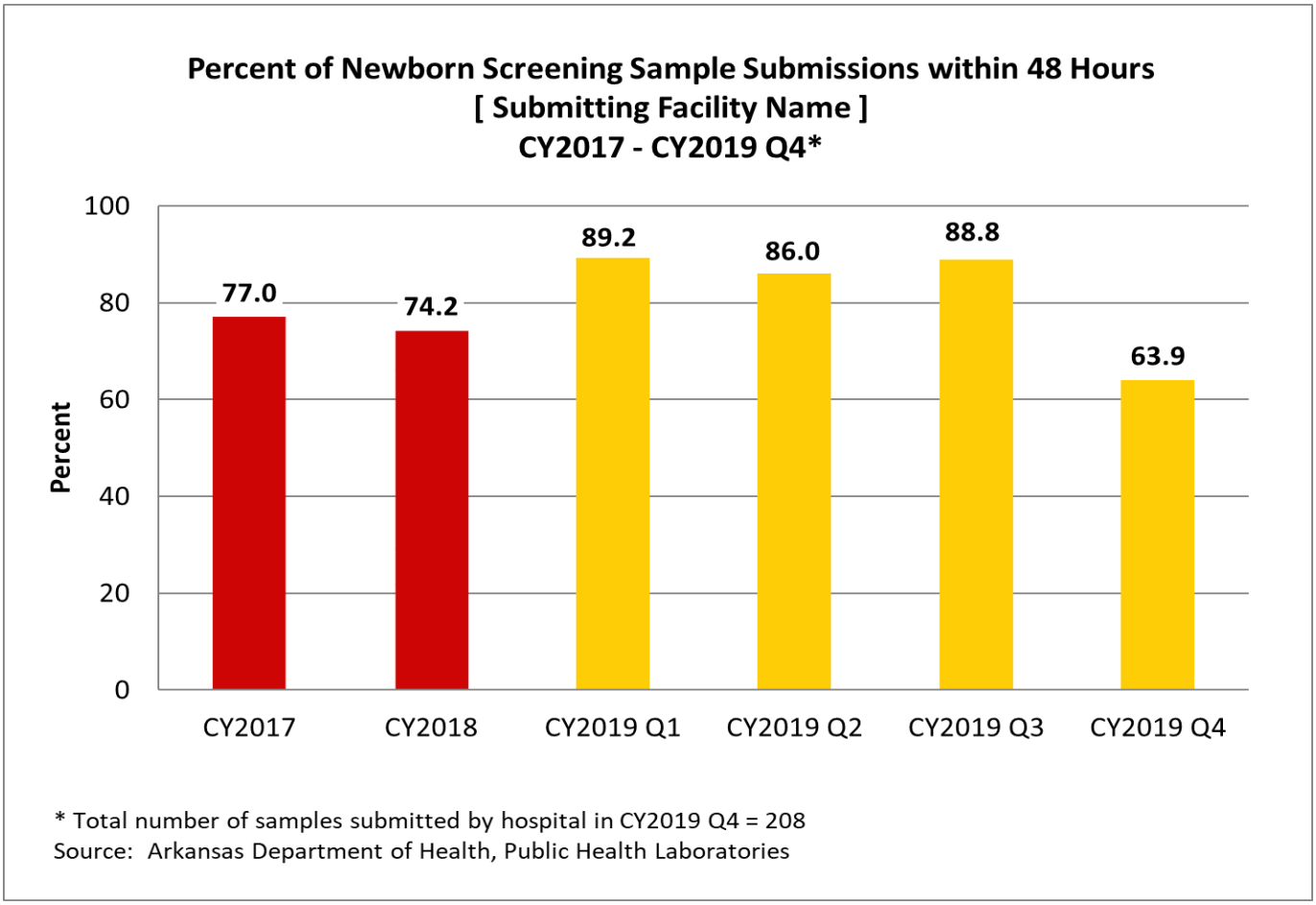


Figure 2

Figure 2 illustrates an example of the quarterly report sent to the hospital administrators, lab and Nursery/NICU managers at each birthing facility and Arkansas Children's Hospital (ACH).

In addition to the quarterly reports, a yearly report representative of all 40 facilities is distributed. This anonymized bar graph report shows placement among peers, with the specific hospital's bar in red, along with the average submission for the state.

Figures 3 and 4 identify state average in 2018 of 85.9% compared to the average in 2019 of 86.4% which indicates an 0.5% improvement.

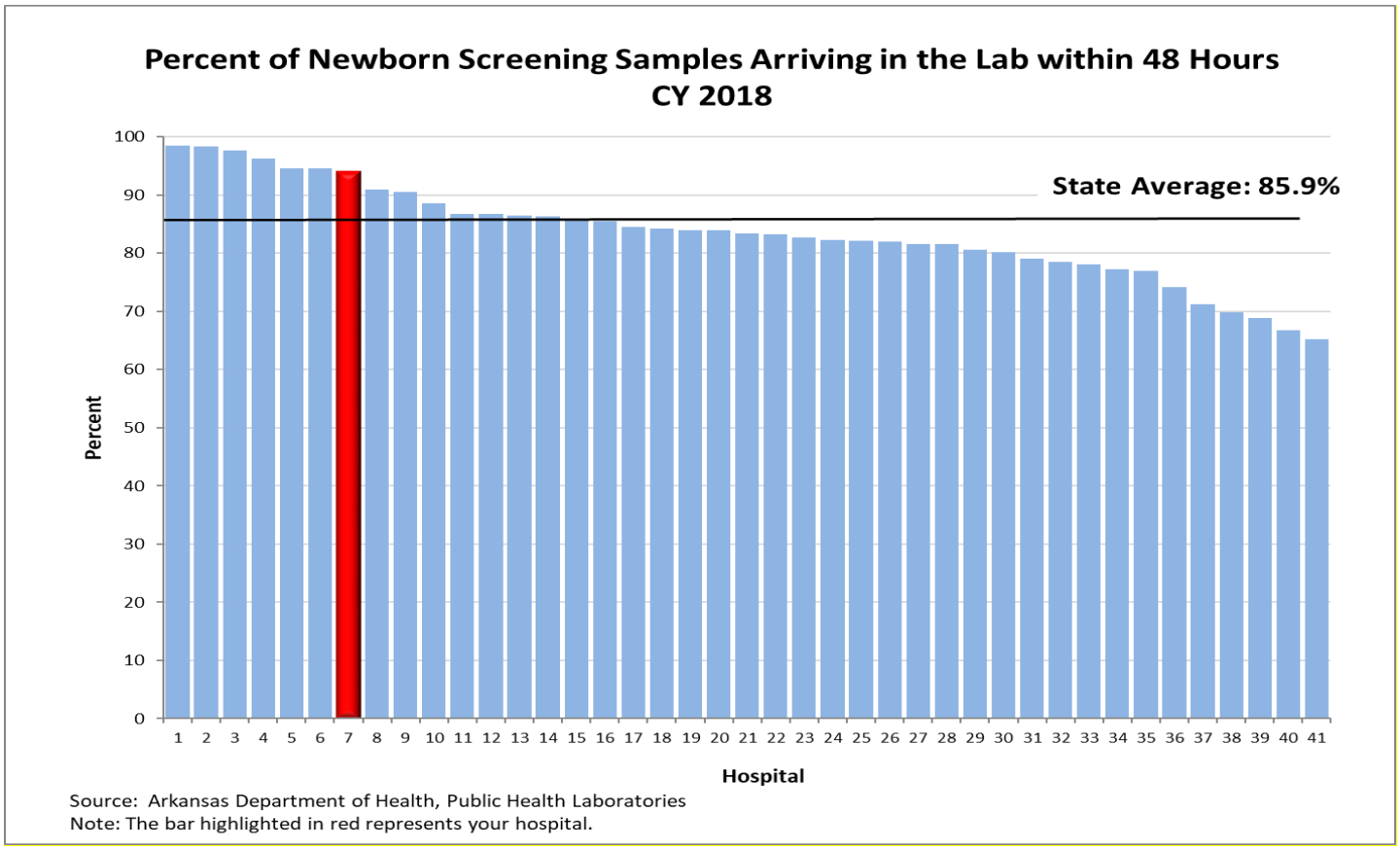


Figure 3

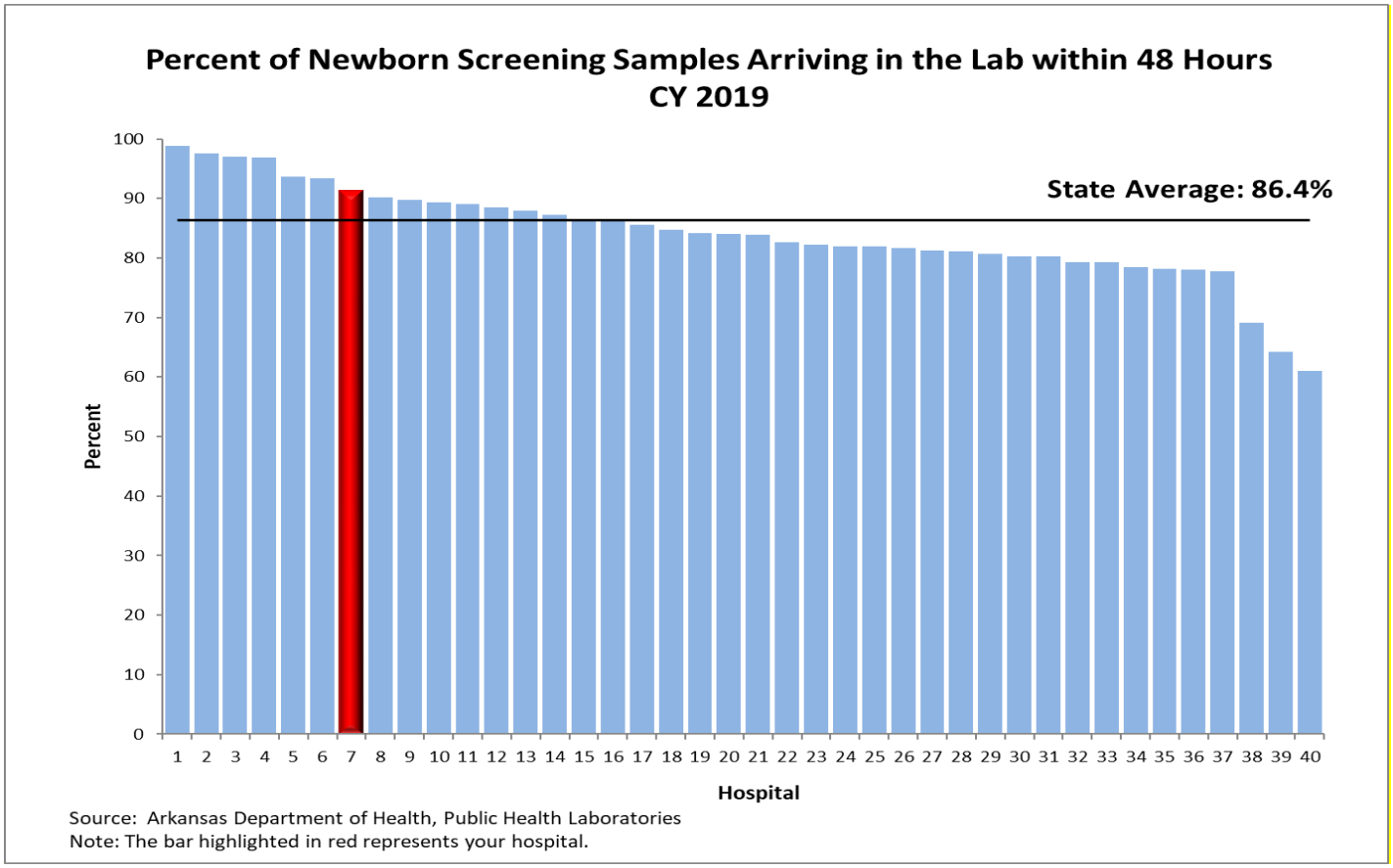


Figure 4

The data for the birthing facilities and Arkansas Children's Hospital are also monitored monthly. Data is compiled by the NBS lab manager and submitted to the AR State Genetic Nurse Coordinator/Hospital educator for use in outreach to the facility when needed. Any facility that does not meet the goal of 80% of the specimens reaching the lab within 48 hours is contacted to discuss any issues related to timely specimen submission.

Comparison 2018 to 2019 Statistics:

The chart below provides a quick comparison view:

Comparison years:	2018	2019
Births	35,738	35,243
*Specimens Received	40,538	39,908
Unsatisfactory specimens not tested	153 (0.4%)	185 (0.5%)
Test Completed	345,223	345,395
Presumptive positive tests	7,097	9,142
Confirmed cases	101	104
Leading confirmed disorder	Congenital Hypothyroidism 48/101	Congenital Hypothyroidism 63/104

Figure 5

In Figure 5, the asterisk (*) identifies the difference in the number of specimens received versus the number of births is due to repeat submissions of unsatisfactory specimens, borderline results, and compliance with Neonatal Intensive Care Unit (NICU) recommendations.

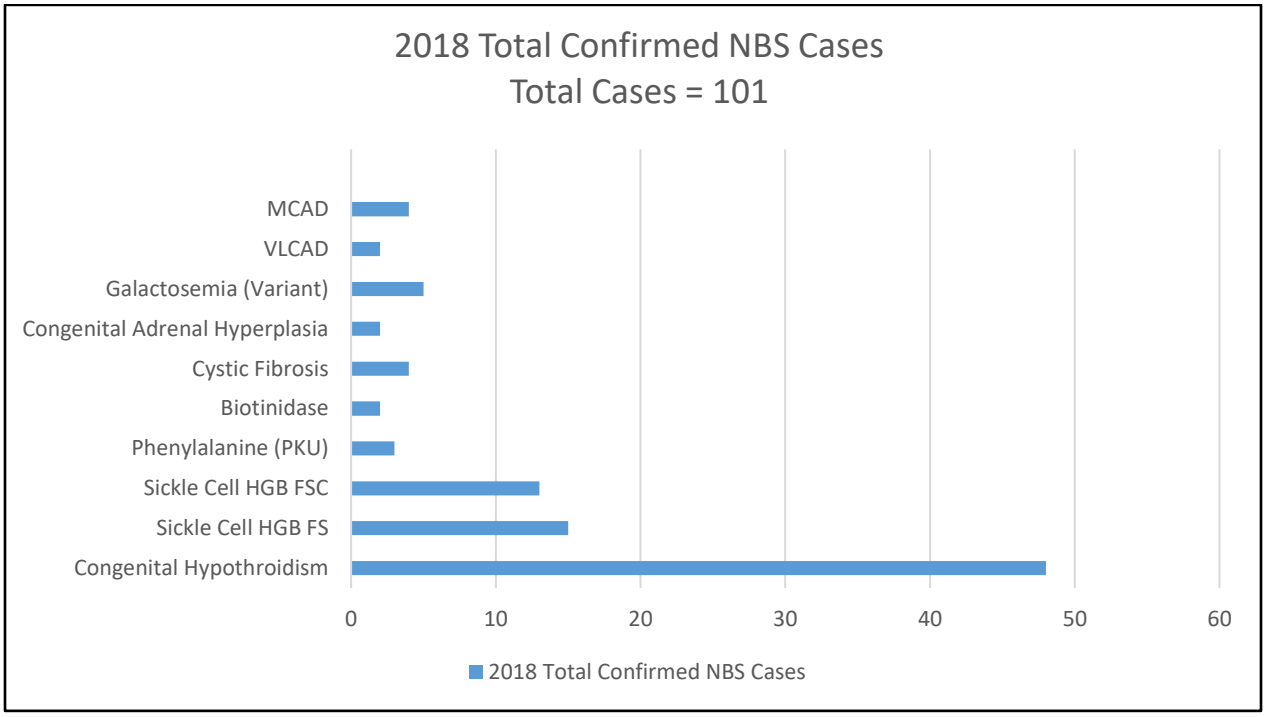


Figure 6

Figure 6: Total 2018 NBS Confirmed Cases =101

Congenital Hypothyroidism – 48 out of 101 = 48%
Sickle Cell HGB FS – 15 out of 101 = 15%
Sickle Cell HGB FSC – 13 out of 101= 13%
Phenylalanine – 3 out of 101 = 3%
Biotinidase – 2 out of 101= 2%
Cystic Fibrosis – 4 out of 101 = 4%
Congenital Adrenal Hyperplasia – 2 out of 101 = 2%
Galactosemia (Variant) – 5 out of 101 = 5%
VLCAD – 2 out of 101 = 2%
MCAD – 4 out of 101 = 4%

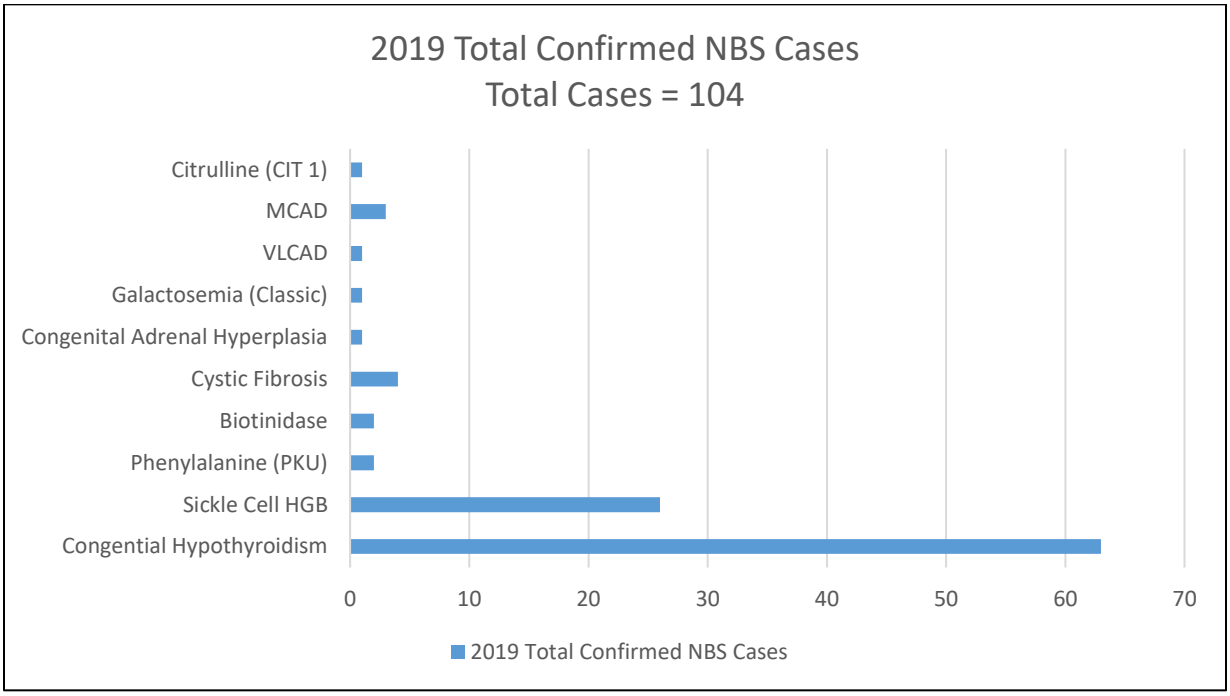


Figure 7

Figure 7: Total 2019 NBS Confirmed Cases =104

Congenital Hypothyroidism – 63 out of 104 = 61%
Sickle Cell HGB – 26 out of 104 = 23%
Phenylalanine – 2 out of 104 = 2%
Biotinidase – 2 out of 104 = 2%
Cystic Fibrosis – 4 out of 104 = 4%
Congenital Adrenal Hyperplasia – 1 out of 104 = 1%
Galactosemia (Classic) – 1 out of 104 = 1%
VLCAD – 1 out of 104 = 1%
MCAD – 3 out of 104 = 3%
Citrulline (CIT 1) – 1 out of 104 = 1%

Figures 6 (2018) and 7 (2019) illustrates a breakdown of the confirmed cases for both years.

In 2018, 48% and in 2019, 61% of total confirmed cases were Congenital Hypothyroidism (CH). Congenital Hypothyroidism (CH) cases were the greatest number of confirmed cases in both years.

NBS Saturday call cases 2018 verses 2019:

In 2018, 129 critical cases were identified during the Saturday call versus 91 critical cases in 2019. The month of January in 2018 presented the largest number of critical cases (28) identified.

The months of March and August in 2019 had the largest number of critical cases (12 each) identified on Saturdays.

Figures 8 (2018) and Figure 9 (2019) provide a visual picture of the number of critical cases reported on Saturdays:

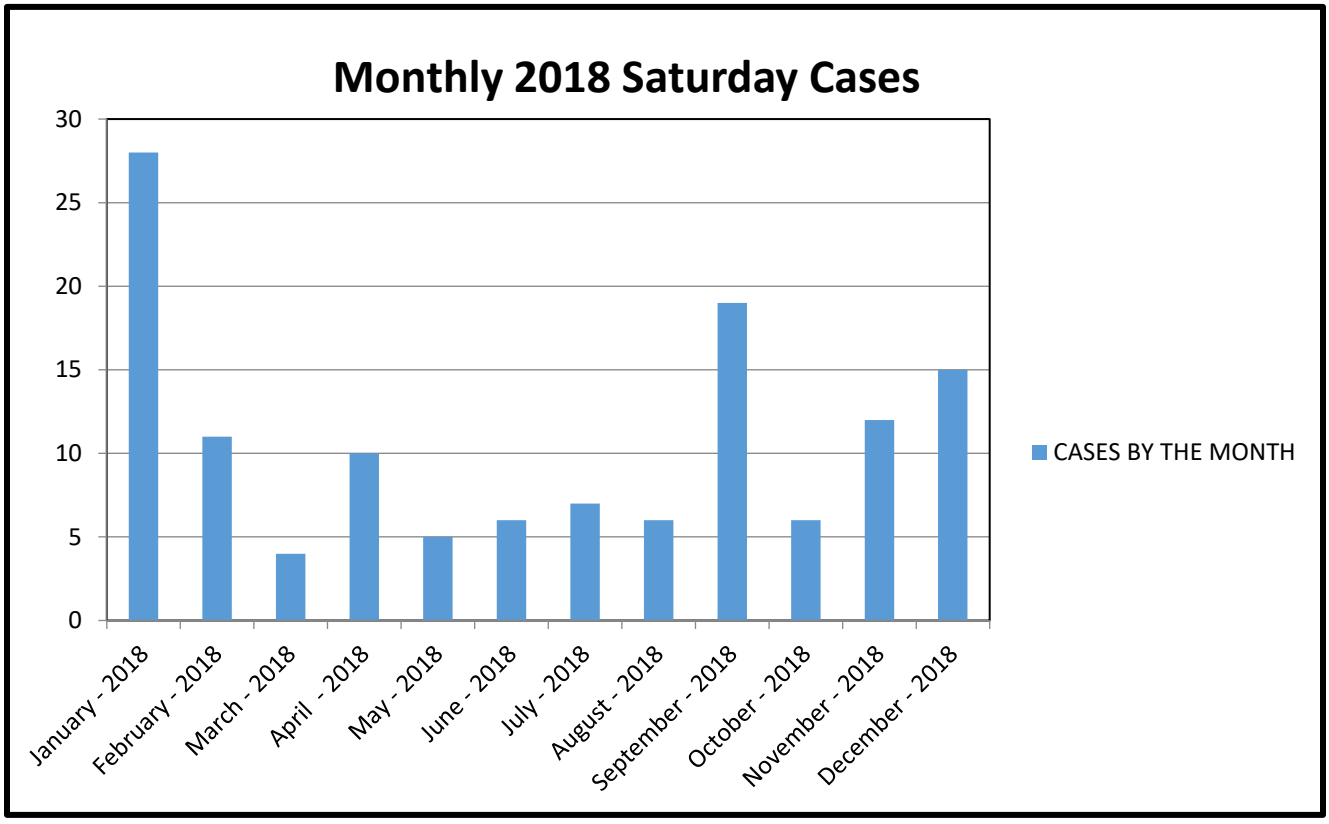


Figure 8

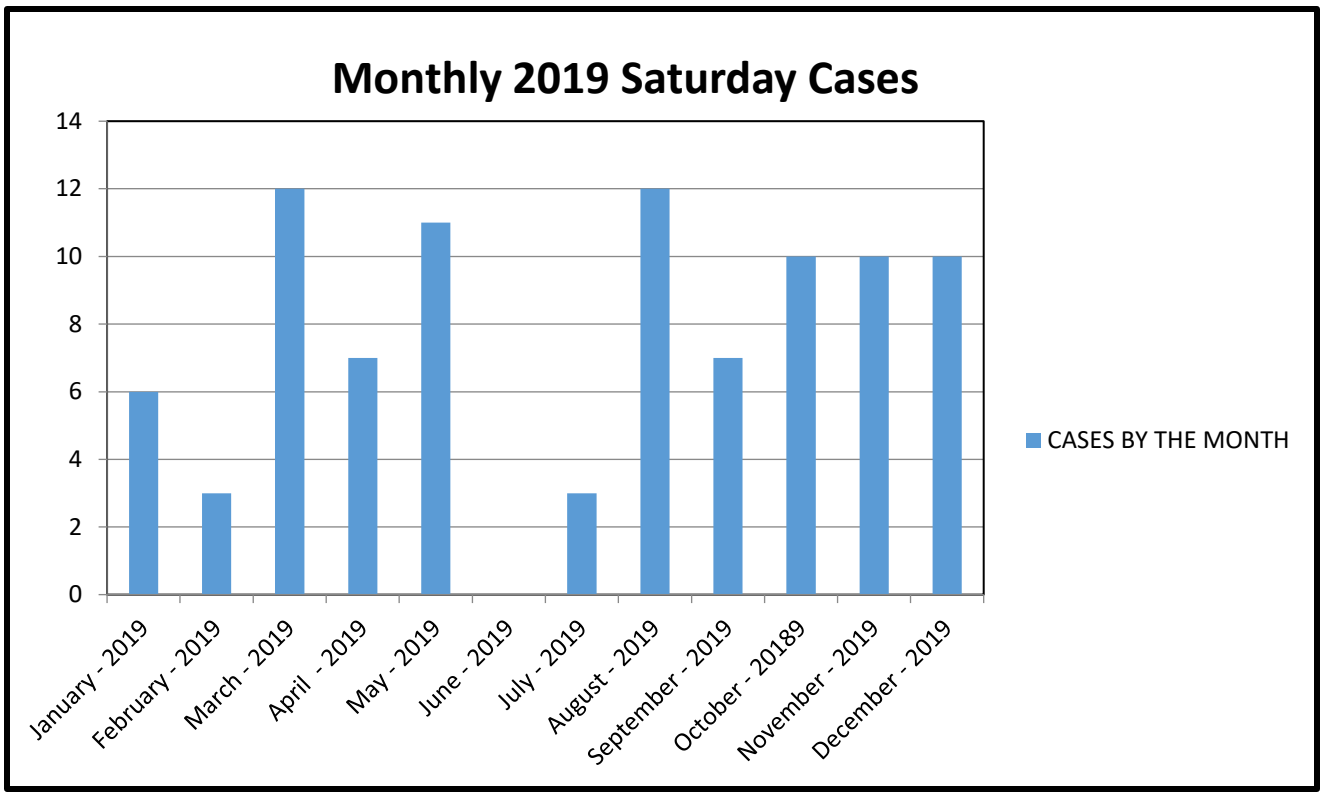


Figure 9

NBS Education:

The NBS program works to provides various types of annual education opportunities for birthing facility staff, Licensed Lay Midwives, and other providers to increase awareness of the NBS protocols and processes. These educational opportunities are offered as an onsite in-service, online course, and annual conferences. Participants are eligible to earn nursing contact hours. Additionally, NBS staff actively participates in professional opportunities offered to NBS programs nationally.

Child and Adolescent Health Section: Educational Opportunities:
AR Dept. of Health Approved Provider Unit #AR-222-8: Feb.1, 2019 - Jan.31, 2022

- Newborn Screening – www.healthy.arkansas.gov
  - Type "Newborn Screening" in the Search for Services space
  - Select Health Professionals from the menu on the left side of the page
  - Click on Video to the right of page
- Critical Congenital Heart Disease (CCHD) – WWW.ARCHILDRENS.ORG
  - In the search window type in CCHD.
  - Dr. Bryan Burke, Jr. is the speaker
  - Click on "CCHD Screening in Newborns with Pulse Oximetry"

- Infant Hearing – http://www.infanthearing.org/nhstc/index.html
    - "Newborn Hearing Screening Training Curriculum" provides options for e-learn with contact hours or without.
- Arkansas TRAIN Learning Network: https://www.train.org/ADH/welcome
    - Create your account if you do not have one.

CHANGE in terminology: nursing continuing education credits are now called CONTACT HOURS not CNE contact hours!

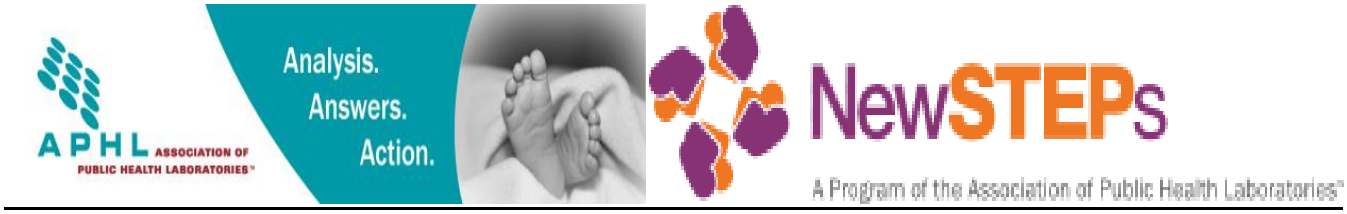
- Courses available on TRAIN for NURSES as of 01. 01. 2020 for **contact hours**:
    - AR NBS-protecting Babies Through a Simple test: Course #1053880 (1.5 contact hrs.)
    - Medical Aspects of NBS: Course #1075665 (2.0 contact hrs.)
    - Introduction to CAH: Course# 1076702 (2.0 contact hrs.)
    - CCHD Screening in Arkansas: Course #1087269 (2.0 contact hrs.)
    - ERAVE Enduring for Hospital Users: Course #1086116 (2.0 contact hrs.)-***NOTE this course is taken after the learner has trained on site with the hospital trainer).***
    - ERAVE Enduring for Hospital Users (**LIVE)Course # 1087154**(2.0 contact hours)-**NOTE this course is taken after the learner has attended a LIVE training (ZOOM or face to face) with Traci Massery, ADH ERAVE trainer.**

AR Licensed Lay Midwives: The ADH Women's Health Section promotes NBS education by including a "Newborn Screening Tool Kit". The program manager ensures each new Licensed Lay Midwife will receive a NBS Tool Kit along with their certificate and welcome packet.

Educational Brochures: NBS brochures are available in three different languages: English, Spanish and Marshallese. They are available to download and print on the ADH website: ***https://www.healthy.arkansas.gov/programs-services/topics/newborn-screening*** **.** Additionally, the NBS program distributes English and Spanish brochures directly to healthcare providers upon request. The Marshallese brochure is available to be downloaded from the website.

Educational Toolkits: Toolkits are provided to the birthing facilities, midwives, and ADH/Local Health Units. If at any time, changes are made to the toolkits, updates are sent out to all recipients by the NBS and Lab Managers.

Plans to add more educational modules to AR TRAIN, an affiliate of the Train Learning Network powered by the Public Health Foundation, will be set up for hospitals, midwives, and local health unit staff to utilize.



In 2018 and 2019 NBS staff completed professional development by participating in conferences offered by including the Association of Public Health Laboratories (APHL) and Newborn Screening Technical Assistance and Evaluation Program (NewSTEPS). These organizations are proactive in providing education opportunities with limited cost to participants.

Local Health Unit (LHU) Reports

Quarterly reports are compiled and sent to the Maternal/Child Health (MCH) Specialists in each region. These reports consist of timeliness data and unsatisfactory specimen information for each LHU that submitted a specimen for screening.

NBS Workgroup: The workgroup meets quarterly to provide educational activities, reports of submission data and current program strategies in newborn screening. This workgroup is comprised of ADH NBS program and Lab staff, ACH specialty physicians, QI representatives and other stakeholders with an interest in newborn screening. In mid-2018, a sub-committee was created within the NBS Workgroup to begin the work of developing the protocol/procedures for upcoming additions to the Recommended Uniform Screening Panel (RUSP).

Arkansas Genetic Health Committee (AGHC): Dr. Brad Schaefer, ACH/ADH NBS Medical Director, chairs the committee which met three times in 2018. The AGHC serves the function of an independent advisory committee to the Arkansas Department of Health, UAMS, State of Arkansas Legislature and other organizations involved in providing genetic services within the state. Committee activities may promote, expand, or coordinate genetic services with the goal of prevention or improvement of genetic conditions. This committee has a responsibility to the citizens of the state, especially those that would require genetic services. They monitor care provided and give advice when appropriate. In addition, they facilitate communication between genetic service providers to assure that services are coordinated among providers throughout the state. AGHC is the overall committee that will oversee the function of the sub-committees, facilitate communication with the legislative branch, coordinate with other regional programs, and oversee educational opportunities for professionals and laymen including families and patients with genetic conditions. It includes members from various professions, parents, patients, service organizations, charitable organizations, and any other interested parties. Media may also be actively involved as deemed appropriate. Members are nominated and approved by the governing body. The AGHC committee was instrumental in the success of Phenylketonuria (PKU) Act 1096 of 2017 for adult PKU food to be covered under a patient's insurance.

Newborn Screening Plans for 2020
The laboratory plans to add four new disorders to the screening panel in 2020.

Mucopolysaccharidosis type 1 (MPS1)

The mucopolysaccharidoses (MPS) are a group of inherited diseases, each caused by a different enzyme deficiency. Seven distinct clinical types and numerous subtypes of the mucopolysaccharidoses have been identified. They are named after the doctor who first identified them.

X-linked Adrenoleukodystrophy

Adrenoleukodystrophy (ALD) occurs when certain fats (very long chain fatty acids, or VLCFAs) cannot be broken down in the body. These fats build up and affect how the body normally functions. This disease largely affects the nervous system and the adrenal glands. When an individual has ALD, the buildup of VLCFAs may disrupt the fatty covering (myelin) of the nerve cells in the brain and spinal cord causing the fatty covering (myelin) to breakdown, which reduces the ability of the nerves to relay information to the brain. Without myelin, the nervous system cannot function properly, causing for example, difficulties swallowing or weakness in the legs. However, these symptoms vary depending on the type and age of onset and other factors which are not well understood. In addition, the build of VLCFAs damages the outer layer of the adrenal glands (adrenal cortex). ALD affects males more severely and is much more common in males as x-linked conditions are very rare in females. However, 20-40% of women who are carriers have symptoms in adulthood.

Pompe Disease

Pompe is an inherited condition that affects many different parts of the body. It is considered a lysosomal storage disorder because people with Pompe have lysosomes (the recycling center of each cell) that cannot break down certain types of complex sugars. This causes undigested sugar molecules and other harmful substances to build up in cells throughout the body, and ultimately, organ failure.

Spinal Muscular Atrophy (SMA)

Spinal muscular atrophy (SMA) is a genetic condition that causes muscle weakness and atrophy (when muscles get smaller). SMA can affect a child's ability to crawl, walk, sit up, and control head movements. Severe SMA can damage the muscles used for breathing and swallowing. There are four types of SMA. Some types of SMA show up earlier and are more severe than others. All types of SMA need ongoing treatment by a medical care team. There's no cure for SMA, but early treatment can help children with SMA live a better life.

The Arkansas NBS program is constantly working to better serve Arkansas families and health professionals.

Mission: To protect and improve the health and well-being of all infants, children, and youth in Arkansas.

Vision: Infants, children and youth in Arkansas are healthy and reach full developmental and academic potential.

Arkansas Department of Health
Newborn Screening Program

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