Section 9

Congenital and Perinatal Conditions

Birth Defects

Every 4½ minutes, a baby is born with a birth defect in the U.S. Major birth defects are conditions present at birth that cause structural changes in one or more parts of the body. They can have a serious adverse effect on health, development, or functional ability. Birth defects are one of the leading causes of infant mortality, causing one in five infant deaths. In Florida, there are approximately 220,000 live births annually and 1 out of every 28 babies is born with a major birth defect.
Despite their substantial impact, only 35% of birth defects have a known cause and research suggests a complex interaction between genetic and environmental factors. In 1997, the Florida Legislature provided funding to the Florida Department of Health to operate and manage a statewide population-based birth defects registry. Per subsection 381.0031(4), Florida Statutes and further specified in Florida Administrative Code Rules 64D-3.029 and 64D-3.035, birth defects are reportable to the Florida Birth Defects Registry (FBDR).

FBDR surveillance data are used for:

- Tracking and detecting trends in birth defects.
- Identifying when and where birth defects can possibly be prevented.
- Providing the basis for studies on the genetic and environmental causes of birth defects.
- Planning and evaluating the impact of efforts to prevent birth defects.
- Helping Florida's families whose infants and children need appropriate medical, educational and social services.

The FBDR is a statewide, population-based passive surveillance program with information on more than 100,000 infants born with serious birth defects. Data are collected on live infants born to mothers residing in Florida who are diagnosed with one or more structural, genetic, or other specified birth outcomes in the first year of life. The FBDR's passive case ascertainment methodology involves the linkage of multiple secondary source datasets including the Florida Division of Public Health Statistics and Performance Management birth records, the Agency for Health Care Administration hospital inpatient and ambulatory discharge databases, Regional Perinatal Intensive Care Centers data, Children's Medical Services (CMS) case management records, and CMS Early Steps data. There is an inherent delay in FBDR data since they include all outcomes through the first year of life. At the time this report was published, the most recent FBDR data available were from 2014 (Table 1).

Table 1. Average Annual Number of Birth Defects and Prevalence Rates (Per 10,000 Live Births) of Selected Birth Defects, Florida, 2010-2014

Central Nervous System Defects	Number of Birth Defects	Prevalence Rate
Anencephalus	17	0.8
Spina bifida without anencephalus	59	2.8
Cardiovascular Defects		
Transposition of the great arteries	51	2.4
Tetralogy of Fallot	105	4.9
Atrioventricular septal defect	88	4.1
Hypoplastic left heart syndrome	69	3.2
Orofacial Defects		
Cleft lip with cleft palate	106	5.0
Cleft palate without cleft lip	110	5.1
Musculoskeletal Defects		
All limb deficiencies (reduction deformities)	81	3.8
Gastroschisis	100	4.7
Chromosmal Defects		
Trisomy 21 (Down syndrome)	289	13.5

For more information, please visit www.fbdr.org.

Neonatal Abstinence Syndrome

Neonatal abstinence syndrome (NAS) occurs in a newborn who was exposed to addictive opiate drugs while in the mother's womb. The most common opiate drugs that are associated with NAS are heroin, codeine, oxycodone (oxycontin), methadone, and buprenorphine. Symptoms of withdrawal depend on the drug involved. Symptoms can begin within one to three days after birth, or may take up to 10 days to appear. Symptoms may include blotchy skin coloring (mottling), diarrhea, excessive or high-pitched crying, excessive sucking, fever, hyperactive reflexes, increased muscle tone, irritability, jitteriness, poor feeding, rapid breathing, seizures, sleep problems, slow weight gain, stuffy nose, sneezing, sweating, trembling (tremors), or vomiting.

In June 2014, NAS became a reportable condition per Florida Administrative Code Rule 64D-3.029. The FBDR is currently conducting enhanced surveillance for NAS. This surveillance incorporates multi-source passive case finding efforts and trained abstractor review of maternal and infant hospital medical records in order to capture all relevant clinical information to classify potential NAS cases, determine specific agents to which the mother and infant were exposed, and to obtain a more complete understanding of this public health issue.

Based on data collected by the FBDR, the rate of NAS in Florida increased dramatically from 1998 to 2010, followed by a slower rate of increase from 2011 (66.7 cases per 10,000 live births) to 2013 (69.2 cases per 10,000 live births). However, in 2014, the rate of NAS increased to 76.6 per 10,000 live births, an 11% increase from 2013. In 2014, NAS rates per 10,000 live births were substantially higher among non-Hispanic white infants (156.2) compared to non-Hispanic black infants (26.6) and Hispanic infants (20.2).

Currently, there is substantial variation in the diagnosis and reporting of NAS across institutions, providers, and surveillance systems. These inconsistencies result in concern about the reliability of NAS data. However, it also represents an opportunity for further establishment of a standardized set of recommendations and guidelines for clinical diagnosis, data collection, surveillance, and reporting. A limitation of using a passive surveillance methodology (without medical record confirmation) to identify NAS cases is the likelihood for misclassification. Misclassification can result in false negatives (the failure to capture an infant born with NAS) or false positives (identifying an infant as a NAS case who does not meet the clinical definition of NAS). Another limitation is the timeliness of the data. There is an inherent delay in FBDR data since they include all outcomes through the first year of life. At the time this report was published, the most recent FBDR data available were from 2014.

Despite limitations, use of FBDR and other existing surveillance systems provides insight into the epidemic of prescription drug abuse and its effects on babies and allows community leaders to obtain a more complete understanding of this important public health issue to support response to local concerns.

For current NAS surveillance data, please visit www.fbdr.org.

Perinatal HIV

Perinatal HIV transmission, also known as mother-to-child HIV transmission, can happen at any time during pregnancy, labor, delivery, and ingestion of breast milk. The Centers for Disease Control and Prevention (CDC) recommends that all women who are pregnant or planning to become pregnant be tested for HIV before pregnancy and as early as possible during every pregnancy. Without treatment, 25-30% of babies born to HIV-positive mothers will become infected with HIV. However, if mothers are aware of their HIV status and treated along with their infants, the chances of the infant acquiring HIV is only 1-2%. Children living with HIV frequently are slow to reach important developmental milestones such as crawling, walking, and speaking. Many do not gain weight or grow normally. Like adults living with HIV, children with HIV also develop life-threatening opportunistic infections. The types of infections are different for children than adults, with serious bacterial infections occurring more often among children. Florida law requires that women residing in Florida be tested for HIV at their initial prenatal care visit, again at 28-32 weeks of pregnancy, and at labor and delivery. This testing requirement allows Florida providers to address any potential missed opportunities to treat during the prenatal period.

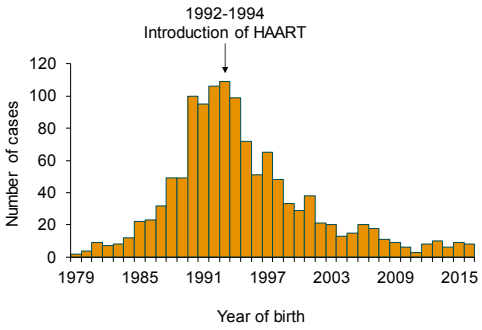
Perinatally acquired HIV prevention in Florida is focused on:

- Prevention services for women of child bearing age.
- Ensuring services for pregnant women living with HIV and their newborns.
- Education and technical assistance for clinicians who treat pregnant women.

Perinatally acquired HIV births in Florida have decreased by 93% from 1993 to 2016. Initiation of highly active antiretroviral therapy (HAART) between 1992-1994 played a significant role in this decrease (Figure 1).

There were 483 infants perinatally exposed to HIV in 2016, but only eight perinatal HIV cases, seven pediatric HIV cases, and one pediatric AIDS case were identified (Map 1). There were several possible missed opportunities where interventions could have taken place to prevent transmission among the 87 infants perinatally infected with HIV from 2007 to 2016 (Figure 2).

Figure 1. Perinatally Acquired HIV Cases Born in Florida by Year of Birth, 1979-2016



Map 1. Infants Perinatally Exposed to HIV by County, Florida, 2016 (N=483)

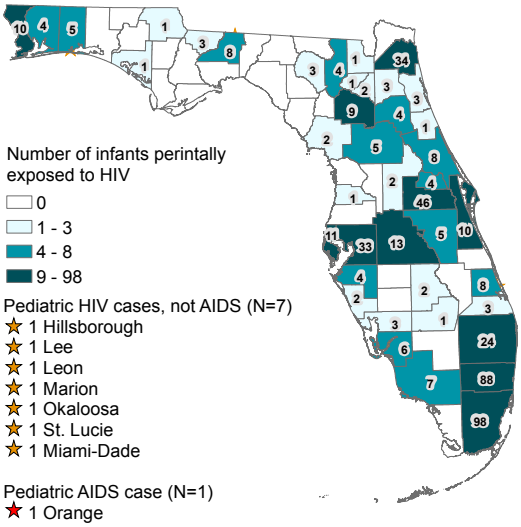
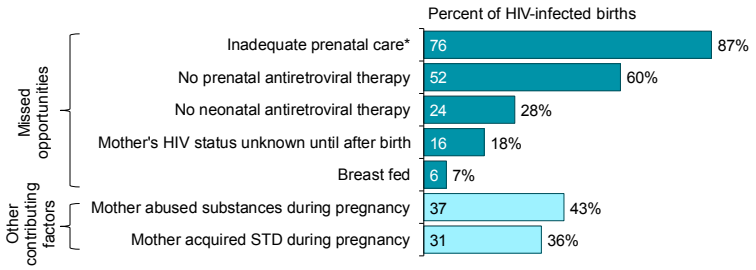


Figure 2. Possible Missed Opportunities for HIV Prevention Among 87 Infants Perinatally Infected With HIV, Florida, 2007-2016



Note that an infant can be included in multiple categories.
* Inadequate prenatal care means prenatal care after fourth month and less than five visits.

For more information about perinatal prevention services, please visit www.floridahealth.gov/diseases-and-conditions/aids/prevention/topwa1.html.

Perinatal Hepatitis B

Hepatitis B virus (HBV) infection in a pregnant woman poses a serious risk to her infant at birth. Without post-exposure prophylaxis (PEP), approximately 40% of infants born to HBV-infected mothers in the U.S. will develop chronic HBV infection, approximately one-fourth of whom will eventually die from chronic liver disease. Perinatal HBV transmission can be prevented by identifying HBV-infected pregnant women and providing hepatitis B immune globulin and hepatitis B vaccine to their infants within 12 hours of birth. Preventing perinatal HBV transmission is an integral part of the national strategy to eliminate hepatitis B in the U.S.

National guidelines call for the following:

- Universal screening of pregnant women for HBV surface antigen during each pregnancy.
- Case management of HBV-positive mothers and their infants.
- Provision of immunoprophylaxis for infants born to infected mothers, including hepatitis B vaccine and hepatitis B immune globulin.
- Routine hepatitis B vaccination for all infants, with the first dose administered at birth.

Please see Hepatitis B, Surface Antigen in Pregnant Women in Section 2: Data Summaries for Selected Reportable Diseases/Conditions of Frequent Occurrence for additional information on HBV surveillance in pregnant women. The 2015 National Immunization Survey estimates that HBV vaccination coverage for birth dose administered from birth through 3 days of age was 72.4% in the U.S. and 53.2% in Florida.[1] Birthing hospitals have a standing order to administer the birth dose; however, pediatricians sometimes choose to wait to give the first dose in their private offices. With lower-than-expected vaccination rates, Florida is currently working with the American Academy of Pediatrics to provide education reminding health care providers that the recommendation is now to provide the birth dose within 24 hours to help decrease HBV infections in newborns. Despite low compliance with administering the birth dose of HBV vaccine, only 10 perinatal hepatitis B cases have been reported over the past 10 years, with the last case occurring in 2014.

1 Hill HA, Elam-Evans LD, Yankey D, Singleton JA, Dietz V. 2016. Vaccination Coverage Among Children Aged 19–35 Months — United States, 2015. Morbidity and Mortality Weekly Report, 65 (39):1065-1071. Available at www.cdc.gov/mmwr/volumes/65/wr/mm6539a4.htm.

Perinatal Hepatitis C

Hepatitis C virus (HCV) infection is a leading cause of liver-related morbidity and mortality. Transmission of HCV is primarily via parenteral blood exposure, and HCV can be transmitted vertically from mother to child. Compared to vertical transmission for infants born to HBV-infected mothers, the rate of vertical transmission for HCV is much lower. Vertical transmission occurs in only ~6% of infants born to HCV-infected mothers, though that rate can double for women who are also infected with HIV or who have high HCV viral loads.[1] According to the Centers for Disease Control and Prevention (CDC), the rate of women of childbearing age testing positive for HCV increased by 22% across the U.S. between 2011 and 2014.[2] CDC recommends that health care providers assess all pregnant women for risk factors associated with hepatitis C and test those who may be at risk.[2] CDC also recommends testing for all infants born to HCV-infected mothers.[2] Having a pediatric specialist can assist in monitoring disease progression in babies and, when needed, aid in intervention. These children should be targeted for vaccination against hepatitis A and B and specialists should monitor any medication that could potentially harm the already fragile liver. More research is needed to better understand whether or not treatment for hepatitis C is safe for pregnant women and children. Florida enhanced efforts to identify these mothers and infants and started performing outreach to those who are at highest risk of transmission. Infants born to HCV-infected mothers should be tested for HCV at the first well-baby visit and at two months, and followed up for any adverse health outcomes.

The incidence of chronic hepatitis C in women of childbearing age has increased dramatically over the past 10 years in Florida (Figure 3). Changes in treatment options for HCV have led to an increased focus on identifying HCV infections. Given the large number of chronic hepatitis C cases reported and limited county health department resources, there have been concerns regarding data completeness and case ascertainment in the past. Earlier data are less reliable. Over the past few years,

improvements in electronic laboratory reporting (ELR) and increased focus on surveillance are believed to have improved case ascertainment. Automated case classification and reporting logic in the surveillance application have improved data quality and sensitivity. In 2014, reporting requirements were updated to include mandatory reporting of all positive and negative hepatitis results, as well as all liver function tests, to support the identification of acute hepatitis C cases. ELR has continued to expand and in 2016, 96.1% of all chronic HCV laboratory results were received by the Department electronically. In 2016, incidence increased even more, primarily due to a change in case definition that expanded the case classification criteria. The number of chronic hepatitis C cases identified in women of childbearing age increased 80% from 2012 to 2016. Despite this very large increase, the incidence of babies infected with HCV has not increased (Figure 4). In an effort to improve case ascertainment in this age group, Florida developed and implemented a surveillance case definition for perinatal hepatitis C in 2016. Previously these cases would have been captured within the chronic hepatitis C case definition. See Table 2 for additional information on perinatal hepatitis C cases reported in 2016.

Figure 3. Number of Reported Chronic Hepatitis C Cases Among Women of Childbearing Age (15-44 Years Old), Florida

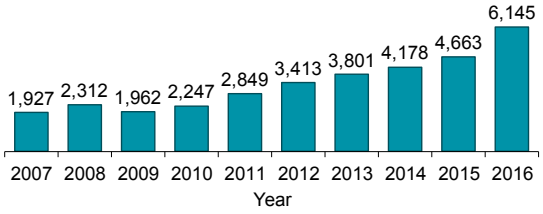


Figure 4. Number of Reported Chronic Hepatitis C Cases Among Children ≤2 Years Old (2007-2015) and Perinatal Hepatitis C Cases (2016), Florida

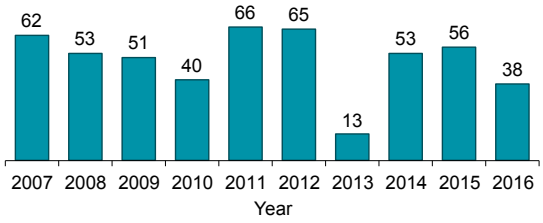


Table 2. Characteristics of Perinatal Hepatitis C Cases Reported in 2016, Florida

Summary	
Number of cases in 2016	38

Gender	Number	(Percent)
Female	20	(52.6)
Male	18	(47.4)
Unknown gender	0	(0.0)

Race	Number	(Percent)
White	25	(65.8)
Black	0	(0.0)
Other	4	(10.5)
Unknown race	9	(23.7)

Ethnicity	Number	(Percent)
Non-Hispanic	24	(63.2)
Hispanic	3	(7.9)
Unknown ethnicity	11	(28.9)

Case Classification	Number	(Percent)
Confirmed	38	(100.0)
Probable	0	(0.0)

Outcome	Number	(Percent)
Interviewed	6	(16.0)
Hospitalized	0	(0.0)
Died	0	(0.0)

Location Where Exposed	Number	(Percent)
Florida	28	(73.7)
Unknown	10	(26.3)

County of Residence	Number	(Percent)
Duval	5	(13.2)
Pinellas	5	(13.2)
Palm Beach	4	(10.5)
Brevard	3	(7.9)
Broward	2	(5.3)
Hillsborough	2	(5.3)
Sarasota	2	(5.3)
Collier	1	(2.6)
Escambia	1	(2.6)
Highlands	1	(2.6)
Indian River	1	(2.6)
Lee	1	(2.6)
Leon	1	(2.6)
Martin	1	(2.6)
Okaloosa	1	(2.6)
Orange	1	(2.6)
Osceola	1	(2.6)
Seminole	1	(2.6)
St. Johns	1	(2.6)
St. Lucie	1	(2.6)
Sumter	1	(2.6)
Volusia	1	(2.6)

1 Koneru A, Nelson N, Hariri S, Canary L, Sanders KJ, Maxwell JF, et al. 2016. Increased Hepatitis C Virus (HCV) Detection in Women of Childbearing Age and Potential Risk for Vertical Transmission — United States and Kentucky, 2011–2014. *Morbidity and Mortality Weekly Report*, 65(28):705-710. Available at www.cdc.gov/mmwr/volumes/65/wr/mm6528a2.htm.

2 Centers for Disease Control and Prevention. Increases in Hepatitis C Threaten Young Women and Babies. Available at www.cdc.gov/nchhstp/newsroom/2016/hcv-perinatal-press-release.html.

Congenital Zika Syndrome

Zika virus emerged in 2016 as a serious public health threat in Florida resulting in a large-scale response by the Florida Department of Health. Over 1,000 imported cases and 285 locally acquired cases were identified in Florida in 2016. Zika is primarily spread through the bite of the *Aedes aegypti* mosquito. Although mosquito transmission is most common, Zika virus has the potential to spread through perinatal or sexual transmission, and rarely through blood transfusions and organ or tissue donations. Illness is characterized by rash, fever, arthralgia, and conjunctivitis. Only about one in five people are symptomatic. Severe disease requiring hospitalization is uncommon. Zika virus infection during pregnancy, including asymptomatic infections, has been linked to fetal abnormalities, including microcephaly. The full spectrum of fetal outcomes resulting from Zika virus infection is yet to be determined and is being studied. Congenital Zika infections can occur in infants, with or without symptoms. An infant with at least one of the following characteristics is considered a case of congenital Zika syndrome: severe microcephaly, decreased brain tissue (including intracranial calcifications), damage to the back of the eye, congenital contractures, or hypertonia.

Surveillance for congenital Zika infections and associated birth defects primarily occurs through identifying pregnant women possibly exposed to Zika virus during pregnancy. On August 3, 2016, the Governor directed the Department to provide free Zika virus infection risk assessment and testing to pregnant women in all Florida counties. All pregnant women with laboratory evidence of Zika infection were followed throughout pregnancy. In 2016, 12 pregnant women were found to have detectable viral ribonucleic acid (RNA) in their serum for two weeks or longer (prolonged viremia). Three gave birth in 2016 and nine gave birth in 2017. One woman with prolonged viremia gave birth to an infant who tested positive for Zika virus, but appeared healthy at birth. No women with prolonged viremia delivered infants with congenital Zika syndrome. Other testing methods during pregnancy, such as amniocentesis, have not been fully evaluated and test specificity and sensitivity is undetermined. It is also unknown what proportion of infected fetuses will have abnormalities. There was only one positive amniotic fluid result in 2016 and that pregnancy resulted in a Zika-related fetal death in 2017.

Zika testing was authorized for all infants born to mothers with laboratory evidence of Zika infection during pregnancy, for infants with abnormal clinical findings potentially related to congenital Zika virus infection, and for infants born to mothers with a history of residence or travel to areas with active Zika virus transmission during pregnancy. Testing was performed at the Department's Bureau of Public Health Laboratories. Placenta samples were collected from women not previously tested or who did not have conclusive test results during pregnancy. Placenta samples testing positive for Zika RNA are indicative of infection in the mother, but not necessarily the infant. Forty-two placenta samples were tested in 2016; three were Zika RNA-positive. The first congenital Zika syndrome case in Florida was born to a mother whose Zika infection was confirmed through placenta testing. For pregnancies that resulted in fetal or infant death, attempts were made to collect samples for Zika virus testing. Tissues from one fetal death tested positive for Zika RNA in 2016. Tissues from one fetal death and cerebrospinal fluid from another fetal death tested positive for Zika virus in 2017.

All infants born to Zika-positive mothers were followed until 12 months of age. Regional Zika Pregnancy Registry Coordinators and other county-level epidemiology staff worked with health care providers to collect medical records at birth and at pediatric visits that occurred at 2, 6, and 12 months. All medical records were reviewed by abstractors from the Florida Birth Defects Registry (FBDR) program. Any infant born with laboratory evidence of Zika infection or with abnormalities associated with Zika virus were referred to Early Steps, an early intervention program in Florida. Early Steps serves these infants from birth until 36 months of age. Infants exposed to Zika virus who did not exhibit abnormalities at birth may later exhibit physical abnormalities or developmental delay. The Department's Bureau of Epidemiology, FBDR, and Early Steps worked together to identify any Zika-related abnormality that developed after birth, and ensured affected infants had access to appropriate programs. The Department is funding additional research on the long-term effects of Zika on infants.

Pregnant women and infants with evidence of Zika infection who moved out of Florida were followed by their new state or territory of residence so that follow-up could continue. Likewise, any pregnant women or infants who moved to Florida were followed during pregnancy and received full infant follow-up.

In 2016, 299 Zika infections were identified in pregnant women. Of those pregnant women, 182 had pregnancy outcomes in 2016 and 103 in 2017 (14 pregnant women were lost to follow-up). Five congenital Zika infections were identified, and four of those infants were classified as congenital Zika syndrome cases (one infant was asymptomatic). Of the four congenital Zika syndrome cases, three had microcephaly and two had intracranial calcifications.

Due to the complexities of Zika testing, tracking of pregnant women, and delay in identifying abnormalities that may develop over time, these numbers may not reflect the true incidence of disease. As we continue to learn more about Zika virus, additional infants may be identified as congenital Zika syndrome cases that were not identified at birth. Any infants born in 2017 or identified as congenital Zika cases in 2017 will be included in the *2017 Florida Morbidity Statistics Report*. See Table 3 for additional information on Zika infections in pregnant women and infants.

For more information on Zika virus disease and infection, please see 2016 Focus: Zika Virus in Florida in the Introduction and Section 2: Data Summaries for Selected Reportable Diseases/ Conditions of Frequent Occurrence.

Table 3. Characteristics of Zika Infections in Pregnant Women and Infants, Florida

Summary		Number
Number of Zika infections in pregnant women		299
Confirmed		52
Probable		247
Congenital Zika syndrome		4
Confirmed		2
Probable		2
Congenital Zika infection (asymptomatic)		1
Probable		1
Pregnancy Outcomes	**2016**	**2017**
Live births	176	103
Infants tested	140	95
Infants with laboratory evidence of Zika	4	1
Non-live births	6	4
Fetuses tested	1	2
Fetuses with laboratory evidence of Zika	1	2
Lost to follow-up	6	8
Transfers		**Number**
Pregnant women with Zika infections transferred to Florida		1
Pregnant women with Zika infections transferred out of Florida		5