Department of Health and Human Services
Maine People Living Safe, Healthy and Productive Lives
Paul R. LePage, Governor
Mary C. Mayhew, Commissioner

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January 26, 2016

Senator Eric L. Brakey, Chair
Representative Drew Gattine, Chair
Joint Standing Committee on Health and Human Services
100 State House Station
Augusta, Maine 04333-0100

Dear Senator Brakey and Representative Gattine:

Enclosed please find the 2015 Annual Report to the Legislature for the Maine CDC Birth Defects Program submitted by the Department of Health and Human Services. This report is required under Title 22 of the M.R.S.A., Chapter 1687. The report discusses the Birth Defects Program's activities and accomplishments in 2015 as well as planned activities for 2016.

Thank you for the opportunity to provide the Joint Committee on Health and Human Services with this Birth Defects Program report.

Sincerely,

Mary C. Mayhew
Commissioner

MCM/klv

Enclosure

Maine CDC Birth Defects Program

January 1, 2015 - December 31, 2015

Submitted to the Joint Standing Committee on Health and Human Services

2015 Annual Report

Maine Center for Disease Control and Prevention
An Office of the Department of Health and Human Services
Paul R. LePage, Governor
Mary C. Mayhew, Commissioner

Table of Contents

Executive Summary	3
Birth Defects Program Context	5
Birth Defects Program Summary	7
Birth Defects Program Tracking System	9
Public Awareness	11
Accomplishments and Future Direction	11
References	12
Appendix A – Reportable Birth Defects	13

EXECUTIVE SUMMARY

December 2015

Background

In May, 1999 the Department of Health and Human Services was authorized to collect information on birth defects in Maine by statute (Title 22: Health and Welfare: Subtitle: Facilities for Children and Adults; Chapter 1687; Birth Defects Program; §8941-§13375). The Maine CDC Birth Defects Program was established within the Department of Health and Human Services, Maine Center for Disease Control and Prevention (Maine CDC).

Purpose

The overall objectives of the Maine CDC Birth Defects Program (BDP), using a public health approach, are:

- to assess the full impact of birth defects on Maine children and their families,
- to improve access to specialty services for families and locate resources for emotional and economic support, which includes referring the infant with a confirmed birth defect to the Department of Education Part C – Child Development Services (CDS) program,
- to monitor trends related to prevalence of selected birth defects in Maine and
- to educate the provider community and the general public on prevention strategies to decrease the incidence of birth defects in Maine.

Highlights

This 2015 annual report summarizes the current activities of the Maine CDC Birth Defects Program, the ongoing and upcoming activities for calendar year 2016 and shows selected birth defects counts and birth prevalence for the years 2008 – 2012. In 2015, the Maine CDC BDP:

- Contacted families with a baby with a confirmed birth defect to offer support and information regarding birth defects.
- Completed and submitted the Annual Report of selected birth defects to the U.S. Centers for Disease Control and Prevention, National Birth Defect Prevention Network.
- Continued to make referrals for babies with a confirmed birth defect to the Maine Department of Education/Child Development Services – Individuals with Disabilities Education Act (IDEA) Part C Agency.
- Met regularly with the Maine CDC Environmental Public Health Tracking Unit to discuss updates to the Environmental Tracking Portal related to birth defects.
- Met regularly with the two abstractors that work with the Maine CDC BDP to develop skills. Information discussed included:
    - Specific criteria and definition for the current list of birth defects being abstracted
    - Issues around access to medical records
    - Discussion specific to critical congenital heart defects (CCHD)

- Provided educational materials about the prevention of birth defects to parents, health providers and other interested parties.
- Provided an educational presentation about the Maine CDC Birth Defects, cleft lip and palate and newborn screening programs to hospital staff and public health nurses in northern Maine.
- Convened the Maine CDC Birth Defects Advisory Committee three times to discuss relevant birth defect program issues and activities.
- Participated in the New England Birth Defects Consortium (NEBDC). The NEBDC is a regional collaboration of New England states (Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island and Vermont) established in 2008 to improve services for infants and children in New England with birth defects by promoting regional collaboration in data sharing, research, prevention and health care quality activities, http://www.nebirthdefects.org/. The current initiative focuses on collecting data in Region I regarding risk factors related to critical congenital heart defects. This initiative was completed in the summer of 2015.
- Participated in the annual National Birth Defects Prevention Network Meeting which occurred in October 2015.
- Met regularly with ChildLINK staff to discuss any issues with the database as well as to work toward expanding and increasing its use.

For more information on the Maine CDC Birth Defects Program:
Contact Diane Haberman, Program Coordinator, diane.haberman@maine.gov or 207-287-8424
http://www.maine.gov/dhhs/mecdc/population-health/mch/cshn/birth-defects/index.html

Program Context

Background

A birth defect is defined as an abnormal condition that occurs before or at the time of birth. Birth defects include a wide range of abnormalities with varying levels of impact. Some birth defects are serious and can result in death, while others are less severe and can be treated with appropriate medical services. Birth defects may be caused by genetic factors, environmental, drug or medication exposures while others remain unexplained. Birth defects can cause both mental and physical disabilities that affect children and their families for life.[1]

Birth defects affect about one in every 33 babies born in the United States each year. They are the leading cause of infant deaths, accounting for more than 20 percent of all infant deaths. Babies born with birth defects have a greater chance of illness and long-term disability than babies born without birth defects.[2] In order to prevent birth defects, it is essential to know what types of birth defects are occurring. A population-based birth defects surveillance program that uses multiple sources of data allows a surveillance program to accurately quantify morbidity and mortality, detect temporal trends and assess the financial burden on families and State programs that birth defects may cause. Many children who survive have a lifetime of major expenses from essential services such as, specialty medical care, special education, rehabilitation and developmental services.

The Maine Birth Defects Program (BDP) is committed to fulfilling its mission that all infants with birth defects are identified early and referred to an established network of services, in order to achieve optimal health and develop to their fullest potential. The BDP is a comprehensive surveillance program that benefits the residents of Maine through the early identification of infants who have birth defects. Early identification ensures timely and appropriate access to systems of care that are family-centered, culturally competent and community-based. It is the intention of the BDP to participate fully in epidemiological investigations as a means of informing public policy, to develop prevention strategies in order to reduce birth defects and to assess for timely referrals and follow-up care to reduce mortality and morbidity among children identified with birth defects.

Legislation and Rules

Legislation supporting the Maine BDP has and continues to define the purpose of the program. The statutes defining these roles and responsibilities are listed below.

May 1999, Public Law (P.L.) 1322, 22 M.R.S.A. c. 1687, established the BDP within the Maine Department of Health and Human Services. Program rules were formally adopted April 2003 outlining reporting responsibilities and access to medical records. Mandated reporting began May 2003.

May 2008 – Rules were updated to include three additional reportable birth defects.

April 2011 – Rules were amended to include the 45 birth defects recommended by the U. S. CDC and the National Birth Defects Prevention Network. Referral to the Part C Agency, Child Developmental Services was also included.

Stakeholders

The following is a brief listing of organizations that have a strong association with the Birth Defects Program. The March of Dimes and the National Birth Defects Prevention Network are also key national-level partners.

Consumers • Parents and families	**Early Intervention Agencies** • Department of Education – Child Development Services
External groups involved with screening, follow-up and diagnosis • Hospitals and their staff • Nurses • Primary care providers • Specialist physicians • Genetic counselors	**Other DHHS Programs** • Newborn Hearing Screening Program • Newborn Bloodspot Screening Program • Environmental Public Health Tracking • Data, Research and Vital Statistics • Office of MaineCare Services

Birth Defects Advisory Committee

The Advisory Committee meets at least annually and at times more frequently. The Committee consists of interested parties including parents, health professionals, outside agencies including the Maine Chapter of the March of Dimes and other State, community and private sector agencies, as well as the Maine CDC Children with Special Health Needs staff. The group provides consultation to the BDP on development, implementation and evaluation of program policies, procedures and activities.

Summary of Activities

The Birth Defects Advisory Committee met three times this past year. Items discussed included:

- Review of options to share existing data with interested parties,
- Methods to reach out to families and other interested parties to spread the word about birth defects prevention,
- Methods of outreach to parents at the birth hospital with information and support and
- Updates to implementation of critical congenital heart defects screening and follow up within New England and Maine.

Birth Defects Program Summary

Program Description

The Maine Birth Defects Program began developing a birth defects surveillance system in 1999 with funding from the U.S. Centers for Disease Control and Prevention (U.S.CDC). The Maine CDC Birth Defects Program was established to identify newborns with birth defects, ensure that they receive appropriate specialty services and to monitor birth defect trends.

As a surveillance unit, the Birth Defects Program began passive case ascertainment with confirmation of cases by active case ascertainment on May 1, 2003. Passive case ascertainment with active case ascertainment is an approach whereby the surveillance program receives case reports of birth defects from a variety of data sources and then follows up with a review of the case. As required by statute, those entities licensed under Title 22: Hospitals and Title 32: Licensed Professionals are required to provide or make available health records and information relating to the occurrence of birth defects. Passive data sources include hospital case reports, birth and death certificates and medical discharge records using diagnostic codes. Once a case is identified as a possible reportable birth defect, the case is assigned to an abstractor. The abstractor visits the hospital to review records to confirm the presence of a birth defect.

The Birth Defects Program (BDP) recently updated the listing of reportable birth defects to reflect the birth defects surveillance guidelines developed by the CDC National Birth Defects Prevention Network. The current listing of reportable birth defects may be found in Appendix A.

The BDP receives medical discharge data electronically from all birthing hospitals at this time. The BDP sends all hospitals a monthly electronic reminder to submit data.

In order for a case to be considered by the BDP, it must meet one or more of the following criteria:

- ◊ Infant was live born, stillborn or prenatally diagnosed, with a gestational age of greater than 20 weeks,
- ◊ Fetuses less than 20 weeks gestation but with a prenatal diagnosis,
- ◊ Birth occurred in Maine and the mother was a Maine resident,
- ◊ Diagnosis was made before the infant reached 1 year of age and
- ◊ Birth defect is included in the Maine CDC BDP list of reportable birth defects.

Potential cases are identified through weekly downloads of both the electronic birth and infant death certificates and medical records discharge data. Once a potential case is identified, abstraction is performed using a comprehensive electronic abstraction method. Information collected includes the nature and details of the birth defect, demographics, mother's health history, prenatal information, cytogenic and laboratory data, family history and, when available, father's history, and mother's exposure to illegal drugs, medications, smoking or alcohol.

The data collected by the abstractors is reviewed and entered in the birth defects surveillance and tracking system, called ChildLINK. ChildLINK was built to connect existing State information systems with data obtained from hospitals, health care providers and others

mandated to report birth defects. Once a child is confirmed to have a birth defect, the child's family is notified by mail of services available to them.

The BDP reports on and gathers information on 45 birth defects (see Appendix A for a complete listing of reportable birth defects). Cases are confirmed usually within the first three months after birth. However, this time frame can be longer depending on when the birth defect was reported and the ability of the abstractors to gather the necessary information from the birth hospital. There is also a lag time in verifying a birth defect because, by law, a birth defect can be diagnosed and reported within the first year of life.

Personnel and Funding Sources

The Birth Defects Program consists of one full-time coordinator and two part-time contractors for the provision of abstraction services. The Birth Defects coordinator position is funded through the Maternal and Child Health Block Grant. The two abstractor contracts are funded by the State General Fund.

Goals and Activities

The Birth Defects Program gathers data about infants born each year with certain birth defects diagnosed within the first year of life. The statute requires that the "Program:"

- Collect, analyze and distribute information to identify the birth defects with regard to the following: causes, risk factors and strategies for prevention and the provision of services,
- Establish a system for data collection that identifies prevalence and incidence rates by region and population group and identifies the morbidity and mortality rates resulting from birth defects,
- Contact families to provide information about available resources and services and
- Conduct investigations to determine the nature and extent of the disease or known or suspected causes of birth defects.
- Examples of Maine BDP activity:
  - Collection and abstraction of data on 45 birth defects. The BDP submits data annually to the National Birth Defects Prevention Network and U.S. CDC.
  - Collaboration with the Maine CDC Environmental Public Health Tracking Unit to show 12 of the 45 birth defects on the Environmental Health Tracking Portal.
  - Connection with a family once a baby is identified with a confirmed birth defect. The baby identified with a confirmed birth defect is also referred to the Maine Department of Education Part C Agency (CDS) for follow-up for early intervention services.
  - Participation with the New England Birth Defects Consortium. The Consortium has collected regional data on risks factors in critical congenital heart defects in the summer of 2015. A report is currently being written on the outcome of this work.

Birth Defects Tracking System

Tracking System

The Birth Defects Program began collaborating with the University of Maine at Orono in 2001 to develop and implement a comprehensive surveillance and tracking system. Today, ChildLINK tracks approximately 13,000 infants born in Maine each year. ChildLINK links birth defect data with multiple data sources that include birth and death certificates, hospital discharge data and metabolic and newborn hearing screening data. ChildLINK maintains security/confidentiality of all records by assigning permission to access the system on an individual basis. Access is monitored by Symantec on a 24/7 basis. By linking information from these existing data sources, ChildLINK can:

- Help assure that children with birth defects and their families receive information about resources and services that may be of assistance and
- Provide valuable public health data to State and national policy makers.

Process Overview

Once a month, hospitals send to the BDP a discharge report reflecting the previous month's birth defects data identified at the hospital. An automatic reminder is sent from ChildLINK to the hospital if the data is not sent in by the fifteenth of the month. When the system receives the report, the report is reviewed for proper formatting and then uploaded to the ChildLINK system. The data is then linked to the birth certificate of the corresponding child.

Statistical Reports

The Birth Defects Program annually submits data for a specific span of years to the U.S. Centers for Disease Control and Prevention, National Birth Defects Prevention Network. The most recent data submitted to the U.S. CDC was 2008 – 2012 which will be available for publication in February 2016. The 24-month lag time in U.S. CDC data is based on the definition of a birth defect. By definition, a birth defect meets criteria and is included in the BDP count if the birth defect is identified within the first year of life. This means that the BDP may still be gathering data as late as December of the year after the birth of the baby. Consequently time is needed to abstract the data necessary to confirm the birth defect.

Birth Defects Counts and Prevalence Rates (2008-2012), Maine				
Condition	Summary	Five year count	Prevalence per 10,000 Live Births	Notes
Anencephalus	One in 4,4915 live births	13	2.0	
Cleft lip with and without cleft palate	One in 1,521 live births	42	6.6	
Cleft palate without cleft lip	One in 1,486 live births	43	6.7	
Coarctation of aorta	One in 2,061 live births	31	4.9	
Common truncus	One in 12,779 live births	5	0.8	
Down syndrome (Trisomy 21)	One in 743 live births	86	13.5	
Encephalocele	One in 21,299 live births	3	0.5	
Gastroschisis	One in 1,452 live births	44	6.9	1
Hypoplastic left heart syndrome	One in 2,778 live births	23	3.6	
Hypospadias*	One in 161 live births	204	62.2	
Omphalocele	One in 5,809 live births	11	1.7	2
Pulmonary valve atresia	One in 12,779 live births	5	0.8	
Limb deficiencies (Reduction defects)	One in 3,550 live births	18	2.8	
Spina bifida without anencephalus	One in 2,778 live births	23	3.6	
Tetralogy of Fallot	One in 2,556 live births	25	3.9	3
Transposition of great arteries – All	One in 2,904 live births	22	3.4	
dextro-Transpositon of great arteries (d-TGA)	One in 2,904 live births	22	3.4	
Tricuspid valve atresia and stenosis	One in 10,650 live births	6	0.9	
Total Live Births 2008-2012		*63,897*		
Total Male Live Births (2008-2012)		*32,786*		

Notes

1. **Gastrochisis is coded 756.73. Cases are also abstracted to determine diagnosis**
2. **Omphalocele is coded 756.72. Cases are also abstracted to determine diagnosis**
3. **Includes pulmonary atresia with septal defect**

***Hypospadias: prevalence per 10,000 male live births; Total Male live Births are provided for Hypospadias rates.**
***Each year approximately 12,800 babies are born in Maine.**

General comments
Case-finding is limited to babies born in Maine to Maine residents and to birth defects identified in the first year of life.
A baby born with multiple conditions, adding up the number of defects will not yield the number of babies born with defects. In addition, this list represents only a portion of reportable birth defects collected and tracked in Maine.
National data can be found at the following link: http://www.nbdpn.org/annual_reports.php

Public Awareness

Education and Informational Materials

The Birth Defects Program has a range of materials including brochures to send to interested parties and stakeholders that include prevention information regarding folic acid use, information on the various birth defects and other topics of interest regarding birth defects. The materials can be accessed at the Maine DHHS CDC website at the following internet address: http://www.maine.gov/dhhs/mecdc/population-health/cshn/birth-defects/index.html

Calendar Year 2015 Program Accomplishments and 2016 Future Direction

Accomplishments

- As of December 31, 2015 it is estimated that 78 infants with a confirmed birth defect will have been referred to the Maine Department of Education – Part C Agency (CDS).
- Program met regularly with abstractors to discuss ways to make the abstraction process more complete. This dialogue helped to clarify questions on the abstraction form and a fostered a more clear understanding of the birth defects criteria among the abstractors. Regular meetings also allowed time to discuss other issues related to birth defects.
- Program visited birth hospitals and shared information regarding birth defects, prevention, and the Birth Defects Program in collaboration with the Maine CDC Cleft Lip and Palate Program director.
- Program used social media and other options to raise awareness regarding prevention activities and birth defects to stakeholders, families and other interested parties.
- Program began discussions with hospital administrators to allow birth defects abstractors to abstract individual medical records remotely. Currently one hospital (Miles Memorial Hospital) allows for remote access.
- The completed critical congenital heart defect project is looking at risk factors in diagnosing a CCHD with the New England Birth Defects Consortium. A written report will be made available once the consortium work is completed.

Future Direction

- Work collaboratively with the other New England states on projects of mutual interest that will allow for increased understanding of the full impact of birth defects on Maine children and their families as well throughout New England. Due to the small numbers of births in Maine and other New England states working together provides for a greater number of cases and, consequently, more accurate data.
- Explore increasingly effective ways to share prevention information with stakeholders and interested parties using data collected through the abstraction process.

- Continue to work with the Maine CDC Environmental Health Tracking Unit to expand the number of birth defects being reported on the portal.
- Update the Birth Defect Program Rules in FY 2016 to include seven critical congenital heart defects.
- Begin outreach education with midwives regarding the rules for critical congenital heart defects adopted in the summer of 2015.
- Begin a pilot project with an initial group of birth hospitals to make sure the electronic submission of data to the ChildLINK database is working without problems.

References

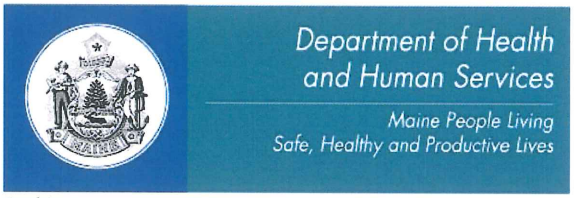
[1] Trust for America's Health "Birth defects and Developmental Disabilities: A Major Public Health Challenge"
[2] http://www.cdc.gov/node.do/id0900f38000dffe

Appendix A

Reportable Birth Defects Included in Case Definition for 2015

Birth Defect	ICD-9-CM Codes
Central Nervous System	
Anencephalus	740.0 – 740.1
Spina Bifida without anencephalus	741.0, 741.9 w/o 740.0 – 740.10
Hydrocephalus without spina bifida	742.3 w/o 741.0, 741.9
Encephalocele	742.0
Microcephalus	742.1
Eye	
Anophthalmia/microphthalmia	743.0, 743.1
Congenital cataract	743.30 – 743.34
Aniridia	743.45
Ear	
Anotia/microtia	744.01, 744.23
Cardiovascular	
Common truncus	745.0
Transposition of great arteries	745.10, 745.11, 745.12, 745.19
Tetralogy of Fallot	745.2
Ventricular septal defect	745.4
Atrial septal defect	745.5
Endocardial cushion defect	745.60,745.61, 745.69
Pulmonary valve atresia and stenosis	746.01, 746.02
Tricuspid valve atresia and stenosis	746.1
Ebstein's anomaly	746.2
Aortic valve stenosis	746.3
Hypoplastic left heart syndrome	746.7
Patent ductus arteriosus	747.7
Coarctation of aorta	747.10
Orofacial	
Cleft palate without cleft lip	749.0
Cleft lip with and without cleft palate	749.1, 749.2
Choanal atresia	748.0
Gastrointestinal	
Esophageal atresia/tracheoesophageal fistula	750.3
Rectal and large intestinal atresia/stenosis	751.2
Pyloric stenosis	750.5
Hirshsprung's disease (congenital megacolon)	751.3
Biliary atresia	751.61
Genitourinary	
Renal agenesis/hypoplasia	753.0
Bladder exstrophy	753.5

Birth Defect	ICD-9-CM Codes
Obstructive genitourinary defect	753.2, 753.6
Hypospadias and Epispadias	752.61, 752.62
Musculoskeletal	
Reduction deformity, upper limbs	755.20 – 755.29
Reduction deformity, lower limbs	755.30 – 755.39
Gastroschisis	756.79
Omphalocele	756.79
Congenital hip dislocation	754.30, 754.31, 754.35
Diaphragmatic hernia	756.6
Chromosomal	
Trisomy 13	758.1
Down syndrome (Trisomy 21)	758.0
Trisomy 18	758.2
Other	
Fetal alcohol syndrome	760.71
Amniotic bands	No code



Paul R. LePage, Governor Mary C. Mayhew, Commissioner

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