Department of Health and Human Services
Maine People Living
Safe, Healthy and Productive Lives

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January 30, 2013

Senator Margaret M. Craven, Chair
Representative Richard R. Farnsworth, Chair
Members, Joint Standing Committee on Health and Human Services
#100 State House Station
Augusta, Maine 04333-0100

Dear Senator Craven, Representative Farnsworth and Members of the Joint Standing Committee on Health and Human Services:

Attached is the 2012 Annual Report to the Legislature for the Maine 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 Maine CDC Birth Defects Program's activities and accomplishments in 2012 as well as planned activities for 2013.

Thank you for the opportunity to provide the Joint Committee on Health and Human Services with a report on the activities and accomplishments of the Maine CDC Birth Defects Program.

Sincerely,

Mary C. Mayhew
Commissioner

MCM/klv

Attachment

Maine CDC Birth Defects Program

January 1, 2012 - December 31, 2012

Submitted to the Joint Standing Committee on Health and Human Services

2012 Annual Report

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EXECUTIVE SUMMARY

Background

Effective May 1999, State of Maine statutory language (Title 22: Health and Welfare: Subtitle: Facilities for Children and Adults; Chapter 1687; Birth effects Program; §8941-§13375) authorized the Department of Health and Human Services to collect information on birth defects in Maine. The Maine CDC Birth Defects Program was established within the Department of Health and Human Services, Maine Center for Disease Control and Prevention, Children with Special Health Needs Program. The program started collecting data in 2003.

Purpose

The overall purpose of the Maine CDC Birth Defects Program (MBDP), using a public health approach, is 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; refer the infant with a confirmed birth defect to the Department of Education Part C – Children's Developmental Services (CDS) Program; 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 2012 annual report summarizes the current activities of the MBDP as well as the ongoing and upcoming activities for calendar year 2013. It also shows selected birth defects counts and birth prevalence for the years 2004 – 2008.
Some of the activities MBDP undertook in 2012:

- Contact families with a baby with a confirmed birth defect by letter to offer support and information regarding birth defects.
- Follow-up with families via telephone call to ensure families had an opportunity to have their questions or concerns addressed.
- Refer babies with a confirmed birth defect to the Part C Agency (CDS).
- Meet regularly with the Maine CDC Environmental Public Health Tracking Unit to discuss specific issues relating to the Environmental Tracking Portal and birth defects.
- Meet regularly with the 2 abstractors that work with the MBDP to develop skills and share information pertinent to birth defects and the abstraction process.
- Provide educational materials regarding the prevention of birth defects to parents, health providers and other interested parties.
- Provide educational presentations to Hospital staff as well as other interested parties including parents regarding the MBDP and prevention activities.
- Meet with the advisory board once to discuss issues relating to the MBDP.
- Work in collaboration with the New Hampshire Birth Conditions Program on a research project looking at the relationship between arsenic and birth defects in the two states.
- Regular discussions with the epidemiology staff related to birth defects data.

- Currently working on updating the abstraction process and discussing ways to present data collected.
- Attended the annual National Birth Defects Prevention Network Meeting.
- Meet regularly with ChildLINK staff to discuss the database system and work to expand and increase its use.

For more information on the Maine CDC Birth Defects Program:
Contact Diane Haberman, Program Coordinator, daine.haberman@maine.gov or 207-287-8424
http://www.mainepublichealth.gov/BirthDefects

Program Overview

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% 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 the surveillance program to accurately quantify morbidity and mortality, detect temporal trends, and assess the financial burden that birth defects may cause. Many children who survive have a lifetime of major expenses. In addition, specialty medical care, special education, rehabilitation and developmental services are essential.

The Maine CDC Birth Defects Program (Maine CDC 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 MBDP 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 Maine CDC 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 CDC Birth Defects Program 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, establishes the Maine CDC Birth Defects Program (Maine CDC 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.

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

May 2008 – Rules were updated to include 3 more reportable birth defects.

April 2011 – Rules were amended to include the 45 birth defects recommended by the CDC and the National Birth Defects Prevention Network. Referral to the Part C Agency (CDS) was also included.

Stakeholders

The following is a brief listing of organizations that have a strong association with the Maine CDC Birth Defects Program. In addition, at the national level, the March of Dimes and the national Birth Defects Prevention Network are key 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 ○ Specialists physicians ○ Genetic Counselors	• **Other State Programs** ○ Maine CDC Newborn Hearing Screening Program ○ Maine CDC Newborn Bloodspot Screening Program ○ Maine CDC Environmental Health Tracking Unit ○ Maine CDC Data, Research and Vital Statistics ○ Office of MaineCare

State Advisory Committee

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

Summary of Activities

The Committee met once this past year. Items discussed included the Maine CDC BDP's participation with the New Hampshire Birth Conditions Program in investigating the possible link of arsenic and the development of birth defects, referral of children confirmed with a birth defect to CDS, and contacting families by letter and telephone by the Maine CDC BDP. Other topics discussed included other ways to reach out to families and other interested parties to spread the word about prevention, Critical Congenital Heart Defect (CCHD) and what is being done to identify and follow-up within New England and Maine, abstracting all 45 birth defects on the NBDPN/CDC list and updating the Birth Defects Website. The State Advisory Committee will start meeting quarterly.

Maine CDC Birth Defects Program Summary

Program Description

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

The Maine CDC Birth Defects Program began passive case ascertainment with confirmation 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. 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 ICD-9 codes. Once a case is identified as a possible reportable birth defect, the case is assigned to an abstractor. The abstractor goes to the hospital to review records to confirm the presents of a birth defect; active case ascertainment.

The Maine CDC Birth Defects Program 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 Maine CDC BDP receives medical discharge data electronically from 28 of the 30 birth hospitals. Maine CDC BDP is currently working with 1 of the remaining hospitals to submit data electronically. The hospital is having difficulty with its computer system and the Maine CDC BDP will continue to work with them to make electronic data submission possible. The remaining hospital has a policy that it will not send protected health information electronically and submits it data in hardcopy. To ensure compliance, the Maine CDC BDP sends each hospital a monthly electronic reminder to submit data.

In order for a case to be considered by the Maine CDC BDP it must meet 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,
- ◊ The birth must occur in Maine and the mother must be a Maine resident,
- ◊ The diagnosis was made before the infant reached 1-year of age, and
- ◊ The 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 record 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; father's history when available; and mother's exposure to illegal drugs, medications, smoking, and 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 link existing state information systems with data obtained from hospitals, health care providers and others mandated to report. Once a child is confirmed to have a birth defect the family is notified by mail of services available to them.

Currently, the Maine CDC Birth Defects Program reports and gathers information on 45 birth defects, see Appendix A for a complete listing of reportable birth defects. These 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 detected and reported and the ability of the abstractors to gather the necessary information from the birth hospital. Some birth defects do not manifest right away. There may also be a lag time in verifying a birth defect because, by law, a birth defect can be diagnosed within the first year of life.

Funding Sources

The Maine CDC Birth Defects Coordinator position is funded through the Maternal and Child Health Block Grant. Abstractor positions are funded in part by a U.S. Center for Disease Control and Prevention Environmental Health Tracking Grant and the State's General Fund.

Personnel

The Maine Birth Defects Program consists of one full-time Coordinator and two part-time contractors. The MBDP contracts with Maine Medical Center and a private individual for part-time abstraction services.

Goals, Activities and Achievements

The Maine CDC 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 regards to the following: causes, risk factors and strategies for prevention and the provision of services,
- ◊ Establish a system for data collection that identify prevalence and incidence rates by region and population group and identify the morbidity and mortality rates resulting from birth defects, and
- ◊ Contact families to provide information about available resources and services,

◊ Conduct investigations to determine the nature, and extent of the disease or known or suspected causes of birth defects. Examples of investigation include:

- Maine collects data on 45 birth defects and collects further information through the abstraction process on 22 of those birth defects. Maine CDC BDP is in the process of expanding the abstraction process to include all 45 conditions. The Maine CDC BDP submits data annually to the National Birth Defects Prevention Network and U.S. CDC.
- The Maine CDC BDP works in collaboration with the Maine CDC Environmental Health Tracking Unit to show 12 of the 22 birth defects on the Environmental Health Tracking Portal.
- The Maine CDC BDP connects with a family once a baby is identified with a confirmed birth defect with an initial letter. The family is then contacted by phone to follow-up with the family to identify any unmet needs and to connect them to the resources that they may need. The baby identified with a confirmed birth defect is also referred to the Department of Education Part C Agency (CDS) for follow-up for early intervention services.
- The Maine CDC BDP is currently working with the New Hampshire Birth Conditions Program in investigating the effects of arsenic on the development of birth defects.

Maine CDC Birth Defects Tracking System

Tracking System

Maine CDC DBP 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 date, metabolic screen data, and newborn hearing screening data. By linking information from these existing data sources ChildLINK can: 1) help ensure that children with birth defects and their families receive information about resources and services that may be of assistance; and, 2) provide valuable public health data to state and national policy makers. 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.

Process Overview

Once a month, hospitals send in a discharge report reflecting the previous month's birth defects data identified at the hospital. If the data is not sent in by the 15th of the month, an automatic reminder is sent from ChildLINK to the hospital's contact person. Information gathered is put into an excel format and then submitted electronically to the ChildLINK database system. Once 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

Birth Defects data is submitted annually to the National Birth Defects Prevention Network and U.S. CDC. The most recent data years are below. At the end of the calendar year 2012, data will be available covering 2004 – 2009. The 15-month lag time in data being available is based on the definition of a birth defect. By definition, a birth defect meets criteria and is included in the Maine CDC Birth Defects count if the birth defect is identified within the first year of life. This means that the Maine CDC BDP may still be gathering data as late as December of the year after the requested year. Consequently time is needed to abstract the data necessary to confirm the birth defect. The 2011 birth defects data will not be available until March 2013.

Secondly, the U.S. CDC requests data for a specific span of years and once requested, the data is not published until the end of the request year. The 2010 birth defects data was recently sent to the U.S. CDC.

Maine

Birth Defects Counts and Prevalence 2004-2008 (Prevalence per 10,000 Live Births)

Defect	Maternal Race/Ethnicity						
	Non-Hispanic White	Non-Hispanic Black or African	Hispanic	Asian or Pacific Islander	American Indian or Alaskan Native	Total**	Notes (+)
Anencephalus	6 *0.94*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	8 *1.17*	
Cleft lip with and without cleft palate	55 *8.61*	0 *0.00*	0 *0.00*	1 *9.13*	1 *18.15*	59 *8.66*	
Cleft palate without cleft lip	62 *9.70*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	62 *9.10*	
Coarctation of aorta	31 *4.85*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	31 *4.55*	
Common truncus	5 *0.78*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	5 *0.73*	
Down syndrome (Trisomy 21)	79 *12.37*	1 *7.03*	1 *10.22*	0 *0.00*	1 *18.15*	88 *12.91*	
Encephalocele	3 *0.47*	0 *0.00*	1 *10.22*	0 *0.00*	0 *0.00*	4 *0.49*	
Gastroschisis	36 *5.63*	0 *0.00*	0 *0.00*	1 *9.13*	0 *0.00*	39 *5.72*	+
Hypoplastic left heart syndrome	19 *2.97*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	20 *2.936*	
Hypospadias*	26 *7.92*	1 *13.55*	1 *19.31*	0 *0.00*	0 *0.00*	28 *7.98*	+
Omphalocele	16 *2.50*	0 *0.00*	0 *0.00*	1 *9.13*	0 *0.00*	19 *2.79*	+
Pulmonary valve atresia and stenosis	3 *0.47*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	3 *0.44*	+
Reduction deformity, lower limbs	1 *0.16*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	1 *0.15*	
Spina bifida without anencephalus	23 *3.60*	1 *7.03*	0 *0.00*	0 *0.00*	0 *0.00*	24 *3.52*	
Tetralogy of Fallot	21 *3.29*	0 *0.00*	1 *10.22*	0 *0.00*	0 *0.00*	23 *3.37*	+
Transposition of great arteries	35 *5.48*	1 *7.03*	0 *0.00*	0 *0.00*	1 *18.15*	37 *5.43*	+
Tricuspid valve atresia and stenosis	2 *0.31*	0 *0.00*	0 *0.00*	0 *0.00*	0 *0.00*	2 *0.29*	+
Total Live Births	**63887**	**1423**	**978**	**1095**	**551**	**68158**	
Total Male Live Births	**32849**	**738**	**518**	**566**	**291**	**32049**	+

*Hypospadias: prevalence per 10,000 male live births **Total includes unknown race

Maine

Trisomy Counts and Prevalence by Maternal Age 2004-2008 (Prevalence per 10,000 Live Births)

Defect	Age		
	<35	35 and >	Total**
Down syndrome (Trisomy 21)	46 *7.85*	42 *44.04*	88 *12.92*
Total Live Births	**58613**	**9537**	**68126**

**Total includes unknown age

Notes(+)

1. Hypospadias: Aggregate and Total Male Live Births only include 2008.
2. Omphalocele is coded 756.72 and Gastroschisis is coded 756.73. Cases are also abstracted to determine diagnosis.
3. Pulmonary valve atresia and stenosis: Aggregate only includes Pulmonary Valve Atresia.
4. Tetralogy of Fallot: Includes pulmonary atresia with septal defect.
5. Transposition of great arteries: We have been including 745.130, 745.140, 745.150, 745.180 through 2011.
6. Tricuspid valve atresia and stenosis: We are not including 746.106 or 746.105.
7. Unless otherwise noted, birth defect counts include live births plus stillbirths.
8. Casefinding is limited to babies born in Maine to Maine residents.
9. Race/ethnicity is based on maternal race and ancestry fields on the birth certificate.
10. Total live birth counts and male live birth counts are of live births born in Maine to Maine residents.

Public Awareness

Education and Informational Materials
The Maine CDC Birth Defects Program has a range of materials including brochures that are sent to interested parties and stakeholders; including prevention information regarding folic acid use, information on the various birth defects and other topics of interest regarding birth defects.

Website
http://www.maine.gov/dhhs/boh/cshn/birth_defects/index.html

Maine CDC Birth Defects Program
Accomplishments and Future Direction

Accomplishments

- Reduced backlog of families to be contacted by phone after an initial letter was sent to them to offer support.
- Continued arsenic and birth defects project with the New Hampshire's Birth Condition Program.
- Referred 43 babies with confirmed birth defects to the Department of Education - Part C Agency (CDS)
- Continued to meet regularly with abstractors to discuss ways to make the abstraction process more complete and to discuss other issues related to birth defects.
- Continued to educate the provider community by going to birth hospitals and sharing information regarding birth defects, prevention and the Maine CDC Birth Defects Program in collaboration with the Maine CDC Cleft Lip and Palate Program Director.
- Continued to use social media and other options to get information out regarding prevention activities and birth defects to stakeholders, families and other interested parties.

Future Direction

- Continue to work on expanding the ability to follow-up with families to offer support by phone on a timely basis.
- Continue to work in collaboration with the other New England States on projects of mutual interest.
- Expand the number of birth defect cases being abstracted to include all 45 birth defects.
- Continue to explore ways to share information with stakeholders and interested parties in regards to the information gathered through the abstraction process including risk factors.
- Continue to explore ways to share prevention information with stakeholders and interested parties.
- Continue to work with the Maine CDC Environmental Health Tracking Unit to expand the number of birth defects being reported on the portal.
- Expand the number of times the Maine CDC Birth Defects Advisory Committee meets from one time a year to three to four times a year.