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Maine Birth Defects Program

2008 Annual Report

Department of Health and Human Services
Maine Center for Disease Control and Prevention
Division of Family Health
Children with Special Health Needs

Forward

In May 1999, the 119th Maine State Legislature enacted Public Law 1322 establishing the Maine Birth Defects Program (MBDP) within the Department of Health and Human Services, Maine Center for Disease Control and Prevention, Division of Family Health. The legislation allows the MBDP to collect, analyze and distribute information on causes, risk factors, and strategies for prevention, and to maintain a central registry for cases of birth defects. Legislation also requires reporting from those licensed under Title 22 (hospitals) and Title 32 (Licensed Professionals) to make available health records and information relating to the occurrence of birth defects. In April 2003, the Department of Health and Human Services, Maine Birth Defects Program adopted formal rules outlining reporting responsibilities and access to medical records. Mandated reporting began May 1, 2003.

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 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 that birth defects may cause. Many children who survive have a lifetime of major expenses. In addition specialty medical care, special education,

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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/id/0900f3ec8000dffe

rehabilitation and developmental services are essential. Millions of dollars are spent each year for the care of babies born with birth defects. In 1992, it is estimated that the total lifetime cost for caring for children with birth defects in the United States exceeded $ 1.4 billion dollars annually. Many of the known birth defects are preventable or outcomes can be improved through early identification, treatment and services.

The Maine Birth Defects Program

The MBDP is committed to fulfilling our 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 goal of the MBDP is to reduce the prevalence, mortality, and morbidity of birth defects in Maine through a comprehensive surveillance system tied to an extensive referral and service system for children and families impacted by birth defects.

The MBDP is a comprehensive surveillance system that benefits the citizens of Maine through the early identification of 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 MBDP to take the lead in the integration of child health data across programs, 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 the mortality and morbidity among children identified with birth defects.

The current list of identifiable birth defects was developed by key partners that included: genetic providers, neonatologists, pediatric cardiologist and surgeons. The following reportable conditions were selected because these major defects require immediate specialty care, are evident in the first year of life, span a wide range of severity, etiology and treatment, and most are among the leading cause of infant and fetal deaths in Maine. The reportable conditions include: Anencephaly, Spina Bifida, Encephalocele, Transposition of the Great Vessels, Double Outlet Right Ventricle, Tetralogy of Fallot, Single Ventricle, Tricuspid Atresia, Hypoplastic Left Heart, Coarctation of the Aorta, Interrupted Aortic Arch, Cleft Lip, Cleft Palate with Cleft Lip, Gastroschisis, Down Syndrome, Omphalocele and Truncus Arteriosis.

Rules that outline reporting responsibilities and access to medical records were formally adopted in April 2003 and mandated reporting began May 1, 2003. As adopted, hospitals and providers who diagnose birth defects in infants and fetuses are required to report to the MBDP "either within 24 hours of the time the infant is discharged or transferred, or within 7 days of the diagnosis or confirmation of a reportable defect". Additionally, rules require hospitals to identify a contact person in the Neonatal Intensive Care Nursery (NICU), Newborn Nursery (NBN), and Medical Records Departments to facilitate reporting and access to medical records. These hospital-based contacts provide additional information when a case report is received, for example to provide missing demographic data.

Administration of the MBDP

The Maine Birth Defects Program (MBDP) is housed with the Maine Department of Health and Human Services, Maine Center for Disease Control and Prevention, Division of Family Health, Children with Special Health Needs/Genetics Program.

Staffing and Funding

The MBDP staff consists of a half-time Birth Defects Screening Coordinator. Program staff, maintenance of the tracking database ChildLINK and other activities are currently funded through the Federal Maternal and Child Health Block Grant.

Reporting and Tracking System

The MBDP uses a combination of both passive and active surveillance. Active surveillance by definition is "a surveillance approach to case identification that is based on surveillance staff being engaged intensively in all activities related to finding and confirming potential birth defects cases." Passive surveillance by definition is "a surveillance approach to case identification whereby birth defects program receive case reports from data sources." [3] For a case to be included in the MBDP, 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

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[3] National Birth Defects Prevention Network June 2004 *Guidelines for Conducting Birth Defects Surveillance"*, Chapter 6, Section 2.

- 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 MBDP list of reportable defects.

Reportable Birth Defects

The MBDP recently completed analyzing data from May 1, 2003 – December 31, 2006. During that period there were 52,210 births, of those 197 infants had a confirmed birth defect. It should be noted that an infant or fetus can have multiple birth defects and each is counted as a separate individual infant with confirmed birth defects.

Table 1: Confirmed Birth Defects in Maine – May 1, 2003 – December 31, 2006

Reportable Birth Defect	**Count**
Anencephaly	6
Cleft Lip	8
Cleft Palate	34
Cleft Palate with Cleft Lip	25
Coarctation of Aorta	13
Double Outlet Right Ventricle	6
Down Syndrome	43
Gastroschisis	19
Hypoplastic Left Heart Syndrome	13
Interrupted Aorta Arch	2
Omphalocele	11
Other	28
Single Ventricle	1
Spina Bifida	9
Tetralogy of Fallot	14
Transposition of Great Vessels	16
Total	**197**

With Maine’s relatively low birth rate (approximately 14,000 births/year), it may take several years of data gathering to determine the occurrence rates of selected birth defects. The MBDP will use the data to provide information to the public and health care professionals about factors that may reduce or increase a woman’s chance of having a baby born with a birth defect.

Activities for CY2007

- Hired ½ time Birth Defects Coordinator

- Notified birthing facilities of the new Birth Defects Coordinator to re-establish formal collaborations
- Submitted data to National Birth Defects and Prevention Network at Center for Disease Control Atlanta
- Confirmed birth defects from 2005 and 2006.

Activities for CY2008

- Continue to work towards electronic submission of all hospital discharge data.
- Submit rule change to include 3 additional reportable birth defects – Reduction Deformity – Upper Limb, Reduction Deformity – Lower Limb, Trisomy 13, and Trisomy 18.
- Provide technical assistance to Maine Medical Center's abstraction specialist
- Attend national Birth Defects Conference in Washington, DC
- Apply for CDC birth defects grant if appropriate.
- Analyze and confirm birth defects from 2007

Summary

The MBDP's comprehensive surveillance system benefits the citizens of Maine through early identification of birth defects. Early identification of birth defects has the potential to assure that infants are connected to appropriate health care services that provide families with resources that can prepare for their child's future.

A long-term goal of the MBDP is to monitor trends in birth defects, to participate fully in epidemiological investigations as a means of informing public policy, and to assess for timely referrals and follow-up care to reduce mortality and morbidity among children identified with birth defects. With Maine's relatively low birth rate (approximately 14,000 births/year), it may take several years of data gathering to determine the occurrence rates of selected birth defects.