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

Department of Health and Human Services
Commissioner's Office
221 State Street
11 State House Station
Augusta, Maine 04333-0011
Tel.: (207) 287-3707; Fax (207) 287-3005
TTY Users: Dial 711 (Maine Relay)

January 30, 2015

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

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

Attached is the 2014 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 2014 as well as planned activities for 2015.

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.

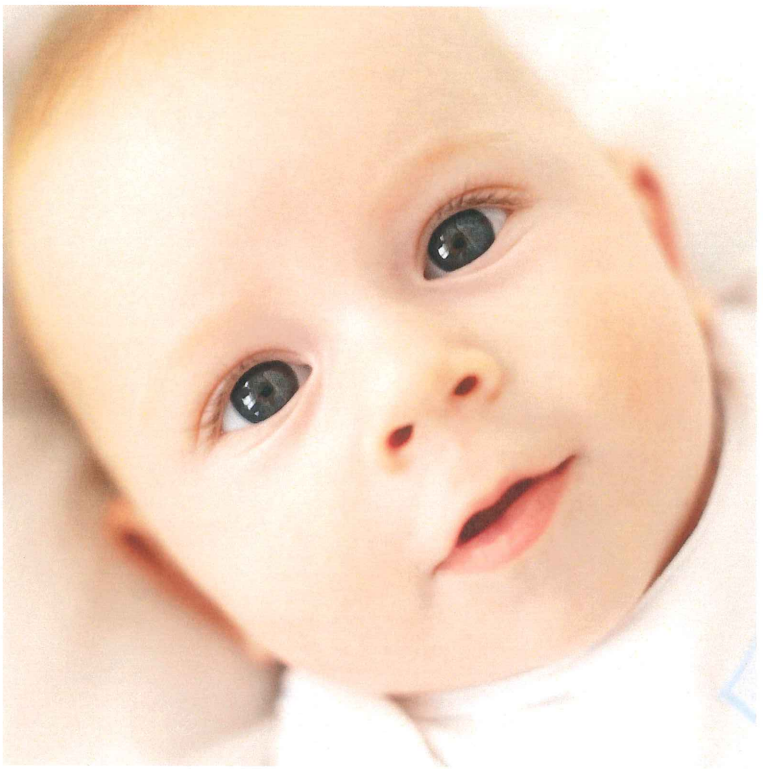
Sincerely,

Mary C. Mayhew
Commissioner

MCM/klv

Attachment

Maine CDC Birth Defects Program



January 1, 2014 - December 31, 2014

Submitted to the Joint Standing Committee on Health and Human Services

2014 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

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EXECUTIVE SUMMARY
December 2014

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, the Maine Center for Disease Control and Prevention.

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 2014 annual report summarizes the current activities of the Maine CDC Birth Defects Program (BDP), the ongoing and upcoming activities for calendar year 2015 and shows selected birth defects counts and birth prevalence for the years 2006 – 2010. In 2014, the Maine CDC BDP:

- Hired a new abstractor for the northern Maine allowing the Maine CDC Birth Defects Program to continue to abstract and confirm birth defects in the northern half of the state.
- Contracted 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 (CDS) – 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 and share information pertinent to birth defects and the abstraction process.
- Provided educational materials about the prevention of birth defects to parents, health providers and other interested parties.

- Provided educational presentations about the Maine CDC Birth Defects and Cleft lip and Palate Programs to staff at two hospitals.
- Met with the Maine CDC Birth Defects Advisory Committee four times to discuss relevant birth defect program issues.
- 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 the Region I regarding risk factors related to critical congenital heart defects (CCHD).*
- Completed, in partnership with the Maine CDC Division of Population Health Epidemiology staff a quality improvement project to review data elements currently being abstracted by Maine CDC BDP staff and to assess the accuracy of the abstraction process including re-abstraction of the data and follow up.
- Participated in the annual National Birth Defects Prevention Network Meeting which occurred in a virtual format this year.
- 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, 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 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 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 Maine CDC 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 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 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 Maine CDC BDP within the Maine Department of Health and Human Services. Program rules were formally

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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

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 (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. 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**
	  ○ 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 Services

Maine CDC 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 Maine CDC BDP on development, implementation and evaluation of program policies, procedures and activities.

Summary of Activities

The MECDC Birth Defects Committee met four times this past year. Items discussed included: a) the quality improvement project being worked on by the Maine CDC BDP and the Maine CDC epidemiology staff regarding the abstraction process; b) ways to reach out to families and other interested parties to spread the word about birth defects prevention; c) reaching out to parents at the birth hospital with information and support; and d) Critical Congenital Heart Defects (CCHD) and what is being done to identify and follow up within New England and Maine.

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 U.S. Centers for Disease Control and Prevention (CDC). The Maine CDC Birth Defects Program was established within the Maine CDC to identify newborns with birth defects, ensure that they receive appropriate specialty services and to monitor birth defect trends.

As a surveillance unit, the Maine CDC 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 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 all birthing hospitals at this time. Maine CDC BDP sends all hospitals a monthly electronic reminder to submit data.

In order for a case to be considered by the Maine CDC 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 and is housed at the University of Maine. 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.

Currently, the Maine CDC Birth Defects Program reports on 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 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 Maine CDC Birth Defects Program consists of one full-time coordinator and two part-time contractors. The Maine CDC BDP contracts with Maine Medical Center and a private individual for part-time abstraction services.

The Maine CDC Birth Defects Coordinator position is funded through the Maternal and Child Health Block Grant. The two abstractor positions are funded by contracts through the State General Fund.

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,
- 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 CDC BDP activity:

- Collection and abstracts of data on 45 birth defects. The Maine CDC BDP submits data annually to the National Birth Defects Prevention Network and U.S. CDC.
- Collaboration with the Maine CDC Environmental 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 Department of Education Part C Agency (CDS) for follow-up for early intervention services.
- Work with the New England Birth Defects Consortium. Currently the Consortium is working on collecting regional data on risks factors in critical congenital heart 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 data and metabolic and newborn hearing screening data. By linking information from these existing data sources ChildLINK can: 1) help assure 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 to the Program 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 15th of the month. This information 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

The Maine CDC 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 Federal CDC was 2006 – 2010 which will be available for publication in February 2015. The 24-month lag time in Federal CDC data is based on the definition of a birth defect. By definition, a birth defect meets criteria and is included in the Maine CDC BDP 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 birth of the baby. Consequently time is needed to abstract the data necessary to confirm the birth defect.

Birth Defects Counts and Prevalence Rates (2006-2010), Maine

Condition	Summary	Five year count	Prevalence per 10,000 Live Births	Notes
Anencephalus	One in 7,401 live births	9	1.4	
Cleft lip with and without cleft palate	One in 1,257 live births	53	8	
Cleft palate without cleft lip	One in 1,514 live births	44	6.6	
Coarctation of aorta	One in 2,220 live births	30	4.5	
Common truncus	One in 60,550 live births	7	1.1	
Down syndrome (Trisomy 21)	One in 822 live births	81	12.2	
Encephalocele	One in 22,202 live births	3	0.5	
Gastroschisis	One in 1,625 live births	41	6.2	1
Hypoplastic left heart syndrome	One in 3,172 live births	21	3.2	
Hypospadias*	One in 160 male live births	125	62.5	2
Omphalocele	One in 4,440 live births	15	2.3	3
Pulmonary valve atresia	One in 8,326 live births	8	0.6	
Reduction deformity, lower limbs	One in 33,103 live births	2	0.5	2
Reduction deformity, upper limbs	One in 22,202 live births	3	0.8	2
Spina bifida without Anencephalus	One in 2,896 live births	23	3.5	
Tetralogy of Fallot	One in 2,896 live births	23	3.5	4
Transposition of great arteries – All	One in 2,081 live births	32	4.8	
dextro-Transpositon of great arteries (d-TGA)	One in 3,172 live births	21	3.2	
Tricuspid valve atresia and stenosis	One in 13,321 live births	5	0.8	
Total Live Births 2006-2010		66,605		
Total Male Live Births (2008-2010)		19,995		

Notes

1. Gastrochisis is coded 756.73. Cases are also abstracted to determine diagnosis
2. Surveillance for this condition began with 2008 births
3. Omphalocele is coded 756.72. Cases are also abstracted to determine diagnosis
4. Includes pulmonary atresia with septal defect

*Hypospadias: prevalence per 10,000 male live births; Total Male live Births are provided for Hypospadias rates.

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 Maine CDC 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 CDC website at the following internet address: http://www.maine.gov/dhhs/mecdc/population-health/cshn/birth-defects/index.html

Maine CDC Birth Defects Program
Calendar Year 2014 Accomplishments and Future Direction

Accomplishments

- In 2013 a total of 43 infants with a confirmed birth defect were referred to the Department of Education - Part C Agency (CDS). Of those, a total of 13 children are currently open with a plan and the remaining 30 children are closed for the following reasons: unable to contact; parent declined services; closed a screening; not eligible for Part C and/or moved out of state.
- As of December 31, 2014 nine infants with a confirmed birth defect were referred to the Department of Education – Part C Agency (CDS). Of those, a total of two are currently open with a plan and the remaining 7 are closed for the following reasons: unable to contact; parent declined; closed a screening, no services required and/or moved out of state.
    - The small number of cases referred is due to the fact that the MECDC Birth Defects Program has not finished abstracting all cases for 2014.
- 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 visiting 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 raise awareness regarding prevention activities and birth defects to stakeholders, families and other interested parties.
- Began discussions with hospital administrators to allow birth defects abstractors to abstract individual medical records remotely. Currently two hospitals (Maine Medical Center and eastern Maine Medical Center) allow remote access to individual medical records.

Future Direction

- Continue to expand 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. The group is currently looking at ways to collect data on risk factors for Critical Congenital Heart Defects (CCHD).
- Continue to explore effective ways to share information with stakeholders and interested parties in regard to the information gathered through the abstraction process including risk factors.
- Continue to explore effective 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. https://gateway.maine.gov/cognos/cgi-bin/cognosisapi.dll?b_action=cognosViewer&ui.action=run&ui.object=%2fcontent%2ffolder[%40name%3d%27CDC%20EOHP%20EPHT%20AVR%27]%2freportView[%40name%3d%27Maine%20Environmental%20Public%20Health%20Tracking%20%28EPHT%29%20Network%20-%20Public%20Data%20Portal%27]&cv.header=false&cv.toolbar=false
- Work with birth hospitals towards allowing abstractors to have remote access to medical records.
- Program rules will be updated to include all 7 Critical Congenital Heart Defects (CCHD)
- Finalize rules for Critical Congenital Heart Defects (CCHD) Rules are currently in draft form and will be going out for public comment soon.

Appendix A
Reportable Birth Defects Included in Case Definition for 2014

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
Transpolition 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 (include only if weight => 2500 grams or note if unable to exclude ,2500 grams infants)	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
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