Louisiana Birth Defects Monitoring Network

2019 Annual Legislative Report

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May 2019

LOUISIANA DEPARTMENT OF HEALTH

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Introduction

Louisiana Birth Defects Monitoring Network (LBDMN) is responsible for the surveillance of birth defects in Louisiana's children. Mandated in 2004, it was the intent of the legislature to "establish a system to collect, analyze, and disseminate data regarding birth defects in the state and to provide information to families of children born with birth defects regarding services available in their community and the development of appropriate prevention programs." (*LA R.S. 40:31.43; and LAC Title 48, Part V, Subpart 55, Chapters 161 & 163 et al.).* See Appendix 1

In addition to fulfilling the annual legislative reporting requirement, the following report details data findings and performance measure assessments for our stakeholders. The categories include:

- Traditional case definition findings from 2013-2015 births;
- Special Project Report: Surveillance of Birth Defects Potentially Associated with Maternal Zika Virus Exposure among 2016-2017 Births; and
- Annual Data Quality Assessment Report Summary for the 2018 calendar year.

Our Mission

The mission of the Louisiana Birth Defects Monitoring Network (LBDMN) is to prevent birth defects and birth defect-related disabilities in Louisiana's children.

What We Do

LBDMN staff conduct active surveillance of birth defects in children born in Louisiana. Monitoring the health status of newborns provides population-based data to help inform policies, educate the public, support efforts of community partners, and prevent new occurrences of birth defects. LBDMN evaluates concerns about unexpected groups of birth defects as well as the effectiveness of preventive interventions. Regionally assigned Data Collection Specialists (DCS) statewide evaluate patient discharge information of newborns until age three years from all birthing hospitals in Louisiana, as well as at Children's Hospital and Tulane University Medical Center in New Orleans. LBDMN maintains a private and confidential database of children affected by congenital structural, functional, and/or genetic birth defects. De-identified medical record data are analyzed statistically for patterns and trends over time.

Who We Serve

LBDMN provides specific birth outcome data for the approximately 63,000 annual births in Louisiana. The program assists:

- Families of infants birth until three years of age with birth defects, by linking them with appropriate medical, educational, public health, and peer support resources;
- Men and women of reproductive age, by providing preventive education regarding birth defects;
- Policy makers, by identifying environmental risk factors and other causes potentially linked to specific birth conditions, and identifying preventive strategies to decrease birth defects;
- Researchers from the Centers for Disease Control and Prevention (CDC), universities, and other states investigating possible causes of specific birth defects.

Total Served

Approximately 1,500 children with specified birth defects are identified annually, which is an average of 300 per 10,000 live births. Since 2005, LBDMN has investigated potential birth defects among 24,737 children (unduplicated) between ages 0-3.

Eligibility Criteria

LBDMN case definition criteria include all of the following:

- The child must have a major structural, functional, or genetic birth defect. Major defects are generally those that can adversely affect the child's health and development. Children who have minor defects posing no significant health or social burdens are excluded.
- The mother's residence at the time of the birth must be the state of Louisiana as determined by the mother's hospital records, or if still in question, by vital records birth registration data.
- Diagnosis of the qualifying condition must be made before the child's third birthday.
- Pregnancy outcomes include only live births with a gestational age at birth of at least 20 weeks. In the absence of an age estimate, the infant must have a birth weight of at least 350 grams.

Services Provided

LBDMN provides:

- Active public health surveillance of hospital discharges of newborns until three years of age for major structural, functional, or genetic birth defects.
- Referral to services for families of children birth until three years of age identified with specified birth defects.
- Prevention of future birth defects through public awareness campaigns in partnership with national, state, and local stakeholders such as CDC, National Birth Defects Prevention Network, Louisiana Chapters of the American Academy of Pediatrics and the American College of Obstetricians and Gynecologists, March of Dimes, regional Families Helping Families, and Spina Bifida of Louisiana. Campaigns include education on the importance of management of chronic conditions such as diabetes and hypertension; folic acid consumption; dangers of fetal alcohol, opioid, and tobacco exposure; infection control to prevent risks of associated birth defects.

Funding Sources

Total Annual Federal Funding State Fiscal Year 2018: $638,000 (Title V MCH Block Grant)

Operations

LBDMN operates within the Louisiana Department of Health, Office of Public Health, Bureau of Family Health as one of four programs serving Children and Youth with Special Health Care Needs and their families.

Advisory Board

As mandated in the establishing law, *LA R.S. 40:31.43*, LBDMN is guided by an advisory board of volunteer stakeholders appointed by the secretary of LDH including the following:

(1) One pediatrician from a list of names submitted by the Louisiana State Medical Society
(2) One board-certified clinical geneticist from a list of names submitted by Ochsner Clinic

(3) One board-certified clinical geneticist from a list of names submitted by Tulane University Medical Center
(4) One board-certified clinical geneticist from a list of names submitted by Louisiana State University Health Sciences Center- New Orleans
(5) One board-certified clinical geneticist from a list of names submitted by Louisiana State University Health Sciences Center- Shreveport
(6) One maternal/fetal medicine physician from a list of names submitted by the March of Dimes
(7) One parent representative from a list of names compiled from various parent groups or by individual application
(8) One consumer representative from a list of names compiled from various consumer groups or by individual application
(9) One epidemiologist employed by or contracted to the department

The role of the LBDMN advisory board as prescribed in the law is *"to make recommendations on the implementation and continuing operation of the surveillance system."* The advisory board meets in person annually. Other contacts throughout the year are via email or teleconference as necessary.

Staff

LDH-OPH Bureau of Family Health contributes staff time to support LBDMN operations, including the Principal Investigator (10%) and direct supervisor of the LBDMN Program Manager; a CDC-funded Senior Epidemiologist (5%); and the LBDMN Staff Epidemiologist (60%). Surveillance staff contracted through the Louisiana Public Health Institute at 100% time commitment to LBDMN include the Program Manager, RN Case Review Clinical Coding Specialist, and six regional DCS. See Appendix 2 for Organizational Chart.

Methodology

LBDMN has conducted birth defects surveillance in Louisiana since 2005 using active case ascertainment methodologies. This means multiple data sources are used to identify potential cases of interest, which may fit within the case definition. Once potential cases are qualified, these sources are reviewed to abstract data, validate abstractions, and track children with birth defects who meet case definition at any time from birth up to their third birthday. Hospital medical records are the primary source for data collection. DCS obtain discharge indices from hospitals to identify potential cases by billing codes (ICD code). Other secondary data sources include Medicaid, Hospital Inpatient Discharge Data (LAHIDD), as well as birth, death, and fetal death record data from the Louisiana Vital Records Electronic Event Registration System (LEERS).

All abstracted data are reviewed for completeness and coding accuracy by a Registered Nurse Case Review Clinical Coding Specialist and/or the LBDMN Program Manager before data are accepted into the Registry and are available for reporting. The surveillance data are stored and managed in a custom web-based database integrated with LEERS birth and death certificates as well as Early Hearing Detection and Intervention (LA-EHDI) data.

For each case, ICD billing codes are converted into CDC clinical coding system, based on the British Pediatric Association and Classification of Diseases and the ICD-9/10-CM, which is used to classify birth defects for data analyses and reporting. Prevalence rate of birth defects is calculated as the number of

children with birth defects per 10,000 total live births. There is an exception for hypospadias and Turner Syndrome, which is limited to males and females respectively and interpreted as the rate among live born males and females, respectively. The 95% confidence interval (CI) is calculated with the assumption that the number of children with birth defects followed a Poisson Distribution. Please refer to Appendix 3 for the Case Ascertainment\Review\Quality Assurance flow chart.

Findings

Not all defects are evident at birth; therefore, LBDMN follows children until three years of age allowing adequate time for proper diagnosis. The data is finalized every 3.5 years to more accurately present the number of birth defects that have occurred within a calendar year.

Traditional Case Definition Findings

The following tables represent data from births in 2013-2015 calendar years. Only live births with birth weight >= 350 grams or gestational age >= 20 weeks were included. The data in this report are limited to children born to Louisiana residents and birth occurrence in state. Of 180,641 children born between 2013 and 2015, 5,016 children were diagnosed with at least one birth defect, yielding an overall prevalence of 277.7 per 10,000 live births or 2.8 % (annual average is 300/10,000). Among children with birth defects, cardiovascular system defects (about 48%) are the most common followed by defects of the genitourinary, musculoskeletal, chromosomal, orofacial, gastrointestinal, central nervous, eye, and ear, face, and neck systems. (Table 1)

Table 1: Distribution (%) of total birth defects by organ and chromosome system among children born with birth defects, 2013-2015 birth cohort (n = 5,016)

Organ and chromosome system	Number	Percent*
Cardiovascular	2,407	48.0
Genitourinary	1,438	28.7
Musculoskeletal	534	10.6
Chromosomal	479	9.5
Oro-facial	298	5.9
Gastrointestinal	277	5.5
Central Nervous	253	5.0
Eye	57	1.1
Ear, Face, Neck	30	0.6

**Because one child may have more than one birth defect, the total percents are greater than 100% when totaled*

The four most common birth defects with a prevalence greater than 10 per 10,000 live births among children born in 2013-2015 included atrial septal defect (81.3), hypospadias (75.8), ventricular septal defect (48.1), and Down Syndrome (12.0). Stratified by organ and chromosome system, the most common birth defects were: cardiovascular: atrial septal defects and ventricular septal defects; genitourinary: hypospadias; central nervous: microcephalus, spina bifida, and hydrocephalus; eye: congenital cataract and anophthalmia/microphthalmia; ear, face, and neck: anotia/microtia; orofacial: cleft lip and cleft palate; gastrointestinal: rectal/large intestinal atresia or stenosis; musculoskeletal: clubfoot, craniosynostosis, and gastroschisis; and chromosomal: Down syndrome (Table 2).

Table 2: Distribution (%) and prevalence (per 10,000 live births) of specific birth defects by organ and chromosome system, 2013-2015 birth cohort (N = 180,641)

System	Birth defects	Number	%	Rate	CI95%
Central Nervous (n = 253)	Microcephalus	109	43.1	6.0	5.0, 7.3
	Spina bifida (without anencephalus)	67	26.5	3.7	2.9, 4.7
	Hydrocephalus (without Spina Bifida)	42	16.6	2.3	1.7, 3.1
	Anencephalus	21	8.3	1.2	0.7, 1.8
	Encephalocele	15	5.9	0.8	0.5, 1.4
Eye (n = 57)	Congenital cataract	33	57.9	1.8	1.3, 2.6
	Anophthalmia/microphthalmia	24	42.1	1.3	0.9, 2.0
Ear, face, neck (n = 30)	Anotia/microtia	26	86.7	1.4	0.9, 2.1
Cardiovascular (n = 2407)	Atrial septal defect	1468	61.0	81.3	77.2, 85.5
	Ventricular septal defect	869	36.1	48.1	45.0, 51.4
	Atrioventricular septal defect (Endocardial cushion defect)	125	5.2	6.9	5.8, 8.2
	Pulmonary valve atresia and stenosis	110	4.6	6.1	5.0, 7.3
	Tetralogy of Fallot (TOF)	100	4.2	5.5	4.5, 6.7
	Coarctation of the aorta	92	3.8	5.1	4.1, 6.2
	Transposition of the great arteries (TGA)	55	2.3	3.0	2.3, 4.0
	Hypoplastic left heart syndrome	47	2.0	2.6	1.9, 3.5
	Dextro-transposition of great arteries	36	1.5	2.0	1.4, 2.8
	Aortic valve stenosis	22	0.9	1.2	0.8, 1.8
	Tricuspid valve atresia and stenosis	14	0.6	0.8	0.4, 1.3
	Total anomalous pulmonary venous connection	6	0.2	0.3	0.1, 0.7
	Ebstein anomaly	14	0.6	0.8	0.4, 1.3
	Common truncus (truncus arteriosus)	7	0.3	0.4	0.2, 0.8
Gastrointestinal (n = 277)	Rectal and large intestinal atresia/stenosis	84	30.3	4.7	3.7, 5.8
	Esophageal atresia/tracheoesophageal fistula	39	14.1	2.2	1.5, 3.0
	Small intestinal atresia/stenosis	42	15.2	2.3	1.7, 3.1
	Biliary atresia	16	5.8	0.9	0.5, 1.4
Oro-facial (n = 298)	Cleft palate without cleft lip	132	44.3	7.3	6.1, 8.7
	Cleft lip with cleft palate	94	31.5	5.2	4.2, 6.4
	Cleft lip without cleft palate	50	16.8	2.8	2.1, 3.6
	Choanal atresia	25	8.4	1.4	0.9, 2.0
Genitourinary (n = 1438)	Hypospadias*	698	48.5	75.8	70.3, 81.7
	Renal agenesis/hypoplasia	72	5.0	4.0	3.1, 5.0
	Congenital Posterior Urethral Valves*	60	4.2	6.5	5.0, 8.4
Musculoskeletal (n = 534)	Clubfoot	160	30.0	8.9	7.5, 10.3
	Craniosynostosis	136	25.5	7.5	6.3, 8.9
	Gastroschisis	76	14.2	4.2	3.3, 5.3
	Diaphragmatic hernia	44	8.2	2.4	1.8, 3.3
	Limb deficiencies (reduction defects)	66	12.4	3.7	2.8, 4.6
	Omphalocele	43	8.1	2.4	1.7, 3.2
Chromosomal (n = 316)	Trisomy 21 (Down Syndrome)	216	45.1	12.0	10.4, 13.7
	Trisomy 18	37	7.7	2.0	1.4, 2.8
	Deletion 22 q11	27	5.6	1.5	1.0, 2.2
	Trisomy 13	16	3.3	0.9	0.5, 1.4
	Turner Syndrome**	14	2.9	1.6	0.9, 2.7

**Prevalence limited to males (92,029); **Prevalence limited to females (88,612)*

Prevalence of specific birth defects by organ and chromosome system and race/ethnicity are presented in Table 3.

Table 3: Prevalence of specific birth defects by organ and chromosome system and race/ethnicity, 2013-2015

Defects	White/Non-Hispanic		Black/Non-Hispanic		Hispanic		Other- Non-Hispanic	
	n	Prevalence, CI95%	n	Prevalence, CI95%	n	Prevalence, CI95%	n	Prevalence, CI95%
Central nervous system								
Anencephalus	11	1.2, 0.6-2.1	9	1.3, 0.6-2.5	-		0	
Spina bifida without anencephalus	38	4.1, 2.9-5.6	25	3.7, 2.4-5.4	-		-	
Hydrocephalus without Spina Bifida	19	2.0, 1.2-3.2	22	3.2, 2.0-4.9	-		0	
Encephalocele	9	1.0, 0.4-1.8	5	0.7, 0.2-1.7	0		-	
Microcephalus	44	4.7, 3.4-6.3	55	8.1, 6.1-10.6	-		6	4.8, 1.8-10.4
Eyes								
Anophthalmia/microphthalmia	12	1.3, 0.7-2.3	10	1.5, 0.7-2.7	0		-	
Congenital cataract	19	2.0, 1.2-3.2	12	1.8, 0.9-3.1	0	-	-	
Ears, face, and, neck								
Anotia/microtia	15	1.6, 0.9-2.7	7	1.0, 0.4-2.1	-		-	
Cardiovascular system								
Transposition of the great arteries (TGA)	26	2.8, 1.8-4.1	19	2.8, 1.7-4.4	9	7.2, 3.3-13.6	-	
Tetralogy of Fallot (TOF)	48	5.2, 3.8-6.8	39	5.8, 4.1-7.9	10	8.0, 3.8-14.7	-	
Ventricular septal defect	460	49.4, 45.0-54.1	297	43.8, 39.0-49.1	72	57.4, 44.9-72.3	40	31.9, 22.8-43.5
Atrial septal defect	668	71.8, 66.4-77.4	664	97.9, 90.6-105.6	79	63.0, 49.9-78.6	57	45.5, 34.4-58.9
Atrioventricular septal defect (Endocardial cushion defect)	60	6.4, 4.9-8.3	53	7.8, 5.9-10.2	7	5.6, 2.2-11.5	5	4.0, 1.3-9.3
Pulmonary valve atresia and stenosis	53	5.7, 4.3-7.4	48	7.1,5.2-9.4	6	4.8,1.8-10.4	-	
Tricuspid valve atresia and stenosis	8	0.9, 0.4-1.7	5	0.7, 0.2-1.7	0		-	
Ebstein anomaly	9	1.0, 0.4-1.8	-		0		-	
Aortic valve stenosis	18	1.9, 1.1-3.1	-		0		0	
Hypoplastic left heart syndrome	23	2.5, 1.6-3.7	22	3.2, 2.0-4.9	-		0	
Coarctation of the aorta	50	5.4,4.0-7.1	31	4.6, 3.1-6.5	7	5.6, 2.2-11.5	-	
Dextro-transposition of great arteries	16	1.7, 1.0-2.8	13	1.9, 1.0-3.3	7	5.6, 2.2-11.5	0	
Pulmonary valve atresia	-		-		0		0	
Orofacial system								
Cleft palate without cleft lip	78	8.4, 6.6-10.5	34	5.0, 3.5-7.0	13	10.4, 5.5-17.7	7	5.6, 2.2-11.5
Choanal atresia	13	1.4, 0.7-2.4	9	1.3, 0.6-2.5	-		-	
Cleft lip without cleft palate	40	4.3, 3.1-5.9	6	0.9, 0.3-1.9	-		-	
Cleft lip with cleft palate	53	5.7, 4.3-7.4	33	4.9, 3.3-6.8	6	4.8, 1.8-10.4	-	
Gastrointestinal system								
Esophageal atresia/tracheoesophageal fistula	18	1.9, 1.1-3.1	12	1.8, 0.9-3.1	-		5	4.0, 1.3-9.3
Rectal and large intestinal atresia/stenosis	42	4.5, 3.3-6.1	34	5.0, 3.5-7.0	5	4.0, 1.3-9.3	-	
Biliary atresia	7	0.8, 0.3-1.5	6	0.9, 0.3-1.9	-		-	
Small intestinal atresia/stenosis	14	1.5, 0.8-2.5	19	2.8, 1.7-4.4	8	6.4, 2.8-12.6	-	
Genitourinary system								
Renal agenesis/hypoplasia	41	4.4, 3.2-6.0	26	3.8, 2.5-5.6	-		-	
Congenital Posterior Urethral Valves	34	7.1, 4.9-9.9	25	7.3, 4.7-10.8	0		-	
Hypospadias	421	88.0, 79.8-96.9	229	67.0, 58.6-76.3	26	40.9, 26.7-60.0	22	59.5, 37.3-90.1
Musculoskeletal system								
Gastroschisis	35	3.8, 2.6-5.2	28	4.1, 2.7-6.0	10	8.0, 3.8-14.7	-	
Omphalocele	19	2.0, 1.2-3.2	21	3.1, 1.9-4.7	0		-	
Diaphragmatic hernia	21	2.3, 1.4-3.4	15	2.2, 1.2-3.6	6	4.8, 1.8-10.4	-	
Limb deficiencies (reduction defects)	37	4.0, 2.8-5.5	22	3.2, 2.0-4.9	-		-	
Craniosynostosis	91	9.8, 7.9-12.0	40	5.9, 4.2-8.0	0		5	4.0, 1.3-9.3
Clubfoot	88	9.5. 7.6-11.6	57	8.4, 6.4-10.9	12	9.6, 4.9-16.7	-	
Chromosomal system								
Trisomy 13	8	0.9, 0.4-1.7	6	0.9, 0.3-1.9	-			
Trisomy 21 (Down Syndrome)	113	12.1, 10.0-14.6	65	9.6, 7.4-12.2	26	20.7, 13.6-30.4	12	9.6, 4.9-16.7
Trisomy 18	18	1.9, 1.1-3.1	15	2.2, 1.2-3.6	-		-	
Turner Syndrome	-		7	2.1, 0.8-4.3	-		-	
Deletion 22 q11	15	1.6, 0.9-2.7	9	1.3, 0.6-2.5	-		0	

Special Project:
Surveillance of Birth Defects Potentially Associated with Maternal Zika Virus Among 2016-2017 Births

In response to the emerging public health concern surrounding Zika virus exposure, in August 2016 LBDMN received CDC grant funding to conduct rapid surveillance of birth defects known to be associated with maternal Zika exposure and referrals to coordinated care for children born with those birth defects.

Funding enabled LBDMN to accomplish infrastructure enhancements in collaboration with the Louisiana Bureau of Vital Statistics and Louisiana Newborn Hearing Screening and Early Hearing Detection and Intervention (LA-EHDI) programs. These improvements include the following:

- Addition of Zika Virus to the list of Congenital Infections as a risk factor for Congenital or Late-Onset Hearing Loss on the LA-EHDI Newborn Hearing Screening Report Form;
- Addition of Zika Virus to the list of Infections Present and/or Treated During This Pregnancy on the Louisiana Birth Certificate application (LEERS);
- Addition of Microcephaly, Other Central Nervous System Anomalies, Microphthalmia, and Arthrogryposis to the list of Congenital Anomalies on LEERS;
- Integration of LA-EHDI and LEERS databases into the LBDMN database for data accessibility. These administrative data sources broaden and speed case ascertainment activity.

Zika Birth Defects Surveillance (ZBDS), through LBDMN, tracked specified birth defects *regardless of viral exposure* in order to determine baseline prevalence rates. Note: The Zika Pregnancy Registry in the Infectious Disease Epidemiology Section within the Bureau of Infectious Diseases tracks cases of mothers and infants with exposure to Zika virus.

Eligibility Criteria/Case Definition:

- This project included births from January 1, 2016-June 30, 2017.
- Pregnancy outcomes included live births and fetal deaths occurring in Louisiana with one of 26 categories of birth defects associated with Congenital Zika Syndrome as specified by the CDC's ZBDS Case Inclusion Guidance.
- Louisiana residents as determined by the mother's hospital records, or if unclear, by vital records birth registration forms.
- Diagnosis of the qualifying condition must have been prior to the child's first birthday.
- Neither the mother nor the baby need to have been tested positive or exposed to Zika virus for inclusion.

Cases were ascertained from the following datasets: Louisiana State Bureau of Vital Statistics records including births, deaths, and fetal deaths, Medicaid, and 49 of 51 eligible reporting hospitals. Among these births and fetal deaths, LBDMN reported 254 qualifying birth defects to CDC (3 fetal deaths and 251 live births) ***none of which had maternal exposure to Zika.***

Table 4: Zika associated birth defects, Louisiana January 2016 - June 2017

Birth defects	Number
Brain abnormalities with and without microcephaly	***141***
Confirmed or possible congenital microcephaly	45
Intracranial calcifications	20
Cerebral/cortical atrophy	14
Abnormal cortical gyral patterns	37
Corpus callosum abnormalities	15
Cerebellar abnormalities	14
Porencephaly	10
Hydranencephaly	14
Ventriculomegaly / hydrocephaly	43
Fetal brain disruption sequence	24
Other major brain abnormalities	72
Neural tube defects (NTDs) and holoprosencephaly	***63***
Anencephaly / Acrania	16
Encephalocele	9
Spina bifida	25
Holoprosencephaly/Arhinencephaly	14
Structural eye abnormalities	***25***
Microphthalmia / Anophthalmia	7
Coloboma	3
Cataract	13
Intraocular calcifications	2
Chorioretinal anomalies involving the macula	2
Optic nerve atrophy, pallor, and other optic nerve abnormalities	4
Congenital contractures and joint	16
Congenital deafness	27

The impact of Zika virus exposure has been low to non-existent in Louisiana resulting in no live births or fetal deaths of children affected by Congenital Zika Syndrome to follow beyond birth. Although rapid surveillance of Zika associated birth defects has ended, LBDMN continues to track a select few of these birth defects as part of its traditional case definition surveillance activities.

Data Quality Assessment Summary for 2018 Calendar Year

CDC monitors national birth defects surveillance through a branch called the National Center for Birth Defects and Developmental Disabilities (NCBDDD). NCBDDD coordinates standards for state birth defects programs through the National Birth Defects Prevention Network (NBDPN). The NBDPN Standards Workgroup produces an annual assessment report summary on Data Quality for population-based birth defects surveillance systems.

Performance standards are used to improve and standardize operations, outcomes, and surveillance functions across state programs, thereby making data comparable at the state, multi-state, and national levels. Eleven data quality measures around completeness, timeliness, and accuracy are associated with three performance levels (1) Rudimentary, (2) Essential, or (3) Optimal.

In 2018, Louisiana met level 1 criteria, rudimentary, in both measures of timeliness. Improving in this quality measure has been the aim of the program since 2015. In order to see consistent improvements, LBDMN has made systemic changes in workflow processes including:

- Implementation of a web-based LEERS integrated database in 2015;
- Adding 3 supplemental datasets for case ascertainment in 2017;
- Securing electronic submission of monthly discharge reports from 49 of 51 eligible reporting facilities in 2017/18;
- Securing remote access for abstracting medical records in 39 reporting facilities in 2017/18;
- Expanding the case definition to include fetal death; and
- Adopting a tiered abstraction strategy approach beginning in 2019 for future data cohorts.

See Appendix 4 for complete 2018 NBDPN Data Quality Assessment Report Summary.

Conclusion

Of 180,641 children born in Louisiana between 2013 and 2015, 5,016 children were diagnosed with at least one birth defect, yielding an overall prevalence of 277.7 per 10,000 live births or 2.8 % (annual average is 300/10,000). Among children with birth defects, cardiovascular system defects (about 48%) are the most common followed by defects of the genitourinary, musculoskeletal, chromosomal, orofacial, gastrointestinal, central nervous, eye, ear, face, and neck systems.

Louisiana Birth Defects Monitoring Network incorporates evidence based best public health surveillance practices including current technology and advanced methodologies to improve systems and data quality to identify, understand, and prevent birth defects and to make referrals to improve quality of life of families in Louisiana.

Section 2 – Appendix

Appendix 1: LA Revised Statute (LA R.S.) 40:31.43; and Louisiana Administrative Code (LAC) Title 48, Part V, Subpart 55, Chapters 161 & 163 et al.

Appendix 2: Office of Public Health Organizational Chart

Appendix 3: Case Ascertainment\Review\Quality Assurance Chart

Appendix 4: 2018 NBDPN Data Quality Assessment Summary

Appendix 5: Birth Defects Codes and Descriptions

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LOUISIANA REVISED STATUTES TITLE 40, PART VII, SECTIONS 31.41–31.48

Part VII. Louisiana Birth Defects Surveillance System

§31.41. Legislative intent

It is the intent of the legislature to establish a system to collect, analyze, and disseminate data regarding birth defects in the state and to provide information to families of children born with birth defects regarding services available in their community and the development of appropriate prevention programs.

Acts 2001, No. 194,§ 1.

§31.42. Definitions

As used in this Part, the following definitions shall apply unless the content clearly states otherwise:

(1) "Advisory board" means the advisory board of the birth defects surveillance system.

(2) "Birth defect" means an abnormality of structure, function, or metabolism that develops during prenatal, perinatal, or early postnatal life that is diagnosed before a child reaches three years of age.

(3) "Department" means the Department of Health and Hospitals.

(4) "Office" means the Office of Public Health within the Department of Health and Hospitals.

(5) "Reporting source" means any physician, nurse, allied health professional, hospital, laboratory, and any other facility or agent which directly or indirectly provides medical services or other health care to a child affected by a birth defect.

(6) "Secretary" means the secretary of the Department of Health and Hospitals.

(7) "Surveillance system" means the process that is used to collect data about children with birth defects.

Acts 2001, No. 194,§ 1.

§31.43. Louisiana Birth Defects Surveillance System

A. The department shall establish a birth defects surveillance system within the Office of Public Health to collect, analyze, interpret, and disseminate data relative to birth defects in Louisiana.

B. In establishing the surveillance system, the department shall require reporting sources to report information on birth defects to the office. However, reporting sources shall not collect or report information on birth defects of a child to the office whenever there is a written objection by the parent or legal guardian that collecting and reporting such information would conflict with their religious tenets or practices.

C. The system has the authority to collaborate with other interstate and interagency efforts as they relate to the surveillance system.

Acts 2001, No. 194,§ 1.

§31.44. Confidentiality

Notwithstanding any other provision of the law to the contrary, individual identifying data in the surveillance system shall be confidential and shall not be subject to discovery. Such data shall not be released unless express written informed consent of a parent or legal guardian has been obtained. Data gathered by the office shall be used only for the purposes set forth in this Part.

Acts 2001, No. 194,§ 1.

§31.45. Report

The department shall produce an annual report on the results obtained through the surveillance system to be submitted to the advisory board, the secretary, and the House and Senate Committees on Health and Welfare.

Acts 2001, No. 194,§ 1.

§31.46. Advisory board

A. The secretary shall establish an advisory board to make recommendations on the implementation and continuing operation of the surveillance system.

B. The secretary shall appoint nine members, each of whom shall have an expressed interest in a birth defects surveillance system, and shall be appointed in the following manner:

(1) One pediatrician from a list of names submitted by the Louisiana State Medical Society.

(2) One board-certified clinical geneticist from a list of names submitted by Ochsner Clinic.

(3) One board-certified clinical geneticist from a list of names submitted by Tulane University Medical Center.

(4) One board-certified clinical geneticist from a list of names submitted by Louisiana State University Health Sciences Center-New Orleans.

(5) One board-certified clinical geneticist from a list of names submitted by Louisiana State University Health Sciences Center - Shreveport.

(6) One maternal/fetal medicine physician from a list of names submitted by the March of Dimes.

(7) One parent representative from a list of names compiled from various parent groups or by individual application.

(8) One consumer representative from a list of names compiled from various consumer groups or by individual application.

(9) One epidemiologist employed by or contracted to the department.

C. Each member shall serve at the pleasure of the secretary.

D. Vacancies shall be filled in the manner of the original appointment.

E. The members of the advisory board shall serve on a voluntary basis and shall receive no compensation for their services.

F. The members of the advisory board shall elect from their membership a chairman and a vice chairman.

Acts 2001, No. 194,§ 1.

§31.47. Cooperation by other state entities

All departments, commissions, boards, agencies, officers, and institutions of the state and all subdivisions thereof shall cooperate with the office in carrying out the purposes of this Part.

Acts 2001, No. 194,§ 1.

§31.48. Rules and regulations

The department shall promulgate rules and regulations in accordance with the Administrative Procedure Act to implement the provisions of this Part.

Acts 2001, No. 194,§ 1.

RULE

Department of Health and Hospitals
Office of Public Health

Birth Defects Surveillance System
(LAC 48:V.Chapters 161 and 163)

In accordance with the applicable provision of the Administrative Procedure Act R.S. 49:950 et seq. and the Birth Defects Surveillance System R.S. 40.31.41 through 31.48 et seq., notice is hereby given that the Department of Health and Hospitals, Office of Public Health has adopted procedures for the surveillance of birth defects for all children under age 3, for provision of information on appropriate follow-up services to families of children identified as having birth defects, and for protection of the confidentiality of information about children who become part of the birth defects registry as well as the privacy of these individuals and their families.

Title 48
PUBLIC HEALTHCGENERAL
Part V. Public Health Services
Subpart 55. Birth Defects Surveillance System

Chapter 161. General Provisions

§16101. Definitions

Advisory Board C the nine-member advisory board of the program.

Birth Defect C an abnormality of structure, function or metabolism that develops during prenatal, perinatal or early postnatal life that is diagnosed before a child reaches 3 years of age.

Case Finding C the process used to identify potential birth defects cases for inclusion into the central registry or central database of the Louisiana Birth Defects Monitoring Network.

CSHS C the Children's Special Health Services Program within the Office of Public Health.

Confidential Information C information collected through the Louisiana Birth Defects Monitoring Network that is private and protected under state and federal laws.

Director C the program director for the Louisiana Birth Defects Monitoring Network.

Department C the Department of Health and Hospitals.

LBDMN C the Louisiana Birth Defects Monitoring Network, which the office will establish to collect information about children with birth defects. The LBDMN is established to carry out the directives of the Louisiana Birth Defects Surveillance System, which was created under Louisiana Revised Statutes 40.31.41-31.48.

Office C the Office of Public Health within the Department of Health and Hospitals.

Registry C the centralized database where data collected through the LBDMN is housed.

Reporting Source C any physician, nurse or allied health professional, hospital, laboratory, and any other facility or agent directly or indirectly responsible for providing medical services to an individual affected by a birth defect.

AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1019 (May 2004).

Chapter 163. Program Procedures

§16301. Procedures for Identification and Referral of Children with Birth Defects

A. The program will include the following.

1. Reporting sources required to report pursuant to the rule shall allow personnel from the department or its contractors to abstract information from the mother's and infant's files on their demographic characteristics, family history of birth defects, and outcomes of that and other pregnancies by that mother, according to the case definition used in LBDMN.

2. The chief operating officer, administrator, manager, director, and/or

person in charge of each reporting source shall appoint one staff member as a contact person for the LBDMN surveillance activities. That staff member should be responsible for coordinating scheduled visits by LBDMN staff to review logs, discharge indices, and other casefinding sources, and will be responsible for arranging medical records review visits and record management.

3. LBDMN staff and the contact individual at the reporting source shall establish a schedule of case-finding and record review visits. This schedule shall take into account the capabilities of each individual reporting source in responding to data/information requests, as well as the need for timely case-finding and reporting for the LBDMN.

4. Potential cases are obtained/ abstracted through review of medical records, logs, indices, appointment rosters, and other records.

5. The original medical records and other materials provided by the reporting source shall not be removed from that facility. Copies and other data should be made in compliance with existing federal and state laws and regulations.

6. The office will require information from a reporting source to be collected on a birth defects reporting form. This may be an electronic or paper form, as determined by LBDMN procedures.

7. The office will maintain a centralized database to include information reported on the birth defects reporting form.

8. The office will notify parents of infants and children identified of available early intervention services in their community.

B. Implementation

1. All reporting sources must comply with Act 194 of 2001 and these rules by July 1, 2004.

AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1019 (May 2004).

§16303. Reporting Requirements

A. The office shall determine the health care facilities and providers which shall be required to report all birth defects, the types of conditions or defects that shall be reported, the type of information that shall be contained in the confidential report and the method for making the report.

B. To ensure an accurate source of data necessary to investigate the incidence, prevalence, and trends of birth defects, a reporting source shall make available to the program staff, office staff, or authorized agent medical records or other information upon request that relates to the occurrence of a birth defect.

C. The department secretary may require, in lieu of active case finding, reporting sources identifying and diagnosing birth defects to report the birth defects to the program within 30 days of diagnosis.

AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1020 (May 2004).

§16305. Confidentiality

A. Except as specifically authorized by this Chapter, information furnished to a LBDMN employee or to an authorized agent of the office that relates to cases or suspected cases of a birth defect is confidential and may be used only for the purposes outlined in this Chapter.

B. Information relating to individual cases or individual suspected cases of birth defects is not public information and shall not be released or made public except as provided by this Chapter.

C. The LBDMN may release information:

1. for summary reporting purposes, if released without personal identifiers;

2. to medical personnel, appropriate state agencies, health authorities, regional directors, and public officers of parishes and municipalities as necessary to comply with this Chapter and board rules relating to the identification, monitoring, and referral of children with birth defects;

3. to appropriate federal agencies, as authorized by law and provided that the information contains no personal identifiers.

D. No reporting source shall be held civilly or criminally liable for conveying confidential information, except in a case of gross negligence or willful misconduct.

E. A board member, the secretary of the department, an employee of the LBDMN or office, or an authorized agent may not be examined in a civil, criminal, special, or other proceeding as to the existence or contents of pertinent records of or reports or information about a child identified or monitored for a birth defect without the consent of the child's parents, managing conservator, guardian, or legally authorized representative.

F. All employees or authorized agents of the LBDMN or office given access to medical or registry records shall agree, in writing, to maintain confidentiality of information about children with birth defects and to protect the privacy of individuals and families who become part of the LBDMN registry.

AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1020 (May 2004).

§16307. Access to Information from the Central Registry

A. The LBDMN or other authorized persons may conduct investigations of cases or suspected cases in the LBDMN registry.

B. Access to the central registry information is limited to LBDMN personnel. Other persons with a valid scientific research interest may be granted access to the information upon approval by program director, the board, and the Department's Institutional Review Board. These persons must satisfy any requirements stipulated by the board, and must receive Institutional Review Board permission to obtain the data.

C. All persons granted access to confidential information and data shall agree, in writing, to maintain confidentiality, and shall be subject to civil penalties and/or internal proceedings and penalties if confidentiality is violated. Penalties may include denial of future access to confidential information.

D. The department and LBDMN shall maintain a listing of each person who is given access to confidential information in the LBDMN registry. The listing is public information and shall be made available to the public during the office's normal hours of operation. The listing shall include:

1. the name of the person authorizing access;

2. the name, title, and organizational affiliation of each person who is granted access;

3. the dates of access;

4. the specific information requested;

5. the specific purpose for which the information was used;

6. results of independent research.

E. Progress reports and reports of findings generated from approved studies shall be submitted to the LBDMN staff and board annually or at the conclusion of the project, if the duration is shorter than 12 months.

F. All persons granted access to LBDMN information and data shall certify the destruction of data at the conclusion of the project.

AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1020 (May 2004).

§16309. Program Operation

A. The office shall monitor reporting sources for compliance with all sections of this statute.

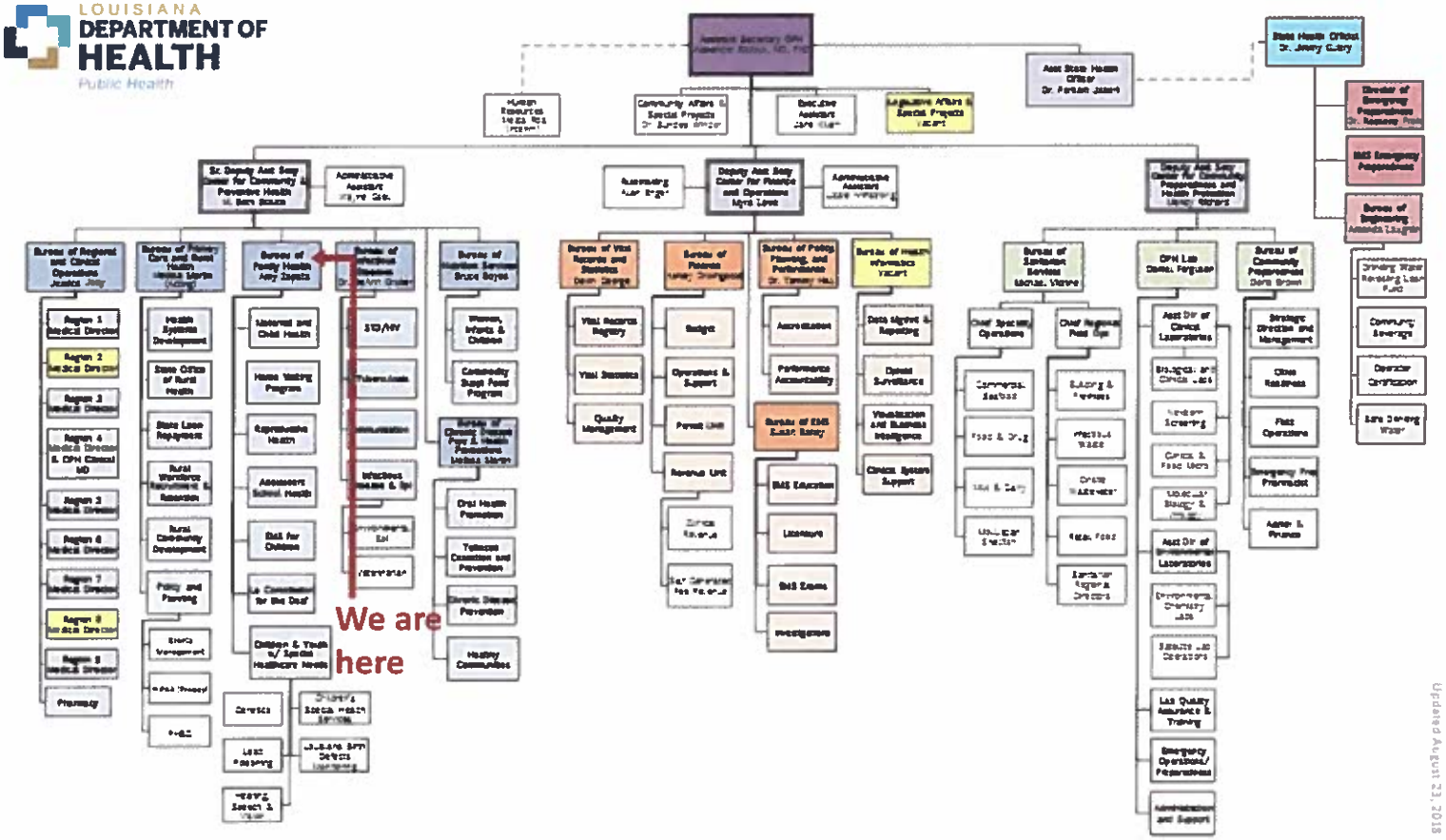
AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1021 (May 2004).

Frederick P. Cerise, M.D., M.P.H.
Secretary

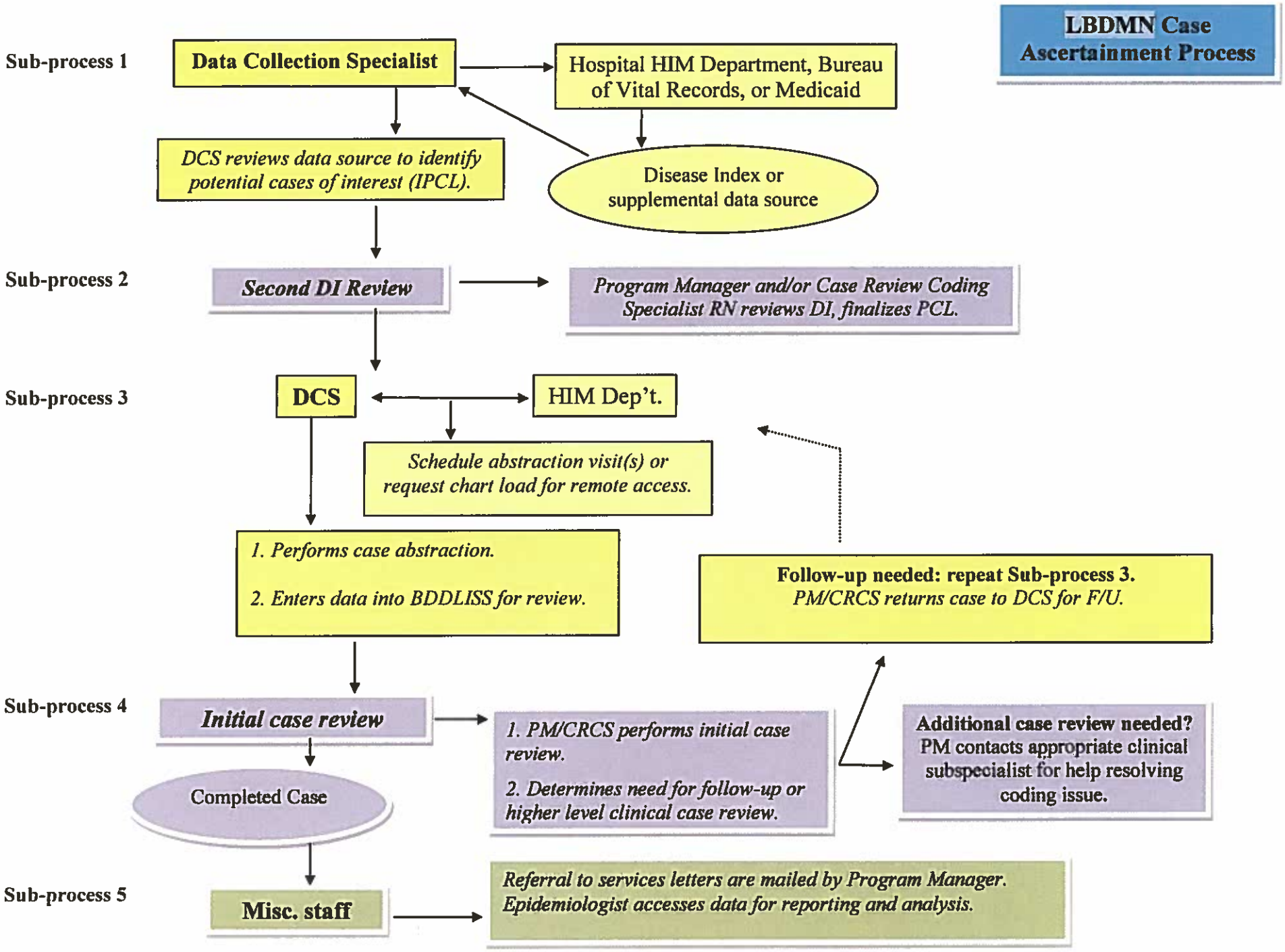
0405#050

LOUISIANA
DEPARTMENT OF
HEALTH
Public Health

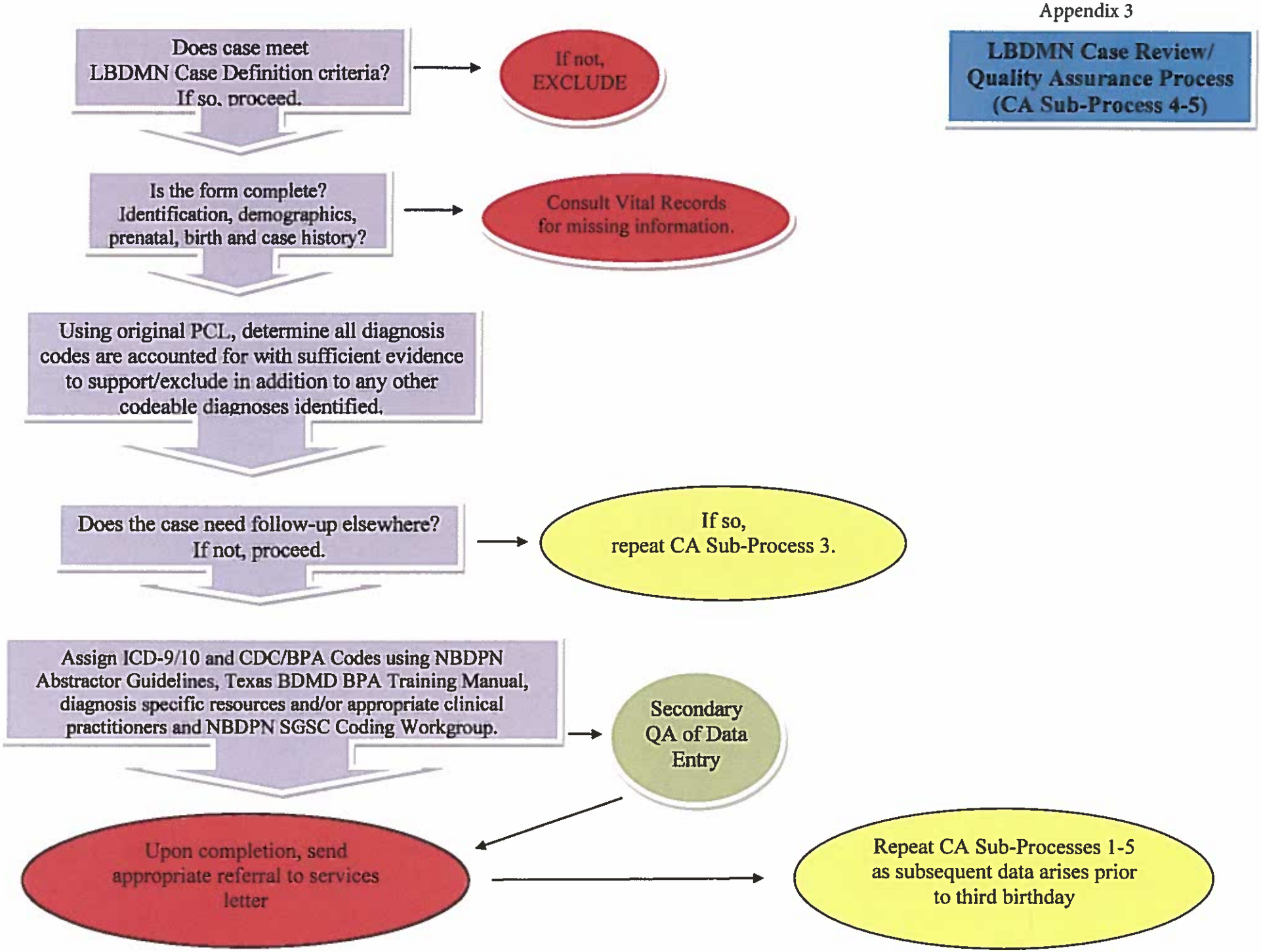


Updated August 23, 2018

Appendix 2


LBDMN Case Ascertainment Process
Sub-process 1
Data Collection Specialist
Hospital HIM Department, Bureau of Vital Records, or Medicaid
DCS reviews data source to identify potential cases of interest (IPCL).
Disease Index or supplemental data source
Sub-process 2
Second DI Review
Program Manager and/or Case Review Coding Specialist RN reviews DI, finalizes PCL.
Sub-process 3
DCS
HIM Dep't.
Schedule abstraction visit(s) or request chart load for remote access.
1. Performs case abstraction.
2. Enters data into BDDLISS for review.
Follow-up needed: repeat Sub-process 3.
PM/CRCS returns case to DCS for F/U.
Sub-process 4
Initial case review
1. PM/CRCS performs initial case review.
2. Determines need for follow-up or higher level clinical case review.
Additional case review needed?
PM contacts appropriate clinical subspecialist for help resolving coding issue.
Completed Case
Sub-process 5
Misc. staff
Referral to services letters are mailed by Program Manager.
Epidemiologist accesses data for reporting and analysis.

Appendix 3


LBDMN Case Review/ Quality Assurance Process (CA Sub-Process 4-5)
Does case meet LBDMN Case Definition criteria? If so, proceed.
If not, EXCLUDE
Is the form complete? Identification, demographics, prenatal, birth and case history?
Consult Vital Records for missing information.
Using original PCL, determine all diagnosis codes are accounted for with sufficient evidence to support/exclude in addition to any other codeable diagnoses identified.
Does the case need follow-up elsewhere? If not, proceed.
If so, repeat CA Sub-Process 3.
Assign ICD-9/10 and CDC/BPA Codes using NBDPN Abstractor Guidelines, Texas BDMD BPA Training Manual, diagnosis specific resources and/or appropriate clinical practitioners and NBDPN SGSC Coding Workgroup.
Secondary QA of Data Entry
Upon completion, send appropriate referral to services letter
Repeat CA Sub-Processes 1-5 as subsequent data arises prior to third birthday

LOUISIANA

NBDPN DATA QUALITY ASSESSMENT REPORT SUMMARY

2018

Data Quality Assessment for Population-based Birth Defects Surveillance System

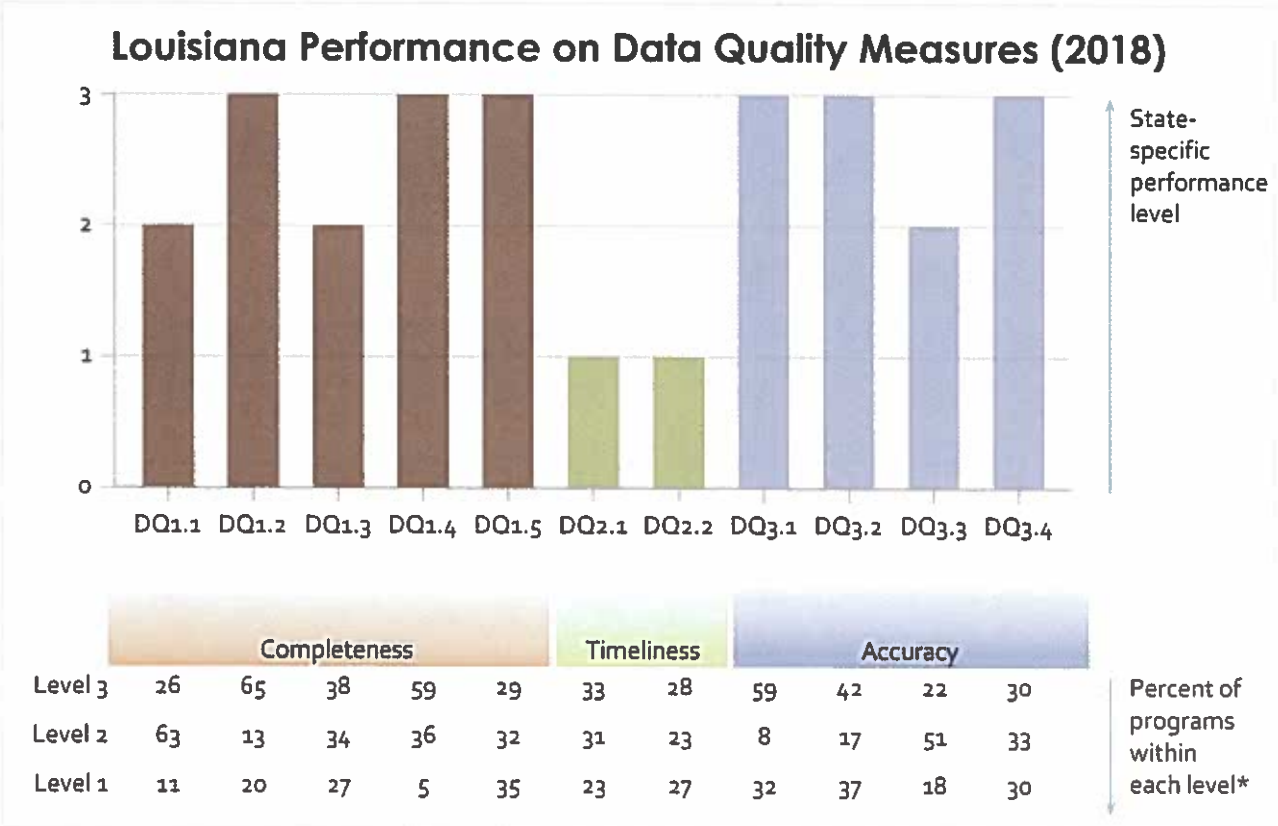
Performance standards for birth defects surveillance are intended to improve and standardize operations, outcomes and surveillance functions across state programs, thereby making data more consistent and useful for a variety of purposes at the state, multi-state and national levels. The eleven measures reflecting data quality (DQ) were developed around completeness, timeliness and accuracy attributes (see Appendix 1). Three performance levels were associated with each measure:

Level 1: Rudimentary level of performance by a surveillance program
Level 2: Essential level of performance by a surveillance program
Level 3: Optimal level of performance by a surveillance program

The expectation is that the majority of programs would be able to achieve a Level 2 on all measures.

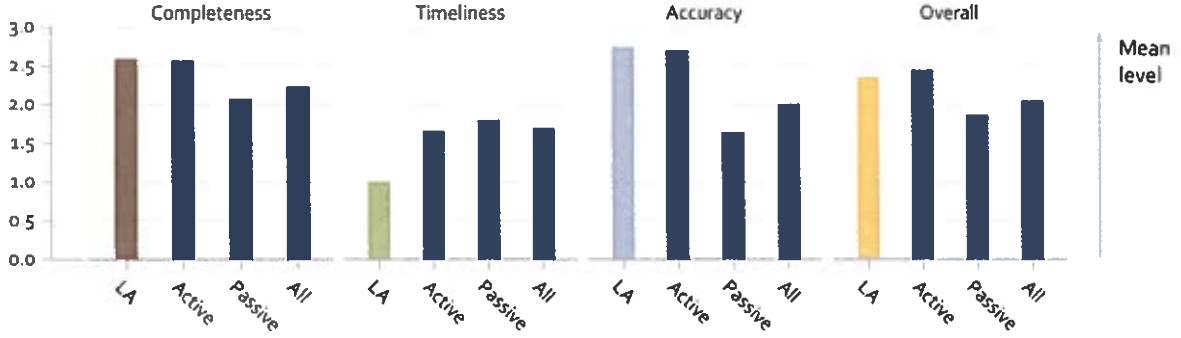
Program-specific Performance Measure Scores

The self-reported measure's performance level for your program with comparative national percentages are shown in the figure below. These results reflect the responses NBDPN received from state programs' completion of the 2018 Data Quality Self-Assessment Tool. The percent of programs by performance level is calculated for all programs in the U.S. that met inclusion criteria (n=46, NOTE: Includes Zika state based birth defects surveillance programs). The calculation by measure and performance level excludes those programs who could not meet level 1 for DQ1.1 or an overall mean score on all measures of at least one (n=1).

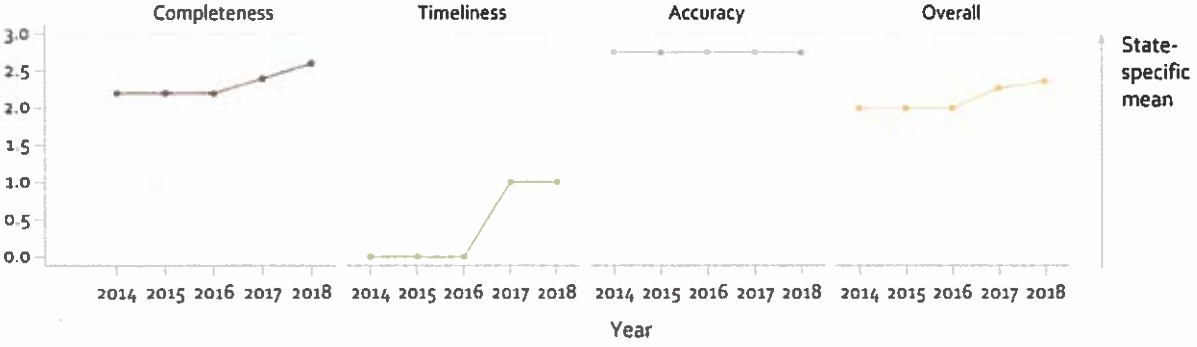


*Percent of states who could not meet Level 1 for each Data Quality Measure (DQ) is not shown.

State-specific Performance on Data Quality Measures, by Attribute and Case-finding Methodology (2018)



Temporal Changes in State-specific Performance on Data Quality Measures, by Attribute (2014-2018)



Next Steps

1. State Programs
   - Put processes in place to ensure programs have the support necessary to achieve national standards
   - Serve as champions who raise awareness about the value of national standards for data quality
   - Identify and convey your program's need for resources to achieve national standards

2. NBDPN
   - Data Quality Standards
     - Generate a report for each program that compares their performance levels on each measure to mean levels across all programs and across programs with the same approach to case-finding
     - Conduct ongoing reassessment and improvement
   - Develop Data Utility Standards
   - Incorporate Standards into the Birth Defects Surveillance Manual
   - Facilitate trainings to improve programs' ability to evaluate and enhance data quality

NBDPN DATA QUALITY ASSESSMENT REPORT SUMMARY

Appendix 1: Data Quality Measures

Completeness

DQ1.1: Types of data sources used systematically and routinely to identify potential cases at a population-based level

DQ1.2: Birth defects included using standard NBDPN case definitions

DQ1.3: Pregnancy outcomes included

DQ1.4: Systematic and routine identification of cases during ascertainment period (age of diagnosis)

DQ1.5: Data elements collected

Timeliness

DQ2.1: Time of case data completion for NBDPN *core* list

DQ2.2: Time of case data completion for NBDPN *recommended* list

Accuracy

DQ3.1: Data quality procedures for verification of cases diagnosis

DQ3.2: Scope of birth defects verified

DQ3.3: Level of expertise for individuals who perform case diagnosis verification

DQ3.4: Database quality assurance process

Louisiana Birth Defects Monitoring Network
National Birth Defects Prevention Network
Code Lists for Congenital Anomalies
ICD-9 CM to CDC/BPA to ICD-10 CM Crosswalks

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
Central Nervous System				
Anencephalus	740.0 – 740.1	740.00 – 740.10	Q00.0 - Q00.1	Level 1
Encephalocele	742.0	742.00 – 742.09	Q01.0 – Q01.9	Level 2
Holoprosencephaly	742.2	742.26	Q04.2	Level 2
Spina bifida without anencephalus	741.0, 741.9 w/o 740.0 - 740.10	741.00 – 741.99 w/o 740.0 – 740.10	Q05.0 - Q05.9, Q07.01, Q07.03 w/o Q00.0 - Q00.1	Level 1
Eye				
Aniridia	743.45	743.42	Q13.1	Level 2
Anophthalmia/microphthalmia	743.0, 743.1	743.00 – 743.10	Q11.0 – Q11.2	Level 2
Congenital cataract	743.30 – 743.34	743.32	Q12.0	Level 2
Ear				
Anotia/microtia	744.01, 744.23	744.01, 744.21	Q16.0, Q17.2	Level 2
Cardiovascular				
Aortic valve stenosis	746.3	746.30	Q23.0	Level 2
Atrial septal defect	745.5	745.51 – 745.59	Q21.1	Level 2
Atrioventricular septal defect (Endocardial cushion defect)	745.60, .61, .69	745.60 – 745.69	Q21.2	Level 1
Coarctation of the aorta	747.10	747.10 – 747.19	Q25.1	Level 2 CCHD - secondary target
Common truncus (truncus arteriosus or TA)	745.0	745.0	Q20.0	Level 2 CCHD - primary target
Double outlet right ventricle (DORV)	745.11	745.13 – 745.15	Q20.1	Level 2 CCHD - secondary target

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
Ebstein anomaly	746.2	746.20	Q22.5	Level 2 CCHD - secondary target
Hypoplastic left heart syndrome	746.7	746.70	Q23.4	Level 1 CCHD - primary target
Interrupted aortic arch (IAA)	747.11	747.215 - 747.217	Q25.2, Q25.4	Level 2 CCHD - secondary target
Pulmonary valve atresia and stenosis	746.01 (pulmonary valve atresia), 746.02 (pulmonary valve stenosis) (Note: for CCHD, 746.01 only (pulmonary atresia, intact ventricular septum))	746.00 (pulmonary valve atresia), 746.01 (pulmonary valve stenosis) (Note: for CCHD, 746.00 only (pulmonary atresia, intact ventricular septum))	Q22.0, Q22.1 (Note: for CCHD, Q22.0 only (pulmonary atresia, intact ventricular septum))	Level 2 CCHD - primary target
Single Ventricle	745.3	745.3	Q20.4	Level 2 CCHD - secondary target
Tetralogy of Fallot (TOF)	745.2	745.20 – 745.21, 747.31 (Note: code 746.84 has been removed)	Q21.3	Level 1 CCHD - primary target
Total anomalous pulmonary venous return (TAPVR)	747.41	747.42	Q26.2	Level 2 CCHD - primary target
Transposition of the great arteries (TGA)	745.10, 745.12, 745.19 (Note: for CCHD, 745.10 only (d-TGA only))	745.10 – 745.12, 745.18 – 745.19 (Note: for CCHD, 745.10 (TGA complete, no VSD), 745.11 (TGA incomplete, with VSD), 745.19 (unspecified	Q20.3, Q20.5 (Note: for CCHD, Q20.3 only)	Level 1 CCHD - primary target

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
		TGA))		
Tricuspid valve atresia and stenosis	746.1	746.100 (tricuspid atresia), 746.106 (tricuspid stenosis) (excluding 746.105 – tricuspid insufficiency) (Note: for CCHD, 746.100 only. Only tricuspid atresia is a CCHD. Many cases of tricuspid stenosis are not critical.)	Q22.4	Level 2 CCHD - primary target
Ventricular septal defect	745.4	745.40 – 745.49 (excluding 745.487, 745.498)	Q21.0	Level 2
Orofacial				
Choanal atresia	748.0	748.00	Q30.0	Level 2
Cleft lip with cleft palate	749.2	749.20 – 749.29	Q37.0 – Q37.9	Level 1
Cleft lip without cleft palate	749.1	749.10-749.19	Q36.0 – Q36.9	Level 1
Cleft palate without cleft lip	749.0	749.00 – 749.09	Q35.1 – Q35.9	Level 1
Gastrointestinal				
Biliary atresia	751.61	751.65	Q44.2 - Q44.3	Level 2

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
Esophageal atresia/tracheoesophageal fistula	750.3	750.30 – 750.35	Q39.0 – Q39.4	Level 2
Rectal and large intestinal atresia/stenosis	751.2	751.20 – 751.24	Q42.0 – Q42.9	Level 2
Small intestinal atresia/stenosis	751.1	751.10-751.19	Q41.0 – Q41.9	Level 2
Genitourinary				
Bladder exstrophy	753.5	753.50	Q64.10, Q64.19	Level 2
Cloacal exstrophy	751.5	751.555	Q64.12	Level 2
Congenital Posterior Urethral Valves	753.6	753.6	Q64.2	Level 2
Atresia & stenosis of urethra & bladder neck	753.6	753.600 753.610 753.620 753.630 753.690	Q64.31 Q64.32 Q64.33 Q64.39	LBDMN
Hypospadias	752.61	752.60 – 752.62 (excluding 752.61 and 752.621)	Q54.0 – Q54.9 (excluding Q54.4)	Level 2
Renal agenesis/hypoplasia	753.0	753.00 – 753.01	Q60.0 – Q60.6	Level 2
Unspecified obstructive defect of renal pelvis and ureter	753.20	753.290	Q62.39, Q62.10	LBDMN
Congenital obstruction of ureteropelvic junction	753.21	753.210	Q62.11	LBDMN
Congenital obstruction of ureterovesicle junction	753.22	753.210	Q62.12	LBDMN
Congenital ureterocele	753.23	753.290	Q62.31	LBDMN
Hydronephrosis	591	753.200	N13.1 N13.2 N13.30 N13.39	LBDMN
Hydroureter	593.3	753.220	N13.4	LBDMN
Musculoskeletal				

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
Clubfoot	754.51, 754.70	754.50, 754.73	Q66.0, Q66.89	Level 2
Craniosynostosis	No specific code	756.00-756.03	Q75.0	Level 2
Diaphragmatic hernia	756.6	756.60 – 756.62	Q79.0, Q79.1	Level 2
Gastroschisis	756.73 (as of 10/1/09; previous years, it was in a shared code 756.79 with omphalocele)	756.71	Q79.3	Level 1
Limb deficiencies (reduction defects)	755.2 – 755.4	755.20 – 755.49	Q71.0 – Q71.9, Q72.0 – Q72.9, Q73.0 – Q73.8	Level 1
Omphalocele	756.72 (as of 10/1/09; previous years, it was in a shared code 756.79 with gastroschisis)	756.70	Q79.2	Level 2
Chromosomal				
Deletion 22 q11(VCF syndrome)	758.32	758.37	Q93.81	Level 2
Trisomy 13 (Patau syndrome)	758.1	758.10 – 758.19	Q91.4 – Q91.7	Level 2
Trisomy 18 (Edward syndrome)	758.2	758.20 – 758.29	Q91.0 – Q91.3	Level 2
Trisomy 21 (Down syndrome)	758.0	758.00 – 758.09	Q90.0 – Q90.9	Level 1
Turner syndrome	758.6	758.60-758.69	Q96.0-.9	Level 2
Cri-du-chat syndrome (5p deletion)	758.31	758.310	Q93.4	LBDMN
Other Microdeletions	758.33	758.300	Q93.88	LBDMN
Other autosomal deletions	758.39	758.320 758.330 758.340 758.350 758.360	Q93.3 Q93.7 Q93.89	LBDMN

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
		758.380		
		758.390		
Balanced autosomal translocation in normal individual	758.4	758.400	Q95.0	LBDMN
Other conditions due to autosomal anomalies	758.5	758.500	Q99.8	LBDMN
		758.510		
		758.520		
		758.530		
		758.540		
		758.580		
		758.585		
		758.586		
		758.590		
Klinefelter's syndrome	758.7	758.700	Q98.4	LBDMN
		758.710		
		758.790		
Other conditions due to sex chromosome anomalies	758.8	758.800	Q97.0	LBDMN
	758.81	758.810	Q97.1	
		758.820	Q97.2	
		758.830	Q97.8	
		758.840	Q98.5	
		758.850	Q98.7	
		758.860	Q98.8	
		758.880	Q99.8	
Other conditions due to chromosome anomalies	758.89	758.890	Q99.8	LBDMN
Conditions due to anomaly of unspecified chromosome	758.9	758.900	Q99.9	LBDMN
		759.910		
		758.920		
		758.930		
		758.990		
Anomalies of spleen	759.0	759.000	Q89.01	LBDMN
		759.005	Q89.09	
		759.010		
		759.020		
		759.030		
		759.040		
		759.050		
		759.080		
		759.090		
Anomalies of adrenal gland	759.1	759.100	Q89.1	LBDMN
		759.110		
		759.120		
		759.130		
		759.180		

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
		759.190		
Anomalies of other endocrine glands	759.2	759.200 759.210 759.220 759.230 759.240 759.280 759.290	Q89.2	LBDMN
Situs inversus	759.3	759.300 759.310 759.320 759.330 759.340 759.390	Q89.3	LBDMN
Conjoined twins	759.4	759.400 759.410 759.420 759.430 759.440 759.480 759.490	Q89.4	LBDMN
Tuberous sclerosis	759.5	759.500	Q85.1	LBDMN
Other hamartoses, NEC	759.6	759.600 759.610 759.620 759.630 759.680 759.690	Q85.8	LBDMN
Multiple congenital anomalies, so described	759.7	759.700	Q89.7	LBDMN
Prader-Willi syndrome	759.81	759.870	Q87.1	LBDMN
Marfan syndrome, Stickler	759.82	759.860	Q87.40	LBDMN
Fragile X syndrome	759.83	759.880	Q99.2	LBDMN
Other specified anomalies	759.89	759.890 759.800 759.820 759.840	E78.71 E78.72 Q87.2 Q87.3 Q87.5 Q87.81 Q87.89 Q89.8	LBDMN
Congenital anomaly,	759.9	759.900	Q89.9	LBDMN

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
unspecified		759.910 759.990		

* NBDPN Standard Levels: Level 1 - core conditions; Level 2 - recommended conditions
*LBDMN: additional conditions specified by LBDMN Advisory Board of Directors
*CCHD: critical congenital heart defect