Louisiana Birth Defects Monitoring Network

2020 Annual Legislative Report

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LOUISIANA DEPARTMENT OF HEALTH

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Introduction

The Louisiana Birth Defects Monitoring Network (LBDMN) within the Bureau of Family Health (BFH), Office of Public Health (OPH), Louisiana Department of Health (LDH) is responsible for 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" (Louisiana Revised Statute (R.S.) 40:31.43 and Louisiana Administrative Code (LAC Title 48, Part V, Subpart 55, Chapters 161 & 163 et al. – see Appendix A).

In addition to fulfilling the annual legislative reporting requirement, the following report summarizes the performance of this important system that monitors key birth outcomes in the state and presents key findings.

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. LBDMN 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 inform Louisiana families of resources to improve their quality of life.

What We Do

The LBDMN conducts active surveillance of birth defects in children born in Louisiana. Monitoring the health status of newborns provides population-based data to inform policies, educate the public, and support efforts in the state to improve maternal and child health outcomes and prevent new occurrences of birth defects. LBDMN evaluates concerns about unexpected groups of birth defects (cluster investigation) as well as the effectiveness of preventive interventions.

Who We Serve

As a part of the Bureau of Family Health's system of monitoring birth outcomes in the state, the LBDMN supports:

- Policy makers, by identifying risk factors such as maternal exposures and chronic conditions potentially linked to specific birth conditions, and identifying preventive strategies to decrease birth defects;
- Families of infants with birth defects, from birth through three years of age, by informing them of appropriate medical, educational, public health, and peer support resources available in their region;
- Men and women of reproductive age, by providing preventive education regarding birth defects via our website;
- Researchers from the Centers for Disease Control and Prevention (CDC), universities, and other states investigating possible causes of specific birth defects.

Approximately 1,500 children with specified birth defects are identified annually, averaging 300 per 10,000 live births. Since 2005, LBDMN has investigated potential birth defects among 28,347 children. 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, operating within the LDH OPH Bureau of Family Health is a core activity to identify and support children and youth with special health care needs (CYSHCN) and their families.

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 2019: $644,550 (Title V Maternal and Child Health (MCH) Block Grant)

Operations

Role of the LDH OPH Bureau of Family Health

This public health activity is supported by high level epidemiologists and health policy leaders and is carried out by a statewide network of regionally-assigned Data Collection Specialists (DCS) who evaluate patient discharge information of newborns until three years of age. Records are reviewed from all birthing hospitals in Louisiana, as well as at Children's Hospital and Tulane University Medical Center in New Orleans. The LBDMN maintains a longitudinal data system of all children born in Louisiana diagnosed with a congenital structural, functional, and/or genetic birth defects. De-identified medical record data are analyzed statistically for patterns and trends over time. The program also informs

families of resources for children from birth until three years of age identified with specified birth defects.

Role of the 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 Med. Center
(4) One board-certified clinical geneticist from a list of names submitted by Louisiana State University (LSU) Health Sciences Center (HSC) - New Orleans
(5) One board-certified clinical geneticist from a list of names submitted by LSU HSC - Shreveport
(6) One maternal/fetal medicine physician from a list of names submitted by the March of Dimes
(7) One parent representative identified through 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.

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 that may fit within the case definition. Once potential cases are deemed within the case definition, these records 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 diagnostic billing codes (International Classification of Diseases (ICD) Codes). Other secondary data sources include Medicaid, Louisiana 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 diagnostic 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 B 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. We allow an additional six months for records to be processed by hospitals, reported to us, and abstracted to more accurately capture all diagnoses identified in those born in each calendar year.

The following tables represent data from births in 2014-2016 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 169,699 children born between 2014 and 2016, 5,202 children were diagnosed with at least one birth defect, yielding an overall prevalence of 306.5 per 10,000 live births or 3.1%. Among children with birth defects, cardiovascular system defects (48.4%) were the most common followed (in order of occurrence) by defects of the genitourinary, musculoskeletal, chromosomal, orofacial, central nervous, gastrointestinal, eye, and ear/face/neck systems. Other birth defects contributed about 5% (Table 1).

According to the Centers for Disease Control and Prevention, birth defects affect one in every 33 babies or 300 per 10,000 (about 3% of all babies) born in the United States each year.

Table 1: Type of birth defects by organ and chromosome system among children with birth defects, 2014-2016 (n = 5202)

Organ and chromosomal system	Number	Percent
Cardiovascular	2,517	48.4
Genitourinary	1,516	29.1
Musculoskeletal	645	12.4
Chromosomal	504	9.7
Oro-facial	330	6.3
Central nervous	269	5.2
Gastrointestinal	197	3.8
Eye	59	1.1
Ear, face, and neck	59	1.1
Other	257	4.9

*** Because one child may have more than one birth defect, the total percent is greater than 100% when totaled.*

The six most common specific birth defects overall, regardless of the organ or chromosomal system to which it belongs, with a prevalence greater than 10 per 10,000 live births among children born in 2014-2016 included: atrial septal defect (91.3), hypospadias (82.8), ventricular septal defect (53.9), clubfoot (13.9), Down syndrome (12.8), and craniosynostosis (11.7). Stratified by organ and chromosomal system, the most common birth defects were: for cardiovascular: atrial septal defects and ventricular septal defects; for genitourinary: hypospadias; for central nervous: spina bifida; for eyes: congenital cataract and anophthalmia/microphthalmia; for ear, face, and neck: anotia/microtia; for orofacial: cleft

lip and cleft palate; for gastrointestinal: rectal, large, and small intestinal atresia or stenosis; for musculoskeletal: clubfoot and craniosynostosis; and chromosomal: Down syndrome (Table 2).

Table 2: Occurrence of specific birth defects by organ and chromosomal system, 2014-2016 (N = 169,699)

System	Birth defects	Number	%	Prev.	CI95%
Central nervous	Spina bifida without anencephalus	69	25.7	4.1	3.2, 5.1
(n = 269)	Anencephalus	23	8.6	1.4	0.9, 2.0
	Holoprosencephaly	23	8.6	1.4	0.9, 2.0
	Encephalocele	18	6.7	1.1	0.6, 1.7
Eyes	Congenital cataract	34	57.6	2.0	1.4, 2.8
(n = 59)	Anophthalmia/microphthalmia	22	37.3	1.3	0.8, 2.0
Ear, face, neck	Anotia/microtia	34	57.6	2.0	1.4, 2.8
(n = 59)					
Cardiovascular	Atrial septal defect	1549	61.5	91.3	86.8, 95.9
(n= 2517)	Ventricular septal defect	900	35.8	53.0	49.6, 56.6
	Atrioventricular septal defect	157	6.2	9.3	7.9, 10.8
	Pulmonary valve atresia and stenosis	112	4.4	6.6	5.4, 7.9
	Tetralogy of Fallot	96	3.8	5.7	4.6, 6.9
	Coarctation of the aorta	87	3.5	5.1	4.1, 6.3
	Double outlet right ventricle	54	2.1	3.2	2.4, 4.2
	Transposition of the great arteries	45	1.8	2.7	1.9, 3.5
	Hypoplastic left heart syndrome	45	1.8	2.7	1.9, 3.5
	Dextro-transposition of great arteries	39	1.5	2.3	1.6, 3.1
	Aortic valve stenosis	36	1.4	2.1	1.5, 2.9
	Tricuspid valve atresia and stenosis	17	0.7	1.0	0.6, 1.6
	Ebstein anomaly	17	0.7	1.0	0.6, 1.6
	Tricuspid valve atresia	14	0.6	0.8	0.5, 1.4
	Total anomalous pulmonary venous connection	14	0.6	0.8	0.5, 1.4
	Interrupted aortic arch	14	0.6	0.8	0.5, 1.4
	Pulmonary valve atresia	13	0.5	0.8	0.4, 1.3
	Single Ventricle	11	0.4	0.6	0.3, 1.2
Oro-facial	Cleft palate without cleft lip	161	48.8	9.5	8.1, 11.1
(n = 330)	Cleft lip with cleft palate	95	28.8	5.6	4.5, 6.8
	Cleft lip without cleft palate	62	18.8	3.7	2.8, 4.7
	Choanal atresia	22	6.7	1.3	0.8, 2.0
Gastrointestinal	Rectal and large intestinal atresia/stenosis	92	46.7	5.4	4.4, 6.6
(n = 197)	Small intestinal atresia/stenosis	64	32.5	3.8	2.9, 4.8
	Esophageal atresia/tracheoesophageal fistula	40	20.3	2.4	1.7, 3.2
	Biliary atresia	15	7.6	0.9	0.5, 1.5
Genitourinary	Hypospadias*	715	47.2	82.8	76.9, 89.1
(n = 1516)	Renal agenesis/hypoplasia	70	4.6	4.1	3.2, 5.2
	Congenital posterior urethral valves*	34	2.2	3.8	2.7, 5.5
Musculoskeletal	Clubfoot	236	36.6	13.9	12.2, 15.8
(n = 645)	Craniosynostosis	199	30.9	11.7	10.2, 13.5
	Limb deficiencies (reduction defects)	70	10.9	4.1	3.2, 5.2
	Gastroschisis	65	10.1	3.8	3.0, 4.9
	Omphalocele	50	7.8	2.9	2.2, 3.9
	Diaphragmatic hernia	38	5.9	2.2	1.6, 3.1
Chromosomal	Trisomy 21 (Down syndrome)	218	43.3	12.8	11.2, 14.7
(n = 504)	Trisomy 18	34	6.7	2.0	1.4, 2.8
	Deletion 22 q11	26	5.2	1.5	1.0, 2.2
	Trisomy 13	15	3.0	0.9	0.5, 1.5
	Turner syndrome**	12	2.4	1.4	0.7, 2.5

**Prevalence limited to male (86,338); **Prevalence limited to female (83,360)*

Stratified by race and ethnicity, birth defects, both overall and by organ and chromosomal system, were similar for non-Hispanic white (NHW) and non-Hispanic black (NHB). The five most common birth

defects with a prevalence greater than 10 per 10,000 live births in both groups included atrial septal defect (NHW: 82.0 vs. NHB: 106.3), hypospadias (NHW: 82.2 vs. NHB: 74.0), ventricular septal defect (NHW: 56.5 vs. NHB: 46.5), clubfoot (NHW: 14.8 vs. NHB: 13.1), and Down syndrome (NHW: 11.8 vs. NHB: 13.1). In addition, cleft palate without cleft lip (11.6), craniosynostosis (15.0), and atrioventricular septal defect (11.5) were seen with a prevalence greater than 10 per 10,000 live births in non-Hispanic white and non-Hispanic black, respectively (Table 3).

Table 3: Occurrence of specific birth defects by organ and chromosomal system and race and ethnicity, 2014-2016

	Non-Hispanic White		Non-Hispanic Black		Hispanic		Non-Hispanic Other	
Defects	n	Prevalence, CI95%	n	Prevalence, CI95%	n	Prevalence, CI95%	n	Prevalence, CI95%
Central nervous system								
Spina bifida without anencephalus	36	4.2, 2.9-5.8	27	4.3, 2.8-6.2	-		-	
Anencephalus	11	1.3, 0.6-2.3	9	1.4, 0.7-2.7	-		0	
Encephalocele	11	1.3, 0.6-2.3	5	0.8, 0.3-1.8	0		-	
Holoprosencephaly	6	0.7, 0.3-1.5	16	2.5, 1.4-4.1	-		0	
Eyes								
Congenital cataract	14	1.6, 0.9-2.7	16	2.5, 1.4-4.1	-		-	
Anophthalmia/microphthalmia	12	1.4, 0.7-2.4	8	1.3, 0.5-2.5	0		-	
Ear, face, neck								
Anotia/microtia	18	2.1, 1.2-3.3	8	1.3, 0.5-2.5	5	4.1, 1.3-9.7	-	
Cardiovascular system								
Atrial septal defect	707	82.0, 76.0-88.2	672	106.3, 98.4-114.7	82	67.8, 54.0-84.2	57	86.8, 65.7-112.4
Ventricular septal defect	487	56.5, 51.6-61.7	294	46.5, 41.3-52.1	72	59.6, 46.6-75.0	35	53.3, 37.1-74.1
Atrioventricular septal defect	67	7.8, 6.0-9.9	73	11.5, 9.1-14.5	8	6.6, 2.9-13.0	-	
Coarctation of the aorta	50	5.8, 4.3-7.6	30	4.7, 3.2-6.8	-		-	
Tetralogy of Fallot	47	5.4, 4.0-7.2	33	5.2, 3.6-7.3	10	8.3, 4.0-15.2	5	7.6, 2.5-17.8
Pulmonary valve atresia and stenosis	46	5.3, 3.9-7.1	52	8.2, 6.1-10.8	6	5.0, 1.8-10.8	5	7.6, 2.5-17.8
Double outlet right ventricle	26	3.0, 2.0-4.4	21	3.3, 2.1-5.1	-		-	
Aortic valve stenosis	25	2.9, 1.9-4.3	8	1.3, 0.5-2.5	-		-	
Hypoplastic left heart syndrome	20	2.3, 1.4-3.6	19	3.0, 1.8-4.7	-		-	
Transposition of the great arteries	17	2.0, 1.1-3.2	16	2.5, 1.4-4.1	9	7.4, 3.4-14.1	-	
Dextro-transposition of great arteries	14	1.6, 0.9-2.7	15	2.4, 1.3-3.9	8	6.6, 2.9-13.0	-	
Tricuspid valve atresia and stenosis	8	0.9, 0.4-1.8	6	0.9, 0.3-2.1	0		-	
Ebstein anomaly	8	0.9, 0.4-1.8	5	0.8, 0.3-1.8	-		-	
Tricuspid valve atresia	7	0.8, 0.3-1.7	-		0		-	
Interrupted aortic arch	7	0.8, 0.3-1.7	6	0.9, 0.3-2.1	0		-	
Total anomalous pulmonary venous connection	5	0.6, 0.2-1.4	5	0.8, 0.3-1.8	-		-	
Single ventricle	-		6	0.9, 0.3-2.1	0		-	
Pulmonary valve atresia	-		7	1.1, 0.4-2.3	-		-	
Oro-facial system								
Cleft palate without cleft lip	100	11.6, 9.4-14.1	41	6.5, 4.7-8.8	14	11.6, 6.3-19.4	6	9.1, 3.4-19.9
Cleft lip with cleft palate	56	6.5, 4.9-8.4	27	4.3, 2.8-6.2	8	6.6, 2.9-13.0	-	
Cleft lip without cleft palate	44	5.1, 3.7-6.8	14	2.2, 1.2-3.7	-		-	
Choanal atresia	10	1.2, 0.6-2.1	9	1.4, 0.7-2.7	0		-	
Gastrointestinal system								
Rectal and large intestinal atresia or stenosis	51	5.9, 4.4-7.8	31	4.9, 3.3-7.0	5	4.1, 1.3-9.7	-	
Small intestinal atresia or stenosis	26	3.0, 2.0-4.4	28	4.4, 2.9-6.4	7	5.8, 2.3-11.9	-	
Esophageal atresia or tracheoesophageal fistula	18	2.1, 1.2-3.3	15	2.4, 1.3-3.9	-		-	
Biliary atresia	6	0.7, 0.3-1.5	6	0.9, 0.3-2.1	-		-	

Genitourinary system								
Hypospadias	421	82.8, 76.9-89.1	236	74.0, 64.9-84.1	29	47.0, 31.5-67.4	20	59.5, 36.3-91.9
Renal agenesis/hypoplasia	40	4.6, 3.3-6.3	25	4.0, 2.6-5.8	-		-	
Congenital Posterior Urethral Valves	16	3.9, 2.7-5.5	14	4.4, 2.4-7.4	-		-	
Musculoskeletal system								
Craniosynostosis	129	15.0, 12.5-17.8	60	9.5, 7.2-12.2	-		5	7.6, 2.5-17.8
Clubfoot	128	14.8, 12.4-17.6	83	13.1, 10.5-16.3	21	17.4, 10.8-26.6	-	
Gastroschisis	33	3.8, 2.6-5.4	22	3.5, 2.2-5.3	7	5.8, 2.3-11.9	-	
Limb deficiencies (reduction defects)	32	3.7, 2.5-5.2	28	4.4, 2.9-6.4	6	5.0, 1.8-10.8	-	
Omphalocele	21	2.4, 1.5-3.7	26	4.1, 2.7-6.0	0		-	
Diaphragmatic hernia	20	2.3, 1.4-3.6	10	1.6, 0.8-2.9	6	5.0, 1.8-10.8	0	
Chromosomal system								
Trisomy 21 (Down syndrome)	102	11.8, 9.6-14.4	83	13.1, 10.5-16.3	19	15.7, 9.5-24.5	8	12.2, 5.3-24.0
Trisomy 18	16	1.9, 1.1-3.0	16	2.5, 1.4-4.1	-		-	
Trisomy 13	6	0.7, 0.3-1.5	8	1.3, 0.5-2.5	0		-	
Turner syndrome	-		5	1.6, 0.5-3.7	-		0	
Deletion 22 q11	13	1.5, 0.8-2.6	9	1.4, 0.7-2.7	-		-	

- Case numbers between one and four are not shown

Data Quality Assessment Summary for 2019 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 2019, Louisiana ranked among the nation's top active surveillance programs in completeness, accuracy, and overall quality. However, we met only 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 three 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
- In 2019, successfully adopting a tiered abstraction strategy approach beginning with the 2016 birth cohort and for future data cohorts.

See Appendix C for complete 2019 NBDPN Data Quality Assessment Report Summary.

Conclusion

Of 169,699 children born in Louisiana between 2014 and 2016, 5,202 were diagnosed with at least one birth defect, yielding an overall prevalence of 305.6 per 10,000 live births or 3.1% (US, 3.0%). Among children with birth defects, cardiovascular system defects (about 48%) were the most common.

LBDMN incorporates evidence-based best public health surveillance practices including current technology and advanced methodologies to improve systems and data quality to identify and understand birth defects. LBDMN partners with Louisiana's Environmental Public Health Tracking Network to publish birth defects data on both its state and national portals along with other core sets of health, exposure, and hazards data, information summaries, and tools to enable analysis, visualization and reporting of insights drawn from data. Data are available to environmental and public health practitioners, healthcare providers, community members, policy makers, and others to make information-driven decisions that affect their health. To further prevention, LBDMN participates in campaigns to inform men and women of reproductive age of healthy prenatal lifestyle choices such as daily consumption of 400 micrograms of folic acid daily; reducing exposures to infections and toxins; and controlling chronic conditions such diabetes and hypertension. Finally, LBDMN works within the Bureau of Family Health to inform Louisiana families of children impacted by birth defects of resources to improve their quality of life.

The Louisiana Birth Defects Monitoring Network's surveillance system and public health actions as described above fulfil the legislative mandate of Louisiana Revised Statutes Title 40, Part VII, Sections 31.41–31.48 to maintain "*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.*"

Appendix

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

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 case-finding 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.

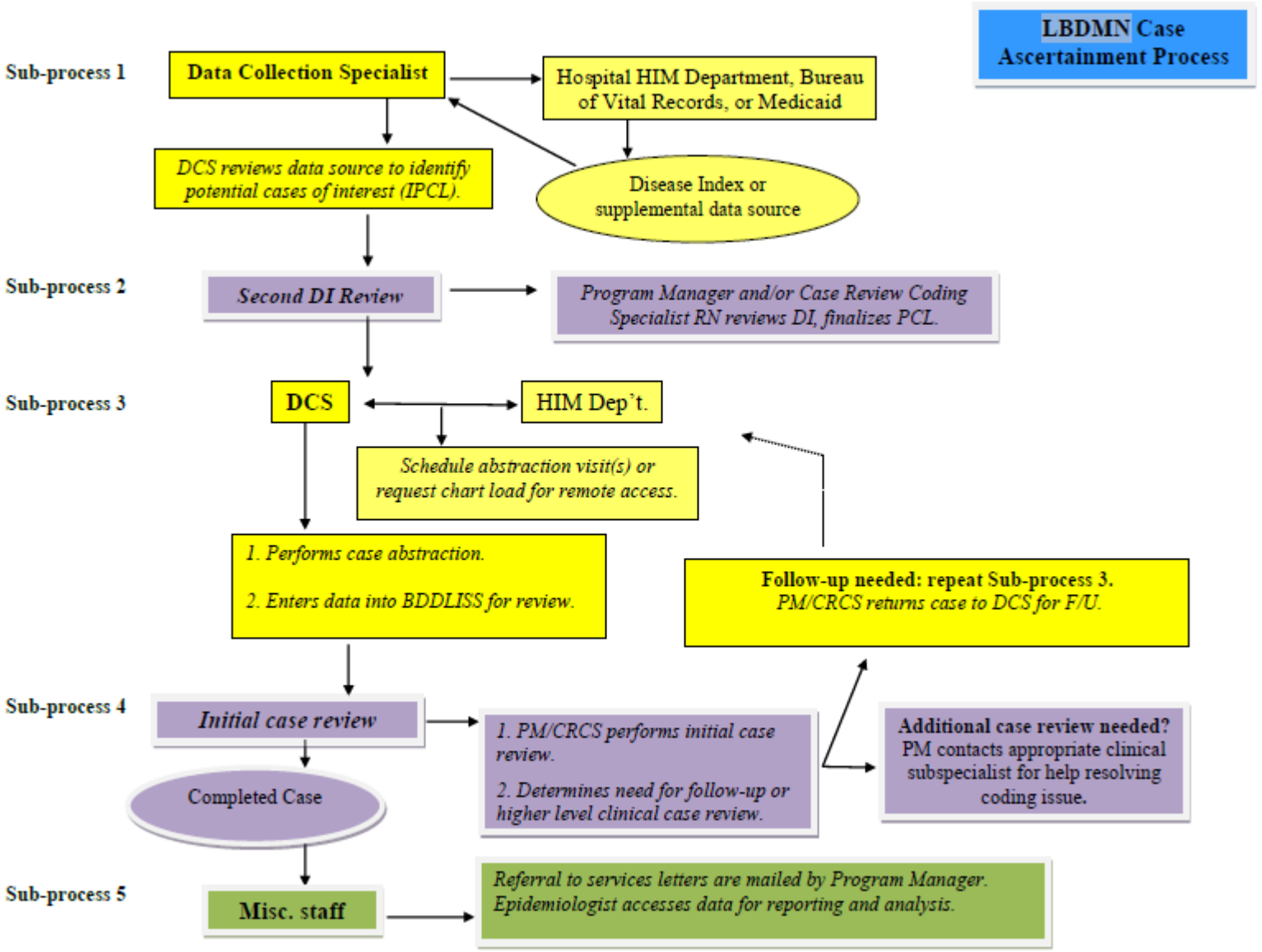
AUTHORITY NOTE: Promulgated in accordance with R.S. 40.31.48.

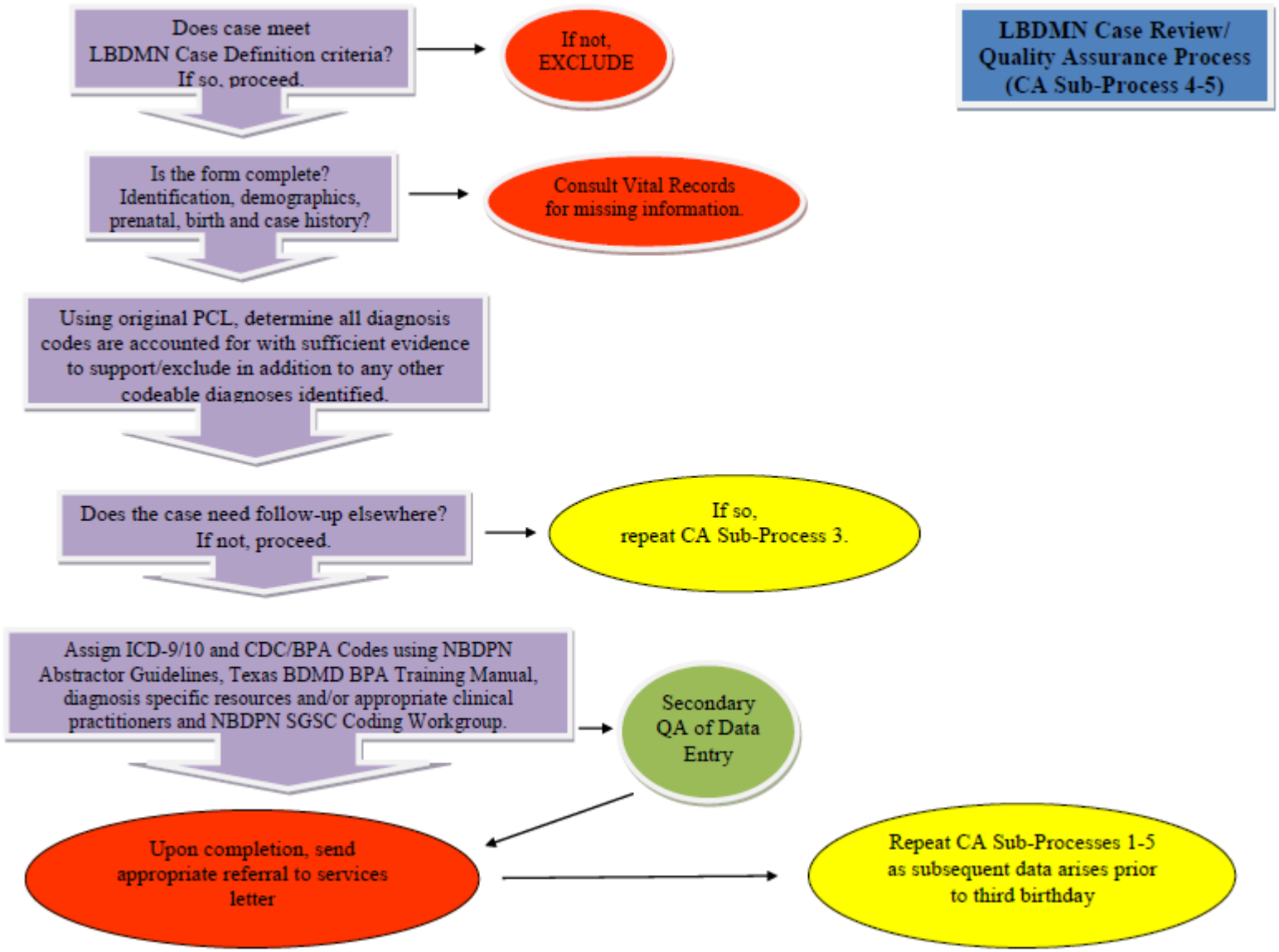
HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 30:1021 (May 2004).

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

0405#050

Appendix B: Case Ascertainment\Review\Quality Assurance Chart




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

Appendix C: 2019 NBDPN Data Quality Assessment Summary

LOUISIANA

NBDPN DATA QUALITY ASSESSMENT REPORT SUMMARY

2019

Data Quality Assessment for Population-based Birth Defects Surveillance System

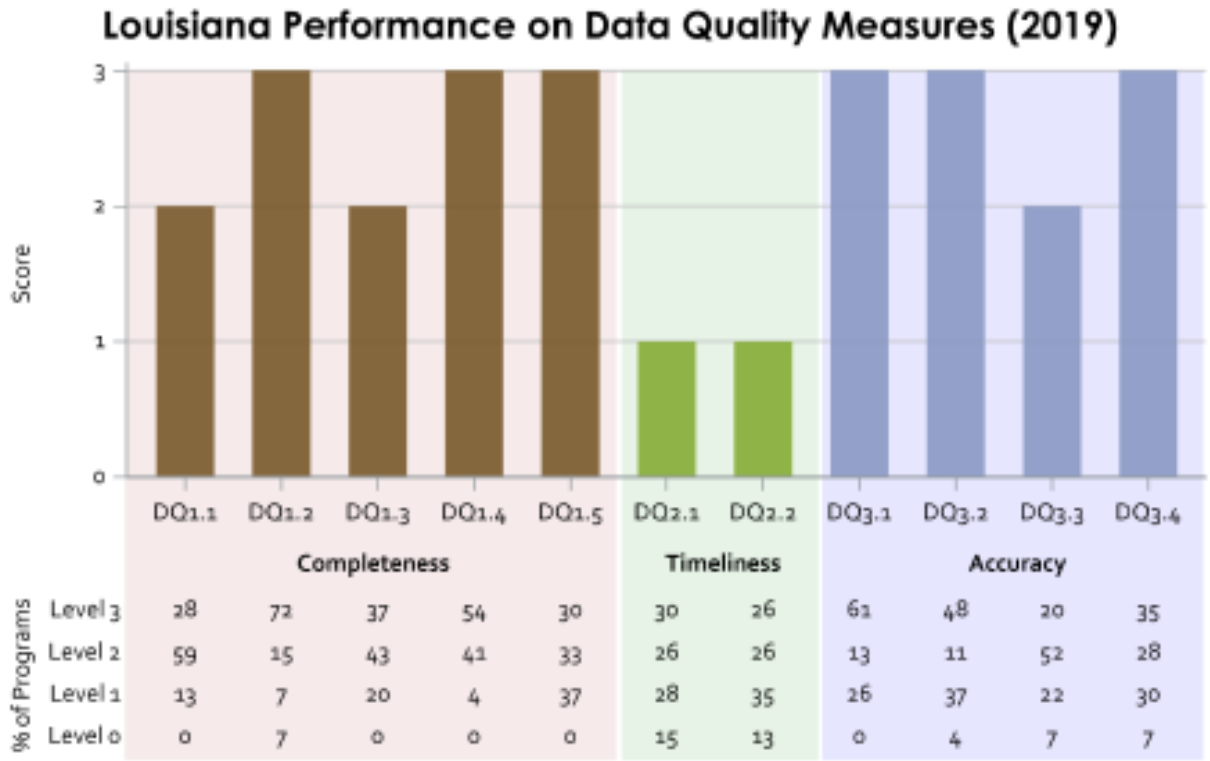
Performance standards for birth defects surveillance are intended to improve and standardize operations, outcomes and surveillance functions across state programs, thereby making data more consistent and useful for a variety of purposes at the state, multi-state and national levels. The eleven measures 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

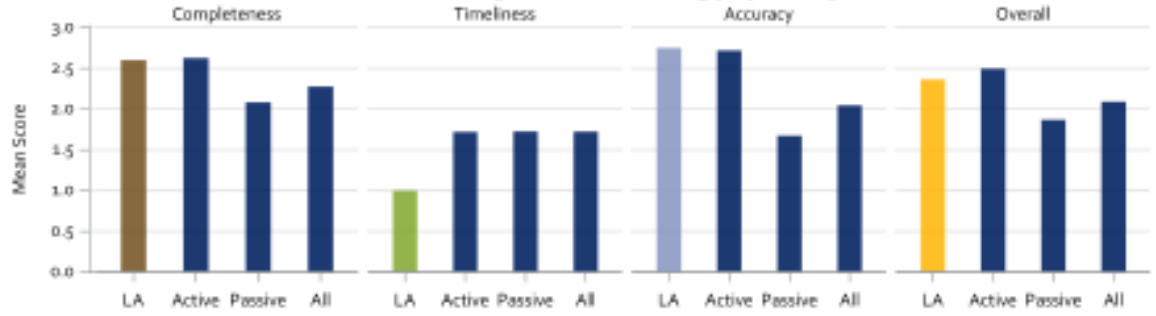
The expectation is that the majority of programs would be able to achieve a Level 2 on all measures.

Program-specific Performance Measure Scores

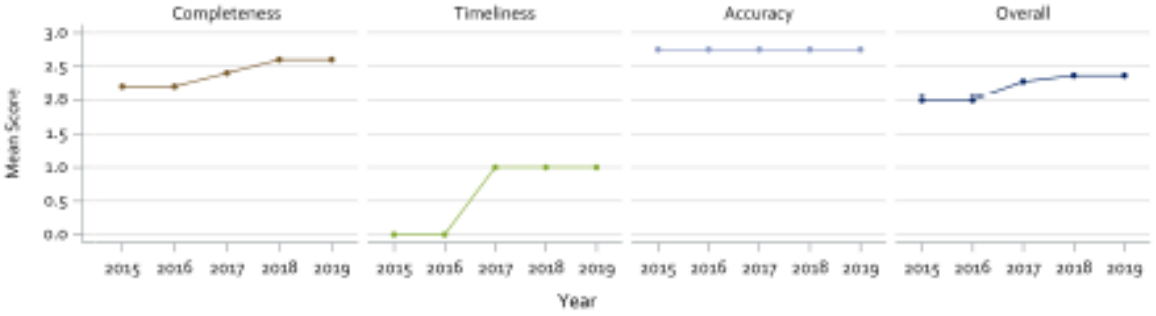
The self-reported measure's performance level for your program with comparative national percentages are shown in the figure below. These results reflect the responses NBDPN received from state programs' completion of the 2019 Data Quality Self-Assessment Tool. The percent of programs by performance level is calculated for all programs in the U.S. that submitted forms in the current year, that met a level 1 for DQ1.1 and achieved an overall mean score of 1 for all measures (n=46).



State-specific Performance on Data Quality Measures, by Attribute and Case-finding Methodology (2019)



Temporal Changes in State-specific Performance on Data Quality Measures, by Attribute (2015-2019)



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
   - Provide program-specific and overall summary reports
   - Assess how well the current tool measures program data quality
   - Develop Data Utility Standards
   - Incorporate Standards into the Birth Defects Surveillance Manual
   - Facilitate trainings to improve programs' ability to evaluate and enhance data quality

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

For questions about **NBDPN standards**, e-mail standards@nbdpn.org.

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

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
Double outlet right ventricle (DORV)	745.11	745.13 – 745.15	Q20.1	Level 2 CCHD - secondary target
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,	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*
		with VSD), 745.19 (unspecified 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	758.500 758.510 758.520 758.530 758.540 758.580 758.585 758.586 758.590	Q99.8	LBDMN
Klinefelter's syndrome	758.7	758.700 758.710 758.790	Q98.4	LBDMN
Other conditions due to sex chromosome anomalies	758.8 758.81	758.800 758.810 758.820 758.830 758.840 758.850 758.860 758.880	Q97.0 Q97.1 Q97.2 Q97.8 Q98.5 Q98.7 Q98.8 Q99.8	LBDMN
Other conditions due to chromosome anomalies	758.89	758.890	Q99.8	LBDMN
Conditions due to anomaly of unspecified chromosome	758.9	758.900 759.910 758.920 758.930 758.990	Q99.9	LBDMN
Anomalies of spleen	759.0	759.000 759.005 759.010 759.020 759.030 759.040 759.050 759.080 759.090	Q89.01 Q89.09	LBDMN
Anomalies of adrenal gland	759.1	759.100 759.110 759.120 759.130 759.180 759.190	Q89.1	LBDMN

Birth Defects	ICD-9-CM Codes	CDC/BPA Codes- (internal use only)	ICD-10-CM Codes	Standard Level*
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, unspecified	759.9	759.900 759.910 759.990	Q89.9	LBDMN

* 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

Note: As of January 2014, the following conditions were dropped from the NBDPN/LBDMN list:

- **ICD-9 760.71 - Amniotic bands**
- **ICD-9 754.30-31, 754.35 - Congenital hip dislocation**
- **ICD-9 752.62 - Epispadias**
- **ICD-9 762.8 - Fetus or newborn affected by maternal alcohol use**
- **ICD-9 751.3 - Hirschsprung disease (congenital megacolon)**
- **ICD-9 742.3 - Hydrocephalus without spina bifida**
- **ICD-9 742.1 – Microcephalus* LBDMN chose to continue surveillance due to Zika concerns**
- **ICD-9 747.0 - Patent ductus arteriosus**
- **ICD-9 750.5 - Pyloric stenosis**

The following conditions were added and are included in the tables above:

- Cloacal Exstrophy
- Clubfoot
- Congenital posterior urethral valves (not new to LBDMN list)
  - Craniosynostosis
  - Deletion 22q11 (not new to LBDMN list)
  - Double outlet right ventricle (DORV) (not new to LBDMN list)
- Holoprosencephaly
- Interrupted aortic arch (IAA) (not new to LBDMN list)
- Single ventricle
- Small intestine atresia/stenosis
- Turner syndrome (not new to LBDMN list)

The following conditions were merged and are included in the tables above:
Reduction deformity, lower limbs; reduction deformity, upper limbs. Merged to limb deficiencies (reduction defects).

The following conditions were separated and are included in the tables above:
Cleft lip with and without cleft palate separated to cleft lip with cleft palate; cleft lip without cleft palate.

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