Louisiana

Birth Defects

Monitoring Network

2021 Annual Legislative Report

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LOUISIANA DEPARTMENT OF HEALTH

Report prepared by:

Julie Johnston, Program Manager, LDH-OPH-Bureau of Family Health (BFH)-Louisiana Birth Defects Monitoring Network (LBDMN)

Tri Tran, MD, MPH, Senior Epidemiologist, LDH-OPH-BFH

Dionka Pierce, MPH, Program Manager, LDH-OPH-BFH

Editors:

Amy Zapata, MPH, Director, LDH-OPH-BFH

Jane Herwehe, MPH, Lead, Data to Action Team, LDH-OPH-BFH

Acknowledgements:

The Louisiana Birth Defects Monitoring Network's surveillance system and public health actions as described in this report fulfill 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."

Funding for LBDMN surveillance is provided through the federal Title V Maternal and Child Health (MCH) Block Grant.

We recognize the LBDMN Data Collection Specialists and Case Review Coding Specialist who abstracted the medical records to collect these data. Their dedication and hard work made this report possible.

We are thankful for the volunteer members of the LBDMN Advisory Board, who provide clinical expertise for the operations and management of the birth defects surveillance system.

Lastly, we honor the families of children impacted by birth defects represented in this report. It is our sincere hope that the activities of LBDMN will improve the systems of care serving Louisiana's families.

Contents

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 2001, 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).

The following report summarizes the key findings related to the prevalence of birth defects and performance of this essential surveillance system.

Our Mission

The mission of LBDMN is to collect, analyze, and disseminate high quality, timely, actionable data to inform policy and to improve Louisiana's maternal health system to eliminate preventable birth defects, mitigate disability, and connect families with resources to improve their quality of life.

What We Do

The LBDMN incorporates evidence-based public health surveillance best practices including current technology and advanced methodologies to conduct 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 can evaluate 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 3 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 birth defects prevention education materials 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 295 per 10,000 live births. Since 2005, LBDMN has investigated potential birth defects among 33,447 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 3rd 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 is a core activity of the Bureau of Family Health's Title V Maternal Child Health programs 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 3 years of age for major structural, functional, or genetic birth defects.
- A partnership with the BFH Family Resource Center to link families of children under 3 years of age with specified birth defects health, social, and developmental resources.
- 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 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 to prevent risks of associated birth defects.

Operations

Role of the LDH OPH Bureau of Family Health

This public health activity is supported by senior 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 3 years of age. Records are reviewed from all birthing hospitals in Louisiana, as well as at Children's Hospital, Ochsner Medical Center, and Tulane University Medical Center in New Orleans. The LBDMN maintains a longitudinal data system of all children born in Louisiana diagnosed with a structural, functional, and/or genetic birth defect. De-identified medical record data are analyzed statistically for patterns and trends over time. In the 2021 referral project, BFH Family Resource Center will link families to health, social services, and developmental resources for children identified with specified birth defects.

Role of the Advisory Board

As mandated in the authorizing statute, LA R.S. 40:31.43, LBDMN is guided by an advisory board of volunteer stakeholders appointed by the Secretary of LDH.

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 on the third Friday of January, June, and October. The focus of 2021-2022 board activities is moving birth defects data to action through data analysis. This will help identify evidence-based practices to address trends with systems-level strategies. These strategies will aid in advancing prevention efforts and interventions to increase health and developmental outcomes for children identified with birth defects. Planned activities include recruitment of additional members to diversify expertise including pediatric cardiology, hospital health information management, and rural health access, as well as representation from the new Our Lady of the Lake Children's Hospital; and implementing board case review to identify areas in need of recommendations for primary and secondary prevention.

In addition to providing annual reports to the Louisiana Legislature, LBDMN data are included in annual reports produced by the National Birth Defects Prevention Network, special reports such as the Morbidity and Mortality Weekly Report exploring gastroschisis trends and maternal opioid prescription rates of 2006-2015, and *Prevalence of individual brain and eye defects potentially related to Zika virus in pregnancy in 22 U.S. states and territories, January 2016 – June 2017*, and manuscripts including *Short Interpregnancy Interval and Prevalence of Birth Defects: A multi-state study* to be published in the January 2022 special issue of *Birth Defects Research.*

Methodology

LBDMN contacts health providers to find cases and collect data. Potential cases of interest are identified from hospital discharge indices, 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). Medical and vital statistic records are reviewed to collect and validate data among children diagnosed from birth up to their 3rd birthday.

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. Data are stored and managed in a database integrated with LEERS birth and death certificates as well as Early Hearing Detection and Intervention (LA-EHDI) data.

Not all defects are evident at birth; therefore, LBDMN includes children diagnosed before their 3rd birthday allowing adequate time to capture all birth defects within our case definition. Additional time is allowed for records to be processed by hospitals, reported to us, and abstracted to more accurately capture all diagnoses identified among those born in each calendar year. Please refer to Appendix A for the Case Ascertainment\Review\Quality Assurance flow chart.

Diagnoses are converted into the CDC clinical coding system, based on the British Pediatric Association and Classification of Diseases and the ICD-10CM. Prevalence rate 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. Data are reported in three-year increments to have an adequate number of cases to be representative at the state level when stratified by race/ethnicity and type of major birth defects.

Findings

Of 179,510 children born between 2015 and 2017, 5,296 children were diagnosed with at least one birth defect, yielding an overall prevalence of 295 per 10,000 live births or 2.95%. Among children with birth

defects, cardiovascular system defects (50.3%) 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).

Table 1: Type of birth defects by organ and chromosome system among children with birth defects, 2015-2017 (n = 5,296)

Organ and chromosome system	Number	Percent*
Cardiovascular	2,662	50.3
Genitourinary	1,532	28.9
Musculoskeletal	633	12.0
Chromosomal	507	9.6
Oro-facial	318	6.0
Central nervous	279	5.3
Gastrointestinal	180	3.4
Eye	77	1.5
Ear, face, and neck	61	1.2
Other	251	4.7

** Because one child may have more than one birth defect, the aggregate percentages are 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 2015-2017 included atrial septal defect (96.3), ventricular septal defect (49.0), hypospadias (79.8), clubfoot (14.3), Down syndrome (10.8), and craniosynostosis (10.4). 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 for chromosomal: Down syndrome (Table 2).

Table 2: Occurrence of specific birth defects by organ and chromosomal system, 2015-2017 (N = 179,510)

System	Birth defects	Number	%	Prev.	95%CI
	Total	**5,296**		**295.0**	**287.1, 303.1**
Central nervous (n = 279)	Spina bifida without anencephalus	50	17.9	2.8	2.1, 3.7
	Holoprosencephaly	25	9.0	1.4	0.9, 2.1
	Encephalocele	19	6.8	1.1	0.6, 1.7
	Anencephalus	17	6.1	0.9	0.6, 1.5
Eyes (n = 61)	Congenital cataract	31	50.8	1.7	1.2, 2.5
	Anophthalmia/microphthalmia	22	36.1	1.2	0.8, 1.9
Ear, face, neck (n = 77)	Anotia/microtia	39	50.6	2.2	1.5, 3.0
Cardiovascular (n= 2662)	Atrial septal defect	1,729	65.0	96.3	91.8, 101.0
	Ventricular septal defect	880	33.1	49.0	45.8, 52.4
	Atrioventricular septal defect	131	4.9	7.3	6.1, 87
	Pulmonary valve atresia and stenosis	119	4.5	6.6	5.5, 7.9
	Coarctation of the aorta	88	3.3	4.9	3.9, 6.0
	Tetralogy of Fallot (TOF)	87	3.3	4.8	3.9, 6.0
	Double outlet right ventricle (DORV)	41	1.5	2.3	1.6, 3.2
	Transposition of the great arteries (TGA)	40	1.5	2.2	1.6, 3.0
	Dextro-transposition of great arteries	38	1.4	2.1	1.5, 2.9
	Hypoplastic left heart syndrome	35	1.3	1.9	1.4, 2.7
	Aortic valve stenosis	28	1.1	1.6	1.0, 2.3
	Total anomalous pulmonary venous connection	17	0.6	0.9	0.6, 1.5
	Single ventricle	16	0.6	0.9	0.5, 1.4
	Tricuspid valve atresia and stenosis	14	0.5	0.8	0.4, 1.3
	Ebstein anomaly	14	0.5	0.8	0.4, 1.3
	Pulmonary valve atresia	13	0.5	0.7	0.4, 1.2
	Tricuspid valve atresia	10	0.4	0.6	0.3, 1.0
	Interrupted aortic arch (IAA)	9	0.3	0.5	0.2, 1.0
	Common truncus (truncus arteriosus or TA)	7	0.3	0.4	0.2, 0.8
Oro-facial (n = 318)	Cleft palate without cleft lip	164	51.1	9.1	7.8, 10.6
	Cleft lip with cleft palate	88	27.4	4.9	3.9, 6.0
	Cleft lip without cleft palate	56	17.4	3.1	2.4, 4.1
	Choanal atresia	22	6.9	1.2	0.8, 1.9
Gastrointestinal (n = 180)	Rectal and large intestinal atresia/stenosis	76	42.2	4.2	3.3, 5.3
	Small intestinal atresia/stenosis	65	36.1	3.6	2.8, 4.6
	Esophageal atresia/tracheoesophageal fistula	34	18.9	1.9	1.3, 2.6
	Biliary atresia	15	8.3	0.8	0.5, 1.4
Genitourinary (n = 1532)	Hypospadias*	728	47.5	79.8	74.1, 85.5
	Renal agenesis/hypoplasia	72	4.7	4.0	3.1, 5.1
	Congenital posterior urethral valves*	33	2.2	3.6	2.5, 5.1
Musculoskeletal (n = 633)	Clubfoot	256	40.4	14.3	12.6, 16.1
	Craniosynostosis	186	29.4	10.4	8.9, 12.0
	Limb deficiencies (reduction defects)	65	10.3	3.6	2.8, 4.6
	Gastroschisis	57	9.0	3.2	2.4, 4.1
	Omphalocele	40	6.3	2.2	1.6, 3.0
	Diaphragmatic hernia	37	5.8	2.1	1.5, 2.8
Chromosomal (n = 507)	Trisomy 21 (Down syndrome)	194	38.3	10.8	9.3, 12.4
	Trisomy 18	33	6.5	1.8	1.3, 2.6
	Deletion 22 q11	26	5.1	1.4	0.9, 2.1
	Trisomy 13	12	2.4	0.7	0.3, 1.2
	Turner syndrome**	7	1.4	0.8	0.3, 1.6

**Prevalence limited to male (91,277); **Prevalence limited to female (88,233)*

Stratified by race and ethnicity, the total prevalence of birth defects was a bit higher in non-Hispanic whites (NHW) (306.9) than in non-Hispanic Blacks (NHB) (289.2). The five most common birth defects with a prevalence equal or greater than 10 per 10,000 live births in both groups included atrial septal defect (NHW: 87.8 vs. NHB: 114.3), hypospadias (NHW: 91.9 vs. NHB: 72.0), ventricular septal defect (NHW: 52.1 vs. NHB: 42.9), clubfoot (NHW: 15.8 vs. NHB: 13.2), and Down syndrome (NHW: 10.0 vs. NHB: 11.0). In addition, cleft palate without cleft lip (11.2) and craniosynostosis (13.4) were seen with a prevalence greater than 10 per 10,000 live births in NHW (Table 3).

Table 3: Occurrence of specific birth defects by organ and chromosomal system and race and ethnicity, 2015-2017

	Non-Hispanic White		**Non-Hispanic Black**		**Hispanic**		**Non-Hispanic Other**	
Defects	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**
Total	2843	306.9, 295.8-318.4	1913	289.2, 276.4-302.5	347	263.9, 236.8-293.2	187	253.2, 218.2-292.2
Central nervous system								
Spina bifida without anencephalus	29	3.1, 2.1- 4.5	18	2.7, 1.6-4.3	-		-	
Anencephalus	9	1.0, 0.4-1.8	5	0.8, 0.2-1.8	-		0	
Encephalocele	9	1.0, 0.4-1.8	10	1.5, 0.7-2.8	0		0	
Holoprosencephaly	8	0.9, 0.4-1.7	14	2.1, 1.2-3.6	-		0	
Eyes								
Congenital cataract	10	1.1, 0.5-2.0	12	1.8, 0.9-3.2	5	3.8, 1.2-8.9	-	
Anophthalmia/microphthalmia	11	1.2, 0.6-2.1	9	1.4, 0.6-2.6	0		-	
Ear, face, neck								
Anotia/microtia	19	2.1, 1.2-3.2	10	1.5, 0.7-2.8	8	6.1, 2.6-12.0	-	
Cardiovascular system								
Atrial septal defect	813	87.8, 81.8-94.0	756	114.3, 106.3-122.7	98	74.5, 60.5-90.8	58	78.5, 59.6-101.5
Ventricular septal defect	483	52.1, 47.6-57.0	284	42.9, 38.1-48.2	82	62.4, 49.6-77.4	31	42.0, 28.5-59.6
Atrioventricular septal defect	61	6.6, 5.0-8.5	60	9.1, 6.9-11.7	7	5.3, 2.1-11.0	-	
Coarctation of the aorta	52	5.6, 4.2-7.4	32	4.8, 3.3-6.8	-		-	
Tetralogy of Fallot	50	5.4, 4.0-7.1	28	4.2, 2.8-6.1	5	3.8, 1.2-8.9	-	
Pulmonary valve atresia and stenosis	59	6.4, 4.8-8.2	48	7.3, 5.4-9.6	8	6.1, 2.6-12.0	-	
Double outlet right ventricle	23	2.5, 1.6-3.7	15	2.3, 1.3-3.7	-		-	
Aortic valve stenosis	19	2.1, 1.2-3.2	5	0.8, 0.2-1.8	-		-	
Hypoplastic left heart syndrome	20	2.2, 1.3-3.3	12	1.8, 0.9-3.2	-		-	
Transposition of the great arteries	23	2.5, 1.6-3.7	12	1.8, 0.9-3.2	-		-	
Dextro-transposition of great arteries	21	2.3, 1.4-3.5	12	1.8, 0.9-3.2	-		-	
Tricuspid valve atresia and stenosis	6	0.6, 0.2-1.4	8	1.2, 0.5-2.4	0		0	
Ebstein anomaly	8	0.9, 0.4-1.7	5	0.8, 0.2-1.8	-		0	
Tricuspid valve atresia	5	0.5, 0.2-1.3	5	0.8, 0.2-1.8	0		0	
Interrupted aortic arch	5	0.5, 0.2-1.3	-		0		-	
Total anomalous pulmonary venous connection	7	0.8, 0.3-1.6	8	1.2, 0.5-2.4	-		-	
Single ventricle	9	1.0, 0.4-1.8	6	0.9, 0.3-2.0	0		-	
Pulmonary valve atresia	5	0.5, 0.2-1.3	8	1.2, 0.5-2.4	0		0	
Oro-facial system								
Cleft palate without cleft lip	104	11.2, 9.2-13.6	41	6.2, 4.4-8.4	13	9.9, 5.3-16.9	6	8.1, 3.0-17.7
Cleft lip with cleft palate	55	5.9, 4.5-7.7	21	3.2, 2.0-4.9	10	7.6, 3.6-14.0	-	
Cleft lip without cleft palate	35	3.8, 2.6-5.3	16	2.4, 1.4-3.9	-		-	
Choanal atresia	14	1.5, 0.8-2.5	5	0.8, 0.2-1.8	-		-	

Table 3: Occurrence of specific birth defects by organ and chromosomal system and race and ethnicity, 2015-2017 (continued)

	Non-Hispanic White		Non-Hispanic Black		Hispanic		Non-Hispanic Other	
Defects	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**	**n**	**Prevalence 95%CI**
Gastrointestinal system								
Rectal and large intestinal atresia or stenosis	42	4.5, 3.3-6.1	27	4.1, 2.7-5.9	5	3.8, 1.2-8.9	-	
Small intestinal atresia or stenosis	28	3.0, 2.0-4.4	27	4.1, 2.7-5.9	7	5.3, 2.1-11.0	-	
Esophageal atresia or tracheoesophageal fistula	15	1.6, 0.9-2.7	14	2.1, 1.2-3.6	-		-	
Biliary atresia	5	0.5, 0.2-1.3	7	1.1, 0.4-2.2	-		-	
Genitourinary system								
Hypospadias	435	91.9, 83.4-100.9	240	72.0, 63.1-81.7	31	46.0, 31.3-65.3	22	58.9, 36.9-89.2
Renal agenesis/hypoplasia	44	4.8, 3.5-6.4	20	3.0, 1.8-4.7	5	3.8, 1.2-8.9	-	
Congenital Posterior Urethral Valves	15	3.2, 1.8- 5.2	15	4.5, 2.5-7.4	-		0	
Musculoskeletal system								
Clubfoot	146	15.8, 13.3-18.5	87	13.2, 10.5-16.2	19	14.4, 8.7-22.6	-	
Craniosynostosis	124	13.4, 11.1-16.0	47	7.1, 5.2-9.4	9	6.8, 3.1-13.0	5	6.8, 2.2-15.8
Gastroschisis	30	3.2, 2.2-4.6	18	2.7, 1.6-4.3	6	4.6, 1.7-9.9	-	
Limb deficiencies (reduction defects)	27	2.9, 1.9-4.2	29	4.4, 2.9-6.3	7	5.3, 2.1-11.0	-	
Omphalocele	18	1.9, 1.2-3.1	20	3.0, 1.8-4.7	0		-	
Diaphragmatic hernia	22	2.4, 1.5-3.6	9	1.4, 0.6-2.6	6	4.6, 1.7-9.9	0	
Chromosomal system								
Trisomy 21 (Down syndrome)	93	10.0, 8.1-12.3	73	11.0, 8.7-13.9	19	14.4, 8.7-22.6	8	10.8, 4.7-21.3
Trisomy 18	20	2.2, 1.3-3.3	11	1.7, 0.8-3.0	-		-	
Deletion 22 q11	15	1.6, 0.9-2.7	7	1.1, 0.4-2.2	-		-	
Trisomy 13	5	0.5, 0.2-1.3	7	1.1, 0.4-2.2	0		0	
Turner syndrome	0		5	1.5, 0.5-3.6	-		-	

- Case number not shown between one and four

2020 Performance Assessment and Improvements

LBDMN follows national standards and guidelines for birth defects surveillance. 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 Data Quality Standards and Assessment Tools 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.

Using NBDPN data quality assessments, Louisiana ranks among the nation's top active surveillance programs in completeness, accuracy, and overall quality.

In 2020, LBDMN critically assessed each facet of operations to adopt performance improvement measures to improve timeliness of workflow alignment, referral to resources, advisory board activities, and legislative reporting.

Notable updates include:

- Adopting a new approach for data collection to only abstract NBDPN core and recommended cases, eliminating 29 codes that were not reportable to any federal, state, or stakeholder partners
- Implementation of automated case-finding process for initial potential cases
- Expanded remote access to electronic medical records for abstractions
- Partnering with the BFH Family Resource Center to facilitate referral to health, social, and developmental resources for families

All of these efficiencies will allow timelier reporting of core and recommended cases from birth years 2018-2020 in the 2022 annual report. With data from three birth years, the advisory board can begin to implement special projects such as grouping types of cases for review to identify recommendations for better surveillance quality or evidence-based interventions to address systemic gaps to improve testing and follow-up for moms and children affected by birth defects.

Summary

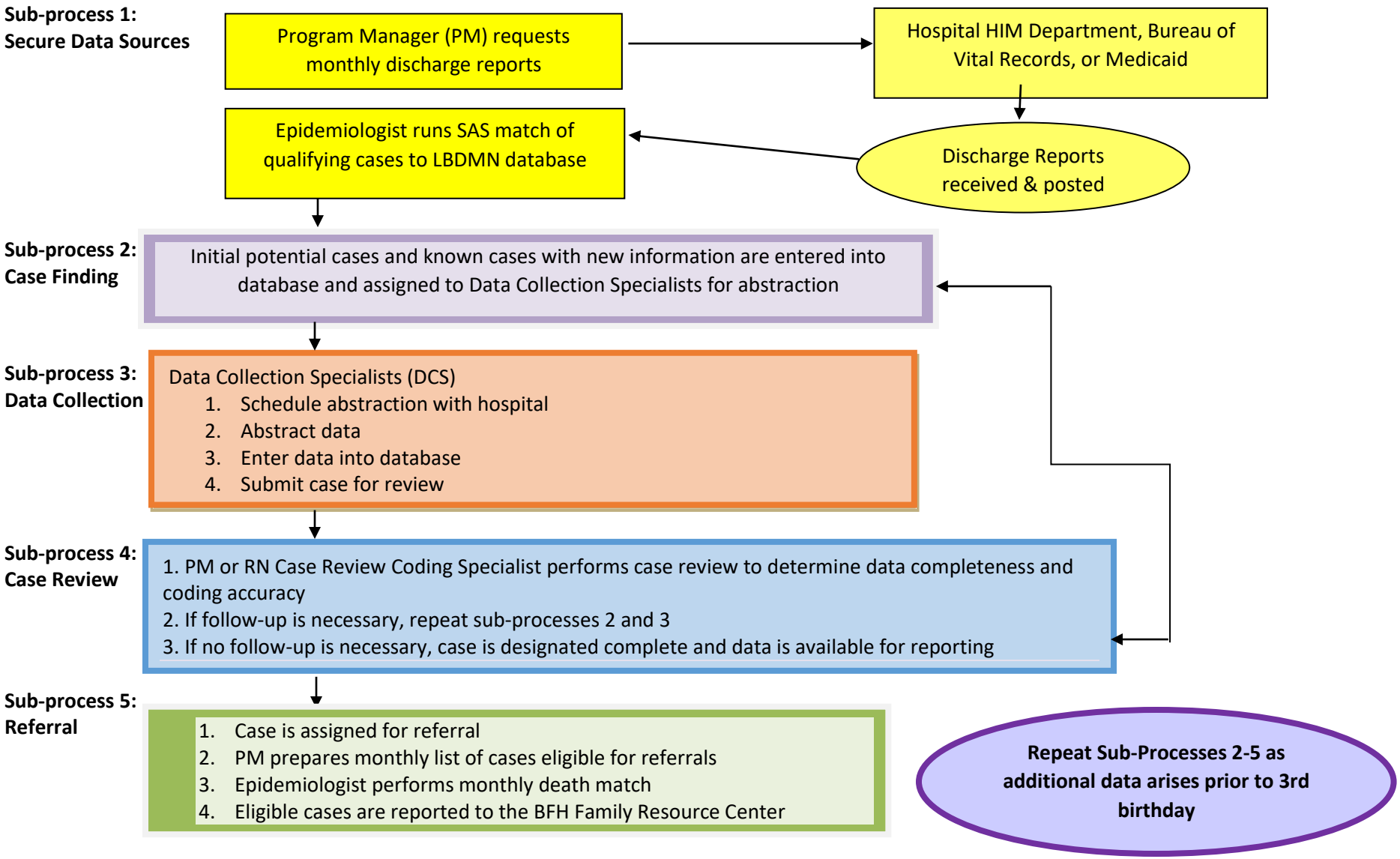
Of 179,510 children born in Louisiana between 2015 and 2017, 5,296 were diagnosed with at least one birth defect, yielding an overall prevalence of 295 per 10,000 live births or 2.95%, which is a slight 0.15% decrease over last year. According to the CDC, the U.S. average is about 3% of all babies born each year. Among Louisiana children with birth defects born in 2015-2017, cardiovascular system defects (about 50%) were the most common, which is a 2% increase over last year.

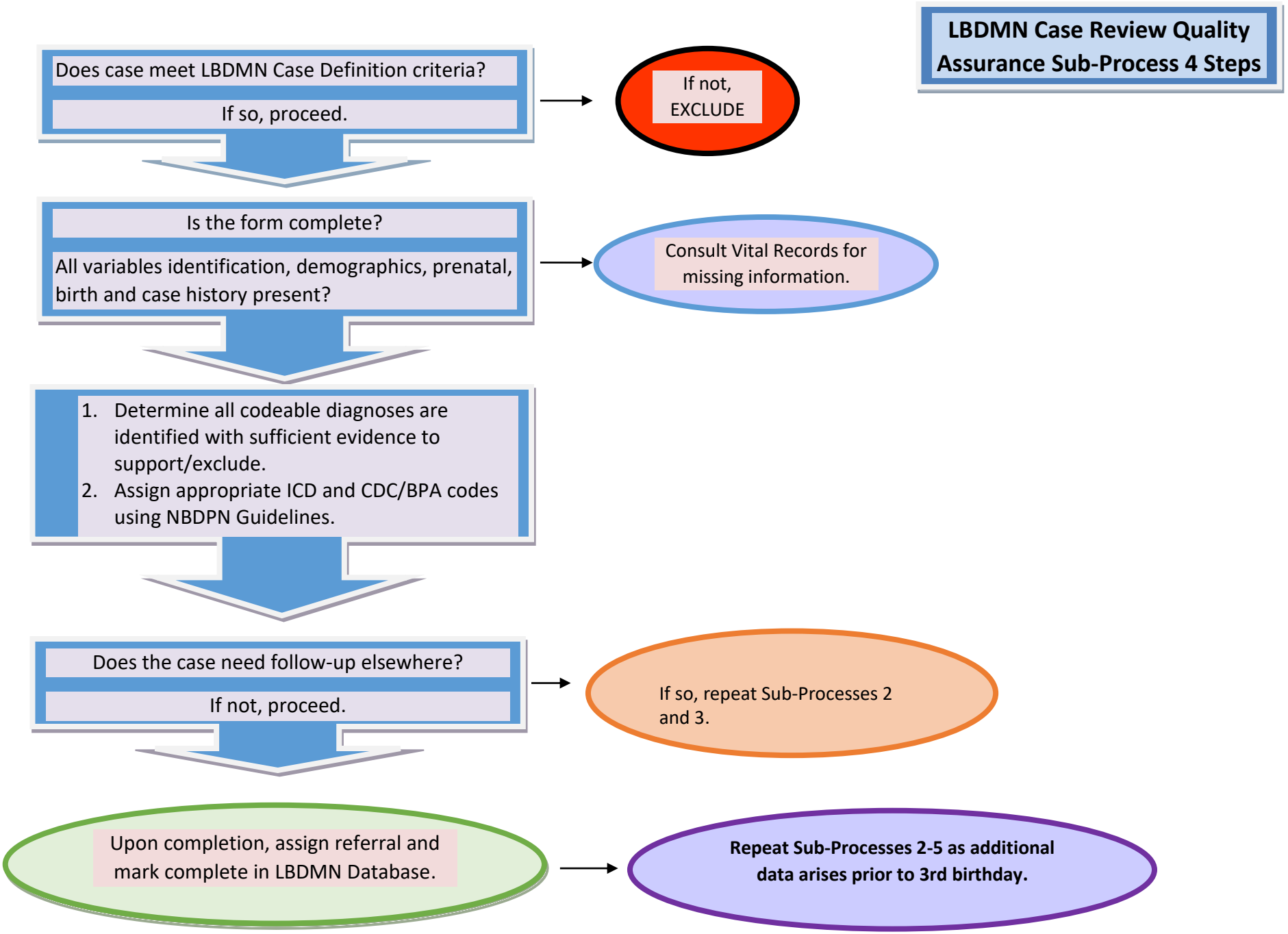
As the established statewide mechanism for tracking and monitoring birth defects, LBDMN incorporates evidence-based public health surveillance best practices and continues to seek opportunities for quality improvement. Recent updates to data collection approaches, automation of processes, increased remote access to medical records, and a partnership to increase timely referrals to services for families are expected to enhance program operations. The program and advisory board are developing protocols for review of preventable cases of birth defects so that this surveillance system can begin to generate actionable recommendations for systems level changes and positively impact the occurrence of preventable birth defects in the future.

LBDMN data are available at partnersforfamilyhealth.org, at the National Birth Defects Prevention Network, from CDC Data & Statistics on Birth Defects, and on the Louisiana Environmental Public Health Tracking Network health data portal to enable analysis, visualization, and reporting. These data can be accessed by environmental and public health practitioners, healthcare providers, community members, policy makers, and others to make data-driven decisions that affect the health of Louisiana citizens.

Appendix

Appendix A: Case Ascertainment/Review/Quality Assurance Process Chart




LBDMN Case Review Quality Assurance Sub-Process 4 Steps
Does case meet LBDMN Case Definition criteria?
If so, proceed.
If not, EXCLUDE
Is the form complete?
All variables identification, demographics, prenatal, birth and case history present?
Consult Vital Records for missing information.
1. Determine all codeable diagnoses are identified with sufficient evidence to support/exclude.
2. Assign appropriate ICD and CDC/BPA codes using NBDPN Guidelines.
Does the case need follow-up elsewhere?
If not, proceed.
If so, repeat Sub-Processes 2 and 3.
Upon completion, assign referral and mark complete in LBDMN Database.
Repeat Sub-Processes 2-5 as additional data arises prior to 3rd birthday.

Appendix B: Birth Defects Codes and Descriptions

ICD-10 CM CODES BY STANDARD LEVEL	DESCRIPTION	2018-2020 CASE DEFINITION	CDC CODES	REFERRAL
CORE				
Q00.0-Q00.1	Anencephalus	CASE FINDING & CODING	740.000 –740.100	NO
Q05.0-Q05.9	Spina bifida without anencephalus	CASE FINDING & CODING	741.000; 741.020 -741.090; 741.900-	YES
Q07.01		CASE FINDING & CODING	741.010	YES
Q07.03 w/o Q00.0 - Q00.1		CASE FINDING & CODING	741.010	YES
Q20.0	Common Truncus	CASE FINDING & CODING	745.000	YES
Q20.3, Q20.5	TGA	CASE FINDING & CODING	745.100-745.120, 745.180-745.190	YES
Q21.2	AVSD	CASE FINDING & CODING	745.600-745.690	YES
Q21.3	TOF	CASE FINDING & CODING	74.20-745.21	YES
Q23.4	HLHS	CASE FINDING & CODING	746.700	YES
Q26.2	TAPVR	CASE FINDING & CODING	747.420	YES
Q35.1 - Q35.9	Cleft palate without Cleft lip	CASE FINDING & CODING	749.000-749.090	YES
Q36.0 - Q36.9	Cleft lip without Cleft palate	CASE FINDING & CODING	749.100-749.190	YES
Q37.0 - Q37.9	Cleft lip with Cleft palate	CASE FINDING & CODING	749.200-749.290	YES
Q71.0 - Q71.9	Limb reduction defects	CASE FINDING & CODING	755.200-755.290	YES
Q72.0 - Q72.9		CASE FINDING & CODING	755.300-755.390	YES
Q73.0 - Q73.8		CASE FINDING & CODING	755.400-755.490	YES
Q79.2	Omphalocele	CASE FINDING & CODING	756.700	YES
Q79.3	Gastroschisis	CASE FINDING & CODING	756.710	YES
Q90.0 - Q90.9	Trisomy 21	CASE FINDING & CODING	758.000-758.090	YES
RECOMMENDED				
Q01.0 - Q01.9	Encephalocele	CASE FINDING & CODING	742.000-742.090	YES
Q02	Microcephaly	CASE FINDING & CODING	742.100	YES
Q04.2	Holoprosencephaly*	CASE FINDING & CODING	742.260	YES
Q11.0 - Q11.2	Anophthalmia/Microphthalmia	CASE FINDING & CODING	743.000-743.100	YES
Q12.0	Congenital cataract*	CASE FINDING & CODING	743.320-743.326	YES
Q13.1	Aniridia	CASE FINDING & CODING	743.420	YES
Q16.0, Q17.2	Anotia/microtia	CASE FINDING & CODING	744.010, 744.210	YES
Q20.1	DORV	CASE FINDING & CODING	745.130-745.150	YES
Q20.4	Single ventricle	CASE FINDING & CODING	745.300	YES
Q21.0	VSD	CASE FINDING & CODING	745.400-745.490	YES
Q21.1	ASD	CASE FINDING & CODING	745.500-745.590	YES 745.510 only
Q22.0, Q22.1	Pulmonary valve atresia and stenosis	CASE FINDING & CODING	746.000, 746.010	YES
Q22.4	Tricuspid valve atresia and stenosis	CASE FINDING & CODING	746.100, 746.106	YES
Q22.5	Ebstein anomaly	CASE FINDING & CODING	746.200	YES
Q23.0	Aortic valve stenosis	CASE FINDING & CODING	746.300	YES

Q25.1	COA	CASE FINDING & CODING	747.100-747.190	YES
Q25.2, Q25.4	IAA	CASE FINDING & CODING	747.215-747.217	YES
Q30.0	Choanal atresia	CASE FINDING & CODING	748.000	YES
Q39.0 - Q39.4	TEF/ EA	CASE FINDING & CODING	750.300-750.350	YES
Q41.0 - Q41.9	Small intestine atresia/stenosis	CASE FINDING & CODING	751.100-751.190	YES
Q42.0 - Q42.9	Rectal and large intestine atresia/ stenosis	CASE FINDING & CODING	751.200-751.240	YES (exclude Imperforate anus)
Q44.2 - Q44.3	Biliary atresia*	CASE FINDING & CODING	751.650	YES
Q54.0 - Q54.9 (not Q54.4)	Hypospadias	CASE FINDING & CODING	752.600-752.607;752.620;752.625-752.627	NO
Q60.0 - Q60.6	Renal agenesis/ hypoplasia	CASE FINDING & CODING	753.000-753.010	YES
Q64.10, Q64.19	Bladder exstrophy	CASE FINDING & CODING	753.500	YES
Q64.12	Cloacal exstrophy	CASE FINDING & CODING	751.550	YES
Q64.2	PUV	CASE FINDING & CODING	753.600	YES
Q66.0, Q66.89	Clubfoot	CASE FINDING & CODING	754.500-754.004; 754.730-754.734	YES
Q75.0	Craniosynostosis*	CASE FINDING & CODING	756.000-756.030	YES
Q79.0, Q79.1	Diaphragmatic hernia	CASE FINDING & CODING	756.610-756.616	YES
Q91.0 - Q91.3	Trisomy 18	CASE FINDING & CODING	758.200-758.295	91% mortality rate
Q91.4 - Q91.7	Trisomy 13	CASE FINDING & CODING	758.100-758.190	91% mortality rate
Q93.81	22q11 deletion*	CASE FINDING & CODING	758.370	YES
Q96.0 - Q96.9	Turner syndrome*	CASE FINDING & CODING	758.600-758.690	YES
LBDMN				
Q87.40	Marfan syndrome, Stickler	CODING	759.860	YES
Q87.81	Alport syndrome	CODING	759.870	YES
Q93.3	Other autosomal deletions	CODING	758.380	YES
Q93.4	Cri du chat syndrome (5p deletion)	CODING	758.310	YES
Q93.59	Other deletions of part of a chromosome	CODING	758.390	YES
Q93.7	Deletions with other complex rearrangements	CODING	758.390	YES
Q93.88	Other microdeletions	CODING	758.380	YES
Q93.89	Other deletions from the autosomes	CODING	758.380	YES
Q97.0	Other conditions due to sex chromosome anomalies	CODING	758.8**	YES
Q97.1	Female with more than three X chromosomes	CODING	758.850	YES
Q97.2	Mosaicism, lines with various numbers of X chromosomes	CODING	758.800, 758.820, 758.830	YES
Q97.8	Other specified sexchromosome abnormalities, female phenotype	CODING	758.810	YES
Q98.4	Klinefelter's syndrome	CODING	758.700, 758.710, 758.790	YES

Q98.5	Karyotype 47,XYY	CODING	758.840	YES
Q98.7	Male with sex chromosome mosaicism	CODING	758.840	YES
Q98.8	Other specified sex chromosome abnormalities, male phenotype	CODING	758.820	YES
Q99.2	Fragile X syndrome	CODING	758.880	YES
Q99.8	Other conditions due to autosomal material	CODING	758.580	YES
Q99.8	Other conditions due to chromosome anomalies	CODING	758.880	YES
Q99.9	Conditions due to anomaly of unspecified chromosome	CODING	758.890	YES

Louisiana Department of Health
628 North Fourth Street, Baton Rouge, Louisiana 70802
(225) 342-9500
www.ldh.la.gov

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