Rhode Island

Rhode Island Birth Defects Program (RIBDP)

Purpose: Surveillance, Referral to Services, Referral to Prevention/Intervention Services
Partner: Hospitals, Environmental Agencies/Organizations, Advocacy Groups, Universities, Community Nursing Services, Early Childhood Prevention Programs, Families
Program status: Currently collecting data
Start year: 2000
Earliest year of available data: 2002
Organizational location: Department of Health (Center for Health Data and Analysis)
Population covered annually: 10,800
Statewide: Yes
Current legislation or rule: Title 23, Chapter 13.3 of Rhode Island General Laws requires the development of a birth defects surveillance, reporting, and information system that will a) describe the occurrence of birth defects in children up to age five; b) detect trends of morbidity and mortality; and c) identify newborns and children with birth defects to intervene on a timely basis for treatment.
Legislation year enacted: 2003

Case Definition
Outcomes covered: All birth defects and genetic diseases
Pregnancy outcome: Livebirths (All gestational ages and birth weights), Fetal deaths - stillbirths, spontaneous abortions, etc. (All gestational ages), Elective terminations (All gestational ages)
Age: Birth up to 5 years
Residence: RI maternal residence

Surveillance Methods
Case ascertainment: Combination of active and passive case ascertainment
Vital records: Birth certificates, Death certificates, Matched birth/death file
Other state based registries: Programs for children with special needs, Newborn hearing screening program, Newborn metabolic screening program, RI has an integrated child health information system called KIDSNET, which links data from 10 programs including: Newborn Developmental Risk Screening, Newborn Bloodspot Screening, Newborn Hearing Screening, Home Visiting, Immunization, etc.
Delivery hospitals: Discharge summaries
Pediatric & tertiary care hospitals: Discharge summaries, Specialty outpatient clinics
Other specialty facilities: Prenatal diagnostic facilities (ultrasound, etc.), Cytogenetic laboratories, Genetic counseling/clinical genetic facilities, Maternal serum screening facilities
Other sources: Physician reports

Case Ascertainment
Conditions warranting chart review in newborn period: Any chart with an ICD-9-CM code 740-759/ICD-10-CM code Q00-Q99, Any chart with a selected list of ICD-9-CM codes outside 740-759/ICD-10-CM codes outside Q00-Q99, All stillborn infants, All elective abortions, All infants in NICU or special care nursery, All prenatally diagnosed or suspected cases, Chart reviews are conducted for infants born at the regional perinatal center and the 4 other maternity hospitals who were identified with an ICD-9-CM code 740-759 and 760.71 or an ICD-10 Q code and other sentinel conditions
Conditions warranting chart review beyond the newborn period: Any infant with a codable defect
Coding: ICD-9-CM/ICD-10-CM

Data Collected
Infant/fetus: Identification information (name, address, date-of-birth, etc.), Demographic information (race/ethnicity, sex, etc.), Birth measurements (weight, gestation, Apgars, etc.), Tests and procedures, Infant complications, Birth defect diagnostic information
Mother: Identification information (name, address, date-of-birth, etc.), Demographic information (race/ethnicity, sex, etc.), Gravidity/parity, Illnesses/conditions, Prenatal care, Prenatal diagnostic information, Pregnancy/delivery complications, Family history

Data Collection Methods and Storage
Data collection: Printed abstract/report submitted by other agencies (hospitals, etc.), Electronic file/report filled out by staff at facility (laptop, web-based, etc.), Electronic file/report submitted by other agencies (hospitals, etc.)
Database collection and storage: Access, Oracle

Data Analysis
Data analysis software: SAS, Access
Quality assurance: Validity checks, Re-abstraction of cases, Double-checking of assigned codes, Comparison/verification between multiple data sources, Data/hospital audits, Timeliness
Data use and analysis: Routine statistical monitoring, Public health program evaluation, Baseline rates, Rates by demographic and other variables, Monitoring outbreaks and cluster investigations, Time trends, Observed vs. expected analyses, Epidemiological studies (using only program data), Needs assessment, Service delivery, Referral, Grant proposals, Education/public awareness, Prevention projects

System Integration
System links: Link to other state registries/databases, KIDSNET, hospital discharge data
System integration: Integrated into KIDSNET for web-based provider reporting

Funding
Funding source: 10% General state funds, 10% MCH funds, 80% CDC grant

Other
Web site: www.health.ri.gov/programs/birthdefects
Surveillance reports on file: 2020 Rhode Island Birth Defects Data Book

Contacts
Samara Viner-Brown, MS
Rhode Island Department of Health
3 Capitol Hl
Providence, RI 2908
Phone: **(401)222-5122**
Email: **samara.vinerbrown@health.ri.gov**

Kristen St. John, MPH
Rhode Island Department of Health
3 Capitol Hl
Providence, RI 2908
Phone: (401)222-5123
Email: Kristen.stjohn@health.ri.gov

DATA TABLES

Rhode Island

Birth Defects Counts and Prevalence 2014 - 2018 (Prevalence per 10,000 Live Births)

	Maternal Race/Ethnicity						
Defect	**White, Non-Hispanic**	**Black, Non-Hispanic**	**Hispanic**	**Asian or Pacific Islander, Non-Hispanic**	**American Indian or Alaska Native, Non-Hispanic**	**Total***	**Notes**
Anencephalus	5	1	0	0	0	6	
	1.6	***2.4***	***0.0***	***0.0***	***0.0***	***1.1***	
Anophthalmia/microphthalmia	2	3	3	0	0	9	
	0.7	***7.2***	***2.2***	***0.0***	***0.0***	***1.7***	
Anotia/microtia	7	0	6	1	0	14	
	2.3	***0.0***	***4.5***	***3.8***	***0.0***	***2.6***	
Aortic valve stenosis	3	1	3	0	0	9	
	1.0	***2.4***	***2.2***	***0.0***	***0.0***	***1.7***	
Atrial septal defect	63	12	39	3	0	131	
	20.8	***28.8***	***29.2***	***11.5***	***0.0***	***24.5***	
Atrioventricular septal defect (Endocardial cushion defect)	10	3	2	0	0	17	
	3.3	***7.2***	***1.5***	***0.0***	***0.0***	***3.2***	
Biliary atresia	2	0	0	0	0	2	
	0.7	***0.0***	***0.0***	***0.0***	***0.0***	***0.4***	
Bladder exstrophy	1	0	0	0	0	1	
	0.3	***0.0***	***0.0***	***0.0***	***0.0***	***0.2***	
Choanal atresia	2	0	3	0	0	6	
	0.7	***0.0***	***2.2***	***0.0***	***0.0***	***1.1***	
Cleft lip alone	13	1	6	0	0	20	
	4.3	***2.4***	***4.5***	***0.0***	***0.0***	***3.7***	
Cleft lip with cleft palate	15	0	11	2	0	30	
	4.9	***0.0***	***8.2***	***7.6***	***0.0***	***5.6***	
Cleft palate alone	15	2	9	1	0	28	
	4.9	***4.8***	***6.7***	***3.8***	***0.0***	***5.2***	
Cloacal exstrophy	0	0	1	0	0	1	
	0.0	***0.0***	***0.7***	***0.0***	***0.0***	***0.2***	
Clubfoot	54	4	23	4	1	92	
	17.8	***9.6***	***17.2***	***15.3***	***35.6***	***17.2***	
Coarctation of the aorta	9	1	4	0	0	19	
	3.0	***2.4***	***3.0***	***0.0***	***0.0***	***3.5***	
Common truncus (truncus arteriosus)	2	1	0	0	0	3	
	0.7	***2.4***	***0.0***	***0.0***	***0.0***	***0.6***	
Congenital cataract	5	0	3	2	0	10	
	1.6	***0.0***	***2.2***	***7.6***	***0.0***	***1.9***	
Congenital posterior urethral valves	2	1	0	0	0	4	1
	1.3	***4.9***	***0.0***	***0.0***	***0.0***	***1.5***	
Craniosynostosis	4	1	2	0	0	8	
	1.3	***2.4***	***1.5***	***0.0***	***0.0***	***1.5***	
Deletion 22q11.2	0	0	0	0	0	2	
	0.0	***0.0***	***0.0***	***0.0***	***0.0***	***0.4***	
Diaphragmatic hernia	3	0	5	1	0	11	
	1.0	***0.0***	***3.7***	***3.8***	***0.0***	***2.1***	
Double outlet right ventricle	5	0	2	0	0	10	
	1.6	***0.0***	***1.5***	***0.0***	***0.0***	***1.9***	
Ebstein anomaly	1	1	0	0	0	3	
	0.3	***2.4***	***0.0***	***0.0***	***0.0***	***0.6***	
Encephalocele	1	0	1	0	0	3	
	0.3	***0.0***	***0.7***	***0.0***	***0.0***	***0.6***	
Esophageal atresia/tracheoesophageal fistula	9	0	2	0	0	11	
	3.0	***0.0***	***1.5***	***0.0***	***0.0***	***2.1***	
Gastroschisis	19	0	12	0	0	31	
	6.3	***0.0***	***9.0***	***0.0***	***0.0***	***5.8***	
Holoprosencephaly	1	0	1	0	0	3	
	0.3	***0.0***	***0.7***	***0.0***	***0.0***	***0.6***	
Hypoplastic left heart syndrome	3	1	3	1	0	10	
	1.0	***2.4***	***2.2***	***3.8***	***0.0***	***1.9***	
Hypospadias	134	27	39	2	2	219	1
	85.9	***132.7***	***58.4***	***14.8***	***137.9***	***80.5***	
Interrupted aortic arch	0	1	0	0	0	1	
	0.0	***2.4***	***0.0***	***0.0***	***0.0***	***0.2***	

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Birth Defects Counts and Prevalence 2014 - 2018 (Prevalence per 10,000 Live Births)

	Maternal Race/Ethnicity						
Defect	**White, Non-Hispanic**	**Black, Non-Hispanic**	**Hispanic**	**Asian or Pacific Islander, Non-Hispanic**	**American Indian or Alaska Native, Non-Hispanic**	**Total***	**Notes**
Limb deficiencies (reduction defects)	7	1	8	0	0	18	
	2.3	***2.4***	***6.0***	***0.0***	***0.0***	***3.4***	
Omphalocele	5	0	4	1	0	12	
	1.6	***0.0***	***3.0***	***3.8***	***0.0***	***2.2***	
Pulmonary valve atresia and stenosis	18	6	5	3	0	36	
	5.9	***14.4***	***3.7***	***11.5***	***0.0***	***6.7***	
Pulmonary valve atresia	6	3	3	1	0	15	
	2.0	***7.2***	***2.2***	***3.8***	***0.0***	***2.8***	
Rectal and large intestinal atresia/stenosis	8	0	11	1	0	21	
	2.6	***0.0***	***8.2***	***3.8***	***0.0***	***3.9***	
Renal agenesis/hypoplasia	14	3	4	0	0	22	
	4.6	***7.2***	***3.0***	***0.0***	***0.0***	***4.1***	
Single ventricle	2	0	0	0	0	4	
	0.7	***0.0***	***0.0***	***0.0***	***0.0***	***0.7***	
Small intestinal atresia/stenosis	14	3	5	3	0	25	
	4.6	***7.2***	***3.7***	***11.5***	***0.0***	***4.7***	
Spina bifida without anencephalus	8	2	8	2	0	21	
	2.6	***4.8***	***6.0***	***7.6***	***0.0***	***3.9***	
Tetralogy of Fallot	12	1	5	0	0	20	
	4.0	***2.4***	***3.7***	***0.0***	***0.0***	***3.7***	
Total anomalous pulmonary venous connection	1	0	1	0	0	2	
	0.3	***0.0***	***0.7***	***0.0***	***0.0***	***0.4***	
Transposition of the great arteries (TGA)	4	0	3	0	0	10	
	1.3	***0.0***	***2.2***	***0.0***	***0.0***	***1.9***	
Dextro-transposition of great arteries (d-TGA)	4	0	1	0	0	5	
	1.3	***0.0***	***0.7***	***0.0***	***0.0***	***0.9***	
Tricuspid valve atresia and stenosis	2	0	0	0	0	2	
	0.7	***0.0***	***0.0***	***0.0***	***0.0***	***0.4***	
Tricuspid valve atresia	2	0	0	0	0	2	
	0.7	***0.0***	***0.0***	***0.0***	***0.0***	***0.4***	
Trisomy 13	4	1	1	0	0	6	
	1.3	***2.4***	***0.7***	***0.0***	***0.0***	***1.1***	
Trisomy 18	12	0	6	0	0	19	
	4.0	***0.0***	***4.5***	***0.0***	***0.0***	***3.5***	
Trisomy 21 (Down syndrome)	44	12	28	2	0	98	
	14.5	***28.8***	***21.0***	***7.6***	***0.0***	***18.3***	
Turner syndrome	2	1	1	1	0	6	2
	1.4	***4.7***	***1.5***	***7.9***	***0.0***	***2.3***	
Ventricular septal defect	135	17	52	9	0	222	3
	44.5	***40.9***	***39.0***	***34.4***	***0.0***	***41.4***	
Total live births	**30,355**	**4,161**	**13,339**	**2,615**	**281**	**53,577**	**4**
Male live births	**15,595**	**2,035**	**6,676**	**1,349**	**145**	**27,220**	
Female live births	**14,760**	**2,125**	**6,663**	**1,271**	**135**	**26,354**	

Rhode Island

Birth Defects Counts and Prevalence 2014 - 2018 (Prevalence per 10,000 Live Births)

	Maternal Age (Years)			
Defect	**Less than 35**	**35+**	**Total***	**Notes**
Gastroschisis	30	1	31	
	6.9	***1.0***	***5.8***	
Trisomy 13	2	4	6	
	0.5	***3.9***	***1.1***	
Trisomy 18	7	12	19	
	1.6	***11.6***	***3.5***	
Trisomy 21 (Down syndrome)	35	61	98	
	8.1	***59.0***	***18.3***	
Total live births	**43,231**	**10,343**	**53,577**	**4**

Notes

1. Data for this condition include male and unknown gender cases only. Prevalence is calculated per 10,000 male live births.
2. Data for this condition include female and unknown gender cases only. Prevalance is calculated per 10,000 female live births.
3. Data for conditions include probable cases.
4. Data for total live births include unknown gender.

General comments

*Data for totals include unknown and/or other.
-Data for conditions include stillbirths and terminations at all gestational ages.