Potential Data Sources

Addressing Congenital Heart Defects: Occurrence, Populations and Risk Factors

Centers for Disease Control and Prevention (CDC) Birth Defects Data and Statistics
http://www.cdc.gov/ncbddd/birthdefects/data.html
Resources documenting the public health burden of birth defects and their complications in the United States. Statistics on the occurrence of some of the more common birth defects and links to additional information and resources.

Congenital Heart Defects
http://www.cdc.gov/Features/HeartDefects/
The CDC works to identify causes and prevention opportunities for birth defects, including congenital heart defects, by applying a public health approach that incorporates three essential elements: surveillance or disease tracking, research to identify causes, and prevention research and programs.

Pregnancy Risk Assessment Monitoring System (PRAMS)
http://www.cdc.gov/prams/
PRAMS is a surveillance project of the CDC and state health departments. PRAMS collects state-specific, population-based data on maternal attitudes and experiences before, during, and shortly after pregnancy. A web-based query system (CPONDER) can be used to access national and state-level PRAMS data. Data sets are available for analysis. There is some variation from state to state. Some states do not participate. For more detailed data requests, also contact your state program. PRAMS data is a potential source for evaluating pregnancy experience regarding maternal risk factors that contribute to congenital heart defects.

National Survey of Children with Special Health Care Needs (NSCSHCN)
http://www.cshcndata.org/
This is a subset of the National Survey of Children's Health, a telephone survey of US households sponsored by the Maternal and Child Health Bureau of the Health Resources and Services Administration. The survey is to assess the prevalence and impact of special health care needs among children. Data are collected on indicators such as condition impact on activity and schooling, health insurance coverage and access. Find national and state profiles by factors such as sex, race and health condition.

Birth Defects Research Part A: Clinical and Molecular Teratology
One issue each year (typically December) is devoted to birth defects surveillance, in collaboration with the National Birth Defects Prevention Network. This issue provides a directory of state birth defects surveillance programs with program descriptions, contacts and state birth defects prevalence rates of selected birth defects. Original articles regarding birth defects surveillance are also included.

June 2011 (Volume 9 Issue 6) is a special issue focusing on congenital heart defects.
Find this issue online at **http://onlinelibrary.wiley.com/doi/10.1002/bdra.v91.6/issuetoc.**

National Birth Defects Prevention Network, Birth Defects Prevention Month 2012
References for statistics regarding prevalence and public health impact of CHDs

Title	*Reference*	*Data source*	*Condition*	*Focus*	*Results*
1 Prevalence of congenital heart defects in Metropolitan Atlanta 1998-2005.	Reller, M.D., et.al., J Pediatr. 2008; 153(6):807-813.	Metropolitan Atlanta Congenital Defects Program 1998-2005	CHDs (3240 cases)	Prevalence 81.4/10,000 live births overall	CHD birth prevalence overall and of specific CHDs. Most common were septal defects. Sample birth prevalence estimates = 27.5 (muscular VSD) and 10.3 (secundum ASD) per 10,000 live births. Least common were complex heart defects. Sample birth prevalence estimates = 4.7 (Tetralogy of Fallot) and 2.3 (Transposition of Great Arteries) per 10,000 live births.
2 CDC. Trends in infant mortality attributable to birth defects—United States, 1980–1995.	MMWR 1998;47:773–8.	CDC: U.S. Public use multiple-cause mortality data and National Vital Health Statistics 1980-1995	CHDs overall and select CHDs Other structural birth defects	Infant mortality (IM) vs. infant mortality due to birth defects (IMBD)	IM overall decline of 39.8%; IMBD decline of 34.2%. CHDs were the single largest contributor to IMBD. The largest specific cause of cardiovascular IMBD, hypoplastic left heart syndrome (HLHS), mortality declined a little (~5%). IMBD for other CHDs declined substantially (e.g., complex CHD - transposition of the great vessels by ~36% and simple CHD - ventricular septal defect by ~61%), probably due to better treatment.
3 Racial difference by gestational age in neonatal deaths attributable to congenital heart defects--United States, 2003-2006	MMWR 2010;59:1208-11.	2003-2006	CHDs	Neonatal mortality (<28 days) and health disparities	Overall similar neonatal mortality due to CHDS for infants of white and infants of black mothers (2.0 vs. 2.1 per 10,000 live births). Among preterm infants: higher for white than for black (6.8 vs. 4.5) Among full term (37-44 weeks ga) infants: slightly lower for white than for black (1.3 vs. 1.5).
4 Hospitalizations for birth defects, 2004. Agency for Healthcare Research and Quality (AHRQ).	Russo, C. A. (Thomson Medstat) and Elixhauser, A. January 2007. Healthcare Cost and Utilization Project HCUP Statistical Brief #24	HCUP 2004 Nationwide Inpatient Sample (NIS).	CHDs Other structural birth defects	Health care costs	Highest aggregate costs were for stays related to cardiac and circulatory congenital anomalies, about $1.4 billion—more than half of all hospital costs for birth defects. Cardiac and circulatory congenital anomalies accounted for more than one-third of all hospital stays for birth defects and had the highest in-hospital mortality rate.

5 Improving the quality of surveillance data on congenital heart defects in the metropolitan Atlanta congenital defects program.	Riehle-Colarusso T, Strickland MJ, Reller MD, Mahle WT, Botto LD, Siffel C, Atkinson M, Correa A. Birth Defects Res A *Clin Mol Teratol*. 2007 Nov;79(11):743-53. PMID:17990334	Metropolitan Atlanta Congenital Defects Program 1968-2003	CHDs (7749 cases) Review; standardized nomenclature	Surveillance	Application of clinical CHD nomenclature improved the clinical accuracy of surveillance data by eliminating normal physiologic variants and obligatory shunt lesions. Classification aggregated specific CHDs into groups appropriate for research and surveillance.
6 The contribution of chromosomal abnormalities to congenital heart defects: a population-based study	Hartman RJ, Rasmussen SA, Botto LD, Riehle-Colarusso T, Martin CL, Cragan JD, Shin M, Correa A *Pediatr Cardiol*. 2011. PMID: 21728077	Metropolitan Atlanta Congenital Defects Program 1994-2005	CHDs (4430 cases) Review; chromosomal anomaly (CA); aneuploidy plus	Etiology and comorbidity Live births and stillbirths	Among 4430 infants with CHDs, 547 (~12%) also had a chromosome anomaly. Prior studies report 9-18% of CHDs associated with chromosomal anomaly. Provides CHDs most frequently (interrupted aortic arch (IAA) type B; atrioventricular spetal defect (AVSD) = 66-70%) and least frequently (heterotaxy; Ebstein anomaly = 2-3%) associated with CA. Provides CAs most frequently observed: tri 21 (~53%), tri 18 (~13%); del 22q11.2 (~12%); and tri13 (~6%). CA was significantly more frequent in those with multiple CHDs, stillbirth and maternal age ≥ 35 yrs (trisomy only).
7 Pulse oximetry screening at 4 hours of age to detect critical congenital heart defects	Sendelbach DM, Jackson GL, Lai SS, Fixler DE, Stehel EK, and Engle WD, *Pediatrics* 2008;122;e815; DOI: 10.1542	University of Texas Southwestern Medical Center, Dallas, Texas	Critical CHDs (severe, complex, cyanotic)	Universal newborn screening via pulse oximetry (POx <96%)	15,233 newborns in newborn nursery screened by POx at 4hrs after birth. Inborn, term and late preterm, born 3-1-2006 through 2-28-2007, largely Hispanic. Four infants with CCHD were diagnosed clinically. POx was low for three of four. No CCHD was detected via POx alone, e.g., with normal clinical examination. Conclusion – POx did not improve detection of CCHD.
8 Pulse oximetry screening as a complementary strategy to detect critical congenital heart defects	Meberg A, Andreassen A, Brunvand L, Markestad T, Moster D, Nietsch L, Silberg IE,Skålevik JE, *Acta Pædiatrica* 2009 DOI:10.1111	Norway (multi-facility collaborative covering all Norwegian newborns)	Critical CHDs (severe, complex, cyanotic)	Universal newborn screening via pulse oximetry (POx <95%)	50,008 of 116057 live born infants screened by POx in the first day of life. Screened infants were more likely to be diagnosed with CCHD before discharge: 44/50 (88%) vs. 37/48 (77%). Two infants who failed POx screening were inadvertently discharged without further investigation.

National Birth Defects Prevention Network, Birth Defects Prevention Month 2012
References for statistics regarding <u>risk factors</u> for CHDs

Article	***Reference***	***Data source***	***Risk factor***	***Risk of CHD [per calculated odds ratios (ORs) and confidence intervals (CIs)]***
9 Maternal smoking and congenital heart defects in the Baltimore-Washington Infant Study.	Alverson CJ, Strickland MJ, Gilboa SM, Correa A. 2011. *Pediatrics* 127(3):e647-53.	Baltimore-Washington Infant Study Group 1981-1989	Smoking tobacco moderate	Adjusted ORs and CIs for associations between maternal smoking during the first trimester and selected CHDs. ORs for certain CHDs: ASD (secundum) = 1.36; pulmonary valve stenosis = 1.35; l-TGA = 1.79; and truncus arteriosis = 1,90.
10 Diabetes mellitus and birth defects.	Correa et al. 2008. *Am. J Obstet Gynecol* 199:237-e1-237.e9.	National Birth Defects Prevention Study. 1997-2003	Diabetes mellitus (pre-existing, pre-gestational) and GDM moderate to strong	Adjusted ORs and CIs for associations between diabetes (DM and GDM) and selected CHDs, both isolated and multiple and for other selected birth defects. ORs for isolated CHDs in association with DM ranged from 1.44 (pulmonary valve stenosis) to 12.36 (Atrial VSD); ORs for multiple defects with DM ranged from 4.17 (ASD, unspecified) to 71.97 (d-TGA). ORs for CHDs in association with GDM were slightly increased (1.10-2.16).
11 Associations between maternal fever and influenza and congenital heart defects.	Oster ME, Riehle-Colarusso T, Alverson CJ, Correa A. *J Pediatr*. 2011 Jun;158(6):990-5.	Baltimore-Washington Infant Study Group 1981-1989	Fever Influenza	There were significant associations between fever and influenza and specific CHDs, namely right-sided obstructive defects (fever: OR, 2.04; 95% CI, 1.27 to 3.27; influenza: OR, 1.75; 95% CI, 1.16 to 2.62) and atrioventricular septal defects in infants with Down syndrome (fever: OR, 1.92; 95%CI, 1.10 to 3.38; influenza: OR, 1.66;95% CI, 1.04 to 2.63). Maternal antipyretic use in the setting of fever or influenza tended to decrease these associations.
12 Association between prepregnancy body mass index and congenital heart defects.	Gilboa SM, Correa A, Botto LD, Rasmussen SA, Waller DK, Hobbs CA, Cleves MA, Riehle-Colarusso TJ; National Birth Defects Prevention Study. 2010. *Am J Obstet Gynecol* 202(1):51.e1-51.e10.	National Birth Defects Prevention Study. 1997-2004 (6440 cases of CHD)	Overweight and Obesity (BMI) moderate	Adjusted ORs and CIs for associations between high BMI ($\geq$25.0 kg/m^2), all CHDs and select CHDs. ORs for all CHDs combined were 1.16, 1.15, and 1.31 for overweight status, moderate obesity, and severe obesity, respectively. Specific CHDs associated with high BMI were conotruncal defects (tetralogy of Fallot), total anomalous pulmonary venous return, hypoplastic left heart syndrome, right ventricular outflow tract (RVOT) defects (pulmonary valve stenosis), and septal defects (secundum atrial septal defect).
13 Preconceptional folate intake and malformations of the cardiac outflow tract.	Scanlon KS, Ferencz C, Loffredo CA, Wilson PD, Correa-Villaseñor A, Khoury MJ, Willett WC. 1998. Baltimore-Washington Infant Study Group. *Epidemiology* 9(1):95-8.	Baltimore-Washington Infant Study Group.	Folic acid intake protective	Maternal folic acid intake was inversely correlated with presence of outflow tract anomalies and positively correlated with absence of outflow tract anomalies.

14 Noninherited risk factors and congenital cardiovascular defects: current knowledge Scientific statement from the American Heart Association Council on Cardiovascular Disease in the Young	Jenkins KJ, Correa A, Feinstein JA, Botto L, Britt AE, Daniels SR, Elixson M, Warnes CA, Webb CL; American Heart Association Council on Cardiovascular Disease in the Young. 2007. *Circulation* 115(23):2995-3014.	Endorsed by American Academy of Pediatrics.	Multiple noninherited risk factors review	A detailed review of noninherted risk factors for CHDs in general and for specific CHDs. Includes reported risk ranges. Includes review of evidence for protective effect of folic acid, multivitamin use in periconceptional period.
15 Maternal treatment with opioid analgesics and risk for birth defects.	Broussard C.S., Rasmussen S.A., Reefhuis J, et.al. *Am J Obstet Gynecol* 2011;204:314.e1-11.	National Birth Defects Prevention Study 1997-2005	Opioids (prescription; pain treatment) moderate	Prior association found between codeine use in early pregnancy and increased risk for CHDs. Study found moderate increase in risk for several specific types of heart defects, but not all types (ORs up to 2.7). Some increase in risk for additional (non cardiac) birth defects also found.

Critical Congenital Heart Defects (CCHD)

The Critical Congenital Heart Defects (CCHD), sometimes called cyanotic heart defects, are severe congenital heart defects requiring surgery or catheter intervention in the first year of life. They present with hypoxemia in most or all cases.

CCHD Lesions:

Heart Defect	Prevalence (per 10,000 live births)*	Hypoxemia	Ductus Arteriosus Dependent
Outflow tract defects			
Tetralogy of Fallot (TOF)	6.1	Most	Few
Transposition of the great arteries (TGA) (D-TGA =dextro; complete transposition of the great arteries; transposition of the great vessels)	4.0	All	Few
Truncus arteriosus (*common truncus*)	1.0	All	None
Total anomalous pulmonary venous connection (TAPVC)	1.2	All	None
Right obstructive defects			
Tricuspid Atresia	0.5	All	Some
Pulmonary atresia, intact septum (*right ventricle hypoplasia, imperforate pulmonary valve and pulmonary artery anomalies*)	0.8	All	All
Left obstructive defects			
Hypoplastic left heart (HLHS)	3.3	All	All
CHDs sometimes presenting with hypoxia			
Coarctation of the aorta	4.7	Some	Some
Aortic arch atresia or hypoplasia (supravalvular aortic stenosis)	1.0	Some	All
Aortic valve stenosis (critical)	1.6	Few	Some
Double-outlet right ventricle	1.7	Some	Some
Ebstein anomaly	0.6	Some	Some
Pulmonic stenosis, atresia (*pulmonary valve atresia, stenosis*)	6.3	Some	Some
Other major heart defects	12.4	Some	Some

Prevalence Rates from the Metropolitan Atlanta Congenital Defects Program. Table adapted from *Role of Pulse Oximetry in Examining Newborns for Congenital Heart Disease: A Scientific Statement From the American Heart Association and American Academy of Pediatrics.* Circulation 2009;120;447-458; DOI: 10.1161 /CIRCULATIONHA. 109.192576. Downloaded from circ.ahajournals.org at MDCH September 23, 2010