Understanding the “Who”,“When”, and “Where”of Severe Congenital Heart Defects among a 10-year Cohort of Iowa Births

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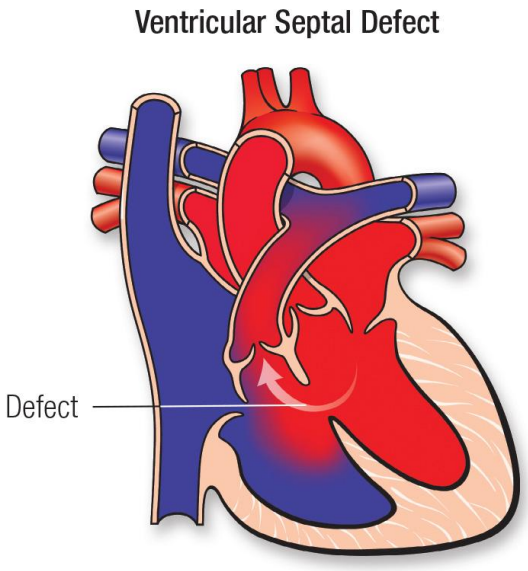
NIH National Heart, Lung, and Blood Institute

Grant # HL-147231

The University of Iowa

What are Congenital Heart Defects (CHDs)?

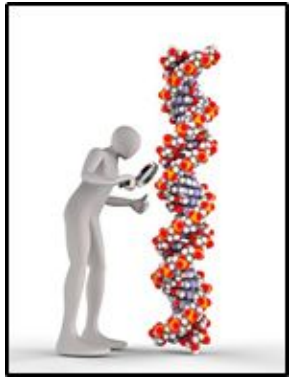
- Major structural defect that is present at birth
- Problem with the formation of the structure of the heart or major heart vessels *in utero*
    - Most commonly a hole between both ventricles of the heart
- Range from defects that self-resolve to those that are lethal
- Some CHDs may not be detected until adulthood



Risk Factors

Multifactorial etiopathogenesis

- Genes
- Environmental exposures (broadly defined)
    - Examples include maternal obesity, diabetes, tobacco use, alcohol use, medication use during pregnancy, pesticides, psychosocial factors (e.g. socioeconomic status, prenatal stress)

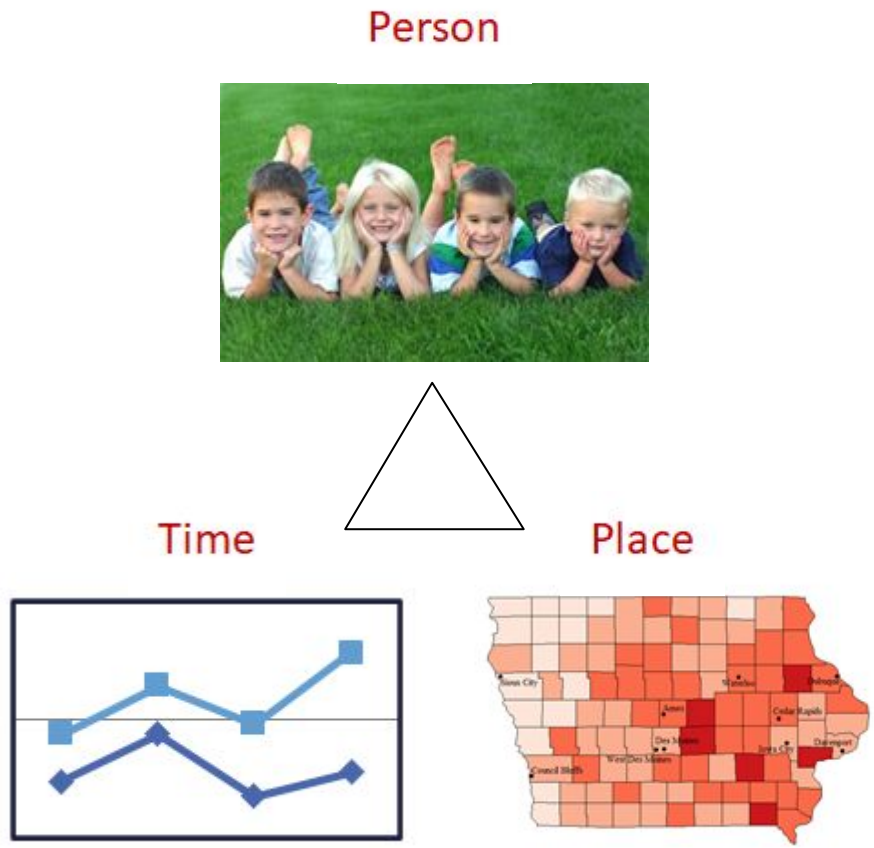


Significance

- Most prevalent group of birth defects in the United States, affecting about 1% of all births
- Leading cause of defect-associated infant mortality, morbidity, and healthcare costs
- CHDs or their sequelae may require care well into adulthood
- As such, CHDs represent a major public health burden across the life span
- To date, lack of population-based surveillance for CHDs across the lifespan in the United States that encompasses data on children, adolescents, and adults living with CHDs

Objectives

- Who
- When
- Where



Methods

Iowa Registry for Congenital and Inherited Disorders

IRCID:

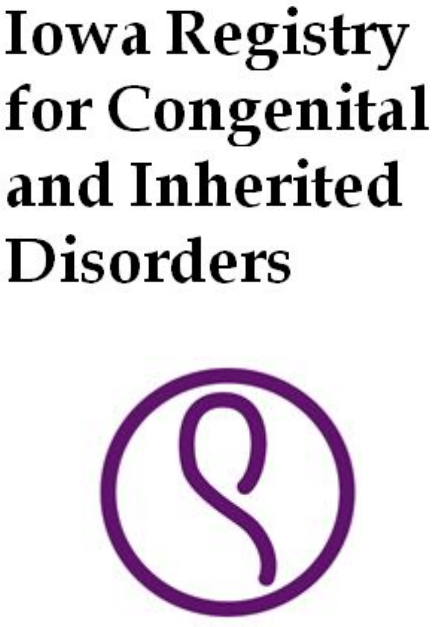
- Established in 1983 (Iowa Department of Public Health)
- Case children are live births, stillbirths, or electively terminated with a least one CHD (mother is Iowa resident at delivery)
- Conducts surveillance in all Iowa hospitals and neighboring states

Case Population:

- Children diagnosed with severe CHDs from IRCID (2010 - 2019)
- n = 776

Comparison population:

- All Iowa live births and fetal deaths (2010-2019)
- n = 406955



Child and Parental Characteristics

Child:

- Sex
- Birth Weight
- Gestational Age
- Birth year
- County location

Mother:

- Age
- Race
- Ethnicity
- Education level
- County location

Father:

- Age
- Race
- Ethnicity

Statistical Analysis

- Descriptive statistics
  - Frequency
  - Percentage
  - Birth Prevalence
  - Prevalence Ratio
- Spatial statistics
  - Moran’s I test statistic
  - Simultaneous autoregressive modeling
  - Mapping

Spatial Statistics

- **Autocorrelation**: the correlation between observations from a single random variable as a function of the time separation between them
- **Spatial autocorrelation**: a function of distance and direction
- **Moran's I test statistic**:
  - Assuming normally distributed data and large sample size
- **Simultaneous autoregressive model (SAR)**:
  - Accounts for spatial autocorrelation
  - Maps the fitted values

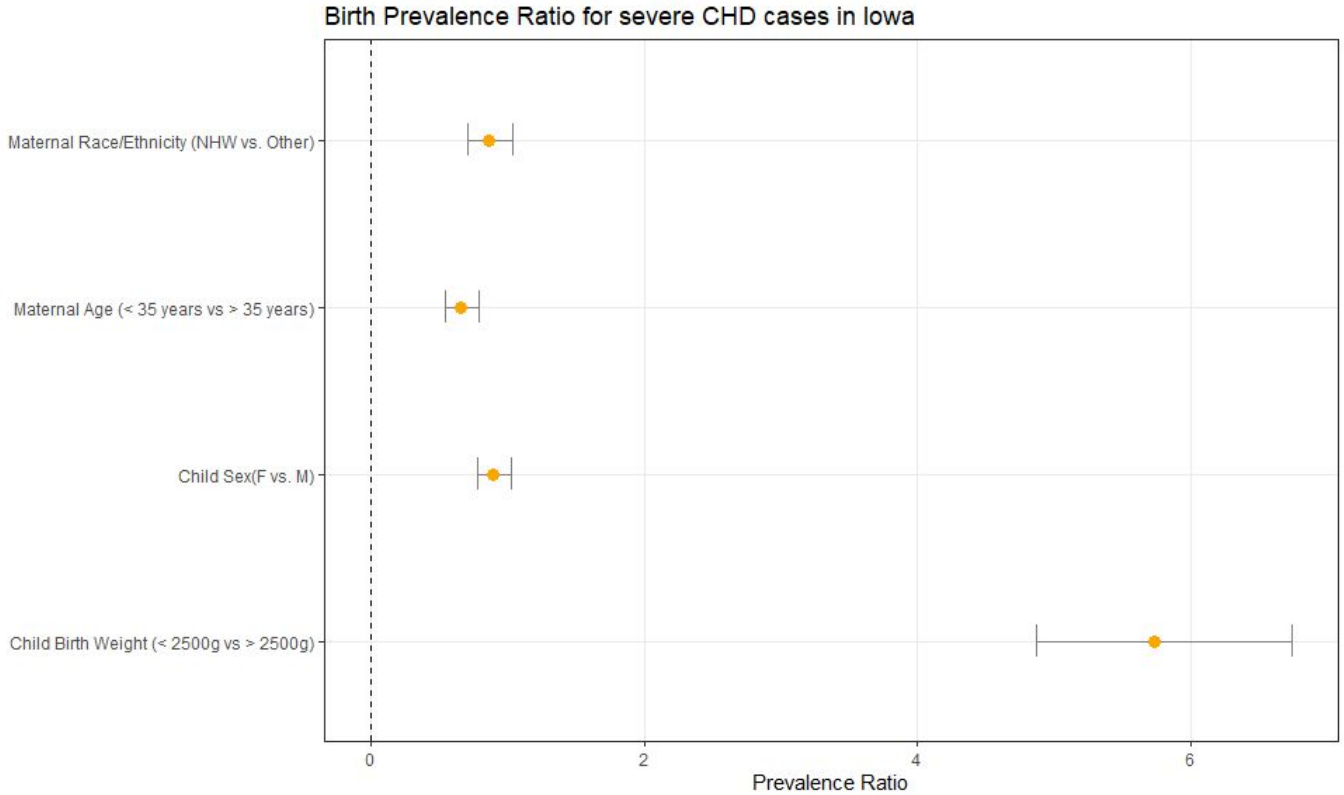
$$z = \frac{I - E(I)}{\sqrt{var(I)}}$$

Results

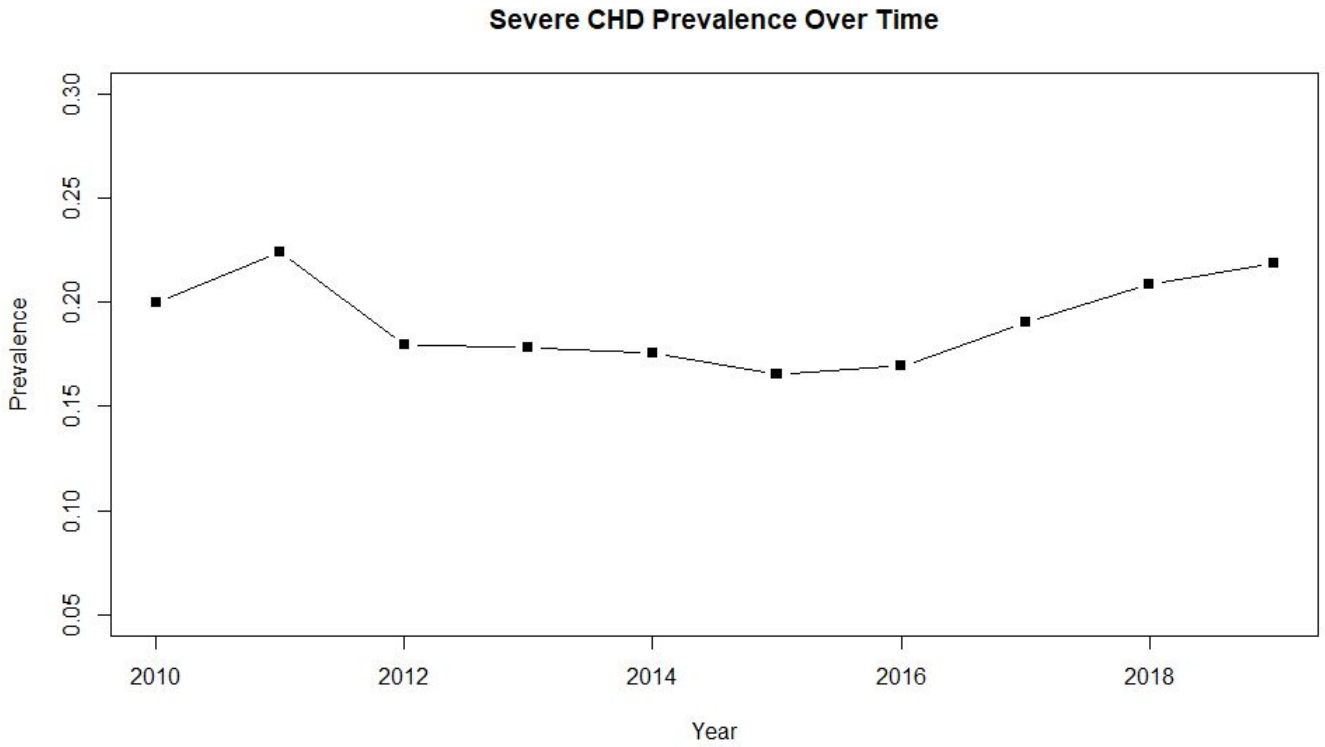
Severe CHDs

- Complete Atrioventricular Canal (CAVC) - 24.5%
- Tetralogy of Fallot (TOF) - 17.4%
- Hypoplastic left heart syndrome (HLHS) - 16.5%
- Double outlet ventricle (DOV) - 10.8%
- Dextro-Transposition of the Great Arteries (dTGA) - 10.2%
- Pulmonary Atresia - 5.3%
- Total anomalous pulmonary venous return (TAPVR) - 3.9%
- Truncus Arteriosus - 3.2%
- Single Ventricle - 2.7%
- Tricuspid Atresia - 2.6%
- Interrupted Aortic Arch (IAA) - 2.3%
- Congenitally corrected transposition of the great arteries (CCTGA) - 0.6%

Descriptive Results

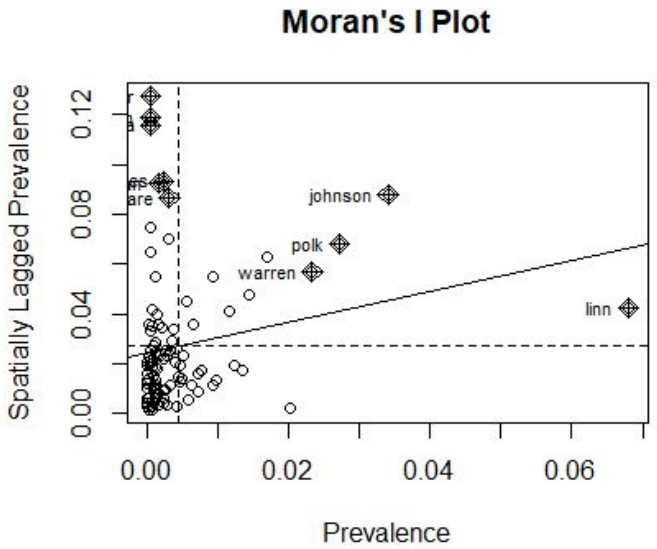


Birth Prevalence Over Time

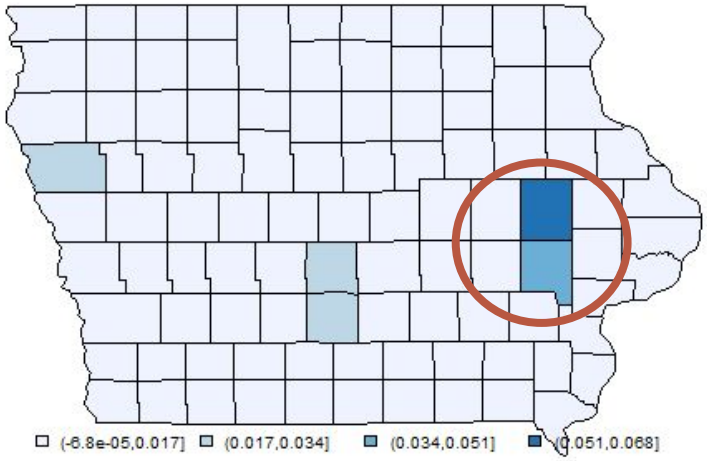


Spatial Results

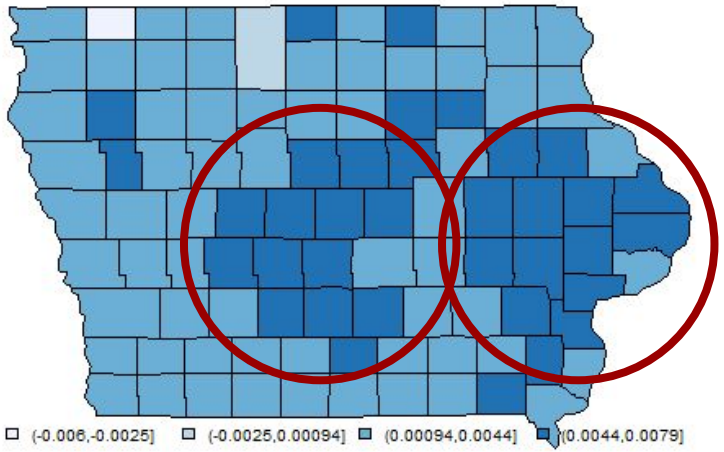
- $\mathbf{H_0}$: observations are spatially independent
- $\mathbf{H_a}$: observations are autocorrelated
- Moran's I statistic result : **0.093 (p-value = 0.014)**
- Using alpha = 0.05, we reject the null hypothesis that the observations are spatially independent (p = 0.014)
- Need to adjust for spatial autocorrelation using SAR model



Spatial Results



Crude



Adjusted

Conclusions

WHO

- Males have higher birth prevalence than females
- Children with mothers with a maternal age below 35 years had higher birth prevalence than children with mothers with a maternal age above 35 years
- Children with mothers of Other race/ethnicities had higher birth prevalence than children with Non-Hispanic White mothers
- Higher birth prevalence with those who have a low birth weight

WHEN

- Cases tended to trend evenly across 2010-2019

WHERE

- Birth prevalence seemed to be high both in urban and rural areas

Snapshot of severe CHD prevalence and spatial distribution across the most recent decade

Future Directions

- Expand study population to children with other types of CHDs to examine range of severity
- Extend follow-up of case children into the fifth decade of life
- Examine residence level spatial analysis into adulthood to improve understanding of care burden across the state
- Comprehensive population-based data for CHDs across the lifespan in the United States that encompasses data on children, adolescents, and adults living with CHDs

Congenital Heart Defect Surveillance across Time And Regions (CHD STAR)

- Diagnosis of severe, shunt, valve, or other CHD
- Residence in Iowa at some time point following diagnosis
- Birthdate on or after January 1, 1965 and on or before December 31, 2019
- Follow-up through December 31, 2019 or until out-migration or death

References

American Heart Association. "Ventricular Septal Defect." Diagram. *American Heart Association*, 2021, www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/ventricular-septal-defect-vsd.

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Thank You/Acknowledgement

- Our project team:
    - Dr. Jake Oleson, Dr. Paul Romitti, Ali Zorn , Dr. Kristin Conway
- Dr. Gideon Zamba
- Scott Cleven and Melissa Jay
- Terry Kirk
- Biostatistics Department

Questions?