Original Article

Academic Outcomes in Children With Congenital Heart Defects

A Population-Based Cohort Study

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Background—Most studies evaluating neurocognitive outcomes in children with congenital heart defects (CHD) have focused on high-risk patients or used specialized, resource-intensive testing. To determine the association of CHD with academic outcomes and compare outcomes according to the severity of CHD, we linked state educational records with a birth defects registry and birth certificates.

Methods and Results—We performed a retrospective cohort study using data from the North Carolina Birth Defects Monitoring Program, North Carolina Department of Public Instruction, and North Carolina Department of Health and Human Services vital records. We performed logistic regression, adjusting for maternal education, race/ethnicity, enrollment in public pre-Kindergarten, and gestational age, to determine the association of CHD with not meeting standards on reading and math end-of-grade examinations in third grade in 2006 to 2012. Of 5624 subjects with CHD and 10 832 with no structural birth defects, 2807 (50%) and 6355 (59%) were linked, respectively. Children with CHD had 1.24× the odds of not meeting standards in either reading or math (95% confidence interval, 1.12–1.37), with 44.6% of children with CHD not meeting standards in at least one of these areas compared with 37.5% without CHD. Although children with both critical and noncritical CHD had poorer outcomes, those with critical CHD were significantly more likely to receive exceptional services compared with the noncritical group (adjusted odds ratio, 1.46; 95% confidence interval, 1.15–1.86).

Conclusions—Children with all types of CHD have poorer academic outcomes compared with their peers. Evaluation for exceptional services should be considered in children with any type of CHD. **(*Circ Cardiovasc Qual Outcomes*. 2017;10:e003074. DOI: 10.1161/CIRCOUTCOMES.116.003074.)**

Key Words: birth certificates ■ child ■ education ■ gestational age ■ heart defects, congenital ■ registries

As infant morbidity and mortality for children with congenital heart defects (CHD) has decreased over the past 40 years, there has been an increased focus on improving the overall quality of life for these children.[1,2] One essential component of improving the lives of these children is understanding their neurocognitive outcomes.[3] Indeed, the American Heart Association in 2012 issued guidelines for the surveillance, screening, evaluation, reevaluation, and management of such outcomes in children with CHD.[4]

To date, however, most studies evaluating neurocognitive outcomes in children with CHD have focused only on high-risk patients or have used specialized, resource-intensive testing. On the basis of the rigorous developmental and psychological testing methods such as the Bayley Scales of Infant Development, Wechsler Intelligence Scale for Children, Psychomotor Development Index, or the Developmental Test of Visual-Motor Integration, we know that children with critical heart defects such as hypoplastic left heart syndrome or transposition of the great arteries who require surgery in infancy are at increased risk of poor neurocognitive outcomes in childhood.[5–14] However, CHD patients requiring surgery in infancy represent only a subset of all CHD patients, and performing such testing on the entire population of children with CHD is not feasible or cost-effective. Furthermore, the results of these tests do not necessarily translate into the real-world outcomes that are of greatest interest to parents, most notably school performance. Thus, there are important knowledge gaps on the neurocognitive outcomes among children with less severe forms of CHD and on the school performance of children with CHD.

By linking state educational records with birth defects registries and birth certificates, we can address these knowledge gaps by conducting large-scale neurocognitive studies in patients with known birth defects.[15–17] The purpose of our study, therefore, was to use state educational records to determine the association of CHD with academic outcomes and

Received June 10, 2016; accepted January 13, 2017.

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WHAT IS KNOWN

- Children with CHD are at risk for poor neurocognitive outcomes. Current American Heart Association guidelines recommend routine neurocognitive screening only for those children with critical CHD, that is, those with cyanosis or who required surgery during infancy.

WHAT THE STUDY ADDS

- Children with CHD have poorer performance on end-of-grade testing in reading and math in the third grade compared with children without birth defects.
- Poor testing performance is seen not only in children with critical CHD but also in those with noncritical CHD.
- Children with noncritical CHD may benefit from better recognition and treatment of potential neurocognitive deficits

to compare such outcomes according to the severity of CHD. We hypothesized that children with CHD would have poorer academic performance than children without CHD, and that children with more severe forms of CHD would have poorer academic performance than children with less severe forms.

Methods

Study Design

We performed a population-based retrospective cohort study using data from the North Carolina Birth Defects Monitoring Program (NCBDMP), the North Carolina Department of Public Instruction (NCDPI), and the North Carolina Department of Health and Human Services vital records. Since 1987, the NCBDMP has collected information about major structural birth defects in live births identified ≤1 year of age via an active, population-based surveillance system.[18] For classification of heart defects, the NCBDMP uses the system developed by the Centers for Disease Control and Prevention based on the British Paediatric Association codes, a system that has been shown to be superior to International Classification of Diseases, Ninth Revision, for the identification of heart defects.[19] The NCDPI collects information on end-of-grade testing, academic promotion, and receipt of exceptional services (individualized educational plans for children with disabilities) for all children in public schools in NC. In NC, end-of-grade testing in reading and math is administered to all public school students in third through eighth grades, with students being required to demonstrate grade-level proficiency in grades 3, 5, and 8 for academic promotion. Because the study used existing data only, it was considered exempt by the North Carolina Department of Public Health Institutional Review Board, and only individuals authorized by the North Carolina Department of Public Health had access to confidential information.

Cohorts

All children born in the years 1998 to 2003 and identified with CHD via International Classification of Diseases, Ninth Revision codes in the NCBDMP were linked to the educational records from NCDPI and birth certificate records from NC. Staff at NCDPI matched records from the NCBDMP and school records using a deterministic linkage algorithm in SAS, using child's first and last name, date of birth, and sex as matching variables. Matches were verified using the last 4 digits of the child's social security number. Children with the known chromosomal anomalies were excluded because these children often have other severe developmental disabilities that would likely impact the outcomes of interest. Cases of CHD were further subclassified as critical or noncritical according to the diagnosis. In a manner consistent with similar analyses in this population, a case was deemed critical if a child had one or more of the following: coarctation of the aorta, d-transposition of the great arteries, double outlet right ventricle, Ebstein anomaly, hypoplastic left heart syndrome, interrupted aortic arch, pulmonary atresia, single ventricle, tetralogy of Fallot, total anomalous pulmonary venous return, tricuspid atresia, or truncus arteriosus.[1] A non-CHD group was generated by a random sampling of NC birth certificates of children born 1998 to 2003 who were not known to have any birth defects, and these records were similarly linked with the educational records from the NCDPI. Reasons for inability to link include migration out of the state, not being in public school in the third grade, taking alternate testing instead of standardized testing, change in name, or death before third grade.

Outcomes of Interest

Our primary outcome of interest was performance, reported as achievement levels, on the reading and math end-of-grade examinations administered in NC public schools at the end of third grade in years 2006 to 2012. Achievement scores were classified as either a level 1 to 2, indicating that the student has limited or partial command of the NC grade-level content matter, or a 3 to 4, indicating a solid or superior command of grade-level content. We dichotomized achievement scores as does not meet grade-level proficiency standard (levels 1 and 2) versus meets grade-level proficiency standard (levels 3 and 4). Performance in the third grade was chosen because (1) this period is the earliest that standardized mathematics and reading comprehension examinations are administered in NC and (2) the performance in the third grade is a well-established predictor of long-term success, including graduating from high school.[20] Our secondary outcomes of interest included retention at the end of third grade and receipt of exceptional services in the third grade.

Statistical Analyses

After describing the distribution of confounders among the cohorts, we estimated the odds of each of our outcomes and 95% confidence intervals among children with CHD compared with children without a structural birth defect using logistic regression in our primary analysis. In subsequent secondary analyses, we stratified our analyses by critical versus noncritical CHD comparing outcomes among those with critical CHD or noncritical CHD versus those without structural birth defects, and those with critical CHD versus those with noncritical CHD. Finally, we modeled our outcomes of interest among those children with and without additional noncardiac defects versus those without any structural birth defects. In each model, we performed a complete case analysis and included the following covariates of interest chosen a priori: maternal education level (obtained from the birth certificate), race/ethnicity (obtained from the birth certificate), enrollment in public pre-Kindergarten (obtained from NCDPI records), and gestational age (obtained from the birth certificate). All analyses were performed using SAS 9.3 (Cary, NC).

Results

Of the 5624 subjects with CHD and 10 832 with no structural birth defects, 2807 (50%) and 6355 (59%) were linked, respectively, to the third-grade educational records and comprise our study cohorts. Among the CHD group, those who did not link with a third-grade educational record were more likely to be Hispanic or non-Hispanic white than those who did link, and the 2 CHD groups were similar in respect to sex, maternal education, and gestational age. In comparing the linked CHD group with the cohort of children without structural birth defects, the 2 groups were similar with respect to

Table 1. Descriptive Characteristics of Children With Congenital Heart Defects and Children Without a Known Structural Birth Defect Born in NC From 1998 to 2003 and Matched With Third-Grade NC School Records From 2006 to 2012

	Congenital Heart Defects (n=2807)	No Structural Birth Defects (n=6355)	*P* Value
Sex			0.66
Male	1423 (51%)	3190 (50%)	
Female	1384 (49%)	3165 (50%)	
Race/ethnicity			<0.001
White, non-Hispanic	1518 (54%)	3684 (58%)	
Black, non-Hispanic	889 (32%)	1812 (29%)	
Hispanic	314 (11%)	573 (9%)	
Other/missing	86 (3%)	286 (5%)	
Maternal education			0.007
Did not complete high school	702 (25%)	1424 (22%)	
Completed high school	2102 (75%)	4919 (77%)	
Missing	3 (0.1%)	12 (0.2%)	
Public pre-Kindergarten			0.16
Yes	164 (6%)	326 (5%)	
No	2643 (94%)	6029 (95%)	
Gestational age, mean (SD)	36.5 (4.2)	38.7 (2.1)	<0.001
Missing	1	2	

sex and percentage enrollment in public pre-Kindergarten. Those with CHD were less likely to be non-Hispanic white, less likely to have mothers who completed high school by the time of delivery, and were born on average 2 weeks earlier than those without CHD (Table 1). Among the CHD cohort, the most common diagnoses were atrial septal defect and ventricular septal defect (Table 2).

In the comparison of those with CHD versus those without any known structural birth defects, those with CHD had poorer outcomes (Table 3). Children with CHD were more likely not to meet standards in both reading and math, with 44.6% of those in the CHD group not meeting standards in at least one of these areas when compared with 37.5% in the non-CHD group. Similarly, those with CHD were more likely to have received exceptional services (20.5% versus 12.5%). Although a higher percentage of children with CHD were retained in the third grade, this did not meet statistical significance.

We then performed separate analyses comparing children with critical CHD and noncritical CHD separately to those without known structural birth defects (Table 4). Of the 2807 with CHD, 463 (16.5%) had critical CHD, and 2344 had only noncritical CHD. Both those with critical CHD and noncritical CHD were more likely than children without structural birth defects to not meet standards in at least reading or math (adjusted Odds ratio [OR], 1.43 and 1.20, respectively) and to have received exceptional services (adjusted OR, 2.24 and 1.51, respectively). Although there were slightly higher

Table 2. Phenotypes of Congenital Heart Defects Among Children With Third-Grade Reading or Math Test Scores in NC Between 2006 and 2012

	n=2807* (%)
Aortic stenosis	72 (2.6)
Atrial septal defect	1554 (55.4)
Atrioventricular septal defect	34 (1.2)
Coarctation of aorta	143 (5.1)
Double outlet right ventricle	48 (1.7)
Ebstein anomaly	9 (0.3)
Hypoplastic left heart syndrome	33 (1.2)
Interrupted aortic arch	7 (0.3)
Pulmonary atresia	38 (1.4)
Pulmonary stenosis	320 (11.4)
Single ventricle	20 (0.7)
Tetralogy of Fallot	157 (5.6)
Total anomalous pulmonary venous return	15 (0.5)
Transposition of great arteries	61 (2.2)
Tricuspid atresia	23 (0.8)
Truncus arteriosus	24 (0.9)
Ventricular septal defect	1327 (47.3)

*Children may have >1 congenital heart defects.

percentages of children with critical CHD and noncritical CHD being retained in the third grade when compared with those without any structural birth defects, these differences did not meet statistical significance. When we compared those with critical CHD to those with noncritical CHD, we found that those with critical CHD were slightly more likely to fail reading or math and much more likely to receive exceptional services. There was no difference in the third-grade retention.

Table 3. Third-Grade Educational Outcomes for Children With Congenital Heart Defects Versus Children Without a Known Structural Birth Defect

	No Structural Birth Defects (ref)	Congenital Heart Defect	OR* (95% CI)
End-of-grade tests: does not meet standards			
Reading	n=6302; 31.3%	n=2780; 39.9%	1.38 (1.21–1.53)
Math	n=6326; 21.1%	n=2798; 25.5%	1.14 (1.01–1.28)
Either	n=6341; 37.5%	n=2803; 44.6%	1.24 (1.12–1.37)
Both	n=6287; 14.8%	n=2775; 20.8%	1.37 (1.20–1.56)
Third-grade retention	n=6341; 2.0%	n=2803; 2.8%	1.31 (0.97–1.79)
Receipt of exceptional services	n=6341; 12.5%	n=2803; 20.5%	1.64 (1.44–1.86)

Sample size varied for each comparison and is indicated by the n in each cell. CI indicates confidence interval; and OR, odds ratio.

*Adjusted for maternal education, race/ethnicity, public pre-Kindergarten enrollment, and gestational age.

Table 4. Third-Grade Educational Outcomes for Children With Critical Versus Noncritical Congenital Heart Defects

	No Structural Birth Defects	Critical CHD	Noncritical CHD	Critical CHD vs No Structural Birth Defects	Noncritical CHD vs No Structural Birth Defects	Critical CHD vs Noncritical CHD
				OR* (95% CI)	OR* (95% CI)	OR* (95% CI)
End-of-grade tests: does not meet standards						
Reading	n=6302; 31.3%	n=456; 40.8%	n=2324; 39.8%	1.59 (1.29–1.96)	1.34 (1.19–1.50)	1.18 (0.95–1.47)
Math	n=6326; 21.1%	n=461; 28.1%	n=2337; 25.0%	1.48 (1.18–1.86)	1.07 (0.95–1.22)	1.38 (1.08–1.75)
Either	n=6341; 37.5%	n=463; 45.6%	n=2340; 44.4%	1.43 (1.17–1.76)	1.20 (1.08–1.34)	1.19 (1.12–1.47)
Both	n=6287; 14.8%	n=454; 23.3%	n=2321; 20.4%	1.82 (1.43–2.32)	1.28 (1.11–1.47)	1.41 (1.09–1.82)
Third-grade retention	n=6341; 2.0%	n=463; 2.8%	n=2340; 2.8%	1.44 (0.80–2.60)	1.29 (0.93–1.80)	1.10 (0.59–2.03)
Exceptional services	n=6341; 12.5%	n=463; 25.1%	n=2340; 19.6%	2.24 (1.79–2.81)	1.51 (1.87–1.73)	1.46 (1.15–1.86)

Sample size varied for each comparison and is indicated by the n in each cell. CHD indicates congenital heart defects; CI, confidence interval; and OR, odds ratio.
*Adjusted for maternal education, race/ethnicity, and public pre-Kindergarten enrollment.

In our final analyses, we subdivided those with CHD into those without any other structural birth defects and those with other noncardiac defects (Table 5). Of the 2807 with CHD, 2215 did not have any additional structural birth defects and 592 did (21.1%). Compared with those without any structural birth defects, both groups were more likely not to meet standards in reading or math (adjusted OR, 1.19 and 1.48) and to receive exceptional services (adjusted OR, 1.46 and 2.40) with the larger effect sizes in the group with additional noncardiac defects. Those with only CHD were not more likely than children without any known structural birth defects to be retained in third grade, but those with CHD and additional structural birth defects were (adjusted OR, 1.73). When we compared those with CHD with additional noncardiac defects to those with only CHD, we found that those with additional noncardiac defects were slightly more likely to fail reading or math and much more likely to receive exceptional services. There was no difference in the third-grade retention.

Discussion

In this study, the largest of its kind to study educational outcomes in children with CHD, we found that children with CHD have poorer performance on end-of-grade testing in reading and math in the third grade and have higher receipt of exceptional services. Interestingly, this poorer performance in reading and math was seen in both the critical and noncritical CHD cohorts, yet children with noncritical CHD were less likely to receive exceptional services than those with critical CHD.

Although this study confirms the findings of previous studies that children with critical heart defects have poorer neurocognitive outcomes than children without birth defects, it raises the notion that children with milder defects may, too, have significant challenges. Historically, it has been thought that the majority of children with mild or moderate CHD would not have any developmental disabilities.[13] Indeed, in the 2012 American Heart Association guidelines for the evaluation and management of neurodevelopmental outcomes in

Table 5. Third-Grade Educational Outcomes for Children With Congenital Heart Defects With and Without Other Noncardiac Birth Defects

	No Structural Birth Defects	CHD With Other Noncardiac Defects	CHD Only	CHD With Noncardiac Defects vs No Structural Birth Defects	CHD Only vs No Structural Birth Defects	CHD With Noncardiac Defects vs CHD Only
				OR* (95% CI)	OR* (95% CI)	OR* (95% CI)
End-of-grade tests: does not meet standards						
Reading	n=6302; 31.3%	n=583; 44.3%	n=2197; 38.8%	1.70 (1.39–2.05)	1.32 (1.18–1.47)	1.31 (1.07–1.59)
Math	n=6326; 21.1%	n=590; 28.7%	n=2208; 24.7%	1.36 (1.10–1.68)	1.08 (0.95–1.23)	1.24 (1.00–1.54)
Either	n=6341; 37.5%	n=590; 48.3%	n=2213; 43.6%	1.48 (1.22–1.79)	1.19 (1.06–1.33)	1.24 (1.02–1.51)
Both	n=6287; 14.8%	n=583; 24.7%	n=2192; 19.8%	1.71 (1.36–2.15)	1.28 (1.11–1.47)	1.36 (1.08–1.71)
Third-grade retention	n=6341; 2.0%	n=590; 3.4%	n=2213; 2.7%	1.73 (1.03–2.93)	1.21 (0.87–1.70)	1.24 (0.73–2.11)
Exceptional services	n=6341; 12.5%	n=590; 28.7%	n=2213; 18.3%	2.40 (1.95–2.97)	1.46 (1.27–1.68)	1.72 (1.39–2.14)

Sample size varied for each comparison and is indicated by the n in each cell. CHD indicates congenital heart defects; CI, confidence interval; and OR, odds ratio.
*Adjusted for maternal education, race/ethnicity, and public pre-Kindergarten enrollment.

children with CHD, the only heart defects deemed to place a child at high risk for a developmental disorder or disability were those with cyanosis or that required surgery during infancy.[4] In our study, there were similar rates of not meeting standards in reading or math (≈45%) between the critical and noncritical CHD groups and similar rates of being retained in the third grade (≈3%). However, 25% of the children with critical CHD were receiving exceptional services, but only 20% of those with noncritical CHD were receiving such services. It seems that the academic challenges faced by children with noncritical CHD may be underappreciated.

Previous studies using educational records to examine outcomes in children with CHD have found similar results to ours. In a study of 256 patients in Arkansas, Mulkey et al[21] found that children with CHD had lower scores on school-age achievement tests and an 8-fold increase in the receipt of special education services. As this study was limited to children who had surgery in the first year of life, comparisons on outcomes for children with milder forms of CHD are not possible. In a recent Centers for Disease Control and Prevention study of the use of special education services among children in Metropolitan Atlanta, Riehle-Colarusso et al[22] found that children with CHD were 50% more likely to receive such services when compared with children without birth defects, with children with critical CHD having only a slightly higher likelihood when compared with those with noncritical CHD. This study did not examine school testing or grade retention. Finally, in an evaluation of children in Georgia who had undergone surgery for CHD, Oster et al[23] found that children with a history of CHD surgery had poorer performance on math and social studies testing than their peers without CHD, but that there was no difference between those in the higher surgical risk categories compared with those in the lower.

The rationale as to why children with CHD may be at risk for poor neurocognitive outcomes is complex. There are many theories to explain this increased risk, including hypoxia, use of cardiac bypass, and type of anesthesia, with these effects particularly pronounced in the immature neonatal brain.[12] However, children with milder forms of CHD do not typically have hypoxia or surgical risk factors in infancy. What children with noncritical CHD may share with their critical CHD counterparts, however, is a similar genetic predisposition for certain outcomes. The impact of genotype on survival and clinical outcomes has been well established.[24–26] Now, however, it has been suggested that certain genotypes may be associated not only with CHD but also with neurodevelopment. Homsy et al[27] studied the exome sequencing of 1213 CHD parent–offspring trios and found that there were shared genetic contributions for CHD and neurodevelopmental disabilities. Similar genetic predisposition to certain neurodevelopmental outcomes among differing cardiac phenotypes may help explain our findings.

This study is not without its limitations. First, our study is limited to children who attend public schools. Children in private schools or home schools are not tracked via these educational methods, and we thus have no data on that subset of the population. Self-selection of public school by families of children with CHD for the purposes such as obtaining exceptional services or because of financial constraints already on these families from chronic illness may skew our results. Second, we do not have information on children who are not able to take the end-of-grade testing. Children who are not able to take the standard end-of-grade testing may be placed in alternative testing. Although we know what portion during this timeframe underwent alternate testing (11% of children with CHD and 3% of children with no structural birth defects), data on the outcomes of such testing are not available, and thus these children were not included in our cohorts. Finally, the NCBDMP collects information on the diagnosis of CHD, but we have no information on the surgical interventions or clinical outcomes. Thus, we are not able to investigate whether certain types of treatments, surgeries, or other interventions may be associated with educational outcomes; such investigations are certainly warranted in future studies.

By linking 3 large databases in NC, we were able to address important knowledge gaps on the neurocognitive outcomes of children with CHD. These children face significant challenges in school performance, and these challenges are not limited to only that subset with critical CHD. Parents and providers of children with all types of CHD should be cognizant of the academic obstacles these children may face and should consider evaluations for exceptional services.

Sources of Funding

Funding for this project was provided via an award from the National Institutes of Health National Heart, Lung, and Blood Institute Pediatric Heart Network Clinical Scholars program.

Disclosures

None.

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Circ Cardiovasc Qual Outcomes. 2017;10:
doi: 10.1161/CIRCOUTCOMES.116.003074
Circulation: Cardiovascular Quality and Outcomes is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 1941-7705. Online ISSN: 1941-7713