Distribution Agreement

In presenting this thesis or dissertation as a partial fulfillment of the requirements for an advanced degree from Emory University, I hereby grant to Emory University and its agents the non-exclusive license to archive, make accessible, and display my thesis or dissertation in whole or in part in all forms of media, now or hereafter known, including display on the world wide web. I understand that I may select some access restrictions as part of the online submission of this thesis or dissertation. I retain all ownership rights to the copyright of the thesis or dissertation. I also retain the right to use in future works (such as articles or books) all or part of this thesis or dissertation.

Signature:

Rachel Schwarz

Date

Isolated, Non-Syndromic Craniosynostosis and Special Education Use in Metropolitan Atlanta

By

Rachel Schwarz Master of Public Health

Epidemiology

Penelope P. Howards, Ph.D Committee Chair

Suzanne Gilboa, Ph.D Committee Member

Isolated, Non-Syndromic Craniosynostosis and Special Education Use in Metropolitan Atlanta

By

Rachel Schwarz

B.S. Miami University 2014

Thesis Committee Chair: Penelope P. Howards, Ph.D

An abstract of A thesis submitted to the Faculty of the Rollins School of Public Health of Emory University in partial fulfillment of the requirements for the degree of Master of Public Health in Epidemiology 2016

Abstract

Isolated, Non-Syndromic Craniosynostosis and Special Education Use in Metropolitan Atlanta

By Rachel Schwarz

Background: Craniosynostosis (CS) is known to result in delayed neurodevelopment. However, there are no population-based data on the utilization of special education services among children with CS compared to children without birth defects. Methods: We linked data from the Metropolitan Atlanta Congenital Defects program and live birth certificates to create a cohort of children born from 1989-2004 with CS and children born with no major birth defects. To identify children receiving special education services, we linked birth certificate number to the Special Education Database of Metropolitan Atlanta from 1994-2012. Available data on special education exceptionalities only included the primary exceptionality assigned upon the most recent year of enrollment in special education. We used logistic regression to calculate odds ratios (ORs) and 95% confidence intervals (CIs) comparing the odds of receiving special education services among children with CS to the odds of receiving special education services among children in the same birth cohort with no major birth defects. We also assessed the association between CS and common specific special education exceptionalities. Results: Among all children with CS (n=217), 19.4% were enrolled in special education for at least one year between ages 3 through 10, while only 10.7% of children with no major birth defects (n=6,059) were in special education. Children with CS had nearly twice the odds (adjusted OR: 1.74 [95% CI: 1.19, 2.54]) of being enrolled in special education compared with children without any major birth defects. Children with sagittal CS had nearly twice the odds (1.89 [1.09, 3.27]) and children with metopic CS had nearly four times the odds (4.04 [2.08, 7.87]) of being enrolled in special education compared with children with no major birth defects. **Conclusion:** Children with CS receive special education services more than children without birth defects. These results can better prepare providers, educators, and parents to help children with CS seek out early intervention special educational services. Additional work to understand longer-term developmental outcomes among children with CS is needed.

Isolated, Non-Syndromic Craniosynostosis and Special Education Use in Metropolitan Atlanta

By

Rachel Schwarz

B.S. Miami University 2014

Thesis Committee Chair: Penelope P. Howards, Ph.D

A thesis submitted to the Faculty of the Rollins School of Public Health of Emory University in partial fulfillment of the requirements for the degree of Master of Public Health in Epidemiology 2016

Chapter 1	1
Background	1
Types of Craniosynostosis	
Etiology of Craniosynostosis	2
Craniosynostosis and Neurodevelopment	2
Special Education Services and Exceptionalities	
Other Birth Defects and Special Education Services	5
Craniosynostosis and Special Education Services	
References	7
Chapter 2	9
Introduction	9
Methods	
Study Population	
Receipt of Special Education Services	
Analysis	
Results	
Discussion	
Conclusions	17
References	
Table 1	
Figure 1	
Table 2	21
Table 3	
Table 4	

Table of Contents

Chapter 1

Background

It is estimated that birth defects affect 3% of all births in the United States.¹ Craniosynostosis (CS) is a birth defect causing the premature fusion of one or more cranial sutures, resulting in altered growth of the skull. The prevalence of CS is estimated at between 1 in 2,100 to 1 in 2,500 live births in the US.^{2,3} Children born with CS have a 15-year survival rate of 97.6% (95% CI: 84.3-99.7).⁴ Craniosynostosis results in delayed neurodevelopment.^{5,6} Such delays in neurodevelopment might be manifested by increased participation in special education compared to children without birth defects; however, there is no available literature on craniosynostosis and the receipt of special education. The purpose of this thesis was to investigate this hypothesis using population-based data from metropolitan Atlanta, Georgia.

Types of Craniosynostosis

CS is a birth defect in which there is premature fusion of one or more cranial sutures, resulting in altered growth of the skull; the head may be triangular, broad and flattened, or long and narrow.⁷ CS can occur as an isolated (non-syndromic) defect, a non-syndromic defect with other accompanying defects or abnormalities, or as part of a genetic syndrome accompanied by other congenital anomalies involving midface retrusion, limb abnormalities, and brain anomalies.⁸ Recognized genetic syndromes include Apert, Crouzon, Pfeiffer, Muenke, and Saethre-Chotzen syndromes. It is estimated that syndromic cases of CS account for 25-40% of all CS cases and isolated cases account for 60-75% of all CS cases, ^{9,10} CS cases are classified by the location of the fused cranial suture(s): sagittal, coronal, metopic, lambdoid, or multiple sutures. The sagittal suture is affected in 40-55% of cases, the

coronal suture in 20-25% of cases, the metopic suture in 14% of cases, the lambdoid suture in 3-5% of cases, and complex, multi-sutures in 5% of cases.¹¹

Etiology of Craniosynostosis

Although the etiology of non-syndromic CS is unknown, it has been associated with male sex,¹² white race/ethnicity,¹² low birth weight and preterm delivery,¹² maternal residence at high altitude,¹³ maternal use of nitrosatable drugs during pregnancy,¹⁴ maternal use of fertility treatments,¹⁵ and heavy maternal smoking during pregnancy.¹⁶ It is theorized that genetics may be a predisposing factor, because CS carries a sibling recurrence risk of 2% for sagittal and metopic synostosis, 5% for unicoronal synostosis, and 10% for bicoronal and complex synostosis.⁹

Craniosynostosis and Neurodevelopment

Craniosynostosis may increase intracranial pressure. It has been reported that average intracranial pressure in infants and newborns ranges from 0.7-5.1 mmHg and reaches 15 mmHg by adulthood.¹⁷ Research assessing intracranial pressure in those with CS has found that, in small samples (n=41 and 13), approximately 93% of infants and children presented with higher than average intracranial pressure, and near 20% had intracranial pressure greater than 15 mmHg.^{17,18}

The hypothesis that these increases in intracranial pressure among infants and children with CS may result in developmental delays may have been first motivated by neurodevelopmental outcomes in preterm infants with hydrocephalus, a condition characterized by accumulation of cerebrospinal fluid resulting in elevated intracranial pressure. In a sample of 68 infants with hydrocephalus, 25% showed mild developmental delay as measured by the Denver Developmental Screening Test (DDST) after 5 years of follow up.¹⁹

A number of studies have assessed the neurodevelopment and intellectual functioning of children with CS through IQ measurement and test administration. It was reported that school-age children with CS achieved significantly lower scores, 3.9-4.2 points less, on measures of IQ and math achievement than demographically similar children without CS.³ On the Preschool Language Scale (PLS-3), the odds of being delayed on receptive and expressive communication scales were twice as high in children with CS compared to children without CS, adjusted for familial socioeconomic status, age at assessment, race/ethnicity, maternal IQ, and recruitment site.⁴ Factors associated with the extent of intellectual disability in children with CS include severity of skull deformity, subtype of craniosynostosis, genetics, age at surgery, and age at evaluation.²⁰ There is no available literature that tracks children born with CS into adulthood to assess long term developmental outcomes.

Special Education Services and Exceptionalities

Children are ascertained for special education services and exceptionalities after an educational assessment process has been performed by the school system when children are between 3 and 10 years of age. Once a child has been determined eligible for special education services and assigned an exceptionality, the school system develops and implements an individualized education program (IEP) that addresses the specific learning needs of the child. Special education exceptionalities include mild to severe intellectual disability, profound intellectual disability, behavioral disorders, learning disorders, orthopedic impairments, hearing loss, deafness, vision loss, blindness, severe emotional disorder, significant developmental delay, and other health problems.²¹ Services provided by educational institutions can include speech-language pathology, audiology services, psychological services, physical and occupational therapy, therapeutic recreation, mobility

services, school health services, social work services, and parent counseling and training.²² In 2003, about 4.2 million children received special education or early intervention services, with children in the West and South being less likely to receive these services compared to children in the Northeast.²³

Special educations services and exceptionalities seem to result in small, but significant gains in learning-related behavior.²⁴ Learning-related behavior includes listening and following directions, working independently, participating in groups, and organizing work materials. Analyses of nationally-representative data indicate that receipt of special education services has either a negative or statistically non-significant impact on children's learning, but teacher ratings indicate a significant positive effect on children's learning-related behaviors by 0.9 to 0.14 points on a 4 point scale.²⁴ Long-term follow up of children's learning-related behaviors from kindergarten to sixth grade indicates that teacher ratings of learning-related behaviors can be an important indicator of later academic success.²⁵

Similarly, special educations services have resulted in gains in academic performance. Analysis of 767,763 students from The University of Texas at Dallas' Texas Schools Project suggests that one year of special-education improves performance by 0.1 standard deviations on average, or an increase 3-4 percentile points. This gain in performance closes over 10% of the achievement gap between those identified with learning disabilities and regular education students.²⁶ Furthermore, children with learning disabilities who remain in special education for at least two years have fives times greater mathematics test score gains compared with similar children who remain in special education for one year or less.²⁶

Other Birth Defects and Special Education Services

In 2012, 6.4 million children and youth received special education services, corresponding to 13 percent of total public school enrollment.²⁷ Between 2009 and 2010, 37.5% of the total special education population had a diagnosed learning disorder, 21.8% had a speech or language impairment, 5.7% had a developmental delay, and 7.1% had an intellectual disability.²⁸ Many studies have assessed special education exceptionalities and receipt of special education in relation to birth defects. In children with an orofacial cleft, 25.9% were in special education for at least one year between ages 3 through 10 compared to 8% of children without a major birth defect, with the most common exceptionalities being severe intellectual disability, speech and language disorder, and orthopedic impairment. Approximately 14% of the children with an orofacial cleft who used special education had significant developmental delay, which was over five times more likely in these children than in children with no major birth defects who used special education.²¹ Children with isolated gastrointestinal anomalies were 20% more likely than children with no major birth defects to receive special education (adjusted risk ratio: 1.2 [95% confidence interval: 1.04-1.37]).²⁹ The most common special education exceptionalities for these children were speech and language disorders and learning disorders. Children with congenital heart defects were 50% more likely to receive special education services than those without any major birth defects (adjusted prevalence risk ratio: 1.5 [95% confidence interval: 1.4–1.7]).³⁰ Among children with an orofacial cleft, a larger percentage of non-Hispanic black (29.8%) and Hispanic (31.3%) children were in special education than non-Hispanic white (24.0%) children.²¹ To date, no similar analysis exists for children with CS.

Timing of entry into special education by children with birth defects is also an important factor in enrollment. One could theorize that younger children are more receptive

to multiple forms of learning, potentially leading to more cognitive development. Additionally, children in Georgia are only mandated to remain in school between ages 6-16, but beginning special education prior to age 6 could yield more years of learning and subsequently more educational attainment.³¹ The median age at entry into special education was 6 years for children with an isolated orofacial cleft, compared with the median age at entry into special education for children with no major birth defects at 7 years.²¹

Craniosynostosis and Special Education Services

To date, there is no available literature on the receipt of special education services in children with CS compared to children with no major birth defects. This study investigated the prevalence of special education use among children with CS compared to children in the same birth cohort with no major birth defects. This analysis will provide an evidence-based information for planning appropriate educational services (early intervention programs) for children with CS and the development of guidance for families of services that might be of benefit to affected children. Supplementary study analyses will examine if any racial or ethnic differences exist in participation in special education and if there are any common exceptionalities associated with children with CS.

References

- 1. Centers for Disease Control and Prevention (CDC). (2008). Update on overall prevalence of major birth defects--Atlanta, Georgia, 1978-2005. *MMWR Morbidity and mortality weekly report*, 57(1), 1.
- Lajeunie, E., Merrer, M.L., Bonaïti-Pellie, C., Marchac, D., & Renier, D. (1995). Genetic study of nonsyndromic coronal craniosynostosis. *Am J Med Genet*, 55(4), 500-504.
- Boulet, S. L., Rasmussen, S. A., & Honein, M. A. (2008). A population-based study of craniosynostosis in metropolitan Atlanta, 1989–2003. *Am J Med Genet A*, 146A(8), 984-991.
- Tennant, P. W., Pearce, M. S., Bythell, M., & Rankin, J. (2010). 20-year survival of children born with congenital anomalies: a population-based study. *The lancet*, 375(9715), 649-656.
- Speltz, M. L., Collett, B. R., Wallace, E. R., Starr, J. R., Cradock, M. M., Buono, L., . . . Kapp-Simon, K. (2015). Intellectual and academic functioning of school-age children with single-suture craniosynostosis. *Pediatrics*, 135(3), e615-23.
- 6. Starr, J. R., Collett, B. R., Gaither, R., Kapp-Simon, K. A., Cradock, M. M., Cunningham, M. L., & Speltz, M. L. (2012). Multicenter study of neurodevelopment in 3-year-old children with and without single-suture craniosynostosis. *Archives of Pediatrics & Adolescent Medicine*, 166(6), 536-542.
- 7. Johnson, D., & Wilkie, A.O.M. (2011). Craniosynostosis. Eur J Hum Genet, 19(4), 369-376.
- 8. Maliepaard, M., Mathijssen, I.M., Oosterlaan, J., & Okkerse, J.M. (2014). Intellectual, behavioral, and emotional functioning in children with syndromic craniosynostosis. *Pediatrics, 133*(6), 1608-1615.
- Wilkie, A.O., et al. (2007). Clinical dividends from the molecular genetic diagnosis of craniosynostosis. Am J Med Genet A, 143A(16), 1941-1949.
- Greenwood, J., Flodman, P., Osann, K., Boyadjiev, S. A., & Kimonis, V. (2014). Familial incidence and associated symptoms in a population of individuals with nonsyndromic craniosynostosis. *Genet Med*, 16(4), 302-310.
- Slater, B.J., Lenton, K.A., Kwan, M.D., Gupta, D.M., Wan, D.C., & Longaker, M.T. (2008). Cranial sutures: a brief review. *Plast Reconstr Surg*, 121(4), 170–178.
- Sanchez-Lara, P.A., Carmichael, S.L., Graham, J.M., Lammer, E.J., Shaw, G.M., Ma, C., Rasmussen, S.A. (2010). Fetal constraint as a potential risk factor for craniosynostosis. *Am J Med Genet A*, 152(2), 394–400.
- Alderman, B. W., Zamudio, S., Baron, A. E., Joshua, S. C., Fernbach, S. K., Greene, C., & Mangione, E. J. (1995). Increased risk of craniosynostosis with higher antenatal maternal altitude. *International journal of epidemiology*, 24(2), 420-426.
- Gardner, J. S., Guyard-Boileau, B., Alderman, B. W., Fernbach, S. K., Greene, C., & Mangione, E. J. (1998). Maternal exposure to prescription and non-prescription pharmaceuticals or drugs of abuse and risk of craniosynostosis. *International journal of epidemiology*, 27(1), 64-67.
- 15. Carmichael, S. L., Ma, C., Rasmussen, S. A., Cunningham, M. L., Browne, M. L.,... Shaw, G. M. (2015). Craniosynostosis and risk factors related to thyroid dysfunction. *Am J Med Genet A*, 167(4), 701-707.
- Carmichael, S.L., Ma, C., Rasmussen, S.A., Honein, M.A., Lammer, E.J., & Shaw, G.M. (2008). Craniosynostosis and maternal smoking. *Birth Defects Research Part A: Clinical and Molecular Teratology*, 82(2), 78-85.
- 17. Fok, H. J., Jones, B. M., Gault, D. G., Andar, U., & Hayward, R. (1992). Relationship between intracranial pressure and intracranial volume in craniosynostosis. *British Journal of Plastic Surgery*, 45(5), 394-397.

- Gault, D. T., Renier, D., Marchac, D., & Jones, B. M. (1992). Intracranial pressure and intracranial volume in children with craniosynostosis. *Plastic and reconstructive surgery*, 90(3), 377-381.
- Resch, B., Gerdermann, A., Maurer, U., Ritschl, E., & Müller, W. (1996). Neurodevelopmental outcome of hydrocephalus following intra-/periventricular hemorrhage in preterm infants: short-and long-term results. *Child's Nervous System*, 12(1), 27-33.
- Knight, S. J., Anderson, V. A., Spencer-Smith, M. M., & Da Costa, A. C. (2014). Neurodevelopmental outcomes in infants and children with single-suture craniosynostosis: a systematic review. *Developmental neuropsychology*, 39(3), 159-186.
- Yazdy, M. M., Autry, A. R., Honein, M. A., & Frias, J. L. (2008). Use of special education services by children with orofacial clefts. *Birth Defects Research Part A: Clinical and Molecular Teratology*, 82(3), 147-154.
- 22. Aron, L., & Loprest, P. (2012). Disability and the education system. The future of Children, 22(1), 97-122.
- Schiller, J. S., Adams, P. F., & Nelson, Z. C. (2005). Summary health statistics for the US population: National Health Interview Survey, 2003. Vital and health statistics. Series 10, Data from the National Health Survey, (224), 1-104.
- 24. Morgan, P. L., Frisco, M. L., Farkas, G., & Hibel, J. (2008). A propensity score matching analysis of the effects of special education services. *The Journal of special education*, *43*(4), 236-254.
- 25. McClelland, M. M., Acock, A. C., & Morrison, F. J. (2006). The impact of kindergarten learning-related skills on academic trajectories at the end of elementary school. *Early Childhood Research Quarterly*, 21(4), 471-490.
- Hanushek, E. A., Kain, J. F., & Rivkin, S. G. (2002). Inferring program effects for special populations: Does special education raise achievement for students with disabilities?. *Review of Economics and Statistics*, 84(4), 584-599.
- Snyder, T. D., & Dillow, S. A. (2013). Digest of Education Statistics, 2012. NCES 2014-015. National Center for Education Statistics.
- Scull, J., & Winkler, A. M. (2011). Shifting Trends in Special Education. *Thomas B. Fordham Institute*. Retrieved from http://edex.s3-us-west 2.amazonaws.com/publication/pdfs/ShiftingTrendsinSpecialEducation_7.pdf
- Hamrick, S. E., Strickland, M. J., Shapira, S. K., Autry, A., & Schendel, D. (2010). Use of special education services among children with and without congenital gastrointestinal anomalies. *American journal on intellectual and developmental disabilities*, 115(5), 421-432.
- Colarusso, T., Andrew, A., Razzaghi, H., Boyle, C., Mahle, W., Braun, K. V. N., & Correa, A. (2014). Receipt of Special Education Services Among Children with Congenital Heart Defects in Atlanta, Georgia. *Circulation*, 130(Suppl 2), A15517-A15517.
- 31. U.S. Department of Education: National Center for Education Statistics. (2015). [Table 5.1. Compulsory school attendance laws, minimum and maximum age limits for required free education, by state: 2015]. Retrieved from <u>https://nces.ed.gov/programs/statereform/tab5_1.asp</u>

Chapter 2

Isolated, Non-Syndromic Craniosynostosis and Special Education Use Introduction

Craniosynostosis (CS) is a birth defect characterized by the premature fusion of one or more cranial sutures, resulting in altered growth of the skull. The prevalence of CS in the United States is estimated at between 1 in 2,100 to 1 in 2,500 live births.^{1,2} Cases of CS are confirmed by radiographic imaging (computerized axial tomography scan) and classified by the location of the fused cranial suture(s): sagittal, coronal, metopic, lambdoid, or multiple sutures. CS is classified as single-suture if only one of the cranial sutures is fused. The sagittal suture is affected in 40-55% of cases, the coronal suture in 20-25% of cases, the metopic suture in 14% of cases, the lambdoid suture in 3-5% of cases, and complex, multi-sutures in 5% of cases.³ CS can occur as an isolated (non-syndromic) defect, a non-syndromic defect with other accompanying defects or abnormalities, or as part of a genetic syndrome accompanied by other congenital anomalies involving midface retrusion, limb abnormalities, and brain anomalies.⁴ It is estimated that syndromic defects account for 25-40% of all cases of CS and isolated defects account for 60-75% of all cases of CS.^{5,6} Recognized genetic syndromes include Apert, Crouzon, Pfeiffer, Muenke, and Saethre-Chotzen syndromes.

Craniosynostosis may increase intracranial pressure, resulting in developmental delays and neurological disorders. It has been reported that school-age children with CS achieved significantly lower scores, 3.9-4.2 points fewer, on measures of IQ and math achievement than demographically similar children without any major malformations.⁷ Factors associated with the extent of intellectual disability in children with CS include severity of skull deformity, subtype of craniosynostosis, presence or absence of a genetic syndrome, age at surgery, and age at evaluation.⁸

To date, there is no available literature on the receipt of special education services in children with CS compared to children with no major birth defects. This study investigates the prevalence of receipt of special education services among children with CS compared to children in the same birth cohort with no major birth defects. This analysis will help develop an evidence base for planning appropriate educational services (early intervention programs) for children with CS and in better advising families of services that might be of benefit to their children.

Methods

Study Population

The study cohort, identified from birth certificates, was comprised of children born from 1989 to 2004 to women residing in the following metropolitan Atlanta, Georgia counties: Clayton, Cobb, DeKalb, Fulton, and Gwinnett. It included children with CS and a random sample of contemporaneous children whose birth records did not link to the Metropolitan Atlanta Congenital Defects Program (MACDP) (i.e., children without any major birth defects) as the reference cohort. Children with CS within this cohort were identified by the MACDP using the 9th revision of the 1979 British Pediatric Association (BPA) Classification of Diseases and the World Health Organization's 1979 International Classification of Diseases Clinical Modification (ICD-9-CM) coding systems.⁹ MACDP is an ongoing, population-based surveillance system for fetuses, infants, and children with major structural birth defects that uses multiple sources of case ascertainment (e.g. birth hospitals, pediatric hospitals, specialty clinics, and perinatal offices).⁹ MACDP abstractors actively ascertain cases by reviewing records at medical facilities for the presence of any congenital anomaly ascertained by MACDP. A clinical geneticist reviewed the abstracted information on identified children with CS to ensure they were correctly classified as having CS, and to determine the affected suture type. Children with any non-syndromic CS subtype (sagittal, metopic, lambdoid, coronal, multiple, and unspecified suture types) were included in the study.

Children were excluded from the study if any of the following criteria were met: (1) the child was from a multiple birth, (2) the child did not survive past 3 years of age, or (3) the child was diagnosed with a major birth defect other than CS. Additionally, children were excluded from the CS case population if any of the following criteria were met: (1) the child was diagnosed with syndromic CS or (2) the diagnosis was not confirmed by radiographic imaging (computerized axial tomography scan).

This study was approved by the Emory University Institutional Review Board.

Receipt of Special Education Services

The Special Education Database of Metropolitan Atlanta (SEDMA) links children identified by special education departments of nine public school districts in metropolitan Atlanta longitudinally through their years of receiving special education services.¹⁰ All of the metropolitan Atlanta counties included in the MACDP catchment area are included in SEDMA. Children are identified for special education services and exceptionalities after an educational assessment process has been performed by the school system.¹¹ Once a child has been determined to be eligible for special education services and assigned an exceptionality, the school system develops and implements an Individualized Education Program (IEP) that addresses the specific learning needs of the child. Special education exceptionalities include mild to severe intellectual disability, profound intellectual disability, behavioral disorders, learning disorders, orthopedic impairments, hearing loss, deafness, vision loss, blindness, severe emotional disorder, significant developmental delay, and other health problems. To identify which children in our study cohort received special education services, we linked our study cohort to SEDMA data from 1994-2012 using birth certificate number. We limited our cohort to those between ages 3 and 10; 3 years is the earliest age at which children can receive special education services through the public school system in metropolitan Atlanta and 10 years is likely the maximum age that children will be first identified for special education services.¹¹

Available data on special education exceptionalities only included the primary exceptionality assigned upon most recent year of enrollment in special education. Explanatory variables obtained from birth certificate data included the following: birth weight (<1500 g, 1500–2499 g, \geq 2500 g), maternal race/ethnicity (non-Hispanic white, non-Hispanic black, other [Hispanic, Asian/Pacific Islander, American Indian/Alaskan, multiracial]), maternal education at delivery (some high school, high school graduate, college, post-college), sex, and maternal age at delivery (<20 years, 20–24 years, 25–29 years, 30–34 years, \geq 35 years). These variables were chosen because of their association with special education participation in the literature.

Analysis

In descriptive analyses, we calculated frequency distributions of maternal and infant characteristics of children aged 3-10 years with CS compared to children with no major birth defects who were enrolled at least one year in special education in metropolitan Atlanta. Descriptive analyses included the analysis of the proportion of children in special education stratified by maternal race/ethnicity and the proportion of children in special education stratified by type of CS.

To estimate the association between CS and participation in special education, we used logistic regression to calculate both crude and adjusted odds ratios (ORs) and 95%

confidence intervals (CIs). We compared the odds of receiving special education services among children with CS to the odds of receiving special education services among children in the same birth cohort with no major birth defects. We also assessed the association between receiving special education services and type of CS (compared to both the cohort of infants without birth defects and compared to the cohort of infants with sagittal CS, the most common subtype). Finally, we assessed the association between CS and receiving common specific special education exceptionalities. All multivariable models included adjustment for the following *a priori* confounders that were identified from relevant literature: birth weight, maternal race/ethnicity, maternal education at delivery, sex, and maternal age at delivery.

All analyses were performed using SAS version 9.3 (SAS Institute Inc. Cary, NC).

Results

The study included 6,059 children with no major birth defects and 217 children with CS born in metropolitan Atlanta between 1989 and 2004 (Table 1). Compared with children without major birth defects, children with CS were more likely to be male, weigh greater than 2,500 grams at birth, and be born to older, non-Hispanic white, and more educated mothers. The majority of children with CS were classified as having a fused sagittal or metopic suture.

Children with CS born to non-Hispanic black mothers had a higher proportion enrolled in special education than children with CS born to non-Hispanic white mothers or mothers of other race/ethnicities (non-Hispanic black (26.1%), non-Hispanic white (18.0%), other (14.3%)) (Figure 1). The distribution of maternal race/ethnicity for children enrolled in special education was similar for children without any major birth defects (non-Hispanic white (9.5%), non-Hispanic black (11.7%), other (11.5%)). Among all children with CS, 19.4% were enrolled in special education for at least one year between ages 3 through 10, while only 10.7% of children with no major birth defects were in special education (Table 2). Children with CS had 1.74 (95% CI: 1.19, 2.54) times the odds of being enrolled in special education compared with children without any major birth defects, controlling for infant characteristics (birth weight and sex) and maternal characteristics (maternal race/ethnicity, maternal education at delivery, and maternal age at delivery) (Table 3).

Among children receiving any special education services, speech/language disorders were the most common exceptionality with 42.9% of children with CS and 42.3% of children with no major birth defects enrolled for speech/language disorders (Table 2). Children with CS were much more likely to be enrolled in special education for an intellectual disability than children without any major birth defects. After adjustment for infant characteristics and maternal characteristics, children with CS had 11.27 (95% CI: 4.16, 30.54) times the odds of being enrolled in special education for any degree of intellectual disability than children without any major birth defects (21.4% vs. 5.0%). There were no other strong differences in enrollment in special education exceptionalities between children with CS and children without any major birth defects after adjustment for explanatory variables.

The odds of participation in special education differed between children with varying types of CS and children without any major birth defects. Children with sagittal CS had 1.89 (95% CI: 1.09, 3.27) and children with metopic CS had 4.04 (95% CI: 2.08, 7.87) times the odds of being enrolled in special education compared with children with no major birth defects (Table 4). Although those with metopic CS had 2.14 (95% CI: .91, 5.03) the odds of

being enrolled in special education compared with children with sagittal CS, there were no other meaningful differences in the odds of participation between any of the other types of CS (e.g. coronal vs. sagittal).

Discussion

In our study of 217 children with CS, 19.4% were enrolled in special education for at least one year between ages 3 through 10, while only 10.7% of a random sample of 6,051 children with no major birth defects were enrolled in special education. We found that children with CS had nearly twice the odds of being enrolled in special education than children without any major birth defects. Upon examination of type of CS, children with sagittal CS had nearly twice the odds and children with metopic CS had nearly four times the odds of being enrolled in special education than children with no major birth defects. Upon examination of type of CS, children with sagittal CS had nearly twice the odds and children with metopic CS had nearly four times the odds of being enrolled in special education compared with children with no major birth defects. Although those with metopic CS had twice the odds of being enrolled in special education compared with children with sagittal CS, there were no other significant differences in the odds of participation between any of the other types of CS, though the number of children in some sub-groups of CS were small. The most common exceptionality for children with CS was a speech/language disorder followed by an intellectual disability.

These findings are consistent with other studies which have assessed special education exceptionalities and receipt of special education in relation to birth defects. Children with isolated gastrointestinal anomalies were 20% more likely than children with no major birth defects to receive special education (adjusted risk ratio = 1.2, 95% CI: 1.04-1.37) and children with congenital heart defects were 50% more likely to receive special education services than those without any major birth defects (adjusted prevalence risk ratio = 1.5; 95% CI: 1.4-1.7).^{10,12} Among children with an orofacial cleft, a larger percentage of non-Hispanic black (29.8%) and Hispanic (31.3%) children were in special education than non-Hispanic

white (24.0%) children.¹¹ We also found that a larger percentage of non-Hispanic black children with CS (26.1%) were in special education, but a smaller proportion of non-Hispanic white (18.0%) and other race children (14.3%) with CS were in special education.

This study has several strengths. Children with CS were identified from a populationbased, active-ascertainment birth defects surveillance program with careful case review that determined whether the CS was syndromic or not, whether it was accompanied by other major birth defects, and what the affected suture was. Birth certificate data were used to identify a large sample of contemporaneous children without major birth defects for comparison. The linkage of data between SEDMA and MACDP provides the first population-based assessment of special education use by children with CS.

Although these findings provide new insight into special education use by children with CS, there are still a number of limitations. We had a small sample size due to the rarity of CS in the population, leading to low power and imprecise estimates for some results. As a result of our small sample size, many categories had to be collapsed, such as "other" race and intellectual disability, which included mild intellectual disability, moderate intellectual disability, severe intellectual disability, and profound intellectual disability. Thus, this analysis was not able to examine CS and its relation to severity of intellectual disability or special education use by those other than Non-Hispanic white and Non-Hispanic black children. Additionally, available data on special education exceptionalities only included the primary exceptionality assigned upon most recent year of enrollment in special education rather than information from all years of enrollment in special education. Thus, we were unable to examine factors such as age at entry into special education. Another limitation is that we lack migration data for the five counties of metropolitan Atlanta included in the MACDP (Clayton, Cobb, DeKalb, Fulton, and Gwinnett). We do not know how many children in the MACDP and those in the sample of controls migrated out of the five Atlanta counties and were not available for linkage to SEDMA.¹⁰ Finally, it is important to highlight that SEDMA data links children identified by special education departments of nine public school districts in metropolitan Atlanta. Any receipt of special education services through private schooling or homeschooling was not captured by SEDMA. In 2011, an estimated 1.77 million students were homeschooled and 4.94 million students attended private school in the US.^{14,15} Because our analysis only includes those enrolled in public school, there is potential for underestimation of the true proportion of children with CS receiving special education services.

Conclusions

To our knowledge this is the first population-based study to assess the use of special education by children with CS. The results of this study suggest that children with CS have higher odds of enrolling in special education for certain exceptionalities than children without any major birth defects. These findings highlight the need for special education services for children with CS. Future analyses should examine the timing of enrollment in special education for those with CS and track children with CS in special education into adulthood to assess long term developmental outcomes.

References

- Boulet, S. L., Rasmussen, S. A., & Honein, M. A. (2008). A population-based study of craniosynostosis in metropolitan Atlanta, 1989–2003. *American Journal of Medical Genetics Part A*, 146(8), 984-991.
- Lajeunie, E., Merrer, M. L., Bonaïti-Pellie, C., Marchac, D., & Renier, D. (1995). Genetic study of nonsyndromic coronal craniosynostosis. *American journal of medical genetics*, 55(4), 500-504.
- Slater, B. J., Lenton, K. A., Kwan, M. D., Gupta, D. M., Wan, D. C., & Longaker, M. T. (2008). Cranial sutures: a brief review. *Plastic and reconstructive surgery*, 121(4), 170e-178e.
- 4. Maliepaard, M., Mathijssen, I. M., Oosterlaan, J., & Okkerse, J. M. (2014). Intellectual, behavioral, and emotional functioning in children with syndromic craniosynostosis. *Pediatrics*, 133(6), e1608-e1615.
- Wilkie, A.O., et al. (2007). Clinical dividends from the molecular genetic diagnosis of craniosynostosis. Am J Med Genet A, 143A(16), 1941-1949. doi:10.1002/ajmg.a.31905
- Greenwood, J., Flodman, P., Osann, K., Boyadjiev, S. A., & Kimonis, V. (2014). Familial incidence and associated symptoms in a population of individuals with nonsyndromic craniosynostosis. *Genet Med*, 16(4), 302-310. doi:10.1038/gim.2013.134
- Speltz, M. L., Collett, B. R., Wallace, E. R., Starr, J. R., Cradock, M. M., Buono, L., . . . Kapp-Simon, K. (2015). Intellectual and academic functioning of school-age children with single-suture craniosynostosis. *Pediatrics*, 135(3), e615-e623.
- Knight, S. J., Anderson, V. A., Spencer-Smith, M. M., & Da Costa, A. C. (2014). Neurodevelopmental outcomes in infants and children with single-suture craniosynostosis: a systematic review. *Developmental neuropsychology*, 39(3), 159-186.
- Correa, A., Cragan, J. D., Kucik, J. E., Alverson, C. J., Gilboa, S. M., Balakrishnan, R., et al. (2007). Metropolitan Atlanta Congenital Defects Program. *Birth Defects Research Part A: Clinical and Molecular Teratology*, 79(2), ii-186.
- Colarusso, T., Andrew, A., Razzaghi, H., Boyle, C., Mahle, W., Braun, K. V. N., & Correa, A. (2014). Receipt of Special Education Services Among Children with Congenital Heart Defects in Atlanta, Georgia. *Circulation*, 130(Suppl 2), A15517-A15517.
- Yazdy, M. M., Autry, A. R., Honein, M. A., & Frias, J. L. (2008). Use of special education services by children with orofacial clefts. *Birth Defects Research Part A: Clinical and Molecular Teratology*, 82(3), 147-154.
- Hamrick, S. E., Strickland, M. J., Shapira, S. K., Autry, A., & Schendel, D. (2010). Use of special education services among children with and without congenital gastrointestinal anomalies. *American journal on intellectual* and developmental disabilities, 115(5), 421-432. doi: 10.1352/1944-7558-115-5.421
- Carmichael, S. L., Ma, C., Rasmussen, S. A., Cunningham, M. L., Browne, M. L.,... Shaw, G. M. (2015). Craniosynostosis and risk factors related to thyroid dysfunction. *Am J Med Genet A*, 167(4), 701-707. doi:10.1002/ajmg.a.36953
- Broughman, S. P., & Swaim, N. L. (2013). Characteristics of private schools in the United States: Results from the 2011–12 Private School Universe Survey (NCES 2013-316). US Department of Education. Washington, DC: National Center for Education Statistics.
- Noel, A., Stark, P., & Redford, J. (2013). Parent and Family Involvement in Education, from the National Household Education Surveys Program of 2012. First Look. NCES 2013-028. National Center for Education Statistics.

	Children with	Children with no major	
	CS	birth defects	
	n=217 (%)	n=6,059 (%)	p-value
Sex			*
Male	146 (67.3)	3,082 (50.9)	< 0.001
Female	71 (32.7)	2,977 (49.1)	
Maternal race			
Non-Hispanic white	150 (69.1)	2,563 (42.4)	<.0001
Non-Hispanic black	46 (21.2)	1,888 (31.2)	
Other	21 (9.7)	1,608 (26.6)	
Maternal education			
Some high school	31 (14.5)	1,336 (22.3)	< 0.001
High school	45 (21.0)	1,698 (28.3)	
College	104 (48.6)	2,168 (36.2)	
Graduate school	34 (15.9)	789 (13.2)	
Missing	3	68	
Maternal age at delivery			
(years)			
<19	12 (5.5)	691 (11.4)	< 0.001
20-24	31 (14.3)	1,545 (25.5)	
25-29	66 (30.4)	1,706 (28.2)	
30-34	58 (26.7)	1,394 (23.0)	
35+	50 (23.0)	723 (11.9)	
Birth weight (grams)			
<1,500	10 (4.7)	27 (0.7)	< 0.001
1500-2499	17 (8.0)	221 (5.7)	
2,500+	186 (87.3)	3,652 (93.6)	
Missing	4	2,159	
Type of CS			
Sagittal	87 (40.1)	-	
Metopic	40 (18.4)	-	
Lambdoid	39 (17.8)	-	
Coronal	33 (15.2)	-	
Multiple	16 (7.4)	-	
NOS	2 (0.9)		

Table 1. Characteristics of children aged 3-10 with craniosynostosis (CS)compared to children with no major birth defects born in metropolitan Atlanta,1994-2012



Figure 1. Proportion (%) of children aged 3-10 with and without CS enrolled in special education, stratified by maternal race/ethnicity, metropolitan Atlanta, 1994-2012

	,	Children with no
	Children with CS	major birth defects
Special education category	n=217(%)	n=6,059 (%)
Any special education services	42 (19.4)	645 (10.7)
Speech/Language Disorder	18 (42.9)	273 (42.3)
Learning Disorder	7 (16.7)	167 (25.9)
Intellectual Disability	9 (21.4)	32 (5.0)
Other Health Problem	7 (16.7)	74 (11.5)
Significantly Developmentally Delayed	1 (2.4)	32 (5.0)
Behavior Disorder	0 (0.0)	54 (8.4)
Orthopedically Impaired	0 (0.0)	4 (0.6)
Visually Impaired/Blind	0 (0.0)	4 (0.6)
Hearing Impaired/Deaf	0(0.0)	2 (0.3)
Severe Emotional Disorder	0 (0.0)	1 (0.2)
Deaf/Blind	0 (0.0)	0 (0.0)
Missing	0 (0.0)	2 (0.3)

Table 2. Proportion (%) of children aged 3-10 with and without CS enrolled inspecial education by exceptionality, metropolitan Atlanta, 1994-2012

	Unadjusted Odds Ratio	Adjusted Odds Ratio
	(95% CI)	(95% CI) ^a
Special education category		
Any special education services	2.02 (1.43, 2.85)	1.74 (1.19, 2.54)
Speech/Language Disorder	1.92 (1.17, 3.15)	1.61 (0.94, 2.74)
Learning Disorder	1.18 (0.55, 2.54)	1.31 (0.58, 2.92)
Intellectual Disability	8.15 (3.84, 17.29)	11.27 (4.16, 30.54)
Other Health Problem	2.70 (1.23, 5.92)	1.70 (0.73, 3.93)
Significantly Developmentally		
Delayed	-	-
Behavior Disorder	-	-
Orthopedically Impaired	-	-
Visually Impaired/Blind	-	-
Hearing Impaired/Deaf	-	-
Severe Emotional Disorder	-	-
Deaf/Blind	-	-

Table 3. Crude and adjusted odds ratios for the association between craniosynostosis (CS) and enrollment in special education, by exceptionality, among children age 3-10 in metropolitan Atlanta, 1994-2012

a. Adjusted for birth weight, maternal race/ethnicity, maternal education at delivery, maternal age at delivery, and infant sex

among children age 5-10 in metropolitan Atlanta, 1994-2012				
		Unadjusted Odds Ratio	Unadjusted Odds	
	n=687	(95% CI) ^a	Ratio (95% CI) ^b	
Children with no major birth				
defects	645	ref	-	
Children with CS	42	2.02 (1.43, 2.85)	-	
Sagittal	16	1.89 (1.09, 3.27)	ref	
Metopic	13	4.04 (2.08, 7.87)	2.14 (.91, 5.03)	
Lambdoid	6	1.53 (0.64, 3.66)	0.81 (0.29, 2.25)	
Coronal	5	1.50 (0.58, 3.90)	0.79 (0.27, 2.37)	
Multiple	2	-	-	

Table 4. Participation in special education stratified by type of craniosynostosis (CS) among children age 3-10 in metropolitan Atlanta, 1994-2012

a. Unadjusted odds ratio for participation in CS for at least one year comparing children with various types of CS to children with no major birth defects

b.Unadjusted odds ratio for participation in CS for at least one year comparing children with metopic, lambdoid, and coronal CS to children with sagittal CS