HHS Public Access

Author manuscript

Cardiol Young. Author manuscript; available in PMC 2016 February 01.

Published in final edited form as:

Cardiol Young. 2016 February ; 26(2): 250–256. doi:10.1017/S1047951115000062.

Influence of Congenital Heart Defect on Psychosocial and Neurodevelopmental Outcomes in Children with Down Syndrome

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Abstract

Objective—To evaluate the family psychosocial outcomes of children with Down syndrome and atrioventricular septal defect, and examine the impact of these variables on the child's neurodevelopmental outcome.

Methods—This was a cross-sectional study that consisted of 57 children with Down syndrome (20 cases and 37 controls), approximately 12–14 months of age. In both groups, we assessed the development of the child, the quality of the child's home environment, and parenting stress.

Results—Compared with the Down syndrome without congenital heart defect group, the atrioventricular septal defect group revealed lower scores in all developmental domains, less optimal home environments, and higher parental stress. Significant differences in development were seen in the areas of cognition (p=0.04), expressive language (p=0.05), and gross motor (p<0.01). The Home Observation for Measurement of the Environment revealed significant differences in emotional and verbal responsiveness of the mother between the two groups. The Parenting Stress Index revealed the Down syndrome with atrioventricular septal defect group had a significantly higher child demandingness subdomain scores compared to the Down syndrome without congenital heart defect group.

Conclusions—The diagnosis of a congenital heart defect in addition to the diagnosis of Down syndrome may provide additional stress to the child and parents, elevating parental concern and disrupting family dynamics, resulting in further neurodevelopmental deficits. Finding that parental stress and home environment may play a role in the neurodevelopmental outcomes may prompt

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Conflict of Interest

None

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the Emory University institutional committees.

new family-directed interventions and anticipatory guidance for the families of children with Down syndrome who have a congenital heart defect.

Keywords

Down syndrome; Congenital Heart Defects; Neurodevelopmental Outcomes; Psychosocial

Introduction

Down syndrome, or trisomy 21, is the leading genetic cause of intellectual disability, with an incidence of 1 in 691 live births, which means approximately 6000 infants with Down syndrome are born annually in the United States.[1] Children with Down syndrome are at increased risk for a congenital heart defect (CHD), with a reported prevalence of 41–56% compared with 1%–5% in the general pediatric population.[2–5] Atrioventricular septal defect, the most common form of CHD in Down syndrome, occurs in 31–61% of children with Down syndrome and CHD, but is observed in only 1 in 10,000 live births in those without Down syndrome.[6–8] This represents a 2000-fold increase in risk for atrioventricular septal defect among newborns with DS compared to the general pediatric population.[2–5,9]

Survival rates in the Down syndrome population have increased tremendously, from a median age of 25 years in 1983 to almost 60 years currently.[10] Routine echocardiogram screenings of all newborns with Down syndrome has improved detection of CHD, which has contributed to the growing number of early survivors. Additionally, rapid advancements in both surgical procedures and perioperative care have also decreased the mortality rate and improved survival rates among individuals with Down syndrome and CHD.[11–12] Indeed, the morbidity and mortality rates in all children (Down syndrome and non-Down syndrome) with atrioventricular septal defect is approximately 3%, with only 2.7% requiring additional operations. In children with Down syndrome and atrioventricular septal defect, the 5-year postoperative survival rate is approximately 90%.[13–16] Therefore, it is becoming increasingly important to evaluate the impact of atrioventricular septal defect and/or its treatment on their neurodevelopmental outcomes and family psychosocial functioning (e.g., parenting stress and impact on the family).

Neurodevelopmental outcomes in typically developing children with CHD have been studied extensively. Neurodevelopmental studies in this population have shown that they have more problems with reasoning, learning, executive function, inattention, language skills, and social skills compared to peers without CHD.[17–19] Notably, research on neurodevelopmental outcomes among individuals with Down syndrome and CHD is limited to two studies, which confirm that children with Down syndrome and CHD have an increased risk of developmental deficits, particularly in the language domain, compared to children with Down syndrome without CHD.[20,21]

Despite our extensive knowledge about the cognitive and behavioral phenotypes of Down syndrome, the influence of CHD in this population is often under recognized in neurodevelopmental and family outcome studies. There has been no study to evaluate the family psychosocial outcomes associated with children with Down syndrome and CHD, and the impact of these variables the child's neurodevelopmental outcome. Thus, this study is

the first to determine whether maternal and family factors are important in mediating neuropsychological outcomes for children with Down syndrome and CHD. Our findings will help clinicians (e.g., cardiologists and pediatricians) and parents pinpoint the most effective interventional therapies and resources that are responsive to the needs of children with Down syndrome and CHD and their families.

Materials and Methods

This was a cross-sectional study that consisted of children with Down syndrome post atrioventricular septal defect repair (cases) and children with Down syndrome with a structurally normal heart (controls). Participants were recruited through the Down Syndrome Clinic at Emory University in Atlanta, Georgia. The subjects' parent gave written consent for participation, as approved by the Emory University Institutional Review Board.

The participants included 57 children with Down syndrome (20 with atrioventricular septal defect and 37 without a congenital heart defect), approximately 12–14 months of age. This age range was selected because it allows parents to adjust to the needs of their newborn and also provides time to allow a child with an atrioventricular septal defect to recover from cardiac surgery (surgical repair of atrioventricular septal defect is typically performed when the child is roughly 10 pounds and/or 6 months of age). Additionally, this age range represents an ideal period of time to implement interventions for the child and family in the event that our study reveals developmental deficits and lower family psychosocial outcomes. Participants with Down syndrome and atrioventricular septal defect were ascertained after the cardiac repair. We only considered atrioventricular septal defects for this study to make our study sample as homogeneous as possible with respect to their cardiac status. Furthermore, the operative techniques and timing of repair of other types of CHDs, such as ventricular septal defects and atrial septal defects, differed when compared to atrioventricular septal defects. Controls were participants with Down syndrome who has a structurally normal heart as documented by echocardiograms. Subjects were eligible for participation if he/she had a confirmed trisomy 21 based on chromosomal karyotype, was delivered > 34 weeks gestational age, had vision and hearing within normal limits and parents spoke English as the primary language. Subjects with other medical conditions (e.g., congenital hypothyroidism, gut abnormalities, seizures, congenital infections, ophthalmologic and hearing complications) were excluded from the study. Subjects were also excluded with a gestational age of less than 34 weeks at delivery, 5-min Apgar score less than 7, or intrauterine exposure to substance abuse.

The child's development was assessed using the Bayley Scales of Infant and Development, Third Edition or Bayley-III.[22] The Bayley-III provides subscale scores on cognitive, language, and motor developmental indexes, with a mean of 100 and a standard deviation of 15. Mothers completed the Parenting Stress Index, a self-reported questionnaire that consists of 101 items to assess parental stress levels.[23] The questionnaire examines stress related to parental characteristics (Parent Domain score) and child characteristics (Child Domain score). The Parent Domain score is based on the parent's perception related to competence, isolation, attachment, health, role restriction, depression, and spouse/parenting partner

relationship. The Child Domain score is based on the child's adaptability, distractibility/hyperactivity, parent-child interaction, demandingness, mood, and acceptability.

The quality of the child's home environment was assessed by the Disability Adapted Infant/Toddler Home Observation for Measurement of the Environment-Developmental Delay or HOME assessment.[24] The HOME assessment has six subscales: 1) Emotional and Verbal Responsiveness, 2) Avoidance and Restriction, 3) Organization and Physical Environment, 4) Learning Materials, 5) Parental Involvement with Child, and 6) Opportunities for Variety in Daily Life. The HOME assessment is administered at the home with the parent and child present and consists of a combination of interview and observation by a qualified psychometrician. The questionnaire is the standard research instrument for assessment of environmental factors associated with development in children with disabilities. Of note, the psychometricians who administered the Bayley-III and HOME assessment were not aware of the child's cardiac status.

Statistics

The two cohorts were compared for variables such as child age, demographic information, and birth characteristics using Chi square tests, T-test, and Wilcoxon sign-rank tests as appropriate. Raw outcome scores from the Bayley-III, HOME, and Parenting Stress Index were analyzed between groups using the same methods. It has been noted that developmental quotients instead of standard scores can be used to track the developmental progress of intellectually disabled children against their chronological ages to compare their levels of functioning in different developmental domains.[25] The developmental quotient in children with intellectual disabilities correlated well with intellectual quotient scores later in life.[26] Therefore, we calculated the developmental quotient for each domain by dividing the developmental age by the chronological age and multiplying by 100. For the Bayley-III, we report the scores in the domains of cognition, gross motor, fine motor, receptive language, and expressive language. In order to account for potential confounding between groups, outcome scores from the Bayley-III, HOME, and Parenting Stress Index were also analyzed using multiple linear regression with case status as the main predictor and adjusting for child age, sex, and race as well as parental age, education level, and annual household income. The confounders included in the final model included only those variables whose removal altered the β estimate of the main predictor (case status) more than 10%. Total scores from the HOME assessment and Parenting Stress Index were also tested as significant predictors of Bayley-III scores.

Results

As indicated in Table 1, the Down syndrome with CHD group (n=20) was tested at approximately the same age as controls (n=37), though the cases were slightly older at 13.4 ± 1.1 months compared to 12.8 ± 0.8 months (p=0.03). The notable differences in demographics between the two groups were: 1) maternal education, with significantly more mothers with a bachelor's degree or higher among the control group (82.6%) compared to the case group (55.6%) (p=0.03) and 2) race, with more control subjects identifying as white (62.2%) compared to controls (60%) (p=0.03). Birth information among the two groups was

also relatively similar, however the case group had significantly lower Apgar scores (8.5 ± 0.6) at 5 minutes compared to the control group (8.9 ± 0.4) (p=0.01).

Analysis of the raw Bayley-III scores (Table 2) reveals that the composite scores are lower across all domains (cognition, receptive language, expressive language, gross motor, and fine motor) for the Down syndrome with CHD group compared to the Down syndrome without CHD cohort. In the receptive language domain, the Down syndrome with CHD cohort score (53.5 ± 19.5) was significantly lower than the control (66.9 ± 21.4) (p=0.02). The gross motor scores were also significantly different, with score of 53.1 ± 7.7 for those with CHD and the Down syndrome without CHD group score of 59.4 ± 7.7 (p<0.01). In the cognitive domain, the CHD cohort score (72.1 ± 14.0) was lower than the Down syndrome without CHD score (79.3 ± 13.5), though this difference did not reach statistical significance (p=0.06). After adjusting for confounders (e.g., demographic information), the CHD group scored significantly lower in the areas of cognition (p=0.04), expressive language (p=0.05), and gross motor (p<0.01) compared to the Down syndrome without CHD group. However, if we also adjust for the HOME and Parenting Stress Index scores, only the gross motor scores are significantly different between the two groups (p=0.02) indicating that parental stress and the home environment are a significant confounder between the case and control groups with regards to neurodevelopmental outcomes.

In the HOME analysis (Table 3), the Down syndrome with CHD group scored lower in all subscales compared to the Down syndrome without CHD group. The responsivity score, which includes emotional and verbal responsiveness of the mother, was 8.9 ± 2.3 in the Down syndrome with CHD group and 10.7 ± 1.7 in the DS without CHD group (p<0.01), with mothers of children with Down syndrome without CHD group demonstrating higher levels of responsiveness/interaction with their children. This association remains significant after adjusting for potential confounders (e.g., parental education level and income). There were no statistically significant differences in acceptivity, organization, learning material, involvement, or variety between mothers of the two groups. Overall, the total HOME score was significantly higher for the Down syndrome without CHD group compared to the Down syndrome with CHD group (p=0.01), with the Down syndrome without CHD group scoring 47.7 ± 4.8 compared to 43.8 ± 5.7 for the Down syndrome with CHD group. This result indicates a better overall home environment for the control group. However, this association does not remain significant after adjusting for potential confounders.

The overall Parenting Stress Index score (Table 4) is higher for the Down syndrome with CHD group (198.8 ± 26.9) compared to the Down syndrome without CHD group (183.2 ± 43.0), although the differences were not statistically significant (p=0.94). Furthermore, in the child demandingness subscale, the Down syndrome with CHD group scored 16.9 ± 3.9, significantly higher than the control group's score of 15.0 ± 4.3 (p=0.03). However, the association did not remain after adjusting for confounders. Among the other subdomain scores, there were no statistically significant differences between the two groups in either the child or parent domains. However, among the parent domain scores, differences in isolation between the two groups were noted which did not reach statistical significance (p=0.07).

Discussion

Neurodevelopmental outcomes in typically developing children with CHD have shown specific deficits in cognitive function, behavior, and learning.[17–19] Furthermore, a diagnosis of CHD may impact the family psychosocial outcomes, and family, in particular maternal factors (e.g., stress) may be more important determinants of neurodevelopmental outcomes than are operative management.[27,28] Studies have also shown that parents of children with Down syndrome experience greater parental stress than do parents of age-matched typically developing children.[28] However, these studies do not take into account co-morbid medical problems such as CHD. Our study is the first to determine if maternal and family function is important in contributing to outcomes in neurodevelopment for children with Down syndrome and CHD.

Based on the Bayley-III scores among the two groups, our results support previous studies showing significant differences in neurodevelopment outcomes between the two groups.[20,21] The Down syndrome with CHD group had greater developmental delay in all domains (motor, cognitive, language) compared to the Down syndrome without CHD group (Table 2). The differences in receptive language (p=0.02) and gross motor (p<0.01) scores were the only significantly different scores. After adjusting for demographic and parental variables (e.g., education and income), the scores for cognition (p=0.04), expressive language (p=0.05), and gross motor skills (p<0.01) remain significant between the two groups. However, when accounting for parental stress and home environment based on the Parenting Stress Index and HOME assessment scores, only gross motor skills (p=0.02) remains significantly different between the two groups, indicating there is a possible relationship with the level of stress and home environment to the child's neurodevelopmental outcomes.

The diagnosis of a CHD in addition to the diagnosis of Down syndrome may provide additional stress to the child and parents, elevating parental concern and disrupting family dynamics, resulting in further developmental deficits. Based on the Parenting Stress Index, parents of children with Down syndrome with CHD consistently reported more stress than parents of those with Down syndrome without CHD, in both Child and Parent subdomain factors (Table 4). The only significant child subdomain was child demandingness (p=0.03), and in the parent subdomain, isolation differed between the two groups (p=0.07). This finding is consistent with the extra parental care in the CHD group, who may require medications, specialized feeding routine, and additional appointments with specialists and therapists after cardiac surgical repair. The overall Parenting Stress Index score was also higher among the Down syndrome with CHD group, but the difference was not statistically significant (p=0.16); thus, the presence or absence of CHD does not significantly affect the total level of parental stress among mothers of children with Down syndrome. This finding may be due parents completing our study early in their experience in caring for a child with Down syndrome. Further studies should be conducted as these children age to assess whether continued increased medical needs related to CHD lead to increased levels of parental stress.

Based on the HOME assessment (Table 3), the Down syndrome with CHD group differed significantly from the control group in the area of verbal and emotional responsiveness

($p<0.01$) and total score ($p=0.01$), with the Down syndrome with CHD group having lower scores, which may be indicative of a lower quality of home environment for the children in this group. Mothers of children with Down syndrome without CHD demonstrate higher levels of responsiveness, both emotional and verbal, to their children than mothers of children with Down syndrome with CHD. Children with CHD in general are fragile which may lead parents to have decreased play activity and/or interaction. When looked at in conjunction with the Parenting Stress Index results, it may be possible that the increased demandingness seen in children with Down syndrome with CHD plays a role in the level of responsiveness. The differences in responsiveness remain significant ($p<0.01$) after adjusting for potential confounders (e.g., race, education). However, the difference in total score does not remain significant after adjusting for potential confounders, suggesting that the CHD status alone is not responsible for the lower overall home environment score in the Down syndrome with CHD group.

In terms of our study limitation, we did not evaluate the impact of other CHD subtypes on neurodevelopmental outcomes and family functioning. As our study is cross-sectional, further longitudinal study is needed to determine the trajectory of neurodevelopmental outcomes and parental/family functioning in children with Down syndrome with CHD compared cohort with Down syndrome without CHD. Additionally, future studies should consider the neurodevelopmental trajectory of children with Down syndrome with other CHD subtypes; such as ventricular septal defect and atrial septal defect.

Our results provide evidence that clinicians must recognize the developmental needs of those who have a CHD, as well as consider the needs of the family. Children with Down syndrome with CHD have greater delays than their peers with Down syndrome without CHD. Furthermore, it is possible that mothers and families of children with CHD are more prone to increased stress and other challenges at home, putting their children at even greater risk for developmental delay. Even when the cardiac defect is repaired, parents continue to experience personal stress and/or lack guidance in setting appropriate expectations for their child which may lead to decreased parent-child interaction. Finding that parental stress and home environment partially account for some of these differences in neurodevelopmental outcome between the two groups may prompt new family-directed interventions and anticipatory guidance for the families of children with Down syndrome with CHD. For example, pediatricians and cardiologists evaluating families of children with Down syndrome who have a CHD may ask about parental stress and family support as part of the routine care.

Acknowledgments

We thank the parents and children for their participation in this study.

Financial Support

Grant sponsors: NIH/NICHD; Grant number: 1K23HD058043-01A1, 5R01HD074346-02, and 1U54NS091859-01(JV).

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Table 1

Demographic of Subjects with DS + AVSD compared to DS − CHD

	DS + AVSD (n = 20)	DS − CHD (n = 37)	Association with case status
Chronological age in months	13.4 ± 1.1	12.8 ± 0.8	p=0.03
Males : Females	9 : 11	23 : 14	p=0.21
Whites : Other	12 : 8	23 : 14	p=0.03
Maternal Age at Delivery (years)	34.9 ± 7.6	32.6 ± 5.8	p=0.21
Paternal Age at Delivery (years)	37.8 ± 8.2	34.6 ± 6.7	p=0.16
Total Family Annual Income[†] **<$50,000 : ≥ $50,000**	6 : 10	8 : 26	p=0.31
Paternal Education[*] **< bachelors : ≥ bachelors**	10 : 8	14 : 21	p=0.28
Maternal Education[*] **< bachelors : ≥ bachelors**	8 : 10	6 : 29	p=0.03
Apgars1[*]	7.3 ± 1.7	7.8 ± 1.2	p=0.15
Apgars5[*]	8.5 ± 0.6	8.9 ± 0.4	p=0.01
Gestational Age	37.1 ± 1.9	37.8 ± 1.4	p=0.09

DS = Down syndrome; AVSD = atrioventricular septal defect

[*] Data are missing for two participants in the DS + AVSD group and two participants in the DS − CHD group

[†] Data are missing for four participants in the DS + AVSD group and three participants in the DS − CHD

Table 2

Bayley III scores for DS + CHD and DS − CHD

	DS + CHD (n = 20)	DS − CHD (n=37)	T-test*	Linear regression	
				Model 1†	Model 2^
Cognition	72.1 ± 14.0	79.3 ± 13.5	p=0.06	p=0.04	p=0.18
Receptive Language	53.5 ± 19.5	66.9 ± 21.4	p=0.02	p=0.10	p=0.17
Expressive Language	60.4 ± 20.3	66.8 ± 21.1	p=0.27	p=0.05	p=0.06
Fine Motor	64.9 ± 11.7	67.6 ± 12.9	p=0.43	p=0.91	p=0.48
Gross Motor	53.1 ± 7.7	59.4 ± 7.7	p<0.01	p<0.01	p=0.02

*Unadjusted analyses

†Models adjusted for covariates: child age at testing, child sex/gender, child race, maternal age at testing and level of education, paternal age at testing and level of education, and household income

^Models adjusted for the covariates above as well as HOME assessment and Parenting Stress Index scores

Table 3

HOME assessment scores for DS + CHD and DS − CHD

	DS + CHD (n = 20)	DS − CHD (n = 37)	T-test*	Linear regression†
Responsivity	8.9 ± 2.3	10.7 ± 1.7	p<0.01	p<0.01
Acceptivity	7.8 ± 0.7	8.1 ± 0.9	p=0.09	p=0.51
Organization	5.6 ± 0.7	5.6 ± 0.9	p=0.86	p=0.73
Learning Material	7.1 ± 1.7	7.9 ± 1.4	p=0.10	p=0.48
Involvement	10.3 ± 1.9	11.3 ± 1.2	p=0.08	p=0.77
Variety	4.0 ± 1.2	4.2 ± 0.9	p=0.68	p=0.64
Total Score	43.8 ± 5.7	47.7 ± 4.8	p=0.01	p=0.49

*Unadjusted analyses

†Models adjusted for covariates: child age at testing, child sex/gender, child race, maternal age at testing and level of education, paternal age at testing and level of education, and household income

Table 4

PSI scores for DS + CHD and DS − CHD

	DS + CHD (n = 20)	DS − CHD (n = 37)	T-test*	Linear regression†
Child Domain	**94.1 ±15.3**	**90.4 ± 14.3**	**p=0.32**	**p=0.66**
Distractability/Hyperactivity	26.6 ± 4.9	26.6 ± 4.8	p=0.93	p=0.90
Adaptability	21.1 ± 4.7	21.7 ± 5.4	p=0.78	p=0.43
Reinforces Parent	7.5 ± 1.8	7.5 ± 2.6	p=0.48	p=0.94
Demandingness	16.9 ± 3.9	15.0 ± 4.3	p=0.03	p= 0.16
Mood	7.5 ± 1.9	7.2 ± 2.2	p=0.39	p=0.15
Acceptability	14.0 ± 3.5	13.1 ± 3.9	p=0.44	p=0.52
Parent Domain	**102.1 ± 16.8**	**99.3 ± 27.0**	**p=0.37**	**p=0.73**
Competence	22.6 ± 4.5	22.6 ± 7.0	p=0.58	p=0.80
Isolation	11.4 ± 2.3	10.1 ± 3.6	p=0.07	p=0.06
Attachment	10.5 ± 3.1	9.5 ± 2.2	p=0.23	p=0.46
Health	11.7 ± 2.5	10.7 ± 3.6	p=0.12	p=0.33
Role Restriction	15.6 ± 3.6	16.8 ± 5.4	p=0.58	p= 0.71
Depression	16.4 ± 4.0	16.5 ± 6.5	p=0.67	p=0.46
Spouse	13.9 ± 3.8	14.3 ± 5.8	p=0.84	p= 0.59
Total Score	198.8 ± 26.9	183.2 ± 43.0	p=0.16	p=0.17
Life Stress Scale	6.9 ± 6.6	7.0 ± 7.2	p=0.94	p=0.67

PSI = Parenting Stress Index

*Unadjusted analyses

†Models adjusted for covariates: child age at testing, child sex/gender, child race, maternal age at testing and level of education, paternal age at testing and level of education, and household income