



Academic Profile of Children with 22q11.2 Deletion Syndrome: Delineating Specific Mathematics and Reading Disabilities

Stephen R. Hooper, Kathleen Curtiss, Kelly Schoch, & Vandana Shashi

Carolina Institute for Developmental Disabilities
University of North Carolina School of Medicine
Duke University Medical Center

Paper presented at the 20th International Scientific Meeting of the Velo-Cardio-Facial Syndrome Educational Foundation, Inc., Dublin, Ireland, July 19, 2013.

Conflicts of Interest

- The investigators have no conflicts of interest to disclose.



Introduction

- Intellectual functioning in children with 22q11DS varies from the low average to mild intellectual disability range.
 - 45% qualify for an Intellectual Disability.
- Additional deficits have been documented in:
 - Sustained attention
 - Visual-spatial abilities, with some reported similarities to nonverbal learning disability
 - Executive functions
- Psychiatric disorders have been noted in up to 60% of children with 22q11DS
 - Attention-Deficit/Hyperactivity Disorder (ADHD)
 - Anxiety disorders
 - Approximately 25% develop major psychiatric illnesses (e.g., schizophrenia, bipolar illness, major depression) in late adolescence/early adulthood



Introduction

- The focus of much of the research with 22q11DS has been on neurocognitive and psychiatric functioning, particularly in light of the fact that this population is at higher risk for severe psychiatric disorders.
- Less focus has been placed on academic functioning of these children which, perhaps predictably, is also an area of difficulty for many.
- The available research has shown specific deficits in mathematics, with relatively better reading and spelling abilities.



Introduction

- Deficits in math in children with 22q11DS have been shown to be related more to procedural difficulties than fact retrieval.
 - Retrieving basic arithmetic facts tends to be faster than when borrowing and carrying numbers is required to complete a calculation
- They also have been shown to experience greater difficulty solving word problems, and comparing the magnitude of numbers.



Introduction

- Studies investigating reading are sparse.
- Available findings are somewhat mixed with respect to specific reading skills:
 - Overall average pre-reading abilities with relatively lower phonological awareness.
 - Significantly better developed word reading skills compared to reading comprehension.
 - Reading comprehension level is equivalent to word reading level.



Current Study

- The current study sought to expand upon what little literature already exists by examining the specific academic profile of children and adolescents with 22q11DS.
- The current exploratory study expanded upon prior research by investigating the prevalence and nature of learning problems in 22q11DS in a larger cohort, comparing their performance to controls, and looking at achievement in specific subject areas within the reading and math academic domains beyond overall reading and math scores.



Study Objectives

- To investigate the overall academic performance of children with 22q11DS compared to controls.
- To investigate the prevalence of LD in children with 22q11DS.
- To provide an in-depth analyses of specific academic skill areas within mathematics and reading in children with 22q11DS.
 - Such a description would provide information that would be helpful in devising academic interventions for children with 22q11DS.



Sample Description

- Sample Size
 - 22q11DS = 85
 - Controls = 76
- Preliminary data analyses revealed no differences between the 22q11DS and control groups on
 - Chronological age, $t(161) = -1.55, p = .124$
 - 22q11DS = 13.57 ± 3.1 years
 - Control = 13.46 ± 2.7 years
 - Socioeconomic status, $t(148) = -0.31, p = .75$
 - 22q11DS = 29.17 ± 14.3
 - Control = 33.5 ± 17.8
 - Gender, $\chi^2(2) = .223, p = .895$
 - 22q11DS = 52.9% male
 - Controls = 53.8% male
 - Ethnicity, $\chi^2(8) = 15.4, p = .052$
 - 22q11DS = 82.4% Caucasian
 - Controls = 70.5% Caucasian



Measures

- Intelligence
 - Wechsler Intelligence Scale for Children-III/IV
- Achievement
 - Wechsler Individual Achievement Test-II/III
 - The WIAT-III, allows for in-depth skills analyses on several subtests that provide a more specific look at academic strengths and weaknesses.



Data Analyses

- In order to investigate the prevalence of significant learning problems in children with 22q11DS, we employed a low achievement model.
 - Bottom quartile compared to their peers in achievement
 - IQ broadly within the average range (≥ 80)
- In order to investigate the performance of children with 22q11DS compared to controls, a multivariate analysis of covariance (MANCOVA) was conducted separately for the reading and math domains.
 - Scores on the Spelling subtest were examined with an analysis of variance (ANCOVA).
- To examine our final question regarding group performance on various academic skill areas, a percent correct variable was calculated for each skill area.
 - The percent correct was based on the number of items correct in the specific skill area divided by the total number of items administered in that area for each individual participant.
 - To compare groups on the various skills areas, t-tests were conducted.



Prevalence of Learning Problems in 22q11.2DS

- Examination of mean academic scores from our sample of children with 22q11DS indicated borderline to low average academic functioning across all tasks.
- Higher overall reading ($M = 83.53 \pm 15.43$) and spelling ($M = 84.34 \pm 16.23$) scores than mathematics ($M = 73.6 \pm 17.68$).
- Performance within each academic domain was consistent, with no significant discrepancies between scores on the individual subtests within an academic domain.



Prevalence of Learning Problems in 22q11.2DS

- Prevalence of learning problems (Standard Score ≤ 90) was high:
 - 67% (57/85) for a reading problem
 - 87% (74/85) for a math problem
 - 36% (31/85) for a spelling problem
- When the requirement of an average IQ was applied (IQ ≥ 80):
 - 3.5% for a reading problem
 - 16.5% for a math problem
 - 2.4% for a spelling problem



Specific Academic Skills

- Several critical academic skills were examined using the WIAT-III specific skills analysis procedures.
- This afforded a more detailed examination of:
 - Reading Comprehension (literal and inferential comprehension)
 - Numerical Operations (basic concepts, basic math, algebra, geometry)
 - Multiplication (1, 2, 3-digit multiplication)
 - Math Reasoning (basic concepts, everyday applications, algebra, geometry)



Conclusions

- Results indicated an overall profile of better reading and spelling than math abilities.
 - These general findings replicate earlier studies.
- In addition, despite a large majority of children with 22q11DS performing in the lowest quartile when compared to their peers, very few met criteria for the low achievement definition of a learning disability.
- There were also many significant differences on the supplemental scores of the WIAT-III, with the exception of very basic skills that would be learned at a young age, and advanced skills in which neither group were proficient.



Conclusions

- From an assessment perspective, academic skills need to be assessed on a regular basis, in detail, and progress monitoring implemented.
- From an intervention perspective:
 - Early intervention for both reading and math skills is necessary
 - Higher-order comprehension skills may require ongoing scaffolding and treatment
 - Higher-order math skills will require ongoing assistance that likely will go beyond typical instruction and tutoring.
- Increased examination of neurological underpinnings to associated learning problems.



Questions??

Stephen R. Hooper, Ph.D.
stephen.hooper@cidd.unc.edu

