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# Intelligence and visual motor integration in 5-year-old children with 22q11-deletion syndrome

Sasja Duijff [a,*], Petra Klaassen [a], Frits Beemer [b], Henriette Swanenburg de Veye [a], Jacob Vorstman [c,d], Gerben Sinnema [a]

[a] Department of Pediatric Psychology, KA 00.004.0, University Medical Centre Utrecht, Postbus 85090, 3508 AB, Utrecht, The Netherlands
[b] Department of Medical Genetics, KC 04.084.2, University Medical Centre Utrecht, Postbus 85090, 3508 AB, Utrecht, The Netherlands
[c] Department of Child and Adolescent Psychiatry, A01.468, University Medical Centre Utrecht, Utrecht, The Netherlands
[d] Rudolf Magnus Institute of Neurosciences, Stratenum Building, Room STR5.203, Universiteitsweg 100, 3584 CG, Utrecht, The Netherlands

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ABSTRACT

The purpose of this study was to explore the relationship between intelligence and visual motor integration skills in 5-year-old children with 22q11-deletion syndrome (22q11DS) ($N = 65$, 43 females, 22 males; mean age 5.6 years (SD 0.2), range 5.23–5.99 years). Sufficient VMI skills seem a prerequisite for IQ testing. Since problems related to these skills are reported in children with 22q11DS, weak VMI skills may contribute to the lower than average IQ scores commonly reported. To investigate if the correlation of VMI and IQ score was mainly influenced by problems with visual perception skills (VP), motor coordination skills (MC) or difficulties with the integration of both skills (VMI), a subgroup ($n = 28$) was also administered the *Beery VMI* supplemental developmental tests. Due to the narrow age range of this study, we were also able to provide an insight into the neurocognitive phenotype of 5-year olds with 22q11DS and the influence of gender, heart disease and origin of deletion on this phenotype. Results show a mean full scale IQ (FSIQ) = 73.0 (SD 10.4) and mean VMI = 86.2 (SD 8.4). A significant correlation between FSIQ and VMI was found ($r = .45$, $p = .000$), with most variation (26%) explained in the performance IQ score ((PIQ), $r = .51$, $p = .000$). VP correlated significantly with FSIQ ($r = .44$, $p = .01$) and PIQ ($r = .49$, $p = .004$). MC was not significantly correlated with IQ (FSIQ, $r = .21$, $p = .15$; PIQ, $r = .28$, $p = .07$), suggesting that problems with motor coordination do not influence results on IQ-tests in a significant way at this age. Girls scored significantly higher on FSIQ and PIQ than boys; cardiac anomalies were not predictive of FSIQ or VMI scores. The results of this study suggest a characteristic neurocognitive phenotype for 5-year olds with 22q11DS. Deficiencies in visual perception and/or processing are negatively correlated with IQ scores, whereas deficiencies in motor skills do not have a relevant negative impact at this age. These findings provide further insight into 22q11DS specific neurocognitive deficiencies.



## 1. Introduction

The 22q11-deletion syndrome (22q11DS) is a genetic syndrome with a supposed incidence of 1 in 4000 live births (De Vriendt, Fryns, & Mortier, 1998; Oskarsdottir, Vujic, & Fasth, 2004). It is also known as velocardiofacial syndrome (VCFS),

* Corresponding author at: WKZ/UMC Utrecht, Department of Pediatric Psychology, Huispost KA.00.004.0, Postbus 85090, 3508 AB, The Netherlands. Tel.: +31 88 755 4111; fax: +31 88 755 5325.
*E-mail addresses:* s.duijff@umcutrecht.nl, 22q11@umcutrecht.nl (S. Duijff), p.w.j.klaassen@umcutrecht.nl (P. Klaassen), f.a.beemer@planet.nl (F. Beemer), h.deveye@umcutrecht.nl (H. Swanenburg de Veye), j.a.s.vorstman@umcutrecht.nl (J. Vorstman), g.sinnema@umcutrecht.nl (G. Sinnema).

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DiGeorge syndrome or Shprintzen syndrome. In 90% of cases the deletion occurs de novo, in 10% the deletion is passed on as an autosomal dominant trait. 22q11DS is characterized by a wide range of features. Common physical manifestations include velopharyngeal insufficiency with or without cleft palate, cardiac anomalies and a characteristic facial appearance (Bassett et al., 2011).

Psychiatric diagnoses are frequently reported and include autism spectrum disorders and attention deficit/hyperactivity disorder; children are also at an increased risk of developing psychotic symptoms and schizophrenia in adolescence (Antshel et al., 2006; Gothelf, 2007; Kates et al., 2007; Murphy, 2005; Vorstman et al., 2006). Also behavioral problems in childhood are often reported (Gerdes et al., 1999; Jansen et al., 2007). Neuropsychological characteristics of children with 22q11DS include an overall delay in cognitive development and IQ scores in the range of 70–85 (De Smedt et al., 2007; Niklasson & Gillberg, 2009). Significant differences between verbal and performance IQ (in favor of verbal IQ) are repeatedly reported (De Smedt et al., 2007; Gothelf, 2007; Moss et al., 1999). Research into the factors contributing to variability in IQ in 22q11DS focus on gender differences, familial versus de novo deletions and medical conditions such as cardiac anomalies. Reports on gender differences are inconclusive (Antshel et al., 2005; De Smedt et al., 2007) and cardiac anomalies are not found to be related with IQ. Inheritance of the deletion has been found to have a (negative) influence on IQ (De Smedt et al., 2007). Common to all reports, however, is that the included patients are characterized by broad age ranges.

Motor deficits are another important characteristic of 22q11DS. Difficulties in gross motor skills, hypotonia and difficulties with coordination and balance are reported in young children (Gerdes et al., 1999; Oskarsdottir, Belfrage, Sandstedt, Viggedal, & Uvebrant, 2005; Roizen et al., 2007; Sobin, Monk, Kiley-Brabeck, Khuri, & Karayiorgou, 2006). In school children (age 5–17 years) with 22q11DS not only gross but also fine motor deficits are present, such as poor eye-hand coordination and graphomotor deficits (Roizen et al., 2010; Van Aken et al., 2007; Aken, Caeyenberghs, Smits-Engelsman, & Swillen, 2009). Visual-motor integration is an important contributor to those academic skills that require a response on paper (e.g., maths, spelling and essays) and requires accurate perception of visual-spatial objects and monitoring one's own movements. Visual-motor integration in 22q11DS as specifically measured by *VMI Beery* has been studied by four authors (Lajiness-O'Neill et al., 2006; Niklasson & Gillberg, 2009; Roizen et al., 2010; Van Aken et al., 2009). Mean *Beery VMI* scores results were reported ranging from more than 2 SD below the mean to .5 SD below the mean. Two authors also assessed VMI motor coordination, resulting in scores 1.5 SD below the mean. One author also investigated visual perception skills (mean: −1 SD). All authors stressed the possible influence of poor motor skills on the test results and Sobin et al. (2006) stress the importance of recognizing these motor deficits in children with 22q11DS for accurate interpretation of a child's performance, both on tests as well as in the classroom. In general, the *Beery VMI* correlates moderately with intelligence tests and shares approximately 25% of variance (Beery & Beery, 2004).

The purpose of this study is to explore the relationship between intelligence and visual motor integration skills in 5-year-old children with 22q11DS. As these children are in the early stage of formal education (copying, drawing, writing), insight into the relations between cognitive and visual motor integration skills is clinically very relevant for accurate interpretation of test results. To avoid too much developmental variability, a narrow age range has been chosen.

The second aim of this study is to analyze the respective roles of visual perception, fine motor coordination skills and/or the (in)ability to integrate both in cognitive performance. Clearly, problems in VMI skills can confound the results on an IQ test if these skills are required but are not yet suitably mastered.

Also, due to the narrow age range of this study, we are able to provide an insight into the neurocognitive phenotype of 5-year olds with 22q11DS. All current reports on, for example, the influence of cardiac problems, origin of deletion, possible NLD profiles and gender differences on IQ are based on wide age ranges while it is very possible that, as suggested by Sobin et al. (2005): 'in this population the patterns of neurocognitive strengths and weaknesses shift with age; future studies will be needed to accurately characterize the neurocognitive phenotype ... at various points of development'.

## 2. Materials and methods

### 2.1. Participants

Participants were recruited through referrals from genetic counselors, cleft clinics and/or pediatric cardiologists from hospitals throughout The Netherlands through postings on the website of the Dutch parent support group VCFS/22q11DS. All participants have a 22q11-deletion as confirmed by FISH analysis (fluorescence in situ hybridization). The procedure was approved by the Dutch Central Committee on Research involving Human Subjects (C.C.M.O.) and is part of an ongoing national study on intelligence and behavior in children with 22q11DS. Written informed consent was obtained from all parents or legal guardians.

In the current study age was an inclusion criterion: all children between the age of 5.0 and 5.99 years were included, resulting in 77 children. Five children could not be assessed due to moderate or severe mental retardation ($n = 5$, all females). Seven children (3 females, 4 males) were unable to speak or speak comprehensibly, these children were assessed with a different, non-verbal intelligence test (SON-R 2.5–7) (Tellegen, Winkel, Wijnberg-Williams, & Laros, 1998). However, to avoid heterogeneity of assessment tools, these results were not used in the current study. The demographics of our final study group are presented in Table 1. Difference in sex distribution is significant, $p = .006$ (binomial test). Cardiac anomalies were found in 35 children (54%) and included Tetralogy of Fallot, ventricular septal defect and interrupted aortic arch. The