Neuropsychological, Learning and Psychosocial Profile of Primary School Aged Children with the Velo-Cardio-Facial Syndrome (22q11 Deletion): Evidence for a Nonverbal Learning Disability?*

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ABSTRACT

In this exploratory study, the neuropsychological and learning profile of nine primary school age children with velo-cardio-facial syndrome (VCFS) was studied by systematic neuropsychological testing. In five out of nine children, the following profile was found: a VIQ-PIQ discrepancy (in favor of the VIQ), significantly better scores (.05 level) for reading (decoding) and spelling compared to arithmetic, deficient tactile-perceptual skills (difficulties mainly on the left side of the body), weak but not deficient visual-perceptual abilities, deficient visual-spatial skills, extremely poor psychomotor skills (gross motor skills more deficient than fine motor skills), problems with processing of new and complex material, poor visual attention, good auditory memory and relatively good language skills. These findings correspond to the pattern of neuropsychological assets and deficits that has been described for the syndrome of nonverbal learning disabilities (NLD) (Rourke, 1987, 1988, 1989, 1995). The psychosocial profile of all nine children with VCFS also correspond to that of children with NLD.

Further studies on the relationship between cognitive function, behavior, psychiatric disorder and abnormalities in brain anatomy in young people with VCFS will be needed. In clinical practice, it is worthwhile exploring in greater depth the neuropsychological functions of children with VCFS to rule out NLD, since they may benefit from specific remediation following the learning principles of the NLD-treatment.

Velo-cardio-facial syndrome, also known as Shprintzen syndrome (Shprintzen et al., 1978), is a relatively common congenital anomaly syndrome estimated to affect between 1 in 4000 to 1 in 5000 individuals (Devriendt et al., 1998). Its major features are a cleft palate or velopharyngeal insufficiency (with hypernasal speech as the most common presenting symptom), cardiac anomalies, a characteristic facial appearance, learning disabilities or mental retardation. The discovery of a submicroscopic deletion in chromosome 22q11 (Scambler, Kelly, & Lindsay, 1992) in the majority of patients has confirmed that VCFS is a specific syndrome. In most patients the deletion occurs de novo, but familial occurrence with an affected parent is noted in 15% of the patients (Swillen et al., 1998). Marked variability in the associated somatic anomalies and in intelligence has been found. Learning difficulties have been reported frequently. Mental retardation (defined as Full Scale IQ < 70 or > 2 Standard Deviations below the mean) is found in 45% of the cases, and the incidence of mental retardation is significantly

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higher in individuals with a familial deletion than in individuals with a \textit{de novo} deletion (Swillen et al., 1997).

In the group of children with learning difficulties (FSIQ $> 70$), higher verbal IQ than performance IQ (Swillen et al., 1997; Golding-Kushner, Weller, & Shprintzen, 1985; Moss et al., 1995), problems with arithmetic, deficits in attention and concentration, and deficits in visual-spatial-motor abilities have been reported (Golding-Kushner et al., 1985; Swillen et al., 1997; Moss et al., 1999). This pattern of deficits seems to be similar to patterns observed in children who have been identified with nonverbal learning disabilities (NLD). Children and adolescents with NLD syndrome display a pattern of neuropsychological assets and deficits characterized by well developed rote verbal skills within the context of relatively poor psychomotor, tactile-perceptual, visual-spatial-organizational, and non-verbal problem-solving abilities (Harnadak & Rourke, 1994; Rourke, 1989; Rourke, 1995). Problems with social perception, judgement and interactive skills are also reported in children with NLD.

Since the original description of the syndrome (Shprintzen et al., 1978), there is now substantial insight into the medical aspects of the condition. However, we are at an early stage in understanding the complex mechanisms in which the learning difficulties arise in this syndrome. In the present study we examined the neuropsychological abilities, academic achievement and psychosocial profile of primary school age children with VCFS.

The research-project has an explorative character.

METHOD

Subjects
Subjects of the study group were recruited from a group of 134 patients followed by the multidisciplinary team for persons with VCFS at the Center for Human Genetics in Leuven.

Inclusion criteria for the study group were: (a) VCFS confirmed by a 22q11 deletion (shown by FISH: fluorescence \textit{in situ} hybridization using probe DO832) ; (b) age between 6–12 years (primary school age); (c) Full Scale IQ (FSIQ) $> 70$ on the most recent intelligence test available.

Thirteen children met the three criteria. Children were recruited by letter and by telephone. Two families did not wish to participate in the study. The original group studied consisted of eleven children. However, at retesting of intelligence, two children had a FSIQ below 70 and were excluded for further analysis. So, nine children (4 boys, 5 girls) remained who fulfilled the three criteria. Their age varied from 6 years 10 months to 12 years 10 months (mean age: 10.5).

Table 1 gives a summary of age at the time of testing, sex, origin of the deletion (familial or \textit{de novo}), important medical data, and FSIQ of the nine subjects.

In all nine children the 22q11 deletion occurred \textit{de novo}, and all nine had a heart defect. In seven of the children the heart defect was successfully corrected; in two children (patient no. 3 and patient no. 7) residual cyanosis remained which limited physical exercise.

One child (patient no. 8) had a cleft palate which was surgically repaired and one child (patient no. 9) had a pharyngoplasty for severe velopharyngeal insufficiency.

Procedures
The children were evaluated individually using an assessment battery including measures of intellectual, neuropsychological, linguistic and academic functioning. Children were tested by experienced examiners, and the test battery was presented in a standardized fashion. Parents filled out a form concerning the behavior and psychosocial functioning of their child. The following measures were administered in order to assess neuropsychological abilities and academic skills:

1. Intelligence – The Wechsler Intelligence Scale for Children-Revised (WISC-R) (Van Haasen et al., 1986);
2. Visual Perceptual Skills – Test of Visual Perceptual Skills (TVPS) (Gardner, 1982);
3. Visual Attention – Bourdon-Vos dot-test (Vos, 1988);
4. Auditory Memory – 15 words from Rey (Rey, 1958);
5. Intermodal (visual and auditory) Memory – subtest Name Learning of the Revised Amsterdam Child Intelligence test (RAKIT) (Bleichrodt, Drenth, Zaal, & Resing, 1984);
6. Tactile Perceptual Skills – Finger localization (Benton, Sivan, de Hamsher, Varney & Spreen, 1994);
Table 1. Medical Data of the Nine Children.

<table>
<thead>
<tr>
<th>Subject</th>
<th>Age (y; m)</th>
<th>Sex</th>
<th>Origin of the Deletion</th>
<th>Heart Defect</th>
<th>VPI</th>
<th>Other Medical Problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6; 10</td>
<td>F</td>
<td>De novo</td>
<td>Truncus arteriosus</td>
<td>++</td>
<td>Feeding problems in infancy; Hypocalcemia; Bilateral VUR;</td>
</tr>
<tr>
<td>2</td>
<td>7; 1</td>
<td>F</td>
<td>De novo</td>
<td>VSD + P.S.</td>
<td>+</td>
<td>Nasal reflux</td>
</tr>
<tr>
<td>3</td>
<td>8; 9</td>
<td>M</td>
<td>De novo</td>
<td>Extreme TOF + right aortic arch</td>
<td>+</td>
<td>Constipation; Feeding problems in infancy</td>
</tr>
<tr>
<td>4</td>
<td>10; 2</td>
<td>M</td>
<td>De novo</td>
<td>TOF + right aortic arch</td>
<td>+</td>
<td>Constipation; Laryngomalacia</td>
</tr>
<tr>
<td>5</td>
<td>11; 6</td>
<td>F</td>
<td>De novo</td>
<td>TOF + right aortic arch</td>
<td>+</td>
<td>Hypocalcemia</td>
</tr>
<tr>
<td>6</td>
<td>12; 6</td>
<td>M</td>
<td>De novo</td>
<td>TOF + right aortic arch</td>
<td>-</td>
<td>Bilateral VUR;</td>
</tr>
<tr>
<td>7</td>
<td>12; 5</td>
<td>F</td>
<td>De novo</td>
<td>TOF + right aortic arch</td>
<td>+</td>
<td>CP</td>
</tr>
<tr>
<td>8</td>
<td>12; 6</td>
<td>F</td>
<td>De novo</td>
<td>VSD + tetralogy of Fallot</td>
<td>+</td>
<td>PP</td>
</tr>
<tr>
<td>9</td>
<td>12; 10</td>
<td>F</td>
<td>De novo</td>
<td>Tetralogy of Fallot</td>
<td>+</td>
<td>PP</td>
</tr>
</tbody>
</table>

Note: TOF: Tetralogy of Fallot; VSD: Ventricular Septum Defect; P.S.: pulmonary stenosis; CP: Cleft Palate; PP: pharyngoplasty; VUR: vesico-ureteral reflux.
To avoid the risk for type 1 error, Bonferonni correction was done (p-value was multiplied by 10).

Table 2. Intelligence Results (WISC-R) of the Group Studied (n = 9).

<table>
<thead>
<tr>
<th>Subject n°</th>
<th>Age (y; m)</th>
<th>Sex</th>
<th>TIQ</th>
<th>TVIQ</th>
<th>TPIQ</th>
<th>F1IQ</th>
<th>F2IQ</th>
<th>F3IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6;10</td>
<td>F</td>
<td>81</td>
<td>93</td>
<td>74</td>
<td>95</td>
<td>76</td>
<td>82</td>
</tr>
<tr>
<td>2</td>
<td>7;1</td>
<td>F</td>
<td>75</td>
<td>82</td>
<td>73</td>
<td>91</td>
<td>67</td>
<td>82</td>
</tr>
<tr>
<td>3</td>
<td>7;9</td>
<td>M</td>
<td>73</td>
<td>73</td>
<td>79</td>
<td>79</td>
<td>82</td>
<td>68</td>
</tr>
<tr>
<td>4</td>
<td>10;2</td>
<td>M</td>
<td>71</td>
<td>76</td>
<td>70</td>
<td>75</td>
<td>70</td>
<td>77</td>
</tr>
<tr>
<td>5</td>
<td>11;6</td>
<td>F</td>
<td>70</td>
<td>73</td>
<td>71</td>
<td>80</td>
<td>69</td>
<td>79</td>
</tr>
<tr>
<td>6</td>
<td>12;0</td>
<td>M</td>
<td>70</td>
<td>76</td>
<td>68</td>
<td>79</td>
<td>70</td>
<td>72</td>
</tr>
<tr>
<td>7</td>
<td>12;5</td>
<td>M</td>
<td>73</td>
<td>73</td>
<td>77</td>
<td>71</td>
<td>75</td>
<td>86</td>
</tr>
<tr>
<td>8</td>
<td>12;6</td>
<td>F</td>
<td>75</td>
<td>81</td>
<td>75</td>
<td>90</td>
<td>74</td>
<td>74</td>
</tr>
<tr>
<td>9</td>
<td>12;10</td>
<td>F</td>
<td>78</td>
<td>94</td>
<td>68</td>
<td>93</td>
<td>59</td>
<td>98</td>
</tr>
<tr>
<td>M (SD)</td>
<td>10;5</td>
<td></td>
<td>74</td>
<td>80.11</td>
<td>72.78</td>
<td>83.66</td>
<td>71.33</td>
<td>79.77</td>
</tr>
</tbody>
</table>

RESULTS

Intellectual Profile
Mean FSIQ was 74 (SD = 3.70).

Profile analysis revealed a non-significant ($t(8) = 2.098; p = .692^1$) difference between VIQ ($M = 80.11$) and PIQ ($M = 72.78$) for the group. Seven children (77.8%) had better results on verbal tasks relative to visuo-spatial tasks, but only two of them had a statistically significant (.05) discrepancy (see table 2). A difference of at least 10 points was considered clinically significant (Rourke, 1971, 1973, in Rourke, 1989). Three children (3/9) had a clinically significant VIQ-PIQ discrepancy of 10 points. The remaining six children (6/9) did not have a clinically significant VIQ-PIQ discrepancy of 10 points.

Analysis of the IQ factor scores, showed a non-statistically ($t(8) = 2.982; p = .181$) but a clinically significant difference (difference > 10 IQ-points) between the verbal comprehension factor F1IQ ($M = 83.67$) and the perceptual-organizational factor F2IQ ($M = 71.33$) (see Fig. 1).

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1 To avoid the risk for type 1 error, Bonferonni correction was done (p-value was multiplied by 10)
1). Five children had an F1IQ more than 10 points higher than the F2IQ; four of them had F1IQ’s that were significantly (.05) better.

The third factor, the freedom from distractibility factor F3IQ \((M = 79.77)\) was not significantly lower than the verbal comprehension F1IQ factor or the perceptual-organizational F2IQ factor.

**Academic Skills**

Seven children were able to do an arithmetic (KRT) and reading test at their school level. At the time of the testing, the two youngest children did not reach a sufficient reading and arithmetic level to perform the tests. Likewise, only six children were able to do the spelling test. Five children were also presented a reading comprehension test. Mean scores for the school assessment battery are given in table 3.

Comparing mean results on the Arithmetic test and those of reading (decoding), the discrepancy is obvious. The difference between the mean results of the KRT and the Brus \((p = .036)\) and of the KRT and the Klepel \((p = .018)\) is more than 1 standard deviation and can be rated clinically and statistically significant. All children \((n = 7)\) had better scores on reading tests in comparison to the arithmetic test. Six of them \((6/7)\) had a discrepancy of at least one standard deviation, in favor of the reading tests.

Only five children were tested on both reading-comprehension and decoding skills. Mean z-scores are better for reading-comprehension than for decoding, but the difference is negligible.

A comparison of the Spelling and Arithmetic results shows a better group mean for spelling. The difference is nearly one standard deviation. The spelling results of three children of the group \((n = 6)\) were at least one standard deviation better than the results on an arithmetic test. The results on the Praxis (Spelling) were statistically not significantly better \((p = .327)\) than those on the KRT (Arithmetic).

Concerning Spelling, Arithmetic and decoding-skills, the results of the group are similar to those of children with NLD. There is an obvious difficulty in completing Arithmetic tests, while
their reading and spelling skills are quite normal (see Fig. 2). On the other hand, in contrast with children with NLD, the children with VCFS in our study group did not have better results on the reading-decoding test than on the reading-comprehension test.

Tactile Perception
Eight out of nine children (8/9) had a total finger localization score within the borderline or clinical range. The 4 youngest children of the group had a percentile 0. Group results were statistically significant, with a $p$-value of < .001 on the $z$-test. The tactile perception was obviously deficient. Their problems are even worse when children are younger. Correlation between total raw scores and age of the children was .921.

Six children had better raw scores on the right-hand side compared to the left-hand side. Two children scored similarly on both sides. Group mean raw scores were also better on the right-hand side. The difference between the right and the left-hand side is statistically significant ($p = .033^1$). Tactile perceptual problems

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**Table 3. Mean Scores on the School Assessment Battery.**

<table>
<thead>
<tr>
<th>Test</th>
<th>Group Mean ($M$)</th>
<th>z-score</th>
<th>Standard Deviation ($SD$)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arithmetic (KRT) ($n = 7$)</td>
<td>-1.429</td>
<td></td>
<td>(0.45)</td>
</tr>
<tr>
<td>Reading (test 1 – Brus) ($n = 7$)</td>
<td>0.043</td>
<td></td>
<td>(0.911)</td>
</tr>
<tr>
<td>Reading (test 2 – Klepel) ($n = 7$)</td>
<td>-0.05</td>
<td></td>
<td>(0.881)</td>
</tr>
<tr>
<td>Reading Comprehension (Bel) ($n = 5$)</td>
<td>0.19</td>
<td></td>
<td>(0.589)</td>
</tr>
<tr>
<td>Spelling (Praxis) ($n = 6$)</td>
<td>-0.5</td>
<td></td>
<td>(1)</td>
</tr>
</tbody>
</table>

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**Fig. 2.** Comparison between reading (decoding), spelling, and arithmetic in the group studied ($n = 7$).
seem therefore to be more obvious on the left side of the body.

The lowest raw scores were on the most complex part of the test (simultaneously touching two fingers behind a box, when the child cannot see his fingers). The difference between the first part (touching one finger while the child is seeing his fingers) and the third and most complex part of the test is statistically significant ($p = .003^1$) as is the difference between the second part (touching one finger while the child cannot see his finger) and the third part ($p = .007^1$).

**Visual Perception**

Three subtests of the TVPS were presented: Visual Form Constancy, Figure–Ground and Visual Closure. Mean scores of the three subtests of all the children ($n = 9$) were calculated. Only one child had a mean $z$-score of at least 0. All the other children had a negative score, of which three (3/9) scored below $–1$. Group mean $z$-score of the three subtests was $–0.79$ which means a weak but not deficient score.

**Psychomotor Skills**

All children had very weak psychomotor skills on the Bruininks-Oseretzky test of motor proficiency. Each child had a total $z$-score of $–1$ or less. The highest total motor percentile reached was percentile 12. Three children had a percentile 1 or even less. Five children (5/9) had a lower score on gross motor skills in comparison to fine motor skills.

On the Visual-motor Integration test, the mean group $z$-score ($z = –0.093$) was situated within the normal range. Six children (6/9) had a standard score below one standard deviation of what could be expected according to their age. The visuo-motor integration skills are deficient for the majority of children with VCFS. Problems with visuo-motor skills increased with age.

**Executive Function**

Fisher, DeLuca, & Rourke (1997) mentioned that deviations of at least one standard deviation for perseverative errors and categories completed can be rated significant. The mean group $z$-score for perseverative errors in our study group was $–0.18$ ($SD = 0.92$) which falls within the normal range. Only one child was making more perseverative errors than could be expected according to age. The other eight children were able to use new strategies if necessary.

Results for categories completed, on the contrary, were weak. Six children (6/9) were not able to fulfill as many categories as their peers. The mean group $z$-score was $–2$ ($SD = 2.2$): we conclude the children had difficulties in finding correct solution strategies.

**Attention**

The ‘Bourdon-Vos dots test’ is a visual attention test. Each child is evaluated for speed and for correctness. There are five speed ($–2 – +2$) and six correctness ($–3 – +2$) categories. A category score of $–1$ or less, can be called deficient. $Z$-scores are available.

With regard to correctness, the mean category-score is $–1.843$ ($SD = 1.855$). Six children (6/9) have category-scores of $–1$ or less. In the speed categories, seven children (7/9) have a $z$-score of $–1$ or $–2$.

Even when the children work slower, they are not working more correctly. Only three children had correctness categories of 0 or better. The other children score in the $–1$, $–2$ or $–3$ category. Four of them were working very impulsively, e.g. not starting at the beginning of the lines, or skipping parts. They all score category $–3$.

**Memory**

The ‘15 words of Rey’, is an auditory memory test. Each child listens to 15 words being read five times and has to repeat as many words as possible after each time. $Z$-scores are available for each time and a total $z$-score can be calculated.

$Z$-scores for the first time they hear the words are variable: 3/9 have a score below $–1$, 3/9 score normal and 3/9 have scores above 1. The mean group $z$-score ($z = 0.48; SD = 1.764$) is within the normal range.

The mean $z$-score for the total group ($z = 0.08; SD = 1.764$) is also situated within the normal range. Only one child has a result that devi-
ates more than one standard deviation from the age-related mean. All the other children (8/9) have normal to above normal results.

**Language**
The 9 children of our study group have scores within the normal range on each subtest of the TVK (Language Tests for Children). Group mean z-scores on each subtest were \(-.6; -.06; -.095; -.045; -.16\).

**Psychosocial Functioning**
The Child Behavior Checklist (CBCL), the Teacher Report Form (TRF) and the Youth Self Report (YSR) each consist of 112 behavior patterns (problem items), and these score as follows: 0 if the item is “not true” for the child; 1 if the item is “somewhat or sometimes true”; and 2 is “very true or often true”.

* T-scores \((M = 50, SD = 10)\) were calculated for each child. For the total problem score, the internalizing (withdrawn, somatic complaints, anxious/depressed) and externalizing (aggressive behavior and delinquent behavior) subscores, a total t-score of 63 or higher (90th percentile or more) is considered to be in the clinical range. * T-scores of 71 or higher (98th percentile or more) for the 8 “small band syndromes” (withdrawn, somatic complaints, anxious/depressed, social problems, thought problems, attention problems, aggressive behavior, delinquent behavior) are found in children with severe behavioral and emotional problems. * T-scores of 66 or higher (> 93rd percentile) are considered to be “of concern”.

All parents filled out the CBCL. The mean group t-score was 62.6 \((SD = 9.30)\), which is at the border of the clinical range \((t > 63)\). Two-thirds of the parents \((6/9)\) score the behavior of their children within the clinical range. The majority of children have internalizing problems: the mean t-score for internalizing behavior problems is 62.7 \((SD = 6.946)\), which is almost 10 points higher than the mean t-score for externalizing behavior problems \((t = 54.8; SD = 14.03)\). Most problems found concerned social skills (problems in relationships with peers), attention problems and withdrawal. Only one child had more externalizing than internalizing problems. She is the youngest girl of the group studied.

Seven teachers filled out the TRF. The mean group t-score on the TRF is 59 \((SD = 5.88)\). According to the teachers, the children with VCFS had more internalizing problem behavior \((t = 56.43; SD = 17.93)\) than externalizing problem behavior \((t = 52.43; SD = 6.68)\). 4/7 of the teachers rated the internalizing problem behavior of their pupils within the clinical range; none of the pupils \((0/7)\) had a clinical score for externalizing problem behavior. The two highest scores on the small band syndromes are on “withdrawn” \((t = 64.428; SD = 9.18)\) and on “social problems” \((t = 62.857; SD = 7.081)\).

All six children of 11 years and older, filled out the YSR. The mean group t-score on the YSR was 55 \((SD = 5.138)\). All had higher scores for internalizing problem behavior patterns \((mean t-score = 54.33; SD = 8.04)\) than for externalizing problem behavior \((mean t-score = 46.5; SD = 6.28)\). The two highest scores on the small band syndromes were on “social problems” \((t = 66.83; SD = 10.048)\) and on “attention problems” \((t = 60; SD = 7.924)\). Four of the six adolescents \((4/6)\) had a score within the borderline or clinical range for “social problems”, two scored within the borderline range for “attention problems”.

**DISCUSSION**

Before interpreting the results of the current investigation, two important considerations should be noted. First, it is interesting to note that all children in this study had the 22q11 deletion *de novo*. In an earlier study (Swillen et al., 1997) we have shown that the incidence of intellectual disability/mental retardation is correlated with the mode of inheritance: mental retardation is more frequent in the group of children who inherited the deletion from one of their parents. Second, the mean FSIQ of our study group is 74 \((range 70–81; SD = 3.70)\). In most other studies on children with a NLD, the intelligence range is 70–100. Our study group is therefore at the bor-
Table 4. Comparison NLD-VCFS.

<table>
<thead>
<tr>
<th>Domains</th>
<th>NLD</th>
<th>VCFS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intelligence</td>
<td>VIQ &gt; PIQ (10 points or &gt;)</td>
<td>VIQ &gt; PIQ (n.s.)</td>
</tr>
<tr>
<td>Academic Skills</td>
<td>R and S &gt; Arithmetic</td>
<td>R and S &gt; Arithmetic (.05)*</td>
</tr>
<tr>
<td>Visual-Perceptual-Organizational Skills</td>
<td>Deficient</td>
<td>Weak but not deficient</td>
</tr>
<tr>
<td>Psychomotor Skills</td>
<td>Deficient</td>
<td>Z-scores of –1 or more*</td>
</tr>
<tr>
<td>Complex and New Material</td>
<td>Problematic</td>
<td>Problematic: Z-scores of –2*</td>
</tr>
<tr>
<td>Tactile-Perceptual Skills</td>
<td>Left side of the body</td>
<td>Right &gt; left hand side (.05)*</td>
</tr>
<tr>
<td>Psychosocial Skills</td>
<td>Poor social and interactive skills</td>
<td>Withdrawal, poor social skills, internalizing problems*</td>
</tr>
</tbody>
</table>

Note. * Criteria are met.

Although Fuerst et al. (1995) have speculated on the relationship of VCFS (22q11 deletion) and NLD, this study is the first to provide in depth systematic data on this issue. In the earlier study (Swillen et al., 1997) we reported that the IQ scores of 11 school-aged (7–16 years old) children with a mean full-scale IQ score in the 70s with verbal IQ scores were significantly higher than the performance IQ scores. These data confirmed a report by Moss et al. (1995) of a verbal/performance “split”. The same researchers (1999) recently reported data on the psychoeducational profile of 33 children and adolescents with the 22q11 deletion and concluded that the IQ and academic profiles are reminiscent of a NLD although achievement was not inconsistent with IQ. The results of our study are summarized in table 4 and show that for many neuropsychological functions, VCFS and NLD have the same deficiencies.

Analysis of the total group shows a clear NLD profile. Like children with NLD, children with VCFS show a VIQ-PIQ discrepancy in their intelligence profile in favor of the verbal IQ. When we compare the verbal comprehension factor (F1IQ; \( M = 83.67 \)) with the perceptual-organizational factor (F2IQ; \( M = 71.33 \)), this discrepancy is clinically significant.

With regard to academic achievement, we found, as with NLD-children, a statistically significant difference (.05 level) between the reading (decoding) skills and arithmetic. Also, the children with VCFS in the group studied performed better on spelling compared to arithmetic. Therefore, as in children with NLD, primary school age children with VCFS exhibited severe difficulties in mechanical arithmetic and average to above-average single-word reading and spelling skills. However, we noted two differences in the learning profile of children with VCFS in comparison to children with a NLD. First, we did not find significantly better results for reading (decoding) in comparison to reading comprehension. It would be interesting to explore the reading comprehension skills of NLD children further. It is our hypothesis that scores of NLD children on a reading comprehension test may depend on the way the test is constructed. We think that NLD children will fail on a reading comprehension test only if the child has to infer the answers from the text. In other words, if the NLD child can find the answers literally in the text, he will perform on the same level as his decoding skills and this may explain the good scores of our group. Second, the children with VCFS did not spell hyperphonetically. Several studies (Sweeny & Rourke, 1978, 1985) have found that children with NLD tend to make phonetically accurate spelling mistakes. These studies were conducted with English-speaking subjects. However the native language of our group was Dutch, a language with a spelling that is phonetically much more accurate than English. It is our clinical experience that the dimension of phonetic accuracy is not as significant in the assessment of Dutch speaking children with NLD, and we believe that this may be accounted
We found the following pattern of strengths and weaknesses for our group on the neuropsychological test battery: deficient tactile-perceptual skills (more problems on the left-hand side of the body), weak but not deficient visual-perceptual abilities, deficient visual-spatial skills, extremely poor psychomotor skills (complex motor skills more deficient than simple motor skills), problems with processing of new and complex material (WCST), poor visual attention, good auditory memory and relatively good language skills. These findings correspond to the pattern of neuropsychological assets and deficits that has been described for the NLD syndrome (Rourke, 1987, 1988b, 1989). It seems, however, that the deficiencies in psychomotor skills of children with VCFS are more pronounced, compared to children with NLD (Cracco, personal communication). Certain medical complications of VCFS such as a congenital heart defect (Tetralogy of Fallot, conotruncal heart defect) or the hypotonia, may in part explain the deficient gross motor skills. The finding of poorer tactile performance for the left than the right hand could suggest that the right hemisphere is more compromised by this condition than the left. However, until now, there are no data from brain imaging studies in children with VCFS that confirm this hypothesis. To date, most structural neuroimaging studies of VCFS individuals have been qualitative and reported the presence of an agenesis of the corpus calosum, a small cerebellum, periventricular white matter abnormalities, white matter hyperintensities, cavum septum pellucidum, smaller total brain volume, and enlarged Sylvian fissures (Mitnick et al., 1994; Lynch et al., 1995; Chow et al., 1999; Bingham et al., 1997).

The psychosocial profile of children with VCFS seems to be similar to that of children with NLD. Parents, teachers and youngsters themselves report more internalizing problem behavior than externalizing problem behavior. All three sets of informants most frequently report social problems (especially problems in relationships with peers), withdrawn behavior, and attention problems. Children with NLD are particularly vulnerable to psychosocial problems. These problems tend to get worse when they grow older and extend well into adulthood with a higher prevalence of psychiatric disorders (Rourke, Young, & Leenaars, 1989). Previous reports (Goldberg et al., 1993; Shprintzen et al., 1992) in adolescents and adults with VCFS found that at least 10% of them will develop psychiatric disorders: symptoms most frequently described are those associated with depression, anxiety disorders and psychotic disorders (including schizophrenia). A good follow-up with special concern for the development of social skills and self-image of VCFS children is indispensable, and parents, teachers and professionals must be alert for behavioral changes.

Because of the small sample, and the lack of a control group, the data from this study should be treated with caution. For future research, multicenter studies on learning problems in VCFS are needed including control groups (children with velopharyngeal insufficiency and learning problems, children with a congenital heart disease and learning problems). However, an exploratory study like the present one, can be the first step in unraveling the underlying complex mechanisms by which the learning problems arise in this syndrome.

In future, studies on the relationship between cognitive function, behavior, psychiatric disorder and abnormalities in brain anatomy in young people with VCFS will be highly needed.

In clinical practice, it is worthwhile exploring in greater depth the neuropsychological functions of children with VCFS to rule out NLD, since they may benefit from specific remediation following the learning principles of the NLD-treatment as described by Rourke (1995).

REFERENCES


