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A Cognitive Neuropsychological Analysis of Mathematical Abilities: The Mccloskey Model Applied to Children with Velocardiofacial Syndrome

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Abstract

Children with velocardiofacial syndrome (VCFS) frequently display mathematics difficulties. Research into the specific mathematical strengths and weaknesses in this population is relatively limited. Previous studies into mathematical abilities in VCFS have generally not included an IQ-matched control group. The aim of this study was to assess mathematical ability in children with VCFS, within a cognitive neuropsychological framework, and to compare their performance to a chronologically age- and IQ-matched control group. Twenty-three children with VCFS (aged 8 to 14 years) were administered a range of tasks designed to assess components of McCloskey, Caramazza and Basili's (1985) cognitive neuropsychological model of mathematics. Children with VCFS did not differ from controls in general mathematical ability, nor did the groups differ on any of the more specific cognitive neuropsychological tasks designed to assess the McCloskey model of mathematics. Results indicate that the weakness in mathematics well-documented in children with VCFS may perhaps reflect overall lowered IQ and associated cognitive dysfunction.

INTRODUCTION

Velocardiofacial syndrome (VCFS) is a genetic disorder affecting approximately 1 per 2,000 to 4,000 live births [1]. VCFS results from a 1.5 to 3Mb micro deletion on the long (q) arm of chromosome 22 [2]. Children with VCFS typically demonstrate lower than average general intellectual abilities, falling within the mild to borderline impaired range. However, verbal abilities are often significantly higher than nonverbal abilities [3,4]. In addition to their intellectual disability, individuals with VCFS also display deficits in a range of more specific cognitive functions, including: attention [4,5], executive functioning [6], emotional processing, visuospatial processing [7], and working memory [5].

Research investigating academic functioning in VCFS suggests relative impairments in mathematics compared to literacy. Moreover, it seems that these mathematical difficulties may be present as young as kindergarten age [8]. Structural and functional imaging studies have highlighted parietal and prefrontal aberrations in the brains of individuals with VCFS [9]. Neuro imaging studies indicate that functions of the parietal lobes may be particularly related to mathematical difficulties in children with VCFS, as is the case in the typically developing population [10]. Associations between short-term memory abilities and mathematics have been found in children with VCFS [11,12], mirroring findings in the normal population. This suggests that short-term memory and mathematics either rely on the same functions, or that they engage similar brain regions.

More recently, research studies into mathematical abilities in VCFS have employed cognitive neuropsychological models [11,13,14]. Such models propose that discrete mental processes (or modules) are relevant for cognitive number processing and calculation [15]. Two leading cognitive neuropsychological models of mathematical processing have been proposed. McCloskey et al. [16], proposed a model of number processing and calculation. Dehaene et al., proposed the second model, known as the triple-code model of number processing.

THE MCCLOSKEY MODEL OF NUMBER PROCESSING AND CALCULATION

The McCloskey model is one of the most widely accepted cognitive neuropsychological models of mathematics and is well-supported by empirical data [16-18]. The model partitions number processing and calculation mechanisms into several functionally distinct components (Figure 1). At the most basic level, a distinction is drawn between the calculation system (Figure 1 i), which encompasses processing components

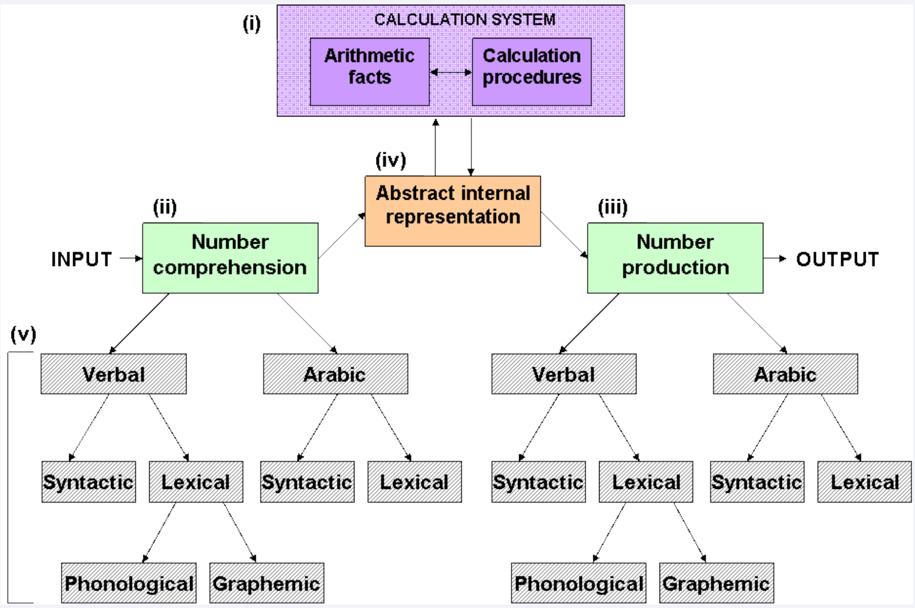

Figure 1 The McCloskey Cognitive Neuropsychological Model of Number Processing and Calculation.

required specifically for carrying out calculations, and the number processing system (Figure 1 ii & iii). Within the number processing system, the mechanisms for number comprehension (Figure 1, ii) are distinct from those for number production (Figure 1, iii). At the core of the model is an abstract internal representation (Figure 1 iv), which is postulated to represent (in abstract form) the basic quantities in a number, such as the power of ten associated with each (e.g. nine tens and three ones for the number 93). The boxes at the bottom of the model (Figure 1v) illustrate the different components of number comprehension and production described by [17].

Contrary to traditional cognitive neuropsychological models, the McCloskey model does not allow each specific component to be tested separately. This is because some of the modules are dependent on the functioning of other modules (for example, calculation procedures rely on intact number processing). Therefore, testing of the McCloskey model involves examination of different streams of processing, which incorporate several different components.

Number processing

Number processing involves both number comprehension and number production. Number comprehension converts numerical inputs into internal abstract representations, which are subsequently available for further processing such as carrying out a calculation. The number production system converts abstract representations into Arabic or verbal format to produce a written or spoken number. The model proposes that within both number comprehension and number production, a number of distinct components exist (Figure 1v). The components for processing Arabic numbers (i.e. numbers in digit form, such as 387) are distinct from those for processing verbal numbers (i.e. numbers written or spoken number words, such as three hundred and eighty seven). Within the verbal and Arabic comprehension and production components, lexical processing is distinguished from syntactic processing. Syntactic processing involves the processing of relations among elements in order to comprehend or produce numbers as a whole. Lexical processing, however, relates to the comprehension and production of the individual elements in a number (e.g. the word seven, or digit *7*). Finally, within the lexical processing mechanisms of the verbal number system, a distinction is drawn between production/ comprehension of spoken numbers (phonological processing components) and written numbers (graphemic processing components).

The calculation system

The calculation system of the McCloskey model (Figure 1,i) is based on a unitary representation of quantity regardless of the code in which the input is presented (for example, verbal or Arabic number forms). The calculation system relies on intact number processing, so that if impairments exist in number comprehension or production, calculation will also be impaired. However, calculation also requires three additional capabilities: 1) processing of operational symbols (*+*) or words (*plus*) that identify the operation to be performed; 2) retrieval of basic arithmetic facts (e.g. times tables); and 3) execution of calculation procedures (e.g. the order in which to proceed in long division).

The McCloskey model has been used to investigate mathematical abilities in girls with Turner syndrome [19], another genetic condition associated with mathematical difficulties. Mathematical deficits in girls with Turner syndrome were primarily procedural in nature, that is, they had difficulty implementing procedural knowledge in calculation (such as, the order in which to proceed in long division). Despite differences between VCFS and Turner syndrome in terms of genetic aetiology, physical manifestation, and global intelligence, both clinical populations exhibit numerical and mathematical impairments as well as visuospatial deficits. Children with VCFS and Turner syndrome also appear to process some mathematically-relevant information in the same way. For example, VCFS and Turner syndrome display similarities in terms of number processing and visual attention [20], and they share at least some aspects of abnormality in parietal regions [21]. This might suggest that the two syndromes also share similar mathematical difficulties, possibly both displaying difficulties in calculation procedures (Figure 1, i).

DEHAENE'S TRIPLE-CODE MODEL OF NUMBER PROCESSING

In contrast to the McCloskey model, Dehaene's model does not separate number processing and calculation, rather it postulates that three systems, or codes, each contribute to different aspects of number processing and calculation [22]. Dehaene's three codes are as follows [23]:

- The verbal system which represents numerals as sequences of words. It is the primary system for accessing arithmetic facts from memory, for example recalling times table facts.
- The quantity system which is a nonverbal semantic representation of the size and distance relations between numbers (e.g. judging which of two numbers is larger). This system subserves semantic knowledge about numerical quantities (e.g. magnitude comparisons) and is also engaged in single-digit calculation when the answer is not stored in long-term memory (and therefore cannot be retrieved by the verbal system).
- The visual Arabic system where numbers are encoded as strings of digits. This system is responsible for multidigit operations.

While the McCloskey and triple-code models envisage mathematical processing in quite different ways, the two models are not entirely incompatible. The primary difference between the two models is the role of the abstract representation of quantity (the abstract internal representation component in McCloskey's model; the quantity system in Dehaene's model). According to the McCloskey model, this representation is activated during all mathematical tasks, including simple number processing. In contrast, the triple-code model stipulates that some mathematical tasks do not involve the quantity system, namely number transcoding and retrieval of arithmetic facts.

Application of dehaene's model to children with VCFS

Dehaene's model of number processing [22] has been adopted as a framework in two cognitive neuropsychological studies of mathematical abilities in children with VCFS. Results from both studies demonstrate that, in terms of Dehaene's model, the quantity system of number processing is primarily affected in children with VCFS, whilst the verbal and visual Arabic system appeared unaffected. Furthermore, in terms of the nature of deficit, De Smedt et al., suggested that mathematical difficulties in children with VCFS are primarily procedural in nature, whereby they displayed difficulties in the application of rules and strategies in calculations. Procedural deficits have also been described in Turner syndrome [19].

Framework for the current study

To date, two studies have examined the mathematical abilities of individuals with VCFS using the triple-code model proposed by Dehaene [22]. The current study employed the framework of the McCloskey model which has not yet been used to investigate mathematical abilities in children with VCFS. Furthermore, as the McCloskey model has previously been applied to girls with Turner syndrome [19], employing identical tests to the current study, this allows for an indirect comparison between the two syndromes.

Study aims and predictions

The overall aim of this study was to investigate mathematical abilities, employing the McCloskey model, in children with VCFS compared to a control group of children matched by chronological age and IQ. Age and IQ are both important predictors of mathematics [24]. In addition, in these study children with VCFS and IQ < 70 were not excluded, whereas previous studies of this nature in VCFS have been limited to children with borderline to normal intelligence [11,13,14].

The first aim of this study was to examine general academic functioning in children with VCFS. Academic functioning was examined using two methods. First, children with VCFS were compared to controls on general mathematical ability to determine whether groups were comparable on a standardised mathematics task (Measures). Second, within-groups discrepancies between mathematics and single word reading were examined. It was predicted that there would be no difference between children with VCFS and controls on standardised tasks of mathematics or single word reading. The performance of children with VCFS on standardised mathematics tasks appear to be consistent with their IQ [3].

The second aim was to examine the mathematical abilities of children with VCFS employing the McCloskey model. The performance of children with VCFS and controls were measured on a range of mathematical tasks specifically designed to assess aspects of number processing and calculation [25], from the McCloskey model (Measures). The following predictions were proposed:

1. Children with VCFS will, on average, display intact performance on the number processing component of McCloskey's model.
2. Within the number processing system, children with VCFS will show a weakness in their ability to accurately judge relative magnitudes of Arabic numbers (number comprehension and internal abstract representation components), as compared to their own performance on other number processing tasks and compared to the control group.
3. Within the calculation system, children with VCFS will display a relative weakness on the arithmetical problems task (calculation procedures component), as compared to their own performance on retrieval of addition and multiplication facts (addition facts component), and relative to controls.

METHOD

Participants

Children with VCFS: Twenty-three children with VCFS (aged 8 to 14 years) were recruited through the database at the Genetics

Clinic at The Children's Hospital at West mead, Sydney, Australia, and through an advertisement posted in the VCFS Foundation of New South Wales newsletter. Children aged 8 to 16 years were considered for inclusion in the study. This age range was chosen because the focus of the study was to examine the mathematical profile of primary and high school students (i.e. up to 16 years) with VCFS. Diagnosis of VCFS was confirmed by fluorescent in-situ hybridization, obtained through medical records, and in the majority of children (95%) the deletion occurred de novo (i.e. no family history of VCFS). Ethics approval was obtained prior to recruitment. Children were excluded if they: were not fluent with English; had frequent seizures (one per month) and/or were on anti-epileptic medication; had been administered the Wechsler Intelligence Scale for Children [26,27] in the last 12 months; were on medication that could affect cognitive functioning (except for stimulant medication).

A recruitment letter and information sheet was mailed to 55 families with children with VCFS from the hospital database. Fifteen families responded (27%) to express their interest in taking part in the study. Four children did not meet the inclusion criteria, and were, therefore, excluded, leaving 11 children with VCFS identified through the hospital database. Fifteen letters were sent to families through the VCFS foundation and 12 replied and were met inclusion criteria. Of the total sample of 33 children recruited through the VCFS foundation and the hospital database, 23 were included in data analysis as 23 suitable control children were obtained to match each participant with VCFS on chronological age and FSIQ.

Controls: The current study compared a group of children with VCFS to chronological age- and IQ-matched controls. In addition to the exclusion criteria used for children with VCFS, controls were selected if they had no diagnosed genetic condition (such as Down's syndrome, Fragile X or Turner Syndrome), or any medical conditions that may impact on cognitive functioning such as diagnosed head injury, brain tumours, hydrocephalus, visual or hearing problems. Children were individually matched to children with VCFS by chronological age, within 9 months (mean age difference = 4.4 months), and on the Full Scale IQ (FSIQ) from the Wechsler Intelligence Scale for Children – 4th Edition ([WISC-IV] [27], within 5 standard IQ points (mean FSIQ difference = 3.0 points). Twenty-three controls were recruited through the NSW Department of Education and Training (DET), Australian Independent Schools (AIS) and Catholic Education Offices (CEOs) in the Sydney metropolitan region.

Controls were recruited through the NSW Department of Education and Training (DET), Australian Independent Schools (AIS) and Catholic Education Offices (CEOs) in the Sydney metropolitan region. One hundred and eighty two DET and AIS schools were randomly selected from a database and were contacted via mail. Of these, 40 declined to take part in the study, 125 did not respond and 17 agreed to pass on study information sheets to parents of children within the specified age range. The same exclusion criteria used for participants with VCFS were adopted for the control group. Thirty five children were identified through this process and were administered an IQ and academic assessment to determine whether they could be matched to a child with VCFS. Of these, 23 children could be matched by age and IQ to a child with VCFS and were therefore included in the study.

The final study group consisted of 23 children with VCFS and an age- and IQ-matched control group of 23 children.

Measures

Intellectual abilities: The Wechsler Intelligence Scale for Children - Fourth Edition [28] was used to assess intellectual abilities. It yields a Full Scale IQ (FSIQ) score as well as a Verbal Comprehension Index (VCI) which assesses verbal intelligence, and a Perceptual Reasoning Index (PRI) which is an index of nonverbal intelligence. It also provides an overall Processing Speed Index and Working Memory Index.

General mathematical ability: Wechsler Individual Achievement Test – Second Edition [28]: The Numerical Operations subtest was used to assess whether group differences existed on a standardised measure of general mathematical ability. Numerical Operations is a paper-and-pen test and includes: numeral writing (such as filling in the missing number of: 1, 2, 3, _, 5), calculations (such as: 34 + 25 or 2 x 34), fractions (such as: ½ + ¼), decimals (such as: 0.45 – 0.24), and algebra (such as: simplify 7a + 3b – 2a) .

Single word reading: Wechsler Individual Achievement Test - Second Edition [28]: The Word Reading subtest was used to gain a standardised measure of single word reading.

Components of number processing and calculation

The tasks (described below) were designed by Temple [25] to systematically probe the number processing and calculation mechanisms specified in the McCloskey model [17]. These tasks were adopted in the current study. Each task was completed by each participant in the order listed below.

Number processing tasks

Reading number words aloud: Forty randomly ordered numbers that were written as words, for example, six hundred and forty-eight. These were presented serially to the child (one number on each card) who was then asked to read the number aloud. The stimulus set comprised of ten single digit items (zero to nine), ten 2-digit numbers, ten 3-digit numbers, and ten 4-digit numbers. The task started with single digit numbers and increased in digit size as the task progressed. An item was scored as correct if the number was read aloud with the correct lexical and syntactic frame (i.e. for the example above, "six hundred and forty-eight" was correct, but "six hundred and forty-nine" or "sixty-four eight" were scored as incorrect).

Reading arabic numbers aloud: Forty stimulus cards were presented, using identical numerals to those in the reading number words aloud task, but numerals were presented in Arabic format (e.g. 648). The participant was asked to read the numbers aloud. An item was scored as correct if the number was read aloud with the correct lexical and syntactic frame (i.e. "six hundred and forty-eight" was correct, but "six hundred and forty-nine" or "sixty-four eight" were scored as incorrect).

Writing arabic numbers: Sixteen numbers (a subset of the 40 described for tasks 1 and 2) were spoken aloud and a written

Arabic number response was required (e.g. examiner said: "three hundred and seventy one" and the child were required to write: 371). An item was scored correct only when all digits were written correctly and in the correct order (i.e. 371 was correct, but 317 was scored as incorrect).

Writing number words: Ten numbers (a subset of the 40 described for tasks 1 and 2) were dictated to the child who was asked to write the number down in words (e.g. converting "seven hundred and eleven" into seven hundred and eleven). An item was scored correct when all required words (i.e. numbers and multipliers) were written correctly and in the right order. Exact spelling was not required, although words were required to be recognizable.

Copying arabic numbers. The child was presented with 10 Arabic numbers (a subset of the 40 described for tasks 1 and 2, for example 371) and asked to copy the number onto a separate sheet of paper (e.g. 371). An item was scored correct only when all digits were copied correctly, that is, in the right order (i.e. 371 was correct, but 317 was scored as incorrect).

Magnitude comparisons: The participant was presented with a list of thirty pairs of random Arabic numbers, and asked to circle the larger of the two numbers (e.g. "which is larger, 45 or 79?"). The position of the larger number was randomised. The percentage of items correct and the total time to complete all items were recorded.

CALCULATION SYSTEM

Addition facts

Single digit addition sums were read out aloud to the child (e.g. 9 x 5) and an oral response was required (e.g. 45). Forty-five sums were presented in total, derived from all combinations of the digits 1 to 9 and the largest digit of the pair was always presented first to facilitate the most efficient addition strategy (i.e. starting with the highest number of a pair, then counting up) should counting be required. Questions were presented in the same sequence for each participant. Response times (RT) to correct answers were recorded on a stopwatch and were rounded up to the nearest second (those RTs less than 1 second were recorded as 1 second).

Multiplication facts

As with the addition facts subtest, the child was required to respond orally to dictated multiplication questions. The stimulus set comprised randomly presented times-table questions in which each of the numbers 2 to 9 was paired with all of the possible numbers 2 to 9, resulting in 64 questions. RTs to the nearest second were recorded only for correct answers.

Arithmetical problems

A pen and paper test comprising 20 arithmetical problems. Problems comprised examples of each of the four arithmetical operations (i.e. addition, subtraction, multiplication, division). Figure 2 displays examples of the types of arithmetical problems presented to the child.

Socio-Economic Status

Socio-Economic Indexes for Areas (SEIFA [a measure of socio-economic status within Australia]) scores were determined for each child based on their postcode.

Procedure

Children were individually assessed in a quiet room at The Children's Hospital at Westmead or at their school by a trained clinical psychologist. Assessments were typically conducted over two 2.5 hour sessions, with rest breaks taken as required. The tasks were presented to participants in the same order as presented in the Measures section.

RESULTS

Demographics and IQ standard scores are presented in Table 1. There was no significant difference between the two groups for age, gender, SES or IQ.

General mathematical and reading ability

There were no significant differences between children with VCFS and controls on Numerical Operations and Word Reading, using paired sample t-tests (Table 2). Analysis of performance on Numerical Operations and Word Reading within each group revealed that both children with VCFS and controls performed significantly better on Word Reading than on Numerical Operations *(*$p < .005$ for both groups).

Number processing and calculation

Group statistics (means, standard errors of means) for each component of number processing and calculation are displayed in Figure 3. Group differences on each task were assessed using the non-parametric, Wilcoxon signed rank test as data were

Table 1: Demographic information and mean IQ index scores for the VCFS and Control Groups .

	VCFS	Controls	Group comparison	Effect size
SES, M(SD)	7.3 (3.0)	7.8 (1.9)	NS (p = .48)	d = .22
Sex (M:F)	11:12	12:11	NS (p = .77)	d = .00
Age, M(SD)	10:7 (23 months) range: 8:1 to 14:10	10:7 (22 months) range: 8:0 to 14:11	NS (p = .94)	d = .08
FSIQ, M(SD)	73.1 (9.8)	73.5 (9.5)	NS (p = .55)	d = .05
VCI, M(SD)	77.7 (11.0)	78.4 (12.1)	NS (p = .75)	d = .07
PRI, M(SD)	73.2 (9.9)	76.0 (11.1)	NS (p = .17)	d = .27

Abbreviations: SES: Socioeconomic Status; M: Mean; SD: Standard Deviation; FSIQ: Full Scale IQ; VCI: Verbal Comprehension Index; PRI: Perceptual Reasoning Index

Table 2: Mean WIAT-II Subtest Scores for VCFS and Control Groups.

WIAT-II Subtest	VCFS M (SD)	Control M (SD)	Group Comparisons
Numerical Operations	71.83 (12.31)	70.73 (11.31)	NS (p = .58)
Word Reading	88.50 (15.98)	81.77 (12.39)	NS (p = .14)

Abbreviations: M: Mean; SD: Standard Deviation

9251 + 7271   82 - 49

21 x 2   17) 85

Figure 2 Examples of arithmetical problems.

skewed. All tests were two-tailed, with a significance level of ά = 0.05. When multiple comparisons were conducted, ά level was adjusted using the Holm procedure to reduce the risk of Type I error [29]. It should be acknowledged that mean and standard errors should only be reported as measures of central tendency when describing approximately normally distributed data [30]. While the data were not normally distributed, means and standard errors are displayed in Figure 3 for ease of interpretation. For completeness, medians and interquartile ranges are reported in Table 3.

Number processing

Between groups: All number processing tasks were performed with a high degree of accuracy (medians > 95% accuracy). All children achieved 100% accuracy on the copying Arabic numbers task, so no further analysis was conducted. Comparisons of performances on the four number transcoding tasks revealed no significant group differences (Holm corrected ά levels, ά^1 = .013, ά^2 = .017, ά^3 = .025, ά^4 = .05; Table 2). There were a number of univariate outliers (greater than 3 standard deviations from the mean) for the number processing tasks in both the control group (n = 7) and the VCFS group (n = 5). Outliers were included in the final analyses, but results did not differ when outliers were excluded from analyses.

For magnitude comparisons, no significant differences were found between groups in terms of task accuracy (p = .88) or total completion time (p = .88). Median total completion time on the magnitude comparisons task for the VCFS group was 88 seconds, and for the control group was 101 seconds.

Within groups: Analysing the groups separately, no significant differences were found for VCFS (p = .55) or controls (p = .10) between performances on the four number transcoding tasks (reading and writing Arabic numbers and number words).

Comparisons between mean performance across the four number transcoding tasks and performance on magnitude comparisons (number comprehension task) revealed no significant within group differences for either the VCFS or control group (VCFS: p = .40; Controls: p = .73).

Calculation System

Between groups: As Figure 3 shows, there were no significant differences between the VCFS group and controls on accuracy of performance on addition facts or multiplication facts tasks (see Table 3). With regard to speed of response, the VCFS group performed significantly faster than controls on the addition facts task (p = .02). No significant group differences were found on speed of response for multiplication facts, despite a trend towards faster performance in the VCFS group (p = .07). The VCFS group performed slightly more accurately than controls on the arithmetical problems task, but this failed to reach significance (p = .11).

Within groups: Both groups performed significantly better on addition facts compared to multiplication facts (p < .001 for all comparisons). For both groups, performance on the arithmetical problems task was significantly weaker compared to performances on addition facts and multiplication facts (p < .001 for all comparisons).

DISCUSSION

Overall, as predicted, groups did not differ on general mathematics as measured by the WIAT-II. Contrary to predictions, children with VCFS did not differ from controls on any of the tasks used to assess the McCloskey model, suggesting no relative strengths or weaknesses in number processing or calculation between the two groups.

Standardized academic measures

While previous literature indicates that children with VCFS display weaknesses in general mathematics, typically researchers have not employed IQ- or age-matched control groups. Therefore, it is difficult to determine whether mathematical ability in VCFS is poor relative to IQ. This is especially important given that age and IQ are significant predictors of general mathematical performance [24]. When the groups were appropriately matched on chronological age and IQ, the groups, as predicted, performed similarly on a general mathematics task. The findings are consistent with those of other studies which found that mathematics scores are consistent with levels of intelligence in children with VCFS [3].

Within each group, the discrepancy between general mathematics and single word reading was examined. Consistent with predictions and with previous research [3,7], children with VCFS displayed significantly superior performance on reading (decoding) than mathematics. Contrary to predictions, however,

Table 3: Median, Interquartile Range (IQR) and Group Difference Significance Levels for Each Component of Number Processing and Calculation.

	VCFS Median (IQR)	Control Median (IQR)	Significance (p-value)
Read NW	100 (5)	97.5 (5)	0.69
Read AN	97.5 (8)	100 (5)	0.57
Write AN	100 (0)	100 (0)	0.44
Write NW	100 (20)	100 (0)	0.41
Copy Arabic	100 (0)	100 (0)	--
Mag Comp	96.7 (3)	96.7 (10)	0.88
Addition	95.6 (11)	93.3 (20)	0.25
Multiplication	60.9 (56)	48.4 (64)	0.90
Arithmetic	30 (45)	10 (40)	0.11

Abbreviations: Read NW: Reading Number Words; Read AN: Reading Arabic Numbers; Write NW: Writing Number Words; Write AN: Writing Arabic Numbers; Copy Arabic: Copying Arabic Numbers; Mag Comp: Magnitude Comparisons; Addition: Addition Facts; Multiplication: Multiplication Facts; Arithmetic: Arithmetical Problems

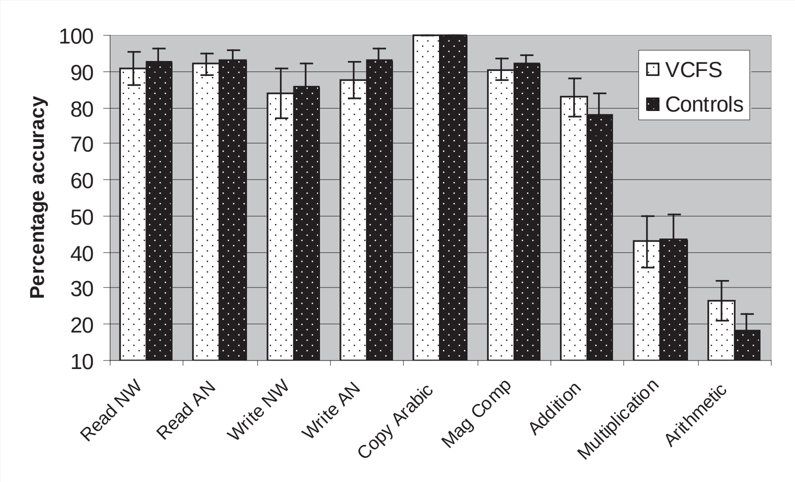

Figure 3 Mean and standard scores for each of the number processing and calculation variables.

the control group also displayed significantly higher performance on reading compared to mathematics. While it is unclear why this was the case, the results may have been skewed by several of the control children who displayed a markedly higher reading score than mathematics score. It may also be that there is a greater focus in schools on reading skills as compared to numeracy, especially for children with an intellectual disability.

McCloskey's model of number processing and calculation

Number processing: Consistent with predictions and consistent with previous research [8,31], both groups achieved a high degree of accuracy on number processing tasks, suggesting intact number comprehension and production. The ceiling effect for number processing tasks in both controls and children with VCFS implies that these are exceptionally easy tasks, particularly in light of the developmental delay of the two groups, and are one of the earliest mathematical competencies that children learn after counting [32].

Although basic number transcoding tasks are straightforward and well-learnt by an early age, magnitude comparisons tasks are more difficult as they require abstraction of numerical relations [33]. Contrary to predictions, both groups performed with a high degree of accuracy on the magnitude comparisons task, with equivalent completion times. Children with VCFS and controls performed equally as well on this task as they did on other tasks of number processing, contrary to predictions. Moreover, our results showed neither reduced accuracy nor increased completion times in children with VCFS, as compared to controls. This finding is in contrast to previous research which has highlighted a weakness in magnitude comparisons in children with VCFS [11,13,14,20,34]. However, this weakness does not relate to reduced accuracy on these tasks. Rather it pertains to an inefficiency of processing information related to quantity, evidenced by slower response times. Group differences may have been evident had individual item response times and accuracy been examined, as has been conducted in previous research [13,34].

Overall, accurate performance on number processing tasks, in both children with VCFS and controls, indicates that the number comprehension, number production, and abstract internal representation components of the McCloskey model (Figure 1, ii, iii, & iv) are intact.

Calculation system: It was predicted that children with VCFS would display a relative weakness on the arithmetical problems task, as compared to their own performance on retrieval of addition and multiplication facts, and relative to controls. With regard to arithmetic fact retrieval, children with VCFS and controls performed addition facts and multiplication facts with equal accuracy. However, children with VCFS were, on average, faster to provide correct answers on the addition facts task compared to controls. According to Dehaene et al. [22], there are two methods for answering these types of task. The first is retrieving the answer from long term memory, which should typically result in an immediate response. The second is by calculating the answer, using a method such as finger counting, which would be associated with response latency. The fact that the VCFS group were significantly faster than controls suggests either that they retrieved a greater number of arithmetic facts from memory than controls, or that they were faster to retrieve these facts from long term memory. This is consistent with previous research which suggests that children with VCFS have well-preserved speeded access to representations in long term memory.

Contrary to predictions, children with VCFS did not differ from controls in their performance on the arithmetical problems task. Children with VCFS and controls were poorer on the arithmetical problems task than on both the addition and multiplication facts tasks. The weakness on the arithmetical problems task in both groups may be due to several factors. First, it may reflect a selective impairment in the calculation procedures component of the McCloskey model, which involves applying procedural knowledge to mathematical problems. Second, the numbers employed on the arithmetic facts tasks were all single-digit, whilst the arithmetical problems task involved multi-digit calculations. The arithmetical problems task, therefore, may simply be more difficult than the arithmetic facts tasks. Given that the children in this study were, on average, developmentally delayed, they may not have yet acquired the cognitive skills necessary to complete these more difficult mathematical problems.

General summary

Overall, group comparisons did not show that the VCFS group were performing poorer than controls on tasks of mathematics. Contrary to predictions, both groups displayed an almost identical profile on the number processing and calculation tasks. This profile was characterised by generally intact number processing, good performance on addition facts but somewhat poorer performance on multiplication facts, and impaired arithmetical problems. Unimpaired number transcoding (reading and writing numbers), intact recall of arithmetic facts and poor calculation procedures in children with VCFS is consistent with previous research. In contrast to previous findings, the present study did not find a weakness in magnitude comparisons. In terms of the McCloskey model, both the number comprehension and number production mechanisms appeared to be generally intact in children with VCFS and controls. As predicted, number facts (addition and multiplication) were performed more accurately than written arithmetical problems (calculation). Intact recall of arithmetic facts in VCFS together with difficulties executing calculation procedures is consistent with previous research [11,13,31] and supports claims that mathematical difficulties are primarily procedural in nature [11,12].

The fact that children with VCFS did not differ from age- and IQ-matched controls suggests that the well-documented mathematical difficulties described in children with VCFS may be explained in the context of their lowered IQ. In fact, children with VCFS may demonstrate certain mathematical strengths compared to their IQ-matched peers, as evidenced by significantly faster response times on average, compared to controls, on addition facts.

Comparison of the McCloskey model in VCFS and turner syndrome

The measures of mathematics administered to children with VCFS in the current study were identical to those previously administered to girls with Turner syndrome [19]. The results obtained by children with VCFS in this study were indirectly compared to those obtained by girls with Turner syndrome [19]. It should be acknowledged that there are significant limitations to this comparison, as the VCFS group in this study (aged 8 years 1 month to 14 years 10 months) had a wider age range than the Turner syndrome group (aged 8 years 11 months to 12 years 1 month). Therefore, results should be interpreted with caution and should serve only as an indirect descriptive comparison of mathematical performance in the two syndromes. Mean FSIQ (as measured by the WISC-R) in the Turner group was within the normal range (M = 90.2; range 80-115), and in children with VCFS was within the borderline range (M = 73.1; SD = 9.8). As emphasised in the current study, IQ is a significant predictor of mathematical performance. Therefore, the higher IQ in the Turner syndrome group may be expected to translate to better performance on mathematical tasks. In terms of mathematical performance, both groups performed with a high degree of accuracy on number processing tasks (group means all > 93% for Turner syndrome and > 95% for VCFS group). Performance on addition and multiplication facts was similar for both Turner and VCFS groups, whereby addition facts were performed with a high degree of accuracy, whilst performances on multiplication facts were somewhat weaker. On the arithmetical problems task, the Turner group had a mean accuracy of approximately 52% [19], which is higher than that found in the VCFS group in the present study, who displayed a mean accuracy of 27%. Given the two groups were quite different in terms of overall intelligence (Turner group approximately 17 IQ points higher than VCFS), it is interesting that children with VCFS performed to the same standard as the Turner group on most tasks. This supports the hypothesis that, despite marked differences in IQ, these two groups are similar in the way that they process numerical information [20], at least in terms of number transcoding, magnitude comparisons and arithmetical facts. Only arithmetical problems were performed more poorly in children with VCFS. This could be because arithmetical problems is a more difficult task developmentally than the other tasks and therefore more cognitively challenging for children with VCFS who have significantly lower IQ scores.

Implications

The results of this study have important educational implications. Overall children with VCFS displayed mathematical skills consistent with their intellectual abilities. Discrepancy between their literacy and mathematical abilities should, perhaps, be interpreted as strength in literacy skills, rather than a weakness in mathematical skills. Overall, the results did not indicate any specific area of mathematical weakness in children with VCFS. Perusal of individual scores for all variables indicated no consistent pattern of performance within the VCFS or control group. Each child displayed a distinct mathematical profile. This emphasises the need for individual assessment of mathematical abilities in children with VCFS, and in children with intellectual impairments in general. Educators should be patient when teaching mathematical concepts to children with VCFS, and should bear in mind that the child's overall cognitive capacity may fall below that of other students.

Limitations

Limitations of the McCloskey model: The McCloskey model offers a useful framework for evaluating different processes of number processing and calculation. However, the McCloskey model has several limitations. First, contrary to typical cognitive neuropsychological models, the model proposed by McCloskey does not enable each module to be tested separately. Rather, tests designed to assess the components of the McCloskey model actually activate several modules. This is a difficulty faced not only when applying the McCloskey model, but also other models of mathematics, including the triple-code model [22].

Second, the framework proposed by McCloskey is essentially a hierarchical model. This means that some modules are more difficult and, therefore, expected to develop at a later stage, than others. In the McCloskey model, basic number processing tasks are much less challenging than tasks of calculation. Researchers examining performances on the McCloskey model should take into account the mental age of a child and, therefore, whether they would be expected to be capable of difficult calculations. Matching a child with VCFS individually to a control child on the basis of chronological age and IQ is a valuable method of accounting for individual differences in mental age. Given the hierarchical nature of the McCloskey model, it would have perhaps been optimal to compare children with VCFS not only to an age- and IQ-matched control group in the current study, but also to an age-matched group of average IQ.

Limitations of study: This study had several limitations. The study employed a control group of children with non-specific intellectual disability. By definition, children with a non-specific intellectual disability are significantly heterogeneous. The fact that close to 40% of the control sample displayed scores which were considered outliers on some tests is a testament to this heterogeneity. Interpretability of the results, therefore, is somewhat limited. It is also possible that a percentage of the control group had undiagnosed genetic conditions. It may have been more appropriate to compare children with VCFS to a control group consisting of children with another genetic condition, such as Turner syndrome, as they may display less heterogeneity than the sample employed in the current study.

Another limitation of the study was that item response times were not recorded with a high degree of precision. Future studies should aim to accurately measure response times in order to examine not only task accuracy, but also patterns of response times within each task. Accurate recording of response times for individual items would be of benefit to conduct more fine-grained analysis on patterns of performance within tasks, particularly for analysis of the distance effect on the magnitude comparisons task. Much of the research investigating magnitude comparisons in children with VCFS has examined the distance effect phenomenon. The distance effect relates to an increased difficulty (evidenced by increased response time) when comparing two numbers that are close together in magnitude (e.g. 5 and 6) as compared to numbers that are far apart in magnitude (e.g. 5 and 9; [35]. A more pronounced distance effect (i.e. exaggerated increase in response time on numbers that are close together in magnitude) in children with VCFS has been reported [13,34]. Unfortunately, the current study was not able to investigate the distance effect because the study did not include an accurate enough measure of response times to individual items [36,37].

A further weakness of the study was that an in-depth error analysis of number processing was not conducted. The McCloskey model breaks number production and number comprehension into several distinct components (Figure 1). While a detailed error analysis was outside the scope of this study, a brief examination of the types of errors made by children with VCFS on basic Arabic number production was conducted. This revealed that the vast majority of errors were of a syntactic nature (such as writing a number in the incorrect order of magnitude, for example: writing 7621 as "700060021"), whilst very few lexical errors were found (which involves substitution of a single incorrect digit or number word in place of a correct one, for example: writing 375 as "378"). This suggests that, where errors in number production exist in children with VCFS, they may be due to difficulties understanding the syntactic properties of numbers. It would be of benefit for future studies to incorporate a thorough error analysis on both number processing and calculation tasks. Categorisation of the types of errors made by children with VCFS may help to further refine our understanding of mathematical difficulties in this population, which is an important step towards designing targeted intervention programmes [38,39].

The age range for this study was limited to children aged 8 to 14 years. While the focus of the current study was to examine basic mathematical processes, it may have been of interest to include older children and adults in order to explore how higher-order mathematical abilities are performed by individuals with VCFS. Higher-order mathematical skills, such as algebra or geometry, may be particularly difficult for people with VCFS given their characteristic deficits in visuospatial and abstract reasoning abilities. Therefore, in an older sample, groups may have differed to a greater degree than what was found in the current study. Further investigation in this area is warranted.

A final limitation from the current study was that instructional environment was not accounted for in this study. The groups may, therefore, not have been well matched in terms of additional exposure to mathematics. This should be considered in future research.

CONCLUSIONS

The results of this study suggest that mathematical difficulties in children with VCFS are predominately consistent with those of children with a non-specific intellectual disability matched to children with VCFS by chronological age and IQ. The contribution of IQ should, therefore, be considered in any future study of mathematics in this population, by incorporating IQ-matched control groups. The study also indicates that cognitive neuropsychological models of mathematics, such as the McCloskey model, are beneficial for determining specific mathematical strengths and weaknesses in children with developmental disabilities such as VCFS.

REFERENCES

1. Tézenas Du Montcel S, Mendizabai H, Aymé S, Lévy A, Philip N. Prevalence of 22q11 microdeletion. J Med Genet. 1996; 33: 719.
2. Driscoll DA, Spinner NB, Budarf ML, McDonald-McGinn DM, Zackai EH, Goldberg RB, et al. Deletions and microdeletions of 22q11.2 in velo-cardio-facial syndrome. Am J Med Genet. 1992; 44: 261-268.
3. Moss EM, Batshaw ML, Solot CB, Gerdes M, McDonald-McGinn DM, Driscoll DA, et al. Psychoeducational profile of the 22q11.2 microdeletion: A complex pattern. J Pediatr. 1999; 134: 193-198.
4. Swillen A, Devriendt K, Legius E, Eyskens B, Dumoulin M, Gewillig M, et al. Intelligence and psychosocial adjustment in velocardiofacial syndrome: a study of 37 children and adolescents with VCFS. J Med Genet. 1997; 34: 453-458.
5. van Amelsvoort TV, Henry J, Morris R, Owen M, Linszen D, Murphy K, et al. Cognitive deficits associated with schizophrenia in velo-cardio-facial syndrome. Schizophr Res. 2004; 70: 223-232.
6. Bish JP, Ferrante SM, McDonald-McGinn D, Zackai E, Simon TJ. Maladaptive conflict monitoring as evidence for executive dysfunction in children with chromosome 22q11.2 Deletion Syndrome. Dev Sci. 2005; 8: 36-43.
7. Bearden CE, Woodin MF, Wang PP, Moss E, McDonald-McGinn D, Zackai E, et al. The neurocognitive phenotype of the 22q11.2 deletion syndrome: Selective deficit in visuo-spatial memory. J Clin Exp Neuropsychol. 2001; 23: 447-464.
8. De Smedt B, Swillen A, Ghesquière P, Devriendt K, Fryns JP. Pre-academic and early academic achievement in children with velocardiofacial syndrome (del22q11.2) of borderline or normal intelligence. Genet Couns. 2003; 14: 15-29.
9. Zinkstok J, van Amelsvoort T. Neuropsychological profile and neuroimaging in patients with 22Q11.2 Deletion Syndrome: a review. Child Neuropsychol. 2005; 11: 21-37.
10. Eliez S, Blasey CM, Menon V, White CD, Schmitt JE, Reiss AL. Functional brain imaging study of mathematical reasoning abilities in velocardiofacial syndrome (del22q11.2). Genetics in Medicine. 2001; 3: 49-55.
11. De Smedt B, Swillen A, Devriendt K, Fryns JP, Verschaffell L, Boets B, et al. Cognitive correlates of mathematical disabilities in children with velo-cardio-facial syndrome. Genet Couns. 2008; 19: 71-94.
12. Wang PP, Woodin MF, Kreps-Falk R, Moss EM. Research on behavioural phenotypes: Velocardiofacial syndrome (deletion 22q11.2). Dev Med Child Neurol. 2000; 42: 422-427.
13. De Smedt B, Swillen A, Devriendt K, Fryns JP, Verschaffel L, and Ghesquiere P. Mathematical disabilities in children with velo-cardio-facial syndrome. Neuropsychologia. 2007; 45: 885-895.
14. De Smedt B, Reynvoet B, Swillen A, Verschaffel L, Boets B, Ghesquiere P. Basic number processing and difficulties in single-digit arithmetic: Evidence from velo-cardio-facial syndrome. Cortex. 2009; 45: 177-188.
15. Deloche G, Willmes K. Cognitive neuropsychological models of adult calculation and number processing: on the role of the surface format of numbers. European Child and Adolescent Psychiatry. 2000; 9: 27-40.
16. McCloskey M. Cognitive mechanisms in numerical processing: evidence from acquired dyscalculia. Cognition. 1992; 44: 107-157.
17. McCloskey M, Caramazza A, Basili A. Cognitive mechanisms in number processing and calculation: evidence from dyscalculia. Brain Cogn. 1985; 4: 171-196.
18. Sokol SM, McCloskey M. Levels of representation in verbal number production. Applied Psycholinguistics. 1988; 9: 267-281.
19. Temple CM and Marriott AJ. Arithmetical ability and disability in Turner's syndrome: A cognitive neuropsychological analysis. Dev Neuropsychol. 1998; 14: 47-67.
20. Simon TJ, Takarae Y, DeBoer T, McDonald-McGinn DM, Zackai EH, Ross JL. Overlapping numerical cognition impairments in children with chromosome 22q11.2 deletion or Turner syndromes. Neuropsychologia. 2008; 46: 82-94.
21. Kates WR, Burnette CP, Jabs EW, Rutberg J, Murphy AM, Grados M, et al. Regional cortical white matter reductions in velocardiofacial syndrome: A volumetric MRI analysis. Biological Psychiatry. 2001; 49: 677-684.
22. Dehaene S, Piazza M, Pinel P, Cohen L. Three parietal circuits for number processing. Cognitive Neuropsychol. 2003; 20: 487-506.
23. Dehaene S and Cohen L. Cerebral pathways for calculation: Double dissociations between Gerstmann s acalculia and subcortical acalculia. Cortex. 1997; 33: 219-250.
24. Floyd RG, Evans JJ, McGrew KS. Relations between measures of Cattell-Horn-Carroll cognitive abilities and mathematics achievement across the school-age years. Psychol Schools. 2003; 40: 155-171.
25. Temple CM. Digit dyslexia: A category-specific disorder in developmental dyscalculia. Cognitive Neuropsychol. 1989; 6: 93-116.
26. Wechsler D. Wechsler Intelligence Scale for Children, Third Edition (WISC-III). San Antonio: The Psychological Corporation. 1991.
27. Wechsler D. Wechsler Intelligence Scale for Children. 4th Edn: Administrative and scoring manual. San Antonio, TX: The Psychological Corporation. 2003.
28. Wechsler D. Wechsler Individual Achievement Test (2nd Edn). San Antonio, TX: The Psychological Corporation. 2002.
29. Holm S. A simple sequentially rejective multiple test procedure. Scandinavian Journal of Statistics. 1979; 6: 65-70.
30. Lang TA, Secic M. How to Report Statistics in Medicine: Annotated Guidelines for Authors, Editors, and Reviewers. American College of Physicians. 2006.
31. De Smedt B, Swillen A, Devriendt K, Fryns JP, Verschaffel L, Ghesquiere P. Mathematical disabilities in young primary school children with velo-cardio-facial syndrome. Genet Couns. 2006; 17: 259-280.
32. Munn P. Symbolic function in pre-schoolers. The development of mathematical skills. Psychology Press. 1998; 43-71.
33. Langdon DW, Warrington EK. The abstraction of numerical relations: A role for the right hemisphere in arithmetic? J Int Neuropsychol Soc.1997; 3: 260-268.
34. Simon TJ, Bearden CE, Mc-Ginn DM, Zackai E. Visuospatial and numerical cognitive deficits in children with chromosome 22q11.2 deletion syndrome. Cortex. 2005; 41: 145-155.
35. Dehaene S, Dupoux E, Mehler J. Is numerical comparison digital?

Analogical and symbolic effects in two-digit number comparison. J Exp Psychol Hum Percept Perform. 1990; 16: 626-641.

36. Zamarian L, Stadelmann E, Nürk HC, Gamboz N, Marksteiner J, Delazer M. Effects of age and mild cognitive impairment on direct and indirect access to arithmetic knowledge. Neuropsychologia. 2007; 45: 1511-1121.

37. McNeil JE, Burgess PW. The selective impairment of arithmetical procedures. Cortex. 2002; 38: 569-587.

38. Shalev RS, Manor O, Auerbach J, Gross-Tsur V. Persistence of developmental dyscalculia: what counts? Results from a 3-year prospective follow-up study. J Pediatr. 1998; 133: 358-362.

39. Temple CM. Cognitive neuropsychology and its application to children. J Child Psychol Psychiatry. 1997; 38: 27-52.

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