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Title: The relationship between visuomotor deficits of children with Neurofibromatosis Type 1 and their ability to function in the areas of writing and attention in school environment

By: Yafit Gilboa

Supervised by: Naomi Josman, Ph.D and Sara Rosenblum, Ph.D

Abstract

Background: Neurofibromatosis type 1 (NF1) is the most common gene disorder with an estimated prevalence of approximately 1 in 3000 -4000. As far as we know Israel is the state with the highest prevalence of the disorder 1:960. NF1 is characterized by neurocutaneous signs, with cafe´-au-lait spots, axillary freckling, cutaneous neurofibromas and iris hamartomas (Lisch nodules) occurring in most patients. NF1 is also a multi-system disorder, with a wide variety of manifestations affecting the eyes, the skeleton, the endocrine system, and the blood vessels, as well as the central and peripheral nervous systems.

Children with NF1 have a unique cognitive profile characterized with slight intelligence decrease. Cognitive problems, attention deficits, and difficulties in visual and linguistic functions are very common among NF1 patients during childhood and clearly represent the most common characteristic. Researchers have demonstrated that up to 81% of children with NF1 are at risk for some type of cognitive deficits. The wide range of cognitive disabilities seen in children with NF1 can be responsible for significant lifetime problems such as academic underachievement, behavioural issues, failure to complete higher education, and limitation of occupation choice. The term "Ecological Validity" refers to an individual's ability to generalize results of controlled experiments to naturally occurring events in the real world. Most previous research in NF1 has been focused on defining the cognitive profile based on formal cognitive testing, with little emphasis on the impact of these deficits on their daily life. Given the broad range of cognitive

deficits that are present in children with NF1, research into functional assessment of cognitive skills and EF remains largely unexplored.

Objectives: The overall goal of this dissertation was to expand our understanding about the functioning at classroom environment of children with NF1, with focus on academic participation, a central area that influence the full participation in school routine. It was taken place using tools and questionnaires with high ecological validity, that objectively measures the functional problem of children with NF1 in their natural environments and were never used with this population.

The specific objectives were:

1. To analyze the process and product of handwriting using digitizing tablet and on line data collection and analysis software (ComPET).
2. To assess the attention function of children with NF1 using a Virtual Classroom that simulate a real classroom.
3. To explore the link between the visual motor integration skills of children with NF1 and their unique handwriting function.
4. To explore the link between parent's reports about attention performance and their function at the Virtual Classroom environment.

Method

Participants: The study group consisted of 30 children diagnosed with NF1, age range 8 years-16 years and 8 months. The control group consisted of 30 typically developing (TD) children matched to the study group by gender and age.

Materials

The handwriting performance of children with NF1 was evaluated with:

1. Coping geometric form to evaluate visual-motor integration skills, using the The Beery–Buktenica Developmental Test of Visual-Motor Integration (VMI)

2. Assessing writing performed through a copying mode using The Hebrew Handwriting Evaluation (HHE).
3. Free style writing task of the Six-Trait Writing Mode.
4. Digitizing tablet and on-line data collection and analysis software, Computerized Penmanship Evaluation Tool (ComPET)

The attention performance and executive functions of children with NF1 was evaluated with:

1. Virtual Classroom (VC)
2. The Conners' Parent Rating Scales— Revised: Long (CPRS-R:L) questionnaire uses to rating symptoms of ADHD

Summary of study contributions

- The results of the children with NF1 in measures of process and product of their handwriting were significantly poorer than for the control group.
- The performance of the NF1 group on the VC was significantly lower than the controls on the number of targets correctly identified (omission errors) and the number of commissions (commission errors).
- For the study group, significant correlations were established between the VMI score and the measures of process and product of the handwriting
- Significant correlations were found between the VC scores and the parent's reports on the questionnaires that asses' attention.
- One discriminate function was found for group classification of all participants. Based on this function, 73.2% of the children correctly classified (72.4% of the study group and 74.1% of the control group). The variable that made the greatest contribution to group membership were (by their Order of importance): The total score of the Six-Trait Writing Model, coping geometric forms using the VMI, The number of targets correctly identified on the VC and the spatial arrangement subtest of the HHE.

Conclusions

- Handwriting product was found to be poorer among children with NF1 in terms of content and spatial arrangement
- The VC appears to be a sensitive and ecologically valid assessment tool for use in the description of attention profile among children with NF1.
- This research adds to the existent literature an evidence to the need of early attention and intervention of teachers and parents of children diagnosed with NF1 in the handwriting and attention difficulties, that could greatly influence their overall academic achievement, social functioning and emotional well-being.