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ABSTRACT

The present study reveals the details of five children with Developmental Gerstmann's syndrome (DGS), a subgroup of Learning disability . There were diversities noticed in terms of their speech and language characteristics. A multidisciplinary team consisting of a Neurologist, a Speech Language Pathologist and a Psychologist at the Institute for Cognitive and Communicative Neuro Sciences (ICCONS) assessed the children.

A dissociation of oral language ability, writing ability and reading ability was noticed among these children. One child showed normal speech and language milestones with normal or better reading ability, but the child had dysgraphia equal in degree to dyscalculia. Three other children presented with a history of developmental dysphasia. Not all children with DGS fail to learn to read at appropriate time. Associated dyslexia was found in 3/5 patients.

Order errors and their script characterized spelling by malorientation of individual letters. All the 5 children described are bilinguals with Malayalam as mother tongue and English as second language. These children made similar errors in both languages. Children classified as DGS are not homogenous and hence will require different remedial strategies. Early identification and intervention of these children is crucial. This is more important in the scenario of Indian culture and education where the students are forced to be biliterate , which further increases the constraints.

Key Words: Gerstmann's syndrome, finger anomia, right-left disorientation, acalculia, agraphia, and apraxia.

INTRODUCTION

Dyscalculia, dysgraphia, right-left confusion and finger agnosia in children with out clear evidence of neurological impairment has been called Developmental Gerstmann's Syndrome (DGS) (Spillane 1942, Kinsbourne and Warrington (1963), Benson and Geshwind (1970) Kinsbourne and Warrington (1963) speculated that the underlying defect in DGS is one of inability to correctly serial order parts of a whole.

These cases were fascinating because of a distinct dissociation of oral communication abilities from written language ability, the association of finger agnosia to acalculia and right left disorientation to agraphia. These cases were selected from the Institute of

Communicative and Cognitive Neurosciences where they were assessed by a multidisciplinary team consisting of a Neurologist, a Speech Language Pathologist and a Clinical Psychologist. All the cases showed a normal intelligence on Wechsler Intelligence Scale for Children (WISC). Right-left discrimination was tested using right-left orientation test (Spreenn and Gaddes 1969), finger agnosia was tested using finger localisation test (Benton 1959). Malayalam Language Test (Rukmini 1994) and Malayalam Articulation Test (Maya 1990) were administered to evaluate the speech and language skills of these children. Age appropriate curriculum based tasks were given to test writing ability, reading ability and mathematical skills. The simplest test for writing was the spontaneous production of letters of the alphabets. Dictated words were used for intermediate testing. In the more advanced, a paragraph on the festival celebrated by them or their hobby was given.

CASE 1 was an eleven year old right handed boy with Malayalam as mother tongue (Malayalam is a dravidian language spoken in the state of Kerala) , born of a nonconsanguinous parentage who discontinued his education in general school system at the age of 6years .Mild delay was reported in the speech and language milestones. General physical examination and systemic evaluations were unremarkable. Neurological evaluations revealed a pleasant, young boy. He had right –left disorientation, finger anomia and agnosia. Other abnormalities noticed include graphesthesia, asteriognosis and two-point indiscrimination. He could not do simple calculations or even count spontaneously. Writing was grossly abnormal and was limited to few alphabets of Malayalam and English and could hardly copy anything. He could neither copy nor spontaneously draw simple geometric figures or line drawings. He couldn't do even simple arithmetic calculation. However his reading ability was good. On WISC the child scored a verbal IQ of 87.3 and a performance IQ of 86.8.

CASE 2 was a six-year-old girl born of nonconsanguinous parentage with the history of severe difficulty in writing. Her motor and language milestones were mildly delayed. Her mother tongue was Malayalam. There was no family history of learning disability or left-handedness.

Physical examination and systemic evaluations were within normal limits. Neurological evaluation revealed asteriognosis, graphesthesia and inability for two-point discrimination. She had right to left disorientation, finger anomia, finger agnosia, acalculia and constructional apraxia. Her expressive language level and receptive language level were age appropriate. However misarticulations of laterals and trills were noticed .The child could read alphabets when presented independently but showed confusion when presented in a group. Writing spontaneously as well as copy writing was grossly affected. She was not able to do simple calculation or copy simple geometric figures. She could count up to 50 with constant cueing. Psychometric evaluation was done using Wechsler Intelligence Scale for Children (WISC). On WISC her verbal IQ and performance IQ were found to be 88.6 and 72.6 respectively.

CASE 3 was a twelve-year old boy studying in the 7^{th}, who was born of a nonconsanguinous parentage. The prenatal, natal and postnatal history were not

significant .The developmental milestones were normal.. He was sent to Kindergarten at the age of 3.5 years. He showed difficulty in writing compared to his peer group. His grandfather, father and sister are reported to be learning disabled. General physical examination and systemic examinations were within normal limits. Neurological examination was unremarkable except for impaired graphesthesia, asteriognosis and two-point discrimination. He had right –left disorientation, finger agnosia, finger anomia, acalculia and constructional apraxia. Detailed speech and language evaluation revealed his auditory verbal comprehension and oral expression to be adequate. He had severe dysgraphia. He mixed up upper case (Capital letters) with lower case (small letters). He made substitution errors when dictation of alphabets of Malayalam and English were given.

Though this subject could write in paragraphs, his writing was characterized by syntactic errors and spelling errors in terms of reversals (e.g. " b" for " d"), omissions (e.g. solder for soldier), sequencing errors (e.g. knoweldge for knowledge), phonological spelling (e.g. hystory for history). This bilingual child whose mother tongue is Malayalam showed similar errors in Malayalam also. Eventhough Malayalam is a phonological language; certain words borrowed from Sanskrit are irregular where there is no phoneme- grapheme correspondence (e.g. channanam-written as chandanam). Phonological errors could be noticed for such words. His reading showed sequential errors (saw for was), confusion between " i " and " l" (e.g. tied as teld), addition of phonemes (e.g. "squench" for "quench"), deletions ("tied" for "tried"). He could not do mental calculation or simple arithmetic calculations. On Wechsler Intelligence Scale for Children (WISC) her verbal IQ and performance IQ were found to be 81.1 and 78.6 respectively.

CASE 4 was a twelve-year-old boy with a history of poor scholastic performance. He was born of a nonconsanguinous parentage at full term. The developmental motor and language milestones were normal. The major problems observed were difficulty in writing, reading, and doing calculations. General physical examination was unremarkable except that he had right-left disorientation and finger anomia, agraphia and acalculia. Detailed speech and language analysis revealed his expressive and comprehensive levels to be adequate for the age .His mother tongue was Malayalam. Writing was characterized by orthographic errors, spelling errors due to substitution and omission, phonological spelling etc. He had confusion between uppercase and lower case. He could not write sentences. His reading showed reversals "bib"for "did", sequential errors "guhe" for "huge". He couldn't draw simple figures. He could not do mental calculation or simple subtractions. On WISC the child scored a verbal IQ of 85.8 and performance IQ of 76.8.

CASE 5 was a 15-year old boy studying in eighth standard. He was born of a nonconsanguinous parentage, one-month post mature. The labour was induced and was forceps assisted. There was no history of birth asphyxia or neonatal infection. All the speech and motor milestones were normal .His mother tongue was Malayalam. He experienced severe difficulty in writing and doing mathematical calculation. His reading comprehension was good. He could not handle money or calculate the balance in transactions. He could tell in terms of coin value rather than the total amount (e.g. 2 fifty paise coin, 3 five-rupee notes etc). He had severe finger agnosia and right left

disorientation, he could not even identify and select the shoes for right leg or left one. General physical examination and systemic evaluations were unremarkable. Neurological evaluation revealed graphesthesia, asteriognosis, right left disorientation and finger agnosia. He had severe dysgraphia and acalculia. He had difficulty in drawing simple geometric figures. His writing was characterized by spelling errors due to sequencing errors, phonological spelling, disorthographia. He mixes up English alphabets and Malayalam alphabets within a word. On WISC the child scored a verbal IQ of 86.5 and performance IQ of 83.2.

DISCUSSION

The five cases described above with the clinical features of Gerstmann's syndrome viz. right-left disorientation, finger anomia, dyscalculia, and severe dysgraphia, showed diversity in terms of the symptoms exhibited by them .Mild delay in the language development was observed in 2 of the cases during the early stages of development which was overcome at a later stage, who also showed severe agraphia .They were not able to copy the alphabets even. Abnormalities in writing were exhibited by the other children also. The errors in writing observed in these cases can be broadly classified into orthographic errors and lexical errors. Orthographic errors include improper margin setting, inability to write in a line, abnormalities in the script such as crude and misshapen letters, abnormal duplication and deletion of cursives, improper orientation of letters, abnormal duplication and deletion of cursives, improper orientation of letters in words, and words in sentences. The lexical errors include errors in spelling, phonological and lexical agraphia, sequencing errors of letters or total inability to write. Critchley (1966) suggested that dysgraphia in Gerstmann syndrome to be an apraxia of writing. Associated dyslexia was seen in three of the children. Hence a dissociation of oral language ability, writing ability and reading ability was noticed among these children.

One of the fascinating features of Gerstmann syndrome is the association of finger agnosia with dyscalculia. This combination recalls the fact that the child learns to count and calculate with the help of his finger. (Critchley 1966,Ross 1991). The use of the word "digit" from the Latin word "digitum", meaning both numeral and finger reflects this association. (Ross 1991). Just as finger agnosia is an inability to recognize the relationship of the fingers to each other in space and to manipulate and arrange them to order, so manipulation of those other digits, the numerals is equally disturbed. This suggests that there is functional and linguistic interdependence and interaction for finger identification, use and calculation.

CONCLUSION

Five children with Developmental Gerstmann syndrome who showed diversities among the group are described. Children classified as DGS are not homogenous and hence will require different remedial strategies.

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