DEVELOPMENTAL GERSTMANN’S SYNDROME
A CASE REPORT & LITERATURE REVIEW

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Abstract
Developmental Gerstmann’s syndrome is a neurological disorder in children characterized by a tetrad of symptoms including dysgraphia, dyscalculia, left-right confusion, and finger agnosia. The incidence and exact cause is unknown. Controversy surrounds the existence, diagnosis and treatment of this condition. The case presented here represents a patient with developmental Gerstmann’s syndrome that was subsequently diagnosed with visual efficiency difficulties and visual perceptual deficiencies. A literature review is included.

Key Words
accommodative insufficiency, directionality, dyscalculia, dysgraphia, finger agnosia, Gerstmann’s syndrome, laterality, perceptual deficiency

INTRODUCTION

Gerstmann’s syndrome (GS) is a neurological disorder that was first described in the 1930’s, and has been subsequently characterized by four signs: dysgraphia, dyscalculia, left-right confusion, and finger agnosia. Controversy exists as to whether GS represents a true syndrome or a collection of symptoms that often occur together. There is a relative absence of reports of this condition in the literature.

It is reported to occur in both adults and children. In adults, the condition is typically acquired (AGS) and may occur after a stroke, in association with damage to the cortex, or in Alzheimer’s disease. The condition is termed developmental Gerstmann’s syndrome (DGS) when these symptoms coexist in a child. While definite causes have been identified in adults, the same cannot be said concerning children. Language deficits that present in AGS are not found in children.

Conversely, reading impairment that occurs frequently in DGS is not observed in AGS. In adults, the condition is typically acquired (AGS) and may occur after a stroke, in association with damage to the cortex, or in Alzheimer’s disease. The condition is termed developmental Gerstmann’s syndrome (DGS) when these symptoms coexist in a child.

While definite causes have been identified in adults, the same cannot be said concerning children. Language deficits that present in AGS are not found in children. Conversely, reading impairment that occurs frequently in DGS is not observed in AGS. In fact, controversy surrounds the very diagnosis of DGS. There is little consensus as to how to make a proper diagnosis as well as the specific tests required. The amount of specific deficits needed to make a diagnosis is also controversial.

Another compounding factor is that several of the tests do not have norms. It is unclear whether the tetrad of findings associated with DGS (Table 1) requires that all four associated symptoms be present for a diagnosis to occur, or if partial expression of the findings is possible.

Signs of DGS
Finger agnosia (aka finger aphasia), one of the four conditions found in DGS, was characterized by Gerstmann. His description was “a loss of the ability to recognize, identify, differentiate, name, select, indicate and orient as to the individual fingers of either hand, the patient’s own, as well as those of another person.” The developmental age when finger naming occurs remains controversial.

Dysgraphia, a writing disability, is characterized by errors in spelling and penmanship. Dysgraphia in DGS is attributed to constructional apraxia, a fifth proposed characteristic of the syndrome. Constructional apraxia is an inability to reproduce simple drawings of shapes, and impairment in tasks that involve constructing patterns using colored blocks. Poor performance in handwriting and constructional tasks is interpreted as representing difficulties in understanding spatial concepts.

Acalculia or dyscalculia is a lack of understanding of the rules for calculation or arithmetic. Deficiencies have been noted with DGS in both verbal and written math calculations. According to one study, children with DGS understand the concepts that form the basis for the math calculations but have poor ability to write and sequence numbers. A contrasting opinion puts forth that the math errors are “due to a Piagetian developmental delay as the child transitions from the preoperational to the concrete operational stage of development.”

The fourth characteristic of DGS is the inability to distinguish right from left on one’s self as well as on another person. This is known as left-right confusion or
lobe damage to either side is implicated,20 has proven difficult, as the multiple skills
in DGS. The search for a specific area brain responsible for the symptoms seen
searchers have not located an area in the
tle manifestation of cerebral palsy. Re-
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tle manifestation of cerebral palsy. Re-
searchers have not located an area in the
brain responsible for the symptoms seen
in DGS. The search for a specific area
has proven difficult, as the multiple skills
deficits shown in DGS are controlled by,
and therefore affected by, multiple brain
loci.3 The incidence of DGS is likewise un-
known. A literature search on PubMed re-
vals a paucity of case reports and no epi-
demiologic studies. It is unknown if there
is a gender or race predilection. Familial
occurrence has also not been documented
with DGS.15 Many consider DGS to be
rare, others feel that it may be more com-
mon than reported.4 Lack of awareness
of the condition, unreliable or unperformed
testing procedures, the overshadowing by
other conditions, as well as behaviors be-
ing attributed to other conditions, are all
cited as potential reasons for a low rate of
diagnoses.4 The diagnosis of DGS itself is compro-
mised. The tests used to diagnose the con-
dition are not standardized, nor are norms
by age and gender available. For example,
in making a diagnosis of finger agnosia,
four types of tasks are assessed.
1) Identification tasks can be evaluated by
showing the examiner which finger
was touched while blindfolded and
then naming the finger.22,24
2) Differentiation tasks that require the
child to identify whether the examiner
touched one or two of their fingers3
3) “In-between tasks” require the child to
identify the number of fingers between
two that were touched.
4) Finger block tasks use blocks that
are shaped so that the fingers form a
specific pattern. The child’s hand is
closed around a block that elevates
some of the fingers. The block is re-
moved, and the eyes that were previ-
ously closed are opened. The child is
asked to identify which block out of a
group of blocks they were holding.3

Of the studies published, the test protocol
was not standardized. Some clinicians re-
ported one type of task used in the di-
agnostic battery, while others use a com-
bination.3 While those diagnosed with DGS exhibit
the diagnostic criteria, there are associ-
ated features that have been documented in
the literature. Average or better than
average intelligence, as well as elevated
verbal intelligence, have been noted in
some reports.3 As much as a 20-point
split has been documented when compar-
ning verbal to performance scores on the
Wechsler Intelligence Scale for Children
(WISC).13 This is the primary measure
of intelligence used in the United States.
Also, most studies report a negative fam-
ily history for learning disabilities or inap-
propriate language development.3
Behavioral problems have been found in
greater numbers in those diagnosed with
DGS. These include attention-deficit-hy-
peractivity disorders and emotional dis-
turbances such as temper tantrums, ag-
gressiveness, spitting and biting.2 Several “soft” neurologic signs have also
been reported in children with DGS. Gait
abnormalities include unusual and uncon-
trollable problems with walking due to
conditions such as vestibular and central
nervous system disorders.29 They may
also show: increased muscle tone, hyper-
active deep tendon reflexes (abnormally
brisk muscle contraction occurring with a
sudden sharp tap to the muscle’s tendon
of insertion),26 extensor plantar responses
(abnormal reflex of dorso-flexion of the
great toe and abduction of the other toes
when the plantar surface of the foot is
stimulated),27 and tremors (rhythmic, in-
voluntary, oscillatory movement of body
parts).28

Similar Conditions
There are several well known syndromes
that have been documented as associated
with DGS. In one study 65% of individu-
als with Fragile X syndrome demonstrat-
ed three or more of the diagnostic charac-
teristics of DGS.22 Williams syndrome, a
rare genetic condition (~ 1/7,500 births),
has both similar and contrasting symp-
toms of DGS. Affected individuals with
William’s syndrome exhibit visuo-spatial
construction problems with elevated ver-
bal intelligence.24 They have symptoms of
inattention and below average perfor-
ance intelligence, but display strong au-
ditory short term memory.29

Table 1. Findings of Developmental Gerstmann’s Syndrome

<table>
<thead>
<tr>
<th>Diagnostic Findings</th>
<th>Associated Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Finger agnosia</td>
<td>Average or better than average intelligence</td>
</tr>
<tr>
<td>Left-right confusion</td>
<td>Elevated verbal intelligence</td>
</tr>
<tr>
<td>Dysgraphia</td>
<td>Negative family history for learning disabilities</td>
</tr>
<tr>
<td>Dyscalculia</td>
<td>Appropriate language development</td>
</tr>
<tr>
<td>Constructional apraxia*</td>
<td>Behavioral problems</td>
</tr>
<tr>
<td></td>
<td>Gait abnormalities</td>
</tr>
<tr>
<td></td>
<td>Increased muscle tone</td>
</tr>
<tr>
<td></td>
<td>Hyperactive tendon reflexes</td>
</tr>
<tr>
<td></td>
<td>Extensor plantar responses</td>
</tr>
<tr>
<td></td>
<td>Tremors</td>
</tr>
</tbody>
</table>

*Hypothesized diagnostic finding

Proposed Neurology

The exact cause of DGS is unknown but
there are several models and hypotheses.
Specific brain areas that have been impli-
cated are the parietal lobe,13 and a sub-
angular lesion. The latter lesion is a focal
ischemic lesion, situated sub-cortically in
the inferior part of the left angular gyrus
and reaching to the superior posterior re-
gion.7,8 While it is theorized that parietal
lobe damage to either side is implicated,20
the left-hemisphere angular gyrus22 and
the right supra-marginal gyrus21 may be
responsible. Although here are no specific
neurological abnormalities documented
in the literature,7 a recent study did find
abnormal EEG recording on four out of
ten patients. Nonspecific findings were
documented, such as hyper-intense le-
sions in the white matter and empty sella
syndrome. This syndrome is a radiologi-
cal finding where spinal fluid is found
within the space created for the pituitary23
It is yet unknown if these abnormalities
are related to DGS.2

Abnormal developmental processes caus-
ing neuro-cognitive skill abnormalities
have been considered as potential causa-
tion of DGS. Kinsbourne17 suggested that
developmental delays were the cause of
DGS and that an underdeveloped “neural
facility” or perinatal trauma caused a sub-
tle manifestation of cerebral palsy. Re-
searchers have not located an area in the
brain responsible for the symptoms seen
in DGS. The search for a specific area
has proven difficult, as the multiple skills

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Patients with nonverbal learning disabilities (visuo-spatial learning deficits, math calculation deficits and deficits with tactile perception) and Asperger’s syndrome also have both similar and dissimilar symptoms to DGS. Unlike DGS however, these two conditions also present with issues relating to nonverbal problem solving, pragmatic speech, and poor spatial judgment. Those with Asperger’s syndrome characteristically show evidence of poor visuo-spatial skills, normal intellectual functioning, elevated verbal intelligence, math deficits, and right hemisphere deficits. In contrast to DGS, they display poor social interactions, circumscribed interests and poor nonverbal communication.

Since the number of reported cases of DGS has been minimal, determination as to the natural history is not well known. Both full and partial resolution has been documented in the literature.4 Even cases of spontaneous resolution have been reported, but some evidence of a learning disability remained.13 Given that adult and teenage cases have been detailed, it is evident that some cases persist and are resistant to remediation.16

Medical and educational treatment is typically symptomatic and supportive. Reading tutoring for dyslexia or graphomotor training for dysgraphia has shown to be mostly unsuccessful. Treatment traditionally includes occupation and speech therapy.5 Compensatory learning techniques such as calculators and computer word processors, bypassing the deficit when possible, may prove to be more effective in the long run. Most importantly, the patient, parents and teachers must be educated on the condition and the possible secondary limitations. This is believed to help alleviate and prevent exacerbation of already existing emotional difficulties.4

CASE REPORT

History

A nine-year-old male presented with complaints of difficulty in school. Currently in fourth grade, he has been receiving speech therapy and attending extended resource special education classes since kindergarten. The patient’s mother indicated that he has difficulty completing assignments particularly in reading, writing, arithmetic and spelling. The patient was delayed in motor and language milestones. The first words were spoken at 2 years of age and two to three word sentences at age 4. The patient has not learned to tie his shoe, nor ride a bicycle or tricycle. Toilet training was also delayed, occurring at 3½ years of age. It was reported that the patient has difficulty relating to others his own age as well as being immature and clumsy. While the pregnancy was uncomplicated, labor was induced due to toxemia. No postnatal complications were reported and the child did not suffer from unusual childhood illnesses. His medical history was positive for multiple ear infections. The patient did not use medications. An uncle was reported to have a seizure disorder and hearing problems, but the patient’s direct family medical history was free of significant medical or emotional problems.

Two years prior, the patient had been diagnosed with Attention Deficit Hyperactivity Disorder (ADHD), Gerstmann’s syndrome and upper body weakness by a neuro-psychologist. That evaluation revealed low average cognitive processing abilities and below average achievement skills. His gross visual-motor integration ranged from severely impaired to average. He exhibited low to mildly impaired receptive vocabulary abilities. Impairments in visual memory, executive functioning and symbolic transfer (transformation of non-verbal signs into verbal representations) were also noted. The Wechsler Abbreviated Scale of Intelligence (WASI) indicated a full scale IQ of 87. Treatments outlined included medical therapy for ADHD, as well as academic strategies and modifications to cope with the deficits.

Optometric Findings

A COVD-Quality of Life-Short Form (COVD-QOL)31-35 was completed producing a score of 41. A score of greater than 20 is considered indicative of a binocular vision dysfunction. The data (Table 2) was obtained at the initial examination. Looking closely at the accommodative data (low PRA, high NRA, low accommodative amplitudes, lag on MEM) a pattern of accommodative insufficiency is revealed. While the reduced Near Point of Convergence (NPC) can indicate convergence insufficiency, retesting with a low plus lens negated this finding. The internal and external structures of the eye were within normal limits. The patient was diagnosed with an accommodative insufficiency and pseudo-convergence insufficiency. The patient was issued a near prescription of +0.50D for use in school and at home. Due to the high score of the COVD-QOL questionnaire, as well as the patient’s history of school difficulties, a perceptual examination was performed following several weeks of wearing the near prescription. A perceptual examination (Table 3) was performed at a later date which indicated a deficiency in laterality and directionality. Dysgraphia was not diagnosed due to at least average performance on two

<table>
<thead>
<tr>
<th>Table 2. Optometric examination data</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Unaided Visual Acuity</strong></td>
</tr>
<tr>
<td>Distance</td>
</tr>
<tr>
<td>Near</td>
</tr>
<tr>
<td><strong>Cover Test-Distance</strong></td>
</tr>
<tr>
<td>Near</td>
</tr>
<tr>
<td>8 exophoria</td>
</tr>
<tr>
<td><strong>Stereo</strong></td>
</tr>
<tr>
<td><strong>Distance Retinoscopy</strong></td>
</tr>
<tr>
<td>OD -0.25-0.25 X 090</td>
</tr>
<tr>
<td>OS Plano</td>
</tr>
<tr>
<td><strong>Manifest Refraction</strong></td>
</tr>
<tr>
<td>OD PL-0.25 X 090</td>
</tr>
<tr>
<td>OS Plano</td>
</tr>
<tr>
<td><strong>Phoria-Distance</strong></td>
</tr>
<tr>
<td>Near</td>
</tr>
<tr>
<td>2 esophoria</td>
</tr>
<tr>
<td><strong>NRA/PRA</strong></td>
</tr>
<tr>
<td><strong>Near Vergences</strong></td>
</tr>
<tr>
<td>BO 12/24/12</td>
</tr>
<tr>
<td><strong>MEM</strong></td>
</tr>
<tr>
<td>OD +1.00</td>
</tr>
<tr>
<td><strong>Accommodative Amplitude</strong></td>
</tr>
<tr>
<td>OD 9.00D</td>
</tr>
<tr>
<td><strong>Accommodative Facility</strong></td>
</tr>
<tr>
<td>OD 10cpm OS 9cpm OU 7cpm (+) harder OD, OS; (-) harder OU</td>
</tr>
<tr>
<td><strong>NPC</strong></td>
</tr>
<tr>
<td>1/13/17/21</td>
</tr>
<tr>
<td>1/17/20</td>
</tr>
<tr>
<td><strong>NPC with probe lens (+0.50D)</strong></td>
</tr>
<tr>
<td>1/5/7 2/5/10</td>
</tr>
</tbody>
</table>

BO=Base out MEM= monocular estimate method
BI= Base in NRA= negative relative accommodation
pcm= cycles per minute PRA= positive relative accommodation

BI= Base in NRA= negative relative accommodation
pcm= cycles per minute PRA= positive relative accommodation

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The patient was entered into a vision therapy program to address both visual skills (accommodative insufficiency) and perceptual deficiencies. It was estimated that 15-20 visits would be needed to remediate the accommodative aspects while 30-40 visits would be needed for laterality and directionality difficulties. Unfortunately, the patient’s mother was unable to continue his scheduled therapy after completing only five visits over a period of 15 weeks. The patient was scheduled to be re-evaluated during his yearly scheduled vision examination.

**Table 3. Perceptual examination data**

<table>
<thead>
<tr>
<th>TEST</th>
<th>CLASSIFICATION</th>
<th>PERCENTILE/AGE EQUIVALENT</th>
<th>Rank</th>
</tr>
</thead>
<tbody>
<tr>
<td>Piaget Right/Left Awareness test</td>
<td>Laterality and Directionality</td>
<td>Age 6 years</td>
<td>Very Weak</td>
</tr>
<tr>
<td>Standing Balance</td>
<td>Assesses body schema and motor planning</td>
<td>&lt; Age 4.0-4.5</td>
<td>Very Weak</td>
</tr>
<tr>
<td>Six Figure Split Form Board</td>
<td>Assesses simultaneous processing and eye-hand coordination</td>
<td>80 %</td>
<td>Strong</td>
</tr>
<tr>
<td>Gardner Reversal Frequency Test --Execution</td>
<td>Requires child to write numbers &amp; letters as called out in random order</td>
<td>1%</td>
<td>Very Weak</td>
</tr>
<tr>
<td>Gardner Reversal Frequency Test --Recognition</td>
<td>Requires child to recognize letters &amp; numbers written backwards/reversed</td>
<td>&lt;1%</td>
<td>Very Weak</td>
</tr>
<tr>
<td>Gardner Reversal Frequency Test --Matching</td>
<td>Evaluates visual attention and discrimination</td>
<td>&lt;17%</td>
<td>Weak</td>
</tr>
<tr>
<td>Developmental Eye Movement Test (DEM): Horizontal</td>
<td>Developmental Eye Movement test: Saccadic eye tracking used for reading</td>
<td>57%</td>
<td>Average</td>
</tr>
<tr>
<td>(DEM) Vertical</td>
<td></td>
<td>62%</td>
<td>Average</td>
</tr>
<tr>
<td>(DEM) Ratio</td>
<td></td>
<td>46%</td>
<td>Average</td>
</tr>
<tr>
<td>(DEM) Errors</td>
<td></td>
<td>76%</td>
<td>Strong</td>
</tr>
<tr>
<td>The Beery-Buktenica Developmental Test of Visual-Motor Integration (VMI)</td>
<td>Visual Motor Integration -simultaneous processing - eye-hand coordination</td>
<td>42%</td>
<td>Average</td>
</tr>
<tr>
<td>VMI-Motor Coordination</td>
<td></td>
<td>39%</td>
<td>Average</td>
</tr>
<tr>
<td>Wold Sentence Copy Test</td>
<td>Requires child to copy a paragraph - Eye hand coordination - Fine Motor Skills - Eye tracking</td>
<td>&gt; 8th grade</td>
<td>Very Strong</td>
</tr>
<tr>
<td>Dyslexia Determination Test (DDT)</td>
<td>Dyslexia Screening Instrument</td>
<td>Above Normal</td>
<td>Very Strong</td>
</tr>
<tr>
<td>Dyslexia Determination Test (DDT)</td>
<td></td>
<td>Above Normal</td>
<td>Very Strong</td>
</tr>
</tbody>
</table>

**DISCUSSION**

This case represents a child diagnosed with a neurological disorder that closely resembles a visual efficiency (accommodative dysfunction) and perceptual dysfunction. Looking closer at our perceptual testing results in relation to the diagnostic criteria for DGS, there are both similarities and dissimilarities. Based on tests such as the Piaget Right/Left Awareness Test and Gardner Reversal Frequency Test, the patient was diagnosed with deficiencies in laterality and directionality and poor coding of words with left-right confusion. These signs are basic tenets of DGS. In contrast, testing in the area of visual motor integration, a component of dysgraphia, was not deficient. Our patient did not show any trouble in any of the copying activities. The Wold Sentence Copy showed proper letter formation, adequate spacing and no letter reversals. Visual motor integration and motor coordination testing both showed results in the average range, negating another diagnostic criterion, constructional apraxia. Finger agnosia and dyscalculia are not areas traditionally tested via optometric perceptual examination so no comparison can be made in these areas.

Why did our testing show a difference in two out of three areas diagnostic for DGS? One explanation is that the testing performed previously under the supervision of the neuro-psychologist occurred several years prior to the perceptual testing. Cases of partial resolution have been documented in the literature, perhaps this case fits into that category. Another possibility is that perhaps the patient simply had perceptual deficiencies and visual efficiency issues at the time of the initial diagnosis. While it is impossible to travel back in time to suggest a referral to an optometrist who specializes in learning-related vision problems; the lack of vision care in this child and many others in documented cases is disappointing. Many of the diagnostic signs are, in fact, related either directly or
indirectly to the visual system. It is possible that children diagnosed with DGS are actually children with significant visual skills and visual information processing disorders. Certainly, the care of a behavioral optometrist should be considered in any diagnosis of DGS.

CONCLUSION
This case report details a patient diagnosed with developmental Gerstmann’s syndrome. The main difference in this case versus others documented in the literature is that this report is the first to demonstrate a visual efficiency problem in a patient diagnosed with DGS. Even though many of the symptoms of DGS are vision related, the need for an examination of the visual system has not been documented in the literature and therefore may not be recommended or encouraged. The question remains as to the prevalence of visual efficiency problems in those with DGS and whether these patients may simply have undiagnosed and untreated visual problems. Regardless of the cause of the visual problem, the behavioral vision care profession can potentially make a huge contribution to the emotional, social and academic well being of these children. Behavioral optometry must be at the forefront of the treatment of these children.

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Date accepted for publication:
August 6, 2008

Journal of Behavioral Optometry Volume 19/2008/Number 4/Page 95