SHORT REPORT

Right hemisphere learning disability associated with left hemisphere dysfunction: anomalous dominance and development

Thomas A Sandson, Dara S Manoach, Bruce H Price, Dorene Rentz, Sandra Weintraub

Abstract

Two patients are described with the social emotional processing disorder, a developmental syndrome usually ascribed to right hemisphere dysfunction. In these two patients however, neurological examinations, EEG, and neuroimaging studies were all consistent with left hemisphere dysfunction. Both patients were left handed and had findings suggestive of anomalous dominance for language. It is proposed that early left hemisphere injury may have resulted in functional reorganisation that allowed sparing of language and motor skills but interfered with the development of functions that the right hemisphere normally subserves.

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Many terms have been used to describe a developmental syndrome characterised by emotional and interpersonal difficulties, deficient paralinguistic communication, and impaired visuospatial and mathematical abilities relative to verbal skills.¹⁻³ We have referred to this syndrome as the social emotional processing disorder (SEPD).⁴ The SEPD has been associated with right hemisphere dysfunction based on the neuropsychological profile, neurological findings, and neurophysiological and neuroimaging studies.¹⁻⁴ We now describe two left handed patients with the emotional, interpersonal, and cognitive features of SEPD and neuro-anatomical evidence of early left hemisphere injury.

Case reports

CASE 1

A 30 year old woman had developed high fevers and generalised seizures at 8 months of age. A right hemiparesis was first noted at that time. At the age of 10, she began to have recurrent complex partial seizures that were refractory to anticonvulsant medications.

She described herself as a shy, withdrawn, and socially isolated child with few friends. She was an average student but needed tutoring in mathematics. Depression has been

diagnosed since her early teens and has been unresponsive to psychotherapy and antidepressant medications.

On examination, she was shy, guarded, and easily embarrassed. She made little eye contact and speech prosody was flattened. Neuropsychological testing (table) showed a 19 point discrepancy between verbal IQ (40th percentile) and performance IQ (8th percentile) as measured by the WAIS-R. Attention was mildly impaired on tests of response inhibition and set maintenance such as the auditory Go-No go test and the Stroop colour interference test. Memory testing with the WMS-R showed a selective impairment of non-verbal memory. Language based functions were normal with the exception of below average performance on the Boston naming test. Many of the errors on the Boston naming test resulted from inaccurate perception of the stimulus picture. Visuospatial perceptual and constructional abilities were impaired. Tests of verbal reasoning and abstraction were performed well. She scored within the normal range on the Beck depression inventory.⁷

Neurological examination showed the ear and thumb to be smaller on the right than on the left. There was mild right upper motor neuron facial weakness. Rapid alternating movements and fine finger movements were slow and clumsy on the right. Deep tendon reflexes were increased on the right and stressed gait produced dystonic posturing of the right hand.

EEGs and EEG telemetry showed a left temporal seizure focus; MRI showed atrophy of the left temporal lobe and dilatation of the left temporal horn (figure).

On intracarotid amobarbitone testing, language and memory were entirely normal after the left internal carotid injection. There was no neglect on a random letter cancellation test. Right internal carotid injection resulted in unresponsiveness for the first 12 minutes, after which the EEG had returned to normal. When she awoke, she had mild left sided neglect on a random letter cancellation test.

A left temporal lobectomy was performed without complication. Pathology showed loss of large pyramidal neurons in the hippocampus and dentate gyrus with fibrillar astrocytosis. There have been no seizures since the surgery but emotional, interpersonal, and

**Behavioural
Neurology Unit, Beth
Israel Hospital and
Harvard Medical
School, Boston, MA,
USA**

T A Sandson
D S Manoach
B H Price
D Rentz
S Weintraub

Correspondence to:
Dr Sandson, Department of
Neurology, Beth Israel
Hospital, 330 Brookline
Ave, Boston, MA 02215,
USA.

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Neuropsychological data*

	Case 1	Case 2		
WAIS-R	Verbal IQ 98 Information 10 Digit span 10 Vocabulary 10 Arithmetic 11 Comprehension 6 Similarities 12	Performance IQ 77 Picture completion 6 Picture arrangement 5 Block design 6 Object assembly 8 Digit symbol 7	Verbal IQ 108 Information 13 Digit span 11 Vocabulary 13 Arithmetic 11 Comprehension 11 Similarities 11	Performance IQ 80 Picture completion 8 Picture arrangement 11 Block design 5 Object assembly 4 Digit symbol 7
Shipley-Hartford	Vocabulary 27 Abstraction 38 Conceptual quotient 123 Estimated IQ 110	Vocabulary 30 Abstraction 32 Conceptual quotient 93 Estimated IQ 111		
Wide range achievement test: spelling	Standard score = 110 75 th percentile grade equivalent >12	Standard score = 111 77 th percentile grade equivalent 10.2		
Wide range achievement test: reading	Standard score = 106 66 th percentile grade equivalent >12	Standard score = 114 82 nd percentile grade equivalent 10.6		
Wide range achievement test: arithmetic	Standard score = 101 53 rd percentile grade equivalent 12B	Standard score = 92 30 th percentile grade equivalent 5.9		
Controlled oral word association test F-A-S	32 (29 th percentile)	42 (69 th percentile)		
Stroop colour interference test (50 items)	72 seconds 3 errors of commission	89 seconds 7 errors of commission		
Boston naming test (total = 60)	45	56		
Wechsler memory scale	WMS-R: Logical memory I 25/50 (47%) Logical memory II 20/50 (41%) Visual reproduction I 35/41 (85%) Visual reproduction II 8/41 (19%)	WMS: Logical memory 18/23 10 minute delay 17/23 Visual reproduction 10/14 10 minute delay 10/14		
Rey-Osterrieth complex figure (total score = 36)	Copy: 36 (99%) Immediate reproduction: 23 (55%)	Copy: 31 (30%) Immediate reproduction: 22 (55%)		
Judgement of line orientation test (total score = 30)	20 (22 nd percentile)	20 (22 nd percentile)		
Facial recognition test (total score = 54)	37 (4 th percentile)	45 (22 nd percentile)		

*References for neuropsychological tests can be found in standard texts.^{5,6}

cognitive difficulties persist. Repeat neuropsychological testing six months after operation was without change except she now scored in the moderate range of depression on the Beck depression inventory.⁷

CASE 2
A 31 year old woman was referred for evaluation of poor organisational skills and chronic depression refractory to psychotherapy and pharmacological intervention. She was noted

to drag her right leg when she was about 18 months old and was diagnosed with cerebral palsy. She did well in all academic subjects except for mathematics, which she failed in the fourth grade. She reported severe depression since early childhood. Her family described her as “introverted” and having a “reserved personality”.

She was noted to have a depressed affect and to avoid direct eye contact. Her speech was monotonic, slow, and measured.

Axial (left) and coronal (right) double echo MRI showing atrophy of the left temporal lobe and dilatation of the left temporal horn tip.

Gestures and body language were restricted. Neuropsychological evaluation (table) showed a 28 point discrepancy between verbal IQ (70th percentile) and performance IQ (9th percentile) as measured by the WAIS-R. There was increased vulnerability to interference on the Stroop color interference test. Dichotic listening for digits showed a left ear advantage (83.3% correct for digits presented to the left ear *v* 66.7% correct for the right ear). Memory testing showed mild difficulty encoding non-verbal information. Language functions were normal. Visuospatial perception and constructions were impaired. Her copy of the Rey-Osterreith complex figure was distorted and drawn in a piecemeal fashion. The interpretation of non-verbal facial expression and body gesture was formally assessed with the audio and video brief exposure portions of the profile of non-verbal sensitivity⁸ and was very impaired.

Neurological examination showed right hemihypoplasia involving the leg more than the arm. The right foot was 1½ shoe sizes smaller than the left. There was a mild right hemiparesis with increased deep tendon reflexes and an extensor plantar response.

An EEG showed brief paroxysmal non-sustained spike and spike slow wave activity from the left midtemporal region. A second EEG was normal. The left hemisphere was shown to be smaller than the right by CT.

Discussion

We describe two female patients with early left hemisphere injury and a developmental syndrome consistent with SEPD. Both patients described a lifelong history of deficient interpersonal relations and chronic depression refractory to pharmacological and psychotherapeutic interventions. Both patients had considerable difficulty with mathematics in school. On formal neuropsychological examination, visuospatial skills and paralinguistic aspects of communication were impaired. Unlike previously described cases of SEPD, however, neurological examinations and neurodiagnostic studies were consistent with left hemisphere injury. Both patients had right sided hemihypoplasia and right sided motor findings. EEGs showed left temporal epileptiform activity and neuroimaging studies left hemisphere abnormalities. Also, intracarotid amobarbitone testing showed normal language after left hemisphere suppression in case 1. Dichotic listening showed a left ear advantage for digit perception suggesting possible anomalous dominance for language in case 2.⁹ Both patients were left handed without a family history of non-right-handedness.

SEPD is a developmental syndrome characterised by (a) interpersonal and emotional difficulties including extreme shyness, social isolation, and chronic, refractory depression, (b) abnormal interpretation and production of paralinguistic communication with avoidance of eye contact, flat or atypical speech prosody, and diminished or exaggerated facial

expression and body gestures, and (c) impaired visuospatial skills with average to superior verbal abilities.² Whereas there is a high incidence of difficulty with arithmetic, several patients have excelled in abstract mathematics.² The discrepancy between our patients' normal performance on the arithmetic section of the WAIS-R and poor performance on the WRAT arithmetic may be related to the WAIS-R being a test of mental calculations whereas the written WRAT may be more sensitive to spatial acalculia.

Unlike the relation between left hemisphere abnormalities and dyslexia, which has been suggested on the basis of neuroanatomical and functional neuroimaging studies,^{10,11} there is little direct evidence linking SEPD to right hemisphere dysfunction. Rudel and Denckla first described the association between SEPD and left sided neurological findings.¹² Voeller found features of SEPD in 15 children who had evidence of right hemisphere injury by CT or neurological examination.³ Grace and Malloy reviewed records from 252 patients at a psychiatric hospital and found 12 patients who met their criteria for right hemisphere learning disability. Three of these patients had right hemisphere abnormalities on EEG and two had right hemisphere abnormalities on CT.¹³ We have described six patients with SEPD and right hemisphere abnormalities on electrophysiological studies.⁴

There are at least four possible explanations for the paradoxical association of SEPD and early left hemisphere injury. First, undetectable right hemisphere damage could be present. A second possible explanation is that injury or epileptiform activity in one hemisphere can exert "abnormal influences" on the remaining hemisphere.¹⁴ This theory has been invoked to explain the improvement in cognitive functioning that is sometimes seen after hemispherectomy. A third possibility is that there could have been reversed cerebral dominance for all cognitive functions before any injury. In this case, early left hemisphere injury alone could result in SEPD. It is not possible to exclude any of these explanations. The first explanation seems unlikely because language would have to develop normally in a damaged right hemisphere. No improvement was seen in any aspect of SEPD in case 1 after left temporal lobectomy as might be expected if the second explanation were correct. The third explanation also seems unlikely as the right hemisphere is usually dominant for directed spatial attention and visuospatial skills even in patients with anomalous dominance for language or handedness.^{15,16}

Finally, it is possible that early left hemisphere injury resulted in displacement of language and hand preference to the right hemisphere and that this reorganisation interfered with the development of cognitive, interpersonal, and emotional functions normally subserved by the right hemisphere. Experimental animal studies have shown that neural reorganisation after early brain injury

can result in the sparing of some functions at the expense of others.^{17 18} In humans, the sequelae of focal brain lesions is influenced by the age and developmental stage at the time of the injury, and early injury may result in considerable sparing of cognitive functions.^{19 20} Little is known about the neuroanatomical substrate of spared cognitive functions associated with early unilateral lesions in humans. It is possible that remodelling takes place analogous to that seen in experimental animals and that these changes interfere with the development of functions that these structures normally subserve. In this way, early left hemisphere injury in our patients could have resulted in pathological left handedness, anomalous dominance for language and "crowding" of the right hemisphere, which resulted in SEPD. The full syndrome of SEPD seems to be an uncommon manifestation of early left hemisphere injury. The "crowding" phenomenon may explain the more common association of early left hemisphere injury and neuropsychological profiles usually ascribed to right hemisphere dysfunction.

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