

Case Reports

Aphasia Following Right Striato-insular Infarction in a Left-handed Child: a Clinico-radiological Study

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Introduction

Acquired aphasia during childhood offers an excellent method of studying inter- and intra-hemispheric specialization for language. However, it is not a common disorder, and the majority of reports have included only a few cases observed over several years, sometimes with different methodologies (*e.g.* Hécaen 1976, Woods and Teuber 1978). Two other potential sources of confusion are the varied aetiologies (trauma, infection, tumour, cerebrovascular accident) and the paucity of clinico-pathological studies. The introduction of the CT scan into clinical practice has not yet greatly modified this picture: lesions were localised by CT scan in only one of the cases reported by Woods and Teuber (1978), and in the child reported by Pohl (1979). Gastaut *et al.* (1979) used CT scan in their study of acute hemiplegia in children, but unfortunately they make no mention of speech disturbance.

Aphasia among left-handed children has been associated with right-hemispheric

damage in 50 per cent of cases (Woods and Teuber 1978), in contrast to only 20 to 30 per cent of left-handed aphasic adults (Brown and Hécaen 1976).

This report describes a left-handed child with acquired aphasia following a right subcortical infarct, and stresses the importance of the localisation of the lesion in understanding the pattern of aphasia and the recovery process.

Case report

C.S., a six-year-old girl, was admitted because of sudden onset of left-sided weakness and speech disturbance. After a few hours of mutism, her spontaneous speech was scanty and lacked fluency, but she could follow two-step oral commands.

The general physical examination was unremarkable and there were no signs of meningeal irritation. Ocular fundi were normal. A defect in the left visual field was observed, together with left central facial weakness, left hemiparesis and left hypoaesthesia to touch, pain and postural sensation. Tendon reflexes were weaker on the left, and a Babinski sign was elicited on that side.

Her history was unremarkable. She had been born at term after a normal labour and there had been no postnatal problems. Psychomotor development had been within normal limits. She was left-handed when assessed by the Edinburgh Handedness Inventory (Oldfield 1971), but because of pressure at school she wrote mainly with her right hand, although her hand-writing

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was far better with her left hand. No other members of the family were left-handed. She had begun primary school five months before the present illness, and already could read and write most simple words, and could do simple calculations.

Normal results were obtained from routine blood and platelet counts, ESR, serum chemistry examinations, tests for blood coagulation, ECG and in an isotopic renogram. CSF examination was normal and cultures were negative. An EEG showed global slowing on the right leads, more marked in the frontal, parietal and temporal areas.

A CT scan was performed on the sixth day of

illness and demonstrated a right-periventricular area of diminished density, involving the internal capsule, lenticular nucleus and the insula. This lesion produced no mass effect, showed no contrast enhancement, and was considered to be a cerebral infarction (Fig. 1). A right-carotid angiogram showed corkscrew irregularities in the supraclinoid segment of the internal carotid artery, and in the proximal part of the middle and anterior cerebral arteries, suggestive of fibromuscular dysplasia. A cerebral Tc^{99m} scan performed on the 34th day was negative.

Detailed neurophysiological examinations were carried out on the third, 18th and 46th days of the

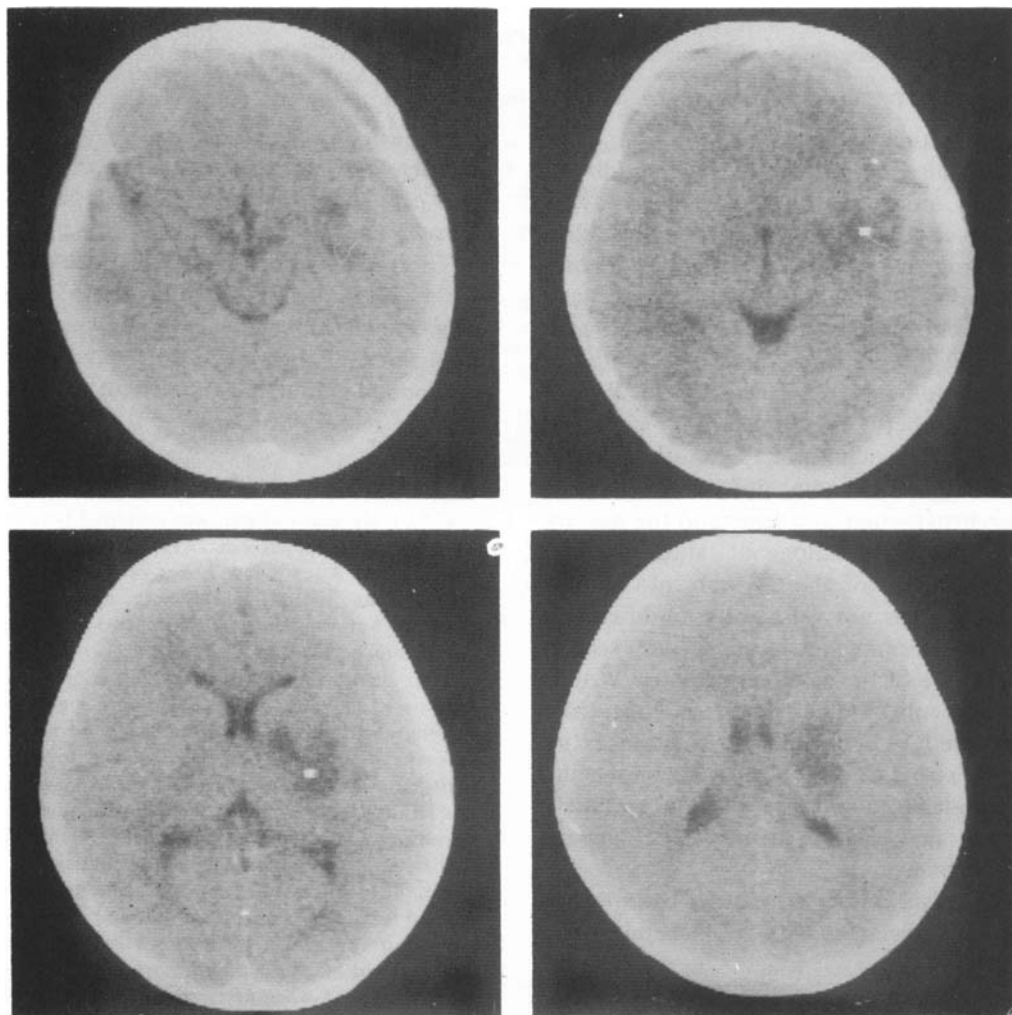


Fig. 1. CT-scan cuts 30mm (*upper left*), 40mm (*upper right*), 50mm (*lower left*) and 60mm (*lower right*) above and parallel with the orbito-meatal line, demonstrating hypodense area in right hemisphere involving internal capsule, lenticular nucleus and insula.

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illness (Table I). At the first examination, C.S. had moderate, non-fluent aphasia and typical right-hemisphere signs such as visual neglect, constructional apraxia and spatial dysgraphia (Fig. 2), and left-channel extinction of dichotic listening. The aphasia improved rapidly, and apart from some articulation errors in spontaneous speech and repetition, the language disturbance had disappeared within about two weeks, as had non-verbal disturbance and left-channel extinction.

Discussion

This patient is similar to other children with acquired aphasia in her initial period of mutism, lack of fluency, good preservation of comprehension and rapid recovery. She also shares some features with adult sinistral aphasia (Brown and Hécaen 1976), namely the relative mildness

of her aphasia, the lack of fluency and the articulatory disturbance.

A peculiar feature of this case is the subcortical location of the lesion producing aphasia. To our knowledge there have been no reports of such lesions producing aphasia during childhood. In adults, striatal and insular lesions have been associated with mutism or with non-fluent aphasia with intact repetition, resembling transcortical aphasia (Benson 1979), and insular lesions are known to cause anarthria (Marie 1926). However, this child had a short period of mutism and non-fluent speech but, in contrast to adult cases, repetition was also disturbed. The subcortical location of the infarct explains the relatively good comprehension

TABLE I
Summary of neuropsychological examinations

	3rd day	18th day	46th day
<i>Verbal functions</i>			
Severity of aphasia ¹	4	5	5
Spontaneous speech	non-fluent articulatory errors literal paraphasia +++	articulatory errors literal paraphasia +	articulatory errors
Praying and singing	disturbed	normal	
Serial speech ²	normal		
Visual naming ²	12/16	14/16	16/16
Visual identification ²	16/16	16/16	16/16
Oral comprehension ²	8/8	8/8	8/8
Token test (22 items) ¹	10	18	17·5
Sentence repetition ¹	2	3	4
Digit repetition ³	2	3	3
Word repetition ²	17/30	21/30	18/30
Reading	many literal errors vowels normal	literal errors	literal errors
Writing (right hand)	spatial dysgraphia	poor handwriting	poor handwriting
<i>Non-verbal functions</i>			
Visual neglect	left +++	0	0
Visual Retention Test Form C (Copying)	1 correct	7	7
<i>Dichotic listening</i>			
18 pairs of common words	14/0	17/16	18/17
19 pairs of digits (right-left ratio)	18/2	18/15	18/17
WISC Verbal		94	105
Performance		98	118
Total		96	112

¹Goodglass and Kaplan (1972); ²Castro-Caldas (1979); ³Benton (1969).

o avô leva a vaca à vila
 no à vila
 o mapa a tia lava a lava
 o mapa leva à vila
 dda a lava a

Fig. 2. C.S. attempting to copy with her right hand. Note left-side visual neglect and spatial dysgraphia.

demonstrated by this child, as the lesion spared the associative auditory areas of the right temporal lobe. The rapid recovery is also consistent with similar adult patients, who also had greater and faster improvement than those with lesions in the classic speech areas (Benson 1979). To explain the rapid improvement in the present case, a 'localisation' factor can be added to the lateralisation and plasticity factors which usually determine the superior recovery among left-handers and children (Brown and Hécaen 1976).

This child's right hemisphere was dominant not only for speech production, as judged by her clinical and dichotic listening patterns, but also for non-verbal functions, as is often the case with left-handed people. She developed a typical right-hemispheric syndrome, consisting of spatial dysgraphia, constructional apraxia and visual neglect. Spatial dysgraphia has also been reported in two other aphasic children with right-hemispheric lesions (Hécaen 1976). Hécaen and Sauget (1971) also found that these right-hemispheric signs were significantly more frequent

among left-handed adults when the lesion was in the right rather than in the left hemisphere.

Dichotic listening may be helpful in assessing improvement of aphasia. When the aphasic disturbance cleared there was improved performance of the left auditory channel, suggesting that language recovery was dependent on right-hemispheric structures.

Acknowledgements: We thank Maria Gabriela Mariano, Speech Therapist, for assistance with the evaluation of this patient, and Maria de Fátima Pereira for typing the manuscript.

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SUMMARY

The case of a six-year-old girl with non-fluent aphasia following a subcortical right-hemispheric cerebrovascular accident is reported. Examination by computerised tomographic scan revealed a right paraventricular area of diminished density involving the internal capsule, lenticular nucleus and the insula. The speech disturbance was followed by typical right-hemispheric signs and by left-channel extinction in dichotic listening tests. The aphasia cleared in two weeks, paralleled by an improvement in left-channel performance, suggesting that language recovery was dependent on right-hemispheric structures.

RÉSUMÉ

Aphasie à la suite d'un infarctissement striato-insulaire droit chez un enfant gaucher : Etude clinique et radiologique

L'article rapporte le cas d'une fillette de six ans ayant présenté une anarthrie à la suite d'un accident vasculocérébral sous-cortical de l'hémisphère droit. L'examen au scanner révéla une région paraventriculaire droite de densité diminuée impliquant la capsule interne, le noyau lenticulaire et l'insula. La perturbation du langage fut suivie de signes typiques hémisphériques droits et par une extinction du canal gauche au cours des épreuves d'écoute dichotique. L'aphasie régresa en deux semaines, avec une amélioration parallèle des performances d'écoute gauche, suggérant que la récupération du langage dépendait des structures hémisphériques droites.

ZUSAMMENFASSUNG

Aphasie nach einem rechtsseitigen striato-insulären Infarkt bei einem linkshändigen Kind: eine klinisch-radiologische Untersuchung

Es wird ein sechsjähriges Mädchen vorgestellt, das nach einem rechtsseitigen cerebrovasculären Zwischenfall eine Aphasie bot. Das Computertomogramm zeigte rechts paraventriculär ein Areal verminderter Dichte, das die interne Kapsel, den N. lenticularis und die Insel einbezog. Nach der Sprachstörung folgten typische rechtsseitige Befunde, sowie linksseitiger Hörverlust beim doppelseitigen Hörtest. Die Aphasie bildete sich innerhalb von zwei Wochen zurück zugleich mit einer Besserung der linksseitigen Hörleistung. Man nimmt daher an, daß die Spracherholung auf Strukturen der rechten Hemisphere beruht.

RESUMEN

Afasia consecutiva a un infarto estriato – insular en un niño zurdo: estudio clínico-radiológico

Se presenta el caso de una niña de seis años de edad con una afasia no fluente consecutiva a un accidente cerebro-vascular subcortical del hemisferio derecho. El examen con TAC reveló una área paraventricular derecha de densidad disminuida que interesaba la cápsula interna, el núcleo lenticular y la ínsula. La alteración del lenguaje fue seguida de signos típicos del hemisferio derecho y de una extinción en el canal izquierdo en los tests de escucha dicóticos. La afasia desapareció en dos semanas, con una mejoría paralela de la realización en el canal izquierdo, lo que sugiere que la recuperación del lenguaje dependió de las estructuras del hemisferio derecho.

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Rhombencephalosynapsis: a Viennese Malformation?

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Introduction

'Rhombencephalosynapsis' is a rare malformation, described in detail by Gross and colleagues (1959, 1978). It is remarkable that the first observation of this malformation (Obersteiner 1916) and the four subsequent cases were all reported in Vienna. The essential features are absence of the incisura cerebelli posterior; fusion of the cerebellar hemispheres; absence of the velum medullare anterius and nuclei fastigii; and fusion of the dentate nuclei, which are shifted towards the midline.

To our knowledge, only one additional case of this syndrome has been reported outside Europe. Kepes *et al.* (1969) described a case with congenital fusion of the thalami, associated with atresia of the

aqueduct and midline fusion of the cerebellar hemispheres. A seventh case is described in the present paper.

Case Report

The child was the male non-identical twin of a 23-year-old primiparous, primigravida mother from a village east of Ulm, Germany. The pregnancy was uneventful until the 38th week, when a caesarean section was performed because of hydrocephalus in one of the twins. The affected boy weighed 3510g, body length was 56.4cm and head circumference was 42.5cm. There was an 'egg-sized', soft, fluctuant, parietal protrusion in the region of the sagittal suture; the latter was dehiscence over an area of 5 x 10cm. Radiological examination of the skull showed craniolacunia and a bifid spine at C1 to C4. Pneumoencephalography done on the third day showed communication between a left paramedian encephalocele and the left lateral ventricle, which was enormously dilated. (CT scans were not available in 1977.) Movements and tonus were reduced, but no other malformations were found. Serology for rubella, cytomegaly, lues and

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