CLINICAL ASPECTS OF ACQUIRED APHASIA AND DYSARTHRIA IN CHILDHOOD

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Verworven afasie en dysarthrie bij kinderen

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Aan de mijnen

General introduction

For the last decade, it has been a common clinical belief that the prognosis of acquired childhood aphasia is good. However, our own clinical experiences were rather conflicting on this point. As a consequence, we re-examined all the children (15) with an acquired aphasia who in a period from 1969-1972 were hospitalized in the University Hospital Rotterdam-Dijkzigt. On re-examination 7 children failed to show complete recovery (VanDongen and Loonen, 1977).

One year later Woods and Teuber published an outstanding study entitled "Changing patterns of childhood aphasia", in which they also contradicted the expectation of a good recovery of acquired childhood aphasia. In addition, they criticized the view supported by the earlier literature that "childhood aphasias differ in kind from adult aphasia being predominantly motor or non-fluent".

The research carried out after 1977 on the classification and the recovery of the different speech disturbances is subject of this thesis.

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CHAPTER 1

ACQUIRED CHILDHOOD APHASIA: A REVIEW

In adults, the term "aphasia" is consistently applied to conditions where language functions become impaired as a consequence of cerebral lesions which usually involve the left hemisphere.
In childhood aphasia, this term has been confusing and controversial for many years. Within the great variety of childhood language abnormalities, 3 disorders have been labeled as aphasic:

1. congenital aphasia.
2. developmental aphasia.
3. acquired aphasia in childhood or "true dysphasia".

Although most authors do not differentiate between congenital aphasia and developmental aphasia, a few explicitly do (Landau *et al.* 1959).

1. CONGENITAL APHASIA has been defined as an isolated language deficit in the absence of hearing-loss, emotional disturbances or intellectual subnormalities as a consequence of early and extensive lesions in the reciprocal thalamocortical projection systems (Vargha-Khadem *et al.* 1985). It has to be emphasized that the use of the term congenital aphasia implies the presence of structural cerebral lesions.
2. In contrast to congenital aphasia, DEVELOPMENTAL APHASIA ("a deficit in the acquisition of normal language functions in children of normal or above intelligence and with adequate hearing ability to permit the perception of verbal sounds", Wyke, 1978) is not associated with a clear pathogenetic mechanism. These children are presumed to have some cerebral dysfunction, but efforts to delineate the neurological status are lacking. To illustrate this issue: Rapin and Wilson (1978) argue that "the failure to develop language is always, except in rare instances of extreme and environmental deprivation, the consequence of neurological dysfunction, affecting pathways which transmit information from the ear to the brain, within the brain itself or from the brain to the muscles of articulation". Trying to specify this dysfunction Rapin and Wilson (1978) state that these children "may not be suffering from a lesion in the central nervous system but rather from a delay in the maturation of relevant neurological systems". In accordance with this statement is their conclusion: "the average non-verbal child whom a child neurologist is asked to examine does not usually have gross evidence of neurological impairment". The so called "soft signs" come to the fore

as shown by a study of Johnston *et al.* (1981). They compared the neurological status of a group of children with developmental aphasia with a matched group of normal children. According to their findings, the language impaired children were less efficient in tasks involving rate of movement, perception of dichhaptic stimuli and left/right identification.

More recently many authors prefer to avoid the term aphasia for this disorder. They argue that this language disorder is not based on focal cerebral damage and that on that account neither the clinical features nor the anatomical correlates can be the same as those of acquired aphasia. Consequently the term developmental aphasia has been changed in "specific language impairment" or "primary language disorder".

3. ACQUIRED APHASIA OR "TRUE" DYSPHASIA. This term will be reserved for children, who have developed language in a normal fashion, but - following brain damage - show a language abnormality (Benson, 1979). Although this definition is very consistent, there are remaining problems. On the one hand, in infancy, it is not possible to fix the end of the preverbal period and the beginning of the verbal period exactly. In addition, there is the theoretical issue of defining how and when language starts: with verbal comprehension or are the first words the signs of language? Consequently, the diagnosis of acquired aphasia in children of 2 years of age or younger is a very difficult one and has to be made with great care. On the other hand, the distinction of childhood language from adult language is a complicated one. On this subject no clear-cut criteria are present.

   After the age of 15, however, language disorders are rarely labeled as "acquired childhood aphasia" but rather as (adult) aphasia, although, as mentioned above, criteria are lacking. The subject of acquired aphasia in childhood has received considerably less attention than has developmental aphasia.

Historical background

The first description of childhood aphasia emerged soon after the classic report of Paul Broca (1861). Clarus (1874) reviewed the literature and - although his definition of aphasia was too wide - (aphasia in idiots!), he presented 47 cases of childhood aphasia. The aphasia was often accompanied by right-sided hemiplegia and convulsions. His conclusions were:

1. acquired aphasia is not rare in childhood.
2. the prognosis is not always favorable. He indicated a permanent loss of speech in approximately 50% of the cases. He suggested that both aetiology and severity of the lesion determine the degree of recovery.
3. there is an ability of the right hemisphere to take over speech in left-hemisphere lesions:

"Das urspruengliche linksseitige Sprachzentrum ist ja hier gaenzlich ausser Funktion, dafuer muss nun das bisher noch ungeuebte rechtsseitige Sprachzentrum in dessen Funktionen eingefuehrt werden".

Eleven years later, Bernhardt (1885) confirmed Clarus' conclusions that aphasia in childhood was not rare, and he also assumed that the right hemisphere can subserve speech. Moreover, he stated that in infantile cerebral hemiplegia, aphasia is in general of the motor "Broca" type. Auditory comprehension of spoken language would be intact. He denies the bad prognosis: "aphasia will be mostly transient, rarely permanent". This conclusion is striking since Bernhardt's last statement is based on the study of 6 children, 4 of them with a follow-up of more than one year; in 2 of the cases speech was still severely disturbed. Nevertheless, the conclusion remained essentially unchallenged until recently.

In 1897, Freud published a monograph on cerebral palsy, in which he remarked on the restricted vocabulary and emphasized that aphasia in children occurred relatively "commonly" in lesions of the right hemisphere, i.e. "crossed aphasia". Crossed aphasia refers to the combination of right hemiparesis with aphasia in a left-handed child or left hemiparesis and aphasia in a right-handed patient. Crossed aphasia in dextrals is extremely rare. Brown and Hécaen (1976) reviewed the literature which yielded only seven well documented cases.

From the above-mentioned literature it is clear that the traditional principles attributed to childhood aphasia had already been formulated in the last century:

- the pattern of aphasia is of the motor type (Broca, non-fluent),
- acquired aphasia after right hemisphere lesion is common,
- the prognosis is favourable,

These principles have been uncritically accepted, although this would imply qualitative differences between children and adults. In the first half of the 20th century interest was focused on adult aphasia. Little has been added to the subject of childhood aphasia. Exceptions were 2 studies in which aphasic children with auditory verbal defects are described (Poetzl, 1926; Brunner and Stengel, 1932). These studies contradicted the earlier suggestion of a dissociation between mechanisms of verbal expression and verbal comprehension. In the early sixties a study of Basser (1962) reemphasized the relatively high incidence of speech disturbance following a right hemisphere lesion in right-handed children as compared with adults. Basser, in accordance with Freud, suggested that both hemispheres participate in the development of speech, and that damage to one of them can cause aphasia. He further speculated that with age and maturation of the cerebral cortex a progressive shift from bilateral representation to unilateral specialization on the left occurs.

Referring to this theory of equipotentiality Lenneberg (1967) specified that in adolescence the right hemisphere had lost its active role in speech and consequently complete recovery of aphasia would be possible only under an age threshold of 11 years at the time of the injury (12 patients retrospectively studied). However,

Krashen (1973) reinterpreted Lenneberg's data and concluded that lateralization to the left hemisphere was completed after the age of 5. Later, this age threshold moved even more downwards. Woods and Carey (1979) studying 25 children stated that left hemisphere lesions acquired after the age of one year leave residual impairment of language functions, if they cause initial aphasia.
In the last decade, there is a growing interest in the subject of childhood aphasia. More recent reports contradict the currently accepted principles supported by the earlier literature, not only concerning the prognosis but also concerning the pattern of aphasia and the role of the right hemisphere in relation to language.

In the literature conflicting statements on the incidence are made. For example Denckla (1979) said: "as the sole 'cortical function' consultant in a large neurological institute, between 1969 and 1976, I saw only seven cases of acquired aphasia in children under 10 years of age". In contrast, Hécaen (1983) stated, that aphasia is more frequent, at least among the youngest children, than in adults. An explanation for this controversy lies in the starting point. Hécaen, referring to children with unilateral cerebral lesion, considered aphasia a frequent symptom. Since unilateral lesions of neoplastic or vascular origin are relatively uncommon in children in comparison to adults, the frequency of aphasia in children is equally low. Exact estimates of the incidence of acquired childhood aphasia can not be given: consecutive series on this subject are completely lacking. An unreliable view as to the frequency of childhood aphasia emerges from the description of series of children with circumscribed aetiology. Issler (1971) reported 20% aphasics in a series of 114 children with an acute hemiplegia and Sloof (1966) mentioned 4 aphasic children in 28 with (left) cerebral hemisphere tumours.

The clinical picture 1897 (Freud) - 1980

Until the sixties a consensus existed about the general nature of this language disorder which can be summarized as follows: "comprehension disorders are rare; expressive language problems, which can range from mutism to mild articulation defects, are predominant". However, this consensus seems to be more apparent than real.
Firstly, the spontaneous speech of aphasic children has been described in rather vague terms, for example: "poverty of speech, reduction of spontaneous speech, unwillingness to speak, reluctance to speak or absence d'incitation à la parole". Especially the last denotations suggest an emotional disturbance rather than a specifically aphasic one.
Secondly, there arise diagnostic problems. How to differentiate in "mild articulation defects" between aphasia and dysarthria? The same problem emerges on the subject of mutism. For example, Todorow (1978) described mutism in children following a

trauma as a psychological reaction to their intolerable condition after the accident. He labeled this as "Dornroschenschlafzustand".
Thirdly, in describing the subsequent reorganization of speech function after traumatic mutism, no aphasic stage need occur, especially not in case of a midbrain syndrome (VanCramon, 1981).

In these studies the clinical investigation of aphasic children had mainly been limited to their spontaneous verbal output. The traditional view on childhood aphasia started to change when more extensive and precise analysis of language disorders was carried out. A first example is the study of Guttmann (1942). For this author the childhood aphasia was still characterized by an absence of spontaneous language. He was however, the first to add some specific features to the clinical picture. Thus, he noted the telegraphic style; the dysarthric difficulties whereas jargon jargon aphasia did not occur. In 1965, Alajouanine and Lhermitte gave a more extensive description of the spontaneous speech, in which they emphasize that - especially in the acute stage - spontaneous speech was nearly absent. In addition, they gave more extensive data about the language disorder and in their series of 32 children they observed that:

1. logorrhoea, frequent in adults with left temporal lesions, is not observed in the child even when clinical signs and instrumental results point to a temporal locus of the lesion.
2. paraphasias, either verbal or phonemic, occur very rarely and when they occur, they predominate among children older than 10 years of age.
3. verbal stereotypes and perseverations are completely lacking.
4. "The syntax is simplified rather than erroneous. Agrammatism such as observed in adults was not present in any of the children ....".

Gloning (1970) analyzed 8 cases of early onset aphasia (pre-school children). He did neither observe neologisms nor jargon aphasia. The results of Assal and Campiche (1973), who studied 18 children with traumatic aphasia, are consistent with these data. They state: "a la relative monotonie de l'aphasie traumatique de l'enfant s'oppose la symptomatologie diversifiée de l'adulte où tous les grandes syndromes classiques et parfois des formes dissociées très pures s'observent". They also mention syntactical problems. The children express themselves in "mots isolées ou phrases très courtes". Less is known of the nature of the articulation problems. Most authors label these as "dysarthric deficits" but they do not specify the type. For example, Alajouanine and Lhermitte (1965) state that the dysarthria in children has no specific features different from dysarthria in adults, provided that the stage of phonetic development of the child at the onset of the aphasia is taken into account. They do not specify this statement in terms of articulatory breakdown or voice qualities.
The assessment of other expressive language modalities as repetition or object naming - frequently used in the examination of adult aphasics - has not been

performed in studies of series of aphasic children. Only Hécaen (1976) incidentally mentions that "disturbances of naming have a great frequency and tend to persist". About the quality of the errors no details are provided. In case studies, repetition and object naming have only exceptionally been examined. Great difficulties in these tasks are common, not only in articulatory aspects but also in syntactical qualities if sentences have to be repeated. Both procedures, important in detecting specific types of aphasia, have been neglected in the assessment of childhood aphasia. Consequently, the variety of language defects demonstrable in childhood aphasia has received insufficient attention. Finally, Alajouanine and Lhermitte (1965) have noted that language disturbances take on different aspects according to age. In their series, the group under the age of 10 had a more severe reduction of verbal expression. Moreover, in this group disorders of articulation relatively frequently occurred. These variations according to age have not been generally confirmed by others (Guttmann, 1942; Hécaen, 1976).

As mentioned above, impaired comprehension of spoken language has been considered a rarely occurring deficit. Earlier reports refer to very simple tasks in assessing this modality "put out your tongue" and so on. Such a request was often correctly carried out, and it was then concluded that comprehension was "normal" or "satisfactory". After the study of Poetzl (1926) who explicitly demonstrated severe verbal comprehension deficits in children and suggested an association between this deficit and the (temporal) locus of the lesion, this deficit was more often described in subsequent studies on childhood aphasia. Guttmann (1942) observed a verbal comprehension deficit in 3 of the 16 aphasic children. He suggested that the size of the lesion was an important factor causing such a deficit. In the study of Alajouanine and Lhermitte (1965) disorders of comprehension were observed in more than one-third of the cases (12 out of 34). They stressed the importance of age at onset of the cerebral lesion. In children who acquired aphasia between 5 and 9 years, understanding of spoken language was nearly always intact.

Finally, until the eighties there are a number of case studies, in most of which disorders of comprehension are prominent. Language disorders are considered a very frequent symptom in spontaneous writing as well as in writing to dictation. About the type of spelling errors, Alajouanine and Lhermitte (1965) stated that "these were often dependent upon phonetic disturbances, showing the links between oral language and its graphic code in the dynamic of writing". It is noteworthy that in their series 4 patients had writing disorders with jargon aphasia. Also, reading disorders are not rare in aphasic children. Reading can be severely disturbed: the children can not recognize or spell the letters of a word.

Recovery

The standard doctrine about childhood aphasia claims that recovery of language function is rapid and complete. However, this claim is largely supported by the earlier literature and contradicted by more recent reports. In 1942, Guttmann threw doubt on this claim. He described 16 aphasic children with mixed aetiology: trauma, brain tumour, abscesses and thrombosis. The follow-up period ranged from 2 months to 2 years. The author concluded that recovery of aphasia was related to the cause, 5 out of the 6 trauma cases recovered. In contrast, there was less improvement in the abscess cases (1 out of 3) and no improvement after thrombosis. Guttmann provided further evidence that recovery was related to an initial motor type of aphasia as well as by the size of the lesion and the fact that "aphasic signs were still present 4 weeks after onset". Additional information with respect to school achievements is not available.
The same holds true for Basser (1962) in a follow-up study of 20 aphasic children. His follow-up ranged from 3 months to 2 years. He noted that "speech loss was not permanent in any case". According to his observations recovery was related to age at onset. A more protracted recovery was observed in those cases in which the lesion occurred before the age of 2. Assessment of this statement is difficult, because in his series many of the children were functioning intellectually at a very low level. Lenneberg (1967) conducted a 2-year follow-up of 6 cases. He reported that recovery was complete for all children who were under the age of 11 at the time of injury. Three of these 6 cases had a traumatic aetiology. Data on school achievement were not provided. Alajouanine and Lhermitte (1965) presented follow-up data on 32 children, ranging in age from 6 to 15 years. Their series included various causes:

- trauma (13)
- intracerebral haematoma (10)
- brain tumour (2)
- occlusion of the arteria cerebri media (2)
- unknown (infectious disease?) (5)

At follow-up, recovery was observed in 23 of the 32 children. The role of aetiology was not evaluated. The authors reported data on school achievement and state that - despite the restitution of language functions - "considering school difficulties, encountered by these children, it appears that subjects involving the use of language were more difficult than mathematics". This raises doubt on the completeness of the recovery. Moreover, they mention that only 2 children "had a social evolution which can be considered as satisfactory, one is a trade booker and the other is in hotel staff". In both cases the aetiology was of traumatic origin. Byers and McLean (1962) studied 10 hemiplegic children suffering from aphasia (vascular origin). After a follow-up period of 1-4 years they describe a complete restitution of speech function although their data show a mild to severe impairment on verbal tests.

Gloning (1970) described 8 pre-school children with acquired aphasia. He related an unfavourable prognosis to severe bilateral cerebral lesions. He does not provide data on his follow-up period.
Assal and Campiche (1973) emphasize as a prognostic sign brainstem dysfunction. The rate of improvement of the aphasic children in the 2 last cited studies was not fully described.
Hécaen (1976) described at first 19 and later (1983) 36 aphasic children in age ranging from 3 to 15 years. Trauma seems the most frequent aetiology, however, no exact data on this subject are available. During the follow-up period which varied from 1 to 8 years 4 of 6 children showed considerable improvement, one became more aphasic (astrocytoma) and one child did not recover at all. Data about a follow-up period from 3 months to one year are available for 13 children. Recovery from aphasia was complete in 6 children; and considerable improvement was noted in 7 children. However, no information is provided on the remaining children, the role of age and aetiology remains unknown. Hécaen expressed the view that persistent aphasic symptoms are linked with the size and the bilaterality of the lesions.
Finally, in 1978 Woods and Teuber published a study, entitled "Changing Patterns of Childhood Aphasia". This study of 65 children with unilateral hemispheric lesion occurring after speech acquisition, conflicted with the earlier notions that acquired aphasia in children is frequently due to right hemisphere lesions. They argue that "before the introduction of penicillin (1930) one third of the total number of childhood aphasias were claimed to have followed right hemisphere involvement" and show that aphasia was frequently listed as a complication of a systemic infectious illness, which, as neuropathological studies have shown, can cause diffuse encephalopathy extending throughout both cerebral hemispheres. Moreover, their arguments gain in strength by the fact that in recent studies the incidence of crossed aphasia dropped to 5% or less (Woods, 1983; Carter *et al.* 1980).

Patterns of recovery

The knowledge of the course of aphasia is limited to incidental observations. In Hécaen's opinion a stage of mutism in traumatic aphasia will be followed by dysarthric difficulties. Alajouanine and Lhermitte (1965) noted that in spontaneous speech phonetic paraphasias disappeared within 6 months after onset. They observed that articulation disorders persisted longer. Naming difficulties (Hécaen, 1976) tend "to persist even after the children returned to school and are mentioned explicitly in school reports". The same applies to writing disorders in contrast with reading disorders. Alajouanine and Lhermitte (1965) contradicted the view of Hécaen (1976) that disorders of verbal auditory comprehension were exclusively present in the early stage and disappeared rapidly. In their series a comprehension

deficit was evident 6 months after the onset of the aphasia in 1/3 of the aphasic children.
Finally, Niebergall *et al.* (1976) found various patterns of recovery in a group of 6 traumatic aphasics (between 10 and 17 years old). He observed a more rapid recovery of verbal comprehension than of object naming as well as the opposite.

In the last decade observations on childhood aphasia differ from the earlier reports in:
a. the clinical picture.
b. the prognosis and recovery.
c. the site of the lesion.

The clinical picture

Woods and Teuber (1978) reported the jargon aphasia in a 5 years-old boy as "an unexpected observation". Subsequent studies agree on the presence of fluent aphasia. Thus Martins *et al.* (1981) found 3 cases of fluent aphasia in a follow-up study of 19 children. Moreover, they refer to a high incidence of auditory comprehension deficits, even in cases of non-fluent aphasics. VanHout *et al.* (1985) presented the results of detailed clinical language examinations in 11 cases with acquired aphasia seen in a 3-year period in a neuropediatric center. They not only reported 2 cases of more marked neologistic jargon aphasia but also of paraphasias. They stated that the presence of paraphasias in the children's verbal output was not the exception but the rule. In addition, verbal perseverations were present while in 7 cases impairment of verbal comprehension ranging from mild to severe was noted. Moreover, in describing a case of acquired conduction aphasia in a child (Martins and Ferro, 1987), evidence is presented that the picture of childhood aphasia was very variable and not limited to the motor type. Cranberg *et al.* (1987) relating CT scan data to severity and type of aphasia in 8 children documented one fluent case. Finally, it has to be emphasized that in many case-reports on the Landau-Kleffner syndrome the presence of neologisms and jargon are emphasized.

Prognosis and recovery

When children with left hemisphere injuries are properly tested for language function and compared with appropriate controls, the recovery of language is less complete than has been suggested in earlier reports. These findings are evidently more in accordance with the observed learning difficulties encountered at school. Woods and Carey (1979) showed that left hemisphere lesions, if they caused initial aphasia, left significant residual impairment on most of the language tasks.

A study of Vargha-Khadem *et al.* (1985) confirmed this finding. These authors concluded that in cases from age 5 onwards left hemisphere lesions the impairments were most marked. From their test data it appeared that this tendency was more evident with respect to naming rather than with respect to auditory comprehension deficits.
The study of Martins *et al.* (1981) in which 19 children, 3 years after the onset of the aphasia were assessed, showed that only 2 had recovered according to the results on standardized language tests.
Goorhuis-Brouwer and Deelman (1983) presented a follow-up study of 10 children over a period of 2 to 5 years. A complete recovery was observed only in 2 children.

Site of lesion

As mentioned above, aphasia in children after right hemisphere lesion is rare. To our knowledge, after the sixties only one fairly extensive report has been published (Assal and Deonna, 1977). In addition, 2 cases with aphasia following a subcortical lesion have been presented. Ferro *et al.* (1982) described a left-handed child with a non-fluent aphasia following a right-sided sub-cortical infarct. Aram *et al.* (1983) studied a case with initial mutism, comprehension deficit, anomia and oral apraxia caused by a left-sided lesion in the putamen, anterior limb of the internal capsule and lateral aspect of the head of the caudate nucleus. Both children recovered satisfactorily. Articulatory disorders and impaired writing persisted for a longer period (6 months).

Conclusions

In the last decade, the traditional ideas on the clinical pattern of acquired childhood aphasia have been considerably modified. Apart from the motor (Broca, non-fluent) type, other types of childhood aphasia have been observed and described.
Fluent aphasia in children has recently been reported in different varieties, among which conduction aphasia. In accordance with this trend is the finding that verbal comprehension deficits are not an exception but can be detected in nearly all aphasic children if appropriate tests are administered.
The optimistic view that recovery of aphasia in children would be rapid and complete, has not been confirmed in recent studies. Now, a number of unfavourable prognostic signs are known: aetiology, size of lesion, type of aphasia and age at onset. Some follow-up studies illustrate the importance of these variables. Standardized language tests can uncover mild but persistent aphasic deficits even many years after the onset of aphasia. This can explain the well-established fact of bad school

achievements in children in spite of their apparent clinical recovery.
Acquired aphasia in right-handed children caused by a right hemisphere lesion (crossed aphasia) seems to be rare; its frequency is nearly identical with that of right-handed adults. Moreover, no case studies have been published in which the lesion was confirmed by CT scan, in contrast with subcortical aphasia, an uncommon symptom in children as well as in adults. Two case reports of this clinical picture have been very well documented by CT scan.

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CHAPTER 2

FLUENT APHASIA IN CHILDHOOD

Introduction

The most salient characteristic of acquired aphasia in childhood is the non-fluency, ranging from marked reduction in speech to absolute mutism (Denckla, 1979). However, the terminology used to describe the aspects of non-fluency may create confusion, as detailed accounts of language functioning in aphasic children are rare. Terms to describe the expressive difficulties of non-fluency are:
- loss of initiation of speech (Hécaen, 1976);
- hypospontaneity of speech (VanHout, 1983);
- reduction of expressive activities (Alajouanine and Lhermitte, 1965);
- Sprachlosigkeit (Poeck, 1975);
- poverty of speech (Guttmann, 1942);
- suppression de la parole (Assal and Campiche, 1973).

Moreover, many authors observed in the initial stage of the aphasia a period of "mutism".

While the above mentioned terms are rather vague, additional labels attempting to specify the nature of this deficit are not clarifying. For example, Alajouanine and Lhermitte (1965) state concerning the syntax: "It is rather simplified than erroneous", but an explanation of this statement is missing. The same holds true for claims as "articulatory difficulties" and "diminution of lexical stock" (Hécaen, 1976, 1983), the verbal output is characteristically "slow and sparse" (Benson, 1979). Moreover, one has to note that mutism, reduction in speech or articulation difficulties do not refer exclusively to aphasic deficits. The term mutism denotes the inability of a person to produce verbal or non-verbal vocal utterances. According to VonCramon (1981) to reinforce this meaning the term "absolute mutism" should be used, while the term "word mutism" is limited to the condition that the patient is not able to produce verbal utterances. In describing the subsequent reorganization of speech function after mutism, no aphasic stage has to occur in adult patients suffering from an acute traumatic midbrain syndrome or as a consequence of callosotomy (Sussman *et al.* 1983).

Is there a possibility to make a differentiation between aphasic and non-aphasic mutism or even psychological mutism? As Geschwind (1974) argues: "If an adult patient is mute but writes normally, you can be almost absolute sure that he is not aphasic". Mutism in the child presents a much greater problem. Children may

become mute under many circumstances, especially under situational stress and the non-writing child who is brought in the hospital with a head injury and does not speak, creates a problem in diagnosis. Also the term "reduction in speech" does not refer specifically to aphasia, while articulation difficulties are frequently described in aphasics as well in children as in adults. However, it can be a sign of dysarthria without aphasia, even in a child with right-sided hemiparesis (Aram *et al.* 1983).

These remarks demonstrate the need for a more exact description (definition) of non-fluent versus fluent aphasia, especially because recent studies have cast doubt on the predominant non-fluent pattern in aphasic children.
How to establish the classification of aphasic speech in a rather well defined fluency/non-fluency dimension? On the basis of a statistical analysis of the spontaneous speech Howes (1964) found a range of 12 to 220 words per minute in aphasics, whereas normal control-persons produced 110-175 words per minute. This implied that there is a number of aphasic patients speaking faster than normals, later on termed as fluent aphasia (Howes and Geschwind, 1964). However, this speaking at a supernormal rate in aphasic patients has not been confirmed in other studies (Wagenaar *et al.* 1975; Kerschensteiner *et al.* 1972). The last mentioned authors investigated whether the fluency/non-fluency dimension of spontaneous speech could be confirmed by mathematical analysis (cluster analysis). They demonstrated that - if the speech production of a group of unselected aphasics was rated by using 6 clinical characteristics (see Table 1) - this procedure yielded 2 distinct groups reflecting "naturally" occurring differences in language behavior and corresponding to the *clinical* syndromes of fluent and non-fluent aphasia. Moreover, they established a rank order of these characteristics according to their power of discriminating between the 2 groups.

Table 1 Criteria for the classification of fluent versus non-fluent speech.

	non-fluent	fluent
1. phrase length	predominantly 1- 2 words	4 word phrases
2. pauses	many	normal
3. prosody	impaired	normal
4. rate of speaking	0-50 words/min	>90 words/min
5. effort	marked	none
6. articulation	markedly dysarthric	normal

For the definition of the variables, see Kerschensteiner *et al.* (1972).

In the period from 1976 until 1983 we studied 33 children with acquired aphasia. The diverse aetiologies are summarized in Table 2.

Table 2 Aetiology of acquired aphasia in children.

	male	female
head injury	8	2
vascular disease	4	4
infectious disease	5	0
brain tumour	1	3
syndrome of Landau and Kleffner	2	4

In 5 patients speech was fluent and we present a detailed report of the language assessment, the CT (respectively technetium) scan abnormalities and relationship of the type of the aphasia to the site of the lesion.

Patients and methods

Patients

The patients were 4 girls, aged 9 to 11 years and a 5 year- old boy. One girl was left-handed (case 4). They all had a normal psycho-motor and speech development. Before the onset of the aphasia none of the children had shown symptoms or signs of neurological disease, neither did any of them have learning problems at the elementary school.

Methods

Computer tomograms

Altogether, 11 CT scans of the patients were performed. Three were made with a second generation scanner and 8 with a third generation scanner. The plane of orientation was chosen approximately parallel to the orbito-meatal line. The thickness of the slices varied from 8 to 13 mm. The number of slices taken at each examination varied from 8 to 12. In general, scanning was carried out according to standard techniques. The interpretation of the pictures was done with standardized window-setting. For the present investigation it was important to localize possible abnormalities in front or behind the sylvian fissure or in a combination of these sites. For this purpose, the sylvian fissure was identified: we took into account slight

differences in the plane of orientation, using the anatomical studies of Matsui and Hirano (1978), and Gado *et al.* (1979) as guides. In case 4 only one technetium scan was available.

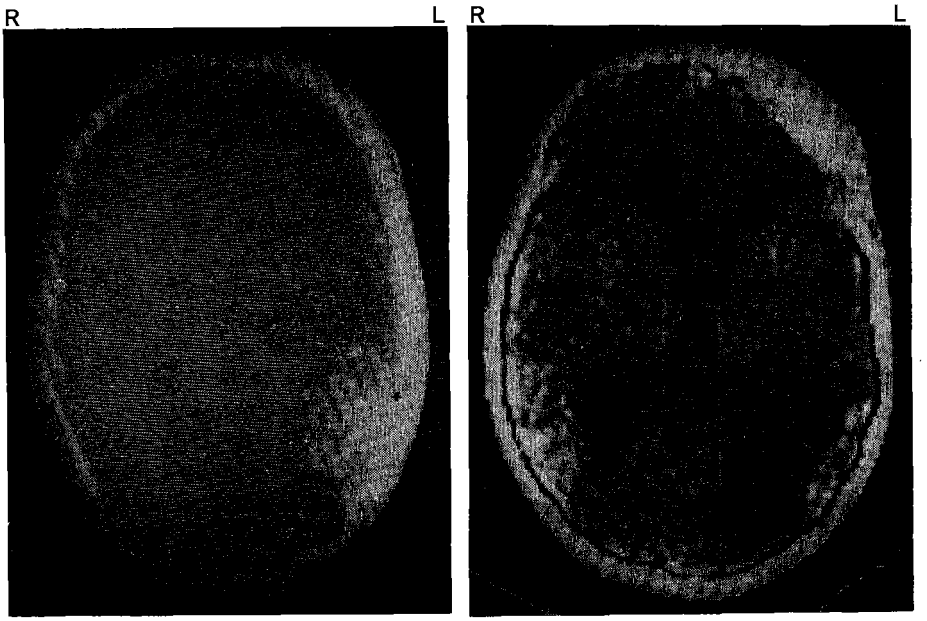
Language testing

The initial language testing was performed as soon as possible after the onset of the aphasia. In each patient follow-up examinations were performed. Furthermore, we collected information about deficits in the use of language at school one to 5 years after the onset of the aphasia.
Fluency is best judged from speech during conversation. Spontaneous speech was recorded in a conversational setting in the presence of the mother. The speech production was rated according to the 6 most specific criteria proposed for the classification of fluent versus non-fluent speech, which are shown in Table 1. In addition to the evaluation of spontaneous speech, language assessment consisted of picture naming (40 items), repetition of words (22 items) and sentences (10 items), reading aloud and writing to dictation, and the Token Test, a sensitive test of ability to comprehend non-redundant verbal information. Finally, a non-verbal intelligence test (Coloured Progressive Matrices) was administered to patients 2, 3 and 4.

Reports of the patients

Patient 1, Born 11.01.1971.

This girl suffered from aplastic anemia. On 03.03.1980 she was found to be dull and she responded inadequately to questions.
On examination, nuchal rigidity was present. Moreover, she was dysphasic, and was hemiplegic on the right side. The CT (Figure 1a) made on the day of the onset of the neurological signs, showed a 3 x 4 cm hyperdense space-occupying lesion, surrounded by a halo of hypodensity located in the left temporal region. The density pattern and the presence of blood in the subarachnoidal space suggested a haematoma in the left hemisphere with extravasation of blood in the subarachnoid space.
The patient was treated for 10 days with suspensions of thrombocytes and corticosteroids in high doses. Seventeen days after the onset of the neurological signs, the extent of the haematoma on CT was grossly reduced. The lateral ventricles were slightly enlarged. After about one month a bone-marrow transplantation was carried out. Two months later she had a series of generalized tonic-clonic fits. Treatment with 2 x 100 mg phenobarbital R daily was started whereafter she remained without convulsions. The hemiplegia gradually recovered. One year after the onset of the neurological signs, a third CT scan (Figure 1b) showed diffuse enlargement of the ventricles and there was a hyperdensity in the periphery, suggesting cortical or subcortical calcification.



a *b*

Figure 1a *Patient 1. Computed tomographic scan at onset shows a haematoma posterior to the sylvian fissure.*

Figure 1b *Scan one year after onset, demonstrating enlarged ventricles, dilated posterior horn, and possible calcification in the left temporo-occipital cortex.*

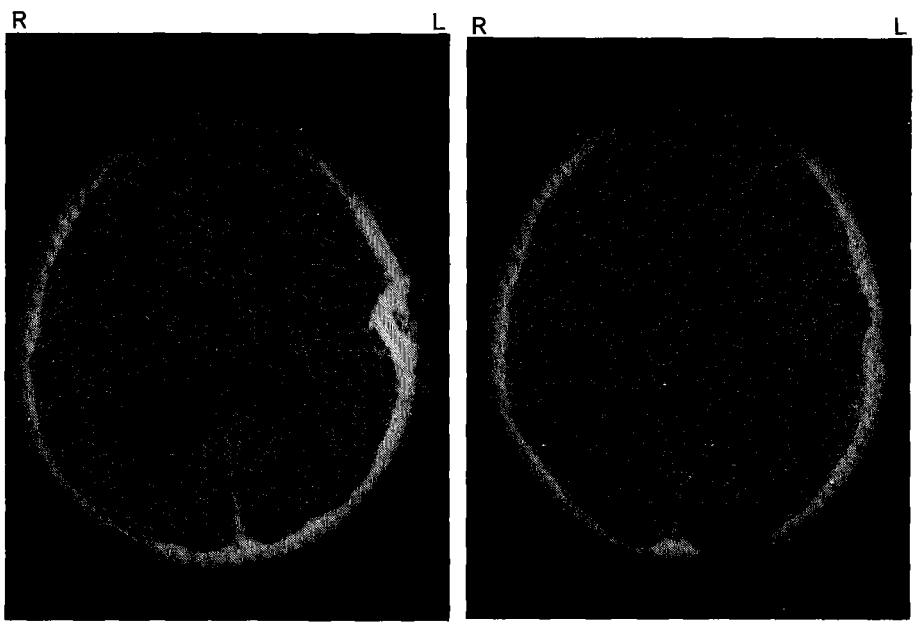
Language testing

Speech was formally evaluated for the first time one month after the onset of the aphasia. The girl was cheerful and talked in a high speech rate (121 words per minute). There were no articulation disturbances. The prosody was normal. Spontaneous speech impressed as empty by the many stereotype sentences. Her utterances were occasionally longer than 12 words. Although neither paraphasias nor neologisms were present during the interview, both abnormalities were observed in the extensive circumscriptions in the picture naming test. The naming difficulties were severe and she stopped the test after 10 trials. She could repeat only 3 monosyllabic words. On the Token Test she could perform only 6 items in the first series. She was able to read aloud only some letters. At that time writing to dictation

could not be examined because of an intravenous infusion in the right arm. The convulsions mentioned above were followed by changes in her behaviour: she showed depressive signs. Inexplicably, the aphasia deteriorated dramatically. The child spoke effortful only simple words in a telegraphic style. She could understand only very simple commands. Repeating words and detailed complementary language examination (Token Test, picture naming) were impossible. After being trained for 6 months by a remedial teacher, she could count from 1 to 10, but she could not read the figures. During the next 2 years virtually no improvement was observed.

Patient 2, Born 30.01.1972.

On June 18th 1982, this patient was hit by a car. She did not lose consciousness. Immediately after the accident she showed speech difficulties.



a *b*

Figure 2a *Patient 2. Computed tomographic scan at onset shows a left temporal depressed fracture, a subgaleal haematoma in the vicinity of the fracture, and a mixed density contusion in the left temporal lobe posterior to the sylvian fissure.*

Figure 2b *Scan 2 months after onset, showing local atrophy in the posterior part of the left temporal lobe.*

On examination, she was a bright girl who seemed to be dysphasic. A large subcutaneous haematoma was palpable at the left temple. Apart from a slight right-sided supranuclear facial paralysis, no other neurological signs were found. CT scan showed a left temporal impression fracture, a subgaleal haematoma in the vicinity of the fracture and a mixed density lesion in the left temporal lobe, situated just underneath the fracture, indicating a mixed density contusion (Figure 2a).
Six days after the accident surgical correction of the impression fracture was performed. The postoperative course was uneventful and 2 weeks after the accident she could be discharged from the hospital with only minimal dysphasic speech. About 2 months after the accident a second CT was made. Now an area of low attenuation was seen in the left posterior temporal region indicating a local atrophy as a result of the contusion (Figure 2b).

Language testing

Detailed language examinations were carried out on the 3rd and the 13th day post onset. In her spontaneous speech she produced many paraphasias. Her speech rate was 96 words per minute. Occasionally, her articulation sounded a little slurry. However, during the greater part of the interview her articulation and prosody were normal. In picture naming she frequently produced phonemic paraphasias. The repetition of polysyllabic nouns was impossible as was the repetition of 3 word sentences. The comprehension of spoken language during the interview was considered normal. In contrast, she obtained a low score on the Token Test (28/61). Her writing to dictation showed many mistakes, some of them reflected paraphasias. Six days after craniotomy her speech rapidly improved. Thirteen days post onset she rarely produced paraphasias in her spontaneous speech and picture naming. However, the recovery of repeating polysyllabic nouns and sentences was less pronounced. The Token Test score was 38/61, demonstrating a receptive disturbance. Reading aloud and writing to dictation demonstrated fewer errors. On the Coloured Progressive Matrices she obtained a score in the 90th percentile.
Two and a half months after the trauma the girl was reexamined. Spontaneous speech was essentially normal, picture naming and repetition of words and sentences were adequate. The Token Test score (54/61) indicated a good verbal comprehension. Reading aloud was performed rapidly without errors. Writing to dictation showed some spelling errors.
Six months after this final examination her tutor reported that there were still difficulties in writing to dictation, especially was there substition of phonemes. This deficit disappeared within the following year.

Patient 3, Born 24.09.1969.

On October 30th 1980 this girl had an accident whilst cycling. She was unconscious for some minutes at most. After being brought home she talked confusedly and was at times somnolent. One day later she occasionally vomited and seemed drowsier. She was then admitted to our department.

On examination, her consciousness was clear. She seemed to give inadequate answers to questions and did not seem to understand instructions well. The neurological examination showed only a slight asymmetry of the reflexes, the right knee and ankle jerks were brisker than those on the left side. During the first three days after admission her behaviour seemed "confused", i.e. dysphasic. On the 4th day she had a focal convulsion with a post-ictal right-sided hemiparesis and hemi-inattention lasting for several hours. Soon thereafter, a spontaneous recovery was observed, and 12 days after the accident she was discharged with only slight wordfinding difficulties. A CT performed 5 days after the accident showed a small area of mixed density (pepper and salt appearance) in the left posterior temporal region (Figure 3).

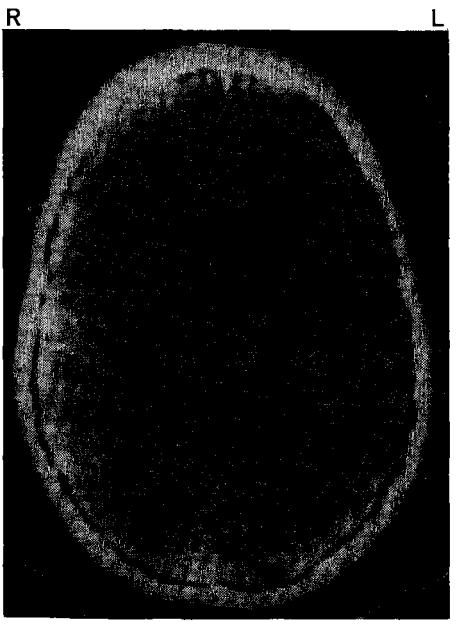


Figure 3 *Patient 3. Computed tomographic scan five days after onset, demonstrating a small, mixed density contusion (arrows) in the left temporal lobe behind the sylvian fissure.*

Language testing

Detailed language assessment was carried out on the 4th, 7th, 10th, 12th and 34th days post onset.

On the 4th day she spontaneously spoke very rapidly: 135 words per minute. Paraphasias frequently occurred, neologisms less often. The speech impressed as empty. Articulation and prosody were normal. She was able to name 6 out of 40 pictures correctly. The high speech rate in the extensive descriptions on this test was striking. Many neologisms were produced. She could only repeat 2 monosyllabic words. Reading aloud was unintelligible on account of frequently occurring neologisms. She was capable of writing only a few simple words. Understanding of verbal commands was considerably limited as reflected in the Token Test score of 13/61. On the Coloured Progressive Matrices Test she obtained a score within the 75th percentile.

On the 7th day the speech rate was 88 words per minute. The number of paraphasias and neologisms had decreased. However, the speech was still empty, and the range of information which could be exchanged was very limited. Moreover, the greater part of polysyllabic words was repeated incorrectly. In picture naming she had a score of 20/40. The Token Test score was 16/61.

On the 10th day, the Token Test score had considerably improved: 35/61. She was now able to read a text aloud. Only complex polysyllabic words gave marked difficulties.

At discharge, 12 days post onset, she had very mild difficulties in understanding a simple conversation in a one-to-one setting. The speech rate was 79 words per minute. Paraphasias and neologisms did not occur, but mild wordfinding difficulties were still present. In the repetition of words, a considerable progress was established. She made only 2 errors. However, she still failed in repeating 5-word sentences. Verbal comprehension had become normal as reflected in a Token Test score of 52/61.

Thirty-four days post onset, the girl was re-examined. Her spontaneous speech had considerably improved and there were no significant limitations in exchanging information. Occasionally, mild wordfinding difficulties occurred. No aphasic errors were noted in repeating words and sentences, picture naming, and Token Test (55/61). Reading was normal.

One year after the trauma, the tutor reported normal school performances with the exception of writing to dictation.

Patient 4, Born 18.02.1965.

Because of renal insufficiency this girl was intermittently haemodialysed from August 1974. On November 13th 1975, she was admitted with headache, vomiting, and restlessness at night. Apart from hypertension with diastolic blood pressures

mostly of 120mm Hg or more, the general examination revealed no abnormalities. It was hardly possible to control the hypertension with drugs. In the night from January 12 to 13 1976 she went into a hypertensive crisis (systolic blood pressure of 290 Hg and diastolic pressure of 190 mm Hg) followed by coma. Neurological examination revealed nuchal rigidity, papiloedema and bilateral extensor plantar responses.
The EEG showed severe diffuse disturbances and a more serious involvement of the left temporal lobe compared to the right. It was concluded that she suffered from brain swelling, and she was treated with mannitol, fluid restriction and anti-hypertensive drugs. In the following days she recovered gradually.
Neurological re-evaluation on January 30th 1976 demonstrated a slight right hemiparesis, a right homonymous hemianopia and dysphasia. Cerebral scintigraphy on March 9th 1976 showed a small zone of increased uptake of Technetium in the paramedian part of the left temporo-occipital region.
In September 1976 she underwent renal transplantation.

Language testing

Two months after the onset of the neurological signs (12.01.1976) a global assessment of the language functions was performed. The child was very inattentive. Moreover, there was a minimal comprehension of spoken language during the interview; echolalia was present. Incidentally in her spontaneous speech simple utterances were present. Picture naming and writing were impossible. In contrast she could repeat and read simple words.
One month after the first language evaluation (02.02.1976) there was a considerable improvement of her speech functions. Her speech rate was 82 words per minute, despite the fact that many pauses were present as a consequence of severe word-finding difficulties. The wordfinding process was usually ended with stereotype remarks as "do know it very well, but I cannot say the correct word". Phonemic paraphasias occurred rarely. Prosody and articulation were normal. Naming to visual confrontation was severely disturbed (5/13) with phonemic paraphasias and neologisms. She was able to repeat 3 syllabic words and 3-word sentences. On the Token Test she could perform 21/61 items which indicated considerable difficulty in comprehending sentences. Writing to dictation showed many errors, reading aloud was hampered by visual neglect due to a right homonymous hemianopia.
Six months post stroke spontaneous speech had improved. Her speech rate was 112 words per minute. She produced utterances of 15 words. Incidentally very mild wordfinding difficulties occurred. In accordance, she completed the picture naming test (one error). Repetition of (5 syllabic) words was perfect; however, repeating of sentences seemed a difficult task to her. She reproduced these sentences using a very

simple structure. On the Token Test, she obtained a score of 45, demonstrating a recovery of language understanding. Reading aloud and writing to dictation were still severely disturbed.
One year post stroke, the child was referred to a school for special education. Language examination revealed a great improvement. Her speech rate was 115, no aphasic errors were noted. Her achievements in the picture naming test as well as repeating of words were normal. However, she still failed in repeating sentences. The Token Test score was nearly normal.
Finally, her teacher reported mild difficulties in writing but severe reading problems.

Patient 5, Born 10.01.1974.

In November 1978 this boy had a haemophilus influenzae meningitis. Initially, he had focal convulsions on the right part of the body. He was unconscious during 8 days. When he regained consciousness, a right hemiparesis and dysphasia were present. Moreover, the right blink reflex was negative and the right corneal reflex was reduced. CT scanning was done one week after the onset of the illness. Almost the whole left hemisphere was hypodense with displacement of the median structures to the right. Moreover, on the right side hypodensity was observed around the ventricles. Cerebral angiography showed a pronounced capillary pattern in the distribution of both the left and the right carotid arteries. A second CT scan was made 2 months post onset and showed widening of the left lateral ventricle (see case 16, chapter 4).
The boy was dismissed 3 months after the onset of the illness with a right hemiparesis, a right homonymous hemianopia and aphasia. In the following years the right arm remained clumsy and became atrophic. In addition, prescription of anticonvulsants was necessary because of psychomotor epilepsy.

Language testing

Language testing in this patient, a right-handed boy, was done for the first time 12 days after the onset of the illness. Initially, the boy did not speak at all. However, showing him a picture book, some pictures elicited a stream of unintelligible sounds. Later on, he incidentally used a meaningful word, but he still jabbered when reacting to verbal commands or objects shown to him. His speech appeared to be phonemic jargon aphasia. During his stay in the hospital, an attempt was made to administer a picture naming test, a repetition test and the Tridimensional Matrix Test (Kreindler *et al.* 1971), a simple test sensitive to receptive verbal disorders. However, he failed completely.
After discharge, he was referred to a Rehabilitation Centre for Children. An intensive therapeutic program was designed for him. The speech therapist reported a very slow progress of the speech functions interfered by behaviour problems.

One year after discharge, a complete language examination was still impossible as a consequence of incooperative behaviour. However, in his spontaneous speech meaningful words occurred frequently. Moreover, the TMT could be administered: the low score on this test reflected his comprehension deficit.
Five years after the onset of the aphasia, detailed language examination could be carried out. His speech rate was 82 words per minute; paraphasias nor neologisms were present, while articulation and prosody were normal. He was able to name 28 out of 40 pictures correctly, indicating moderate wordfinding difficulties. In this test some semantic paraphasias occurred. Repeating of 3 syllabic words was possible. However, he failed completely in repeating simple sentences (3 words). On the Token Test he obtained a score of 19, showing that his understanding of more complex language information had not recovered. His tutor reported that his reading and writing skills were below the level of a 9-year old child.

Discussion

First of all, these 5 cases of childhood aphasia show a great variety of linguistic features and demonstrate the need for a modification of the current clinical picture of childhood aphasia as characteristically non-fluent.
Secondly, these 5 cases are the first extensive reports on children with features of fluent aphasia. Fluent (paraphasic) speech does occur in children, although we agree that the greater part of aphasias in children is of the non-fluent type. The fact that in our series 5/33 have an aphasia with fluent characteristics is in contrast with the reported rarity as mentioned by Lenneberg (1967): "The so-called fluent aphasia is rare or perhaps altogether absent among pediatric patients". This discrepancy can perhaps be explained as follows:

A. In studies of acquired aphasia in children the suspicion of aphasia rose when a right-sided paresis or paralysis was found on examination. In most of these cases an anterior lesion was probably present causing a non-fluent aphasia. Moreover, a selection of aphasic children on the basis of the presence of a hemisyndrome excludes the syndrome of acquired aphasia and convulsive disorder. It has to be noted that in the original discription of this syndrome, jargon aphasia is mentioned (Landau and Kleffner, 1957). Later, observations concerning fluency and neologisms were described (Vande Sandt-Koenderman *et al.* 1984).
B. If a (head) trauma is the underlying cause of aphasia, recovery may be observed within a short time. Two of the cases reported here (2 and 3) demonstrate this as well as other studies (Lange-Cosack and Tepfner, 1973). The fluent characteristics of the aphasia may not be recognized as such.
C. Fluent aphasia can be misinterpreted by considering it as (post-contusional) confusion (see patient 3). The occurrence of paraphasias and neologisms are

indications of the presence of language disturbances. Moreover, the cooperative behaviour of the children easily allows neuropsychological examination.

D. Probably, the introduction of computer tomography plays a role in the early detection of aphasias without other neurological abnormalities. The threshold to perform this non-invasive examination is much lower than for the performance of angiography, pneumencephalography or nuclear brain scanning. Moreover, lesions as contusion or infarction are far more easily demonstrated on a computer tomogram. This could focus the attention of the neurologist on the possibility of fluent aphasia not yet detected or interpreted as such before CT. Concerning the relation between the site of the lesion and the type of aphasia in the patients reported here, 3 of them had a posterior lesion, i.e. in the Wernicke area (see Figure 4).

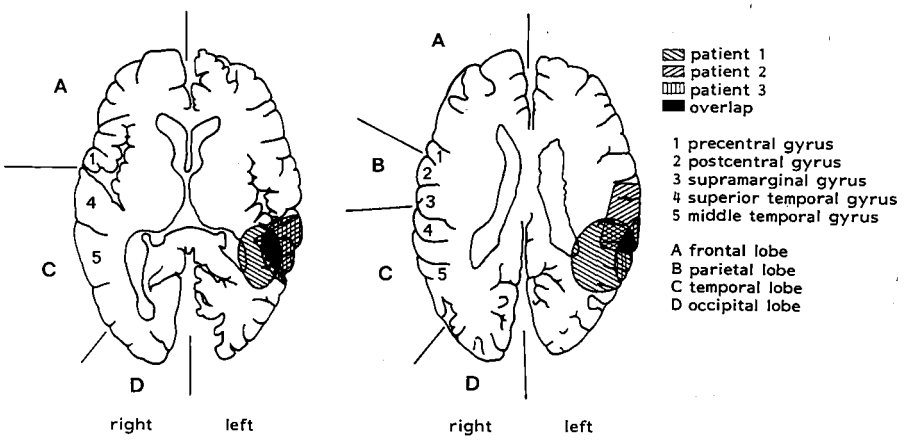


Figure 4 *Lesions evident on computed tomographic scanning at onset in patients 1, 2 and 3. The sylvian fissure was identified using the anatomical studies of Matsui and Hirano (1978) and of Gado et al. (1979) and taking into account slight differences in the plane of orientation.*

One has to be careful about the interpretation of this figure as it suggests that many gyri may be recognized on a computer tomogram. This will not always be possible, and certainly not with the accuracy as shown in the pictures. In most cases the sylvian fissure can be indentified and thus the site of the lesion in relation to this fissure. The relation between the site of the lesion and type of the aphasia seems not to be absolute, that is to say: anterior lesions do not invariably lead to non-fluent aphasia nor do posterior lesions always cause fluent aphasia. We have examined one child with non-fluent aphasia due to a

glioma in the posterior part of the left temporal lobe. In case 5 the phonematic jargon was caused by an extensive infarction in the left hemisphere both anteriorly and posteriorly to the sylvian fissure. In the greater part of the 33 children with acquired aphasia the language disturbances were caused by a large lesion in the left hemisphere, and 28 of them showed, as already mentioned, a non-fluent type of aphasia.

E. The lesion volume was not measured exactly in our patients. There seems to exist some, but no strict, correlation between the volume of the lesion and the prognosis. Knopman *et al.* (1983) found a poor prognosis in adult stroke patients when the lesion was larger than 100 cm^3, but there were cases with small lesions and persisting aphasia. In the patients reported here, the 2 patients with a relatively small lesion caused by head trauma showed a much better recovery than the patients with the larger lesions due to an intracerebral haematoma and postinfectuous cerebral atrophy respectively. In this respect, the series is too small to draw definite conclusions.

Finally, some studies suggested that the clinical picture of acquired aphasia is age-dependent. Geschwind (1974) for instance remarks: "That perhaps the child's Broca area has not adequate practice in language to run as freely as that of adults". This is also emphasized by Lenneberg (1967), who refers to the relation between the level of language development and the type of aphasia. From our series it cannot be analysed to what extent age is an important factor for the development of fluent aphasia since the number of children aged 3 - 5 years is too small. Nevertheless, there is now growing evidence that the classical description of childhood aphasia, that is the clinical picture of the non-fluent child, needs modification.

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CHAPTER 3

NAMING IN APHASIC CHILDREN; ANALYSIS OF PARAPHASIC ERRORS

Introduction

As stated elsewhere (chapter 2) it is generally assumed that aphasic children have primarily expressive difficulties and articulatory problems; they make syntactic errors and speak in a telegraphic style (Ludlow, 1980).
In addition, neologisms are reported not to be present and other paraphasic distortions, especially semantic paraphasias, very rarely occur (Alajouanine and Lhermitte, 1965; Gloning and Hift, 1970; Brown, 1976; Hécaen, 1976, 1983). It is not specified whether these statements are limited to spontaneous speech, or apply to other expressive language modalities such as naming, repetition and reading.
In the last decade this "traditional" picture of childhood aphasia has been considerably challenged as follows:

a. fluent aphasia in children has been observed (Woods and Teuber, 1978; Martins and Ferro, 1982) and extensively described (VanDongen *et al.* 1985; VanHout *et al.* 1985).
b. paraphasias and neologisms are noted not only in the spontaneous speech of children with fluent aphasia but also in children with non-fluent aphasia (Dugas *et al.* 1982; Visch-Brink and VandeSandt-Koenderman, 1984).

The occurrence of neologisms might be associated with the presence of an acute process in the brain (VandeSandt-Koenderman *et al.* 1984). Visch-Brink and VandeSandt-Koenderman (1984) reported a rapid disappearance of neologisms in the spontaneous speech of 2 aphasic children (within 2 days and 6 weeks, respectively, post onset), and conclude that a knowledge of the degree of contextual appropriateness is crucial for a differentiation between neologisms and literal paraphasias.
More formal testing of picture naming, for example via the Boston Naming Test (Kaplan *et al.* 1978), allows a qualitative analysis of naming errors (Knopman *et al.* 1984). This is an important point, as, despite the report that naming disorders frequently occur in childhood aphasia (Hécaen, 1983) such a qualitative analysis of errors is so far not available.

The purpose of the present study was therefore:

1. to analyze incorrect naming responses of aphasic children, focusing on paraphasias, especially neologisms.
2. to examine the distribution of paraphasic errors during the course of aphasia.

Methods

Subjects (Table 1).

We studied the recovery of naming in 6 right-handed children with acquired aphasia. The aphasia examination started, when the children had fully recovered consciousness. Spontaneous speech was - rated on the severity rating scale of the BDAE (Goodglass and Kaplan, 1972) and - classified as fluent or non-fluent, following the criteria of Kerschensteiner *et al.* 1972 (for a more extensive description see VanDongen *et al.* 1985). None of the children demonstrated articulatory disturbances, and no visual abnormalities were present.
In addition, we assessed the linguistic performance in terms of spontaneous speech, naming and Token Test (VanDongen *et al.* 1974), of 3 children with right hemisphere lesions. Before admission to our clinic they had, as had the aphasics, normal school performances.

Table 1 Patient data and linguistic performance at the 1th (completely) performed naming test.

Case	Age	Sex	Etiology	CT localization	Naming test (correct score)	Tokentest (correct score)	Severity (BDAE)	Fluency
1	9	F	head injury	L temporal	15	38	3	F
2	7	M	head injury	L fronto-temporal	20	13	2	NF
3	11	F	head injury	L temporal	8	7	2	F
4	9	M	subdural empyema	L frontal	24	12	1	NF
5	6	F	CVA	L temporal	17	28	1	NF
6	11	M	herpes simplex encephalitis	L fronto-temporal	17	41	3	NF
7	8	F	astrocytoma	R parietal	37	52	-	-
8	9	M	arteriovenous malformation	R frontal	35	48	-	-
9	12	F	intracerebral haematoma	R temporal	39	61	-	-

Procedure

A picture naming test - a modification of the Boston Naming Test consisting of 40 individual line drawings (20x15 cm) representing 1 to 5 syllabic nouns - was administered. Age norms are available for 60 children in the ages of 6 to 13 years attending primary school (Immerzeel, 1985).
The children's speech production was tape-recorded. Only spontaneously given responses were considered to be suitable for analysis. Paraphasic errors were categorized as neologisms, and verbal and literal paraphasias, according to the criteria of Butterworth *et al.* (1984). Other erroneous responses were "circumlocutory" or

"don't know". The data are summarized in Figure 1.
The naming test was repeated on a number of occasions during the days respectively months of recovery in the aphasic children.

Results

A. Aphasic children

The acquired lesions were the result of mild head injury (length of coma <1 hour), cerebrovascular accident, subdural empyema and encephalitis (Table 1). In the initial stage the children appeared to have a severe naming disorder. However, there was a marked difference in the course and recovery of naming between the head-injured and the non-head-injured children.

1. Head-injured children

The head-injured children could be investigated within 3 to 7 days after admission; neologisms, and verbal and literal paraphasias were present in different proportions. The main finding was that neologisms were always present when these children completed the naming test. Moreover, in every succeeding examination there was a decrease of neologisms, which did not appear to apply to the literal and verbal paraphasias (Figure 1). The average decrease in the number of neologisms was analyzed by linear regression showing a decrease of 1,3 ($p=0.002$) for case 1, a decrease of 0,6 ($p=0.06$) for case 2 and a decrease of 0,6 ($p=0.02$) for case 3. All errors disappeared very rapidly. After 6 months, recovery was complete and school achievements were within normal limits. This is in accordance with previous studies, which have shown that traumatic aphasia has a good prognosis (Guttmann, 1942; VanDongen and Loonen, 1977).

2. Non-head-injured children

The non-head-injured children differed in many respects: none of the children could complete the naming test within one month, indicating a more severe aphasia than the head injury group. Case 6 was not able to complete the naming test within one year. Despite the severity of the aphasia, neologisms, and verbal and literal paraphasias occurred less often. Case 5 never used a neologism. In contrast to the head-injured group, the distribution of the error types was irregular over time. Recovery was not complete even after one year. Cases 5 and 6 were referred to a rehabilitation centre for the young where they receive speech therapy.

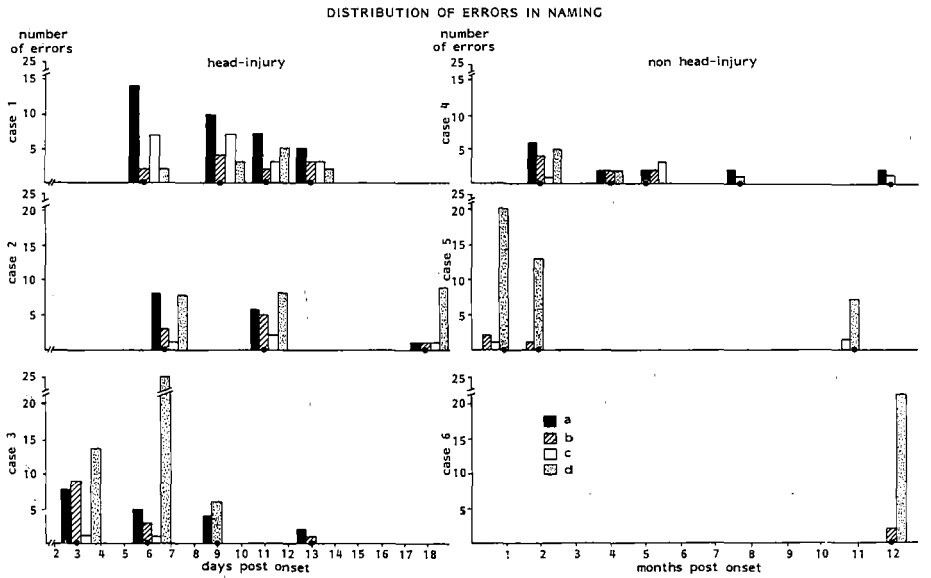

Figure 1 *Distribution of errors in naming of 6 aphasic children.*
a. neologism
b. verbal paraphasia
c. literal paraphasia
d. other erroneous response
•. day of examination

B. Children with right hemisphere lesion

The results of the children with a right hemisphere lesion on the Token Test and the naming test were within normal limits. It has to be emphasized that, as in the normal group, neither neologisms nor other paraphasias occurred. Analysis of the spontaneous speech revealed no disturbances.

Conclusions

In order to describe more completely the features of acquired childhood aphasia, the clinical picture demanded detailed qualitative analysis to determine the relevance of neologisms. Their frequency can be modality-specific (VanHout *et al.* 1985). Consequently, their true significance among other aphasic symptoms can only be assessed after an analysis of paraphasic errors in different modalities. In our study, a comparison of the patterns of errors in spontaneous speech and naming, did not

suggest a common pattern across modalities. For example, the spontaneous speech of case 2 in contrast to that of case 1 and 3 demonstrated more neologistic errors than the naming task.

The naming results of the head-injured children did not indicate an increase of literal paraphasias with a decrease of neologisms as the conduction hypothesis suggests (Kertesz and Benson, 1970): neologisms result from severe phonological distortions of the target word.

Finally, the relationship of neologisms to the severity of aphasia, and locus and size of the lesion could not be evaluated, as our study was limited to a small number of children. The data nevertheless underline the importance of aetiology in that a specific course of recovery of neologisms seems to be associated with head injury.

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CHAPTER 4

ACQUIRED CHILDHOOD APHASIA; COURSE AND OUTCOME

Introduction

The standard doctrine about childhood aphasia claims that recovery of language functions is rapid and complete. Kertesz (1985) states that "very few children will persist with a significant aphasia for any length of time". However, this claim rests largely on quotations from the earlier literature (Basser, 1962; Lenneberg, 1967) and is increasingly contradicted by more recent reports (VanDongen and Loonen, 1977; Woods and Carey, 1979; Satz and Bullard-Bates, 1981; VanHout *et al.* 1985; Cooper and Flowers, 1987).
A number of variables may be related to recovery. It is assumed that the main factors that determine language recovery are: age at onset, aetiology, severity and bilaterality of lesion, and type of aphasia.

Age at onset

Alajouanine and Lhermitte (1965) found no difference in "the speed of recovery in children less than 10 years old versus children of 10 or more years old". According to Lenneberg (1967) the prognosis of acquired aphasia in children is directly related to the age at onset of the aphasia. Aphasias that develop before puberty would clear up completely. In the series of Woods and Teuber (1978) all children who became aphasic before the age of 8 years regained speech but recovery time ranged from one month to more than 2 years. In contrast, in the syndrome of acquired aphasia with convulsive disorder (Landau-Kleffner syndrome = LKS) an opposite relationship is found, that is the older the child at onset the better the outcome (Toso *et al.* 1981; Dulac *et al.* 1983; Bishop, 1985).

Aetiology

Children with a head trauma have been reported to improve more than those with vascular disease (Guttmann, 1942; VanDongen and Loonen, 1977). However, Byers and McLean (1962) reported a complete restitution of speech function in 10 aphasic

children with persistent hemiplegia due to a cerebrovascular lesion at follow-up from 1 - 4 years. Many authors present evidence that the prognosis in children with the syndrome of acquired aphasia with convulsive disorder is poor. After a period of years most of them still show language deficits, which appear to be permanent at adult age (VanHarskamp *et al.* 1978; Mantovani and Landau, 1980; Dugas *et al.* 1982).

Severity and bilaterality of the lesion

Persistent aphasic symptoms have been linked with the severity of the lesion (Guttmann, 1942; Gloning and Hift, 1970; Hécaen, 1983). It has to be emphasized, however, that before cerebral computerized tomography (CT) was available, assessment of the size and location of the lesion remained rather imprecise. Consequently, statements before this period suggesting a relation between the size of the lesion and recovery are of a questionable value. Until now, CT scan data have been reported especially in case studies (Dennis, 1980; Aram *et al.* 1983; VanDongen *et al.* 1985; VanHout *et al.* 1985). In 4 patients, the abnormalities found on CT were described extensively because they were the first reports of children with a subcortical aphasia (Ferro *et al.* 1982; Aram *et al.* 1983), a crossed aphasia (Martins *et al.* 1987), and a conduction aphasia (Martins and Ferro, 1987). In children with acquired aphasia with convulsive disorder normal CT scans are consistently reported despite severe aphasic disturbances (for refs. see Beaumanoir, 1985).

In a group of children clinically classified as aphasic, the effect of the size of a cerebral lesion on the course and the outcome of language functions has not been investigated systematically. In the series of Vargha-Khadem *et al.* (1985), only one of the 10 children with a left hemisphere lesion and hemiparesis was clinically classified as dysphasic. The authors report an absence of a significant relation between severity of cerebral damage and the level of language performance.

The site of the lesion, verified by the localization of depressed fractures of the skull or craniotomy (Guttmann, 1942), is considered as less relevant to recovery. Studying the incidence of paraphasias, VanHout *et al.* (1985) mention CT scan abnormalities, but do not assess the severity of the lesion in 11 children who demonstrated an aphasia acquired by diverse aetiologies.

Cranberg *et al.* (1987) reported 8 children of traumatic and vascular aetiology with acquired aphasia aged 5 - 17 years. Seven of these children showed a non-fluent aphasia and recovered over weeks to months, regardless of the site of the lesion on the CT scan. The effect of the severity of the lesion on the outcome is not discussed by the authors.

It is claimed that if in childhood the left hemisphere is damaged, language can develop in the corresponding area of the right hemisphere. Consequently bilateral

cerebral damage is considered a bad prognostic sign (Collignon *et al.* 1968; Gloning and Hift, 1970; Hécaen, 1983).

Type of aphasia.

Guttmann (1942) emphasized the good prognosis of purely motor (non-fluent) aphasia in young children: combined motor and sensory aphasia had a more serious prognosis. Assal and Campiche (1973) confirmed, but Collignon *et al.* (1968) contradicted this finding. Fluent aphasia in children has been considered to be rare. This can be explained because in the majority of studies the aphasic children were selected according to the presence of a hemiparesis. In most of these cases an anterior lesion was probably present causing a non-fluent aphasia.

The intent of the present study is to investigate the contribution of the variables age at onset, aetiology of the lesion, severity and bilaterality of the lesion, and type of aphasia to the recovery of acquired childhood aphasia.
Assuming a rapid initial recovery, language functions of the children were first assessed at one year post onset. An evaluation of the final recovery of these aphasic children was then made after variable follow-up periods.

Methods

Subjects

In the period 1977-1985 we studied 30 children with acquired aphasia. They were referred to the Department of Neurology, University Hospital Rotterdam-Dijkzigt. CT scan data were available. With the exception of 2 children*, one or more follow-up examinations were carried out in all children who were not completely recovered within one year post onset (Table 1).

* case 12: re-examination could be performed only one month and 5 years post onset.
case 13: re-examination was refused by the child's parents.

Table 1 Age at onset, aetiology of cerebral lesions and clinical findings, CT scan data and Token Test/TMT scores.

case no/ sex	age at onset**	aetiological factors	severity of cerebral lesion	Token Test/ TMT* score at onset
1 M	9.1	contusion; unconscious for 15 min.	N	17
2 F	11.1	contusion; unconscious for 5 min.	NS	13
3 M	3.9	contusion; unconscious for one month; left temporal depressed fracture; persisting right hemiparesis	S	2*
4 M	9.3	contusion; unconscious for 5 days; transitory right hemiparesis	N	22
5 F	10.5	left temporal depressed fracture and local contusion	S	28
6 M	7.3	multiple skull fractures; left parietal depressed fracture; transitory slight right hemiparesis	S	39
7 M	7.6	left temporal contusion; unconscious for some days	S	13
8 M	7.2	contusion; unconscious for one month	S	0*
9 F	9.2	left parieto-occipital intracerebral haematoma; persisting right hemi-paresis secondary to aplastic anaemia	S R+	6
10 F	7.0	atonic attacks from the age of 4 yrs. At 7 yrs. loss of speech and choreiform movements secondary to "Moya-Moya" disease.	NS	26
11 M	13.4	persisting right hemi-paresis secondary to left internal carotid artery occlusion of unknown cause	S R+	39
12 F	8.8	transitory slight right hemiparesis secondary to left frontal sub-cortical haematoma	NS R+	28
13 M	3.5	transitory right hemiparesis following embolism in left middle cerebral artery	S	0*
14 F	6.7	persisting right hemiparesis secondary to left middle cerebral artery infarction of unknown cause	S R+	8

15	M	3.5	transitory right hemiparesis following cerebral infarction in internal capsule and caudate nucleus secondary to "sick sinus" syndrome	S	18*
16	M	4.10	persisting slight right hemiparesis following haemophilus influenzae meningo-encephalitis; residual epilepsy	S R+	0*
17	M	12.7	left frontal and temporo-basal subdural empyema; residual epilepsy	S R+	21
18	M	11.2	transitory right hemiparesis secondary to herpes simplex encephalitis	S	2
19	M	9.8	left fronto-temporal subdural empyema secondary to frontal and maxillary sinusitis	S R+	2
20	M	11.4	severe conduct disorder and residual epilepsy following acute demye-linating encephalo-myelitis	NS R+	0
21	F	6.8	myoclonic absences from the age of 5.8 yrs.	N	5
22	F	4.0	"word deafness" after birth of a sister; atypical absences from the age of 4.5 yrs.	N	0*
23	M	6.0	"word deafness" from the age of 4 yrs. several months later followed by atypical absences	N	2
24	M	4.8	slow motor development; complex partial seizures from the age of 4.3 yrs.	N	0*
25	F	5.11	complex partial seizures from the age of 5.6 yrs.	N	7
26	F	4.7	"word deafness" at the age of 3.6 yrs.	N	4*
27	M	13.0	left temporal oligo-dendroglioma	S	40
28	F	13.6	left temporal grade I astrocytoma	S	41
29	F	9.5	large neuroblastoma in left hemisphere	S	38
30	F	13.6	left fronto-temporal metastasis of adrenal carcinoma	S	40

* = TMT score
** = years.months
R+ = including enlargement of right ventricle and/or sulci of the right hemisphere.
N = normal; NS = non-severe; S = severe (according to Vargha-Khadem *et al.* 1985).

CT scans

With a few exceptions, the CT scans were performed with scanners of the second or third generation. In general, scanning was carried out according to standard techniques. The plane of orientation was chosen approximately parallel to the orbitomeatal line. The number of slices taken at each examination varied from 8 to 12. The interpretation of the pictures was done with standardized window setting. Using the rating scale of Vargha-Khadem *et al.* 1985 (see appendix) the severity of the lesion was rated by a child neurologist who was unaware of the results of the neuropsychological examinations. The selected CT scans were performed 4 weeks post onset or later. From each patient with CT scan abnormalities one representative picture is shown in Figures 1-4. The numbers to the left correspond to the case numbers in Table 1.

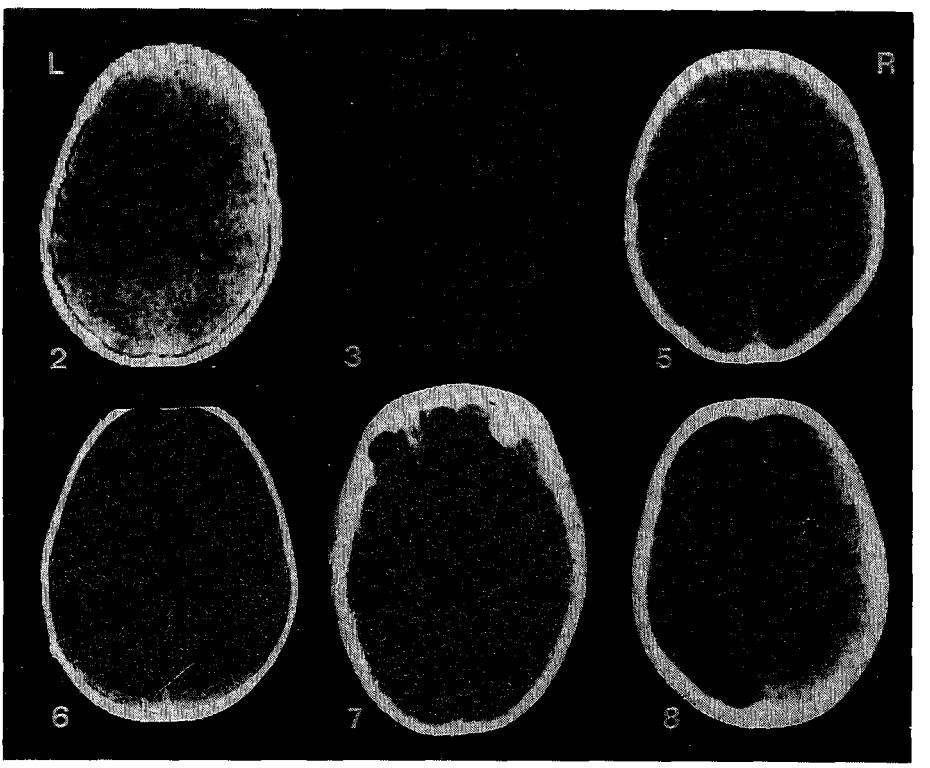

Figure 1 *CT scans of 6 patients with head injury.*

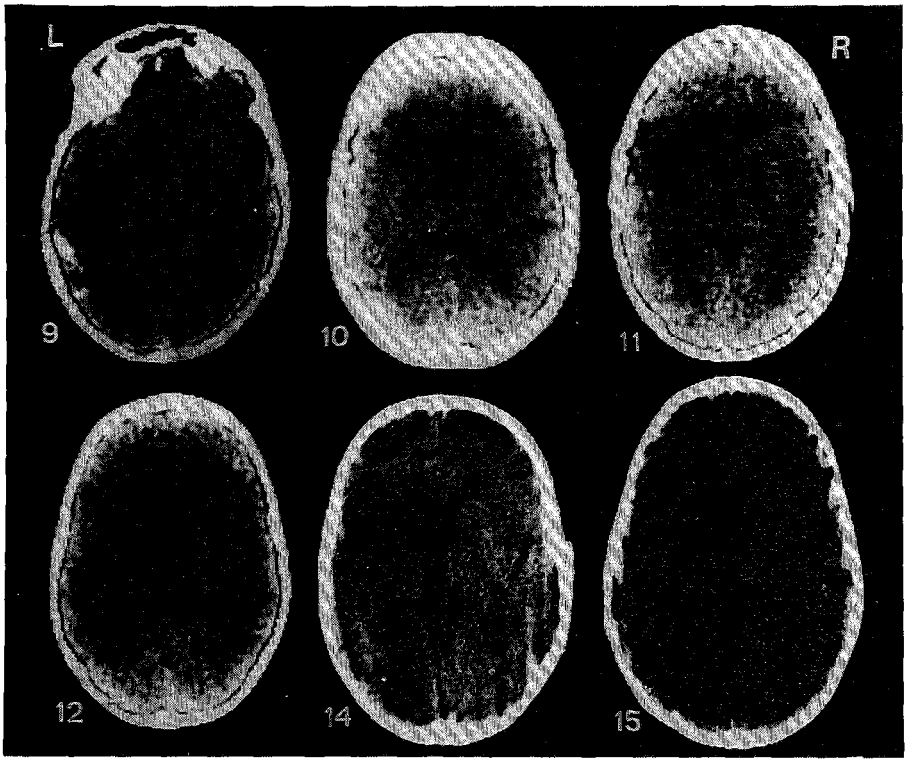

Figure 2 *Ct scans of 6 patients with vascular lesions.*

Language testing

The initial language testing was performed as soon as possible after the onset of the aphasia and follow-up examinations were carried out by the same investigator. Language measures i.e. analysis of spontaneous speech, object naming, repetition and auditory comprehension (Token Test; DeRenzi and Vignolo, 1962) and for toddlers the Tridimensional Matrix Test (Kreindler *et al.* 1971) have been previously published (VanDongen *et al.*1985).

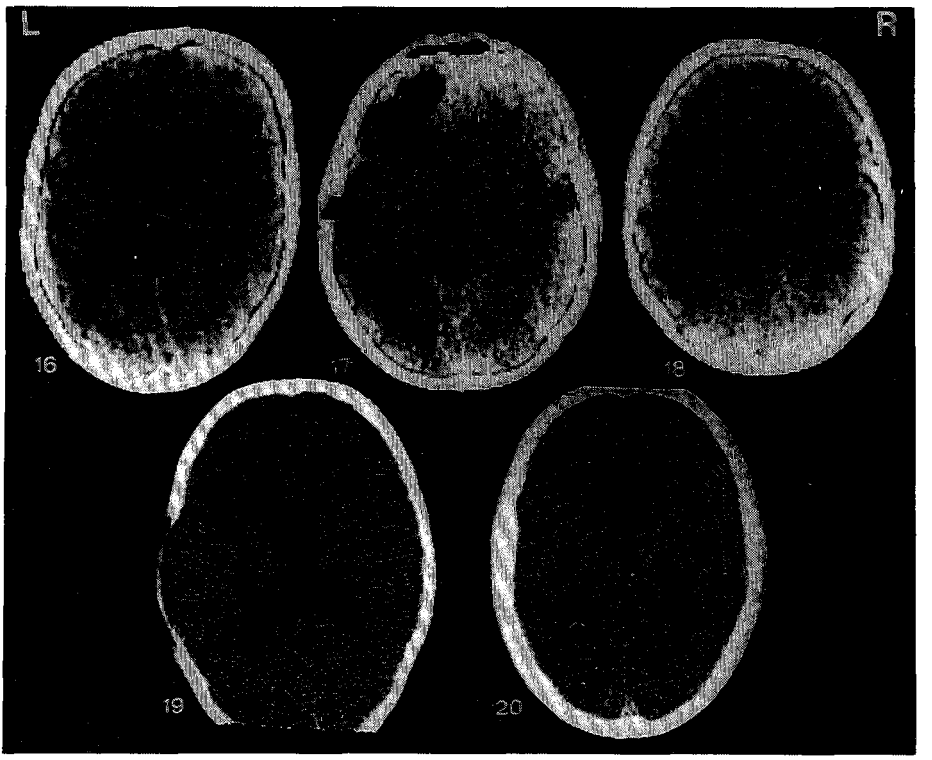

Figure 3 *CT scans of 5 patients with infectious diseases.*

Children were considered as incompletely recovered when:

- aphasic signs were present in spontaneous language: degree 0 - 4 aphasia score on the severity rating scale of Goodglass and Kaplan, 1972 (see appendix).
- the performances on the Token Test or Tridimensional Matrix Test were one or more standard deviations below the average for that child's age (Blauw *et al.* 1987).

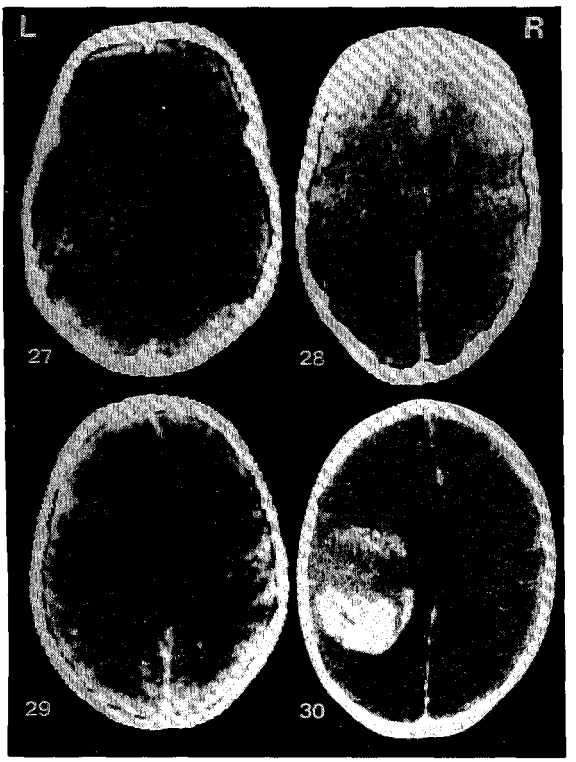

Figure 4 *Ct scans of 4 patients with a cerebral tumour.*

Results

A. Aphasia one year post onset

Analysis of the data (Figure 5) revealed clear differences between the children with a non-progressive lesion and the children with LKS concerning age of onset (range 4 - 6 years) and CT scan ratings (all normal). The 4 children with a cerebral tumour showed a progressive aphasia. They all died about one year post onset. The outcome in the 18 children with head injury, vascular, or infectious disease is therefore reported in Table 2. In view of the small number of patients in the various groups the relation between recovery and the variables "age at onset", "aetiology", "severity of the lesion" and "type of aphasia" could only be evaluated separately. Data of patients with LKS or with a cerebral tumour have been supplemented where relevant.

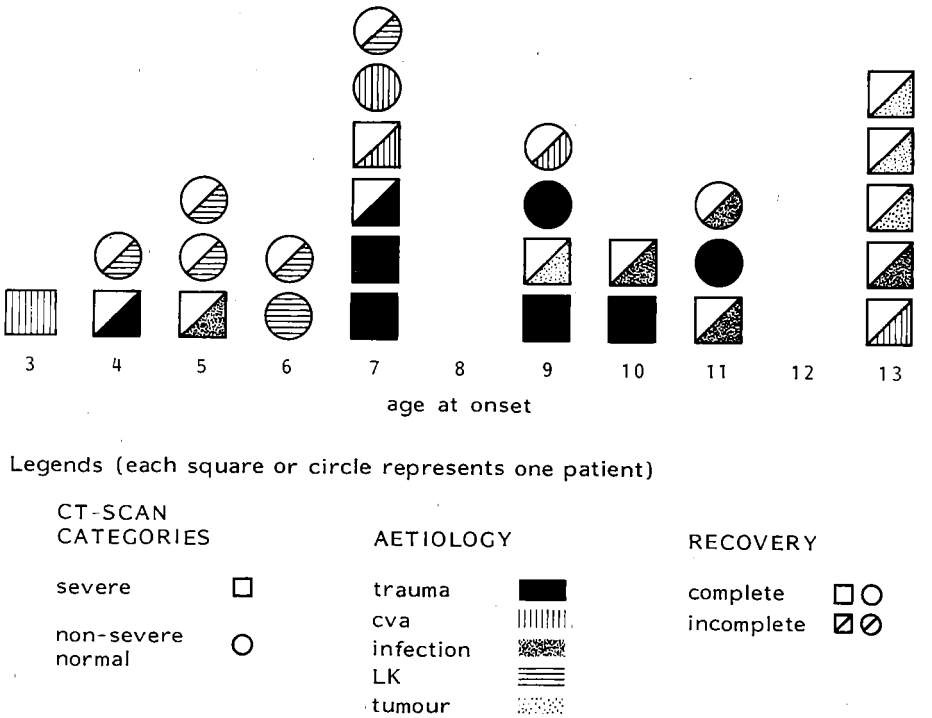

Figure 5 *Severity of the lesions versus outcome one year post-onset in 28 patients.*

The effect of age

The assumption that the earlier in life a lesion is sustained the better the outcome could not be confirmed. Our results indicate that the age limit < 11 years (Lenneberg, 1967) did not differentiate with respect to recovery. The very young children with LKS had a bad prognosis, as in accordance with the literature (Bishop, 1985).

Aetiology

Six of the 8 children with a head injury had a favourable outcome. This was in complete contrast to those with vascular or infectious disease. Furthermore, only one of the 6 children with the LKS recovered completely.

Table 2 Outcome in 18 patients related to age at onset, aetiology, severity of lesion and type of aphasia.

Variables	Recovered	Nonrecovered
Age at onset		
< 11 years	6	7
> 11 years	1	4
*Aetiology**		
head injury	6	2
vascular/infectious disease	1	9
Severity of lesion		
CT severe	4	9
CT non-severe/ normal	3	2
Type of aphasia		
fluent	2	2
non-fluent	5	9

* $p < 0{,}01$ for one tailed Fisher's Exact Test.

Severity and bilaterality of the lesion

In view of the small number of patients, we regrouped them on the basis of severity of lesion. Patients whose CT scans were rated as 1 or 2 comprised the group with a severe lesion and were compared with those patients with a non-severe lesions (categories 3, 4 or 5). Although recovery was frequently observed in children with non-severe/normal CT scans, no level of significance was reached.

Initially all children with a cerebral tumour showed a mild aphasia, although at admission the lesions on CT scan were categorized as severe. However, the aphasic disturbances increased very rapidly during their stay in the hospital. In contrast, all LKS-children demonstrated a severe aphasia at admission despite normal CT scans.

In our series local lesions were limited to the left hemisphere. In addition 7 patients showed a slight enlargement of the right ventricle and/or dilation of the right sulci (Table 1). The 7 children with bilateral hemispheric damage all had a poor outcome. However, this can also be explained by the severity of the lesion. As severity and bilaterality of hemispheric lesion are interrelated, we tested both the effect of severity when correcting for bilaterality, and the effect of bilaterality when correcting for severity (Table 3).

The effect of severity when correcting for bilaterality was not significant (Fisher's Exact Test $p = 0.35$). In contrast, the effect of bilaterality was significant when correcting for severity (Fisher's Exact Test $p = 0.05$).

Table 3 Outcome in 18 patients related to severity and bilaterality of hemispheric lesion.*

Variables	Recovered	Nonrecovered
bilat. hemispheric lesion		
7 (6 S + 1 NS)	0	7
left hemispheric lesion		
11 (7S + 4NS)	6	5
	3S + 3 NS	4S + 1NS

* S and NS: category 1 or 2, respectively 3, 4 or 5 of the rating scale for CT scans (Vargha-Khadem *et al.* 1985).

Type of aphasia

Fluent aphasia was present in 4 children (cases 2, 5, 9 and 16). This type of aphasia did not affect recovery significantly. It seems important that in the 2 recovered children (cases 2 and 5) the aphasia was caused by head injury, an aetiology generally linked to a good prognosis. Conversely an incomplete recovery was present in the 2 children with an unfavourable aetiology i.e. an infectious disease and vascular disease respectively.

B. Aphasia after more than one year (the final outcome)

The results of a follow-up of more than one year were available in 24 children. In view of the variability in age, severity of lesion, and length of follow-up only a general view of the course and outcome can be outlined (Figure 6 and 7).

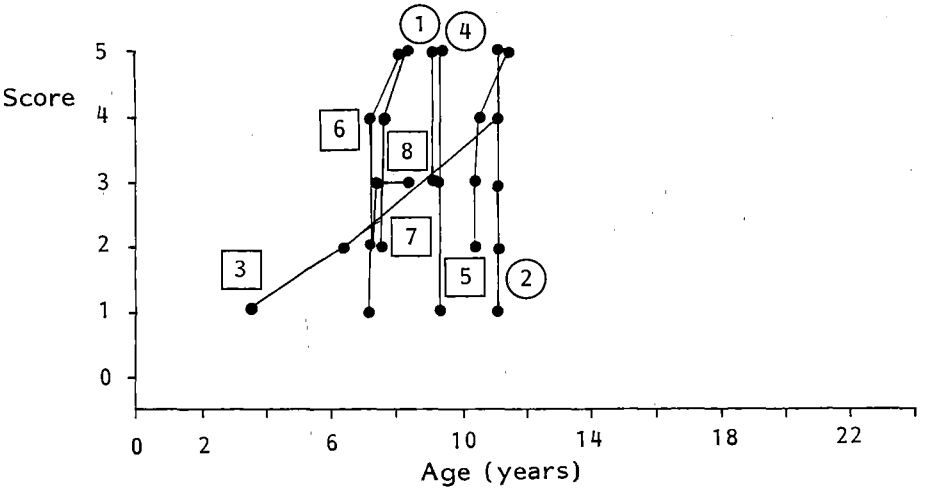

Figure 6a *Course of aphasia in patients with head injury.*

In addition to the above mentioned rapid and favourable recovery, Figure 6a shows a great variety regarding the severity of lesion. Rapid and good recovery appears to be associated with mild cerebral lesions.

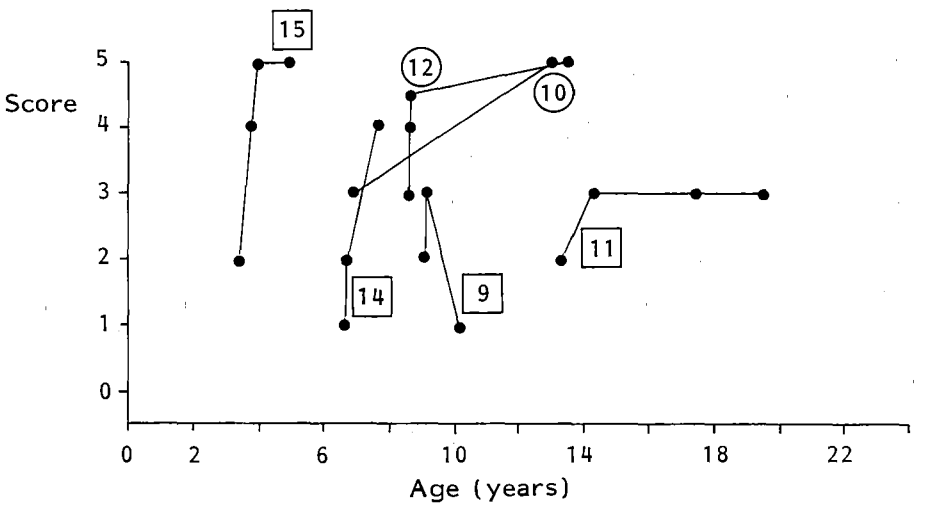

Figure 6b *Course of aphasia in patients with vascular lesions.*

Severe cerebral lesions dominate in this subgroup. Rapid recovery is an exception and is present only in one child who was the youngest one (case 15). Longterm outcome may be very unfavourable as case 11 demonstrates (Figure 6b).

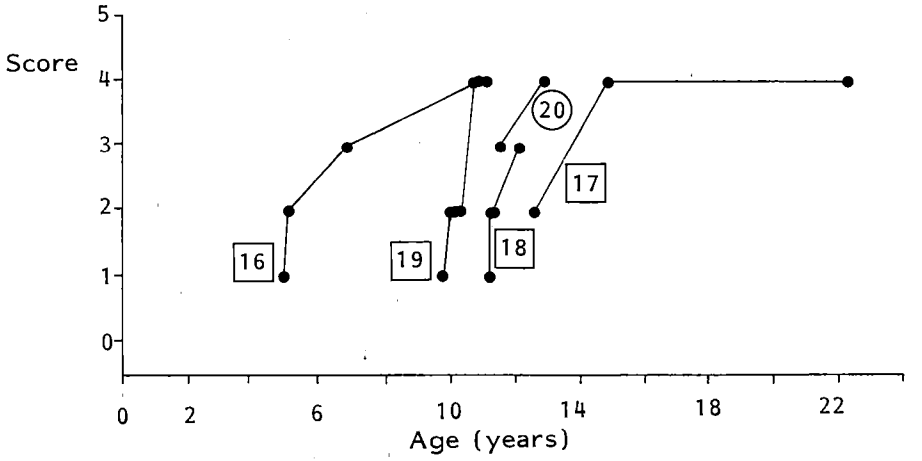

Figure 6c Course of aphasia in patients with infectious disease.

As in the vascular disease group severe cerebral lesions dominate and Figure 6c shows that even in young children the outcome is unfavourable (case 16). Complete recovery did not occur.

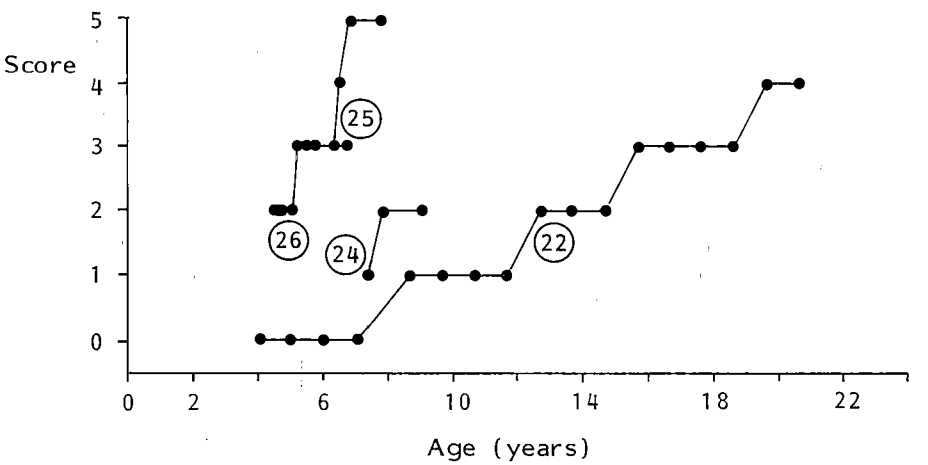

Figure 7a Course of aphasia in 4 patients with LKS; non-remitting course.

Figure 7a demonstrates that despite the normal results of CT scanning, recovery is incomplete even after 17 years follow-up (case 22). Moreover, prediction of outcome is difficult because of the initially fluctuating course of the aphasia with clear

remissions and exacerbations which was observed in 2 of our patients (cases 21 and 23, Figure 7b). Longterm follow-up indicates a slow and consistent recovery process even after recurring aphasia. The age range in this group is small (4-6 years). Consequently the relationship between age at onset of language disorder and final outcome could not be evaluated.

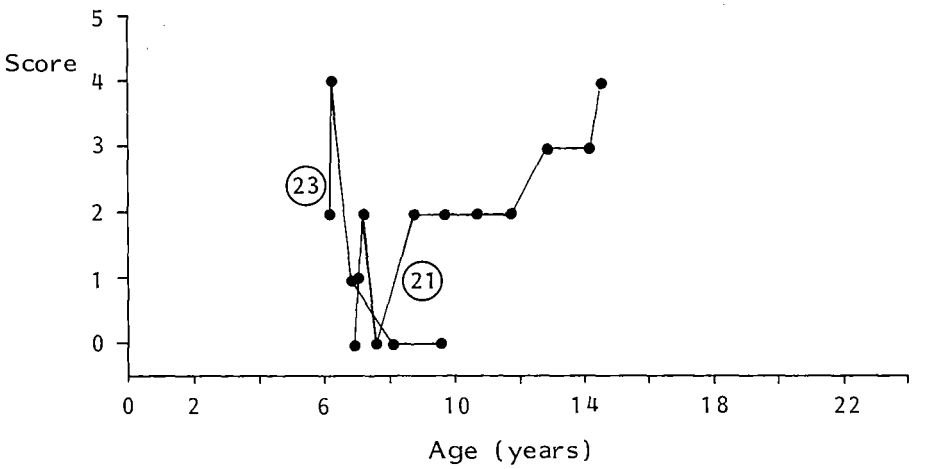

Figure 7b *Course of aphasia in 2 patients with LKS who showed remissions and exacerbations.*

Legends to Figure 6 and 7
Each line represents one patient. Each dot on a line corresponds to an assessment of the language performance.
The ordinate shows the score on the Severity Rating Scale of Goodglass and Kaplan.

○ = *CT scan non-severe/normal (categories 3, 4 or 5, according to Vargha-Khadem et al., 1985).*
□ = *CT scan severe (categories 1 or 2, according to Vargha-Khadem et al. 1985).*

The place of the circles and squares corresponds to the date of the CT scan which was chosen for severity rating.

Discussion

The present study contradicts the common clinical belief that acquired childhood aphasia is ordinarily of short duration: many of the children did not recover completely within one year. Our results demonstrate that with the exception of traumatic aetiology none of the other variables are unequivocally favourable or unfavourable for recovery. Often these variables are interrelated and the outcome will be determined by a complex interaction between a number of factors. These factors are not discriminable at present because the cited studies on childhood aphasia include only small aetiological groups that are not comparable regarding the age of onset and severity of lesions. Despite these limitations, there are some discernible trends regarding outcome. Thus, in the majority of the patients described, one may expect a good count outcome if (mild) head injury is the aetiology. In contrast, a poor outcome is reported in a group limited to infectious disorder (VanHout and Seron, 1983) or the Landau-Kleffner syndrome (Dugas *et al.* 1982).

Our data demonstrate a relation between severity of lesion and aetiology. Normal CT scans were found in all patients with LKS and in 2 patients with head injury. Does this mean that in vascular and infectious disease clinically manifest aphasia is observed only when the lesion is extensive?

The possibility that the intact right hemisphere has contributed to the language recovery has to be considered. However, illustrating the complex interaction of prognostic relevant factors, it seems that bilaterality frequently occurs in infectious and vascular diseases, which are associated with a poor outcome too.

The disagreement in terms of favourable versus unfavourable outcome may be explained by the method of assessment. Usually recovery from aphasia has been estimated in clinical terms for example "readily able to re-engage in free-flowing conversations" (Cranberg *et al.* 1987). It is striking that authors observe complete recovery, or state that the patient had regained normal language or nearly normal language while simultaneously reporting impaired performance on verbal tests. Indeed it has been noted that "diminution of lexical stock is observed in late stages and, even after the children return to school, is mentioned explicitly in school reports" (Hécaen, 1983). In another statement, one reads that "eight years after injury, speech was handicapped by occasional agrammatic language construction and by moderate word-finding difficulty" (Cranberg *et al.* 1987). This discrepancy emphasizes the need for a standardized aphasia examination. Such a procedure would be particularly appropriate in studies of children with left hemisphere lesions without clinically manifest aphasia (Vargha-Khadem *et al.* 1985; Kiessling *et al.* 1983). In such studies, mild verbal deficits in standardized tests are consistently reported even if the lesion is sustained in the early postnatal period.

Another problem in the literature which makes it difficult to predict outcome is the

great variety in length of follow-up in the diverse series of aphasic children reported. The duration of the aphasia in these series has not been related to the various prognostic variables.

In conclusion, despite the fact that this study includes 28 children and is one of the largest series in the literature, no firm prediction about the outcome can be made at present. At best, certain trends can be suggested as our results demonstrate: in contrast to head-injury the other aetiologies are linked to an unfavourable outcome. The same applies to the variable bilaterality of the lesion. The precise weight of the variables severity of the lesion, age at onset and type of aphasia are still unknown.

Appendix

Rating scale for CT scans (Vargha-Khadem *et al.* 1985).

1. Moderate to marked ventricular dilatation and gross loss of substance seen on three or more cuts.
2. Moderate to marked ventricular dilatation with minimal to moderate loss of brain substance seen on less than 3 cuts.
3. Moderate cerebral atrophy indicated by moderate to marked ventricular dilatation seen on 3 or more cuts.
4. Minimal hemisphere atrophy indicated by minimal ventricular dilatation; asymmetry seen on less than 3 cuts.
5. Normal.

Aphasia severity rating scale (Goodglass and Kaplan 1972).

0. No usable speech or auditory comprehension.
1. All communication is through fragmentary expression; great need for inference, questioning and guessing by the listener. The range of information which can be exchanged is limited, and the listener carries the burden of communication.
2. Conversation about familiar topics is possible with help from the listener. There are frequent failures to convey the idea, but the child shares the burden of communication with the examiner.
3. The child can discuss almost all everyday topics with little or no assistance. However, reduction of speech and/or comprehension make conversation about certain material difficult or impossible.
4. Some obvious loss of fluency in speech or facility of comprehension, without significant limitation on ideas expressed or form of expression.
5. Minimal discernible speech handicaps; the child may have subjective difficulties which are not apparent to listener.

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CHAPTER 5

THE LANDAU-KLEFFNER SYNDROME OR "ACQUIRED APHASIA WITH CONVULSIVE DISORDER"

Introduction

In this chapter the literature about the syndrome will be described, as well as our findings in 6 patients and their follow-up.
The Landau-Kleffner syndrome or the syndrome of "acquired aphasia with convulsive disorder" is a rare form (type) of acquired aphasia mostly associated with very heterogeneous epileptic manifestations. The EEG shows bilaterally synchroneous spike-wave discharges, also seen in those children who do not show clinical seizures (Rodriguez and Niedermeyer, 1982).

Table 1 Characteristics of the Landau-Kleffner syndrome.

Initial symptoms:	at onset aphasia, predominantly receptive and mostly epileptic seizures associated with paroxysmal EEG abnormalities occurring in the age group 3-8 years
Secondary symptoms:	behavioral disorders, mild forms of dyspraxia, no signs of intellectual deterioration
Course:	rapid onset and recovery of aphasia or worsening of the aphasia deficit after repeated seizures or progressive aphasic deficit with rare clinical seizures
Treatment:	anticonvulsive medication, cortico-steroids, left temporal lobectomy, speech therapy
Prognosis:	complete recovery in about 30% of the affected patients; recovery probably age related

The syndrome was described by Landau and Kleffner in 1957. This original description concerns 5 cases and can be considered as essential with regard to the clinical picture. In 1980 Mantovani and Landau published a follow-up study of children with this syndrome. In this study the firstly described children were included. In the period 1957-1985 more than 100 similar cases have been reported (Costello and McGee, 1965; Moor House School, 1969; Lanzi and Boiardi, 1970; Calabrese, 1971;

Worster-Drought, 1971; Harms, 1972; Huskisson, 1973; Brissaud and Richardet, 1974; Humphrey *et al.* 1975; Perniola and Pennetta, 1975; Giovanardi Rossi *et al.* 1976; Forster, 1977; Julien *et al.* 1978; Koepp and Lagenstein, 1978; Fejerman and Medina, 1980; Jordan, 1980; Tassinari *et al.* 1982; Braun and Weidner, 1983; Tassinari *et al.* 1985). This means that this type of aphasia has been described more frequently than other forms, although one assumes it is a very unusual condition. Despite the great number of case studies the course and prognosis and pathogenesis remain unclear. This can be illustrated at best by a quotation of Dugas *et al.* (1982) in a study including 12 children with this syndrome: "Le propre du syndrome de Landau-Kleffner est d'échapper à toute systématisation".
Landau and Kleffner (1957) themselves termed the syndrome as acquired aphasia with convulsive disorder. The most recent studies labelled the syndrome as the Landau-Kleffner syndrome. However, other authors have circumscribed this type of aphasia and one encounters the following synonyms:

- auditory agnosia and convulsive disorder (Stein and Curry, 1968)
- acquired aphasia in childhood with seizure disorder (Deonna *et al.* 1977)
- Aphasie im Kindesalter mit EEG Veränderungen (Petersen *et al.* 1978)
- acquired auditory verbal agnosia and seizures in childhood (Cooper and Ferry 1978)
- verbal auditory agnosia in children (Rapin *et al.* 1977)
- acquired epileptic aphasia (Deuel and Lenn, 1977).

The clinical picture

The syndrome occurs in children which achieved developmental milestones at appropriate ages. The first manifestation of the language disturbance (aphasia) is an "apparent word deafness". The parents report a gradual inability to respond to their calls because raising their voices is ineffective. Medical evaluation frequently includes a normal audiogram. The aphasia can progress to (almost) total unresponsiveness to oral communication, while gradually expressive language becomes disturbed too. The children express themselves in a telegraphic style or in very simple sentences. The vocabulary decreases progressively and echolalia, literal and semantic paraphasias may occur (VandeSandt-Koenderman *et al.* 1984). Repetition of simple words may be possible, however, repetition of sentences is very severely distorted by perseverations and paraphasic errors. In most cases the syndrome starts at the age, when the children have not yet learned reading or writing. Incidentally, older children who before the onset of the aphasia could read and write satisfactorily lost these capacities (Rapin *et al.* 1977). A comparison with adult global aphasia can be made for the greater part of these children i.e. no verbal communication is possible.

A complicated factor is the fluctuating course of the aphasia with clear remissions and exacerbations. In addition to the aphasia, extended neuropsychological examination sometimes demonstrates mild forms of apraxia. It has to be emphasized that intellectual deterioration is an unusual feature.

Epileptic manifestations

Although the EEG is always abnormal, not in all children manifest clinical seizures occur. In a review of the literature (Dugas *et al.* 1982) epileptic seizures are mentioned in 56 of 70 cases. The epileptic manifestations are heterogeneous. Generalized seizures with predominantly motor components are most frequently observed. The number of seizures is very variable and a notable finding is a dissociation between the disappearance of clinical seizures and the slight improvement of the language deficits. In most cases anticonvulsant therapy is effective; Mantovani and Landau (1980) report that none of their 9 patients continued to have seizures.

EEG abnormalities

The reported paroxysmal EEG abnormalities may vary: bilateral independant temporal or temporo-parietal spikes, bilateral 1 to 3 Hz spike and wave activity maximal over the temporal regions, generalized sharp waves or spike and wave discharges, multi-local spikes or unilateral spikes (Gomez *et al.* 1983). Although some authors suggest a consistent relation between the course and the prognosis of the language disorder on the one side and the frequency and severity of seizures and EEG abnormalities on the other, most investigators expressed a different view. In the opinion of Holmes *et al.* (1981) "the EEG abnormalities in this syndrome are a epiphenomenon of underlying pathology of brain areas concerned with speech rather than a cause of the speech abnormalities". The same applies to the clinical seizures. In conclusion, the relation between the disappearance of abnormalities in the EEG recorded during wakefulness and the improvement of language deficits remains unclear (see case 1).

Possibly, a more recently described EEG phenomenon may clarify some aspects of this relation. In 1971, Patry *et al.* described 6 children in whom night after night and sometimes over a period of many years sleep induced a dramatic modification of the EEG, very characteristic of petit mal status but without any clinical accompaniment nor changes in normal sleep patterns. Lasting throughout the whole night, it subsides upon awakening. The aetiology in these children is very heterogeneous. Speech problems are considered as a striking feature. However, one has to note that the reported cases of Patry are all mentally defective; some of them started to regress later on (at age 8 respectively 11 years). The initial symptom was an attack of speech arrest.

In lack of better terminology and for descriptive purposes they proposed to call this phenomenon an Electrical Status Epilepticus induced by Sleep (ESES). This term must be questioned because it is difficult to assume, that continuous spikes and waves during sleep without detectable simultaneous clinical signs can be considered as an epileptic status, and because ESES can be found in non-epileptic patients (Morikawa *et al.* 1985). The terms "non-convulsive status epilepticus" (Broughton, 1983) or "non-convulsive status epilepticus encephalographicus during non-REM sleep" have been suggested as an alternative to ESES . The criteria, used for ESES, are twofold (Patry *et al.* 1971):

1. the EEG abnormalities had to become continuous with onset of sleep and cease to be continuous upon arousal. "Continuous" has been defined as at least 85 percent of the slow sleep tracing.
2. ESES has to be recorded on 3 or more occasions over a period of at least one month.

Both criteria are not applied generally. For example, in the view of Billard *et al.* (1982) a sleep paroxysmal activity surpassing 50% can be considered as ESES. Moreover, the same authors state that the densities of paroxysmal activity vary from one session to the next in the same individual. This raises the question of the consistency in time of the phenomenon, which is directly related to the second criterium. Exact data concerning the range of time in which ESES occurs, are totally lacking. One may agree the phenomenon of ESES is rare. Tassinari *et al.* (1985) report that despite worldwide interest in sleep studies in epileptic patients approximately 43 cases of ESES have been described since the study of Patry *et al.* (1971). Tassinari *et al.* (1985) give more details on ESES:

1. it manifests itself in the age range from 4,5 - 14 year.
2. ESES is a self-limited phenomenon. The exact duration cannot be assessed precisely; in one child ESES persisted over a 4 year period.

From a review of the literature, they conclude ESES has not been observed after the age of 15. In the view of Tassinari *et al.* (1982) ESES is not specific to the Landau-Kleffner syndrome. They argue that "a large number of patients with ESES did not show speech disturbances of a kind that would evoke this diagnosis". This statement, however, is not specified. In 1985, Tassinari modified his opinion. In contrast to his previous study, he emphasized not only from his personal series but also from a review of the literature, that ESES "is responsible for appearance of complex and severe neurological impairment, mainly concerning language functions". Dulac *et al.* (1983) gave more details. They described 76 children: 66 from the literature and 10 from a personal series, who developed epileptic seizures, paroxysmal EEG abnormalities and language deterioration after a period of normal development of language i.e. the Landau-Kleffner syndrome. In 7 out of the 10 children from their personal series, they observed the phenomenon of ESES during sleep. However, Dulac's definition of ESES is not in accordance with Patry or Tassinari: a 50% index of spike

and wave complexes during slow sleep is labelled as ESES. Nevertheless, Dulac *et al.* (1983) state that language disturbances are associated with the occurrence of ESES, arguing that ESES interferes with a normal functioning of the "speech areas". Consequently, a progressive loss of language functions will occur during the period of ESES, while the child only can recover after disappearance of ESES.
The most detailed case study has been reported by Kellerman (1978). Referring to the syndrome of Landau-Kleffner, he extensively presented a 6,5 year old boy in which ESES was present. Starting 8 weeks after the onset of the initial symptoms, 15 EEG's were recorded during 16 months. Kellerman reported that: "As soon as the boy fell asleep the EEG showed nearly continuous generalized hypersynchronous activity consisting of synchronous high voltage spike-waves and rhythmical spike-wave variants. Despite this electric epileptic status, there were no clinical symptoms of a seizure".
After this period, no follow-up has been done since further EEG investigations were refused by the child's parents. At the age of 13, the boy had recovered considerably. It is mentioned that reintroduction to a normal grammar school was considered. However, a relation in time between the disappearance of the ESES phenomenon and improvement of the aphasia could not evaluated exactly.

Behavioral disorders

The reported behavioral abnormalities are hyperactivity, agressiveness and depression. The severity of these disturbances ranged from mild to severe: a psychotic condition has been described unfrequently.

Course, outcome and treatment

In contrast to the epileptic phenomena the course of the aphasia is fluctuating (Mantovani and Landau, 1980). This observation has been confirmed by Deuel and Lenn (1977), Rapin *et al.* (1977) and Deonna *et al.* (1977). Deonna *et al.* suggested that the disorder was a heterogeneous one, with at least 3 separate forms, each with a different prognosis (see Table 1). However, no study included a sufficient number of children in order to confirm this hypothesis. Finally, the variability in outcome of the affected children is one of the most puzzling features of this disorder (Mantovani and Landau, 1980). Recently several authors suggested a relationship between outcome and the age at onset of the disorder: the younger the child the worse the long term outcome for language (Toso et al, 1981; Dulac *et al.* 1983; Bishop, 1985).
The therapeutic approach is different: Cole *et al.* (1988) described one child who was treated surgically. Left temporal lobectomy showed a transient improvement of the language function. Drug therapy (anticonvulsants and cortico-steroids) also failed to

alleviate the language deficit (McKinney and McGreal, 1974; Kellermann, 1978; VandeSandt-Koenderman et al, 1984). A great number of children received language therapy. No extensive descriptions of remedial approaches are available.

Aetiology

The aetiology of the syndrome remains unknown. The children show a normal educational-developmental course until the onset of the first epileptic seizures and/or language desintegration. Familial and personal medical history do not reveal relevant data, especially concerning epilepsy and language development. Clinical examination does not show associated neurological signs and in spite of the impressive EEG abnormalities, extensive investigations including cerebral angiography, cerebral computerized tomography, or nuclear magnetic resonance have never yielded any sign of an anatomical lesion. As a consequence, a great number of speculations concerning aetiology has been put forward. The vague relationship between the epileptic seizures and EEG abnormalities and the specific behavioural symptoms (aphasia, apraxia) has not facilitated the search to a common aetiological factor.
Landau and Kleffner (1957) hypothesized that persistant convulsive discharges in the brain tissue mediating language functions may result in a "functional ablation" of these areas. Gascon *et al.* (1973) have observed that in a limited number of children (see case 1, Table 2) an improvement of the language disorder does not occur, when the routine EEG normalized, which does not support the Landau-Kleffner hypothesis.
Since structural abnormalities have until now not been demonstrated and because of the fluctuations of the aphasia, also an inflammatory process has been assumed. However, convincing laboratory findings, indicating such a proces are completely lacking in the literature. Even brain biopsies (McKinney and McGreal, 1974, Cole *et al.* 1988) performed in 2 patients failed to clarify the underlying proces with exception of the case of Lou *et al.* (1977) who reported a biopsy "felt to be suggestive of a subchronic viral encephalitis".

Case reports

In the period from 1969 - 1984 we examined 6 children suffering from this syndrome. In all children the family history was negative, pregnancy and delivery were normal as was the neurological examination. Psycho-motor development, especially language development did not show abnormalities. To all children a (non-verbal) intelligence test was administered, which at that time showed performances within normal limits. Hearing loss is excluded. Moreover, no structural cerebral abnormalities could be demonstrated. Ancillary studies showed EEG abnormalities in all

patients, but arteriograms and CT scans were normal.
One case will be presented extensively. In particular, attention is paid to EEG patterns and the linguistic analysis. The clinical observations of the 5 other children are summarized and mainly focused on the course and outcome of the aphasia. We refer to chapter 2 for a detailed description of the methods of examination.
For details concerning neurological and electro-encephalographic data we refer to Table 3.

Case 1

The patient was a right-handed girl, born in 1965 after an uncomplicated pregnancy and delivery. At the age of one year the child began to walk, and the development of language was normal. There was no history of trauma, infection or epilepsy. In April 1969, however, the parents had the impression that their child had become slightly deaf. Often they had to raise their voices; markedly and frequently they had to repeat their questions before the child understood them. They then observed that the child articulated less well. Her sentences became fragmentary in construction, and after another month she expressed herself by gestures and indicated her wants with arm-movements. In June 1969 her vocabulary had become limited to one word, after much stimulation she could say "daddy".
In September 1969 the parents observed short periods of absent-mindedness when the child blinked her eyes, and objects dropped out of her hands. She was admitted to a pediatric department of a district hospital, where clinical examination showed no abnormalities with the exception of aphasia. The EEG (12.11.1969), however, showed local paroxysmal activity (see Table 2). She was then referred to the neurological department of our hospital for more intensive investigation. Again routine neurological examinations showed no abnormalities. CSF examination, skull X-ray, arteriography and pneumoencephalography were within normal limits, but the EEG (31.12.1969) continued to show abnormalities. Audiometry did not indicate any hearing loss. The girl was given diphenylhydantoin and phenobarbital, and no further seizures occurred. On discharge her speech had not improved. One year later, the child was brought to the hospital because of a generalized seizure. No signs of neurological abnormalities were found. The CT scan was normal, but the EEG showed a spike and slow-wave focus in the right temporal lobe (12.02.1971). To exclude metabolic disturbances, extensive investigations were carried out. Analysis of arylsulfatase, phenylketonuria, amino acid chromatography, and serum lipids revealed no abnormalities, and there was no evidence of toxoplasmosis. Because of difficulties in controlling the seizures, acetazolamide was added to the medication. No further seizures occurred. Psychological examination revealed a WISC performance IQ of 90. There was no colour blindness, and the perception of objects was good, although the copying of drawings was only moderately successful. After discharge, the girl entered a school for deaf children. The medication was

continued and in 1982 limited to Diamox (1 x 125 mg daily), and no relapse has been observed during the follow-up period up to 1984. She remained at this school for the next 10 years. During this period her speech improved slowly (see linguistic analysis Table 2). At the age of 16 (1981), she graduated from this school and attended a college for 2 years, where she studied to be a clerk. Presently she works at a bank establishment.

Methods

The child was frequently re-examined, neurologically and psychologically, and every year an EEG was recorded. Since 1973, the annual re-examination has consisted of an interview with some (open) standard questions, repetition of words and sentences, object naming and wordfluency tests. Auditory comprehension tests were administered, and reading and writing performances were assessed.

A. EEG (see Table 2)

The first EEG showed focal disturbances in the right temporal lobe. After one year, the focal activity had spread to the anterior, and to a less degree, to the posterior part of the right temporal lobe. The EEG recordings in the next year showed a variability in the severity of the focal disturbances, until after December 1971 when they were no longer evident. Bilateral synchronous paroxysms were absent in the first EEG but were present in subsequent recordings. Paroxysms were no longer observed after August 1971. The background activity also showed fluctuations (although to a lesser degree). Nevertheless the background activity over the right hemisphere remained slow; the subsequent EEG recordings made in the years after 1972 showed again background activity over both hemispheres which was too slow for her age. In 1977 the index of intermixed slow activity, which appeared mostly in bursts, often with sharp aspect, decreased considerably. Finally, in 1982 the background activity reached a normal dominant frequency of 10 Hz.

B. Linguistic Analysis

The data are summarized in Table 2.

Spontaneous speech

This was examined by means of standard questions. In 1970 and 1971 the child produced some isolated words ("not" and "that") but only in an emotional state. During the interview she tried to speak, but no meaningful sounds were produced. In 1973 verbal output remained limited to 3 different words, but in 1974 her

vocabulary had increased to 14 different words. Utterances of 2 words occasionally occurred, and the next year utterances of 3 words were counted 5 times in a sample of spontaneous speech. Moreover, she produced 37 different words, and in 1976 there was a gradual progress both in vocabulary (72 words) and in the length of the utterances (6 words). Her sentences were restricted to stereotypes.
In the next years, the child became more and more capable of producing longer utterances. Over the period 1977 - 1983 the degree of syntactic complexity increased. Qualitative aspects of recovery of spontaneous speech could be observed. Since 1980, pauses between words and sentences had considerably decreased (for a more complete analysis, see VanDongen *et al.* in press). The same applied to perseverative tendencies.

Repetition

In 1971 the girl was able to repeat an article or a digit. Thereafter there was very slow improvement. In 1973 and 1974 she succeeded in repeating words of 3 syllables, and by 1975 she was sometimes able to repeat words of 2 or 3 syllables correctly. In 1976 words of 2 syllables were correctly produced, and words of 4 syllables were repeated correctly about half the time. In 1975 repetition of simple sentences was still very difficult for her. She was able to repeat a sentence of 2 words. A reduction of content words and the omission of syllables and articles were observed in attempting to repeat longer sentences.
After 1976, only incidental errors were produced in repeating words consisting of more than 4 syllables. Repetition of sentences, however, remained a difficult task for her, even 14 years after the onset of the aphasia. Although she succeeded in repeating a sentence of 9 words, a simplification consistently occurred in reproducing. The omission of function words remained together with syntactic errors.

Wordfluency

In 1973 and 1974 the child could produce only one name of an animal in one minute, but during the next year a rapid recovery was observed. At the examinations in 1975 and 1976 she produced 14 and 17 names of animals respectively. After 1976 she performed according to age (Snijders and Verhage, 1962).

Object naming

She showed relatively one of her best performances on this test. In 1973 she was able to name 8 of the 10 objects presented, and in the following years her naming ability increased noticeably. Furthermore, when she had wordfinding difficulties, she frequently produced a good circumlocution of the target word.

Table 2 Data of the linguistic analysis and EEG-recordings (1969-1983).

LINGUISTIC ANALYSIS						EEG-RECORDINGS				
	SPONTANEOUS SPEECH					Background activity		Spike-wave activity		
Date	Vocabulary	Utterances	Repetition	Word Fluency	Tokentest	Slow	Irregular	Focal	Bilat. synchr.	Details
12.11.69	0	emotional utterances	0	0	not possible	+	+	+++	–	Spike-wave discharges in right temporal lobe
31.12.69	1	emotional utterances	0	0	0	±	+	++	+	Besides the focus in the right temporal lobe bilateral synchronous spike and wave complexes in the temporal lobe
12.01.71	–	---	–	–	–	+	+	+	+++	Focus in the right temporal lobe extends to the posterior region with generalized bilateral synchronous spike and wave activity
12.02.71	2	some emotional utterances	articles	0	0	±	–	+++	–	Focus in the right temporal lobe extends to anterior and posterior region: bilateral synchr. activity is absent
26.02.71	2	emotional utterances	articles	0	0	+	+	+	+++	Focus in the right temporal lobe: paroxysms of bilateral synchronous spike and wave activity
11.03.71	–	---	–	–	–	++	±	++	+	Slower background activity: more focal activity right temporal: less bilateral synchronous activity
19.08.71	–	---	–	–	–	±	+	+	+	Decrease of focal activity in right temporal lobe
06.12.71	–	---	–	–	–	±	±	–	–	Nearly normal EEG pattern
12.10.72	–	---	–	–	–	++	++	–	–	Marked disturbed background activity especially in the right temporal lobe: no focal discharges
17.12.73	3	---	words of 3 syllables	1	4	+	±	–	–	Improvement of the pattern: background is only slightly disturbed in the right temporal region

Date	Vocabulary	Utterances	Repetition	Word Fluency	Tokentest	Background activity Slow	Background activity Irregular	Spike-wave activity Focal	Spike-wave activity Bilat. synchr.	Details
23.12.74	14	utterances of 2 words	words of 3 syllables	1	9	+	±	–	–	Basically unchanged
29.12.75	37	utterances of 3 words	words of 4 syllables	14	31	+	+	–	–	No marked differences in the background activity over both hemispheres
27.12.76	72	utterances of 5 or 6 words, stereotype phrases	words of 4 syllables, sentences of 2 words	17	31	+	+	–	–	Slow activity 3-6 Hz predominantly over the right hemisphere
27.12.77	108	only simple grammatical sentences	words of more than 5 syll. sentences of 5 words	22	35	+	+	–	–	Basically unchanged
28.12.78	125	decrease of syntactical errors	no error in words, sentences of 3 words	26	43	+	+	–	–	Basically unchanged
27.12.79	119		sentences of 3 words	26	47	+	±	–	–	Bursts of sharp activity over fronto-temp. regions, predominantly right-sided
24.12.80	130	increase of syntactic complexity	sentences of 5 words	25	49	+	±	–	–	Less slow activity than before
23.12.81	116		sentences of 5 words	22	48	+	±	–	–	Bursts of sharp activity over fronto-temp. regions
22.12.82	128		sentences of 9 words	22	48	±	–	–	–	Marked faster background activity
21.12.83	132		sentences of 8 words	23	48	–	–	–	–	Incidently sharp transients over the left temporal regions, bursts of sharp activity over fronto-temporal regions. Normal background activity.

+ present
± nearly normal
– absent

Since 1978 a standardized object naming test (a slightly modified version of the Boston naming test) was administered to her (Immerzeel, 1985). She performed this test within the normal range.

Comprehension of spoken language

She appeared to have no comprehension of spoken language. In 1971 she could not correctly carry out simple commands of the first series of the Token Test (despite correct recognition of colours and forms), nor could she understand verbal commands such as "point to the door", and "touch your nose". Two years later she correctly performed 4 items of the first series of the Token Test, although obeying verbal commands (the three paper test of Pierre Marie) was still impossible for her. During the following years there was a certain recovery of understanding of spoken language, as reflected in the Token Test scores. In 1975 and 1976 she correctly performed 31 items of the full length Token Test. The increase of performance on this test gradually continued. Errors mainly manifested themselves in part 5, indicating difficulties in understanding syntactical constructions (Tallal, 1975). In 1983 the total score still demonstrated mild auditory comprehension difficulties (see Table 2).

Reading

In 1973 she could recognize some letters, and after one year she read articles and could spell words of 3 letters. Thereafter there was no rapid progress in reading. In course of time she spelled and read words of 3 and 4 syllables, but while reading a simple story aloud the omission of articles, conjunctions, and suffixes of verbs was frequently observed. However, a gradual recovery was observed and reading rate and prosody could be considered normal in 1983.

Writing

In 1973 she was able to write the Christian names of some of her classmates. In 1975 and 1976 writing to dictation was only possible for very simple sentences, as many errors (reversals and omission of letters) occurred. In the autumn of 1977 she spontaneously wrote short letters to a girlfriend, and in these letters the omission of articles, prepositions, and (to a less extent) verbs, was most striking. In 1981 spontaneous writing, for example a letter to a friend, still demonstrated simple sentences and incidentally problems with syntactic rules. A comparison of the 2 letters revealed not only an increase in the length of utterances (range 2-7 vs. 3-17 words in 1977 and 1981 respectively), but also an increase of syntactically correct sentences. Up to now, writing letters is a complex task for her.
In conclusion, 14 years after the onset of the aphasia subtle language deficits persist, but do not interfere with her professional life as a clerk.

Case 2

The patient, a right-handed girl, was born in July 1970. When she was 5 years old, psychological examination revealed a WIPPSI IQ of 91. She spoke in complete sentences. The comprehension of spoken language was good.
In March 1976, when she was 5 years and 8 months old, she had nocturnal seizures with twitching of the muscles of the eyes and face and the production of strange sounds.
One year later, her speech deteriorated. She spoke less and her articulation was poor. In a two-month period spontaneous speech disappeared almost completely. In July, when she was 7 years old, she was admitted to our department. On admission the only words she could say were "yes" and "no", "mummy" and "daddy". She hardly produced any sound either. Hyperactivity was present. The Raven IQ was above average. It was impossible to administer formal tests for language comprehension, because she did not cooperate in testing situations requiring linguistic performance.
In August 1977, after 10 days of prednisone therapy, she became "talkative": she started shouting and "babbling". This babbling was not very differentiated, most of the time she used the poorly articulated syllables "ti" and "ta", which she combined to different strings of varying length, besides, she spoke in a very high pitch. However, she did not start to say meaningful words and the prednisone treatment was stopped. The babbling and shouting disappeared almost immediately, and 4 days later she was given this drug again. Although another period of babbling was expected, her verbal behaviour did not change in the first 6 weeks. Then, early October, still under prednisone medication, she suddenly made considerable progress: she started to use meaningful words again. At first her articulation was very poor. Halfway through October it was possible for the first time to make a tape-recording of a conversation with the patient. From then on her spontaneous speech was tape-recorded monthly (for a more complete linguistic analysis, see VandeSandt-Koenderman, *et al.* 1984). Intensive speech therapy was started. In November she was placed in a rehabilitation clinic for children, where speech therapy and prednisone were continued. No side effects of the prednisone were observed.
Suddenly, in March 1978 she had a grand-mal seizure and another period of language breakdown occurred. Prednisone medication was increased but the breakdown of language continued. This time, beside her spontaneous speech problems, there was a reduction of language comprehension as well. She was considered globally aphasic by the end of July 1978. Although her behaviour suggested a global intellectual deterioration, the Coloured Progressive Matrices score (Raven, 1975) surprisingly held the same level.
The prednisone treatment, which had been given for 10 months, was gradually

stopped and the global aphasia lasted for about 7 months. In the mean time she had been dismissed from the rehabilitation centre (April 1978). From then on she attended a school for special education. Then, completely unexpected, again a rapid recovery occurred in May 1979, but this time without steroid drugs. She became again talkative. The recovery of the language functions continued. An assessment of her teacher (April 1980) was illustrative. Although he observed manifest word-finding difficulties in the spontaneous speech, sentences of simple grammatical structure were now present. In addition, comprehension of spoken language manifested itself, when the teacher spoke in a slow rate. She learned to spell simple words. Repetition of words was possible; the same applied to sentences of 3 words. The increase of recovery was slow but continuous: the level of the spontaneous speech grew as well for vocabulary as syntactic capacities. In the next years the school teacher reported a considerable progress of reading and writing. A repeated neuropsychological examination (11.02.85) demonstrated an increase of all language aspects in the spontaneous speech. The child expressed herself in longer utterances; wordfinding problems were less frequent. A mild verbal comprehension disorder was still present (Token Test score = 47), which also could be detected in the repetition of more complex sentences. She had to spell when she read words of 3 or more syllables frequently. Writing under dictation was still difficult. Eight years after the onset of the syndrome aphasic disturbances could still be demonstrated.

Case 3

A right-handed girl, born 12.11.1977. Development was reported to be unremarkable until May 1983. In that period the parents noted that she was staring for some seconds and sometimes shaked her head. In the next months these clinical seizures increased. In October 1983 her expressive language deteriorated; in December 1983 word deafness was observed. Moreover, she articulated less well. She was referred to a hospital for epileptic children; a sleep EEG was recorded (see Table 3). Anti-epileptic therapy was given and seizures stopped in February 1984. In April 1984, a sudden onset of language recovery especially in the expressive modalities developed. One month later the auditory comprehension improved considerably. A neuropsychological examination dd. 28.05.84 demonstrated wordfinding problems as well as the spontaneous speech as in an object namingtest were present. Repetition of simple sentences was not possible. Verbal commands administered in a slow speech rate are, however, understood well.
In August 1984 she was introduced in a primary school. A neuropsychological examination (July '85) did not demonstrate aphasic deficits at all.
Until now (1987) no learning difficulties were reported. On the contrary she was one of the brightest pupils of her class of a primary school.

Case 4

A right-handed girl, born 15.01.79. In June 1983 the parents noted a "word deafness". Often, the child requested to repeat questions "what do you say"?. Meaningfull sounds (telephone ringing) were well recognized. Gradually she started to react to gestures while the child expressed her wishes with gestures later on too. In December 1983 the expressive language worsened considerably: the child's speech was telegraphic; but she articulated well. In the next months her speech deteriorated still more and in March 1984 she recognized less well meaningfull sounds. At neuropsychological examination dd. 3.9.84. she rarely spoke in sentences describing the pictures. Repetition of simple words was not possible; in contrast her performances on a object naming test were mildly disturbed. Comprehension of spoken language was severely disturbed. In October 1984 the teacher of a kindergarten observed a progress in spontaneous speech and understanding of spoken language. A neuropsychological investigation confirmed this impression. There was a recovery of the expressive language especially. The course of the aphasia was favourable: in 1985 the level of verbal comprehension increased, although repetition of words and sentences remained a difficult tasks to her. At the primary school, the child learned to read and write. An examination in November 1986 demonstrated an increase of syntactical complexity in spontaneous speech. Repetition of words of 2 and 3 syllables and simple sentences progressed, while the child regained more and more receptive language capacities, although performances on the Tridimensional Matrix Test and the Token Test were low.

Case 5

A right-handed boy, born 03.06.76. At the age of 3 years, periods of staring were observed and there was a language break-down. An EEG (September 1980) showed focal epileptic activity. The epileptic manifestations were not influenced by anticonvulsant drugs, the subsequent EEG recordings showed a marked increase of epileptic activity. In July 1982 he suddenly showed signs of recovery. His parents reported that speech had recovered completely. However, since August 1983 his speech gradually began to deteriorate. Neuropsychological examination showed a severe language comprehension deficit; the spontaneous language was limited to emotional utterances. The EEG during sleep showed continuous generalized hypersynchronous activity (ESES) without clinical evidence of seizure. The aphasia showed fluctuation. A repeated examination (April 1984) revealed a slight improvement. He spoke in stereotypes: repetition of words was possible; he understood now simple verbal commands. When examined 11.7.85 his performances were basically unchanged.

Case 6

In 1981 a right-handed boy, born 30.03.77, appeared to be "deaf" and not alert. After a mild head injury petit mal attacks were observed frequently. The EEG was severely disturbed by continuous spike-wave complexes (1982). Drug therapy was not effective and he was referred to an institution of epileptic children. At admission his vocabulary was limited. All-night polygraphic EEG-recording was conducted (March '83): a spikes and waves (SW) index >85% during slow sleep was recorded (ESES). In this period his communication skills disappeared; he seemed to be mutistic. In April 1983, his alertness and the comprehension of spoken language increased. In neuropsychological examination, dd. June '83 a dissociation between expressive and receptive language capacities became manifest. Expressive language was limited to meaningless sounds; he was unable to repeat words. In contrast, he could perform the most complex items of the Tridimensional Matrix Test. He obeyed the complex verbal command of his mother as for example "open the door, when you have given me the book". In the next week there was a dramatic improvement. Because the child spoke too much, his voice became hoarse. Repeated neuropsychological examination (7.7.83) showed a talkative boy. In his spontaneous speech wordfinding problems, literal and semantic paraphasias occurred. He expressed himself in complex syntactic sentences. He articulated very well. Repetition of short sentences was difficult for him. The comprehension of spoken language during the interview seemed to be intact, although the Token Test demonstrated mild disturbances of verbal comprehension. The boy was dismissed and entered a school for special education (LOM-school). The parents reported a continuous progress of language functions until October 1983, when language deteriorated. Within 3 months no communication was possible. Despite intensive speech therapy in a school for deaf children his condition did not change. Until 1987 he did not react to verbal stimulation and did not speak. Continuously, he was playing with puzzles, a box of bricks and so on. EEG recording showed the characteristic features of continuous spikes and waves appearing and persisting every time the child slept.

Table 3 Landau-Kleffner subject data: sex, age, initial deficits, epilepsy and course of aphasia.

Pat. nr.	sex	birth date	age at onset	date of onset	first sign	epileptic symptomatology	epileptic activity EEG	ESES	course aphasia	follow-up period in years
1	F	12.04.65	4	April '69	Comprehension deficit	absences since Sept. '69 generalized seizure Jan. '71 Since Feb. '71 no epilepsy	absent since Dec. '72	–	slow and continuous improvement	17
2	F	26.07.70	5,8	March '76	epilepsy	atypical absences since March '76; grand mal seizure focal motor insult March '78, since Aug. '86 no epilepsy	absent since July '84	–	fluctuating course. Since May '79 continuous improvement	8
3	F	12.11.77	5,6	May '83	epilepsy	partial epilepsy since May '83 no epilepsy since Feb. '84	absent since March '84	no Feb. '84	rapid recovery since April '84	3
4	F	15.01.79	4,7	June '83	comprehension deficit	absences twice observed	absent since June '86	no June '86	slow and continuous improvement since Oct. '84	3
5	M	03.06.76	3,1	July '79	simultaneously epilepsy and language breakdown	atypical absences minor status epilepticus	present Oct. '87	present Oct. '83	fluctuating course	3
6	M	30.03.77	4	March '81	comprehension deficit	psychomotor epilepsy, grand mal insults	present Aug. '87	present March '83 Nov. '85	severe deterioration of language functions. Since Jan. '84 no communication skills	4

Discussion

Case 1 offers the opportunity to outline the general problems in diagnosis, course and outcome. The results of this longitudinal study show improvement of both the language functions and the EEG abnormalities. However, synchronicity is lacking. Despite the disappearance of the focal spike-wave complexes and the generalized paroxysms 2 years after the onset of the aphasia, the improvement of the language functions was only minimal. Since 1973 the gradual improvement of these functions has continued while the background activity remained too slow for her age, which might partly be due to the longstanding medication. It may be concluded that no clear-cut relation is present between the disappearance of the above mentioned EEG parameters and the improvement of the language functions. This is in accordance with Dugas *et al.* (1982), although rarely a different assumption has been suggested (Shoumaker *et al.* 1974). Moreover, Dugas *et al.* (1982) stated that in the Landau-Kleffner syndrome the EEG findings typically show a great intra- and interindividual variability. Rodriguez and Niedermeyer (1982) expressed a different view: the EEG shows frequently a normal or mildly slowed background activity and the paroxysms, although often generalized, tended to be lateralized to the mid-temporal regions with a slight preponderance of the left side. It has to be emphasized that in our right-handed patient the spike-wave focus was always on the right. Therefore, when in this patient the right-sided spike and wave focus together with the slowing of the background activity in the same hemisphere are interpreted as local disturbances of cerebral function, aphasia would not be expected.

One could refer to crossed aphasia. However, well-documented crossed aphasia in children is extremely rare (Assal and Deonna, 1977) and consequently language disorders in right-handed children may be expected after structural lesions of the left hemisphere, perhaps irrespective of their age (Woods, 1983; Vargha-Khadem *et al.* 1985). In our case we are concerned with a temporary functional focus of epileptic origin and it is assumed that both the language disorder and the spike-wave complexes during wakefulness are an epiphenomenon of the same underlying process, the pathogenesis of which remains completely obscure.

The recovery of language functions is slow and without exacerbations. However, the rate of improvement of the different language functions varies considerably. While the performances on an object naming test and a wordfluency test were within normal limits in 1977, the spontaneous speech impressed as agrammatical. On the whole, syntactic difficulties seem to play a more important role in the clinical picture of this aphasia than semantic ones. Syntactic errors were not only observed in the spontaneous speech, but also in repetition and spontaneous writing. Moreover, these errors could be demonstrated in auditory comprehension i.e. performances on the Token Test, a test sensitive to mild comprehension disorders. The greater part of

the errors was made on the commands of part 5 of this test in which the more elaborate syntactic structures can be found.
It has to be remembered that syntactic problems are generally not considered the most significant feature of acquired childhood aphasia. Acquired childhood aphasia has been described as non-fluent or motoric (Ludlow, 1980) in which the syntax is rather simplified than erroneous (Alajouanine and Lhermitte, 1965). Recent publications and the cases described in chapter 2 of this thesis, however, plead against this concept (Dennis, 1980; VanDongen *et al.* 1985; VanHout and Lyon, 1986). Finally, in accordance with the follow-up study of Mantovani and Landau (1980), our patient still complains of mild difficulty in understanding usual conversation, particularly if there is background noise. Probably, syntactic problems also play a role here too.

Summarizing, the 6 cases present a great variability of clinical aspects of this syndrome (see Table 3). On the basis of these data, one should be cautious about invoking firm statements concerning course and outcome.
The following comments illustrate this view:

1. the pattern of the aphasic disturbances in the Landau-Kleffner syndrome may be very heterogeneous both at onset and during course of the illness (case 1 vs. case 6).
2. the duration of the follow-up is important for an evaluation of the outcome: longitudinal follow-up of the Landau-Kleffner syndrome shows continuing improvement over many years, even if the degree of aphasia at onset was severe (see case 1 and 2).
3. the occurrence of epileptic activity in an EEG recorded during wakefullness is not a prognostic sign and a normalized EEG does not correspond to the disappearance of the aphasic deficit (cases 1 and 4).
4. for an evaluation of the variable "age at onset" (the younger the child the poorer the outlook) one has to follow these children up into adulthood, when further significant improvement seems unlikely (case 2).
5. the phenomenon of ESES was found in 2 children (cases 5 and 6). At present, both these children are the most severely aphasic ones. However, the follow-up period does not extend over the 4 years. Case 1, of which a nocturnal EEG is absent, demonstrates that considerable recovery is possible after a 4-years period of severely aphasic disturbances. Consequently, an assessment of ESES as a prognostic sign is not yet possible in both children. A necessary improvement of language functions could depend on disappearence of ESES.

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CHAPTER 6

ACQUIRED DYSARTHRIA IN CHILDHOOD: AN ANALYSIS OF DYSARTHRIC FEATURES IN RELATION TO NEUROLOGIC DEFICITS

Introduction

For the last 2 decades, dysarthria has mainly been considered as an articulatory disturbance. A medical dictionary definition is: "imperfect articulation in speech" (Dorland, 1965). Modern usage of the term is more comprehensive and dysarthria has been defined as "a collective name for a group of speech disorders resulting from disturbances in muscular control over the speech mechanisms due to damage of the central or peripheral nervous system" (Darley *et al.* 1969; Darley *et al.* 1975). Darley's definition distinguishes dysarthria from aphasia which is a language disorder acquired as a result of a cerebral lesion (see chapter 1). Besides an evaluation of articulatory problems, disorders of respiration, resonance and prosody are examined.

Recently, much attention has been paid to apraxia of speech (verbal apraxia). In their definition, Morley *et al.* (1954) exclude overtly impaired neurologic function and define this disorder as a deficit in the ability to carry out coordinative movements of the respiratory, laryngeal and oral muscles for articulation. In addition, this speech pattern considerably differs from dysarthria, mainly in the inconsistency of the errors in production of vowels and consonants in words and phrases.

Although in adults dysarthria seems to be a well defined concept, in reality it is not a unitary symptom, but a number of very differentiated speech disorders, characterized by different features. Classifications are based on:

a. aetiology e.g. Parkinson dysarthria,
b. site of the lesion e.g. bulbar dysarthria, cerebellar dysarthria,
c. combinations of a, b as in "spastic-flaccid-hypokinetic" dysarthria in Wilson's disease.

In adults multiple types of dysarthria which sound differently and with a different kind of abnormality of motor function have been demonstrated. In children, acquired dysarthria has not been studied in detail so that until now no comparisons can be made with the adult forms. This means that in children no opinions can be given on the characteristics, course and prognosis of this defect. One of the reasons for the lack of information on the subject of acquired dysarthria in childhood is that many neurologic diseases (Parkinson's disease, multiple sclerosis, Huntington's

disease, amyotrophic lateral sclerosis) which may cause dysarthria, rarely occur in childhood. Consequently, there are few studies of acquired dysarthria in children (Bak *et al.* 1983). Some reports, concerning acquired childhood aphasia (Alajouanine and Lhermitte, 1965) indicate an associated "articulatory disturbance" or "reduced speech intelligibility", but details related to the speech characteristics are not provided. In benign childhood epilepsy with rolandic spikes, anarthria during attacks is a wellknown feature (Lombroso, 1967; Loiseau and Beaussart, 1973); the recovery pattern of this anarthria has not been analysed either.

In the present chapter we report the course of acquired dysarthria speech in 8 children in relation to the neurologic deficits, contrasting patients with bilateral peripheral versus patients with bilateral supranuclear facial palsy.

Methods of examination

A. Speech analysis

During the speech assessment we tried to elicit as much spontaneous speech as possible. The children were also asked to repeat words and sentences. All these tasks were recorded on tape. The speech was judged by 2 independent observers according to slightly modified criteria (Bak *et al.* 1983) as described by Darley *et al.* (1969). For each tape recording, the dysarthric characteristics in the spontaneous and the repeated speech were arranged in 3 categories according to severity: (1) slight, (2) moderate and (3) severe. Eleven characteristics of abnormal speech (of a total of 38 mentioned by Darley *et al.* (1969) were at (ir)regular times encountered in our patients. For each recording, a total score could be determined by adding the scores for these 11 characteristics (maximal score 33). In this way, a rough estimate of the severity of the dysarthria was obtained, facilitating a comparison of the patients and a follow-up of the dysarthria.

B. Neurological examination

Apart from the routine neurologic examination, an elaborate protocol was designed to study the motor and sensory signs in the face, oral cavity, pharynx and tongue. Depending on the item studied, either one of 2 scoring systems was used: an ordinal scale from 1 (severely abnormal) to 4 (normal), or a distinction between abnormal (sign present) and normal (sign absent).

The following items were scored with the ordinal scale:
- opening and closing of the jaws,
- closing of the eyes,
- frowning,
- spreading of the corners of the mouth,
- pouting of the lips,
- wrinkling of the nose,
- protruding the tongue,
- lateral and alternating movements of the tongue,
- chewing, swallowing, blowing and coughing.

The following items were categorized as normal or abnormal:
- position of the mandible at rest (closed or drooping),
- alternating lateral movements of the mandible (possible or impossible),
- position of the corners of the mouth at rest (symmetrical or not),
- position of the uvula and the pharyngeal wall at rest (symmetrical or not),
- movements of the pharynx when vocalizing (present or absent),
- fasciculations of the tongue (absent or present),
- superficial and gnostic sensation in the area of the trigeminal and facial nerves (normal or abnormal),
- pseudobulbar phenomena (absent or present).

All examinations were performed by a childneurologist who at the time was unaware of the results of the speech analysis of the patient. The function of the vocal cords was judged by an ENT surgeon. For each examination, the items belonging to the functional area of one cranial nerve were taken together, and the severity of the deficit was assessed. Complex movements, involving more than one cranial nerve, such as chewing, swallowing, blowing and coughing, were scored separately, as were sensory disturbances and pseudobulbar signs. In this way, 10 neurologic parameters were scored from slightly (1) to moderately (2) to severly (3) abnormal. The total score per examination (max. 30) indicated the severity of the neurologic dysfunction at that time.

C. Patient data

In a consecutive series of 11 children with acquired dysarthria, examined between January 1980 and May 1983, 8 had a bilateral facial palsy. Table 1 gives the main characteristics of these patients; Figures 1, 2 and 3 show the CT appearances of patients 6, 7 and 8, and Figure 4 the angiographic findings in patient 5. All children had a normal speech development and normal school achievements before their illness.

The patients were examined for the first time as soon as their neurologic condition permitted their speech to be recorded. They were lucid and cooperative during the investigations. In patients 5 - 8, an extensive sensory examination was not possible initially, because articulatory problems prevented them from indicating their sensation.
However, later examinations did not reveal any sensory abnormalities in these patients. The patients were divided into a group with a bilateral peripheral facial palsy (1 - 4) and with a bilateral central facial palsy (5 - 8).

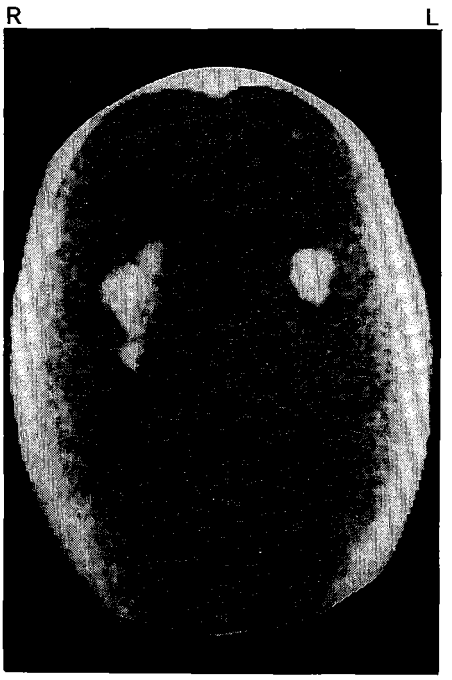

Fig. 1

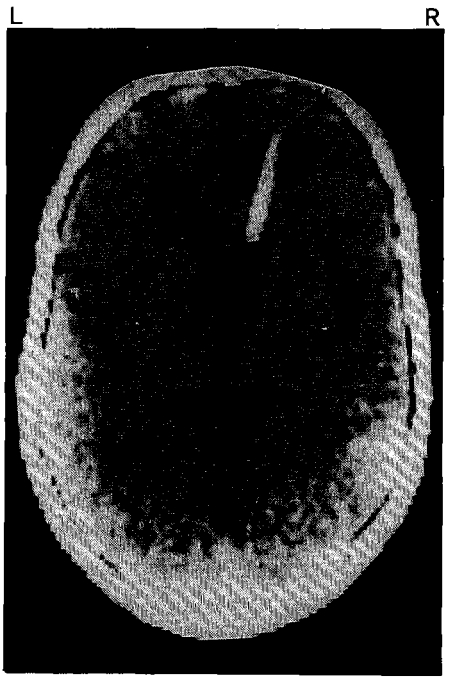

Fig. 2

Figure 1 *Patient 6. Multilobular haematoma in the right cerebral hemisphere and 2 smaller haematomas in the left hemisphere, due to coagulation disturbances.*

Figure 2 *Patient 7. Diffuse supratentorial atrophy and extensive white matter hypodensities in the cerebral hemispheres, due to intraventricular administration of methotrexate via a right frontal ventricular catheter. A local atrophy around the catheter is present, too.*

Table 1 Patient data.

Pt	Sex/age	Diagnosis and localization	Associated neurologic deficits
1	F/15	Bilateral 7th cranial nerve palsy due to leukemic meningeal infiltration	Compression of cauda equina
2	M/12	Bilateral 7th, 9th, and 10th cranial nerve palsies due to Fisher's syndrome	Bilateral 3rd, 4th, and 6th cranial nerve palsies; ataxia, areflexia of the upper extremities
3	M/11	Bilateral 7th and 9th cranial nerve palsies due to leukemic meningeal infiltration	Slight pyramidal weakness of right leg
4	M/11	Bilateral facial and bulbar weakness due to chronic inflammatory polyneuropathy	Flaccid tetraplegia and areflexia
5	M/6	Central facial diplegia and bulbar weakness caused by pontine infarction due to basilar artery occlusion (figure 4)	Left hemiplegia
6	F/13	Bilateral facial and lingual weakness caused by bilateral intracerebral haematomas due to coagulation disturbances caused by anti-leukemic treatment (figure 1)	Bilateral hemiparesis, left > right
7	F/14	Acute methotrexate encephalopathy after intra-ventricular methotrexate for CNS extension of lymphatic leukemia (figure 2)	Bilateral hemiplegia, left > right, with pseudobulbar phenomena
8	M/10	Bilateral cerebral contusion, on the right more than on the left (figure 3)	left hemiplegia, hemianopia, and hemi-inattention; frontal lobe syndrome, pseudobulbar signs

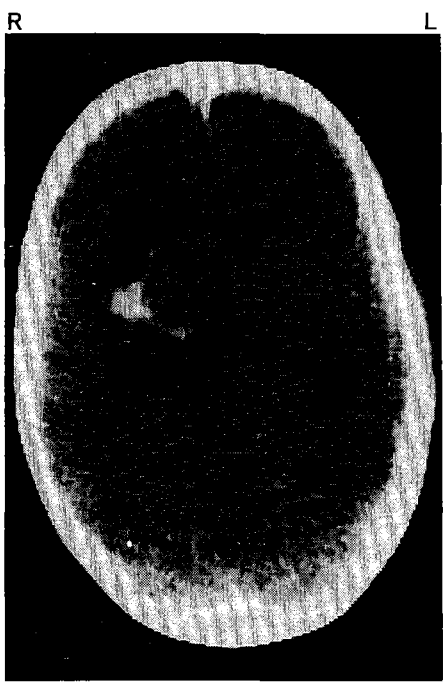

Figure 3 *Patient 8. Large haemorrhagic contusion in the right hemisphere and a smaller hypodense lesion in the lobe of the left hemisphere after a craniocerebral trauma.*

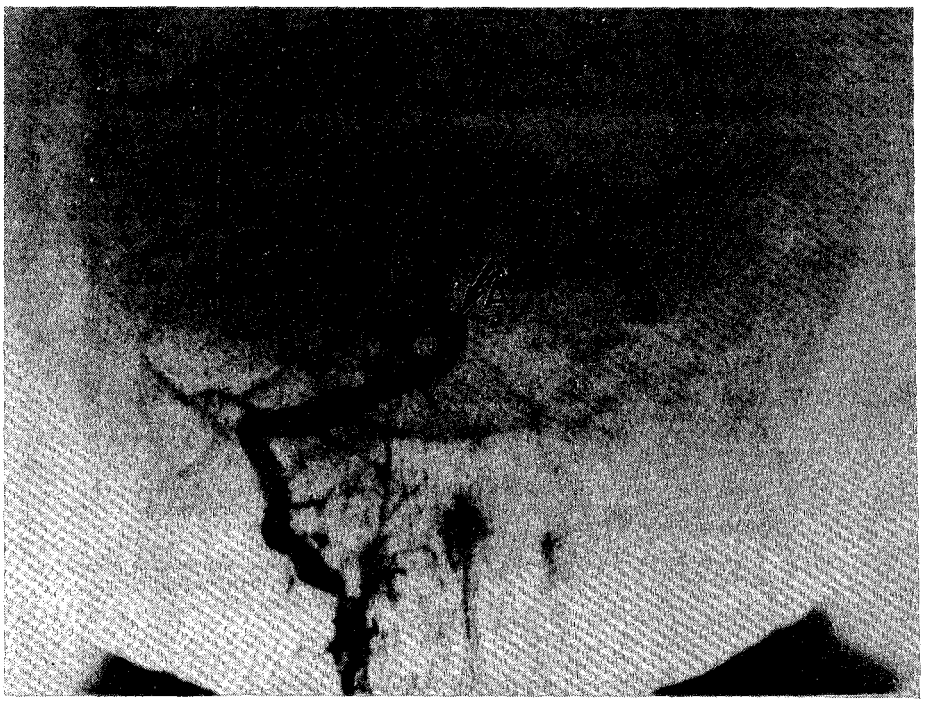

Figure 4 *Patient 5. Right vertebral angiography, showing almost complete occlusion of the basilar artery (A) with collateral flow via the posterior inferior cerebellar artery (B).*

Results

Table 2 summarizes the results of the first neurologic examination and the first speech analysis of the patients. Impairment of the mouth and the tongue movements was the most obvious in the examination of several complex functions, especially blowing. The most pronounced dysarthric features at the first examination were hypernasality and weakened vowels and consonants, followed by reduced stress. Many pauses and reduced stress were the only features present in all patients with a central type of lesion, whereas weakened vowels and consonants was the only feature present in all patients with a peripheral lesion. Disturbances of the voice and of inspiration and expiration were not observed in the group with peripheral lesions, except for the frequent inspiration of patient 4, but he had been dyspneuic shortly before the examination.

Table 2 Neurologic signs and dysarthric features at the time of the first tape recording.

Neurologic signs	Patient no. 1	2	3	4	5	6	7	8
Trigeminal weakness						**	***	*
Bilateral facial weakness UMN					***	***	***	**
Bilateral facial weakness LMN	***	**	***	*				
Pharyngeal weakness		**	*	***	***		**	
Lingual weakness					***	***	***	**
Impairment of								
Chewing					**	***	***	
Swallowing			*	**	**	**	***	
Blowing	**	*	***	**	*	***	***	**
Coughing			**	**	*	***	***	*
Sensory disturbances (face, mouth, tongue)					ne	ne	ne	ne
Pseudobulbar signs							**	**
Audible inspiration						***	**	
Frequent inspiration				***	*		***	
Breathy voice					***		***	**
Harsh voice				**				
Reduced stress				*	**	**	***	***
Dropped vowels/consonants					**		***	
'Weakened' vowels/consonants	*	*	**	**	**		***	**
Prolonged articulation					**			
Hypernasality	*	***		***	***	**	***	
Many pauses					***	**	***	**
Dys/aprosody							***	

*** Severely abnormal
** Moderately abnormal
* Slightly abnormal

ne Not examined
UMN Upper motor neuron
LMN Lower motor neuron

A number of dysarthric features only occurred in the group with a central lesion: dysprosody and prolonged articulation (1 of 4), audible inspiration and dropped vowels and consonants (2/4), breathy voice (3/4) and many pauses (4/4).

Four patients showed a rapid recovery (within 3 weeks) of the dysarthria and the accompanying neurologic signs: patient 1 by antileukemic treatment, patient 4 by plasma infusions (Maas *et al.* 1981), patient 6 without specific therapy except correction of the coagulation disturbances, and patient 7 by withdrawal of intrathecal methotrexate therapy. Unfortunately, patient 1 was not followed according to the neurologic protocol but only from the data in her file. A slow recovery (more than 6 weeks after the first examination) was seen in the other 4 patients: in patient 3 by antileukemic treatment, and in patients 2, 5 and 8 without specific therapeutic measures.

Finally, Figure 5 shows the relation between the severity of the neurologic deficit and the severity of the dysarthria, both at the first examination and during recovery. In patient 5 - 8, with supranuclear lesions, the first neurologic examination was performed while they were anarthric. At that time, the neurologic scores varied from 10 to 25.

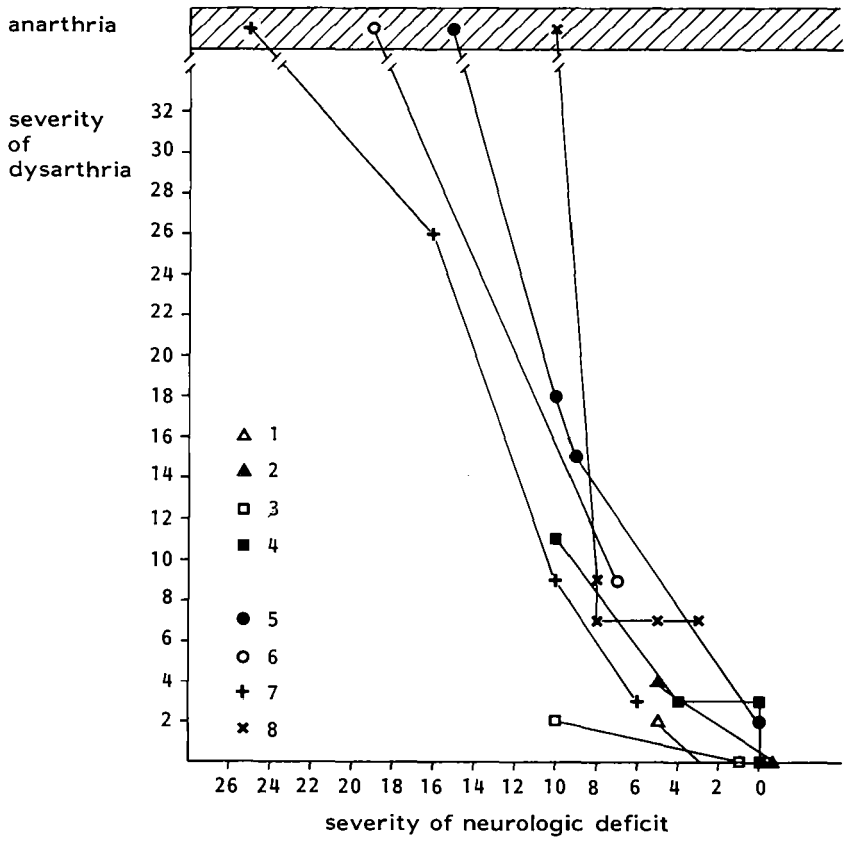

Figure 5 *The recovery of dysarthria in relation to the severity of neurologic dysfunctioning as expressed in total scores.*

Discussion

This study adresses - to the best of our knowledge for the first time - the characteristics of acquired dysarthria in childhood in relation to the neurologic signs. Surprisingly, children with a bilateral peripheral facial paralysis displayed only a small number of dysarthric features. These features were not very conspicuous, except for the hypernasality of patients 2 and 4. The concomitant pharyngeal weakness, or a combination of the facial and the pharyngeal weakness, may have accounted for this.

All children with a bilateral supranuclear facial paresis or paralysis were initially anarthric and later displayed a severe dysarthria, but they also had a severe impairment of the movements of mouth and tongue. Two of these children did not have

pharyngeal weakness (patients 6 and 8); they had the least severe dysarthria in this group.
A relation between (the presence of or absence of) pharyngeal weakness and hypernasality was apparent in patients 2, 4, 5, 7 and 8, but nor in patients 1, 3 and 6. Other dysarthric features showed even less correlation with pharyngeal weakness. The feature "weakened" vowels and consonants seems to be strongly associated with bilateral facial weakness, the only exception being patient 6 from the group with supranuclear lesions (a finding for which we have no explanation).
Lingual weakness only occurred in the group with supranuclear lesions; the best correlation was with the features "many pauses" and "reduced stress", less with "breathy voice". It remains to be determined whether these dysarthric features were indeed merely due to the lingual weakness, or to the supranuclear nature of the lesion in these patients.
During the begin of the illness, the cumulative data suggest a global correlation of the severity of the dysarthric features to the severity of the other neurologic signs (Figure 5). Initially, in the patients with supranuclear lesions the recovery of the dysarthria preceded the recovery of the neurologic signs.They recovered from anarthria to a reasonably intelligible speech with only slight or moderate neurologic improvement otherwise. Surprisingly, anarthria may occur in fully conscious patients with supranuclear lesions with only moderate neurologic scores (10, 15 and 19 in patients 8, 5 and 6 respectively). Moreover, this anarthria recovered while the severity of the other neurologic signs became only slightly less. A cortical control mechanism, possibly frontally located, cannot be the only explanation, since patient 5 suffered from a brainstem infarction. This localization is in accordance with the theory of VonCramon (1981), who suggested that initial aphonia and anarthria, followed by a severe dysarthria, could be caused by lesions in the ponto-mesodiencephalic region, especially the peri-aqueductal grey matter.
Evidently, the initial anarthria makes the speech problems of the patients with supranuclear lesions considerably worse than those of patients with peripheral lesions. On the other hand, the lack of dysarthric findings in children with a bilateral peripheral facial palsy deserves to be mentioned, too, since adults with a bilateral peripheral facial paralysis always show a severe dysarthria.
The overall prognosis of acquired dysarthria in childhood, judged from this small series, seems to be good, even if full neurologic recovery does not occur (as in patients 7 and 8, due to the nature of their illness). The final outcome of the dysarthria cannot, therefore, be considered a prognostic indicator of the persistence of other residual signs.

As far as we stated above neither acquired dysarthria in childhood nor its follow-up has ever been studied in detail. Therefore, we wish to present the case history of a six-year old boy who had a brain-stem infarct caused by occlusion of the basilar

artery and whose dysarthria was a prominent clinical feature (case 5). Follow-up studies are possible until recovery was complete, 7 years after the onset of his disease.

VM was a 6-year old, right-handed boy with normal language and motor development. He attended nursery school and was described as an active child, who maintained good relationships with other children. He was attentive and talked easily. In January 1980, he did not feel well in the morning; he complained of dizziness and staggered while moving around in class. He was taken home where he lost consciousness. The parents observed "shaking" in both legs and that his eyes "looked funny". The same day the boy was admitted to our department.

Neurological examination on admission

On admission, consciousness was clearly reduced. He only opened his eyes when addressed in a loud voice. A left hemiparesis was noted. Skull X-rays and lumbar puncture were normal. A few days later the level of consciousness improved remarkably, but there was still a dense left hemiplegia, affecting face, arm and leg. He could not move his mouth nor protrude his tongue and because of seriously impaired swallowing he had to be fed by nasogastric tube. The boy did not speak, but vocalized when crying and laughing.

An ENT surgeon examining him one week after admission, noted that the swallowing difficulties had disappeared and the pharynx and vocal cords functioned normally, but he still did not speak. Initially, the loss of speech was attributed to aphasia. However, the language comprehension was excellent as assessed by the Token Test (DeRenzi and Vignolo, 1962; VanHarskamp and VanDongen, 1977) and the Tridimensional Matrix Test (Kreindler *et al.* 1971). This finding, together with the disturbed mouth and tongue movements, led us to suspect anarthria. For angiographic investigation see Figure 4. Initially, the CT scan was normal; later on, a small infarct in the left thalamus was noted. Extensive investigations revealed neither source of emboli nor infectious or metabolic causes for the arterial occlusion.

Within the next week the left hemiparesis gradually improved, especially in the leg. He was discharged 9 weeks after the onset of the disease.

Methods of examination

The patient was in hospital for 64 days. The mouth and tongue movements were examined on days nine, 14, 21, 28, 35 and 62. Six audio-tape recordings were of sufficient quality to be included in the study. They were made on days 12, 19, 21, 23, 35 and 62.

During each examination we tried to make the patient talk spontaneously. He was

also asked to repeat words and phrases and to sing a song. All this was recorded on tape. The speech was judged by 2 independent observers. Only 14 of the 38 speech deviations mentioned by Darley *et al.* (1969, 1975) were noted:

1. imprecise consonants,
2. distorted vowels,
3. hypernasality,
4. (continuous) breathy voice,
5. (transient) breathy voice,
6. harsh voice,
7. monopitch,
8. monoloudness,
9. prolonged phonemes,
10. voice stoppages,
11. loudness decay,
12. reduced stress,
13. excess and equal stress,
14. abnormal rate of actual speech.

For each recording, the dysarthric characteristics in spontaneous speech and repetition were arranged according to severity in 3 categories: slight, moderate and severe. The inter-observer agreement was very high (Kendall coefficient of concordance .82; $p < 0.01$).
Singing was more globally rated, as precise criteria are not available.

Results

The recovery of mouth and tongue movements

Table 3 summarizes the recovery of mouth and tongue movements. At the first examination a left central facial paralysis and a right central facial paresis were found. Some faint grimaces remained, which could be interpreted as laughing or looking angry. Imitation of mouth movements was impossible. He could not close his mouth and was constantly drooling.
During the course of the illness, slow improvement in the functions of the lower cranial nerves occurred, as indicated in the table. The pharyngeal functions were fastest to recover and showed no abnormalities by the end of the fourth week. Tongue movements, especially rapid alternating movements, recovered more slowly, but at the last examination only minimal signs of bulbar weakness were present. Despite the fact that upper motor neuron weakness was present on both sides, at no time during the illness pseudo-bulbar reflexes could be elicited or pseudo-bulbar crying or laughing be observed. Cerebellar signs were not present.

Table 3 The recovery of mouth and tongue movements.

	Cranial nerve		
Time	VII	IX/X	XII
Day 9	Facial diplegia improving on the right side	No pharyngeal movements; gag reflex absent on both sides; not able to swallow	No tongue movements
Day 14	Left facial upper motor neuron paralysis; slow improvement of facial expression at rest, but making or imitating mouth movements impossible	Able to swallow soft foods	Slight movement to the left possible, not beyond the teeth
Day 21	Same	Uvula hanging to the left side; gag reflex absent on both sides; vocal cords functioning normally	Slow improvement
Day 28	Only slight facial asymmetry at rest; no movements of the left corner of the mouth	Able to chew and swallow bread	Slowly protruding tongue to the left; no alternating movements
Day 35	Starts to make blowing and whistling movements	No abnormalities	Protrudes tongue to the left and the right; slow alternating movements with many failures
Day 62	No abnormalities	No abnormalities	Slight deviation of the tongue to the left; rapid alternating movements possible

The dysarthria was mainly characterized by:
1. imprecise consonants,
2. distorted vowels,
3. hypernasality,
4. continuous breathy voice,
5. transient breathy voice,
6. harsh voice,
7. monopitch,
8. monoloudness.

The assessment of the spontaneous speech according to dysarthic characteristics

The course of the most important features in spontaneous speech is presented in Figure 6. Twelve days after admission, speech was difficult to understand. He was severely hypernasal, while vowels and consonants were extremely distorted.

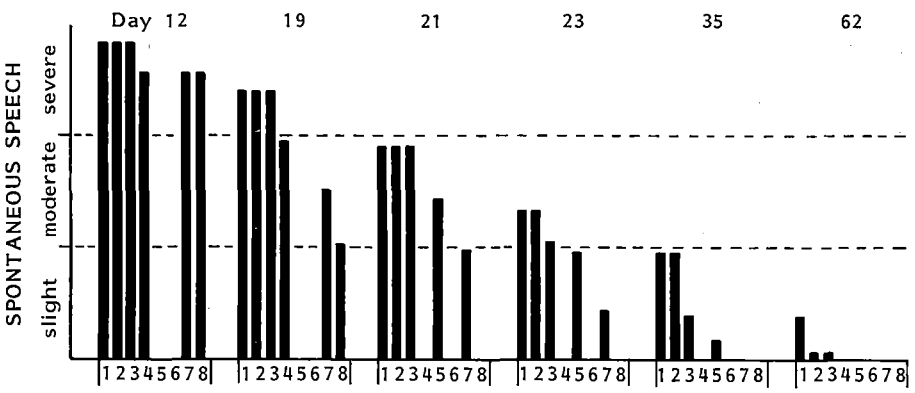

Figure 6 The course of the prominent features of dysarthria in spontaneous speech.

He spoke monotonously; there were no variations in loudness and the voice was continuously breathy. Day 19: the hypernasality, the distorted vowels and the imprecise consonants (the 3 main deviations) remained prominent, although some improvement could be heard. This improvement continued gradually during the next months. Day 21: three-word sentences were heard. The breathiness was no longer continuously present. He interrupted polysyllabic words by breathing and swallowing. Many interjections of "eh,eh" could be heard within the phrases. The rate of speech was very slow. On day 23: the hypernasality was no longer continuously

present and on day 35: the speech deviations, breathy voice and hypernasality had improved considerably. On the last recording (day 62), the consonants were more precise. He was only slightly nasal. Other characteristics were heard occasionally, namely loudness decay (nr. 11, day 12), voice stoppage at the end of sentences (nr. 10), reduced stress (nr. 12) and excess and equal stress (nr. 13, all on day 19).

Repetition of words and phrases

In general, it may be said that the recovery patterns of spontaneous speech and of repetition showed only slight differences. First, not all features could be applied to repetition: monopitch (nr. 7), monoloudness (nr. 8) and reduced stress (nr. 12) were difficult to judge, because of the tendency of the child to imitate the examiner's intonation. Secondly, during the first examination vowels and consonants were less distorted in repetition, and consequently the phonemes were more clearly discernible. Thirdly, speech repetition showed more rapid recovery (Figure 7).

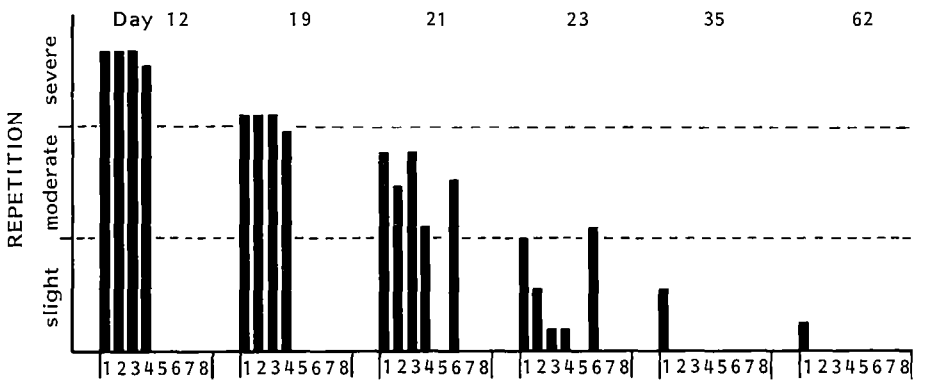

Figure 7 The course of the prominent features of dysarthria in repetition.

Day 12: the child tried to imitate monosyllabic words. A severe hypernasality was present, vowels and consonants were extremely distorted and he spoke in an obviously breathy voice. 21 days after admission, the hypernasality and the defective pronounciation of vowels and consonants were prominent, although the vowels were now beginning to show some improvements. The voice sounded harsh; this disappeared within 2 weeks. 35 and 62 days after admission, the dysarthria was only characterized by slightly imprecise consonants.

The assessment of singing

On day 12, one of the parents sang a well-known children's song to the patient, who, now and then, was able to join in with great difficulty. The words of the song were hardly intelligible. The pattern of recovery was idential to that of spontaneous speech and repetition. However, the melody could be well recognized in an early stage of recovery (day 21).

Conclusion

The dysarthria of this boy was mainly determined by imprecise consonants, distorted vowels, hypernasality and a breathy voice. During the course of his recovery, these dysarthric features were continuously prominent. Some characteristics of the dysarthria could be recognized better in the spontaneous speech than in repetition. No explanation can be given that a harsh voice was heard in repetition at only 2 examinations (day 21 and 23).
During the first 3 weeks, there was a remarkable improvement of the 4 main dysarthric features. Imprecise consonants were the only abnormality at the last examination. A speech analysis of this boy, 7 years post-onset showed no signs of dysarthria. The boy has finished the primary school without learning problems. At the moment, he is attending a secondary school.

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SUMMARY

This thesis focuses on acquired aphasia and dysarthria in childhood which until now have been rarely studied in detail.

In *chapter 1* the literature on features, course and prognosis is reviewed. Until the last decade it was generally believed that acquired aphasia in childhood:
a. is invariably non-fluent,
b. is of short duration,
c. does not persist if it occurs before the age of 12 years.

This chapter points out that this currently accepted doctrine is largely supported by the earlier literature, but has been contradicted by more recent reports, including the present study.

In *chapter 2* the view that acquired aphasia is always non-fluent and devoid of paraphasias is challenged. Rated for a number of characteristics proposed for the classification of fluent speech in adult aphasics, the language defect in 5 right-handed children is extensively described. These 5 patients demonstrate that before 12 years of age a language deficit resembling fluent aphasia in adults can occur. The results contradict the statement of Lenneberg (1967) that "the so-called fluent aphasia is rare or perhaps altogether absent amongst pediatric patients".

In *chapter 3* the clinical picture of acquired aphasia is described in more detail. An analysis of naming errors during recovery shows that neologisms, literal and verbal paraphasias do occur. The cause of the lesion determines the recovery course of neologisms, but no other errors.

In *chapter 4* the effects of a number of variables on course and outcome were investigated: age at onset, aetiology, severity and bilaterality of lesion, and type of aphasia. Analysis of spontaneous speech and tests of auditory verbal comprehension were used to determine the presence of aphasia. The severity of the cerebral lesion was assessed by means of a 5-point rating scale for CT scans. It is concluded that most of the children have not recovered completely one year post-onset. The recovery was significantly different according to the cause of the lesion. Complete recovery was seen in the majority of our traumatic cases. In contrast, bilaterality of the cerebral lesion showed a reverse correlation with recovery.
Concerning the course after more than one year only a general outline could be given.

In *chapter 5* the literature on the characteristics of a particular type of language deficit "acquired aphasia with convulsive disorder" or "the syndrome of Landau-Kleffner" is reviewed. In addition, clinical and electro-encephalographic features of

6 personal cases are presented and the significance of Electrical Status Epilepticus induced by Sleep, (ESES) for the prognosis of aphasia is discussed.

Chapter 6 is devoted to the analysis of the dysarthria of 4 children with bilateral supranuclear facial palsy and 4 with bilateral peripheral facial palsy. The children with peripheral lesions had only moderate dysarthria, characterized mainly by weakened vowels and consonants and by hypernasality. In contrast, children with supranuclear lesions were anarthric at first, followed by severe dysarthria with reduced stress and many pauses. Although there was a relation between severity of dysarthria and neurological disorders, anarthria may be seen in children with bilateral supranuclear lesions and only slight neurological disability.

SAMENVATTING

Deze studie richt zich op twee, zowel in de neurologie als in de taal- en spraakpathologie, weinig bekende en beschreven neurologische symptomen, te weten verworven afasie en dysarthrie bij kinderen.

In *hoofdstuk 1* worden de algemene opinies, die tot aan het eind van de jaren '70 bestonden over verworven afasie aan een kritische beschouwing onderworpen. Tot een tiental jaar geleden was men van mening dat:
a. de afasie altijd non-fluent is,
b. van korte duur is,
c. volledig herstelt, indien ontstaan vóór het twaalfde jaar.

Daarnaast geeft dit hoofdstuk een algemeen overzicht van de veranderde inzichten in het klinisch beeld, het beloop en herstel van deze taalstoornis. Hierin zijn ook de meest recente gegevens (tot en met 1987) betrokkken.

In *hoofdstuk 2* wordt aangetoond, dat de verworven afasie bij kinderen niet tot het non-fluent type beperkt blijft. Indien men criteria voor de classificatie van non-fluent/fluent taalgebruik bij volwassen afatische patiënten toepast op de spontane taal van kinderen met een verworven afasie, dan vindt men in ons verzamelde patiëntenmateriaal bij 5 kinderen een taalgebruik passend bij een fluent afasie. Deze gegevens zijn in tegenspraak met de mening van Lenneberg (1967): "The so-called fluent aphasia is rare or perhaps altogether absent amongst pediatric patients". Voor deze discrepantie worden in hoofdstuk 2 een aantal verklaringen gegeven.

In *hoofdstuk 3* wordt het klinisch beeld van verworven kinderafasie nader gedifferentieerd. In het algemeen wordt aangenomen, dat neologismen bij kinderen niet aanwezig zijn en parafasieën, met name semantische parafasieën, zelden zullen voorkomen. Aan de hand de resultaten van een benoemtaak wordt deze opinie weerlegd. Een analyse van de benoem-fouten gedurende het ziektebeloop toont aan, dat neologismen, literale en verbale parafasieën niet alleen voorkomen, maar dat er ook een relatie lijkt te bestaan met de ziekte die de afasie veroorzaakt. De betekenis van plaats en grootte van de lesie kan, ten aanzien van het optreden van neologismen en andere parafasieën, niet geëvalueerd worden.

In *hoofdstuk 4* worden uitspraken weerlegd, dat de afasie bij kinderen van korte duur zou zijn en, indien ontstaan vóór het twaalfde jaar, volledig zou herstellen. Uit ons vervolgonderzoek van 28 afatische kinderen blijkt dat het merendeel niet binnen een jaar na het ontstaan van de afasie hersteld is. Verbetering blijkt afhankelijk van de aetiologie: traumatische afasieën hebben de beste prognose. De frequent verkondigde meningen, dat "hoe jonger het kind de afasie verwerft, hoe sneller en beter

het herstel" en "hoe groter de lesie, hoe slechter de prognose" kunnen niet bevestigd worden.

In dit hoofdstuk komen eveneens de gevonden interacties onder andere tussen leeftijd en aetiologie ter sprake. Geconcludeerd wordt dat de bestudering van grote groepen afatische kinderen noodzakelijk is om de bijdrage van de verschillende prognostische variabelen (leeftijd, aetiologie, grootte van de lesie, etc.) afzonderlijk te evalueren. Het ontbreken van dergelijke studies maakt de waarde van uitspraken ten aanzien van de prognose dubieus.

In *hoofdstuk 5* wordt een overzicht van een bijzondere vorm van afasie gegeven: het syndroom van Landau-Kleffner, of "acquired aphasia with convulsive disorder". De eigen ervaring met 6 patiëntjes met betrekking tot de diagnostiek en beloop worden beschreven. Tevens wordt aandacht besteed aan de electro-encephalografische afwijkingen en met name een fenomeen waaraan de laatste jaren aandacht is besteed, namelijk de "Electrical Status Epilepticus induced by Sleep", (ESES). Dit fenomeen wordt bij kinderen met het bovengenoemde syndroom bij polygrafisch slaaponderzoek gevonden. Het verdwijnen van ESES wordt als essentiële voorwaarde voor herstel van de taal beschouwd. Gezien het nog korte vervolg van de door ons beschreven 2 kinderen met dit fenomeen, kan hierover geen uitspraak gedaan worden.

In het laatste *hoofdstuk* wordt het beloop van verworven dysarthrie bij 8 kinderen in relatie tot de neurologische uitvalsverschijnselen beschreven. De prognose van verworven dysarthrie op kinderleeftijd lijkt goed te zijn, zelfs indien het herstel van de overige neurologische defecten niet volledig is. De betrekkelijkheid van dit resultaat moet echter benadrukt worden, aangezien er slechts een beperkt aantal aandoeningen, waarbij het symtoom dysarthrie kan voorkomen, in dit onderzoek betrokken is. Bovendien ontbreken bij onze patiëntjes diverse typen dysarthrie.

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Op 1 januari 1969 trad hij in dienst van de Medische Faculteit Rotterdam, waar hij tot heden werkzaam is aan de afdeling Neurologie (Prof. dr. A. Staal) van het Academisch Ziekenhuis Rotterdam-Dijkzigt.