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FOREIGN ACCENT SYNDROME AS A DEVELOPMENTAL MOTOR SPEECH DISORDER

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Abbreviated Title: Developmental FAS

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ABSTRACT

Introduction: Foreign Accent Syndrome (FAS) is a relatively rare motor speech disorder in which the pronunciation of a patient is perceived by listeners of the same language community as distinctly foreign. FAS has been well-documented in adult patients with etiologically heterogeneous, though mostly vascular brain lesions affecting the motor speech network of the language dominant hemisphere. In addition, reports exist of adult patients in whom FAS was due to a psychiatric illness. Although FAS has been reported in children, such accounts are rare and have remained largely anecdotal in that there have been no formally documented cases of FAS as a developmental motor speech disorder. **Methods and Results:** For the first time, we describe the clinical, cognitive and neurolinguistic findings in two patients who in the absence of a history of psychiatric illness or acquired brain damage already presented with FAS at an early stage of speech and language development. In the first patient “developmental FAS” was associated with a dysharmonic distribution of neurocognitive test results indicating slight underdevelopment of visuo-spatial skills and visual memory. The second patient presented with “developmental FAS” associated with specific language impairment (SLI). Independent support for a diagnosis of FAS in both patients was obtained in an accent attribution experiment in which groups of native speakers of (Belgian) Dutch assessed the type of foreign accent of a sample of the patients’ conversational speech. Both patients were judged as non-native speakers of Dutch by the majority of participants who predominantly identified the accent as French. **Conclusion:** This paper for the first time documents two patients who presented with FAS on a developmental basis. The finding that FAS does not only occur in the context of acquired brain damage or psychogenic illness but also exists as developmental motor speech impairment requires a re-definition of FAS as a clinical syndrome.

Keywords: Foreign Accent Syndrome, Specific Language Impairment, Apraxia of Speech, Language, Motor Speech, Cognition

1. INTRODUCTION

The condition in which listeners of the same language community perceive a motor speech disorder as a foreign speech accent was first described in 1907 by the French neurologist Pierre Marie in a patient who developed a regional accent when recovering from anarthria following a subcortical left hemisphere stroke. Since then, more than 60 adult patients with a wide variety, mostly vascular, etiologies have been described who presented FAS either in isolation or, more commonly, in association with other speech and language disorders such as aphasia, apraxia of speech (AoS) or dysarthria. In the majority of patients, FAS resulted from lesions in the perisylvian speech regions involving the prerolandic motor cortex (BA 4), the frontal motor association cortex (BA 6 or 44) or the striatum (Dankovicova et al., 2001). Only few adult patients have been described who developed FAS in the absence of structural brain damage (e.g. Critchley, 1962; Moonis et al., 1996; Coelho and Robb, 2001; Hwang et al., 2001; Reeves and Norton, 2001; Van Borsel et al., 2005; Verhoeven et al., 2005; Laures-Gore et al., 2006; Ryalls and Whiteside, 2006; Poulin et al., 2007; Reeves et al., 2007). In most of these cases FAS resulted from a psychogenic cause (psychogenic FAS).

A full hundred years of multidisciplinary research has neither been able to identify the pathophysiological substrate of this syndrome nor to identify a coherent system in the speech errors that may separate FAS unambiguously from AoS (anarthria, verbal apraxia, speech apraxia) or ataxic dysarthria. It has to be conceded that FAS, AoS and ataxic dysarthria share a slow, monotonous, staccato, scanned, indistinct, remarkably irregular, jerky, explosive, slurred, and laboured verbal output. On the basis of this close semiological resemblance it has been argued that FAS is a sub-type of AoS (Whiteside and Varley, 1998). In addition, a direct link of FAS and AoS with cerebellar speech pathology is also suggested by earlier terminology for FAS - '*ataxia* of the prosody faculty' (Monrad-Krohn, 1947) - as well as for AoS - '*ataxic* aphasia' or '*cortical dysarthria*' (Whitty, 1964). Since the motor speech symptoms of these conditions basically result from distorted articulatory planning and coordination processes, it has been hypothesized that the

cerebellum may also be crucially implicated in the pathophysiology of FAS and AoS (Whitaker, 1982; Cole, 1971; Mariën et al., 2006; Mariën and Verhoeven, 2007).

Although FAS has been well documented in adult patients, reports of FAS as an acquired motor speech disorder following structural brain damage are very rare in a pediatric population and have remained largely anecdotal (Jha, 2007). In addition, there are to the best of our knowledge, no formal reports in which FAS is described as developmental motor speech planning disorder. This paper is the first description of the clinical, cognitive and neurolinguistic findings in two patients who did not acquire FAS in a context of structural brain damage or psychiatric illness but who already presented FAS in an early stage of speech and language development. In the first (adult) patient, FAS was noted in association with developmental AoS, while the second (pediatric) patient presented with FAS in association with specific language impairment (SLI).

2. CASE REPORTS

2.1. Case 1 (TL)

2.1.1. Clinical history

TL is a 29-year-old right-handed woman who is a native speaker of (Belgian) Dutch (Verhoeven, 2005). She consulted the neurological department because of a ‘strange pronunciation’ that had been characteristic of her speech since early childhood. Apart from the perceptual impression of a foreign accent, developmental milestones were unremarkable. Medical history was not contributive. She was born at term after normal gestation and labor and there had been no perinatal or postnatal problems. Scholarly achievements had always been average and there was no family history of developmental language problems or learning disability. This patient’s parents and siblings were monolingual speakers of (Belgian) Dutch. She had an educational level of 14 years and worked as a secretary. At school she had learned French, English and German, but she only seldom used these languages and their use was restricted to professional contexts. After careful

heteroanamnestic inquiry, no indications were found for an acute onset of deviant speech features: close relatives reported that the foreign accent had always been characteristic of her speech and had not changed over time. Neurological examination was unremarkable. Brain MRI and an EEG were normal.

2.1.2. Neuropsychology and Neurolinguistics

Formal assessment of handedness by means of the Edinburgh Inventory (Oldfield, 1971) showed a strong and consistent right hand preference, reflected by a laterality quotient of +100. The Wechsler adult intelligence scale (WAIS-III; Wechsler, 1997) revealed a normal total intelligence quotient (IQ) of 88 with a discrepancy of 16 points between the verbal (VIQ=97) and performance level (PIQ=81; -1.2 SD) (Table 1). At the performance level, deviant scaled scores (≥ -2 SD) were obtained for ‘picture completion’ and ‘picture arrangement’. The Wechsler Memory Scale-Revised (Wechsler, 1987) was also characterized by a significant discrepancy between the verbal (=100) and visual memory index (=74; -1.7 SD). Low scores were obtained on verbal and visual working memory tasks (digit span). The Rey-Osterrieth figure (Osterrieth, 1944) as well as the copying tasks of the Birmingham Object Recognition Battery (BORB) (Riddoch and Humphreys, 1993) and Hierarchic Dementia Scale (HDS) (Cole and Dastoor, 1987) were normal. The ability to conceptually rearrange pictures was unimpaired (Hooper, 1983). Visual object identification and visual semantics were normal (Riddoch and Humphreys, 1993; Mariën et al., 1998). As demonstrated by the Wisconsin Card Sorting Test (Heaton et al, 1993), frontal planning and problem solving were normal. The ability to inhibit a competing and more automatic response set was also normal (Golden, 1978). On tasks evaluating visual search and sequencing the patient scored in the lower range (Reitan, 1958). Mood and behaviour were normal. No indications were found for mood or behavioural abnormalities during the clinical assessments and careful inquiry of the patient’s close relatives confirmed these findings. Examination of bucco-labio-lingual praxis

by means of a Dutch checklist for oral-verbal dyspraxia (Erlings-van Deurse et al., 1993) did not reveal any abnormalities. Formal language investigations by means of standardized test batteries and a semantic and phonological verbal fluency task (unpublished norms) were well within the normal range (Graetz et al, 1992; De Renzi and Vignolo, 1964; Mariën et al., 1998).

[INSERT TABLE 1]

2.2. Case 2 (KL)

2.2.1. Clinical History

KL, a 7-year-old right-handed boy diagnosed at the age of five with SLI was referred to our department by his speech and language therapist because only little therapeutic progress had been made over the two years of treatment. Except for a delayed onset (around age 3) and subsequently deviant development of expressive language skills, developmental milestones were normal. He was born at term after normal gestation and labour, and there had been no perinatal or postnatal problems. Medical history was unremarkable. Both parents were monolingual speakers of Dutch. No family history of developmental disorders or learning disabilities was reported. He successfully attended the first grade of primary school and his scholastic achievements were above average. Repeat neurological examinations, including awake and sleep EEG recordings, were normal. Repeat MRI of the brain did not disclose any structural abnormality. A Tc-99m-ECD SPECT perfusion scan was performed. Trans-axial images with a pixel size of 3.56 mm were anatomically standardized using SPM and compared to a standard normal and SD image obtained from 15 normal ECD perfusion studies. Using a 31 ROI template the Z-scores (SD) were then calculated for each region. A regional Z-score of >2.0 is considered significant. In comparison to normal database findings a significant decrease of perfusion was found in the vermis (-1.96 SD), both lentiform nuclei (left -2.25; right -2.30 SD) and the left thalamus (-3.30 SD). In addition a significant bilateral occipital hypoperfusion was observed.

2.2.2. Neuropsychology and Neurolinguistics

A strong and consistent right hand preference was confirmed by the Edinburgh Inventory (Oldfield, 1971) which yielded a laterality quotient of +100. As reflected by a total WISC-III IQ of 124, a verbal IQ of 115 and a performance IQ of 128, assessment of general intelligence was normal (Wechsler, 2002) (Table 2). At both the verbal and the performance level the subtest scores were well within the normal range. In addition, the neuropsychological test battery did not disclose any indications of a cognitive deficit within the nonverbal domain. As demonstrated by normal results on the WISC-III performance subtests, the HDS subtests and the Rey-Osterrieth figure, visuo-spatial and visuo-constructive skills were normal. Ideomotor and ideational praxis were also intact. Normal results on the Dutch checklist for oral-verbal dyspraxia (Erlings-van Deurse et al., 1993) indicated that the volitional control of nonspeech oral movements was normal. Orientation, visual, tactile and finger gnosis, spatial and verbal problem solving, concentration, working memory and verbal memory (PINOK, Vieijra et al., 1992) were normal. However, neurolinguistic examinations revealed a nonfluent speech disorder characterized by phonemic paraphasias, repetition difficulties at the syntactic level (AAT sentence repetition) and expressive agrammatism. As shown in table 2, the patient obtained normal results at the expressive level for visual confrontation naming (BNT and Peabody Picture Vocabulary Test-III-NL (Dunn, 2005)) and repetition of phonemes, monosyllabic words and compounds (AAT). Subtest results of the Taaltest voor Kinderen (Language Test for Children) (TvK) (Van Bon and Hoekstra, 1982) revealed that morphosyntactic and syntactic processes were distorted at the expressive level. In spontaneous speech also, the inappropriate use of infinitives and the lack of finite verbs, auxiliary verbs, subordinate clauses, function words and articles resulted in a telegraphic style. By contrast, formal investigations of receptive language (TvK) and written language skills (Dudal, 1998) were normal.

[INSERT TABLE 2]

3. ACCENT ATTRIBUTION

External support for the diagnosis of FAS in both patients was obtained in an accent attribution experiment in which 123 native speakers of Dutch (42 MA level psychology students, 37 MA speech and language pathology students and 44 specialist teachers of Dutch as a Foreign Language) assessed the type of foreign accent of a sample of both patients' conversational speech. The patients' speech samples were incorporated amongst other speech samples of five non-native speakers of Dutch from different linguistic backgrounds (English, French, German, Asian), four other FAS patients and five native speakers of Dutch who served as controls. Some of the FAS speakers have been described in detail in Verhoeven et al. (2005) and Mariën & Verhoeven (2007). The speech samples that were used in the experiment were obtained in informal interviews with the subjects. They were asked to speak freely about general topics such as their hobbies, holidays, etc. These interviews were recorded by means of a Marantz Solid State Recorder (PMD660) and an AKG head-mounted condensor microphone (CL444). All the recordings were made in a quiet setting without any disturbing background noise. From these recordings, one representative speech sample was selected for each speaker. From a content point of view, care was taken that the speech samples did not contain any indications about the speakers' nationality or linguistic background. The duration of the individual speech samples varied between 0'25'' and 1'25''. After recording, the speech samples were digitized with a sampling frequency of 44,100 Hz by means of the signal processing package PRAAT (Boersma & Weenink, 2007). The speech samples of all the speakers were assembled into a digital tape. The sequential order of the samples was pseudo-random: consecutive FAS samples or consecutive control accents of the same type were not allowed. In order to give the listening panel enough time to complete an accent-assessment form for the speakers, each speech sample was repeated twice. In the accent attribution task, listeners had to freely write down the perceived accent. The speech samples were played to the judges in three different sessions which were organised at their respective

institutions. First, the judges read the instructions to the test and provided information about their sociological background. Then participants heard two practice speech samples to familiarize themselves with the task. These samples did not re-occur in the real test and the listening panel's judgements on these were not included in any way in the results. After the practice sessions, the tape was stopped for questions. Subsequently, the speech sample of each speaker was played to the listeners twice. After that, listeners were given 3 minutes to complete the questionnaire for the corresponding speech sample. Since the details of this experiment will be reported elsewhere, only the results of TL and KL will be discussed with respect to the other FAS speakers.

In order to visualise the strength of the association between the FAS speakers in the accent attribution test and the actually attributed accents, a correspondence analysis was carried out (Clausen, 1998). Correspondence analysis is a technique which analyses “the association between two or more categorical variables by representing the categories of the variables as points in a low-dimensional space. Categories with similar distributions will be represented as points that are close in space, and categories that have very dissimilar distributions will be positioned far apart” (Clausen, 1998: 2). In the present analysis the first categorical variable is constituted by the different FAS-speakers (patients TL and KL inclusive), while the second categorical variable represents the different accents attributed by the judges in the listening panel. The results of this analysis are given in figure 1.

[INSERT FIGURE 1]

In figure 1, the horizontal axis (c2) separates the individual FAS speakers i.e. the further they are apart on the plot, the more dissimilar they are in terms of accent attribution. The vertical axis represents the different attributed accents. As a result of this, the distances between the different FAS-speakers (crosses) and the accents (squares) represent the strength of association between the individual speakers and the attributed accents. Thus it can be seen that the listening panel has

generally attributed a wide variety of accents to the FAS speakers (FAS 1-4). However, the correspondence analysis indicates differential associations between individual FAS speakers and the attributed accents. Speaker FAS 1 is most strongly associated with a Moroccan accent, while FAS 2 has a strong association with English and Dutch (Holland). FAS 3 is mainly identified as Spanish while FAS4 has the strongest association with German and Indian. Patients TL and KL are most strongly identified as having a French accent by the members of the listening panel. In terms of percentages of accent attribution, patient TL was judged as a non-native speaker of Dutch by the majority of the listeners (62.49%). The different foreign accents attributed to the patient's speech varied with a clear dominance of French: French accounted for 48.13 % of the judgements. The other foreign accents attributed were German (7.00%), Scandinavian (2.57 %), Moroccan (1.77%), Southern European (1.77 %), English (0.80%) and Eastern European (0.80%). In a considerable number of cases, TL was also identified as a speaker of (Belgian) Dutch (37.16 %). Patient KL was considered as a non-native speaker of Dutch by 56.4%. The foreign accents attributed here varied with a clear dominance of French: French accounted for 45.77% of the judgements. The other foreign accents attributed were Moroccan (6.40%), Southern European (1.41%), African (1.41%), South american (1.41%) and Asian (1.41%). It was noted that this patient was considered as a native speaker of (Belgian) Dutch by 43.60% of the participants in the listening panel.

4. PHONETIC CHARACTERISTICS

Three minutes of spontaneous speech of patient TL were subjected to detailed phonetic error analysis, while patient KL's speech was perceptually assessed on the basis of a 2.5 minute sample of his spontaneous speech. These were the speech samples a section of which had been taken for perceptual assessment by the listening panel. In the first instance, all the words in the samples of both patient TL and patient KL were transcribed orthographically by a neurolinguist (PM) and an

experienced phonetician (JV), who worked independently of each other. Subsequently, both researchers indicated the words in the samples the pronunciation of which they considered to deviate from the pronunciation norms of (Belgian) Dutch. On the basis of these assessments, an interrater reliability measure was calculated. For patient TL, Cohen's Kappa (Cohen, 1960) amounted to 0.94, while for patient KL Kappa was 0.96. Thus, there was excellent agreement between both raters as to the words in both speech samples which were realised with a deviant pronunciation.

In the second stage of the analysis, the words on which there was agreement between the two raters concerning their deviant pronunciation were further assessed perceptually by the phonetician, who noted the errors.

4.1. Patient TL

In terms of consonant articulation, it was found that the propositus had a clear palatal articulatory setting (Laver, 1980), whereas this setting tends to be velar in Standard Dutch (Honikman, 1964; Verhoeven, 1994). As a result, the velar fricative sounds normally articulated at the back of the oral cavity such as /x/ and /V/ were realised too anteriorly as pre-velar or palatal. It was furthermore noted that the alveolar sounds /s, t, d, l, n/ were most often pronounced too anteriorly as dental, i.e. as [s^l, t^l, d^l, l^l, n^l]. The trill sound was consistently realised as a uvular trill. Besides this, the patient's speech contained inconsistent consonant deletions, which resulted in consonant cluster simplification (v'RtROk' > v'tROk' 'left'; vo...Rlo...p'R > vo...lo...p'R 'predecessor'), instances of consonant insertion (InVe...#PRslAc's > InVe...#PRsklAc's 'slide rulers'), instances of anticipation (a...vOt/e...t' V'no...m' > a...vOtxe...t' V'no...m' 'had dinner'), instances in which a voiceless sound was erroneously voiced (IntR'sAnt > IntR'zAnt 'interesting'). Furthermore, clear prolongation of the duration of the trill was sometimes noted

(o...v'RHAndIV' > o...v'R.....HAndIV' 'to hand over'). Finally, syllable contraction was observed which seemed to result from articulatory breakdown (V'1Yk > VYk 'happiness').

As far as the articulation of vowels is concerned, there was little quality difference between the high front vowels /i/, /e/ and /I/ and one instance of a vowel substitution (v'Rza...m'la...R v'Rza...m'le...R 'collector') was observed. In addition, most vowels were characterized by a strong onset with creaky voice. In terms of voice quality, an extremely tense laryngeal setting was noted which caused the voice to sound generally metallic. As far as prosody is concerned, speech rate was normal (3.89 syllables/second) as compared to benchmark figures for Dutch in Verhoeven et al. (2004). Speech rate was quantified as articulation rate, i.e. the average rate across measured interpause stretches. A high incidence of pausing was noted. Sentence intonation contours were well-formed although there was one instance of wrong sentence stress placement. Speech rhythm was perceptually assessed to be syllable-timed and staccato rather than the expected stress-timing in Dutch. Finally, instances of self-correction (re...k'nmAS ... re...k'nmASin' 'calculator'), regular facial grimaces and groping behaviour were found.

If we have to relate the observed errors to the perception of a French accent by the majority of the listening panel, the contributing factors are the erratic pausing behaviour, the use of a uvular trill, the palatal articulatory setting, the minimal distinctions between the high front vowels /i/, /e/ and /I/, and the syllable-timed, staccato speech rhythm.

4.2. Patient KL

Phonetic error analysis of a spontaneous speech sample of patient KL showed a number of deviant speech patterns which are generally considered typical for French learners of Dutch such as a very consistent and pronounced uvular /r/, the consistent replacement of word-initial /h/ by glottal stops and the lack of a clear distinction between the vowels /i/ and /I/. In addition to this,

the patient had a palatal articulatory setting which is also typical for French, not for Dutch. The alveolar fricative /s/ was more often than not realised too anteriorly. In addition, a number of sound deletions were noted /na...R bIne > na... bIne 'inwards'). As mentioned above, the patient's language was characterized by grammatical errors, which may have contributed significantly to the impression of a foreign accent.

As far as KL's prosody was concerned, articulation rate amounted to 2.14 syllables/second, which is considerably lower than the benchmark figures in Verhoeven et al. (2005). No abnormalities were found in word accentuation, sentence intonation and speech rhythm were normal.

5. DISCUSSION

This paper describes two patients with a clear history of developmental speech and language impairment presenting clinical characteristics consistent with FAS. Indeed, in both patients, phonetic analyses revealed deviant motor speech patterns which were perceived as a foreign accent by the majority of listeners of the same language community in an accent attribution experiment. The accent of both patients was predominantly identified as French and this is consistent with the kind of errors identified in their spontaneous speech.

Although at least 60 FAS patients have been reported in the literature, the cases described here are unique for several reasons. Firstly, these patients did not acquire FAS following acute or chronic brain injury and this is unlike all other patients reported in the literature so far (except for the psychogenic patients): MRI of the brain did not reveal any evidence for a structural brain lesion at the supratentorial or infratentorial level. In addition, there is no evidence for a progressive course or exacerbations of the motor speech symptoms and no indications were found for a psychogenic cause. Secondly, as confirmed by carefully obtained heteroanamnestic information from near relatives both these patients presented with consistent foreign accent speech

qualities since early speech and language development and this has never been formally reported before. There only have been anecdotal reports of FAS in a pediatric population which mirror the incidence of FAS in the adult population, i.e. FAS as a result of brain damage. A recent example of this is one of an English boy who ‘lost his strong Yorkshire accent’ and who started ‘speaking the Queen’s English’ after an operation for a subdural empyema (Jha, 2007). In addition to this patient, there have been informal accounts of FAS in autistic spectrum disorder in which the foreign accent qualities were related to unusual prosody (Peppé, 2007 personal communication). Arguably, such patients may fall within the developmental spectrum of FAS.

From a semiological point of view the deviant speech symptoms of patient TL seem to suggest that her FAS may be closely related to AoS. This is suggested by a convincing number of typical AoS features in this patient’s speech such as the inconsistency in speech sound errors, the difficulty in sequencing speech movements, the deletion of consonants in clusters, the contraction of syllables following articulatory breakdown, the extremely tense laryngeal setting, the high incidence of pausing, the wrong placement of sentence stress, the syllable-timed and staccato speech rhythm, the self-corrections, the regular facial grimaces and groping behaviour. The association between AoS and FAS has been frequently observed in adult patients and research indicates that FAS may well be considered as a sub-type of AoS (Whiteside and Varley, 1998). Patient KL on the other hand presented with FAS in the context of a developmental nonfluent output disorder consistent with a diagnosis of SLI of the phonological-syntactic type (Allen et al., 1986). In addition to expressive agrammatism and disrupted morphosyntactic skills, phonemic paraphasias as well as articulatory errors disrupted the patient’s oral-verbal output at the phonological and syntactic level. A possible association between FAS and SLI has not been reported before. Despite the necessity of functional neuroimaging findings to unravel the neurobiological mechanisms underlying developmental speech and language disorders, functional neuroimaging studies in the literature on SLI are scarce (Lee et al. 2002; Ors et al., 2005; Hwang

et al., 2006; Im et al., 2007). Methodological limitations do not currently allow sound conclusions but SPECT and PET studies have shown that in the absence of structural brain abnormalities SLI children may frequently present with abnormal metabolic and RCB findings especially at the subcortical level. A quantified Tc-99m-ECD SPECT perfusion study in our SLI patient KL confirmed a significantly decreased perfusion which was most pronounced at the subcortical level. As a result, our findings further support the hypothesis that subcortical structures might constitute a crucial factor in the aetiology of SLI (Ors et al., 2005; Im et al., 2007). The significantly decreased perfusion in the vermis might add to the hypothesis that the cerebellum is crucially implicated in neural network of articulatory planning and as such also in FAS (Whitaker, 1982; Cole, 1971; Mariën et al., 2006; Mariën and Verhoeven, 2007).

At the neurocognitive level, a consistent asymmetry in the cognitive profile of patient TL - characterized by significantly lower results in the nonverbal domain- seems to indicate that developmental problems extended beyond the purely articulatory level. Although they did not have any significant scholastic or clinical repercussions, “underdevelopment” of visuo-perceptual problem solving skills and visual memory was found. Data on neurocognitive functions are only exceptionally reported in the FAS literature and the association of domain specific nonverbal difficulties and FAS has not been reported as an essential aspect of FAS. By contrast, cognitive investigations in patient KL revealed that the developmental problems were strictly confined to the oral-verbal domain.

The observations in this paper require the traditional definition of FAS as an essentially acquired motor speech disorder to be revised to include patients with a developmental speech and language pathology. They furthermore suggest that two entirely different origins of FAS (acquired versus developmental) may lead to the same impression of foreignness in listeners.

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Table 1: Neurocognitive test results of patient TL

NEUROCOGNITIVE TESTS	<i>Score / Maximum (Scaled Score)</i>	<i>Percentile</i>	<i>Mean</i>	<i>±1SD</i>
<i>Mini Mental State Examination</i>	30/30	-	29	1.3
<i>Intelligence</i>				
Wechsler Global IQ (GIQ)	88	-	100	15
Wechsler Verbal IQ (VIQ)	97	-	100	15
Wechsler Performance IQ (PIQ)	81	-	100	15
<i>Memory</i>				
Wechsler Memory Scale (WMS-R)				
Visual Memory Index	74	-	100	15
Verbal Memory Index	100	-	100	15
Global Memory Index	91	-	100	15
- Information	6 / 6	-	-	-
- Orientation	7 / 7	-	-	-
- Mental Control	6 / 6	-	-	-
- Logical Memory (A+B)	28 / 43	57	26	-
- Visual Paired Associates (I-III)	6 / 18	-	-	-
- Verbal Paired Associates (I-III)	19 / 24	-	-	-
- Visual Reproduction	31 / 41	29	33-34	-
- Digit Span Forward	6 / 12	12	-	-
- Digit Span Backward	5 / 12	26	-	-
- Visual Memory Span Forward	7 / 14	22	-	-
- Visual Memory Span Backward	6 / 12	14	-	-
Delayed Recall Index	90	-	100	15
- Logical Memory (A+B)	27 / 43	72	22	-
- Visual Paired Associates	2 / 6	-	-	-
- Verbal Paired Associates	7 / 8	-	-	-
- Visual Reproduction	28 / 41	27	31	-
Hierarchic Dementia Scale (HDS)				
Memory : Biographic : it. 17	10 / 10	-	10	0
<i>Language</i>				
Token Test	0 errors / 50	-	2.28	2.75
Boston Naming Test (BNT)	49 / 60	-	52.8	3.7
Verbal Fluency				
Semantic Fluency	58	-	54.9	3.51
- Animals, 1 minute	19	-	-	-
- Transportation, 1minute	12	-	-	-
- Vegetables, 1 minute	12	-	-	-

- Clothing, 1 minute	15	-	-	-
Total number of perseverations	0	-	-	-
Total number of intrusions	0	-	-	-
Phonological Fluency	32	-	42.6	13.01
- Phoneme F, 1 minute	5	-	-	-
- Phoneme A, 1 minute	10	-	-	-
- Phoneme S, 1 minute	17	-	-	-
Total number perseverations	0	-	-	-
Total number intrusions	0	-	-	-
<i>Executive Functioning</i>				
Wisconsin Card Sorting	6/64	-	5	-
Stroop Colour-Word Test				
Card I	60"	15	48"	-
Card II	66"	40	63"	-
Card III	89"	60	99"	-
Tower of Hanoi	23 moves 0 errors	-	-	-
Trail Making				
Part A	38"	40	32	-
Part B	89"	30	69	-
<i>Praxis</i>				
Rey-Osterrieth Figure	33 / 36	-	35	3
HDS Ideational: item 5	10 / 10	-	9.79	0.17
HDS Ideomotor: item 3	10 / 10	-	9.94	0.23
HDS Drawing: item 15	10 / 10	-	9.81	0.52
HDS Constructional: item 12	10 / 10	-	10	0
<i>Visual Perception</i>				
Judgment of Line Orientation	28 / 30	-	25.3	-
Hooper VOT	24 / 30	-	26	5
Birmingham Object Recognition Battery				
- Copying	normal	-	-	-
- Length match task - A	24 / 30	-	26.9	1.6
- Size match task - A	23 / 30	-	27.3	2.4
- Orientation match task - A	22 / 30	-	24.8	2.6
- Position of gap match task - A	39 / 40	-	35.1	4.0
- Minimal feature match	25 / 25	-	23.3	2.0
- Foreshortened match	25 / 25	-	21.6	2.6
- Object decision	25 / 32	-	27.0	2.2
- Item match	32 / 32	-	30	2.2
- Association match	30 / 30	-	27.5	2.4

Legend: - = no normative data or values available

Table 2: Cognitive Test Results of patient KL

NEUROCOGNITIVE TESTS	<i>Score / Maximum</i>	<i>Percentile</i>	<i>Mean</i>	<i>±1SD</i>
Intelligence (WISC-III)				
Global IQ	124	-	100	15
Verbal IQ	115	-	100	15
Performance IQ	128	-	100	15
Language				
Boston Naming Test	40 / 60	-	33.0	4.8
Taaltest voor Kinderen				
Auditory Synthesis	21 / 29	69	-	-
Auditory Discrimination	36 / 47	71	-	-
Morphology	15 / 36	15	-	-
Semantics	26 / 51	79	-	-
Syntax	8 / 27	2	-	-
Peabody Picture Vocabulary Test III	103	75	-	-
Word Comprehension Quotient	110	-	100	15
Aachener Aphasia Test – Repetition	134 / 144	-	144.1	8.07
Phonemes	30 / 30	-	28.91	2.09
Monosyllabic words	28 / 30	-	29.22	1.32
Loan- & foreign words	28 / 30	-	28.94	2.31
Compounds	26 / 30	-	28.45	2.22
Sentences	22 / 30	-	28.55	1.90
Reading & Dictational Writing				
Word Reading	42 / 120	80	-	-
Dictational Writing				
letters	20 / 20	60	-	-
words	20 / 20	90	-	-
sentences	20 / 20	80	-	-
Verbal Memory				
PINOK: 15 words	40/75	86	32	-
Praxis				
Rey-Osterrieth Figure	33 / 36	-	35	3
HDS Ideational: item 5	10 / 10	-	9.79	0.17
HDS Ideomotor: item 3	10 / 10	-	9.94	0.23
HDS Drawing: item 15	10 / 10	-	9.81	0.52
HDS Constructional: item 12	10 / 10	-	10	0

Legend: - = no normative data or values available