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The Posterior Fossa and Foreign Accent Syndrome: Report of Two New Cases and Review of the Literature

Stefanie Keulen^{1,2} · Peter Mariën^{1,3} · Kim van Dun¹ · Roelien Bastiaanse² ·
Mario Manto⁴ · Jo Verhoeven^{5,6}

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Abstract Foreign accent syndrome is a rare motor speech disorder that causes patients to speak their language with a non-native accent. In the neurogenic condition, the disorder develops after lesions in the language dominant hemisphere, often affecting Broca's area, the insula, the supplementary motor area and the primary motor cortex. Here, we present two new cases of FAS after posterior fossa lesions. The first case is a 44-year-old, right-handed, Dutch-speaking man who suffered motor speech disturbances and a left hemiplegia after a pontine infarction. Quantified SPECT showed a bilateral hypoperfusion in the inferior lateral prefrontal and medial inferior frontal regions as well as a significant left cerebellar hypoperfusion. Further clinical investigations led to an additional diagnosis of brainstem cognitive affective syndrome which closely relates to Schmahmann's syndrome. The second patient was a 72-year-old right-handed polyglot English man who suffered a stroke in the vascular territory of the left posterior inferior cerebellar artery

(PICA) and developed a foreign accent in his mother tongue (English) and in a later learnt language (Dutch). In this paper, we discuss how the occurrence of this peculiar motor speech disorder can be related to a lesion affecting the posterior fossa structures.

Keywords Foreign accent syndrome · Posterior fossa · Cerebellum · SPECT

Introduction

Foreign Accent Syndrome

During the last century, more than 170 reports of patients with foreign accent syndrome (FAS) have been published but the disorder is probably much more frequent than currently estimated. Due to speech changes at the segmental (speech segments, e.g., phonemes) and suprasegmental level (contrastive aspects marking speech that are not identified as segments, but rather define the segments, e.g., rhythm, intonation), this rare condition causes the patient with FAS to be perceived by speakers of the same language community as a non-native speaker. FAS has been related to various etiologies including stroke, cerebral trauma, multiple sclerosis, dementia, and psychiatric disorders. In 2010, Verhoeven and Mariën [1] distinguished three different sub-types of FAS: neurogenic FAS [2], including developmental cases [3], but also psychogenic FAS [4] and a mixed-type FAS [5].

Here, we focus on the neurogenic type, which has been the most widely reported variant of FAS. Although most of the patients with this subtype acquired FAS after stroke [e.g., 2; 6, case 2; 7, 8, 9, case 2] or brain trauma [e.g., 10; 11, case 2; 12, cases 4 & 5], the lesion sites do not unequivocally point towards a single neuroanatomical substrate. In the majority of the

✉ Peter Mariën
peter.marien5@telenet.be

¹ Clinical and Experimental Neurolinguistics, CLIN, Vrije Universiteit Brussel, Brussels, Belgium

² Center for Language and Cognition Groningen (CLCG), Rijksuniversiteit Groningen, Groningen, The Netherlands

³ Department of Neurology and Memory Clinic, ZNA Middelheim Hospital, Lindendreef 1, B-2020 Antwerp, Belgium

⁴ Unité d'Étude du Mouvement (UEM), FNRS, ULB Erasme, Brussels, Belgium

⁵ Department of Language and Communication Science, City University, London, UK

⁶ Computational Linguistics and Psycholinguistics Research Center (CLIPS), Universiteit Antwerpen, Antwerp, Belgium

vascular cases, the causative lesions were located in the language dominant (left) hemisphere, especially in the frontal [e.g., 13, case 3; 14; 15], fronto-temporal [e.g., 16–18], and fronto-temporo-parietal regions [10, 19, 20]. Although handedness was not formally investigated, “crossed FAS” has been reported in three dextral patients after right-hemisphere damage [21, case 3; 13, case 2; 22, 23]. As evidenced by SPECT, the cerebellum was functionally affected (hypoperfusion) in three cases after frontal lesions (mechanism of diaschisis) [2, 9, case 2, 24]. In five other cases, FAS was directly attributed to posterior fossa lesions [6, cases 1 and 2; 23, 25–27].

In this study, we present a survey of the literature and two new cases of FAS after a posterior fossa lesion: the first patient suffered a stroke in the right pons and has been described as an example of brainstem cognitive affective syndrome by Mariën and D’aes [28]. The second patient incurred FAS after a stroke in the left posterior cerebellum.

Posterior Fossa and FAS: Two New Cases

Case 1

A 44-year-old, right-handed, Dutch-speaking Belgian man with an educational level of 11 years was admitted to the hospital on April 20th, 2012 after an acute loss of consciousness followed by motor speech disturbances and a left hemiplegia. The patient had used “poppers;” alkyl nitrites which, when aspirated, arouse euphoric feelings and relax sphincter muscles to enhance sexual experience. CT and electrocardiography on admission were normal. Five days later, the patient was transferred to the Neurology department of ZNA Middelheim Hospital. Neurological examinations on admission revealed FAS and paralytic dysarthria, a left facial nerve paralysis, tongue protrusion to the left, a left hemiplegia, and a left hypertonia. The patient tested positive for (neurovascular) syphilis (blood and CSF analysis) and was subsequently treated with penicillin for 2 weeks. On admission, the medical staff had the impression that the patient spoke with a Hollandic-like accent. Speech was generally very slow and had a pronounced nasal quality. After he was released from hospital, the patient’s friends also noticed the change in accent. Vowel pronunciation was affected by omissions (especially of schwa) and backing. In addition, diphthongs were often articulated as monophthongs. Consonants were regularly devoiced (esp. fricatives), or omitted (esp. plosives – alveolar and velar). T1-weighted MR imaging of the brain had demonstrated a hypointense lesion at the level of the right pons. T2-weighted axial FLAIR-imaging showed a hyperintense signal in the right pons (Fig. 1).

Quantified 99m Tc ECD SPECT scan in May 2012 showed a bilateral hypoperfusion in the inferior lateral prefrontal (L: -2.2 SD, R: -2.7 SD) and left medial inferior frontal regions

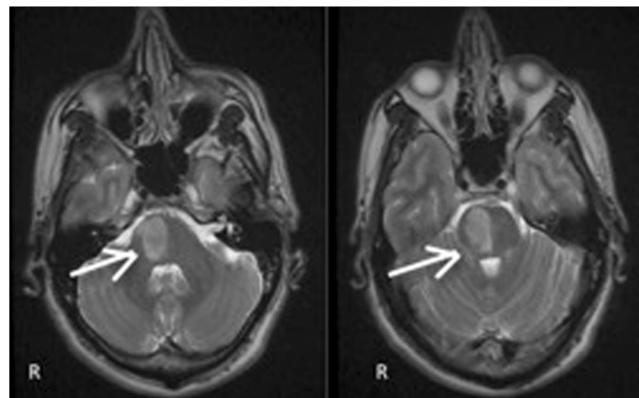


Fig. 1 T2-weighted axial MRI slice of the brain demonstrating an ischemic lesion in the right pons (white arrows)

(-2.8 SD). A significant left cerebellar hypoperfusion was also retained (-2.1 SD) (Fig. 2).

The EEG recording was mildly impaired over both the left and right hemisphere: polyrhythmic activity on the left showed a fast component whereas on the right a slowing of activities was found. Soon after admission, the patient had a grand mal seizure and treatment with valproic acid was started. A transesophageal echocardiogram disclosed mitral insufficiency and a patent foramen ovale with a clear right to left shunt.

Extensive neuropsychological testing was performed in May 2012 (see Table 1). Intelligence, executive and visuospatial functions, memory and frontal behavioural and affective disturbances were investigated. Test results disclosed general cognitive abilities were normal (MMSE: 27/30; -0.5 SD) [29], but the patient had clinically deficient full scale IQ (FSIQ = 73; -1.8 SD) and pathological performance IQ (PIQ = 61, -2.6 SD, with depressed scores for digit symbol substitution (-3 SD), block design (-1.3 SD), matrix reasoning (-1.7 SD), picture completion, (-1.7 SD) and picture arrangement: (-2.7 SD), whereas subtest scores of the verbal IQ were within the normal range (VIQ = 83; -1.1 SD) (Wechsler Adult Intelligence Scale-3rd Edition; [30]). The patient obtained pathological scores on the attention subtests (index 66, -2.3 SD) and delayed recall subtests (index 61, -2.6 SD) of the Wechsler Memory Scale – revised (WMS-R) [31]. Visual memory index amounted to 70 and verbal memory to 80. Index scores on the Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) were normal [32]. Scores on the Trail Making Test were depressed (TMT A: <pc. 10; TMT B: <pc. 10) [33]. On the Wisconsin Card Sorting Test [34], the patient only completed three categories (-2.3 SD; mean 5.58; 1SD = 1.1). A pathological score was found on the Stroop Coloured Word Test (card 1: < pc. 1; card 2: < pc. 1; card 3: pc. 3) [35], and the Frontal Assessment Battery: 13/18 [36]. In summary, the patient demonstrated a borderline pathological full scale IQ, with a significant dissociation between VIQ (-1.1 SD) and PIQ (-2.6 SD), difficulties

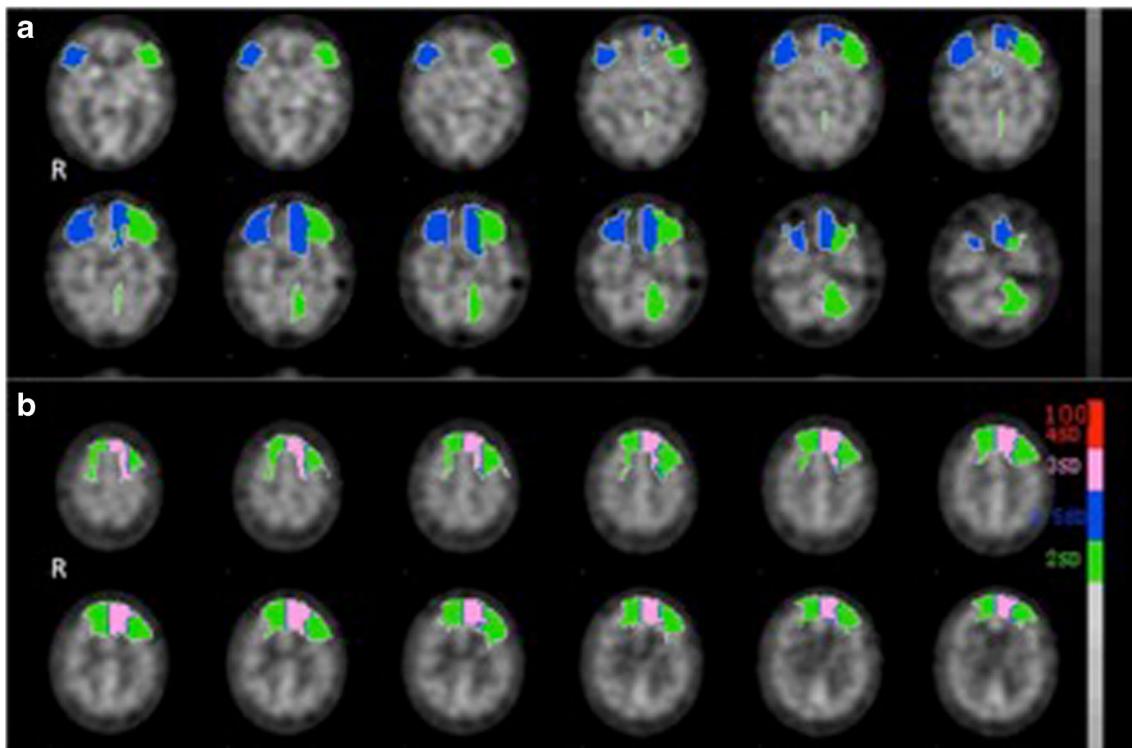


Fig. 2 Quantified ^{99m}Tc ECD SPECT showing a bilateral hypoperfusion in the inferior lateral prefrontal and left medial inferior frontal regions as

well as a significant left cerebellar hypoperfusion (plane a) and a significant bilateral frontal hypoperfusion (plane b)

in set-shifting and maintaining goal oriented strategies (WCST and Stroop test), deficits in visuospatial abilities (TMT-A) and frontal inhibition disturbances (Frontal Assessment Scale).

In October 2012, the patient was re-examined again for intelligence, executive and visuospatial functions, memory, and affective disturbances (same tests as in May 2012). General cognitive abilities were re-tested and MMSE score amounted to 30/30 [29]. Performance IQ was still impaired (PIQ = 65, -2.3 SD). Pathological scores were obtained for digit symbol substitution (-3 SD) and block design (-2 SD) while depressed scores were found for matrix reasoning (-0.7 SD), picture completion (-1.3 SD) and picture arrangement: (-1.3 SD). The verbal IQ (VIQ = 98; -0.1 SD) and full scale IQ (FSIQ = 82; 1.2 SD) were within the normal range. The WMS-R [31] index score for attention remained impaired (68, -2.13 SD) whereas mnemonic capacities had increased significantly: the visual memory index increased to 111, ($+0.73$ SD), the verbal memory index to 101 ($+0.07$ SD); and the delayed recall index to 115 ($+1$ SD). A pathological result was found again for the WCST [34]: the patient completed only three categories. Performance on the Trail Making Test [35] slightly increased to respectively average and low average levels (TMT A < pc. 30; TMT B < pc. 21). A similar observation was made with regard to his performance on the Stroop Colour Word Test: card 1: pc. <25, card 2: < pc. 10; card 3: < pc. 25. Performance on the Frontal Assessment Battery [36] slightly improved to the normal range: 15/18. The patient obtained a defective score of 8 on the Middelheim Frontality

Scale (MFS; cut-off = ≥ 5 , [37]), which confirmed behavioural abnormalities related to apathy, blunting of affect as well as anosognosia. In short, the patient's neuropsychological profile remained stable with respect to executive functions and visuospatial abilities. A dissociation between VIQ (-0.1 SD) and PIQ (-2.3 SD) was still present. However, there was a spectacular increase in mnemonic functioning (the patient now obtained above average scores). The clinical profile was consistent with a diagnosis of brainstem cognitive affective syndrome [28].

Neurolinguistic assessments were carried out to further evaluate speech and language functioning. Results revealed that the patient did not suffer from aphasia. The Comprehensive Aphasia Test (Dutch Version - CAT-NL) [38] showed normal results. For the copying of words, letters were poorly formed, though writing was still legible. The Test for Systematic Detection of Graphomotor Impairment (Systematische opsoring schrijfmotorische problemen (systematic detection of graphomotor problems; SOS-test) [39] was administered and revealed aberrant writing skills in May 2012 (score 8, -4.8 SD). The scores were indicative of an apraxic agraphia. The score was still depressed after retesting in October of the same year (score 5, -2.5 SD).

Case 2

Case 2 is a 72-year-old right-handed polyglot English man, who was admitted to hospital in October 2012 after an acute episode

Table 1 Overview of the neuropsychological investigations: case 1

General	May 2012 Score ($\pm 1SD$ or pc.)	June 2012 Score ($\pm 1SD$ or pc.)	October 2012 Score ($\pm 1SD$ or pc.)	Maximum or # trials [range]	Mean (SD)
Mini Mental State Examination	27 (−0.5)		30 (+0.8)	30	28 (1.9)
WAIS					
Wechsler Full Scale IQ (FSIQ)	73 (−1.8)		82 (−1.2)		100 (15)
Wechsler verbal IQ (VIQ)	83 (−1.1)		98 (−0.1)		100 (15)
Information	11 (+0.3)		9 (−0.3)		10 (3)
Comprehension	4 (−2.0)		10 (0.0)		10 (3)
Digit span	5 (−1.7)		11 (+0.3)		10 (3)
Arithmetic	7 (−1.0)		9 (−0.3)		10 (3)
Similarities	8 (−0.7)		10 (0.0)		10 (3)
Vocabulary	8 (−0.7)		12 (+0.7)		10 (3)
Wechsler Performance IQ (PIQ)	61 (−2.6)		65 (−2.3)		100 (15)
Digit symbol substitution	1 (−3.0)		1 (−3.0)		10 (3)
Picture completion	5 (−1.7)		6 (−1.3)		10 (3)
Block design	6 (−1.3)		4 (−2.0)		10 (3)
Picture arrangement	2 (−2.7)		6 (−1.3)		10 (3)
Matrix reasoning	5 (−1.7)		8 (−0.7)		10 (3)
Memory (WMS-R)	66 (−2.3)	66 (−2.3)	68 (−2.1)		100 (15)
Visual memory	70 (−2.0)	80 (−1.3)	111 (+0.7)		100 (15)
Verbal memory	80 (−1.3)	90 (−0.7)	101 (+0.1)		100 (15)
General memory	73 (−1.8)		104 (+0.3)		100 (15)
Delayed recall	61 (−2.6)	83 (−1.1)	115 (+1.0)		100 (15)
Executive functions					
Wisconsin Card Sorting Test	3 (−2.3)		3 (−2.3)	(64) 128 trials	5.58 (1.1)
Verbal fluency					
Semantic fluency	40 (−1.1)		49 (−0.5)		56.1 (15.1)
Stroop Colour-Word Test					
Card I	126" (< pc. 1)		62" (pc. 15)		
Card II	163" (< pc. 1)		80" (pc. 10)		
Card III	188" (pc. 3)		128" (pc. 15)		
Trail Making Test					
Part A	94" (< pc. 10)		41" (pc. 30)		
Part B	223" (< pc. 10)		99" (pc. 25)		
Behavioural					
Frontal Assessment Battery ^a	13		15	18	
Middelheim Frontality Scale ^b			8		
Language					
Systematische Opsporing Schrijfproblemen (SOS)	8 (−4.8)		5 (−2.5)		1.7 (1.3)
Comprehensive Aphasia Test (CAT)					
Cognitive screening					
Semantic memory		10 (+0.3)		10 [7–10]	9.7 (0.9)
Word fluency		13 (−5.5)			71.9 (10.57)
Recognition memory		9 (−0.7)		10 [4–10]	9.6 (0.9)
Gesture object use		10 (−2.1)		12 [9–12]	11.5 (0.7)
Arithmetic		5 (−0.6)		6 [2–6]	5.5 (0.8)
Language comprehension					
Comprehension of spoken words		30 (+0.9)		30 [20–30]	28.5 (1.7)
Comprehension of written words		30 (+0.5)		30 [24–30]	29.5 (1)

Table 1 (continued)

General	May 2012 Score ($\pm 1SD$ or pc.)	June 2012 Score ($\pm 1SD$ or pc.)	October 2012 Score ($\pm 1SD$ or pc.)	Maximum or # trials [range]	Mean (SD)
Comprehension of spoken sentences		30 (−0.5)		32 [20–32]	30.8 (1.7)
Comprehension of written sentences		30 (−0.2)		32 [20–32]	30.4 (2.2)
Expressive language					
Repetition of digit strings		5 (−0.6)		7 [0–7]	5.6 (1)
Repetition of sentences		6 (+0.5)		6 [4–6]	5.8 (0.4)
Naming objects		46 (+1)		48 [25–48]	43.2 (4.5)
Naming actions		8 (−1.9)		10 [5–10]	9.5 (0.8)
Reading words		48 (+0.5)		48 [40–48]	47.3 (1.5)
Reading complex words		6 (+0.3)		6 [2–6]	5.9 (0.4)
Reading non-words		10 (+0.3)		10 [4–10]	9.6 (1.2)
Writing (copying)		31 (+0.3)		31 [5–31]	30.1 (3.1)

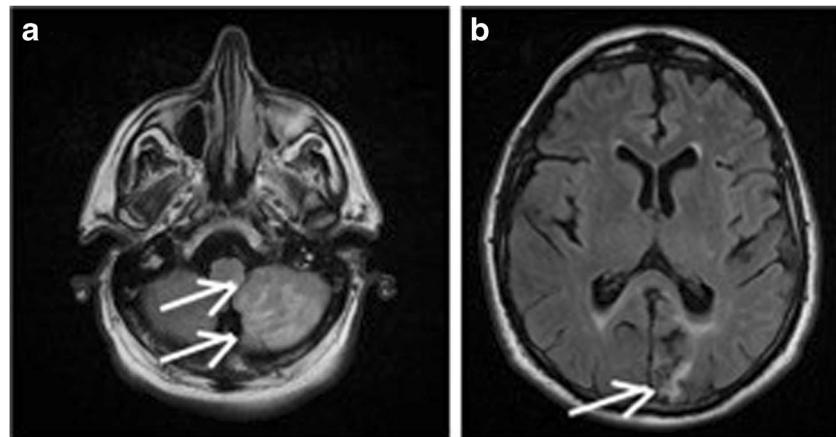
FSIQ full scale intelligence quotient, WMS-R Wechsler Memory Scale-Revised, pc. percentile, PIQ Performance Intelligence Quotient, SD standard deviation, VIQ Verbal Intelligence Quotient

of balance problems, vertigo, and vomiting. On admission to hospital, he presented with left-sided ataxia, dysarthria (slurred speech), and a tendency to fall over to the right side. Although the patient mastered six foreign languages [English (maternal language), French (learnt in school at age 11), German (learnt in school at age 13), Serbo-Croat (learnt after crash course at age 24), Hebrew (Ivrit) (learnt at an intensive course at age 28), and Dutch (moved to Belgium at age 35 and married a Flemish-speaking woman), he could only speak and understand his native language, which was British English. He indicated that he had some very mild word-finding difficulties in English as well. He was completely unable to have a conversation in Dutch or any of the other non-native languages (= differential polyglot aphasia). Medical history included arterial hypertension, coronary heart disease and a left occipital stroke 10 years before this admission. The patient had an educational level of 12 years (grammar school) and had worked as a (war) journalist in different countries. On admission, speech in English was qualified as “slurred”

but during the next days it became marked by a slow rate with sudden accelerations, breathiness, altered word stress, and odd intonation patterns (terminal pitch rise). In terms of segmental changes, diphthongs were backed and consonant articulation became unclear when speech rate increased. Although he severely struggled for everyday words and grammar, his command of Dutch gradually started to return 2 days post-stroke but did not reach premorbid proficiency levels. Minimal progression was made with regard to the other non-native languages. The patient's daughter reported that during the first days post-stroke her father also had started speaking with an “Estuary English” accent, which is typical of the region where the patient grew up. For his job he had commuted between Australia and Europe for more than 12 years, the reason he had adopted an Australian accent. In Dutch, an accent change was found from pre-stroke standard Dutch accent to a regional accent (Antwerp).

T1 and T2-weighted axial FLAIR imaging of the brain showed inhomogeneous hyperintense areas in the left inferior

Fig. 3 T2-weighted axial slice of the brain (a) at the level of the cerebellum, demonstrating (left) a hyperintense area in the left posterior inferior brainstem and postero-inferior portion of left cerebellar hemisphere and (b) demonstrating an old infarction in the calcarine region of the left occipital lobe



cerebellar hemisphere and the postero-inferior left cerebellum, consistent with a recent infarct in the vascular territory of the left posterior inferior cerebellar artery (PICA). A hyperintense area was also observed in the left posterior inferior brainstem (Fig. 3a). In addition, an old vascular lesion was found in the left occipital lobe as well as small vascular lesions in the periventricular white matter (Fig. 3b). MRA revealed atheromatosis of the distal left vertebral artery (no flow was detected) and the right distal vertebral artery and proximal basilar artery (irregular shape).

The neuropsychological investigations were performed in English 1 week after stroke (see Table 2). Neuropsychological tests focused on general cognitive abilities, intelligence, memory, as well as visuospatial and executive functions. A strong and consistent right hand preference was objectified by means of the Edinburgh Handedness Inventory [40] (laterality quotient = +100). General cognitive screening showed normal performance (MMSE: 27/30, SD: −0.33) [29]. The RBANS [32] showed superior visuospatial/constructive skills (index 121, SD: +1.40) and a very superior immediate recall (index 136, SD: +2.40). Language (index 96, SD: −0.27), attention (index 94, SD: −0.4), and delayed recall (index 101, SD: +0.07) were within the normal range. Raven's Coloured Progressive Matrices [41] revealed high average visuospatial problem solving skills (pc. 90). Frontal planning and problem solving were normal as well. The WCST showed normal cognitive flexibility and frontal problem solving (4 categories within 64 trials). The Stroop Colour Word test (pc. 20) showed a low average ability to inhibit a competing and more automatic response. In summary, the patient obtained normal results for all of the administered tests.

As evidenced by the CAT [42], the patient obtained maximum results when tested in his mother tongue. Visual confrontation naming on the BNT [43] was normal for English (57/60, SD: +0.03) but severely pathological for Dutch (BNT: 25/60, SD: −10.64): 13/35 errors were intrusions from English words (foreign words), whereas for 18/35 pictures no response was given. Semantic verbal fluency, assessed by means of a one-minute oral production of words belonging to a specific semantic category, was within the normal range (58 items, SD: +0.31).

A diagnosis of differential polyglot aphasia and FAS following a left cerebellar stroke was made. At the behavioural level, no disturbances were noted. He remained positive and calm.

Survey of the Literature: FAS After Posterior Fossa Lesions

Cole [6] was the first who described two patients with FAS after posterior fossa lesions (Table 3, cases 1 and 2). The first patient was a 29-year-old polyglot man, who lived in Ohio and spoke American English and Yiddish. He developed FAS *presumably* in the context of cerebellar degeneration due to

anoxia. Neurological symptoms consisted of dysphagia, ataxia of arms and legs, spastic-ataxic gait, and bilateral Babinski sign. He suddenly developed an accent, which was perceived as Italian. The second patient was a 58-year-old right-handed, polyglot (English/Hungarian) woman, who suffered an inferior pontine infarction. Post-stroke she developed an Eastern European accent. Initial symptoms included vertigo, diplopia, horizontal gaze palsy, speech difficulties (FAS and dysarthria), and ataxia of the arms and legs. Before the stroke, friends and family considered her accent to be American English, typical of Ohio, where she was a long-time resident. Cole [6] questioned whether there is a relationship between FAS and dysarthria and whether the alterations of speech prosody and rhythm are related to posterior fossa lesions. Robert Joynt commented on these unexpected findings as follows: "...it doesn't appear unusual that any of the brain mechanisms which are used to integrate motor acts may alter force, prosody, and rhythm of speech. It is well that Dr. Cole has pointed this out with posterior fossa lesions" (p. 153). However, he ironically added: "(...) I am enough of a romanticist to think that these subtle inflections and nuances of speech must ultimately stem from the cerebral cortex. For example, if Juliet had said in a monotone, 'Romeo, Romeo, wherefore art thou, Romeo?' I doubt if Romeo would have done himself in for her. But then, I don't think Shakespeare wrote for the cerebellum." (p. 153). In the discussion, it was concluded that distinct forms of dysarthria-like syndromes exist: those due to posterior fossa lesions and those due to frontal lesions.

Forty years later, Dankovičová & Hunt [23] (Table 3, case 3) described a 56-year-old man with a stroke affecting the anterior margin of the mid pons immediately left to the midline. The lesion was discovered on MRI 16 months after the patient suffered the stroke with persistent speech difficulties and a noticeable change of accent, identified mainly as Italian or Greek in a perceptual accent rating experiment. The patient's original accent was typical of his Essex hometown. He was raised in London. His father was an Englishman and his mother was of Greek descent. However, she declared that she had always spoken English with her son. In the light of the patient's speech characteristics, the authors hypothesised that FAS after a pontine lesion may be related to dysarthria (ataxic or bulbar/flaccid).

In 2013, Tran and Mills [26] (Table 3, case 4) described a 60-year-old African-American woman, who presented at the emergency department with an acute change of accent which appeared to her as Jamaican, accompanied by gait disturbances, heaviness of both legs and severe headache. CT scan of the brain on admission was normal but an MRI scan revealed an infarction in the left pons. Magnetic resonance angiography (MRA) confirmed this finding.

In one case, a lesion in the posterior fossa was argued to be—counter intuitively—implicated in the accent *remission*. In 2009, Cohen et al. [25] (Table 3, case 5) described a 58-year-old English-speaking woman who developed FAS after a

Table 2 Overview of the neuropsychological investigations: case 2

General	Score (\pm SD or pc.)	Maximum or #trials	Mean (SD)
Folstein MMSE	27 (-0.3)	30	27.7 (2.1)
RBANS			
Visuospatial/constructive index	121 (+1.4)		100 (15)
Immediate recall index	136 (+2.4)		100 (15)
Language index	96 (-0.3)		100 (15)
Concentration index	94 (-0.4)		100 (15)
Delayed recall index	101 (+0.1)		100 (15)
Raven's colour red progressive matrices	29 (pc. 90)	36	
Executive functions			
Wisconsin Card Sorting Test	4 cat	64 trials	4 cat
Stroop Colour Word Test			
Card 1	45" (pc. 65)		48"
Card 2	46" (pc. 95)		6"
Card 3	128" (pc. 20)		99"
Language			
Boston Naming Test (BNT) NL	25 (-10.6)	60	54.9 (2.8)
Boston Naming Test (BNT) EN	51 (-1.4)	60	54.9 (2.8)
Verbal fluency	58 (+0.3)		53.9 (13.2)
Comprehensive Aphasia Test			
Cognitive screening			
Line bisection	-1 (+0.9)	-1 [-2; -1]	-0.2 (0.9)
Semantic memory	10 (+0.3)	10 [7–10]	9.7 (0.9)
Recognition memory	10 (+0.4)	10 [4–10]	9.6 (0.9)
Gesture object use	10 (-2.14)	10 [9–12]	11.5 (0.7)
Arithmetic	6 (+0.6)	6 [2–6]	5.5 (0.8)
Language comprehension			
Comprehension of spoken words	30 (+0.9)	30 [20–30]	28.5 (1.7)
Comprehension of written words	30 (+0.5)	30 [24–30]	29.5 (1)
Comprehension of spoken sentences	32 (+0.7)	32 [20–32]	30.8 (1.7)
Comprehension of written sentences	32 (+0.7)	32 [20–32]	30.4 (2.2)
Comprehension of spoken paragraphs	4 (+0.7)	4 [0–4]	3.5 (0.7)
Expressive language			
Repetition of words	32 (+0.4)	32 [24–32]	31.4 (1.4)
Repetition of complex words	6 (+0.2)	6 [2–6]	5.9 (0.5)
Repetition of nonwords	10 (+0.6)	10 [0–10]	9 (1.8)
Repetition of digit strings	7 (+1.2)	7 [0–7]	5.6 (1.0)
Repetition of sentences	6 (+0.5)	6 [4–6]	5.8 (0.4)
Naming objects	48 (1)	48 [25–48]	43.2 (4.5)
Naming actions	10 (+0.6)	10 [5–10]	9.5 (0.8)
Reading words	48 (+0.5)	48 [40–48]	47.3 (1.5)
Reading complex words	6 (+0.3)	6 [2–6]	5.9 (0.4)
Reading function words	6 (0)	6 [4–6]	6 (0.2)
Reading non-words	10 (+0.3)	10 [4–10]	9.6 (1.2)
Writing (copying)	31 (+0.3)	31 [5–31]	30.1 (3.1)
Written picture names	23 (+0.7)	23 [8–23]	21.6 (2)

MMSE Mini Mental State Examination, pc. percentile, RBANS Repeatable Battery for the Assessment of Neuropsychological Status, SD standard deviation

left frontoparietal infarction. However, she suddenly *lost* the accent after incurring a right cerebellar hemorrhage. Cohen et al. [25] explained this phenomenon stating that the infarction may have caused a disruption of the “competitive interactions between the two cerebellar hemispheres” (p. 566) and may have led to functional facilitation of the left cerebellar hemisphere in the presence of “maladaptive activation” (p. 566) of the right cerebellar hemisphere.

The case described by Mariën et al. [27] (Table 3, case 6) suffered a cerebellar hemorrhage in the right cerebellar hemisphere, demonstrated on a CT and MRI. The stroke receded without intervention but left the patient with a posterior fossa syndrome and cerebellar cognitive affective syndrome (CCAS)

or Schmahmann’s syndrome [46], a constellation of executive, visuospatial, affective and linguistic disturbances in patients with cerebellar damage. Initially, the patient was mute (akinetic), though later he developed FAS and an ataxic dysarthria. Due to the important cognitive and linguistic impairments as well as personality changes, the researchers also performed a SPECT scan. The functionality of the crossed cerebro-cerebellar connections had definitely been compromised by the lesion as a severe hypoperfusion was demonstrated in the bilateral prefrontal areas, which could account for the executive disturbances, behavioural changes and was reconcilable with the linguistic profile.

Three patients demonstrated FAS after a disruption of the functional connection between the perisylvian speech areas and

the infratentorial regions (i.e., the cerebellum) [2, 9, case 2, 24]. Hwang et al. (2001) [24] (Table 4, case 1) described a 40-year-old, right-handed, Mandarin-Chinese speaking Taiwanese woman, who was admitted to hospital with a sudden change of accent to (American) English as an isolated symptom. Initial physical and neurological examinations, as well as a series of blood tests were normal. The patient displayed focal hyperintensities at the level of the bilateral internal capsules (genua and posterior limbs) on T2-weighted MRI, but these were considered clinically insignificant. The accent receded 3 days after hospitalisation, but the hyperintensities were still visible on repeat MRI 2 years later. In contrast, SPECT performed 1 day after admission revealed a hypoperfusion at the level of the left lateral temporal region and right cerebellum. The authors hypothesised that these functional deficits were due to a transient ischemic attack in the left temporal area. Repeat SPECT 2 years later showed a complete resolution. Hence, the authors argued that the hypoperfusion may have played a significant role in the accent change.

The case described by Verhoeven and Mariën [2], also discussed in Mariën and Verhoeven [9] (here Table 4, case 2) is the first of two stroke patients in whom remission of FAS symptoms was directly related to normalisation of the hypoperfusion in the right cerebellum after focal left frontoparietal damage (diaschisis). The patient was a 53-year-old, right-handed, Dutch-speaking Belgian woman who was admitted to hospital because of an acute motor speech impairment rapidly evolving to mutism associated with a right-sided loss of strength. In addition to mutism, the neurological examination revealed a right hemiparesis and central facial nerve palsy. CT demonstrated an infarction in the left frontoparietal region. Two weeks later, an MRI confirmed ischemic damage in the left middle cerebral artery territory, comprising the inferior frontal gyrus, precentral gyrus, anterior insular cortex, postcentral gyrus and supramarginal gyrus. 99m Tc ECD SPECT at 33 days post-stroke revealed a hypoperfusion in the left frontal motor and parietal cortex, the thalamus and striatum of the left hemisphere as well as in the right cerebellum (no standard deviations provided). Repeat SPECT 3 years after the stroke showed that perfusion deficits at the supratentorial level had only marginally improved, but that perfusion in the right cerebellum had normalised. Parallel to the observed improvement of the perfusion changes at the cerebellar level, a resolution of FAS (French or Russian-like accent) was attested. The second patient was a 61-year-old right-handed Dutch-speaking native Belgian man who suffered a basal ganglia hemorrhage (Table 4, case 3). This patient acutely developed global aphasia, which evolved to conduction aphasia after approximately 1 week. After this transition of aphasic symptoms, the patient's accent was perceived as "North-African." Neurological examinations disclosed a right hemiplegia and right hemianopia. A repeat MRI of the brain 3 weeks later confirmed that the lesion comprised the left putamen, genu

and posterior limb of the internal capsule extending to the insula, temporal and parietal lobe. Quantified 99m Tc ECD SPECT 1 month post-stroke revealed a severe hypoperfusion (more than 2 SD) at the cortical and subcortical level, including the left thalamus, left lentiform nucleus, left medial, lateral temporal region, left motor cortex and right cerebellum. Repeated SPECT completed at 6 months post-stroke demonstrated a complete resolution of the hypoperfusion at the infratentorial level, whilst perfusion deficits at the supratentorial level had only slightly enhanced. Around the same time (i.e., 6 months post-stroke) an improvement in both FAS and aphasic symptoms was observed. The congruent remission of the cerebellar hypoperfusion and the linguistic deficits left the authors to hypothesise on the possibly crucial role of the cerebellum in motor speech planning disorders, including FAS.

Discussion

Two new cases of FAS after a posterior fossa lesion are documented. The first case developed FAS and paralytic dysarthria after an ischemic stroke in the right pons, causing damage to the brainstem nuclei and interrupting the crossed ponto-cerebellar pathway. The combination of linguistic, cognitive and affective changes allowed for a diagnosis of "brainstem cognitive affective syndrome" which closely relates to Schmahmann's syndrome [46]. From an anatomoclinical point of view, the clinical symptoms were consistent with the SPECT findings, demonstrating a hypoperfusion affecting the frontal and prefrontal areas bilaterally as well as in the left cerebellum [28]. The second case developed differential polyglot aphasia and FAS after an infarction in the vascular territory of left PICA. He did not demonstrate any affective or behavioural disturbances, nor any cognitive impairments. At the linguistic level, however, Dutch as well as all the other non-native languages were severely disturbed while his mother tongue (British English) remained clinically unaffected (differential polyglot aphasia). Post-stroke FAS reflected a reversion to a previously learnt accent [50, 51].

Both patients' speech characteristics may also be qualified as typical apraxia of speech (AoS) (slow speech rate, increase in errors when speech rate is increased, attainment of vowels more than consonants, (irregular) initiation difficulty) or as ataxic dysarthria (slow speech rate altered with sudden fast speech rate, disrupted articulation of vowels and consonants, slurring) [52], but do not allow for a clear differentiation. Ataxic dysarthria is the most typical motor speech disorder associated with cerebellar disorders and is mainly characterised by a distorted articulation and prosody [53]. Although ataxic dysarthria is traditionally considered a disorder of motor execution, recent studies have attributed typical characteristics of ataxic dysarthria to a disruption of motor speech programming/planning instead of to a dysfunctional execution of motor speech [9,

Table 3 FAS after posterior fossa lesions

Case	Age/gender/ handedness	Etiology	Lesion localisation	Speech/language characteristics	Original accent	Newly acquired accent	Cognition
1 Cole (1971) (Case 2) [6]	58/F/NI polyglot	Infarction	Clinically suspected inf. pontine infarction	Acute: spastic dysarthric and dysphonic	“Ohioan” (i.e. American English) and Hungarian (polyglot)	Eastern European	Normal mental status, No facial apraxia
2 Cole (1971) (Case 1) [6]	29/M/NI polyglot	Cerebellar anoxia	Clinically suspected cerebellar degeneration secondary to anoxia	Acute: dysarthric, slightly spastic and dysphonic	Ohioan, Yiddish	Italian	No oral apraxia
3 Dankovičová & Hunt (2011) [23]	56/M/R* monolingual	Stroke 1: 2003; Stroke 1 2005; Stroke 2	Stroke 1: CT at 6 m: normal MRI at 16 m: small wedge-shaped area of porencephaly along ant. margin of the mid pons immediately to the midline MRI (2008): post. parietal atrophy sup. to occipital sulcus	Onset (2003): slurred speech; within a few days: improved speech but onset FAS (no formal assessments) After 14 m: BDAE: poor non-verbal oral agility task (8/12), normal verbal agility (14/14) Stroke 2 (2005): no significant change of FAS	British English	Italian, Greek (patient grew up in Essex, to Greek mother who spoke English)	Normal except for non-verbal oral agility (BDAE), apraxia test: transitive limb tasks (9/10), bucco-facial, intransitive limb and whole body tasks (10/10)
4 Tran & Mills (2013) [26]	60/F/NI	Infarction	Initial CT: normal MRI (3w post-onset): subacute infarction in the L hemi-pons and absent flow within L vertebral artery	Initial presentation at ED: speech change was thought to be secondary to a goiter impinging in the recurrent laryngeal nerve 3w after onset symptoms: presented again to ED, complained of on-going altered speech pattern: spoke fluently Jamaican / Italian accent	African-American	Jamaican/Italian	NI
5 Cohen et al. (2009) [25]	58/F/R*	Infarction	MRA head and neck: left pontine infarct (no hemorrhage), irregularity in ant. circulation, diminished flow in the post. circulation (affecting esp. basilar artery) MRA with contrast: high-grade stenosis of basilar artery, fetal origin of the L posterior cerebral artery, and multiple areas of narrowing in both MCA's	*ending most words with ‘-ah’, *epenthesis: adding schwa to end or middle of words BNT = 56/60,	English	English with unlearnt NI accent	
6 Mariën et al. (2013) [27]	72/M/R	Cerebellar stroke (right)	Stroke 1: MRI: stroke L fronto-parietal junction After 3y: stroke 2: MRI: R inf. cerebellar hemorrhage	Onset aphemia; within hours: English with unlearnt accent (FAS) for 3 years Stroke 2: remission FAS, normal speech	Dutch	German	1 m and 6 m post-stroke: depressed MMSE, pathological FSIQ, VIQ and PIQ (WAIS-II), pathological scores on

Table 3 (continued)

Case	Age/gender/ handedness	Etiology	Lesion localisation	Speech/language characteristics	Original accent	Newly acquired accent	Cognition
			deficits bilaterally distributed in the prefrontal brain regions during follow-up MRI (6 months post-stroke) show complete resorption of the cerebellar hemorrhage, atrophy of the right cerebellum (e) and restoration of the intraventricular volumes	After 1 month: spontaneous speech reappeared, FAS and foul language + foreign words (German in his Dutch) Posterior Fossa Syndrome			all subtests of the RBANS, pathological scores on WCST, pathological scores on Stroop and TMT ^a 6 m: decrease in executive + behavioural test CCAS

Ant. anterior, *BDAE* Boston Diagnostic Aphasia Examination [44], *BNT* Boston Naming Test [42], *CT* computed tomography, *ED* emergency department, *F* female, *inf.* inferior, *L* left, *m* month(s), *M* male, *MCA* middle cerebral artery, *MRA* magnetic resonance angiography, *MR* magnetic resonance imaging, *NI* not indicated, *post.* posterior, *R* right, *R** right-handed, *RBANS* Repeatable Battery for the Assessment of Neuropsychological Status [32], *SPECT* single photon emission computed tomography, *sup.* superior, *WAIS-III-NL* Wechsler Adult Intelligence Scale-III—Dutch version [45], *WCST* Wisconsin Card Sorting Test [34], *w* week(s), *y* year(s)

[53]. Since ataxic dysarthria shares some evident semiological characteristics with AoS, a typical speech planning and programming disorder, some authors also hypothesised that ataxic dysarthria may be subserved by a similar pathophysiological mechanism [54]. However, only a handful of cases of AoS have been reported with possible cerebellar involvement. Apart from the FAS case of Verhoeven and Mariën [2] (Table 4, case 2), one other case of AoS has been reported in which the right cerebellum may have played a role. Mariën et al. [55] reported a patient who presented with agraphia and severe AoS after a left cerebral hemisphere infarction with lesions affecting the anterior insular cortex and the opercular part of the left inferior frontal gyrus. SPECT not only showed a hypoperfusion in the insular region but also in the right cerebellar hemisphere. After 1 year, AoS completely receded which was reflected by a reperfusion of the right cerebellar hemisphere.

The case described by Mariën et al. [27] on the other hand, demonstrated ataxic dysarthria, in combination with FAS (and CCAS) after a right cerebellar stroke. The newly reported patients demonstrated FAS in association with a *left* cerebellar hypoperfusion (case 1) and a *left* cerebellar infarction (case 2). It might be hypothesised that disruption of the cerebro-cerebellar network—due to a pontine (case 1) and left cerebellar lesion (case 2)—are responsible in both patients for the planning and organisation deficits at both the linguistic and non-linguistic level (case 2). The first patient presented a significant hypoperfusion of the left inferior medial frontal regions as the result of cerebello-cerebral diaschisis reflecting the loss of excitatory impulses from the cerebellum to the supratentorial areas subserving the planning and organisation of motor and non-motor linguistic and non-linguistic processes, including articulation. For the second patient, no SPECT data were available. Speech production, and more particularly speaking with a monotonous voice, has not only been associated with right but also with left cerebellar hemisphere activations in overt and covert speech tasks [56]. The differential resolution of the aphasia selectively affecting the non-native languages in this patient, in combination with the attested left cerebellar damage, could be related to the theories of the more bilaterally distributed speech and language networks—possibly also those mediating prosody in particular—in bilinguals and polyglots. More right hemisphere structures are recruited in polyglots when they speak non-native languages. These regions are structurally connected with the contralateral left cerebellar hemisphere. In our patient, a reduction or even loss of excitatory impulses from the left cerebellar hemisphere due to stroke might have induced functional disruption of the right cerebral regions crucially implicated in non-native language production [57, 58].

From a pathophysiological perspective, FAS has also been related to an impaired position of vocal tract or increased tension [59, 60]. If increased tension would be one of the primary instigators of FAS-like speech, then this could be an argument in favour of FAS as an executive disturbance: the

Table 4 FAS after frontal lesions with related functional involvement of posterior fossa structures (diaschisis)

Case	Age/gender/ handedness	Etiology	Lesion localisation	Speech/language characteristics	Original accent	Newly acquired accent	Cognition
1 Hwang et al. (2001) [24]	40/F/R* monolingual	Infarction	MRI at onset: normal SPECT on day 1: focal hypoperfusion defect L lateral temporal region and crossed cerebellar diaschisis MRI at 2y: normal SPECT at 2y: normal	No aphasia; dysarthria or AoS but minimal decrease of fluency (by self report), tone errors, FAS; within 3d: almost complete remission of FAS; at 2y: occasional problems with tone in daily conversations	Mandarin	American English-like accent	No memory loss, digit span: normal
2 Verhoeven & Mariën (2002) [2]	53/F/R* polyglot	Infarction	CT/MRI: L hemisphere lesion involving inf. front. gyrus, precentral gyrus, ant. insula, postcentral gyrus and supramarginal gyrus SPECT 33d post-stroke: L frontoparietal hypoperfusion extending to L temporal and adjacent subcortical regions, crossed cerebellar diaschisis SPECT after 3 years: unaltered supratentorial perfusion pattern, improved perfusion in the R cerebellum	At onset: verbal mutism for 1 day evolving to AoS and FAS (no aphasia); 11d–27d post-onset: AoS and FAS (no dysarthria nor aphasia) 36 m post-onset: remission FAS, mild residual AoS	Dutch	French, Russian-like	1 m post-onset: MMSE, RCPM, WAIS-NL, Hierachic Dementia Scale, WMS-R, ROF, WCST, Stroop test, Right-Left Orientation test, Visual Form Discrimination test, Judgment of Line Orientation test: above average cognitive results (IQ, memory, language, praxis, gnosis, concentration, problem solving, executive functions)
3 Mariën & Verhoeven (2007) Case 2 [9]	61/M/R* formal test	Hemorrhage	CT: L hemisphere hemorrhage involving the putamen, genu and post. limb of internal capsule with extension to the post. insula, the medial temporal lobe and the paraventricular white matter of the parietal lobe SPECT: 1 m post-onset: severe hypoperfusion L thalamus, lentiform nucleus, L medial and lateral temporal region and L motor cortex, crossed cerebellar diaschisis; 6 m post-onset: remission R cerebellar hypoperfusion, unchanged perfusion deficits at the supratentorial level	Onset: verbal mutism, global aphasia after 6d: conduction-like aphasia and FAS 1 m post-onset: conduction aphasia and agrammatism, FAS, very high speech rate, omissions and consonant deletions, harsh voice quality, syllable-timed and isochronous speech rhythm, groping AAT (weak oral and written comprehension, very weak repetition, esp. words and sentences; weak naming, weak written language)	Dutch	North-African	1 m post-onset: MMSE (23/30), RPM (IQ: 117), ROF, Right-L-Left orienta- tion task, Visual Form Discrimination task, Judgment of Line Orientation: all normal

AAT Aachen Aphasia Test [47], Ant. anterior; BDAE Boston Diagnostic Aphasia Examination [44], BNT Boston Naming Test [43], CT computed tomography, ED emergency department, F female, inf. inferior, Judgement of Line Orientation Task [48], L left, m month(s), M male, MCA middle cerebral artery, MMSE Mini-Mental State Examination [29], MRA magnetic resonance angiography, MRI magnetic resonance imaging, NI not indicated, post. posterior, R right, R* right-handed, RCPM Raven Coloured Progressive Matrices [41], ROF Rey-Osterrieth Figure [49], SPECT single photon emission computed tomography, sup. superior, visual form discrimination task [48], WAIS-NL Wechsler Adult Intelligence Scale—Dutch version [45], w weeks(s), WCST Wisconsin Card Sorting Test [34], WMS-R Wechsler Memory Scale-Revised, y year(s)

increased muscle tone can be an upper motor neuron sign which characterises patients with (spastic/pseudobulbar) dysarthria. This would constitute an “organic explanation” for FAS. However, current patients both presented with dysarthria types associated with muscle tone weakness: paralytic dysarthria and ataxic dysarthria. This explanation does not seem applicable for current patients. Particularly interesting is the neurocomputational viewpoint on FAS.

Tomasino et al. [61] argued that the deficits of FAS patients are due to deficient sensory feedback (in the case of primarily subcortical damage) or feedforward (in the case of cortical and perilesional subcortical damage) control mechanisms. They explain deficits in the context of the DIVA-model (i.e., Directions into Velocity of Articulators) [62, 63]. The model describes speech production as relying on two subsystems: a feedforward control system and a feedback control system. Applied to speech development, the model can be explained as follows: the phonemes that were acquired at an early age are stored in speech sound maps in the inferior frontal gyrus and are directly linked to articulator position maps stored in the ventral premotor cortex; together they form the feedforward control loop. Via the feedforward control, one is aware of the phonological results of set of motor programs (predictive component) which are transferred to the articulator position and velocity maps. When the initiation maps in the (bilateral) supplementary motor area (SMA) become active, these commands are released. In these commands, the position of the different articulatory variables (jaw height, tongue height, tongue shape etc.) are encoded, which are translated into movements of the vocal tract. The speech sound maps define the auditory and somatosensory target maps in the feedback control system. When speech is articulated, the somatosensory and auditory-perceptual awareness, mediated by subcortical nuclei, activates the feedback control loop, which allows for an update of the feedforward loop and the correction of the articulatory encoding to achieve targets correctly. If the deficits in FAS are explicable, even partly, in the context of the DIVA model, then a role for the cerebellum—affected in both patients—is not inconceivable. The cerebellum plays an important role in “the learning and maintenance of feedforward motor commands” [63, p. 11]. Since it stands in close connection to the motor areas on the one hand, and on the other hand receives input from the somatosensory cortex and auditory areas [64], it is attributed a cardinal role in feedforward motor control via supposed feedback motor learning in response to a miscalibration of intended movement or execution of targets [64]. These feedforward projections are hypothesised to also include the basal ganglia (axons in the dentate nucleus in the cerebellum project to the contralateral (motor) cortex via the thalamus). This is reconcilable with the theories of Reeves and Norton [65], Blumstein and Kurowski [66], Schiff et al. [67] who claim that FAS is due to a disruption of the cortico-striatopallidal-thalamic pathways. Thus, the rhythmic disturbance can be consistent with a defect in the predictions

(feedforward control) exerted by the cerebellar circuitry. This could then also serve as an explanation for the semiological resemblances between FAS and AoS, and by inference FAS, AoS, and ataxic dysarthria. The hypothesis of errors in predictions leading to compensatory substitution mechanisms by the unaffected cerebral areas (including the cerebellum itself) could be put forward. From a cognitive perspective, FAS has been viewed as the result of an impairment in distinguishing between a direct and indirect speech encoding route [68]. The results of the SPECT scan for the first patient demonstrated disturbed physiological communication between the cerebellum and frontal regions, which are responsible for speech planning and programming. For the second patient, the left cerebellar hemisphere was damaged. Here, a maladaptive compensation under the form of an aberrant recovery process [69] or an overcompensation mechanism, as reported when the inhibitory signals from the cerebellum are affected, could also be considered [70]. A maladaptive compensation from the part of the contralateral cerebellar hemisphere, exerting distance effects on the frontal cerebral areas (including for instance) the motor cortex, might be hypothesised. The precise roles of (a) the olivo-cerebellar pathway, a second major inflow tract to the cerebellum with a major role in pace-making activity, and (b) the projections from the cerebellar cortex to cerebellar nuclei remain undetermined. However, these explanations are reminiscent of the case described by Cohen et al. [25] who described a 58-year-old woman who developed FAS after an MCA stroke affecting the left frontoparietal lobe. The authors dwell on how the sudden remission after a second cerebellar stroke in the right inferior posterior lobe can be explained. Cerebellar-parietal connections have been argued to underpin phonological storage, although its exact role remains unclear [71, 72].

In conclusion, it seems that a disruption of the cerebro-cerebellar circuitry following a right pontine lesion and left cerebellar lesion in the posterior lobe, exerting remote functional effects in the cortical areas subserving planning and organisation, may play a foremost important role in the pathophysiology of FAS following posterior fossa lesions. Several hypotheses have been formulated as to how this damage may be related to the onset of FAS. These explanations were not mutually exclusive. It was argued that FAS can be the consequence of disturbed feedforward and/or feedback speech control mechanisms. The cerebellum has been hypothesised to play an important role in feedforward speech control via projections to the frontal cortex. This is relatable to earlier theories claiming that FAS is the result of a disruption affecting the cortico-striatopallidal-thalamic pathways [66–68]. These theories might explain FAS in the first case: as demonstrated by SPECT, damage to the crossed ponto-cerebellar pathway (inflow tract to the cerebellum) induced (pre)frontal dysfunction. On the other hand, the possibility of maladaptive compensation from the part of, possibly, the contralateral cerebellum could be related to the FAS characteristics in case 2.

However, this patient was a polyglot. Hence, the impact of the left cerebellar hemisphere lesion on the speech and language in this patient should also be considered in the context of a more bilaterally distributed language network in polyglots.

In future studies on FAS, DTI and/or fMRI experiments may be a relevant addition with a view to investigate (possibly disrupted) (bi-hemispheric) network activity, as well as fiber tracts. This will allow researchers to hypothesise on FAS prognosis and possibly also on whether the semiological resemblance with (ataxic) dysarthria or AoS is grounded on a shared neural and/or neurophysiological substrate.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

References

- Verhoeven J, Mariën P. Neurogenic foreign accent syndrome: articulatory setting, segments and prosody in a Dutch speaker. *J Neurolinguist*. 2010;23:599–614. doi:10.1016/j.jneuroling.2010.05.004.
- Verhoeven J, Mariën P. Prosodic characteristics of a case of foreign accent syndrome. *Antwerp Papers in Linguistics*. 2002;100:131–48.
- Keulen S, Mariën P, Wackenier P, Jonkers R, Bastiaanse R, Verhoeven J. Developmental foreign accent syndrome: report of a new case. *Front Hum Neurosci* 2016a. 2016a;10(65):1–9. doi:10.3389/fnhum.2016.00065.
- Keulen S, Verhoeven J, De Witte E, De Page L, Bastiaanse R, Mariën P. Foreign accent syndrome as a psychogenic disorder: a review. *Front Hum Neurosci*. 2016b;10(168):1–16. doi:10.3389/fnhum.2016.00168.
- Tailby C, Fankhauser J, Josev EK, Saling MM, Jackson GD. Clinical foreign accent syndrome evolving into a multiplicity of accents. *J Neurolinguist*. 2013;26(3):348–362.
- Cole M. Dysprosody due to posterior fossa lesions. *Trans Am Neurol Assoc*. 1971;96:151–4.
- Kwon M, Kim JS. Change of dialect after stroke: a variant of foreign accent syndrome. *Eur Neurol*. 2006;56:249–52.
- Naidoo R, Warriner EM, Oczkowski W, Sévigny A, Humphreys K. A case of foreign accent syndrome resulting in regional dialect. *The Canadian journal of neurological sciences*. 2008;35:360–5.
- Mariën P, Verhoeven J. Cerebellar involvement in motor speech planning: some further evidence from foreign accent syndrome. *Folia Phoniatrica et Logopaedica*. 2007;59(4):210–7.
- Monrad-Krohn GH. Dysprosody or altered “melody of language”. *Brain*. 1947;70:405–15.
- Nielsen JM, McKeown M. Dysprosody. Report of two cases. *Bull Los Angel Neurol Soc*. 1961;26:157–9.
- Edwards RJ, Patel NK, Pople IK. Foreign accent following brain injury: syndrome or epiphénoménon. *Eur Neurol*. 2005;53(2):87–91.
- Berthier ML, Ruiz A, Massone MI, Starkstein SE, Leiguarda RC. Foreign accent syndrome: behavioural and anatomical findings in recovered and non-recovered patients. *Aphasiology*. 1991;5(2):129–47.
- Denes G, Trumper J, Maddalon M, Romito L. Foreign accent syndrome: an Italian case study. *Proceedings of the ICPHS95*. 1995;2:662–5.
- Munson PD, Heilman B. Foreign accent syndrome: anatomic, physiologic and psychosocial considerations. *S D J Med*. 2005;58(5):187–9.
- Blumstein SE, Alexander PP, Ryalls JH, Katz W, Dworetzky B. On the nature of the foreign accent syndrome: a case study. *Brain Lang*. 1987;31(2):215–44.
- Avila C, González J, Parcet M, Belloch V. Selective alteration of native, but not second language articulation in a patient with foreign accent syndrome. *Clinical Neuroscience and Neuropathology*. 2004;15(4):2267–70.
- Gonzalez-Alvarez J, Parcet-Ibars MA, Avila C, Geffner-Sclarcky D. Una rara alteración del habla de origen neurológico: el síndrome del acento extranjero. *Rev Neurol*. 2003;36:227–34.
- Levy ES, Goral M, Castelluccio de Diesbach C, law II F. Stronger accent following a stroke: the case of a trilingual with aphasia. *Clinical Linguistics Phonetics*. 2011;25(10):815–30.
- Masao R, Martinez A, Cisneros J, Alonso M. Síndrome de acento extranjero. *Arch Neurocienc (Mex)*. 2011;16(3):167–9.
- Critchley M. ‘Regional “accent”, Demotic speech, and aphasia,’ in *Les Editions Acta Medica Belgica*, editors. *Livre Jubilaire Docteur Ludo Van Bogaert*. *Les Editions Acta Medica Belgica*; 1962: 182–191.
- Miller N, O’Sullivan, H. What makes foreign accent syndrome foreign? *Int Clin Phon Linguist Assoc*; 1997, 6th Annu Conf, Nijmegen.
- Dankovičová J, Hunt C. Perception of foreign accent syndrome speech and its relation to segmental characteristics. *Clin Ling Phon*. 2011;25(2):85–120. doi:10.3109/02699206.2010.513027.
- Hwang C, Lin M, Lin S. Pure foreign accent syndrome: a case report. *Acta Neurological Taiwanica*. 2001;10:196–201.
- Cohen DA, Kurowski K, Steven MS, Blumstein SE, Pascual-Leone A. Paradoxical facilitation: the resolution of foreign accent syndrome after cerebellar stroke. *Neurology*. 2009;73:566–7.
- Tran A, Mills L. Clinical communications: adults a case of foreign accent syndrome. *J Emerg Med*. 2013;45(1):26–9. doi:10.1016/j.jemermed.2012.11.015.
- Mariën P, Verslegers L, Moens M, Dua G, Herregods P, Verhoeven J. Posterior fossa syndrome after cerebellar stroke. *Cerebellum*. 2013;12(5):686–91.
- Mariën P, D’aes T. “brainstem cognitive affective syndrome” following disruption of the cerebrocerebellar network. *Cerebellum*. 2015;14(2):221–5.
- Folstein MF, Folstein SE, McHugh PR. Mini mental state examination: a practical method for grading the cognitive state of patients for the clinician. *J Psychiatr Res*. 1975;12:189–98.
- Wechsler D. *Wechsler adult intelligence scale—3rd edition (WAIS-3)*. San Antonio: Harcourt Assessment; 1998.
- Wechsler D. *WMS-R: Wechsler memory scale-revised: manual*. San Diego, CA: Harcourt; 1987.
- Randolph C. Repeatable battery for the assessment of neuropsychological status (RBANS). San Antonio, TX: Psychological Corporation; 1998.
- Reitan R. Validity of the trail making test as an indicator of organic brain damage. *Percept Motor Skill*. 1958;8:271–6.
- Grant DA, Berg EA. Wisconsin card sorting test. Odessa (FL): Psychological Assessment Resources; 1993.
- Stroop J. Studies of interference in serial verbal reaction. *J Exp Psych*. 1935;18:643–62.
- Dubois B, Slachevsky A, Litvan I, Pillon B. The FAB: a frontal assessment battery at bedside. *Neurology*. 2000;55(11):1621–6.

37. De Deyn PP, Engelborghs S, Saerens J, Goeman J, Mariën P, Maertens K, Nagels G, et al. The Middelheim Frontality score: a behavioural assessment scale that discriminates frontotemporal dementia from Alzheimer's disease. *Int J Geriatr Psychiatry*. 2005;20(1):70–9.

38. Visch-Brink E, Vandenborre D, De Smet H-J, Mariën P. Comprehensive Aphasia Test – Nederlandstalige bewerking. Amsterdam: Pearson; 2015.

39. Van Waelvelde H, Hellinckx T, Peersman W, Smits-Engelsman B C. SOS: a screening instrument to identify children with handwriting impairments. *Phys Occup Ther Pediatr*. 2012;32(3):306–19.

40. Oldfield R. The assessment and analysis of handedness: the Edinburgh inventory. *Neuropsychologia*. 1971;9:97–113. doi:10.1016/0028-3932(71)90067-4.

41. Raven JC. Coloured Progressive Matrices. London: HK Lewis; 1976.

42. Howard D, Swinburn K, Porter G. Comprehensive aphasia test. Psychology Press: Routledge; 2004.

43. Kaplan E, Goodglass H, Weintraub S. The Boston naming test. Philadelphia: Lea and Febiger; 1983.

44. Goodglass H, Kaplan E. The Assessment of aphasia and related disorders. 2nd ed. Philadelphia: Lea and Febiger; 1983.

45. Uterwijk J, Klinkenberg E, Kooij A. WAIS-III Nederlandse bewerking. Afname scoringshandleiding. Amsterdam: Harcourt Test Publishers; 2005.

46. Manto M, Mariën P. Schmahmann's syndrome—identification of the third cornerstone of clinical ataxiology. *Cerebellum Ataxias*. 2015;2:2. doi:10.1186/s40673-015-0023-1.

47. Graetz P, De Bleser R, Willmes K. De Akense Afasie Test. Lisse: Swets & Zeitlinger; 1992.

48. Benton AL, deS Hamsher K, Varney NR, Spreen O. Contributions to neuropsychological assessment: A clinical manual. New York: Oxford University Press; 1983.

49. Rey A. L'examen psychologique dans les cas d'encephalopathie traumatique. *Archives de Psychologie*. 1941;28:215–285.

50. Seliger GM, Abrams GM, Horton A. Irish brogue after stroke. *Stroke*. 1992;23:1655–6. doi:10.1161/01.STR.23.11.1655.

51. Roth EJ, Fink K, Cherney LR, Hall KD. Reversion to a previously learnt foreign accent after stroke. *Arch Phys Med Rehabil*. 1997;78:550–2.

52. Duffy JR. Motor speech disorders. Substrates, differential diagnosis, and management. Third edition. St Louis, MN: Elsevier Mosby; 2013.

53. Spencer K, Slocomb D. The neural basis of ataxic dysarthria. *Cerebellum*. 2007;6(1):58–65.

54. Mariën P, Verhoeven J, Engelborghs S, Rooker S, Pickut B, De Deyn PP. A role for the cerebellum in motor speech planning: evidence from foreign accent syndrome. *Clin Neurol Neurosurg*. 2006;108:518–22.

55. Mariën P, Pickut BA, Engelborghs S, Martin J-J, De Deyn PP. Phonological agraphia following a focal anterior insulo-opercular infarction. *Neuropsychologia*. 2001;39(8):845–55.

56. Dogil G, Ackermann H, Grodd W, Haider H, Kamp H, Mayer J, et al. The speaking brain: a tutorial introduction to fMRI experiments in the production of speech, prosody and syntax. *J Neurolinguistics*. 2002;15(1):59–90.

57. Meschyan G, Hernandez AE. Impact of language proficiency and orthographic transparency on bilingual word reading: an fMRI investigation. *NeuroImage*. 2006;29(4):1135–40.

58. Abutalebi J, Green DW. Neuroimaging of language control in bilinguals: neural adaptation and reserve. *Bilingualism: Language and cognition*. 2016;1–10.

59. Kurowski KM, Blumstein SE, Alexander M. The foreign accent syndrome: a reconsideration. *Brain Lang*. 1996;54:1–25.

60. Ingram JCL, McCormack PF, Kennedy M. Phonetic analysis of a case of foreign accent syndrome. *J. Phonet*. 1992;20:457–74.

61. Tomasino B, Marin D, Maier M, Ius T, Budai R, Fabbro F, Skrap M. Foreign accent syndrome: a multimodal mapping study. *Cortex*. 2013;49(1):18–39. doi:10.1016/j.cortex.2011.10.007.

62. Goflinopoulos E, Tourville JA, Guenther FH. The integration of large-scale neural network modeling and functional brain imaging in speech motor control. *NeuroImage*. 2010;52(3):862–74.

63. Tourville JA, Guenther FH. The DIVA model: a neural theory of speech acquisition and production. *Lang Cogn Process*. 2011;26(7):952–81.

64. Schmahmann JD, Pandya DN. The cerebrocerebellar system. *Int Rev Neurobiol*. 1997;41:31–60.

65. Reeves RR, Norton JW. (2001). Foreign accent-like syndrome during psychotic exacerbations. *Neuropsychiatry. Neuropsychology and Behavioral Neurology*. 2001;14:135–8.

66. Blumstein SE, Kurowski K. (2006). The foreign accent syndrome: a perspective. *J Neurolinguistics*. 2006;19:346–55.

67. Schiff HB, Alexander MP, Naeser MA, Galaburda AM. Aphemia: clinical-anatomic correlations. *Arch Neurol*. 1983;40:720–7.

68. Whiteside S, Varley R. A reconceptualization of apraxia of speech: a synthesis of evidence. *Cortex*. 1998;34:221–31.

69. Manto MU, Hildebrand J, Jacquy J. Shift from hypermetria to hypometria in an aberrant recovery following cerebellar infarction. *J Neurol Sci*. 1998;157(1):42–51.

70. Baier B, Müller N, Rhode F, Dieterich M. Vestibular compensation in cerebellar stroke patients. *Eur J Neurol*. 2015;22(2):416–8.

71. Macher K, Böhringer A, Villringer A, Pleger B. Cerebellar-parietal connections underpin phonological storage. *J. Neurosci*. 2014;34(14):5029–37. doi:10.1523/JNEUROSCI.0106-14.2014.

72. Ziegler W. The phonetic cerebellum: cerebellar involvement in speech-sound production. In: Mariën P, Manto M, editors. The linguistic cerebellum. London: Academic Press; 2016. p. 1–32.