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GAWH : Global Aphasia Without Hemiparesis Review and case study

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Global aphasia without hemiparesis (GAWH) is a rare, but very interesting syndrome from the point of view of the dissociation between unimpaired motor function and severe linguistic dysfunction. In such cases, dual lesions in the left cerebral hemisphere would be expected, sparing the motor areas but affecting both anterior and posterior perisylvian language areas. But, several studies show that a dual lesion is not necessary to cause a GAWH. We present a study which demonstrates this, in which patient MVS showed a global aphasia despite the presence of only an anterior lesion. This global aphasia evolved over 1 year to a non-specific aphasia with good writing and repetition. Neurocognitive linguistic assessment at 4 months post onset with the PALPA (Bastiaanse et al, 1995) provided the basis for therapy guidelines.

Definition

Global aphasia is normally associated with large left cerebral perisylvian lesions, which extend from cortical to sub-cortical regions. The fact that motor and language areas are located closely to each other results in the typical contralateral hemiplegia seen in global aphasia. Patients with global aphasia show impairment of all productive and receptive linguistic functions, which typically remains resistant even to intensive speech therapy.

In some cases the hemiplegia is absent or transient, in which case one speaks of GAWH. The question in such cases is whether this syndrome has a better prognosis for recovery of language function than the much more common global aphasia with hemiparesis.

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Neurological accounts

GAWH can be caused by multiple lesions in the perisylvian area: Van Horn et Hawes (1982) ; Tranel et al (1987) ; Van Horn (1987) ; Legatt et al (1987) ; Kawahata et al(1991). In all these studies the authors reported multiple lesions in the anterior and posterior language areas, with a preserved motor area. Embolic encephalopathy is seen as the most common aetiological cause, though Legatt et al (1987) claimed that non-embolic aetiologies are also possible, such as tumour /metastasis, epilepsy, trauma, hemorrhagic stroke.

Other authors have reported GAWH after single lesions in the perisylvian area: Ferro (1983) ; Bougouslavsky (1988) ; Deleval et al (1989) ; Kawahata et al (1991) ; Hirano et al (1990) ; Agarwal et al (1995) ; Silva MT et al (2000) ; Lewis et al (2000). Ferro (1983) reported two cases with a single lesion in the area of the middle cerebral artery (MCA), affecting both Broca's and Wernicke's area. Deleval et al (1989) reported two patients with a single pre-rolandic lesion, including severely disturbed comprehension due to an anterior lesion. The case reported by Kawahata et al (1991) had a single temporo-pariëtal lesion. In the most recent study by Bang et al (2004), we found GAWH - cases due to a single lesion in extra-perisylvian areas, namely prefrontal, pariëto-occipital and subcortical regions as well as in the perisylvian region.

In cases with extra-perisylvian lesions or single perisylvian lesions either anterior or posterior, we would expect perfusion deficits on Single Photon Emission Computed Tomography (SPECT) in the perisylvian language areas. Yet, Bang et al(2004) failed to document extensive perfusion defects in the brain outside the MRI-lesions in the patients with a single lesion, except in one case with a subcortical lesion that showed an extensive perfusion defect in the perisylvian areas on SPECT . These neurological accounts of GAWH are in good agreement with previously reported cases with global aphasia with hemiparesis. Basso et al (1985) and Vignolo et al (1986) already suggested the highly heterogeneous location of the lesion in global aphasia with hemiparesis. The localisation of the lesion is thus not a relevant marker for GAWH.

Gawh and neuroradiology

Keyserlingk et al (1997) compared the CT-scans of people with global aphasia with and without hemiparesis. The motor cortex was free of infarction not only in all GAWH-patients, but also in 7 of the 8 patients with hemiparesis. An infarction of the motor cortex can therefore not be considered to be responsible for the differences between the two groups. Instead, it appears that the extent of the lesion is a valuable parameter for distinguishing the two groups. The frontal display of the CT-scan of people with global aphasia with hemiparesis showed a lesion that extended to the wall of the lateral ventricle, thus including the whole corona radiata with the pyramidal tract. The part of the pyramidal tract, starting in the medial part of the precentral gyrus

and responsible for the innervation of the lower extremities, is interrupted in the central parts of the white matter.

People with global aphasia without hemiparesis, or in those with a transient phase of hemiparesis, a rim of white matter is intact alongside the lateral ventricle and is free of infarction. According to Keyserlingk et al (1997), the internal capsule is supplied mainly by the anterior choroidal artery and the central branches of the medial cerebral artery supply the most lateral part of it.

This should explain the preserved motor function by

1. Leptomeningial anastomosis through which the anterior cerebral artery supplies the medial cerebral artery
2. An incomplete infarction of the medial cerebral artery with an occlusion of the frontal and parietal branches but a preserved blood flow through the central branch.

Linguistic outcome

Although the fact that GAWH patients are quite similar on neurological examination, they are heterogeneous concerning lesion localisation and recovery of language function. Nagaratnam et al (1996) argued that the degree of recovery is dependent on the localisation and the extent of the lesion. GAWH-patients with multiple lesions in both the anterior and the posterior language areas show the most severe global aphasia and the least recovery. This is in contrast to the GAWH patients where a single lesion, either anterior or posterior, causes the global aphasia that show much better language recovery. Accordingly, it is claimed that lesion site and the initial severity of language deficits can predict the outcome in patients with GAWH.

Hanlon et al (1999) distinguished 3 lesion profiles of GAWH:

→ A single temporal lesion without anterior involvement, but with a severely reduced fluency and comprehension deficit

→ A single anterior lesion without temporal involvement, but with a severe comprehension deficit besides the production problems

→ A single lesion with either anterior or posterior involvement.

After 3 months of speech-therapy post-onset the patients with different lesion profiles evolve into different aphasia-subtypes. Lesion analysis showed that persistent GAWH was related to lesioning of the left superior temporal gyrus. Patients with acute GAWH who evolved into a transcortical motor aphasia had lesioning of the left inferior frontal gyrus and adjacent subcortical white matter in common. Patients who evolved into a Wernicke's aphasia were characterised by lesioning of the left precentral and postcentral gyri. These findings indicate that preserved motor function is not predictive of linguistic outcome, whereas the extent and the site of the lesion is a predictive factor.

Case study (mvs)

MVS was a 74-year-old retired professor of African Languages. He had a stenosis of 90 % of the internal carotid artery, for which he had to undergo an endarterectomy in May 1999. The following day he suffered an infarction post-operatively. Two weeks post-onset SPECT scan showed a hypoperfusion of the left frontal cortex and of the right cerebellum. One month post-onset MRI (fig. 1) showed an infarction of the area of the MCA extending to the nucleus lentiformis. The lesion did not extend to the wall of the lateral ventricle.

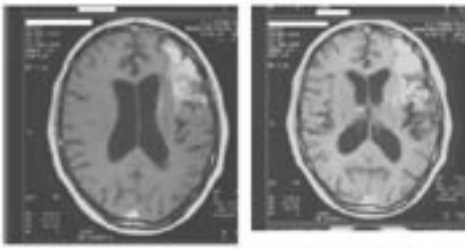


Figure 1. MRI of MVS one-month post onset

This patient showed a transient hemiparesis, which resolved after two days. The first bedside screening showed the linguistic profile of a severe global aphasia with mutism. Two days post – onset he came out of the mutism. We used the principles of Melodic Intonation Therapy, which consists of speaking with a simplified and exaggerated prosody, characterized by a melodic component (high and low) and a rhythmic component (long and short). The reason for his recovery could possibly be found in this method, but the role of spontaneous recovery cannot be ignored. His auditory comprehension was severely disturbed: he was not able to perform on single commands (e.g. Show your nose, show the door). He achieved better on visual association tasks at word-level. (written word – object/picture). He demonstrated severe depression and signs of frustration with his situation.

We did a follow-up of his language function on the basis of two tests, the AAT (Graetz et al, 1992) and the Robert-Van Rumst screening (RVR-screening) (Robert, 1993). In our institution, the second test is used to differentiate global aphasia into 3 groups, each with two subtypes. We distinguish “global global” aphasia, “subglobal “aphasia and “mixed” global aphasia. In each group we have a +type and a –type. The essence of this test is to differentiate the native language from a foreign language. This idea is based on the experiment of Boller and Green (1972). They saw that English native speakers with global aphasia did not change their reactions when they spoke French to them. Also in our institution Robert (1993) spoke foreign languages (Chinese, Polish, German) to the Dutch people with global aphasia. The differences between the foreign languages and the native language vary from large to small. The difference between German and Dutch is much smaller and much more difficult to

differentiate then between Chinese and Dutch. Robert (1993) saw that some patients with global aphasia responded to Chinese stimuli in a similar way as to Dutch stimuli. Other patients could differentiate the Chinese characters and/or spoken words from Dutch written/spoken words, while they had significant problems with the Polish clusters. The patients had to give adequate yes/no answers to the question "is this word/sentence correct Dutch?" to visual and/or auditory stimuli.

A global global aphasia has difficulties to differentiate Chinese characters from Dutch. The difference between the –type and the +type is the patient's behaviour. A –type is apathetic and a +type aggressive. A subglobal aphasia of the –type cannot discriminate Polish clusters (clusters which do not appear in Dutch, e.g. Dobz) from Dutch and a subglobal aphasia of the +type has problems with German clusters (this clusters appear also in Dutch, e.g. Kraft). The third group has no problems at word level with the foreign languages, but has receptive problems with the native language Dutch at sentence level. Robert distinguishes a group of sentences, which were built up with existing words in syntactically correct sentences, although there is no relation between the different parts of the sentence. Each sentence is a collection of words, but without a meaning. Robert named these sentences "hermetic Dutch" (e.g.: the navigation blindfold out of the old / 'de vaart blindt uit'). The mixed global aphasia of the –type has problems with hermetic Dutch. The mixed global aphasia of the +type has no problems with hermetic Dutch, but is not capable to distinguish asemantic Dutch from correct Dutch. The asemantic sentences have a wrong meaning. (e.g.: a rose elephant is flying in the air/'daar vliegt een roze olifant').

On assessment at 3 weeks post-onset MVS was unable to produce any spontaneous speech. Scores on the AAT were shown in Table 1.

Table 1. AAT scores 3 weeks post onset

AAT subtest	Score	Percentiles	Stanines
Token test	50/50	6	-
Repetition	37/150	12	2
Writing	13/90	13	2
Naming	0/120	4	-
Comprehension	37/120	3	1

The score on written language came solely from the subtest reading aloud, any score on dictation (composing and writing) not being possible at this time. In the RVR-screening MVS was able to perform well with the Chinese characters, Polish and German words, but he had problems differentiating hermetic Dutch (3/10) and asemantic Dutch (score: 0/10). The scores on the visual and auditory version were similar. So, according to the RVR-screening, MVS has a mixed global aphasia of the –type.

At 4 months post onset the following AAT scores (Table 2) were found. A statistical analysis with AATP, the psychometrical individual diagnosis of the AAT, revealed a significant improvement on all subtests.

Table 2. AAT scores 4 months post onset

AAT subtest	Score	Percentiles	Stanines
Token test	40/50	35	4
Repetition	94/150	38	4
Writing	78/90	80	7
Naming	58/120	36	3
Comprehension	75/120	35	1

MVS's writing was very good (80 %) compared to 3 weeks post onset (13 %): he wrote words and short sentences in capital letters, without phonological paragraphias. He used his writing capacities as help in spontaneous speech: when he could not say a word or a name he wrote it on a paper and read it afterwards. His spontaneous language was characterized by unintelligible speech. The listener often had to guess the meaning of his utterances. His comprehension was good on word level and relatively good on sentence level. A conversation was possible with the aid of written language and non-verbal gestures.

Fig. 2 shows an example of a conversation about the African countries he had visited. When he couldn't say the name, he first wrote it and read it afterwards.

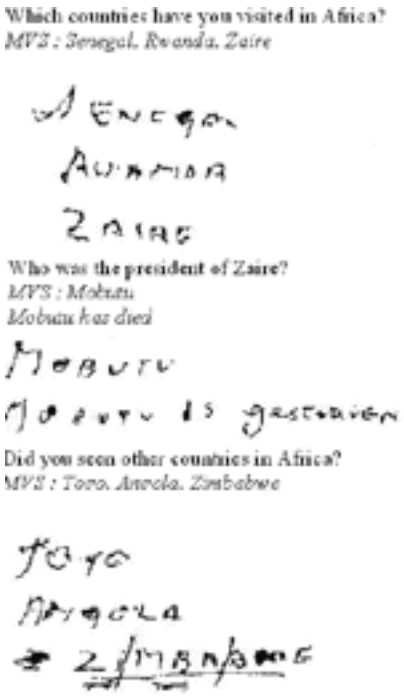


Figure 2. Writing in spontaneous conversation at 4 months post onset

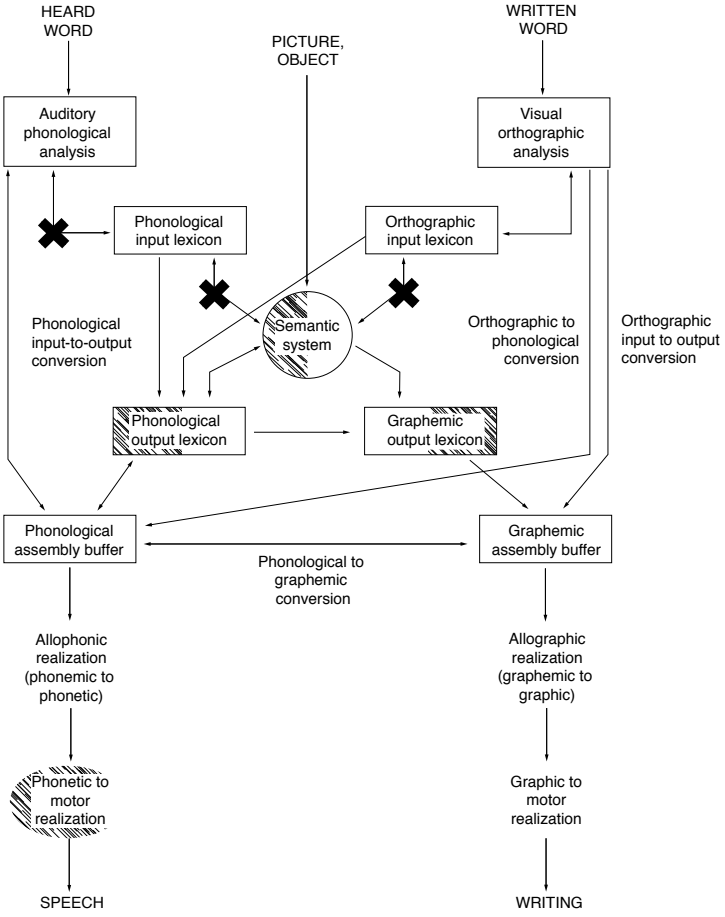
On the RVR-screening he could differentiate hermetic Dutch (score: 10/10) well, but asemantic Dutch remained difficult (score: 2/10). To highlight targets for therapy we carried out neurocognitive linguistic assessment with some PALPA-tasks (Bastiaanse et al, 1995), which results are shown in table 3.

Table 3. Palpa-results 4 months post onset

PALPA - task	**score**
Auditory phonological analysis	
Task 1 : auditory discrimination : minimal pairs, non-existing words	65/72
Task 2 : auditory discrimination : minimal pairs, existing words	72/72
Phonological input lexicon	
Task 5 : auditory lexical decision : imageability and frequency	152/160
Task 6 : auditory lexical decision : morphology	41/60
Orthographic input lexicon	
Task 24 : visual lexical decision : imageability and frequency	154/160
Task 25 : visual lexical decision : morphology	44/60
Semantic system	
Task 45 : word comprehension : matching spoken word - picture	30/40
Task 46 : word comprehension : matching written word - picture	30/40
Task 47 : Synonym judgement (auditory)	55/60
Task 48 : Synonym judgement (visual)	56/60
Task 49 : Semantic word association	High imageable : 8/15 Low imageable : 0/15
Phonological outputlexicon	
Task 52 : verbal naming : frequency	34/40
Graphemic outputlexicon	
Task 51 : graphic naming	38/40

Auditory phonological analysis is disturbed due to a deficit in the bottom-up process. When MVS could retrieve lexical semantic information, as in existing words, he could discriminate well, although when he had no bottom-up information as in the non-existing minimal pairs, he had problems with auditory discrimination. The **phonological /orthographic inputlexicon** was intact, based on the fact that he could differentiate well words with non-existing clusters in Dutch from existing words (cfr. RVR- screening: e.g. 'dobz' versus 'kost'). The scores on tasks 5 and 6 showed that MVS had many problems with compound nonsense words composed of one part existing words (e.g.: 'warmzaam'). These words were labelled by MVS as an existing word, probably due to the disturbed lexical-semantic bottom-up process from the semantic system. Visual scores are better then the auditory results. A **central semantic deficit** was obviously present, because the auditory and the visual information processing were disturbed in the same way: MVS made the same errors in the auditory and visual tasks. MVS made most of all semantic errors and we saw imageability effects: low imageable words were more difficult then high imageable words. The semantic module was still partially intact, because of the bottom-up information to the input lexicons and the relatively good functional comprehension at 4 months post onset. The **phonological output lexicon** and **graphemic output lexicon**

were partially disturbed because he could still retrieve parts of words. His writing was better than his verbal naming. Low frequency words were much more difficult then high frequency words. **Phonetic to motor realisation** was disturbed, based on the presence of an apraxia of speech: his speech was characterized by distortions, slow speech rate, difficult initiation accompanied with visual/auditory groping. The results of this analysis were presented in the model of Ellis & Young (1988), in figure 3. This was the basis to point out the therapy. MVS received lexical-semantic training, with written language as support for the verbal language. Besides semantic training, phonology was also very important, to train the output lexicons. Last but not least, oral production was the ultimate goal of our therapy.



Legend: shading = totally or partially disturbed modules
X = entry disturbances

Figure 3. Language profile of MVS 4 months post onset in the model of ELLIS & YOUNG (1988).

At one-year post onset his AAT profile had developed to that shown in table 4.

Table 4. AAT scores one-year post onset

AAT subtest	Score	Percentiles	Stanines
Token test	43/50	27	3
Repetition	116/150	58	5
Writing	86/90	94	8
Naming	64/120	39	3
Comprehension	88/120	57	3

Fig. 4 shows a good evolution on all speech tasks, but according to AATP, only the changes in the subtest repetition were statistically significant this time. On the RVR-screening, MVS no longer had problems with asemantic Dutch. So, according to this test one can no longer speak of a global aphasia. The alloc-profile assigned a not-classifiable aphasia. The spontaneous writing, illustrated in the text below, was paragrammatic, with intact phonology and morphology.

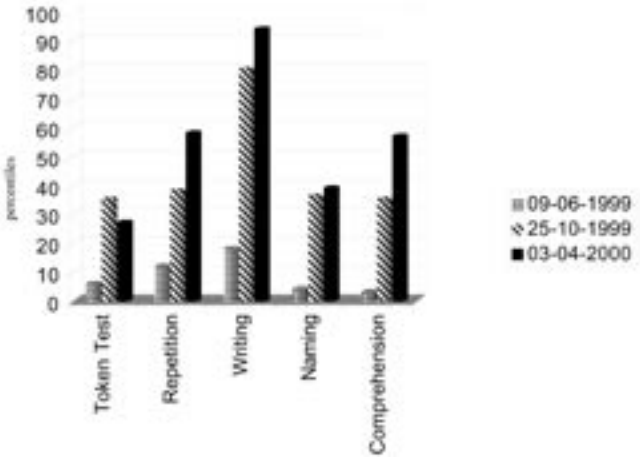


Figure 4. Summary of the AAT-results

Example of MVS's spontaneous writing

Fragment of a letter written to the speech therapist :

> "*Wat is er van het begrijpen van de gesproken taal? Begrijpe wat kan er begrepen worden? Ik kan je het halen wat er van beide individuen gezegd geworden is. Ik kan niet waarop, meer dan drie en meer, ze meer dan aan het vechten zijn...los van alle kwaad en van alle ongemak. Moge ik na deze onvoordelen gelukkig zijn en misschien zeer voordelig, meer dan contextueel, zeer gelukkig meer dan gelukkig zijn.*"

Translation:

> What is it about the comprehension of the spoken language? Understand what can be understood? I can you it get what is there of both individuals said been is. I cannot where up, more then three and more, they are more then fighting… loose from all evil and from all discomfort. May I be after these un-advantages be happy and maybe very advantageous, more then contextual, very happy more than very happy.

On the basis of the frontal lesion localisation, and his relatively good repetition one might think of the diagnosis of transcortical motor aphasia (TMA). According to the AAT – criteria of a TMA, repetition may not be lower then 60%. MVS's repetition scored 58%. The difference between comprehension and repetition may not be more then 20% and spontaneous language must be effortful. MVS's repetition (58%) is one percentile better then his comprehension (57%). All the other subtests must be at least 20% below repetition. In MVS's case the difference between Token Test and repetition is 31 % and the difference between naming and repetition is 19%. But, his writing is 36 % better than his repetition. Therefore, the diagnosis of TMA may not be quite appropriate, especially because of the dissociation between his graphic and oral output. We therefore prefer the term "atypical aphasia" in this case.

Conclusions

As described in many other reports, we argued for the fact that GAWH is a rare type of global aphasia, because of the dissociation between the intact motor function and the severely disturbed linguistic function. We presented a case MVS with GAWH after a single lesion in the left frontal cortex. He evolved after 1-year speech therapy into an atypical aphasia, because of the dissociation between the oral and graphic output. Patients in which there is no clear –cut correlation between the locus of the lesion and the aphasia profile are a challenge to study and describe. A critical objective analysis of the linguistic functions is on the one hand necessary to improve our insight in the brain-language relationships, and on the other hand to point out the guidelines for a successful therapy.

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