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DOES GERSTMANN SYNDROME EXIST?

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SUMMARY

The aim of the study is to present Gerstmann syndrome, manifested as a neuropsychological deficit resulting from the damage to the parietal lobe of the left hemisphere. Here it is discussed based on the studies conducted mainly since the 1950's when it attracted considerable interest, as well as and controversy at the same time. The classic symptoms are briefly described, including the clinical tasks useful in any the diagnosis for during the neuropsychological assessment. The paper also presents recent studies and a alternative different proposal for the understanding of this clinical syndrome.

Josef Gerstmann described a clinical tetrad in his patients, which was later to be known as Gerstmann syndrome. The symptoms included finger agnosia, agraphia, acalculia and left-right disorientation. He associated the above symptoms with damage to the left angular gyrus, hence the alternative a different name for of the syndrome i.e., the angular gyrus syndrome. The existence of the syndrome was questioned for some time, something which was never approved by Gerstmann. Currently, the occurrence of the syndrome is confirmed by studies. However, the full and pure tetrad of the classic symptoms as observed is not common. The clinical picture of the syndrome often usually remains incomplete and is related to other neuropsychological deficits such as aphasia, which frequently occurs. In modern considerations, the language deficiencies of semantic aphasia are not treated as non-Gerstmann syndrome, disturbing its pure form, but are considered to be a part of Gerstmann syndrome as such.

INTRODUCTION

The complexity of understanding the mysteries of the human brain is particularly well illustrated by Gerstmann syndrome (GS). Distinguishing the syndrome caused controversy and disagreement which lasted for many years. The existence of the syndrome was also questioned. The above factors reflected a turbulent history of the syndrome, showing the dialectical evolution of neuropsychological concepts. At the same time, distinguishing the syndrome also resulted in an increased number of studies and conclusions on the structure and functions of the parietal lobe and the recognition of sequelae related to damage to this area of the brain [Rusconi et al. 2010].

JOSEF GERSTMANN AND HIS DISCOVERY OF A NEW SYNDROME

Josef Gerstmann was a physician, now largely remembered for his clinical observations in two areas of neurology. The former was the combination of neuropsychological symptoms defined as GS, which is the subject of the present paper. The latter known as Gerstmann-Sträussler-Scheinker disease (GSS), is an inherited neurodegenerative disease caused by prions, which was described by Gerstmann and colleagues in 1936 [Louis et al. 2017]. Of note, apart from these discoveries, Gerstmann had also other clinical interests and achievements.

Triarhou [2008] presented Gerstmann as one of the pioneers of neurology. The story of the physician reflected a restless and dramatic period of European history at that time. Gerstmann (1887–1969) was born in Austrian Lviv, which he left for Vienna. There he graduated from medical studies in 1912. During World War I, Gerstmann worked as a military doctor in Innsbruck where he treated wounded soldiers of the Austro-Hungarian army, for which he was decorated three times. Between 1918 and 1930 he was a student and co-worker of Julius Wagner von Jauregg who was a Viennese professor of psychiatry and nervous diseases and also the Nobel prize winner for neurophysiology (due to the discovery of the therapeutic value of malaria inoculation in the treatment of dementia paralytica). Gerstmann specialized in nervous diseases and psychiatry and earned the title of professor in 1929. Between 1930 and 1938 Gerstmann was the director of Maria-Theresien-Schlössel, a hospital for nervous and mental diseases in Vienna. However, he was ousted from this position by the Nazis due to his “race”. The growing influence of the Nazis in Europe and the occupation of Austria by the Third Reich (Anschluß) forced him to emigrate due to his Jewish origin (as in the case of other outstanding people of that time, e.g., Otto Loewi). In 1938, Gerstmann and his wife Martha arrived in the United States of America and settled there. They were given support by Paul Schilder, an Austrian psychiatrist and psychoanalyst living in New York who had been a student of Freud. Gerstmann practiced medicine and conducted scientific research in the United States, however, did not achieve a professional position comparable to the one

he had had in Vienna. In 1942 Gerstmann was stripped of his medical degree by the Nazis which was reinstated as late as in 1955. In addition, the property of the scientist was confiscated in Vienna, which led to post-war battles to regain it. Gerstmann did not return to Europe and lived in New York City until his death. The life of Gerstmann was presented in detail by Zeidman et al. [2015].

While still in Vienna in 1924, Gerstmann wrote a paper entitled *Finger agnosia. A selective disturbance of localization on one's own body*. The study initiated a series of seven papers describing the syndrome of four neuropsychological deficits. These papers were published in 1927, 1930, 1931–1932, 1940, 1957 and 1970 (the first four in German whereas the last three in English). Gerstmann gradually isolated a tetrad of symptoms, starting from finger agnosia in 1924. In 1927, Gerstmann defined finger agnosia with agraphia as a new syndrome and included acalculia and left-right disorientation in the syndrome in 1930. Finger agnosia is an inability to recognize fingers that is not the result of sensory deficits. Agraphia is an acquired disorder of writing skills, while acalculia is an acquired impairment of arithmetic skills. Left-right disorientation is a disorder to distinguish right from left in relation to the body scheme. Gerstmann recognized their coexistence as a syndrome, which had not been previously described. The four distinguished symptoms were given a hierarchical structure. The scientist considered finger agnosia and agraphia to be two most crucial features of the syndrome (*Kardinalsymptome*), while acalculia and left-right disorientation were recognized as secondary symptoms, which were less characteristic. In addition, impaired reading skills, constructional apraxia, color anomia, right-sided hemianopia and reduced optokinetic nystagmus were distinguished by Gerstmann as the symptoms that at times co-occurred with the syndrome, however they were not a part of it. Those 'bordering' symptoms are characterized by lower severity and higher variability of prevalence in respect to the symptoms that form this syndrome. Gerstmann did not include agnosia other than finger agnosia, apraxia other than constructive or no form of aphasia as a part of the syndrome, nor did he accept them as 'bordering' symptoms [Gerstmann 1924; Lebrun 2005; Rusconi et al. 2010].

From the 1930's Gerstmann emphasized two aspects of the syndrome, namely functional connections of the components of the syndrome and highly diagnostic localizing value. The scientist treated the combination of the symptoms of the syndrome, which may initially seem surprising, as a consequence of their mutual relationships. The conceptual representation of single fingers and their laterality is important in the writing process and in the acquisition of counting skills, while left-right disorientation is particularly associated with this part of the body. In other words, this clinical tetrad has a basic psychological mechanism (*Grundstőrung*), which explains its occurrence. As a cognitive denominator of the syndrome Gerstmann assumed a selective disorder of the body schema limited to one part of the body i.e. to fingers (known as selective autotopagnosia). Gerstmann believed that the visual-tactile-sensory representation of the fingers did not function normally in the body schema and thus underwent disintegration. This results in the occurrence of symptoms in relation to one's own body, but

also to another person due to the fact that adequate orientation in the scheme of one's own body (fingers) determines the normal perception of the body of another person. Moreover, bearing in mind the results of post-mortem examinations from his own medical practice and the review of literature, Gerstmann considered this syndrome to be the manifestation of the focal, unilateral damage to the left parietal lobe in right-handed patients. The lesion involved the inferior part of the parietal lobe extending to the middle occipital lobe. The researcher paid special attention to the angular gyrus (AG) in the region adjacent to the occipital area. The AG is marked as BA39 area on the Brodmann cytoarchitectonic map of the cerebral cortex [Gerstmann 1957; Lebrun 2005; Rusconi et al. 2010].

In his paper of 1924, Gerstmann described his first case of a 52-year-old female patient with cerebral arteriosclerosis who developed agraphia and acalculia due to left-hemisphere stroke. Aphasia and alexia were not observed. However, in relation to the reading skills, Gerstmann observed that reading a longer text was associated with an increased effort of the patient that was not observed when the patient read shorter texts. The patient made mistakes in relation to her own fingers when she was requested to point them by moving an individual finger defined in the instruction and when was asked to name particular fingers. The mistakes of the patient were also associated with indicating and naming the fingers of the examiner. Gerstmann termed this symptom *finger agnosia* (*fingeragnosie*). The above difficulties were mainly related to fingers. The patient made mistakes less often when she was requested to indicate and name other parts of her body and the examiner's body. The patient had made occasional mistakes by indicating body parts other than fingers before the body side was defined or other parts of the body were ipsilateral to the hand the patient used at the time of pointing body parts. Proper indication of the ipsilateral body part using the hand meant understanding of the meaning of the concepts of the left and right parts of the body. However, the patient had great difficulty in fulfilling the requests related to her own body or the body of the examiner if these requests were connected with differentiation between the body sides, i.e. they included the terms *left* and *right* with respect to the opposite side of the body. The difficulties were related to mistakes that were made when the patient indicated the contralateral body part with her hand i.e. the patient indicated the ipsilateral body part with the proper hand or indicated the proper side and the part of the body with the hand which was inconsistent with the instructions. Difficulties in differentiating body parts of another person also occurred in the reproduction of the movements of the examiner which the patient performed in mirror-image. Additionally, the patient was diagnosed with right-sided hemianopia. Three years later, Gerstmann presented two more cases of patients with a similar tetrad of symptoms although less pronounced compared to his first reported patient. Furthermore, in these two patients Gerstmann found constructional apraxia, color anomia and impaired reading of numbers [Gerstmann 1924; Lebrun 2005; Rusconi et al. 2010].

This syndrome remained a subject of long-term interest of Gerstmann. The scientist was also professionally involved in it after emigrating to the United

States. However, it was the first article in English in 1940 preceded by four different papers in German that made his observations on the syndrome more common. At present, the eponym *Gerstmann syndrome* is commonly used in literature and clinical practice. It was used for the first time by Gerstmann in the title of his paper in 1957. The last article from the series of papers based on his notes was published by his wife in 1970, shortly after his death [Gerstmann 1957; Gerstmann & Gerstmann 1970].

CONTROVERSY SURROUNDING GERSTMANN SYNDROME

In the 1950's and 1960's, the syndrome described by Gerstmann became the subject of controversy. A heated debate was held mainly between Gerstmann and Arthur L. Benton who was an American professor of neuropsychology.

In 1956, Benton and Meyers published a paper under a significant title *An early description of the Gerstmann syndrome*, in which they questioned Gerstmann as the first scientist who had reported finger agnosia and the syndrome itself later named after him. They believed that the discovery had been done earlier by Jules A. Badal, a French professor of ophthalmology. That was not observed by neurologists due to the fact that Badal had published his paper in the ophthalmology journal. In 1888, 36 years before Gerstmann, Badal had described a case of a 31-year-old patient, Valérie Clem affected with eclampsia resulting in a variety of deficits, including the tetrad of symptoms listed later by Gerstmann. For example, the woman confused the names of fingers when she was requested to say which finger of her hand was touched or when she named the fingers in accordance with their order. She had also problems to differentiate between body sides and the sign of the cross was helpful because she knew she used the right hand for it [Benton & Meyers 1956; Lebrun 2005].

Soon, Gerstmann [1957] responded to Benton and Meyers by publishing another paper. The scientist expressed the opinion that in Badal's patient GS was not present and also stressed the fact that he had been the first author reporting finger agnosia. Gerstmann paid attention to visual-spatial disorders of the patient, expressed by considerable problems in orientation in the extrapersonal space (e.g. the location of external objects and their spatial relationships). It did not correspond to the clinical tetrad described by Gerstmann where such deficits did not occur. Gerstmann explained the symptoms of GS by a selective disturbance of the body image but not by the general spatial deficit as it had been adopted by Badal in the case of his patient. Referring to his clinical experience and the experience of other researchers, Gerstmann related the symptoms of the patient to bilateral cerebral pathology in the parietal or parieto-occipital area, but not to the unilateral lesion as in GS. He believed that noticing the tetrad of symptoms of GS in the patient required from the examiner to consider the possibility of the clinical condition different from the conditions previously adopted as typical and to perform careful patient examination. Gerstmann did not feel inclined to accept

the concept of the *incomplete Gerstmann syndrome* in which four specific symptoms were not present and considered the cases of such patients reported in the literature to be a result of insufficient examination.

In turn, Benton [1961] wrote a paper entitled *The fiction of the "Gerstmann syndrome"*. The conclusion in the title was formulated based on his study on a group of 100 patients diagnosed with various cerebral pathologies. Benton examined four skills known to be impaired in GS. Moreover, Benton regarded three domains reported in the literature as dysfunctional in patients with this (often incomplete) syndrome, i.e. constructional praxis, reading and visual memory. In none of the patients Benton found the pure tetrad of GS without the presence of additional deficits. Four symptoms occurred in 10 patients and were a combination of GS and non-GS symptoms. In the subgroup of 12 right-handed patients with the unilateral pathology of the left parietal lobe, Benton diagnosed 3 symptoms of GS in 4 patients that in each case co-existed with other deficits. He did not find the deficits forming GS to co-exist more significantly compared to non-GS deficits. Similarly, none of the other combinations of symptoms considered by Benton occurred significantly more frequently. Therefore Benton concluded that there was no basis for the isolation of GS, which he described as "an artifact of defective and biased observations". Benton believed that the selective attention of researchers searching for the tetrad of specific symptoms affected the method of study and conclusions, resulting in the diagnosis of these deficits and disregard of other deficits.

Another study conducted by Heimburger et al. [1964] supported Benton's criticism of GS. Those authors examined 456 (or 465 - both numbers are provided in their paper) various neurological patients with brain damage, such as cerebrovascular disease, brain tumor or neurotrauma. 111 patients were diagnosed with one or more symptoms of the syndrome. Finger agnosia was the least prevalent symptom. In this subgroup, 23 patients presented with four specific symptoms. In agreement with Benton, those authors did not describe the pure form of the syndrome without non-GS deficits. All patients with all GS components were also diagnosed with the aphasia syndrome, which was not further specified. Those authors found no basis for treating GS as an autonomous entity, but they considered these symptoms to merge with numerous different neurological deficits, mainly dysphasia. In addition, they paid attention to the higher value of symptoms isolated by Gerstmann in relation to inference about the hemispheric lateralization rather than the intrahemispheric location. However, if there were more symptoms of the tetrad, the accuracy of the inference about the intrahemispheric location increased. Patients with four deficits usually presented with damage to the left cerebral hemisphere, more often located in the posterior part of the parietal lobe. However, despite such localization of damage, cases without symptoms indicated by Gerstmann were observed at times. In general, patients with this clinical tetrad had an extensive brain damage extending beyond the AG. It also involved the supramarginal gyrus, the posterior part of the superior temporal gyrus and the anterior part of the occipital lobe. At the same time, lesion to the AG

was not detected in some patients in the post-mortem examination despite previous diagnosis of the tetrad of neuropsychological deficits suggestive of the lesion.

Poeck and Orgass [1966] were of the same opinion after examining 50 patients with cerebral damage. They considered GS to be an arbitrary combination of neuropsychological deficits adopted by Gerstmann, which did not meet the selectivity criterion, co-occurring with non-GS deficits, especially with aphasia.

Concluding the above studies, two facts contributed to the controversy related to the existence of GS i.e. a rare occurrence of the full and pure form. The syndrome was usually diagnosed in the form that did not include the specific tetrad of symptoms. Instead of the full presentation, cognitive deficits of another type coexisted, especially aphasia, constructional apraxia or alexia. In addition, it should be borne in mind that despite the medical development in the 1950's and 1960's, advanced contemporary neuroimaging methods such as computed tomography (CT) or magnetic resonance imaging (MRI) had not been applied to assess brain damage. The presence and the location of a lesion were based on neurological examination, skull radiogram, electroencephalogram (EEG) or pneumoencephalogram. Less frequently the the lesion was confirmed based on observations at the time of neurosurgical procedures or post-mortem examination [Ardila 2014a].

FURTHER STUDIES ON GERSTMANN SYNDROME

Studies on the groups of patients showed that the full and pure GS is not common in clinical practice. However, papers released from the 1980's 1) described cases of individual patients with such combination of symptoms and the confirmation of pathological brain changes using neuroimaging techniques and 2) indicated the possibility of generating the syndrome during direct cortical electrical stimulation.

Case reports of patients with Gerstmann syndrome

In 1983, Roeltgen et al. reported the first case of a patient with the full and pure GS and brain damage confirmed by head CT. In a 62-year-old right-handed man, a focal lesion was observed in the left hemisphere following ischemic cerebral infarction. It involved mainly the superior AG and the posterior supramarginal gyrus with the extension into the posterior and inferior aspects of the superior parietal lobule. During the subacute period of stroke in the clinical tasks, the patient presented with a tetrad of GS. Acalculia was the most prominent symptom with difficulties in arithmetic operations on single-digit numbers (e.g. 9+3=?). Other cognitive dysfunctions were not observed, including aphasia (the Western Aphasia Battery) and constructional apraxia (drawing tasks). Those authors concluded that cerebral damage in the area critical to GS was noted in their patient. A rare diagnosis of the full and pure GS was explained by the fact that brain lesions are rarely restricted as in the case of their patient. Those researchers believed that damage extending outside of this particular brain area would have an influence on the modification of clinical presentation [Roeltgen et al. 1983].

Mayer et al. [1999], 16 years after publishing the article by Roeltgen et al., reported a case study of another patient with the full and pure GS that was considered to be one of the most investigated cases. A 59-year-old right-handed man presented with a tetrad of symptoms of GS due to an ischemic lesion without the presence of other neuropsychological disorders and neurological symptoms. Within 3 months following stroke, the psychological examination was performed with the use of a number of methods, which previously had not been applied for this tetrad. The following were used: the Boston Diagnostic Aphasia Examination, the Rey Auditory-Verbal Learning Test, the Rey-Osterrieth Complex Figure, the Wisconsin Card Sorting Test, the Raven Progressive Matrices, the Wechsler Adult Intelligence Scale-Revised and the Wechsler Memory Scale-Revised. On the basis of the cognitive examination, language, praxis, memory and intelligence disorders were excluded. A selective impairment was demonstrated in the tasks involving number processing. Brain MRI showed a focal ischemic lesion situated subcortically in the inferior part of the left AG. However, the cortical layers were spared. It was related to the interruption of associative fibers from the angular gyrus to the superior parietal area and commissural fibers connecting this lobe with the homologous area of the right hemisphere, i.e. intra- and interhemispheric (interparietal) disconnection. The follow-up MRI examination done 3 years after the stroke revealed atrophy of the splenium of the corpus callosum, corresponding to the interrupted fibers. In addition to the tetrad of GS symptoms, the patient was also diagnosed with medial toe agnosia, i.e. the localization similar to that observed in finger agnosia in this patient and other patients. According to those authors, it excluded the Gerstmann hypothesis of a selective disorder of the body schema as a common factor limited to fingers, but it suggested that the deficit was common for hands and feet. They adopted a hypothesis that the fundamental deficit connecting the symptoms of this syndrome was the disturbance in processing visuospatial images, i.e. performing mental transformations of visuospatial images e.g. during mental rotation. In other words, GS could be diagnosed without this deficit. Those authors considered normal processing of visuospatial images to be an essential function for the realization of the processes that were impaired in this syndrome. In this patient those authors indicated writing errors as the exemplification of the disruption of this transformation. Those errors were limited to four letters, which showed spatial proximity. The errors consisted in the confusion of ‘b’ and ‘p’, ‘d’ and ‘q’ letters when no visual model was available. All of them are formed from two graphical parts (known as letters with two strokes), the structures of which are differentiated by the circle and the adjacent ascending or descending stroke. Top-bottom letter reversal was observed in the patient, however the left-right shift was not detected, which did not occur when these letters were copied. Moreover, reviewing the literature on the syndrome, those authors formulated the conclusions on 1) the left parietal lobe lesion in right-handed patients if GS was present without the accompanying symptoms or with mild severity of symptoms; 2) the left-sided lesion of parietal, temporal and occipital lobes or the left thalamus in right-handed patients if GS co-occurred

with other distinct symptoms such as aphasia, apraxia, sensorimotor deficits; 3) the possibility of the occurrence of this syndrome in left-handed patients with damage to the right parietal lobe; 4) parietal activation in neuroimaging studies when mental visuospatial transformations were performed.

The issue of toe agnosia was previously [1997] discussed by Tucha et al. who diagnosed the full and pure GS in a 72-year-old right-handed woman with glioblastoma multiforme in the left angular and supramarginal gyri. In addition, examination revealed the presence of toe agnosia, which was bilateral and involved all toes. During the investigation, the toes were given the following names: the big, second, third, fourth and little. The patient named her toe that was touched by the examiner or indicated the corresponding toe on the other foot, with or without visual control. Those authors introduced the term *digit agnosia*, which is broader in meaning compared to finger agnosia and indicated the area for further research to determine whether toe agnosia remained a permanent component of GS.

Direct electrical stimulation of the cerebral cortex in the left parietal lobe

Further evidence for the existence of GS was provided by the studies of patients with electrical stimulation of the cerebral cortex.

In 1984, Morris et al. described electrocorticography in a 17-year-old right-handed male with refractory epilepsy. In this diagnostic method, electrodes are placed on the brain surface, which allows recording of neuronal bioelectric activity and identification of the epileptogenic focus, initially detected using electrodes on the cranial skin. It also allows precise cortical mapping, i.e. recognition of functional organization of the cerebral cortex through direct electrical stimulation of the cortex and temporal impairment in cortical functioning. It is used to locate key structures (functionally eloquent cortex), which should be preserved during the resection of the epileptogenic area. Damage to the eloquent area, not necessarily related to speech, results in patient disability. The need for the determination of the location of such brain regions, prior to the main stage of neurosurgical procedure, is related to inter-individual variability that occurs in this respect. In the above patient, the neurological examination and head CT did not show changes. The EEG revealed a left parieto-occipital spike focus. The intracarotid amobarbital procedure (Wada test) showed the dominance of the left hemisphere for speech. Left temporoparietal craniotomy was performed and a subdural electrode array was placed on the posterior cortex. The studies were conducted for 4 weeks, prior to the removal of the epileptogenic area. Electrical stimulation of this region of the cerebral cortex resulted in the occurrence of all GS symptoms within a relatively small area of the cortex located below two electrodes E4 and F4, without revealing other deficits. Below this location, some components of GS syndrome were generated with the non-GS symptoms i.e. anomia, alexia and/or constructional apraxia. On this basis, Morris et al. considered the occurrence of the full and pure GS probable, which was due to a rare and small focal lesion in

the left AG. Coexistence of GS symptoms with the above deficits was indicative of more extensive brain damage involving also the cortical area inferior to the AG. At the same time, a double dissociation was discovered in the tasks of finger gnosis i.e. cortical stimulation using electrode E4 resulted in problems with holding up a particular finger at the request of the examiner without interfering with naming the fingers. However, stimulation of electrodes E5, D4 and D7 resulted in finger anomia with no problems related to holding up fingers at the request of the examiner [Morris et al. 1984].

Roux et al. [2003] paid attention to the insufficient number of studies on neurosurgical patients using electrical stimulation of the AG. To better understand the functional organization of this region, Roux et al. examined 6 right-handed patients (five with left and one with right hemisphere tumors) using the above method 19 years after the observations of Morris et al. [1984]. In all these patients the tumor was located near the AG. Four patients preoperatively showed discrete symptoms of the incomplete GS. This syndrome, however, was not present in the other two patients. None of the subjects was diagnosed with language or apraxic deficits. Direct brain mapping was performed on the side of the tumor prior to its resection as part of awake brain surgery (lightened anesthesia and maintaining verbal contact with the patient during surgery). During cortical electrostimulation, patients performed clinical tasks related to writing, calculating, finger recognition and color-naming sometimes reported as a dysfunctional domain in patients with this syndrome, in addition to the standard object-naming and reading tasks. The intraoperative assessment of the left-right orientation was not performed. In each patient interferences in writing, counting and finger recognition during electrical stimulation of the left and also right AG were observed. The stimulation sites resulting in acalculia and finger agnosia were observed in the supramarginal gyrus and close to the intraparietal sulcus. Therefore, provoking GS symptoms was not limited to the left hemisphere or the AG. The areas resulting in interferences in color- or object-naming and reading were observed in or near the AG (in the superior temporal, middle temporal and supramarginal gyri). Locations specific to one of the analyzed functions were noted. Non-specific locations involved in more than one function were also observed. Concluding their studies, Roux et al. reported that GS and non-GS symptoms could be found during electrical stimulation of the AG, which reflects commonly reported fact of the coexistence of a differentiated combination of symptoms with damage to the AG and uncommon occurrence of the pure form of this syndrome. Such a variant of GS resulted from a cerebral lesion, which did not interfere with other cognitive functions, e.g. language functions were spared, which was considered by Roux et al. a rare clinical condition. Those researchers postulated that brain mapping procedures in the region of the AG should not be limited to the standard word retrieval task [Roux et al. 2003].

Referring to the critical opinion of Benton, on the basis of patient cases reported in the literature and the findings of electrical stimulation of the cerebral cortex, the researcher revised his former view which questioned the existence

of GS in the 1990's. Eventually, Benton [1992] accepted the possibility of the occurrence of the tetrad of this syndrome in the pure form or the form combined with other symptoms with left-sided damage to the parietal, posterior or temporo-parieto-occipital area. Thus, he appreciated "Gerstmann's diagnostic acumen (or his imagination)". Benton noted the possibility of the occurrence of this syndrome as a result of the subcortical lesion located in the left posterior thalamus without the structural damage to the cerebral cortex. At the same time, Benton questioned the validity of distinguishing this syndrome from different possible combinations of symptoms and other lesions of the posterior parietal lobe or the temporo-parieto-occipital junction also generated during direct cortical electrical stimulation. According to Benton, such combinations of symptoms did not gain so much attention as GS, even though they deserved similar attention.

CLASSIC TETRAD OF GERSTMANN SYNDROME

Finger agnosia – finger dysgnosia

The dominant significance of the fingers for everyday human activities is clearly illustrated by the sensory and motor homunculus that is the representation of the human body in the cerebral cortex. Gerstmann [1924, 1957] paid attention to this part of the body describing gnosis disorder related to fingers i.e. impaired identification and differentiation of individual fingers, impaired finger naming and indicating when requested. Performing clinical tasks may be completely or partially impaired and it does not result from the lack of knowledge about fingers (unfamiliarity with the following names: thumb, index finger, middle finger, ring finger, little finger). Finger agnosia is bilateral and is related to the patient's fingers and fingers of another person. Recognition of the other body parts is preserved. Fingers are relatively poorly differentiated structures, which may contribute to the difficulties in their discrimination. At the same time finger position in relation to other fingers is strictly defined. However, the thumb and the little finger are more resistant to recognition disorder. Gerstmann explained it by the fact that the shape and the external location of the fingers distinguished them from other fingers. Patients with finger agnosia usually have the most difficulty with the other three fingers, which was confirmed in further studies [Kinsbourne & Warrington 1962; Mayer et al. 1999].

Gerstmann [1957] and other authors [Roux et al. 2003; Ardila 2014b] attributed a crucial role in the phylogenetic and ontogenetic development of the ability to count and write to the fingers and their conceptual representation. Currently, the decimal number system is commonly used in everyday life. The system is based on ten digits, which corresponds to the number of fingers. The relationship between fingers and counting is evident in some languages, e.g. in English where the word *digit* means a finger and a number. Such a relationship, however, is not observed e.g. in Polish. Neuroimaging studies [Zago et al. 2001; Roux et al. 2003] confirm the relationship between counting and the fingers, indicating the activation of the

precentral gyrus in the part corresponding to the hand and fingers during mathematical processing. In the process of acquiring arithmetic skills, children first count on their fingers, which provides visual-spatial support before doing mental arithmetic. Similar spontaneous behavior can be observed in adult patients with the acquired calculation disorder and the aphasia syndrome (our own observation).

The basic clinical tasks related to finger gnosis referring to the classic Gerstmann's tasks [Benton 1961; Heimburger et al. 1964; Mayer et al. 1999] are as follows:

- naming one's own fingers of both hands, according to the finger order;
- naming individual fingers of each hand or moving fingers, depending on the tactile stimulation i.e. after being touched by the examiner;
- pointing to individual fingers of one's own hands, of the examiner's hand or on the diagram (a schematic representation of both hands) on verbal command.

Kinsbourne and Warrington [1962] in their paper on finger agnosia emphasized the fact that the examination procedures introduced by Gerstmann were clearly dependent on the language, and thus were affected by sensory (receptive) and motor (expressive) aphasia, especially if it was significantly increased. They proposed their own tasks of finger gnosis, limiting the influence of the linguistic component, such as the In-between Test, the Two-Point Finger Test and the Finger Strip Test. These tasks were also recommended by Rusconi et al. [2010].

In the study on various aspects of finger gnosis, Mayer et al. [1999] paid attention to visual control, which could facilitate finger recognition by the presence of specific characteristic features such as a ring, distorted appearance or tattoo. They postulated the use of the tasks of finger gnosis with and without visual control that were based only on the internal visualized representation of the fingers.

In GS, finger agnosia affects both hands. Benton [see Strauss et al. 2006] reported that Henry Head in the 1920's described the unilateral impairment of finger identification that was a type of sensory defect, also resulting from the damage to the parietal lobe and that should be differentiated from a bilateral disorder in GS. However, Brown [see Strauss et al. 2006] paid attention to the possibility of an impairment of the crossed finger localization, as opposed to the uncrossed localization associated with the dysfunction of the corpus callosum e.g. in multiple sclerosis. Then, the patient revealed the difficulties in pointing the finger corresponding to the one that was touched by the examiner on the other hand. It was due to the disorder of interhemispheric transfer of tactile information essential to perform this task.

Agraphia – acquired dysgraphia

Agraphia is a disorder resulting in a loss of writing skills, while acquired dysgraphia is a disturbance of this skill due to brain pathology. As a result, transparency and legibility are lost [Gerstmann, 1957].

The following three types of agraphia are classically reported [Mayer et al. 1999; Roux et al. 2003]:

1) aphasic agraphia, jargonagraphia: paragraphias, i.e. additions, omissions (elisions) or substitutions of letters with well formed letters, pseudowords instead of words; usually a left temporal or temporoparietal lesion;

2) apraxic or apractic agraphia: defective forms of letters, deeply modified letter forms, scrawl in the most severe form, i.e. disorder related to graphomotor pattern of letters; a left parietal or temporoparietal lesion;
3) spatial agraphia: improper management of space on the paper, obvious difficulties in maintaining horizontality in writing, uneven writing or disproportionate use of the sheet space with a large margin i.e. impaired spatial organization (topography) of writing; a right parietal lobe lesion.

The traditional classification of agraphia shows that linguistic and non-linguistic functions are involved in complex writing ability. Depending on the impairment, a newer perspective distinguishes central and peripheral agraphia. According to the literature on GS, the prevailing opinion is that central agraphia is not present in GS unlike peripheral agraphia of the apraxic type [Mayer et al. 1999; Rosca 2007; Ardila 2014a].

Standard clinical tasks for examining writing skills [Benton 1961; Heimburger et al. 1964; Mayer et al. 1999] on the unlined paper include:

- unassisted writing of the following: subject's own first and last names, lowercase and uppercase letters of the alphabet (as the examples of automatic writing), sentences and a text of several lines;
- writing to dictation of letters, one- and multi-syllable words, simple and complex sentences;
- copying of letters, one- and multi-syllable words or sentences.

Acalculia – acquired dyscalculia

Acalculia is an impairment related to loss of mathematical skills whereas acquired dyscalculia is a disturbance of these skills, resulting from brain pathology [Gerstmann 1957]. The use of numerical concepts and arithmetic operations cause difficulties.

As a heterogeneous impairment, it includes the following different categories [Mayer et al. 1999; Roux et al. 2003; Ardila 2014a]:

1) primary acalculia (anarithmia, anarithmetia, arithmetia): a deficit directly related to computational ability; a left parietal lobe lesion, especially of the intraparietal sulcus and the AG;
2) secondary acalculia: calculation impairment is a derivative of the deficit of another cognitive domain related to counting, e.g.:
   - aphasic acalculia due to primary linguistic impairment; left hemispheric lesion;
   - spatial acalculia, which is a disorder of spatial organization of numbers i.e. the visuospatial factor; right hemisphere lesion.

Operations on single-digit numbers are automatic counting whereas the processing of multi-digit numbers requires an adequate interpretation of the spatial organization of individual digits (their order and position), e.g. the meaning of 5 in 57, 75 and 575 [Roux et al. 2003].

The typical clinical tasks used for examining counting skills [Benton 1961; Kinsbourne & Warrington 1962; Mayer et al. 1999] are as follows:

- performing basic mathematical operations of addition, subtraction, multiplication, division in memory and writing using one- to three-digit numbers;
- reading numbers written in the form of digits or letters, reading signs of arithmetic operations;
- writing numbers to dictation using digits or letters, completing the missing mathematical signs in basic arithmetic operations.

Left-right (L-R) disorientation, left-right confusion

The left-right disorientation of the body parts is related to the lateral orientation focused on one's own body or the body of another person. It is also known as somatolateral agnosia. It does not include an impaired ability to distinguish between the sides of objects other than the human body. Nor does it result from the patient's unfamiliarity with the connotation of the words *left-right* [Gerstmann 1957].

Assessment of left-right orientation typically includes [Benton 1961; Heimburger et al. 1964; Mayer et al. 1999]:

- simple commands: indicating the selected body part on the defined side, sidedness of the pointing hand remains undefined, e.g. 'Please show me your left ear';
- complex uncrossed commands: commands to indicate a selected body part on the defined side, movement executed with the ipsilateral hand, e.g. 'Please put your left hand on your left knee';
- complex crossed commands: commands to indicate a selected body part on the defined side, movement executed with the contralateral hand, e.g. 'Please point your left ear with your right hand'.

The above commands are executed with or without visual control and may be related to one's own body, the body of another person or a model of the human body in the form of a picture. In the case of a human body other than one's own, the spatial perspective may be differentiated by the body position in relation to the subject in the same or opposite direction. It is more problematic to meet the crossed commands compared to the uncrossed ones and to distinguish between the sides of another person's body than one's own body, especially if another person is in the opposite position [Rosca 2007; Rusconi et al. 2010].

Heimburger et al. [1964] introduced impracticable commands, e.g. 'Please put your left hand on your left elbow' to distinguish between left-right disorientation and sensory aphasia. The lack of the patient's response which indicates impossibility to fulfill the command is suggestive of impaired speech understanding.

CONTEMPORARY UNDERSTANDING OF GERSTMANN SYNDROME

Rusconi et al. [2009, 2010] referred to the study results of cortical electrostimulation in which the complete GS was not obtained from a single cortical locus

and formulated a hypothesis according to which the syndrome was due to disconnection. The lesion resulted in disconnection of various neural networks involved in cognitive functions, leading to neuropsychological disorders that form this clinical syndrome. Those authors assumed that the occurrence of a specific tetrad was the effect of damage to the structure of separate but spatially convergent fiber pathways and not the result of the disorder of the underlying psychological mechanism expressed through these four symptoms. Therefore, Rusconi et al. did not search for a cognitive explanation of the common expression of symptoms, assuming that such a functional commonality between them did not exist. In other words, the tetrad was structural, not functional. Five healthy right-handed individuals were examined. A small sample was adequate for thorough examination and an in-depth analysis of the results, considering interindividual differences. Studies were conducted with the use of combined structural and functional MRI and activation paradigms in the form of cognitive tasks under experimental and control conditions, corresponding to each of the four functions impaired in GS. The brain activity was recorded during the individual tasks showing the areas in which the damage would most likely result in the occurrence of symptoms of the syndrome. Then, those authors tested for the overlap of cortical activation patterns and fiber bundles associated with the four different domain-related cortical activations. They observed the clear activity of the left parietal cortex in several places during the cognitive tasks. The activity, however, was different, depending on the individuals. The area of cortical activation common for all the analyzed cognitive domains was detected in none of the individuals. In each subject, however, Rusconi et al. discovered a small region of subcortical parietal white matter ('hot spot') connecting various activated parietal cortical areas with the location similar to that previously reported by Mayer et al. [1999]. On that basis, Rusconi et al. concluded that the pure and full GS was the result of intraparietal disconnection. Unlike Gerstmann, those authors did not associate it with a cortical lesion, but with the damage to a focal region of subcortical parietal white matter. The rare prevalence of this clinical condition in its pure and full form accounted for a low number of studies on the syndrome in the literature. Those authors also noted that extensive damage to the dominant parietal lobe led to the symptoms of this syndrome among other neuropsychological deficits (aphasia, apraxia), which made it "virtually impossible" to test for GS [Rusconi et al. 2010].

Osawa and Maeshima [2009] reported a case that confirmed Benton's [1992] observation on the possible occurrence of GS as a consequence of damage to the posterior part of the left thalamus. Those authors diagnosed a complete tetrad of the syndrome due to hemorrhage in the thalamus in the absence of aphasia in a 78-year-old right-handed male patient. Based on structural and functional neuroimaging methods (CT, MRI, single-photon emission computed tomography), damage to the posterior part of the left thalamus was observed. Additionally, decreased regional cerebral blood flow was noted in the left temporoparietal lobe, mainly in the AG. No lesion of the cerebral cortex was observed. Based on neuroimaging results, those authors concluded that projection

fibers connecting the thalamus and the AG were damaged, which secondarily resulted in decreased functioning of the cerebral cortex in the angular gyrus. The set of methods of neuropsychological assessment included psychometric tests useful for the diagnosis toward GS, namely the Finger Localization Test and the Right-Left Orientation Test, developed by Benton and discussed in more detail by Strauss et al. [2006]. It should be noted that much earlier in the Japanese literature Ono et al. [1985] had reported a similar case of a patient with GS due to left thalamic hemorrhage without aphasia.

Most frequently reported cases of patients with this syndrome were related to right-handed patients with a pathological brain change in the left hemisphere. In turn, Nicastro et al. [2017] presented a case of a 57-year-old male patient with GS who was ambidextrous as a result of a switch in handedness (left- to right-handedness). Neuroimaging examination revealed a right cortico-subcortical parietal hemorrhage and arteriovenous malformation (Spetzler-Martin grade III) bleeding into the cerebral tissue. Neurosurgical resection of the malformation was performed in the patient. The clinical presentation of GS showed severe left-right disorientation, moderate acalculia for subtractions, mild finger agnosia with left-side predominance and minor agraphia. Contrary to the prevalent opinion on this syndrome reported in the literature, according to which peripheral agraphia is a component of GS, central agraphia was adopted as a symptom of the tetrad of GS in that patient. It was lexical (surface) agraphia in which the linguistic component is impaired in terms of lexical processing. Ullrich and Roeltgen [2012] presented a similar opinion on this form of agraphia in GS. Those authors described lexical agraphia as frequently coexisting with the aphasia syndrome. The potential explanation for adopting different forms of agraphia can be attributed to the differences in the understanding of the GS model, which originally did not include aphasia, as opposed to the contemporary approach, according to which aphasia is a component of the syndrome.

GS was mostly reported in stroke patients. However, it was also diagnosed in other medical conditions such as brain tumor with epilepsy, multiple sclerosis or mucormycosis.

Shimotake et al. [2008] reported a case of a 34-year-old man diagnosed with astrocytoma located in the left brain hemisphere, mainly in the white matter of the parietal lobe which also involved the AG. In addition, symptomatic epilepsy was diagnosed, causally related to the presence of the tumor. Scalp EEGs showed epilepsy discharges in the left parietal region. The patient presented with transient ictal symptoms of GS, apart from left-right disorientation, which was not observed. The remaining symptoms of this syndrome appeared episodically and were closely related to the occurrence of an epileptic seizure.

Tumefactive multiple sclerosis is another neurological disease that may reveal symptoms of this syndrome. Gnanapavan et al. [2014] reported a case of a 30-year-old right-handed female patient diagnosed with this rare manifestation of multiple sclerosis based on brain biopsy and MRI. MRI showed several focal cystic changes, including the left occipito-parieto-temporal region with the AG.

The patient was diagnosed with the tetrad of GS with subtle symptoms of receptive and expressive dysphasia, and right-sided homonymous hemianopia.

One of the recent reports is a case of a female patient diagnosed with GS in the course of a fungal infection of the central nervous system i.e. aggressive mucormycosis [Stretz et al. 2017].

Zukic et al. [2012] investigated the prevalence of GS within the first week after the occurrence of stroke in acute stroke patients. In a group of 194 patients, a pure tetrad of this syndrome was found in two patients (3.4%) who were right-handed. Head CT revealed the evidence of ischemic lesion in the left parietal or temporoparietal lobe. The findings of that study were consistent with the opinion that the syndrome in a pure and full form remains a unique clinical entity.

Ardila [2014a, 2014b] proposed the reinterpretation of the syndrome ninety years after the first paper of Gerstmann on the syndrome he had isolated. Considering the frequently reported coexistence of aphasia with the symptoms of this syndrome, Ardila proposed a reinterpretation of the syndrome which was based on replacing agraphia with semantic aphasia, with the preservation of the other elements i.e., finger agnosia, acalculia, and left-right disorientation. Ardila excluded agraphia as a deficit usually absent in the incomplete syndrome, which if observed, corresponds to apraxic agraphia, resulting from the damage to the superior parietal lobule rather than to the AG. Ardila also proposed the inclusion of aphasia (fifth component) to the tetrad of the GS. Ardila [2010, 2014a] indicated semantic aphasia which he classified as the one corresponding to transcortical sensory aphasia type II, and post-Rolandic and extra-Sylvian aphasia, according to the neuroanatomical criterion. Ardila et al. [1989] reported that the term *semantic aphasia* was introduced by Henry Head in 1920 to describe the inability to recognize simultaneously the elements of the sentence. Forty years later Alexander R. Luria conducted an in-depth analysis of this type of aphasia. Luria paid attention to the fact that the use of language is related to the positioning of a given word in a system of simultaneous correlations with other words. Therefore, the meaning goes beyond separate words and it is also defined by the relationships among words. Luria considered semantic aphasia to be a deficit of understanding words containing the spatial meaning, i.e. the disorder of spatial operations on the linguistic level. As the manifestations of these dysfunctions in semantic aphasia, Luria mentioned impaired understanding of linguistic structures consisting of successive subordinate clauses (especially with the words *which*, *that, in spite of, as a result of*), structures of the temporal or spatial type (with a preposition, e.g. 'I will be running before lunch, the umbrella is under the chair'), comparative sentences (e.g. 'the elephant is bigger than the ant'), constructions with attributive relations (with the attribute e.g. 'the father's brother, the brother's father'), passive structures ('the earth is illuminated by the sun'), constructions with a double negative (e.g. 'I am not used to not obeying the norms'). These are complex linguistic structures that express logico-grammatical relationships. In other words, patients understand individual words, however, understanding the internal relationships between them is problematic –

patients cannot integrate the elements of a sentence into a whole. Moreover, Ardila [2014b], partly referring to Mayer et al. [1999], indicated an impairment in verbally mediated spatial operations as a fundamental deficit combining the symptoms of the revised Gerstmann model.

In order to help the reader understand these views, we will remind the differences in the functioning of the right and left hemispheres of the brain. The development of the logical and spatial coherence of the self system is conditioned by the proper functioning of the entire brain. This is ensured not only by properly functioning structures, but also by connections within each hemisphere, between both hemispheres and their connections with subcortical structures [Pąchalska 2019]. Differences in the functioning of the left and right hemispheres of the brain within the self system are illustrated in Fig. 1.

It can be seen that the dominant hemisphere of the brain (usually the left in right-handed persons) is closely related to language functions. Therefore, it provides logical coherence possible thanks to linguistic images, which includes language models, grammar and vocabulary, as well as internal narration and dialogue. An important role is also played by the ability of linguistic expression, which is enabled by efficiently functioning articulatory organs and limbs (writing and signalling language statements). Based on this, language texts are created, among which a special role is played by narrative and external dialogue that enables contact with other people. Patterns of neural network connections that evoke thoughts (and thus behaviors) that promote the well-being of the body are permanently encoded, while useless ones disappear [Pąchalska, Kaczmarek, Kropotov 2014].

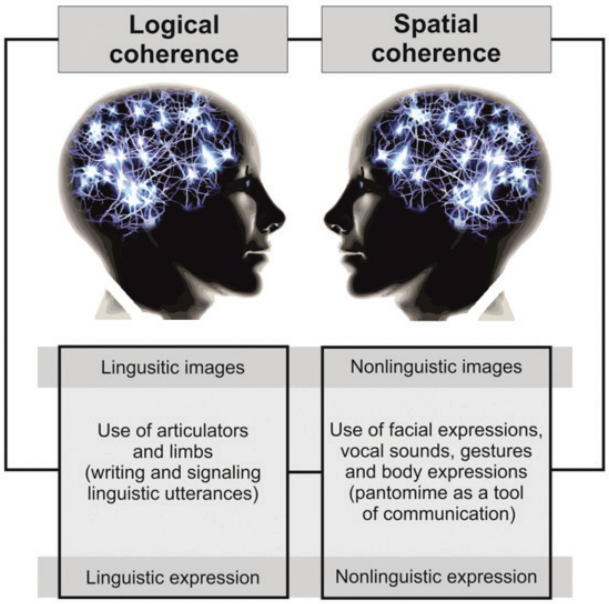

Fig. 1. Differences in the functioning of the left and right hemispheres of the brain
Source: M. Pąchalska 2019

The subdominant hemisphere of the brain is closely related to nonlinguistic functions (generally the right in right-handed persons). Thus, it provides spatial coherence based on nonlinguistic images: image models and "body grammar", i.e., images evoked by facial expressions, gestures and a sequence of movements (pantomime). This enables, through the use of the facial expressions, phonic organs (vocalization), limbs (gestures) and the whole body (pantomime, "body language") nonlinguistic expression. This creates nonlinguistic messages: acoustic (voice, sound) and visual (drawing, gesture) [Pachalska 2019].

Persons with brain damage exhibit disturbances in logical or spatial coherence depending on the location of the damage (structures and neural connections) in the left or right hemisphere of the brain. Linguistic representations are more or less disintegrated, which makes creating language constructions more difficult, as a result of which the process of creating ideas about yourself and the world is disturbed, which is why the image of oneself and, as a result, the whole system of the self is disintegrated [Pąchalska, Kaczmarek, Kropotov 2014].

As observed by Roux et al. [2003], Gerstmann probably did not predict that he had reported one of the most controversial issues of neuropsychology while describing the tetrad of the coexisting symptoms in the 1920's and 1930's. GS has intrigued neurologists and neuropsychologists from the very beginning and has been discussed until now. Critchley [1966] defined it as the enigma that is still a subject of scientific research. At present, the existence of GS is no longer questioned, however, the occurrence of the pure tetrad of this syndrome is rare. The most common opinion, though not conclusive, is that GS results from the structural cortical damage in the region of the left parietal lobe in the AG. It can be the manifestation of subcortical damage i.e. damage to the white matter below the AG or the posterior part of the left thalamus with a secondary impairment of ipsilateral functioning of the AG.

Doubts arise from the combination of symptoms of the classical model. The proposed modification of the model, including aphasia, may affect the change of the opinion on the prevalence of this syndrome. The undisputed scientific achievements of Gerstmann became the inspiration for further research on parietal lobes of the brain, including the AG as an area with a variety of functions and the multisensory center, which integrates information from various sensory modalities [Pąchalska 1999; Seghier 2013].

It should be add, that Gerstmann's scientific achievements have also influenced the binding of the microgenetic theory of symptom formation, which has been confirmed in many scientific studies [Brown 2002, 2015; Pąchalska 2002, 2019; Gazzaniga 2011; Kropotov 2016]. Particularly important for symptom formation following brain damage is modern knowledge about the relationship between consciousness and mental state formation [Pąchalska 2019]. The mental state makes possible to understand the phenomenon of developing (T1) and renewing (T2) this state in time (cf. Fig. 2) in norm and in pathology and the birth of the minimal working self [Pąchalska, Kaczmarek, Kropotov 2014].

In working memory, images are reproduced in subsequent mental states in the order of memory, i.e., in relation to their resemblance to the coming state,

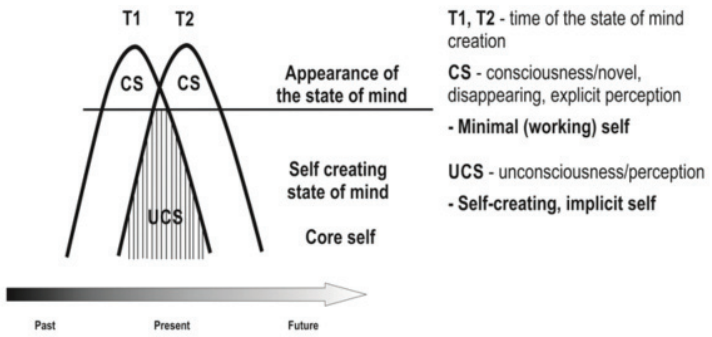

Fig. 2. Developing (T1) and renewing (T2) mental state in time: the birth of the minimal (working) self
Source: Pąchalska 2019

and thus to the possibility of renewing the mental state. In the current state of mind, there are images closer to the perception that takes place, i.e., images from the working memory buffer that have almost reached the character of renewed perception. The brain-mind state in T1 is replaced by the overlapping state T2 before T1 ends in time, i.e., before the next phase occurs. This explains the reoccurrence of the early phases in T1, related to the condition of the body (body and brain), individuality of the person, i.e., Self, character, disposition, capacity of working memory buffers, long-term memory resources and experience, and the durability of basic beliefs, values and personality. Later phases disappear when the whole process of realizing reality is completed to make room for new perceptions. The activity of earlier phases of the mental state in the process of the overlapping of individual phases explains the sense of self continuity in time. It should be emphasized that the early stages of mental state development are components that incorporate later states that are more susceptible to environmental influences. At the same time, the repetition of earlier phases is closely connected with the feeling of a reality that exists [Pąchalska, MacQueen, Brown 2012].

This means that in the process of creating consciousness, one state of mind is replaced by another in a split second, which makes the apparent change replace the previous states of mind by successive states. This overlap of individual states creates a sense of continuity, while their mutual substitution creates a sense of change. It is worth emphasizing that the process of becoming aware of reality may vary depending on the needs, attitude, emotional state and cognitive processes of a person [Kolańczyk 1999], as well as the criterion features of objects with which a given person interacts, and environmental conditions [Pąchalska, Kaczmarek, Kropotov 2014].

Mental states do not constitute a cumulative whole created as a result of separate processes occurring on three levels of microgeny (drives and needs, emotional and cognitive processes), but recreate the course of object (perception) formation in the mind [cf. Pąchalska, Kaczmarek, Kropotov 2014]. And it is the process of creating an object representation that organizes the process of symp-

tom formation in microgenesis. Damage of different brain areas and the disruption of various patterns of neuronal connections underlie the process of formation of certain mental states, which in turn causes the formation of individual disease syndromes, which also includes GS [Pąchalska 2019]. Modern diagnostics based on neuromarkers are also developing, which can confirm the presence of individual syndromes with 90% accuracy, including those differentiating particular types of aphasia [Kropotov 2016; Chantsoulis, Półrola, Góral-Półrola et al. 2017].

It should be emphasized that persons with brain damage (especially to the right hemisphere) may either be underdeveloped, destabilized or have lost one or more components of language, which is evident even in semantic dementia (SD) [Mendez, Ramírez-Bermúdez 2011; Ardila 2014]. The conducted neurophysiological and neuropsychological research of such patients, especially in the later period of the disease [Pąchalska, Kaczmarek, Kropotov 2020], shows impairment of executive components. Besides an inability to recall from the episodic memory, the subject suffers from executive dysfunction. All executive components are impaired (Fig. 3).

Processing in the right inferior temporal cortex was significantly delayed. Processing in the parietal cortex was significantly faster than in norms. This imbalance in timing of the processing in the dorsal and right ventral visual streams created an inability to correctly extract information from the long term episodic memory. Words lose their meaning and SD increases, and self-awareness and meta-consciousness disorders appear. The patient lives in an unreal world and can develop delusional misidentification syndromes (DMS), with the most common being Cotard syndrome, as other authors have also observed [Mendez, Ramírez-Bermúdez 2011]. A closer analysis of the disorders that occur in people with SD, as the disease progresses, shows that as the neurodegenerative disease is progressing over time, both the individual, as well as social and cultural self may disintegrate.

The studies presented here, as well as other neurophysiological studies of people with semantic aphasia, make us aware that neuropsychological symptoms formed as a result of brain damage can be variable [Pąchalska 2019]. This is due to the fact that language is a highly complex brain function, and each person has their own vocabulary, fluency in language use, and their own specific way of selecting words in conversation.

It should be add, that language may be responsible for the unique character of the human mind but, except for verbal thought, many attributes of mind persist when language is lost. More precisely, the actualization of each component of mentality suffices to sustain human psyche, even when there is disruption within a given component. In animals lacking the specialization of the human brain, this commonality of pattern – category/item transition – survives multiple ablations. This has suggested mass action or equipotentiality [Lashley 1951], holographic organization [Pribram 1984] or even the function of the brain and mind not only in space and time but also in a pulsating state in hyperspace, which is presented in the authorial synchronous memory model (cf. Fig. 4).

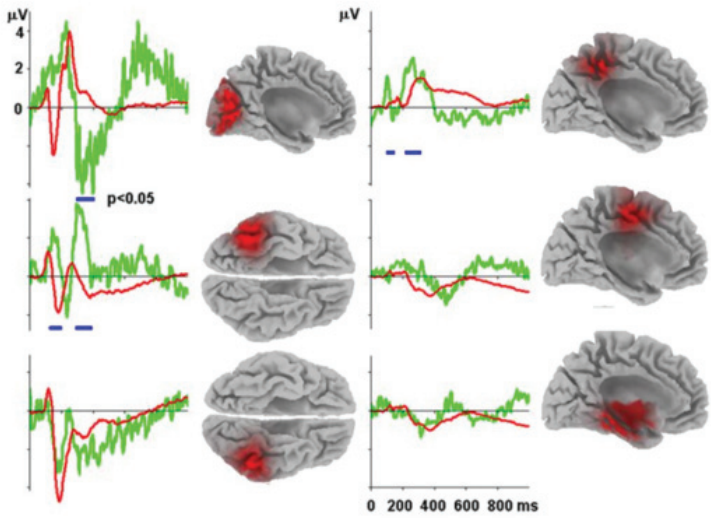

Fig. 3. Impairment of independent components in the patient: all executive components are impaired
Source: Pąchalska, Kaczmarek & Kropotov 2020

The spatial arrangement of the model makes it possible to present on the x and y axes the relationship between the general structure of attention and memory systems (in terms of the number, content and complexity of processed elements) and the period of time necessary to process them. It can be seen that the Attention System buffers data transfers to the working memory system. This system, according to the latest data obtained in neurophysiological studies, processes the smallest number of elements in the shortest possible time: seconds or even milliseconds [Kropotov 2016; Pąchalska, Kaczmarek, Kropotov 2020]. As the number of elements of information processed and / or the duration of the processing exceeds a certain threshold, we gradually move from the attention system (several stimuli, several milliseconds) to the working memory system (several to several dozen stimuli, several milliseconds to several seconds and / or minutes) depending on the capacity of the working memory buffer [see also Kropotov 2016].

In a similar way, there is a transition from the working memory system to the long-term memory system. The boundary of the transition is difficult to determine precisely and most likely it is actually not very sharp. In the human brain, a continuous process takes place, lasting from milliseconds to entire years when information is remembered, stored, reproduced and forgotten. Also semantic and episodic memory is associated with the number, time as well as the content and complexity of processed data [Pąchalska 2019]. The differences between these types of memory mainly concern the content of information. Of course, the longest storage time is characteristic of long-term memory, which is why we put it at the basis of the presented model.

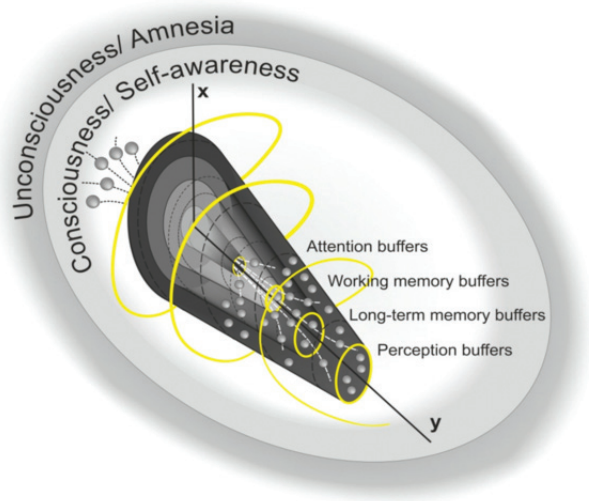

Fig. 4. Synchronous memory model
Source: Pąchalska, Kaczmarek & Kropotov 2014, modified

Brain damage, especially the parietal areas forming neural connections associated with different modalities, destroys slightly different patterns of these connections in each patient, therefore the picture of disorders is variable. This explains the difficulties in describing GS that scholars in the world are facing.

This, in itself, is sufficient to acknowledge the contribution of Professor Josef Gerstmann to the development of neuropsychology and neurology.

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