

UNILATERAL NEGLECT SYNDROME

CLINICAL AND TOPOGRAPHIC STUDY OF 20 SUBJECTS

LUCIANO RIBEIRO PINTO JR.* — SYLVIO SARAIVA **
WILSON LUIZ SANVITO ***

SUMMARY — Twenty patients with unilateral neglect syndrome were studied. They were 10 males and 10 females, and they ranged from 29 to 76 years of age. All were submitted to a CAT scan of the brain. Based on the findings in our sample we drew the following conclusions: the extinction phenomenon was a constant manifestation of unilateral neglect; the line crossing test proved to be most efficient for the identification of visual neglect; the right parietal lobe was the anatomical region most often involved in the unilateral neglect syndrome.

Síndrome da negligência unilateral: estudo clínico e topográfico de 20 casos.

RESUMO — Foram estudados 20 pacientes com a síndrome da negligência unilateral (SNU), de idades variáveis entre 29 e 76 anos, sendo 10 do sexo masculino e 10 do sexo feminino, todos submetidos à tomografia axial computadorizada do crânio. Baseados nesta casuística, concluímos: o fenômeno de extinção foi manifestação constante da SNU; a prova da secção de traços foi a que se revelou mais eficaz na identificação da negligência visual; o lobo parietal direito foi a região anatômica mais freqüentemente comprometida na SNU.

The right hemisphere takes part especially in the function of higher level nervous activities related to the visual-spatial recognition and body image. The disturbance of these functions are at the center of the unilateral neglect syndrome (UNS). Because of the controversial aspects of the UNS we decided to do a prospective study aimed at characterizing the syndrome and establishing the location of the lesions. With this purpose in mind we studied 20 UNS patients.

The first report in the literature to come closes to what is presently called neglect syndrome is the one of Jackson³⁶ in 1876, when the author described a case of visual imperceptiveness in a patient who had a right temporal lobe glioma. The first reference to the extinction phenomenon was made by Bender & Furlow⁸ in 1945; they described a patient who would not identify one of the images when given a double visual stimulus. The same phenomenon may occur after the general sensitivity is stimulated. The most recent systematic studies that brought new contributions to the clinical and physiopathological aspects of the UNS are the ones from Heilman's and Watson's school^{30-34,57,58}. Beginning in the 1970's there have been some studies linking the physiopathology of UNS to biochemical mechanisms, especially the dopaminergic circuits^{24,40-42,55,56}. Unilateral lesions of the dopaminergic paths may cause the neglect to manifest itself in the form of akinesia^{24,40,41}. The UNS is characterized by the presence of the extinction phenomenon, spatial neglect, somatic neglect, and unilateral akinesia. It can present itself in a complete or partial form, and it may be accompanied by other neurological manifestations^{5,6,8,13,28}. Generically, the extinc-

Disciplina de Neurologia da Faculdade de Ciências Médicas da Santa Casa de São Paulo: * Professor Assistente; *** Professor Pleno. Departamento de Neurologia da Faculdade de Medicina da Universidade de São Paulo; ** Professor Livre-Docente.

Dr. Luciano Ribeiro Pinto Jr. — Serviço de Neurologia, Departamento de Medicina, Santa Casa de São Paulo - Rua Cesário Mota Jr. 112 - 01221 São Paulo SP - Brasil

tion phenomenon consists of the lack of perception of one sensitive or sensory stimulus when two stimuli are provided simultaneously. The perception of each stimulus separately must take place as a condition to establish the existence of extinction. In the spatial neglect the patient ignores the existence of half of the spatial field, on the opposite side of the lesion. This deficiency of recognition may reach the visual, auditory, and tactile spheres. In the somatic neglect the patient ignores the existence of half of his body. This phenomenon is also called hemiasomatognosia^{2,13,25,39} and depersonalization. It can be observed in certain patients' actions such as shaving, putting on glasses, putting on shoes or gloves, when he acts only in relation to the non-affected half of his body. The anosognosia, the motor extinction, the motor and sensitive aloesthesia may also be manifestations of somatic neglect. Unilateral akinesia is the inability or delay presented by the patient upon starting or trying to start a movement directed to a specific half of the space, and which cannot be due to a lesion of the peripheral motor neuron or of the pyramidal tract^{32,55}. It is also called intentional neglect³².

SUBJECTS AND METHOD

Our subjects were 20 patients examined either in the Neurology Infirmary of the Department of Medicine of the Santa Casa of São Paulo or in our private practice office.

After the exam of the higher nervous activity we proceeded to the evaluation of the UNS components. The evaluation steps were the followings: 1. Extinction phenomenon — Utilization of the tactile, painful, visual, and auditory double stimuli on the limbs and faces, bilaterally, simultaneously and symmetrically. 2. Spatial neglect — 2.1. Direct observation of patient or report from patient's family. 2.2. Patient's orientation evaluated through his attitude and walking. 2.3 Patient's reading of newspapers, magazines, and sentences. 2.4. Patient's identification of a two-digit number. 2.5. Patient's drawings. 2.6. Line and number crossing. Line crossing: the patient was presented with a piece of paper with 50 vertical lines drawn, and was asked to cross them. Number crossing: the patient was presented with a piece of paper with several numbers written at random on it, and was asked to cross a specific digit. 2.7. Matchsticks retrieval: the patient was presented with matchsticks placed vertically and horizontally on a table, and was asked to retrieve the ones in a certain position only. 2.8 Exam of visual and auditory aloesthesia. 3. Somatic neglect — 3.1. Direct observation of patient or report from patient's family. 3.2. Test for sensitive and motor aloesthesia. 3.3. Test for the presence of anosognosia in cases with hemiplegia. 4. Unilateral akinesia: evaluation through the observation of voluntary movements of each limb separately, or during the simultaneous extension of the upper limbs.

All patients were submitted to a CAT scan of the brain.

RESULTS

All patients had left hemisphere dominance. The extinction phenomenon was observed in all patients: painful extinction in 15 cases, tactile extinction in 12 cases, visual extinction in 11 cases, and auditory extinction in 8 cases. Spatial neglect was present in 16 cases, and somatic neglect in 5 cases (Table 1).

The analysis of specific tests for the study of visual neglect showed abnormalities in 15 patients (Table 2): it became evident in the line crossing in 9 cases (Fig. 1), in the number crossing in 7 cases (Fig. 1), and in the drawings in 5 cases (Fig. 1).

UNS components	Cases	
	Number	%
Extinction phenomenon	20	100
Spatial neglect	16	80
Somatic neglect	5	25

Table 1 — Incidence of unilateral neglect syndrome (UNS) components in 20 patients.

Visual neglect test	Cases		% in 20 cases
	Number	%	
Line crossing	9	60.0	45
Number crossing	7	46.7	35
Drawing	5	33.3	25
Orientation	4	26.7	20
Reading	2	13.3	10
Two-digit identification	2	13.3	10
Matchstick retrieval	1	6.7	5

Table 2 — Incidence of visual-spatial alterations on specific tests for the study of visual neglect in 15 UNS patients.

Location of lesions	Cases	
	Number	%
Right hemisphere	9	45
Right and left hemisphere	4	20
Left hemisphere	2	10
Right thalamus	2	10
Left thalamus	1	5
Right hemisphere and right thalamus	1	5

Table 3 — Location of abnormalities in the CAT scan of the brain of 20 unilateral neglect syndrome patients.

Location of lesions	Cases		% in 20 cases
	Number	%	
Parietal	4	40	20
Parietal and occipital	2	20	10
Parietal, temporal and frontal	1	10	5
Parietal, temporal, frontal, thalamus	1	10	5
Occipital	1	10	5
Temporal	1	10	5

Table 4 — Location of abnormalities in the CAT scan of the brain of 10 patients with right hemisphere involvement.

In the CAT scan of the brain, localized abnormalities of the parietal lobe were observed in 13 cases, and of these 11 were on the right parietal lobe and 2 on the left parietal lobe (Tables 3, 4 and 5). In regard to the nature of the lesions, the tomographic image was one of ischemia in 11 cases, of hemorrhage in 5 cases, and of neoplasm in 3 cases. In one case there was no evidence of localized lesions in the parenchyma.

Location of lesion	Cases Number	% in 20 cases
Parietal, occipital, and temporal	1	5
Parietal, temporal, and frontal	1	5

Table 5 — Location of abnormalities in the CAT scan of the brain of two patients with left hemisphere involvement.

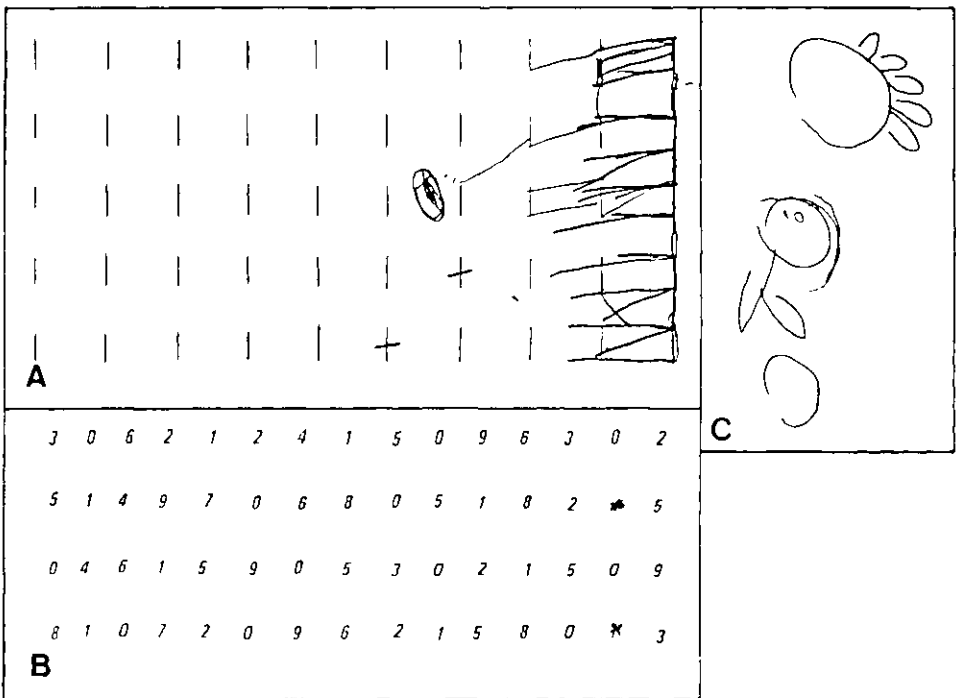


Fig. 1 — Case 12: left visual-spatial neglect observed in the line crossing test (A) and (B) in the specific digit crossing test (number 1). Case 6: left visual-spatial neglect observed in the drawing of a daisy and a doll (C).

COMMENTS

The studies found in the literature generally do not analyse the UNS in its totality. The majority of these studies emphasize the visual-spatial aspects 1,10-12,15,20, 23,27,33,38,44,46,47,50. The extinction phenomenon, and the lateralized alterations of the body image as well, are generally analysed in an isolated fashion 5,7-9,13,16,22,25,28,39, 45,51-53. The finding of the extinction phenomenon in all of our 20 patients supports the studies that consider this phenomenon to be a manifestation of the UNS 32,59. The painful extinction was the one with the highest frequency, a finding which agrees with the thinking of Critchley 16.

In terms of frequency, the visual neglect was the second manifestation found in our patients. The tests of line and number crossing were the ones that demonstrated visual-spatial neglect best. The identification of a two-digit number proved not to be so sensitive a test; however, it has the advantages of being a test of quick execution, and of establishing the differential diagnosis between visual neglect and hemianopia. In its execution it is unlikely that each digit of a pair will be in different visual fields. Furthermore, the spatial neglect results from the relative position of the objects in space, and not from their location in a specific half of the visual field³⁷.

In the UNS the lesion is often located on the right cerebral hemisphere^{3,4,14,18,21,27,44}. In our patients there was a predominance of right hemisphere involvement. Our results also agree with the findings in the literature regarding a higher parietal lobe involvement^{11,16,17,19,23,29,35,43,45,47-49,51,52,54}. The higher incidence of right hemisphere lesions is interpreted as being due to the specialization of the two hemispheres, for the right hemisphere takes part especially in the tridimensional visual mechanisms, in the construction of space^{26,27}, and in the body image^{4,28,51}.

In the genesis of the UNS, the predominance of lesions on the right cerebral hemisphere can also be attributed to other factors. The cortical-limbic-reticular circuit is less organized in the right hemisphere; thus, right hemisphere lesions produce a more pronounced asymmetry in response to orientation than left hemisphere lesions³⁴. The influence of language was the subject of a few studies^{14,54}. The process of verbalization, upon activating the left hemisphere, can increase a neglect that already exists due to right hemisphere lesion.

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