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MOTOR HEMIPLEGIA AND THE CEREBRAL ORGANIZATION OF MOVEMENT IN MAN

I. THE «PYRAMIDAL CONCEPT»: A RESTATEMENT

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SUMMARY — Based on twenty personal cases of pure motor symptoms due to circumscribed encephalic injuries, the pertinent literature is reviewed and the structural and functional supremacy of the pyramidal (corticospinal) and corticonuclear tracts in the human being is emphasized.

Hemiplegia motora e organização cerebral do movimento no homem: I. O «conceito piramidal»: um reposicionamento.

RESUMO — Com base em vinte casos pessoais de sintomas motores puros decorrentes de lesões encefálicas circunscritas, a literatura pertinente é sumariamente revista e a supremacia estrutural e funcional dos feixes piramidais (corticoespinhais) e corticonucleares no ser humano é enfatizada.

In 1984, the author evaluated his initial cases of the locked-in syndrome (LS) in clinical practice. Two of them were reported with the collaboration of Esbérard [110]. During follow-up examinations, it was striking to see the amount of disability those ventrally seated brainstem lesions had inflicted, which seemed even more impressive considering the deficits observed in monkeys bearing similar experimentally placed lesions (see references below). This contrast invited the search for new avenues of working out the subtle ways employed by the brain in order to provide the appropriate means of organizing the multiple aspects of motor control *in man*. In 1984, it was suggested that unilateral pure motor hemiplegia (PMH) and LS were essentially the same syndrome, since both were the consequence of pyramiral tract lesions either unilateral or bilateral. Motor hemiplegic syndromes are precious tools for understanding relevant issues concerning human cerebral motor organization. A systematic investigation of pyramidal tract affection in patients was undertaken. Now, after four years of study of old and new PMH and LS cases, the impression has been gained that more than mere semeiologic esthetics is hidden behind the stereotyped clinical pictures of both syndromes and the capriciousness of the lesions that determine their appearence. Slowly, the conviction has mounted that these two syndromes convey pathophysiologic meaning which should be unravelled if the operational and conceptual knowledge of brain motor mechanisms in health or disease states are to be deepened. It is precisely this pathophysiologic dimension of motor hemiplegias that will be presented and discussed.

Unless otherwise specified, the motor deficit in hemiplegia will be assumed as qualitatively similar to the one observed to occur in hemiparesis, implying identical anatomic correlates for both. Further, the expression pyramidal tract (PT) will be interchangeably

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used with corticospinal tract (CST). It will be also assumed that the corticonuclear tracts are functionally homologous with the CST and both will be implicitly treated as only one.

For the sake of editorial space saving, a complete list of references is appearing at the end of the second and final part of this article.

PATIENTS AND METHODS

The application of CNS imaging (*76*) and physiologic methods were used in selected cases to investigate the system of motor organization in man. Twenty cases of pure motor symptoms produced by cerebral or brainstem lesions as documented by computorized tomography (CT) or nuclear magnetic ressonance (NMR) studies were selected from a larger group of hemiplegics with associated non-motor findings. The group included 8 men and 12 women and the lesions that produced their neurologic pictures were located at cortico--subcortical, subcortical-supracapsular, striato-thalamo-capsular and predominantly ventral brainstem sites. Their ages ranged from 9 to 79 years and most of them had associated medical conditions (Table I). All underwent a standard neurological examination by the author. Patients with persistent signs of disturbed menta functioning, pathologic somnolence or meningeal inflammation were excluded. Some factors were specifically evaluated:

1. Motor deficits were graded according to the ability to perform simple motor acts on command as: a. hemiplegia — none or only feeble movements of the affected parts were possible; b. hemiparesis — sustained movements possible, but less effectively than the non--affected side, best disclosed by the deficitary manoeuvers of Barré and Mingazzini. 2. Spasticity was evaluated according to the presence of the clasp-knife «catching» resistance felt during the elicitation of passive joint movements, and graded as: +, clasp-knife best perceived by the end of movement; ++, clasp-knife already perceived from the start of movement; +++, clasp-knife superimposed on a sustained attitudinal background (Wernicke-Mann attitude), with or without definitive muscular contracture. 3. Muscular reflexes («tendon jerks»), comparatively assessed in relation to the unaffected side, which was taken as «normal» for each patient and given values from 3+ to 4+/6+. Thus, 1+ to 2+ meant

Case	Sex	Age (in 1984)	General Clinical Findings
1. MCP	F	58	moderate AH
2. FLS	M	25	Marfan disease; subacute S. viridans endocarditis
3. AMC	F	9	—
4. PFL	M	56	moderate AH
5. DAA	M	34	craniocerebral trauma
6. MPR	F	50	moderate AH
7. TG	M	47	moderate AH
8. AGS	M	70	moderate AH; pulmonary emphysema
9. LMT	F	23	—
10. ALM	M	63	moderate AH
11. MPG	F	55	moderate AH
12. RR	F	69	moderate AH; diabetes mellitus
13. LC	F	59	moderate AH; diabetes mellitus
14. JJS	M	79	moderate AH; diabetes mellitus
15. AAM	M	64	AH
16. BFA	F	70	AH; diabetes mellitus; coronary disease
17. EPG	F	53	AH
18. MLS	F	34	AH
19. PM	F	76	AH
20. HCP	F	68	AH; bilateral carotid disease; hypertensive cardiomyopathy

Table I — General characteristics of ***patient sample.*** *AH, arterial hypertension.*

T A B L E II

NEUROLOGICAL FINDINGS, LESION SITES AND MAJOR VASCULAR TERRITORIES INVOLVED IN INDIVIDUAL PATIENTS

CASE	MOTOR DEFICIT	SPASTICITY	MUSCULAR REFLEXES	PLANTAR RESPONSE*	CT SCAN **	OTHER FINDINGS***	COURSE	MAJOR ARTERIAL**** TERRITORIES COMPROMISED
1	R hemiplegia sparing the face	***	6+	R_{ex} - L_{fl}	L pallido-capsular infarct	normal SEP	stable (3 yrs)	ACHA
2	L hemiplegia	***	5+	R_{fl} - L_{ex}	R high capsular corona radiata infarct	normal SEP	stable (1 yr)	broderzone of insular (cortical) and LS branches of MCA
3	R hemiparesis with facial involvement	-	5+	R_{ex} - L_{fl}	L high striato-capsular infarct	normal SEPs hypoplasic L int car art + supraclinoid oclusion	stable (5 yrs)	LS branch of MCA
4	R hemiparesis	***	5+	R_{ex} - L_{fl}	L high capsular corona-radiata infarct	-	stable (4 yrs)	LS branch of MCA
5	R Hemiparesis	-	4+	R_{fl} - L_{fl}	small L putaminal hemorrhage	transient mental	complete remission (1 mo)	LS branch of MCA
6	R hemiplegia	***	5+	R_{ex} - L_{fl}	L high capsular infarct	-	stable (4 yrs)	borderzone of cortical and LS branches of MCA
7	R hemiparesis	***	6+	R_{eq} - L_{fl}	L cortico-subcortical precentral infarct + L posterior medial frontal lobe infarct	-	remission to spastic crural monoparesis	rolandic branch of superior division of MCA + collosomarginal
8	L hemiplegia	***	6+	R_{fl} - L_{ex}	R cortico-subcortical posterior frontal lobe infarct	normal SEP	stable (1 yr)	rolandic (cortical) branch of MCA
9	L hemiplegia	***	6+	R_{fl} - L_{ex}	R medial cortico-subcortical	CSF: 50 mononuclear cells + hypergammaglobulinorhachia	remission to spastic crural monoparesis	ACA sparing Heubner's recurrent artery territory
10	R hemiparesis with crural predominance	-	5+	R_{ex} - L_{fl}	L caudate-capsular infarct	-	stable (3 yrs)	Heubner's recurrent artery (ACA)

CASE	MOTOR DEFICIT	SPASTICITY	MUSCULAR REFLEXES	PLANTAR RESPONSE*	CT SCAN **	OTHER FINDINGS***	COURSE	MAJOR ARTERIAL **** TERRITORIES COMPROMISED
11	R hemiparesis	***	6+	R_{ex} - L_{ex}	L ventral pontine hypodensities	normal SEP and BAER L laryngeal paresis	stable(8 mos)	paramedian branch of BA? VA? AICA?
12	mutism + tetraplegia + dysphagia facial diplegia	***	6+	R_{ex} - L_{ex}	post-ischemic ventral pontine atrophy with penducular extension	communicable able to open eyelids and abduct left eyeball normal SEPs and BAER central conductions	stable(4 yrs)	paramedian branches of BA? VAs?
13	mutism + tetraplegia + dysphagia + facial diplegia	***	6+	R_{ex} - L_{fl}	CT not done;presumed ventral pontine infarction at trigeminal level	communicable eye movements full	death in 30 days	paramedian branches of BA? VAs?
14	mutism + tetraplegia + dysphagia + facial diplegia	-	3+	R_{ex} - L_{fl}	ventral pontine infarction	communicable able to open eyelids	death in 15 days	paramedian branches of BA? VAs?
15	amimia,dysphagia,dysarthria (pseudobulbar triad)	-	3+	R_{fl} - L_{fl}	small, symmetric subinsular infarcts	-	partial remission(4 mo)	insular(cortical)branches of MCA
16	R hemiplegia	***	6+	R_{ex} - L_{fl}	L hemisphere infarct	normal SEP	stable (3 mo)	cortical and deep MCA territory
17	R lower facial paresis, dysarthria,dysphagia	-	3+	R_{fl} - L_{fl}	L high capsular-corona radiata infarct	-	remission(24)	borderzone of cortical and LS branches of MCA
18	L hemiplegia	**	5+	R_{fl} - L_{ex}	R putaminal hemorrhage	-	remission(2 mo)	LS branch of MCA
19	R hemiparesis	-	4+	R_{ex} - L_{fl}	capsular hemorrhage (genu)	-	remission(3 mo)	LS branch of MCA or Heubers artery
20	R hemiplegia	***	6+	R_{ex} - L_{fl}	high capsular-corona radiata infarct	normal SEPs	stable (1 yr)	borderzone of cortical and LS branches of MCA

ex - extensor
fl - flexor
eq - equivocal

** Lesion extension was also verified by MR in case 9: CT was normal 48 hours after onset of hemiplegia.NMR made 30 days later.

*** SEP-somato-sensitive evoked potentials.
BAER-brainstem auditory evoked responses.

**** AChA-anterior choroidal artery
ACA -anterior cerebral artery
MCA -middle cerebral artery
LS -lenticulostriate
VA -vertebral artery
BA -basilar artery
AICA-anterior inferior cerebellar artery

hypoactive reflexes and 5+ meant hyperactive reflexes; 6+ meant, in addition, the presence of clonus, Hoffmann sign and reflex irradiation. 4. Babinski sign was defined by an upgoing great toe (*38*) in response to lateral plantar or equivalent (Chaddock, Gordon, Schaefer and Oppenheim) deep cutaneous stimulation, regardless the other toes behavior.

Coordination was assessed by finger-to-nose/heel-to-knee and diadokokynesia tests. Vision and hearing were tested for gross defects. Sensory examinations were performed for light touch with a wisp of cotton; pain with a safety pin bent at right angles; temperature, with test tubes containing cool and warm water; position sense, by passively moving the joints, of toes and fingers in particular, and sequentially asking for the patient's own appreciation of his or her segments positions; vibration sense, by placing a 128 Hz (C) oscillating tuning fork, with weighted ends, over the bony prominences of face (front-malar-chin), shoulder-elbow-fingers and hip-knee-ankles-toes. Patients were asked to keep their eyes closed and explicitly inquired for feelings of paresthesias and dysesthesias.

Each patient had at least one CT scan of the head, except for patient 13, who died before it could be done. All were scanned by a Siemens-Somatom DR2 machine, matrix 256x256, with slices 8 (and eventually 4) mm thick, parallel to the orbito-meatal line. The results were then checked against available correlative anatomical maps (*10,11,28,61*) for verification of lesion location and major arterial territories involved. Whenever feasible, evoked potentials were performed, so as to document the clinical impression that the motor deficit was actually devoid of any somaesthetic subclinical disturbances (*25,92*). Patients 1, 2, 3, 8, 11, 12, 16 and 20 were so examined.

RESULTS

The results presently relevant are shown in table II. Some of these cases have already been reported at length (*111,112*), and the remainder will be dealt with more analytically in a forthcoming paper.

COMMENTS

Pyramidotomy in man vs. pyramidotomy in non-humans — For present purposes, a finding of utmost importance shown in table II is that an identical motor syndrome of contralateral paresis, spasticity with or without contracture, hyperactive tendon jerks and the Babinski sign results from lesions occupying distinct topographies in the cerebrum and brainstem. The resemblance of the full syndromes produced by large hemispheric (case 16), cortico-subcortical (cases 7, 8, 9 and 15), high capsular-corona radiata (cases 4, 6, 17 and 20), striatocapsular (cases 2, 3, 10 and 18), mid-capsular (cases 1, 5, 18 and 19) and ventral brainstem (cases 11, 12, 13 and 14) lesions to each other is compelling evidence that they must be produced by the involvement of a common neural substrate which should be found throughout these distinct but related regions of the neuraxis. It is at this point that human and animal physiology break apart. Substantial misunderstanding has arisen over the last seventy years in large part due to the relative importance different authors have placed upon the structure(s) they believe best suited to fulfill the role of producing the above set of signs. It now seems clear that much confusion would not have happened if animal interspecies differences had previously been considered. In other words, the riddle of motor system organization in man should be primarily evaluated in man himself. It will become clearer that in the nervous system of man the only constant structure which transgresses the brain in the coronal plane, from cortex to medullary pyramids, is the pyramidal pathway. The corollary which follows is that the tetrad of signs described above is a truly human pyramidal tract syndrome. Further, since this syndrome does not change in its essential elements, regardless the level of the neuraxis at which a given lesion is found, any eventual adjacent lesion will certainly have no major importance in the determination of the final motor picture.

The main inference to be drawn from the above considerations is that the syndrome of motor plegias are caused, in a large majority of cases [140], by a lesion of the PTs anywhere along its extent, from cortex to medulla. From the standpoint of pathophysiology, motor plegias of cerebral origin in man give us the clearest proof of the functional supremacy exerted by the PTs in human cerebral motor organization and should be taken as almost ideal experiments probing the effects of PT interruption in man. As presented in the following paragraphs, LS represents, on its turn, the net effect of a bilateral (thus, total) exclusion of the PTs in human behavior.

Residual	Absent
— spasmodic laughing and crying	— standing reflexes
— crude vocalizations and facial grimacing, mainly in response to nociceptive stimulation	— righting reflexes
— yawning	— rhythmic locomotor automatisms
— bruxism	— gestual, pantomimic and skilled movements, including articulatory and manipulative
— chewing and swallowing, in response to food or liquids in mouth	
— sucking and chewing without swallowing, in response to gentle continuous lip or perioral stimulation	
— coughing, in response to direct laryngeal stimulation or to deposition of food or liquids in pharynx	
— decerebration, mostly episodic, spontaneous or in response to nociceptive stimulation	
— autonomous breathing	
— cervical attitudinal reflexes (reciprocal changes in spastic tonus in the four limbs in response to passive head turning)	
— orienting reflexes of head and eyes towards lateralized visual, auditory or tactile stimulli	
— spasticity and hyperactive muscle reflexes, with reflex irradiation and clonus	
— Babinski sign with fanning of toes	
— universal amyotrophy	

Table III — Motor patterns in LS (7, 29, 110).

Absent	Residual
— individual use of fingers for skilled movements	— standing
— spasticity	— locomotor progression
— normal speed of movements	— grasping with hand and fingers
	— attitudinal and righting reflexes
	— hyperactive phasic muscle reflexes
	— hypotonia and hyperextensibility, mainly in hands and feet

Table IV — Motor patterns in the monkey with bilateral pyramidotomy* (37, 63, 82, 148).

* For purpose of clinical interpretations, experimental studies on *apes* would be most desirable, but a perusal of the pertinent literature shows these to be unfortunately rare.

The difference between the human PT syndrome and the one resulting from equivalent lesions inflicted on rats, cats, dogs and even monkeys [37,63,82,148,149,154] is to be stressed. Contrarily to what is shown to be the case in such procedures bilateral injury to the pyramidal system, as found in LS patients, stands as a far more dramatic example of the role played by the PTs in the development of movement in this species. Briefly, in the spastic doubly hemiplegic-mute (anarthric)-dysphagic but communicating LS patient, even righting and postural reflexes, in the sense of Magnus and de Klejn, are lacking and the motor repertoire is drastically restricted to a segmental-plurisegmental pattern of motor integration [7,119]. In table III, the residual motor competencies of three LS patients followed by the author are reported an can be compared to Tower's, Hine's and Lawrence and Kuypers' experimental animals in table IV.

It might perhaps be argued that many elements of the syndrome result, as probably happens in other animals, from "contamination" of the PT affection by lesions of nearby pathways and nuclei. However, there is evidence against this interpretation, the clearest being that, as already stated, an identical motor syndrome undoubtedly occurs in different individuals bearing lesions lodged at a variety of sites occupied by the PTs within the brain, as shown by post-mortem or neuroimaging correlations [16,17,24-27,30-32,41,45-47,53,56,57,60,65,72, 74,84,87,89,93,94,98,108,112,127,129,130,135,139,144 150], anatomic studies on normal brains [131], and direct stimulations during stereotaxic surgical procedures [12,115]. There have been no reported cases of a cortical LS, but this is a plausible possibility, for cases of PMH [156], quadriplegia [85], pseudobulbar palsies [4,20,114] and even monoplegias [9,68,80,159] due to injuries of the proper motor cortical areas have been observed and, as the present article has attempted to demonstrate, *LS is actually a dense, bilateral (double) PMH.* Thus, in a surprisingly ironical way, contemporary clinical neurology seems to be rejoining, in many respects, the main trends of thought developed in the past century by means of the clinicoanatomic method, at least in what concerns the main outcomes of an interruption of the PTs in man. Given the importance of these tracts for the clinician, a brief review of its anatomy is presented at next.

The human pyramidal tract: a selective review with emphasis on some points of clinical relevance — Beginning with the anatomopathologic studies of Türck [151], Charcot [23], Strümpell [143], Babinski [3] and others, the concept of pyramidal tract and pyramidal syndrome had already become established by the dawn of the present century. The condition was characterized by paresis, hyperactive reflexes, spasticity and extensor plantar responses, associated with lesions of the CSTs. This correlation, originally drawn from direct observations made on the brains of amyotrophic lateral sclerosis patients [22,53,77], was subsequently extended to include secondary degenerations of the tracts from an array of other distinct causes [13,127]. The emergence of the pyramidal concept, with its attendant functional and clinical implications, had a strong impact on Neurology some one hundred years ago, promoting deep changes in neurologic thought. There is now ample evidence that the precentral gyrus and the paracentral lobule are the main *source* of the fibers which traverse the bulbar pyramids [8,14, 15,18,23,33,43,47,58,71,77,88,96,97,122,133,144]. Classical writers already knew that restricted lesions of these gyri caused only partial degeneration in corticospinal or corticobulbar fascicles, while total destruction of those areas was accompanied by extensive degeneration of such tracts. Elegant verification of this fact in vivo is now possible by neuroimaging methods [121,142]. It is further recognized that the cells of origin of the PTs lie in area 4 (motor cortex) and 6 (premotor cortex), see references above. Probably a set of fibers also comes from medial area 6 (supplementary motor cortex) [86], but conclusive proof of this in man is still lacking. Claims of PT fibers originating in the parietal lobe have been made, but these are probably scarce and not directly related to the motor syndromes under discussion [78]. The fascicles of the PTs progressively change their *course* along their descending paths [22,64]. The trajectory of foot and leg fibers (see cases 7, 9 and 10), which originate in the paracentral lobule, is the longest of all. In the internal capsule, after traversing the regions of the subcortex (see cases 7, 8, 9, 10, 15 and 16) and corona radiata (see cases 1, 2, 3, 5, 18 and 19), the PT occupies its posterior limb and changes position as it descends, so that it is lodged in the anterior half of the posterior limb in the rostral internal capsule, and in the posterior half of the posterior limb, caudally [131]. At the mesencephalic level, it is seated in the middle three fifths of the peduncle (see case 12), being split in a number of fascicles in the pontine base by the transversely running fibers of the ventral pons (see cases 11, 12, 13 and 14). In the medulla, it emerges again as a compact bundle, adopting the well-known shape of an inverted-based

pyramid. Throughout the descending PT paths along the ventral brainstem, a vast array of fibers leaves them approximately at right angles, heading for local neural structures, in particular the tegmentum and the cranial nerves nuclei [78]. Still according to Kuypers, a special feature of the PT segmental connections in man is the great number of *direct* connections with motoneurons of the brainstem motor nuclei, mainly V, VII, XI and XII, and anterior horns. This *corticomotoneuronal* system is contained in the PTs themselves and made up of axons of the largest diameter coming from the giant Betz cells of Brodmann's area 4. Though it can already be recognized in the monkey, it is not as big as in man and is thus one distinguishing human trait.

Up to this point, all the available evidence shows this design to be fairly symmetrical. From the pyramids down, however, a great deal of interindividual variation occurs regarding the assortment of the whole bulk of the descending fibers in separate tracts inside the cord [75,161]. In "typical" cases, which correspond to more or less 50% of the normal population studied by Yakovlev and Rakic, three main divisions of each PT tract can be identified in each half of the cord. The lateral PT [13,35,138] is crossed and runs in the lateral funiculus down to sacral levels, ending on the most lateral and dorsal motoneuronal and internuncial columns of the anterior horns of the intumescences that supply the limbs [136]. This tract comes mostly from motor cortex projection neurons [8,43]. The ventral PT of Türck [34,90] descends ipsilaterally in the sulcomarginal zone of the cord and is the direct spinal prolongation of the bulbar pyramids themselves. This tract comes from area 6 [49,95] and crosses in part and sequentially in the anterior comissure down to the sacral cord, ending predominantly on the medial neuronal pool of the anterior horns, the motoneuronal components of which supply axial and limb girdle muscles [50]. Finally, the most sparse of the three, the homolateral PT [5,36,81,91], travels along the ipsilateral lateral funiculus intermingled with regional ascending tracts. It distributes its fibers to the same motoneuronal pools of the crossed lateral tract and may come from the supplementary motor area of the ipsilateral hemisphere. This rigid segregation of fibers into distinct tracts according to their respective areas of origin is certainly not decisive for proper functioning, for in individuals in whom the relative bulk of such tracts deviates from this basic design, descending PT axons are probably intermingled with the other extant PTs, since their total fiber number is quite constant for each animal species. In view of the already mentioned considerable interindividual variations, any arrangement of fibers in concise tracts in the cord is surely crucial for the ultimate pattern of motor deficits to be anticipated from any particular PT damage at any height *below* the pyramidal decussation level.