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MOTOR HEMIPLEGIA AND THE CEREBRAL ORGANIZATION OF MOVEMENT IN MAN

II. THE MYTH OF THE HUMAN EXTRAPYRAMIDAL SYSTEM

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SUMMARY — Following a brief review of the concept of extrapyramidal system, clinical and anatomic evidence is presented against its relative prominence in man. It is proposed that the greatest part of those structures traditionally labeled as extrapyramidal effects its respective functional activities by way of the pyramidal tracts themselves. Such structures, centered around the basal nuclei, the cerebellum and possibly, the limbic areas of the prosencephalon are, according to the present suggestion, indeed, **pre** pyramidal. This model is based upon the clinical analysis of patients and agrees with more than one century of anatomic verifications in human brains, favoring the notion of the singularity of the human brain.

Hemiplegia motora e organização cerebral do movimento no homem: II. O mito do sistema extrapiramidal humano

RESUMO — Após revisão sumária do conceito de sistema extrapiramidal, são apresentadas evidências clínicas e anatômicas contra sua importância relativa no homem. Propõe-se que as estruturas tradicionalmente agrupadas sob o rótulo extrapiramidal efetuem a maior parte de suas atividades funcionais, respectivamente, por intermédio dos próprios feixes piramidais. Tais estruturas — centradas nos núcleos da base dos hemisférios cerebrais, no cerebelo e, possivelmente, também, nas áreas límbicas do prosencéfalo —, de acordo com nossa proposição são, na verdade **pre**-piramidais. Esse modelo se baseia na análise clínica de pacientes, está de acordo com mais de um século de verificações anatômicas em cérebros humanos e depõe a favor da noção de singularidade do cérebro humano.

Anatomical and functional reorganization in the human brain — Contrary to the pyramidal system concept [109] which evolved in daily clinical practice, the concept of the 'extrapyramidal system' (EPS) was coined by Prus at a physiology laboratory in 1898 [124]. He investigated the means by which cortically induced epilepsy spread down to segmental levels. Working on dogs, Prus realized that the convulsions would not be affected by bilateral transections of the pyramidal tracts (PTs) at either mesencephalic or pontine levels, which led to the conclusion that epileptic volleys could be transmitted caudally through pathways distinct from the PTs. These, he proposed to call "extrapyramidal" It is noteworthy that Prus believed such EP pathways to follow a sequential multisynaptic course from the cerebral cortex to the spinal cord, due to a number of interruptions in subcortical and brainstem stations. He also suggested that such tracts would be responsible for the 'coordination and association' of movements. Contemporary studies have confirmed and extended Prus' views by showing the anatomical organization of the cortico-reticulospinal tracts [132] and their crucial role in the propagation of cortical epilepsy to the segmental apparatus [149],

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as well as in the integration of body axis balance during standing and locomotion [83]. Unfortunately, these conclusions may not be directly applicable to the human case, for they are entirely derived from animal experimental data.

Prus' studies did not receive widespread attention and lay injustly forgotten by many, even today. The influential work of the Vogts [153], Wilson [157] and Hunt [69] independently revived the concept of an EPS in man, by defining in clinical language the syndromes of the pallidum, the putamen and the corpus striatum, respectively. These notions then gained wide acceptance and were soon incorporated into routine clinical parlance, albeit in a somewhat different meaning, for while Prus envisaged it primarily as a physiological concept, neurologists employed it strictly within a clinical and *presumed* anatomic context. So, in 1917, Hunt conceptualized the EPS as a group of descending tracts with departure points in definite brainstem nuclei — red nucleus, subthalamic nucleus, reticular formation, and locus niger —, in their turn under direct control from the basal ganglia [70]. A final step was taken with the anatomic demonstration of the corticostriate connections, which paved the way for the suggestion that there existed a cortical level of organization at the top of the EP hierarchy [51] (Fig. 1). Such a 'longitudinal' EPS concept — from cortex to cord — was

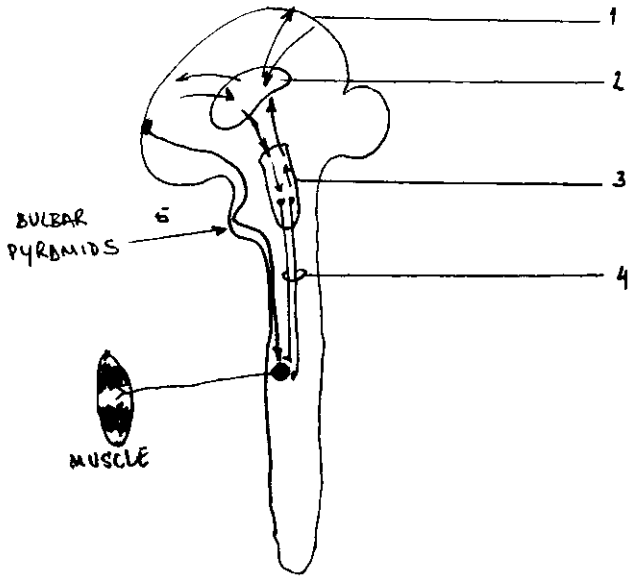

Fig. 1 — Classical schema of extrapyramidal system (EPS). 1 is the cortical EP level; 2, basal ganglia level; 3, brainstem level and 4, descending supra-spinal EP tracts. The cerebellar system, traditionally classified as EP too, is not represented. Note that the EPS, according to this model, is organized *in parallel with the pyramidal system* (5).

then explicitly formulated in clinical terms by Yakovlev in 1944 [160]. More recently, the cortical EPS was further fractionated in view of the development of the 'ventral striopallidum concept' [62], which postulates that the EPS is the main avenue for the expression of limbic-related neural activity as well [54].

LS and PMH add evidence to the notion that what is traditionally thought to be the function of EP pathways are, in fact, *not*, as least in human beings. This led to the conclusion that, *in man, the PTs are the conveyors of most of those functional motor patterns classically attributed to an extrapyramidal mediation.* The residual motor functions which survive experimental pyramidotomy have for a long time been ascribed to the EPS. For several years, an operational model of such a system has been developed and the resultant picture has been transfered to the human case. However, efforts to apply these models to clinical practice have been hampered by apparently contradictory observations: most patients simply do not match the model. The remaining portion of this paper will attempt to show that this presumably happens as a consequence of looking at patients through monkeys' brains...

The available anatomic evidence [79,103] consistently shows that EP pathways in the human cord are meager, scattered and inconspicuous. They all appear to form in brainstem nuclei, but their exact origin remains uncertain, though it seems that most do not descend beyond the upper half of the cervical cord. Presumably, these pathways

are made up of mesencephalic fibers coming from the interstitial nucleus of Cajal (interstitiospinal tract) and the superior colliculus (tectospinal tracts). They are possibly related to the effectuation of some head-and-eyes synergies (see table III). The vestibulospinal and rubrospinal tracts are very rudimentary and may be even lacking in some individuals [106,141]. So the only probably significant EP pathways to reach the caudalmost levels of the human cord are the reticulospinal tracts (RSTs) [100], although they too may be relatively small in man [101,102]. The existence of a 'reticular reflex' type of myoclonus [55], however, speaks for specific roles of RS pathways in relation to the skeletal musculature, besides the more classically acknowledged respiratory and vegetative ones [29,99].

The medial RSTs are the prolongation of the medial longitudinal fasciculus [138,145], running along the sulcomarginal zone of the cord, intermingled with Türck direct PTs and propriospinal fibers [90]. Similarly, the lateral RS fibers do not make up any compact tract in the ventrolateral funiculus, where they descend scattered among other ascending regional fiber tracts [6,118,134]. The terminations of the medial RS fibers at least, are, in a way, complementary to the lateral PT ones, for they preferentially end bilaterally in the medial motoneuronal and internuncial pools of the anterior horns thus indirectly influencing the activity of axial and proximal limb muscles [107]. Then, the only bulky descending supraspinal pathways that can be sharply and readily identified are the PTs [103]. Moreover, the considerable volume of the bidirectional propriospinal paths makes them easily demonstrable in human cords, as well [104].

Thus, as shown by LS cases, if (a) patients deprived of PTs are also deprived of most of those motor patterns traditionally labeled as EP, and if (b) anatomy has denied the existence of any comparatively significant descending supraspinal EP pathways, it is then suggested that in the human brain, after establishing specific thalamic links in non-overlapping specific nuclei, the main bulk of anatomic subsystems of fibers centered in basal ganglia and cerebellum proceed rostrally to the areas of the cerebral cortex from which the PTs originate. The role of the thalamus in this design must be critical for the redistribution of the incoming basal nuclear and cerebellar activity to precisely defined sectors of the precentral 'PT cortex' [1,120,137]. Had one to summarize such concepts in just a few words, it might be said that the 'head nuclei' classically assumed to be extrapyramidal are, indeed, '*pre*pyramidal'.

A few authors in the beginning of the present century had already glimpsed the possibility of a dominant role for the PTs, in contrast to EP ones, in the generation of suprasegmental activity [140,158], but were apparently unable to grasp fully their respective functional importance. Only recently this became a possibility, once the LS as a concept crystalized [48,117,123,152] and could be taken as a reliable model for pyramidotomies in man. Even in lower animal forms, this 'pyramidalization trend' can already be seen to coexist with the more massive EP descending pathways [40], and for mammals, including non-human primates, this stands as a conservative architectural layout. This anatomic schema fits surprisingly well with daily experience, sometimes in an almost exotic fashion [155]: once the PTs are interrupted, the individual is immediately reduced to a segmental (reflex arc) and multisegmental (propriospinal) reflex pattern of motor behavior. In such cases, most cerebellar and basal ganglion activity is presumably prevented from reaching the segmental apparatus of the cord. Uni or bilateral hemiplegia are the semeiologic manifestations of this phenomenon, provided the interruption is complete and occurs above the bulbar pyramidal stage. Such rudimentary motor competence in LS agrees with the anatomic data pertaining to the equally sparse EP pathways in the human cord. Additional evidence can be invoked to strengthen this view, and this will be briefly revisited now.

First, as conventionally recognized by neurosurgeons and neuropathologists, if most of the classic diseases known as 'extrapyramidal' — resting and action tremors, chorea, athetosis, cortical reflex myoclonus, ballismus, torsion dystonia — are to become clinically apparent, a critical amount of structural integrity of the PTs must remain [19,59,71,113,125,128,146,147]. Conversely, a natural or artificial injury inflicted on the PTs will turn a dyskinetic movement into the stereotyped Wernicke-Mann hemiplegic attitude. Similarly, if any of the well known post-hemiplegic disorders of movement is to supervene at all, again a critical amount of the PTs must be functionally patent [21,39]. It is as if the abnormal physiologic activity generated at a given subcortical or brainstem locus, and which emerges embodied in a particular type of

purposeless movement, were effected through the PTs, after being relayed to the precentral motor cortices from specific thalamic links.

Second, the observation that the hemiplegia resulting from surgical hemispherectomies [126] or from cerebral degenerative hemiatrophies with or without anatomic PT involvement [73,116,140] is identical to that which supervenes after a restricted PT injury, even in infancy [163], reassures us that this tract is, indeed, the main way out for the most meaningful lot of all descending suprasegmental activity. This may become even clearer if we contrast the effects of hemispherectomies carried out on other animal species [154], for in them the residual motor abilities is considerably more preserved than in man.

The third statement is implicit in the clinical picture of LS and derives from considerations regarding the mechanism of recovery from a PT lesion in the internal capsule [44], in the ventral pons [42] or in the spinal cord [105]. There is scanty, though concise, evidence that the variable degrees of recovery commonly seen to occur in such cases depend on the PTs of the unaffected side and probably not on EP pathways, as may be the case in some non-human primates [83]. This fact has been cogently demonstrated [2,4,44]. One recently reported patient was rendered hemiplegic by an internal capsular infarct [44], yet was mildly hemiparetic and able to walk less than one month later. Then, the other internal capsule suffered an infarction and she became immediately locked-in, only capable of moving the left toes in response to commands. Unfortunately, her neuro-ophtalmologic examination is not cited on this short report. It appears that in such cases the healthy PT is responsible for recovery, which cannot be obviously assigned to any 'EP pathways' for these simply do not exist at such rostral levels. Besides, once locked-in, patients with verified extensive PT destruction so remain for an indefinite length of time, even for years, again pointing to the exclusion of efficient recovery mechanisms outside the main PTs injury.

Finally, one particular aspect of cerebellar symptoms deserves comment, namely, the well-known ipsilaterality of signs which occur after a cerebellar injury. In man, by far the main set of efferent fibers from the cerebellum is grouped in the superior cerebellar peduncles, which completely decussate in the ventral mesencephalon before heading for specific ventrolateral thalamic nuclei. In turn, efferent fibers ascend to the precentral cortex from the thalamus. As cerebellar symptoms are *ipsi*lateral to a lesion situated at any point from the cerebellar cortex up to the superior peduncle decussation, and *contra*lateral to a lesion lodged at any point from the ventrolateral thalamus [52,67] up to the recipient cortical areas of this nucleus [111], there must be a decussation along the cerebellar pathways other than the superior peduncle one. It is suggested that this second decussation is implicit in the pyramidal decussation itself, the cerebellar motor control being directly exerted at the cortical motor level. This view gains further support from the virtual absence of rubrospinal tracts in man [106,141], thus excluding EP pathways from the mediation of cerebellar function to the segmental apparatus, at least in ways similar to those known to be operative in other mammals.

The above considerations should not be taken to imply an unconditional absence of EP pathways in man. Only their comparative smallness in our species should be stressed. Additionally, most of their functions in other species seems to have been taken over by the PTs, as if a process of structural brain reorganization, for which quantitative data have been collected since the last century (for a review, see Holloway [66]), had happened in the furnaces of phylogenesis. Table 3 gave us some idea of the kind of motor pattern that falls within the domain of the human EPS. The main goal of specifying functions within each one of these fiber systems is still a program-in-execution whose success will depend on the collaborative work in the general field of the clinical neurosciences. As stated elsewhere [112], descending motor systems are, after all, heterogeneous functional systems which may simultaneously conduct distinct types of activity generated at related functional systems within the encephalon. For example, as has been speculated, there may be some 'basal ganglion factors' in the PTs, just as there may be some 'cerebellar' ones, too. Perhaps, the columnar connections of each specialized afferent path within the cortex are as non-overlapping as they have been shown not to be at the thalamic level either and, then, each corresponding efferent PT axon might be differentially tied, in the long run, to a well defined subcortical subsystem [119,162].

Closing remarks — Refinement or challenging of these ideas will probably be soon forthcoming, for they may all be testable today. It seems that a key factor to shape their future development will necessarily lie on the revival of the methods for the study of secondary degenerations within the nervous system, unfortunately seldom used now. An immense treasure in brains has been going to waste for decades because the majority of reported clinicoanatomic case-studies limits itself at pointing to the location and histologic type of the gross lesions found at autopsy. To the clinical researcher, it is not enough to even sharply localize holes within the brain, no matter by which technic: it is as essential to have in hands a trusty Ariadne's thread, a detailed holographic map, as it were, of the linkages set up by the injured sites. Studies such as these, moreover, could be beautiful complements to PET and SPECT measurements eventually made previously in vivo. Thanks to the centenary wisdom embodied in the clinical method and the glamour of modern biomedical technologies, Neurology has become, after all, a fascinating enterprise which can be enjoyed from even apparently trivial phenomena of daily practice, as is the case with hemiplegia. In the near future, patients should also profit from these efforts by means of new and more effective therapeutic strategies devoted to the alleviation of motor symptoms.

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