Hemichorea resulting from a local lesion of the brain. (The syndrome of the body of Luys.) By James Purdon Martin, MD (London). Brain 1927; 50; 637–651; Hemichorea associated with a lesion of the corpus Luysii. By James Purdon Martin and N.S. Alcock. Brain 1934; 57; 504–516; and Hemichorea (hemiballismus) without lesions in the corpus Luysii. By J. Purdon Martin (From the National Hospital, Queen Square, W.C.1) Brain 1957: 80; 1–10.

One half of Brain volume 50 is devoted to papers delivered at a combined meeting of the Section of Neurology of the Royal Society of Medicine and the American Neurological Association, held in London on July 26–28th 1927. James Purdon Martin (1893–1984) presented the case of an arteriosclerotic, hypertensive, syphilitic seaman who developed headache, slept for a few hours and then woke with violent right-sided involuntary movements: ‘when he got up that evening to go to his ship, his right arm was twitching and swinging about widely, and he was throwing his right leg’. Whilst awake, the right arm and leg were tossed about by violent movements originating from all joints but especially the shoulder and hip, and in all directions, producing an amplitude of movement ‘not often seen in chorea’. The head and trunk were involved, to some extent, as were speech, swallowing and respiration. Free and quick, and unhampered by spasticity, the limb movements led to abrasion of the skin on his knuckles and knees. The moving limbs felt hot and sweated. ‘All these movements ceased entirely during sleep’. Motor, sensory and reflex functions were in all other respects normal. Transferred to the National Hospital, Queen Square, he died from pneumonia on the 20th day of the illness. Dr (Joseph Godwin) Greenfield performed the autopsy.

Recent haemorrhage had destroyed the left corpus Luysii, apart from a sliver remaining at the dorsal margin (see Figs 1 and 2); ‘the whole space destroyed by the haemorrhage was almost exactly the position of the body of Luys and of that part of the lenticular bundle (H2) of Forel which lies medial to it; the zona incerta was reached on the medial side and there was a slight bulging into the internal capsule laterally’. Old lacunar infarctions were present in the left part of the pons, dentate nucleus of the cerebellum (bilaterally) and right thalamus. Dr Purdon Martin’s survey of the existing literature on violent hemichorea is much hampered by the nuances of definition adopted by previous commentators; but 15 case reports appearing between 1883 and 1925 do seem relevant. From these, he offers a description of the ‘syndrome of the body of Luys’: wide amplitude proximal chorea on the opposite side to the lesion; mild involvement of the face and bulbar musculature; cessation of the movements during sleep; hypotonia and reduced tendon reflexes; increased temperature and sweating on the affected side; emotional distress and impaired cognitive function on recovery; and a poor prognosis, characteristically with death from pneumonia.

Purdon Martin considers that the pathogenesis of hemichorea depends on involvement of the dorsal part of the body of Luys. His assumption is that fibres descending from the lower part disconnect an intact pathway involving the red nucleus, substantia nigra and corticospinal tract, but with intact influences from the striatum and...
pallidum. In discussion, Dr (SA) Kinnier Wilson clearly felt uneasy with this formulation reminding his younger colleague that the corpus Luysii lesions were destructive and could not therefore directly have given rise to chorea; moreover, he prefers the loss of an ascending influence on the corticospinal pathway as the mechanism. But, in reply, Purdon Martin maintains that he had 'never for a moment lost sight of the fact that choreic movements must arise from surviving structures; and (has not made) the mistake of ascribing positive symptoms to a destructive lesion'.

In 1934, Dr Purdon Martin and Dr NS (Nathaniel Scott) Alcock report another arteriosclerotic patient who developed violent left-sided hemichorea of the arm and leg, after a brief episode of involuntary movement occurring on the same side earlier that day: violent jerking movements caused his arm to strike the chair, and his foot kicked the fender of the fire by which he sat; he took on and off his glass 'by happy shots'. Wild and wide amplitude asynchronous choreic movements of the arm and leg, in all directions, continued; later, he became confused, developed bulbar difficulties, and died within a few hours of admission to the National Hospital on the 8th day of the illness. Dr (Sir Charles) Symonds diagnosed a small haemorrhage in the right corpus Luysii. At autopsy, Dr Greenfield demonstrated just such a lesion, no larger than a pea, more extensive in the lower portion of the nucleus, sparing the anterior and lateral portion and extending medially into the zona incerta and Forel's bundle (H2) but not the ansa lenticularis. There was another minute recent haemorrhage in the right thalamus. The face had been spared, perhaps reflecting the small size of the lesion and somatotopic arrangements within the corpus Luysii. Now, the authors refer to the affected structure as the subthalamic nucleus and—following von Economo—describe the movement as hemiballismus (Fig. 3).
Clearly niggled by Kinnier Wilson’s 1927 interrogation, Dr Purdon Martin and Dr ‘Barney’ Alcock (happily still living and now in his 98th year) engage with matters of pathophysiology in more detail. The corpus Luysii is the critical structure that must be damaged for hemichorea (hemiballismus) to appear; that said, Forel’s H2 bundle and the zona incerta are often, but not invariably, included in the lesion. In debating whether violent hemichorea might develop with lesions elsewhere, they fire a somewhat flaccid salvo at Wilson who had published a case showing atrophy of the post-central cerebral gyrus and a normal corpus Luysii, on the basis that the disease process he described was degenerative and not haemorrhagic (Wilson SAK. Die Pathogenese der unwillkürlichen Bewegungen mit besonderer Berücksichtigung der Pathologie und Pathogenese der Chorea. Deutsche Zeitschrift für Nervenheilkunde 1929: 108; 4–38.) More troubling for them were cases of hemichorea in association with striatal or thalamic lesions; but, in each instance, the area of damage had impinged on the corpus Luysii. Of these, only one—communicated personally by Professor Fragnito who had specifically re-examined tissue from his previously published case (Fragnito O and Scarpini V. Report anatomo-patologico in un caso di emicoreap sintomatica. Rivista di patologia nervosa e mentale 1926: 31; 524 et seq.)—challenged their exclusively Luysian formulation for the origin of acute hemiballismus. On the issue of whether the hemichorea of corpus Luysii lesions could be distinguished from other acute unilateral movement disorders, the authors emphasize their proximal nature, freedom from other motor deficits and violence; however, all these features may be seen, for example, in Huntington’s disease. In short ‘the corpus Luysii is the critical structure damage to which results in hemichorea .. evidence that focal damage to any other structure in the brain results in hemichorea is extremely scanty .. “hemiballismus” consequent on a lesion of the corpus Luysii does not, in our opinion, differ from other choreas except in intensity’.

Thirty years after first writing on hemichorea (hemiballismus), and with the advantage of a complete survey of the literature to 1952 (Martinez Y. Contribution à l’étude de l’Hémiballisme. Paris, 1953) and experimental work in primates (see for example, Carpenter MB. et al. Analysis of choreoid hyperkinesia in the rhesus monkey. Surgical and pharmacological analysis of hyperkinesia resulting from lesions in the subthalamic nucleus of the monkey. Journal of Comparative Neurology 1950: 92; 293–331), James Purdon Martin concedes that hemiballismus may occur with lesions outside the corpus Luysii. The experimental literature shows the critical event to be loss of the bi-directional pathway connecting the corpus Luysii and the globus pallidus, behind and in front of the internal capsule, such that hemiballismus may result from damage to these subthalamic-pallidal pathways and not the nucleus itself. An elderly woman developed hemiballismus 3 months after recovering partially from hemiplegia in the same limbs, and dying 2 months later. Pathological examination by Dr William Blackwood at the National Hospital showed occlusion of the left posterior cerebral artery with infarctions that included fibres passing from the posterior corpus Luysii to the pallidum but not the subthalamic nucleus itself. A woman, aged 68, developed left-sided hemiballismus over a period of 3 weeks; the violence of the movements fluctuated as muscle strength was gradually lost and she died with a dense hemiplegia but no hemichorea. Lesions of several ages were present at autopsy; ‘it is impossible to be dogmatic .. but my interpretation is that the lesion(s) interrupt(ed) a large contingent of fibres from the corpus Luysii passing in the ansa lenticularis to enter (the globus pallidus) directly, from the front, or laterally by way of the external medullary lamina’. A woman of 74 experienced a sudden increase in the extent and severity of involuntary movements, of several weeks duration, affecting the right leg and soon also involving the arm: 6 weeks later, she became aphasic and hemiplegic; the movements ceased and she died soon after. At autopsy, there was occlusion of the left internal carotid artery; the origins of the anterior choroidal and posterior communicating arteries from which the subthalamic nucleus receives its blood supply were patent but flow was now dependent on the (somewhat arteriosclerotic) posterior cerebral circulation (Fig. 4).

Dr Purdon Martin speculates that, in this case, there was relative ischaemia of the corpus Luysii compared with the globus pallidus, thus upsetting the physiological balance of these structures. In that respect, these three cases with observed or inferred damage to the Luysii-pallidal pathways all had less marked hemiballismus than that seen with direct structural lesions of the corpus Luysii; and the severity of the movements may also have been tempered by damage to the
pallidum itself since it is the balance of activity in these two structures that is the basis for hemiballismus. Thus James Purdon Martin felt able, after 30 years, to answer Kinnier Wilson’s question on the physiological basis for this characteristic movement disorder. Hemiballismus does not arise from the destroyed corpus Luysii but as a result of irregular impulses issuing from an intact globus pallidus, freed from restraints normally imposed by its connections with the subthalamic nucleus.

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