HEMIBALLISMUS: AETIOLOGY AND SURGICAL TREATMENT

BY

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Hemiballismus is a relatively uncommon hyperkinesia characterized by vigorous, extensive, and rapidly executed, non-patterned, seemingly purposeless movements involving one side of the body. The movements are almost unceasing during the waking state and, as with other hyperkinesias considered to be of extrapyramidal origin, they cease during sleep.

Clinical Aspects

Cases are on record (Whittier, 1947) in which the abnormal movements have been confined to a single limb ("monoballismus") or to both limbs of both sides ("biballismus") (Martin and Alcock, 1934; von Santha, 1932). In a majority of recorded instances, however, the face, neck, and trunk as well as the limbs appear to have been involved.

The patient usually retains a measure of voluntary control over the affected member so that acts such as walking, putting on slippers, and touching the hand to the face can be performed even if with difficulty. The ballistic movements cannot however be voluntarily checked for more than a few moments at a time. The muscle tonus of the involved limb(s) may be normal but in many instances is slightly diminished (Martin and Alcock), and in consequence of the reduced initial tension of the muscles the deep reflexes of the affected limbs are often moderately diminished.

The vast majority of patients are at or beyond middle life at the time of onset of hemiballismus. However, reports of patients as young as seven (Bonhoeffer, 1930) and eighteen (Bianchi, 1909) years have been recorded. There appears to be no significant difference in sex incidence.

Spontaneous recovery is not to be expected and by far the majority of patients succumb within a few days to several months of progressive exhaustion, cardiac failure, or pneumonia (Grinker and Bucy, 1949; Whittier). A few instances are on record in which the disorder has run an extended chronic course (Touche, 1901; Marcus and Sjögren, 1938), while in one case reported by Lea-Plaza and Uiberall (1945) the abnormal movements are said to have ceased spontaneously after seven weeks. Hemiballismus has also been known to cease following the supervision of a hemorrhagic ictus.

Terminology.—There appears to be among writers on this subject no agreement regarding the precise features of the clinical phenomena to which the term hemiballismus may properly be applied. Various authors have credited Kussmaul and Fischer (1911) with introducing the term hemiballismus to signify the flinging or flapping character of the limb movements, but in general each observer’s concept of the features of hemiballismus represents but an abstraction of his own limited experiences. The consequence is that at most clinical demonstrations of a case alleged to be one of hemiballismus one or more members of the audience are moved to assert that the case in question is not "really" an example of hemiballismus, but one of hemichorea, athetosis, dystonia, or perhaps hysteria. If, on the other hand, the case is presented initially as an instance of hemiathetosis or chorea, similar protests may be raised.

The terms athetosis, chorea, and ballism refer to involuntary, non-patterned movements of bodily members, which are unpredictable in respect of time and form. They may be distinguished from the myorhythmias, myoclonus, the coarse, alternating tremors of Parkinsonism, and the fine tremors of thyrotoxicosis. They are present in the waking state and absent during sleep. They appear both "at rest" and during the execution and maintenance of "voluntary" movements. In our view, the three varieties may be distinguished from one another on the following grounds. In athetosis the movements are relatively slow, vermiform and writhing, and in the limbs are chiefly due to the action of distal muscles. The proximal limb muscles may also participate. Athetotic movements are character-
historically intermittent so that periods of several minutes may elapse without the exhibition of involuntary activity. In chorea the movements are much more sudden and, insofar as the limbs are concerned, are subtended chiefly by action of the more distal rather than the proximal muscles. As in athetosis, choreic movements are frequently intermittent. Ballistic movements resemble choreic movements in that they also are rapidly executed. The muscles of the face, neck and/or trunk may or may not be involved. The proximal muscles of the limbs are regularly involved, with the consequence that the limbs exhibit large excursions. Close inspection however usually reveals involvement also of the more distal limb muscles. In contrast to the intermittent activity characteristic of chorea and athetosis, that of hemiballismus is almost ceaselessly at play during the wakeful state.

**Additional Clinical Manifestations.**—The intellectual, emotional, and sensory functions are seldom deranged in hemiballismus. However, the coexistence of psychological aberrations, vegetative disturbances, hemiparesis, dysphasia and/or oculomotor disorders with hemiballismus has not infrequently been reported. In most such instances necropsy has demonstrated multiple or extended lesions involving neural structures other than those considered responsible for the ballistic movements.

**Aetiology and Pathology**

The pathological process most frequently encountered in hemiballismus is a circumscribed encephalomalacia which is usually the result of cerebral thrombosis. Diabetes mellitus not infrequently underlies the thrombotic process. Somewhat less frequently small circumscribed hemorrhages and emboli appear to be the responsible agents (Martin and Alcock, 1934). Other lesions, including tuberculoma, gumma, metastatic abscess, metastatic neoplasm, encephalitis and the so-called primary degenerative processes of the brain, have been identified. In two instances trauma has been considered the causative agent (Bucy, 1944; Schob, 1920).

The anatomical sites at which lesions have been encountered may be conveniently described under two main headings.

**Lesions Involving the Corpus Subthalamicum.**—The structure most frequently involved in hemiballismus is the corpus subthalamicum contralateral to that side of the body exhibiting abnormal movements. Greiff (1883) is credited with having been the first to observe this correlation. In a carefully executed study forty years later this observation was firmly substantiated by Jakob (1923). Subsequent post-mortem studies have provided over fifty instances in which the corpus Luysi has been damaged, though often along with other neural structures. Indeed, the number of studies in which, like those of Matzdorff (1927) and Thurel and Grenier (1947), the only demonstrable brain lesion is limited strictly to the corpus subthalamicum is very small.

In a review of the literature up to 1934, Martin and Alcock (1934) asserted that no case of hemiballismus had been fully described in which the corpus subthalamicum was not found damaged. This extreme position no longer appears tenable, but Whittier's (1947) observation that hemiballism is the "apparently inevitable symptom in man of destruction of the subthalamic nucleus" is probably quite correct.

**Lesions Involving Other Structures.**—Several cases of hemiballismus have been reported in which the corpus subthalamicum is asserted to have been found intact at necropsy. In these the lesion considered responsible for the hyperkinesia was disclosed in other parts of the brain.

**Lesions of the Afferent or Efferent Fibres of the Corpus Subthalamicum.**—In two of the earliest cases on record, those of Bianchi (1909) and Bonhoeffer (1897), the abnormal movements were imputed (Martin, 1928) to lesions at a level "lower than" the corpus Luysi, presumably situated so as to destroy its efferent connexions to the midbrain. That damage to either the afferent or efferent fibres of the nucleus might produce hemiballismus was postulated in 1939 by Moersch and Kernohan. Subsequently, Papez, Bennett, and Cash (1942) described a case which they considered illustrative of the point. The presence of multiple lesions in their case unfortunately equivocates their interpretation. Kelman (1945), however, recorded the necropsy findings of a hemiballistic subject in which a small metastatic carcinoma from the lung involved the afferent subthalamic tracts.

**Lesions of the Corpus Striatum.**—In 1926 Fragno and Scarpini described the necropsy findings in an 80-year-old male who had suffered from hemiballismus for three years before his death. An encephalomalacic process had markedly damaged the medial portion of the putamen and, to a lesser degree, the head of the caudate nucleus. The corpus Luysi, medial lemniscus, and fields of Forel were reported to be undamaged. Several other investigators (Vogt and Vogt, 1920; Austregesiol

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*This is the only case acknowledged by Martin and Alcock to constitute a valid exception to the doctrine concerning the necessary involvement of the corpus subthalamicum in the pathogenesis of hemiballismus. Its importance theoretically and practically is obvious.
and Gallotti, 1924) have reported cases in which lesions of the corpus striatum were considered responsible for hemiballismus or for hyperkinesias simulating the latter. Certain of these cases have been objected to by Martin and Alcock (1934) on the ground that conclusive evidence clearing the corpus subthalamicum of damage has not been proffered. However, Austregesilo and Borges-Fortes (1937) and Davison and Goodhart (1938) have each reported well-studied cases in which the neostriatum alone has been the seat of damage. Although their cases were published under the titles of "hemichorea" and "monochorea", the descriptions of the abnormal movements conform so closely to those of hemiballismus that differentiation appears difficult if not impossible.

In Case 2 of the present communication, the lesion apparently responsible for the patient's hyperkinesia was disclosed in and adjacent to the right lenticular nucleus, i.e., involving both the neo- and paleostriatum. Histological examination revealed no implication of the corpus Luysi.

Lesions in the Thalamus.—Lewandowsky and Stadelman (1912) reported a case of hemiballismus in which a large lesion was disclosed in the lateral thalamic nuclei extending inferiorly into Forel's Haubenfelde. Unfortunately, however, the authors made no specific report regarding the corpus subthalamicum. In a case described by Nikitin (quoted by von Santha, 1932) a relatively large lesion was encountered in the ventrolateral thalamus. Its ventral border is stated to have extended to within 2 mm. of the dorsal border of the corpus Luysi, the latter structure being evidently uninvoluted. At least two other instances have been recorded in which the apparently responsible lesions were demonstrated in the thalamus at a sufficient distance from the corpus Luysi to vitiate the frequently raised objection that the blood supply of the latter must have been compromised (Malan and Civalieri, 1921; Zontoni, 1908).

Lesions in the Postcentral Gyrus.—In 1929 Wilson reported a case in which degenerative, atrophic changes of the contralateral postcentral gyrus were the only lesions demonstrable at necropsy. Although this report is unique, it must be retained for consideration in view of Wilson's reputation for thoroughness and objectivity.

In brief recapitulation, then, apparently valid cases of hemiballismus have resulted from lesions in the afferent and efferent fibres of the corpus Luysi, the neo- and paleostriatum, the ventral and lateral thalamus and the postcentral gyrus. Although few in number compared with those in which the responsible lesion involves the corpus Luysi, such cases compel reservation concerning the doctrine that hemiballismus is invariably subtended by a lesion of the corpus Luysi.

Thus far animal experimentation has shed but little light on the anatomico-clinical problems related to hemiballismus. Morgan (1927) reported failure to produce hyperkinesia in a series of dogs in which lesions were placed in the corpus subthalamicum and varying adjacent structures. These findings contrast sharply with those in the human in whom damage of the corpus subthalamicum by disease is regularly followed by ballistic movements.

Wilson (1914) and Delmas-Marsalet (1925) reported inability to produce choreic or ballistic movements in laboratory animals by placing experimental lesions in the caudate nucleus. Similar negative evidence from human material was adduced by one of us (Meyers, 1941; 1942a; 1942b). Extirpation of all available portions of the head of the caudate nucleus in nine patients with Parkinsonism failed to produce hyperkinesia of any variety. In certain instances the rostral third of the putamen and in one instance the oral pole of the globus pallidus were removed in addition to the caudate head without producing abnormal movements.

Browder (1948) in an independent study on the surgery of Parkinsonism, has reported ablation of the superior half of the head of the caudate nucleus in a series of humans without the development of hyperkinesia.

Mettler and Carpenter (1949) have recently succeeded in producing choreo-atetoid movements in rhesus monkeys by electrocoagulation of the subthalamus. These investigators are at present endeavouring to assay the effects of "analytic" surgery in these animals. Their work constitutes the first promising animal experimentation in the hyperkinetic area.

**Therapy**

In the endeavour to treat the distressing symptoms of hemiballismus numerous physical and pharmacological agents have been employed. As with most other extrapyramidal disorders, the results of such therapy have been largely disappointing (Meyers, 1942a). The treatment of hemiballismus may be conveniently discussed under three general headings: physical, pharmacological, and surgical.

**Physical Measures.**—Bertrand and Garcin (1933) described a case in which it became necessary to lash the hemiballistic arm to the patient's side in order to prevent self-injury. Wilson described a similar case in which the hyperactive upper limb was secured to an iron frame in order to prevent the patient from raining blows upon herself.
Pharmacological Measures.—It has been a matter of general experience that sedatives, hypnotics, and narcotics prove effective in hemiballismus only when administered in doses sufficiently large to induce sleep. The atropine derivatives have yielded uniformly indifferent results. Stramonium has been said to exert a "somewhat quieting" effect and tobacco is stated to be "useful," but in no instance has it been claimed that these agents have abolished the hyperkinesia.

Peripheral Operations.—Amputation has been resorted to as a measure of desperation in at least one case of hemiballismus (Schaller, 1937), and in another instance paralysis of the brachial plexus by multiple injections of alcohol was employed (Kulenkampff, 1938). In 1931 Jermutowicz reported a salutary result following mechanical stretching of the brachial plexus. A severe paresis of the limb was produced, but the ballistic movements were sufficiently damped to permit the patient to resume work "albeit with difficulty".

Central Operations.—Jermutowicz quite properly anticipated a return of ballistic movements in his patient. It is of considerable historic interest that he proposed among other possibilities extirpation of the contralateral "cortical center for the shoulder and arm". Just such an operation was performed in 1939 by Bucy (1944) for the relief of left upper monoballismus in a 21-year-old man who four years before had sustained a severe head injury. Pneumoencephalography revealed a dilated right lateral ventricle. At operation the right precentral gyrus was identified by electrical stimulation and that portion from which movements of the upper limb were elicited was extirpated together with portions of the frontal gyri lying rostral thereto (Fig. 1). The anterior wall of the central fissure was included in the resection.

The patient's abnormal movements were abolished following this procedure. By the end of the first year they had returned to a slight degree but at the end of the third year they were described as "minimal and usually present only when he is conscious of being observed". Following operation, convulsive seizures supervened for a time. These were ultimately brought under control by barbiturates.

Ballistic movements were temporarily arrested in one case (Papez and others, 1942) by the intramuscular use of curare. Unfortunately, the dose required to achieve this effect (80 units) produced also a generalized paresis. The unpleasant side-effects of curare, erythroidin hydrochloride, and related agents, their short-lived action, and the necessity of often-repeated parenteral administration impose serious limitations on their continued use.

Surgical Measures.—Amputation, peripheral, and central operations have been reported, with varying results.

![Diagram](http://jnnp.bmj.com/content/13/2/115.f1)

Fig. 1.—Diagram illustrating the cortical area resected in Bucy's case of left hemiballismus. (Reprinted through the courtesy of Dr. Paul Bucy and the University of Illinois Press from "The Precentral Motor Cortex", 1944, p. 362, Urbana, Ill.)
The enduring neurological residua included a marked spastic paresis of upper motor neuron type, hyperreflexia and dyspraxia of the previously ballistic limb. Gross movements, however, could be performed and the patient was gainfully employed.

**Linear Cortico-subcortical Section just Anterior to Area 4**: A New Procedure

With the objects of ameliorating hemiballistic movements and preserving all possible function, the writers recently employed a surgical procedure based on a tentative theory of the pathogenesis of hemiballismus.* In essence, the operation consists of subpial section of the cortex and subcortical white matter immediately rostral to Area 4 in such fashion as to interrupt the U-fibres coursing between Areas 4, 3, 1, and 2 posteriorly, and 4-s and 6 anteriorly. Abstracts of two cases treated in this manner follow.

**Case Reports**

**Case I**.—F.B., a 50-year-old woman, was admitted to the University Hospital on Feb. 14, 1948, complaining of ceaseless agitation of the limbs of the left side. The disorder had begun in mid-December, 1947, as irregular twichings of the left half of the face. The movements increased steadily in frequency and amplitude so that at the end of the second week the left upper and lower limbs exhibited violent and for the most part uncontrolled movements. Toward the end of the first month some slowing of speech and a slight weakness of the left lower limb were noted. The patient considered her condition intolerable.

The previous medical history was non-contributory, except that for the past seven years she had been known to have diabetes mellitus. This condition had been well controlled by insulin and diet.

Examination disclosed an intelligent, affable, white woman in good general condition. The blood pressure was 180/100 mm. Hg. The optic fundi disclosed a moderate degree of diabetic retinopathy. The outstanding finding consisted of vigorous, irregular, and rapidly executed movements of flinging type implicating the proximal and distal muscles of the left-sided appendages. The head jerked irregularly towards the left, the jaws champed and the left side of the face exhibited short-lived grimaces. These movements were almost constantly in play during repose and during voluntary activity they often became violent. They were absent only in sleep. Quick-succession movements and the finger-to-nose tests were awkwardly performed on the left side. The right biceps jerk was sluggish, the left absent. The knee jerks were sluggish but equal, and the achilles reflexes were absent. The remainder of the examination was non-contributory.

**Laboratory Studies**.—The blood counts, hemoglobin level, blood Wassermann test and radiographs of the skull and chest revealed no remarkable findings. The fasting blood sugar was 100 mg. % and the urine showed a 3 plus sugar reaction. The cerebrospinal fluid was clear and under an initial pressure of 125 mm. H₂O. Its total protein measured 37 mg. %. The cell count was normal and the Wassermann test negative.

**Special Studies**.—Pre-operative electrographic studies of the right corpus striatum and neighbouring structures (deep leads) were carried out simultaneously with electroencephalographic recordings from standard placements of scalp leads. The technique employed has been previously described (Meyers and others, 1949). The frequency, amplitude, wave-form, and polarity characteristics of the patient’s recordings were compared with data similarly derived from 11 control subjects. They disclosed no distinguishing features (Figs. 2 and 3).

**Operation, March 2, 1948**.—A right fronto-parietal bone flap was reflected under local procaine anesthesia and the excitably motor cortex identified by means of bipolar stimulation with a 60 cps. sine-wave. The threshold was disclosed as 3V and 25 ma. (approx.). The extent of the motor cortex was mapped out with a 3.5V. stimulus and Area 4 was thus envisioned. Toward the end of this procedure a generalized convulsion was inadvertently produced and during the ensuing period of stupor the ballistic movements were absent. When, after some twenty minutes, the patient again became alert, the hyperkinesia reappeared. A subpial incision was then made through the cortex and subcortical U-fibres along a line just anterior to the envisioned Area 4. It was carried to a depth of approximately 2.5 cm. and ran through those portions of the precentral gyrus described by von Bonin as Areas 4a and 4y (Fig. 4). The suppressor area, 4-s, and the premotor cortex, Area 6, were presumed to lie anterior to the line of incision.

During execution of this incision the ballistic movements ceased abruptly and a flaccid, areflexic monoplegia of the left upper limb was demonstrable. A slight degree of voluntary control over the proximal muscles of the extremity was retained. The left half of the face and the left lower limb proved to be only moderately paretic.

**Early Postoperative Course**.—No hyperkinesia was observed at examination eight hours after operation. The left upper limb remained paralyzed but the patient executed fairly vigorous movements of the left lower limb, both spontaneously and to command. The supranuclear left facial paresis remained unaltered. Plantar stimulation evoked a flexor response bilaterally. The left upper limb was both analgesic and anesthetic.

The patient’s subsequent course was generally satisfactory. On the fourth postoperative day she suddenly became unresponsive for a period of one minute. The eyes deviated to the left but no tonic or clonic phenomena appeared. Upon recovery from this episode the patient was immediately able to engage in conversation. She was neither confused nor distressed. A similar phenomenon occurred on the fifth postoperative day. After the institution of phenobarbital and sodium diphenyl hydantoin therapy there was no recurrence. The anticonvulsant drugs were continued until the end of the third month.

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*This pathogenetic theory will be made the subject of a separate communication.
Fig. 2.—EEG of Case I. The scalp electrodes in the standard positions were connected to reference electrodes on the ears (e). The prevailing alpha wave frequency is 8/s., the amplitude ranging between 25 and 50µv. The frequency, amplitude, wave-form and polarity characteristics of the recordings were similar to those obtained in 11 normal controls. LF and RF = left and right frontal; LP and RP = left and right motor; LO and RO = left and right occipital; RT = right temporal zones.

Fig. 3.—EEGs of Case I derived from ring-type pick-up electrodes placed in the right corpus striatum and neighbouring structures. Electrode No. 1 was the deepest of a series of eight, and was located just ventral to the head of the right caudate nucleus. 1-e, 3-e, 5-e, and 7-e indicate that monopolar recording technique was used, the numbered ring being one of a pair of pick-up leads the second member of which was attached to the right ear (e). The mean frequency of waves derived from such sources was 8½/s., with a range of 6½-11. The amplitude ranged between 10 and 60µv. 1-3, 3-5, and 5-7 indicate that bipolar recording technique was used, the rings being connected to each other as numbered. The mean frequency of waves thus recorded was 11½/s., with a range of 8-16. The amplitude ranged between 5-25µv. RP-e = scalp electrode placed over right motor area and connected to a reference electrode on the ear (cf. RP-e, Fig. 2). These electrostriatograms are indistinguishable from those obtained in 11 normal controls.
The paresis of the left lower extremity increased somewhat during the third, fourth, and fifth postoperative days, but repeated examinations failed to disclose evidence of spasticity, hyperreflexia, clonus, or Babinski's sign. Early in the second week the paresis of this limb improved perceptibly. The paralysis of the left upper limb, however, continued unaltered. The patient was discharged to her home on March 19, 1948.

Later Postoperative Course.—At the end of the third month the patient was still free of hyperkinesia. The left facial paresis had completely receded and the functions of the left upper extremity had returned sufficiently to permit of the cutting of food, tying of shoelaces, and similar practical skills. The motor power of the left upper limb was considered approximately half normal and quick-succession movements and the finger-to-nose test could be executed, although awkwardly. The left biceps and triceps jerks were more brisk than those on the right but Hoffman's sign was absent. There was a slight stereognostic defect. Otherwise, the sensory functions on the left side were unimpaired. In walking, the patient exhibited a slight to moderate degree of circumspection. She could, however, walk on her heels and there was no Babinski sign. A residue of the previous hyperkinesia was discernible as short-lived, irregular, incoordinate movements of small amplitude, present chiefly when the patient was walking or executing "voluntary" movements with the left upper limb. In repose, she exhibited no hyperkinesia.

The patient was re-examined at the end of the fifth month and at intervals of three months thereafter. To date, improvement has been progressive and she is in good health, gainfully employed as a housekeeper. Her gait is entirely normal and the left lower limb is entirely free of hyperkinesia. The upper limb exhibits occasional twitches of small amplitude, particularly when the patient is excited. She is able to use both hands for washing, cooking, ironing, sewing, fastening buttons, tying shoelaces, etc. Dyspraxia is apparent only in such complex skills as crocheting. Stereognosis has returned to normal. The tendon jerks continue somewhat overactive, but there is no spasticity and the Hoffman sign cannot be elicited.

Comment.—Without such information as would be afforded by necropsy study it is manifestly impossible to assert with confidence where the pathological lesion responsible for this patient's hyperkinesia was located. Irrespective of its locus, however, it seems reasonable to infer that the salutary effect of operation was achieved by interrupting some neural circuit upon the integrity of which the mediation of ballistic movements depended.

Case II.—J.S., a 71-year-old housewife was admitted to the University Hospital on January 19, 1949, complaining of severe jerking movements of the limbs of the left side of three months' duration. Early in September, 1948, she had experienced unsteadiness and "dizziness" for a week. In mid-October uncontrollable jerking movements appeared, first on the left side of the face and neck and shortly thereafter in the left shoulder and arm. After a few days the movements abated somewhat, but they soon returned with renewed vigour, and within a month possessed also the left lower limb. During the three weeks before being admitted to hospital they became increasingly more vigorous and were almost ceaselessly at play during the waking state. The patient could not voluntarily suppress the movements. The involved muscles felt sore and fatigued but were not painful. Sedatives were required for sleep.

The patient became very irritable and weary, and during the third and fourth months of her illness lost over 25 pounds. For the ten days preceding her admission she had been unable to walk, even with assistance. Anorexia and nausea were prominent symptoms during the week before admission.

Past History.—Dyspnea and palpitation had been present "for many years," and the patient was known to have had arterial hypertension for about a decade. Digitalis therapy had been administered for one and one-half years before the present illness and there had been no symptoms of cardiac failure. She had borne five healthy children. Except for "nervousness" and intermittent attacks of gall bladder colic over the course of 25 years, the past history was unremarkable.

Physical Examination.—The patient appeared ill and harassed. The blood pressure measured 220/80 mm. Hg. The retinal arteries were somewhat narrow, and owing to opacities of the occular media, the optic disc margins appeared hazy. The cranial nerves were clinically intact. A small nodule was palpable in the left lobe of the thyroid gland. The heart was slightly enlarged to the left and exhibited an irregular irregularity of beat. The chest was emphysematous and the radial arteries felt sclerotic.

Almost continuous ballistic movements possessed the left-sided limbs, the head, tongue, and left half of the face. Owing to their presence the tendon reflexes of the left side could not be reliably evaluated; however, they seemed in general less brisk than those of the opposite side. In the same manner, the muscle tone of the hyperkinetic limbs seemed diminished. The ballistic movements (as analyzed by slow-motion cinematography) were manifestly produced by activities of the muscles of the shoulder and hip girdles as well as of the proximal and distal muscles of the extremities themselves. The strength of the left grip was approximately half that exhibited on the right. Tests for gait and coordination could not be carried out.

Operation, Jan. 28, 1949.—Under rectal avertin and light intravenous pentobarbital anaesthesia the motor cortex was outlined and the threshold of that portion corresponding to the upper extremity determined at 2:25 V. and 0:25 ma., using a 60 cps. sine-wave. Stimulation along the anterior margin of the precentral gyrus and the posterior margins of the frontal gyri just anterior thereto with 4:0-5:0 V. evoked no motor discharges but following the application of such stimuli by 8-15 seconds the previously ascertained threshold value of stimulus failed on several trials to evoke its usual response when applied to the motor cortex. The subsequent return of excitability of the latter at 2:25 V. appeared to indicate that no spontaneous heightening of its threshold had taken place. Other portions of the...
exposed cortex failed to show this phenomenon. Although not without reservation, the operators considered that the forward region probably corresponded to the suppressor strip, 4-s. A subpial cortico-subcortical incision was then carried out as described in Case I (Fig. 4).

Fig. 4.—Diagram of lateral aspect of right hemisphere showing the presumed topographic relations of the line of subpial section carried out in Cases I and II. The motor cortex was identified by electrical stimulation, and the disposition of Brodmann’s Area 4 then estimated by visual inspection. The subpial incision followed a line just anterior to Area 4, probably cutting through 4γ and 4α of von Bonin. Area 4-s, the suppressor strip, and Area 6, the premotor cortex, are represented rostral to the line of incision. The pyramidal and parapyramidal corticofugal fibres arising from Area 4 are represented in their course through the depth of the hemisphere. In the inset, the depth of the incision through the subcortical U-fibres of adjoining and near-neighbouring gyri is illustrated. Int.C. = right internal capsule; Crus = right crus cerebri; G.Pre-C. = precentral gyrus; G.Post-C. = postcentral gyrus.

Early Postoperative Course.—Ballistic movements were absent throughout the remainder of the patient’s course. A left hemiplegia of flaccid, areflexic type was demonstrable following recovery from anesthesia. The vital signs reached normal levels on the third postoperative day. At this time a well-patterned Jacksonian seizure of slight to moderate severity and 30–40 seconds’ duration occurred. It began in the left thumb and spread to implicate the entire left upper limb and the left side of the face. Several such episodes occurred on the third and fourth days. They then regressed and were not seen again. The patient was moderately stuporous for the better part of the first week; after this she became more responsive and by the tenth day was fairly rational. Muscle tonus was first detectable by clinical tests in the left lower limb. The tendon reflexes were at this time absent on the left side and normal on the right. The heart rate was 88 per minute and the blood pressure 130/60 mm. Hg.

During the third postoperative week the patient at times seemed bewildered although most of the time her speech was rational. On the seventeenth day she executed weak movements with the left lower extremity and inquired when she might go home. Although examination was considered not completely reliable, her sensory functions appeared essentially undisturbed. The left hemiplegia persisted.

Terminal Course.—On the twenty-sixth postoperative day the patient’s general condition appeared satisfactory. While sitting up eating supper she aspirated a quantity of milk soup. Dypsnea and cyanosis supervened at once. The skin became cool, mottled, and profusely wet with perspiration. The heart rate increased to 140 per minute, and the blood pressure rose to 220/80 mm. Hg. Auscultation revealed no unusual heart sounds, but coarse rales were heard anteriorly over both sides of the chest. Speech was slow and measured. The patient remained conscious throughout and followed verbal directions well. A moderate quantity of thin mucoid material was aspirated by bronchoscopy from the trachea and left upper and lower bronchi. Oxygen therapy was instituted. The acute episode subsided within 45 minutes.

Electrocardiography failed to provide evidence of a coronary occlusion or pulmonary infarct. A few hours later fine and coarse rales were audible over the left lower lobe. Despite the prophylactic administration of penicillin, pneumonia developed and the patient died three days later, on the twenty-ninth postoperative day.

Necropsy.—An acute bronchopneumonic consolidation was revealed throughout the right lung. There was evidence of a generalized arteriosclerosis. All other significant post-mortem findings were confined to the brain. Some residual subpial hemorrhages and localized
Swelling were noted in the gyri adjacent to the line of cortico-subcortical incision. The interface of the incision was well agglutinated. In depth, it closely approximated 2.5 cm. throughout its course. The operative site was not otherwise remarkable.

Microglia were present but on the whole the glial elements, like the neurocytic, appeared devitalized (Fig. 7, a and b). All portions of the right corpus subthalamicum exhibited well-preserved ganglion cells, normal glial elements, and granular ground substance (Fig. 8). The ansa and fasciculus lenticulare disclosed moderate degrees of demyelination traceable into the ventromedial nucleus of the thalamus, the reticular formation of the anterior midbrain and the substantia nigra.

Comment.—The absence of pathological changes in the corpus subthalamicum and the circumscribed character of the pathological findings in the lenticular nucleus opposite the side of the hyperkinesia renders this a critical case. It may of course be denied that this was a genuine example of hemiballismus; if so, criteria other than those set forth above are in need of definition. If, however, the case be accepted as one of hemiballismus, it follows that lesions lying outside the corpus Luysi may subvert the disorder, a circumstance which in its turn suggests that the integrity of a neural circuit, rather than a particular structure (e.g., the corpus Luysi and its centripetal and centrifugal fibres) is normally required to suppress hemiballismus.

Discussion

Bucy’s case and the two cases cited above demonstrate that by the use of relatively simple surgical

Fig. 5.—Gross coronal section of brain of Case II at level of mammillary bodies (seen from behind), showing encephalomalacic shrinking and obscuration of structures in and neighbouring upon the right lenticular nucleus, external capsule, and claustrum. The corpora Luysi appear normal.

The brain was suspended in formalin for a week. Coronal sections were then cut at intervals of approximately 0.5 cm. The ventricular system was moderately dilated. The gross sections revealed only one lesion: a brownish-purple, granular-appearing discoloration within an obviously shrunken lenticular nucleus (Fig. 5). This finding was most intense in those portions of the putamen and both crura of the globus pallidus appearing in the coronal section through the midportion of the mammillary bodies. The internal and external medullary laminae were almost wiped out. The external capsule and claustrum were likewise obscured and the internal capsule appeared to be but slightly involved. More rostrally and caudally, the lenticular nucleus exhibited less intense pathological changes. The corpus subthalamicum exhibited no gross changes in colour, size, shape, or texture (Fig. 6).

Microscopic study of serial sections alternating through Nissl, Weil, and Masson stains confirmed the gross pathological findings. The right putamen and globus pallidus were the seats of well-established encephalomalacic changes. Their neurocytes were in various stages of disintegration. The less severely affected cells exhibited chromatolysis, swelling, and nuclei in eccentric positions. The more severely affected cells appeared as mere smudges. Many cells had evidently completely disappeared. No inflammatory cells or vascular proliferative processes were discernible. The heterogeneous appearance of the ground substance betrayed its general disorganization. A few gemistocytic astrocytes and
procedures ballistic movements can be promptly arrested, or so significantly suppressed as to afford gratifying relief and thus avert exhaustion. In view of the refractoriness of hemiballismus to conservative therapy, the negligible prospect of spontaneous recovery, the severity of the disability, and the frequency with which the illness ends fatally, it seems desirable to recommend early surgical intervention.

It must be acknowledged that by no means all or even a large majority of cases of hemiballismus can be saved by surgical intervention. Pathological inquiry clearly reveals that in some instances the responsible lesions are progressive in nature, coming ultimately to involve those mesencephalic and diencephalic structures upon which life itself depends. In yet other cases multiple lesions develop. Though surgery appears to offer most promise in those cases where there is a well-circumscribed, non-progressive lesion, it seems reasonable to urge operation for all cases on the ground that this provides the only effective therapy at present available.

Summary

1. The excessive, almost ceaseless movements of hemiballismus commonly lead to exhaustion, cardiac failure, and/or pneumonia. The disorder
thus complicated almost always terminates fatally within a few weeks to several months of onset.

2. The lesions responsible for hemiballismus may be located within the contralateral neostriatum, paleostriatum, thalamus or postcentral gyrus as well as in the corpus subthalamicum and/or its afferent and efferent connexions.

3. Spontaneous cessation of hemiballismus is rare and treatment by physical measures, drugs or peripheral surgical procedures has proved to be disappointing. On the other hand, "central "neurosurgical procedures have afforded relief in a few instances. Two cases subjected to linear cortico-subcortical section just anterior to Area 4 are reported. This procedure appears to produce less neurological deficit than does cortical ablation.

4. The "central "neurosurgical procedures (cortical extirpation, linear cortico-subcortical section, and midbrain pyridotomy) constitute the only definitive treatment upon which reliance may be placed for arresting ballistic movements. In view of the grave prognosis of the disorder, surgery should be regarded as a semi-emergency measure.

REFERENCES

Fig. 8.—Photomicrograph of right corpus Luysi showing the integrity of its neurocytic and glial components. All parts of the nucleus were examined and none showed evidence of pathological change. (Nissl stain.)