THREE CASES OF MYOCLONUS ALLEVIATED BY BILATERAL ANSOTOMY, WITH A NOTE ON POSTOPERATIVE ALIBIDO AND IMPOTENCE

RUSSELL MEYERS, M.D.
Division of Neurosurgery, State University of Iowa, Iowa City, Iowa

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The purpose of this paper is (1) to report the therapeutically useful application of bilateral pallido-pallido-fugal section to 3 cases of myoclonus, thus bringing this relatively uncommon hyperkinesia within the compass of neurosurgery; and (2) to record the occurrence of certain sexual deficits which developed postoperatively in 2 of the patients, each of whom had undergone topically homologous 2-staged procedures. The third patient did not exhibit these deficits, presumably because his procedure was implemented nonhomologically.

Case 1. J.D., a service-station operator, aged 35 years, was evaluated on Dec. 14, 1956. He complained of painless, irregular, rapidly executed and uncontrollable jerking movements of the head, neck, trunk and all extremities. These had first appeared in the muscles of the left shoulder 13 years previously. The trunk became severely involved 6 years later, in consequence of which the patient sporadically exhibited vigorous movements of anterior and lateral flexion. The truncal and cephalic movements often were attended by startling, brief vocalizations, reminiscent of a dog's bark.

These phenomena were absent during sleep, least apparent during repose and regularly augmented by emotionally charged experiences. They became an increasing source of embarrassment to the patient in social situations and perceptibly compromised his business pursuits.

The patient had undergone a variety of physical, pharmacologic and psychiatric (including narcoanalytic) treatments. None had proved helpful. The past history revealed that recurrent sore throats and attacks of rheumatic fever in late childhood had resulted in a persistent heart murmur. Chorea, hypersonnolence, headaches and diplopia were denied. At the ages of 28 and 31 years the patient had had illnesses diagnosed as "virus hepatitis."

He had married at the age of 22 and had fathered three healthy children. He and his wife assertedly were very happily mated and, until the time of his admission to the hospital, had had mutually gratifying coitus on an average of three or more times weekly.

Examination. The most conspicuous feature consisted of sporadic, nonpatterned abnormal movements of truncal and appendicular members which exhibited widely varying degrees of excursion.Expiration was often forceful and from time to time produced a bark-like sound. Muscular rigidity was absent. In all other respects, the patient appeared normal neurologically.

Routine blood, serologic and urinary studies and roentgenograms of skull and chest revealed no abnormalities.

The results of the Wechsler-Bellevue and the Minnesota Multiphasic tests were considered normal.

Electroencephalographic recordings (14 leads) made during the wakeful state and Seconal-induced sleep revealed 9-10/sec. occipital alpha; very infrequent left temporal 6/sec. activity, when awake and in light drowsiness; during sleep, bilateral 14/sec. spindles of normal type and mixed slow waves. Once, on spontaneous arousal, 6/sec. positive spikes appeared bilaterally in the parietal and posterior temporal areas. The recording was considered abnormal, with minor slow-wave activity during wakefulness in the left temporal region and bilaterally sharp positive activity during sleep.

Electromyography with coaxial electrodes placed in agonistic and antagonistic muscles most commonly revealed violations of Sherrington's law of reciprocal innervation during myoclonic movements (Fig. 1).

It was decided that bilateral ansotomy should be undertaken in 2 stages at intervals of 3 to 6 months and that the first should be performed to alleviate the movements of the more severely afflicted left side.

1st Operation—Jan. 2, 1957. The writer's intention was to perform a right transventricular...
ansotomy. During local procaine (1 per cent) anesthesia, small right verticofrontal scalp, bone and dural flaps were reflected. At this time the anesthesiologist reported that the patient’s abnormal movements, up to then fully in evidence, had ceased spontaneously. The patient remained alert, responsive and clear.

Acting on the postulate that the abeyance of hyperkinesia would be but transient, the operator continued, making an incision approximately 4 cm. long through the cortex of the anterior portion of the “premotor” region, parallel to and about 2 cm. from the midsagittal line. The incision was deeply developed through the corpus callosum. The anterior horn of the right lateral ventricle was entered and the septum pellucidum, septal vein, caudate head, fornix, choroid plexus and foramen of Monro were visualized. The accessible cerebrospinal fluid was aspirated. A suitable site for incision into the ependyma of the lateral wall of the 3rd ventricle (through which access might be had to the ansa lenticularis as it leaves the medial segment of the globus pallidus) was identified.

Although the patient had remained fully awake, the myoclonic movements did not recur. Attempts to evoke them by having the patient inhale amphetamine hydrochloride were unrewarding and after a delay of 50 minutes it was decided to discontinue or at least defer operation on the ground that, were the abnormal movements to prove enduringly abolished by what had been done thus far, the role of definitive ansotomy would be impossible to evaluate.

Course. During the evening of the day of operation, the myoclonic movements reappeared in part and on the next day they were fully evident, after which they continued with their usual vigor. Recovery from operation was unremarkable.

2nd Operation—Jan. 7, 1957. During general endotracheal anesthesia the scalp, bone and dural flaps made at the first procedure were reflected and access to the 3rd ventricle was again accomplished. No blood clots were encountered. A moderate degree of tissue edema was met and, in order to facilitate exposure of the lateral wall of the 3rd ventricle (preliminary to implementing ansotomy), 0.5 cm. of the fornix was resected at the anterior margin of the foramen of Monro. A cordotomy knife was used to perforate the ependyma of the wall of the 3rd ventricle at the site up to that time used by the writer for introduction of a specially designed leucotome to accomplish ansotomy (Figs. 2 and 3). Section of the ansa was then executed caudal to the anterior commissure.

Postoperative course was uneventful. The abnormal movements of the left side proved gratifyingly reduced. The patient was discharged from the hospital on the 8th postoperative day. Within 2 weeks he resumed work at his service station. The residual hyperkinesia in the limbs of the left side diminished steadily and now was virtually absent. In addition, the abnormal movements of the head, neck and trunk, although still conspicuous, had become perceptibly diminished in frequency, extent and vigor. Myelons of the limbs on the right side persisted much as it had been prior to operation.

Three months following right ansotomy it was timidly ventured by the patient and his wife that, since operation, the patient had experienced a reduction in libido and potency. During the first 3 weeks of the patient’s convalescence at home they had not been greatly concerned over the change in their sexual life, but as time passed they had noted not appreciable improvement and now asked whether a permanent deficit might eventuate. More specifically, the patient no longer exhibited his customary initiative in seeking coitus and spontaneous erections did not occur. When the wife assumed the initiative, her manipulations induced penile erections, but the latter were not turgid and usually proved too short-lived to permit successful intercourse. Ejaculation had been accomplished only twice in 3 months, in each instance with less-than-gratifying orgasm for the partners. The patient was instructed to take methyl testosterone propionate 10 mg., 3 times daily.

Six months following right ansotomy the patient and his wife reiterated their satisfaction with the alleviation of hyperkinesia. Regarding the sexual deficiency, they reported that, although preoperative capacities had not been fully regained, some improvement in the libido and potency had occurred while the patient was taking testosterone. The supply of the steroid had become depleted some 4 weeks prior to the 6th month’s check-up examination. Since then, the patient had relapsed into sexual ineptitude. Testosterone was re-prescribed.

Nine months following right ansotomy, the partners reported that the relative salutary effect of testosterone had been re-achieved. They now inquired whether comparable relief from the residual myoclonic movements might be in order. Regarding left ansotomy, the writer confronted the patient and his wife with the possibility that
Choroid plexus of lat. & 3rd ventricles

Fornix (p. columnae)

Caudate head

Foramen of Munro

Site of penetration

Anterior commissure

Septal region

Olfactory tubercle

Fig. 2. Dissection of human brain to show the right lateral wall of 3rd ventricle and structures neighboring upon the site of penetration of special leucotome in an obliquely downward and lateral direction, directed at the ansa lenticularis and medial segment of globus pallidus. Compare with Figs. 3 and 4.

A more complete and enduring sexual deficit might eventuate as a sequel. Consideration of this gave them considerable pause; however, in view of the persisting social and economic handicaps imposed by the myoclonic movements, they requested left ansotomy.

The patient was readmitted on Nov. 17, 1957 and a work-up comparable to that preceding the first operation was implemented. The speech and psychologic tests revealed no decrement. Electroencephalography was reported essentially unchanged.

3rd Operation—Nov. 19, 1957. The procedure on the left side consisted of steps similar to those described in connection with the right ansotomy. Leucotomic interruption of the left ansa lenticularis was accomplished. In addition, 0.4 cc. of absolute alcohol was slowly injected into the globus pallidus.

Postoperative course was smooth. Myoclonic phenomena were absent in the axial members and rarely observed in the limbs following operation. When present, they were of negligible vigor and excursion. The patient was discharged on the 19th postoperative day.

A follow-up examination was carried out 2 1/2 months later at which time the patient described himself as "feeling fine." His appetite had been excessive and he had gained 11 pounds over his usual preoperative weight of 160 pounds. Extreme satisfaction was expressed at the bilateral alleviation of hyperkinesia which the partners considered as virtually complete. The patient had begun to take an active part in the management of his business. Speech, judgment and memory functions were considered unimpaired. The neurological findings were normal except for a twitch of barely perceptible amplitude which occasionally involved the right hand. Eupraxia, evaluated by the safety-pin test, proved well preserved bilaterally, as were the automatic associated acts involved in walking. The gains were recorded at 125 lbs. on the right, and 120 on the left side, comparing favorably with the preoperative recordings. Alibido and, despite seductive manipulations, complete impotence now prevailed. Methyl testosterone again was prescribed.

At the next examination, 7 1/2 months following the 3rd operation, the gains in regard to hyperkinesia were confirmed. No abnormal movements of any kind were observed.

The patient has been followed at intervals of 4 to 6 months for the 4 1/2 years since the right ansotomy and 3 1/2 years since that on the left. Abnormal movements have remained consistently virtually completely absent; however the states of alibido and impotence continue, uninfluenced by testosterone.

On May 20, 1959, electroencephalography was reported upon by Dr. John Knott as follows: "There are 9 1/2–10 1/2/sec. alpha waves of medium voltage over the occipital regions; scattered
Fig. 3. Coronal section of portion of right hemisphere of human brain seen from in front showing the envisioned site of lesion(s) (shaded area) produced by special leucotome to interrupt the ansa lenticularis and medial segment of the globus pallidus. Compare with Figs. 2 and 4.

Fig. 4. Identification of structures related anatomically to the site of lesion(s) depicted in Fig. 3.
medium-voltage theta waves with several short diffuse bursts of 4-6 sec. activity; during Secobarbital-induced drowsiness, some diffuse medium-voltage slow activity, sometimes sharp. Impression: abnormal record with scattered slow waves but without the previous lateralization.” Electroencephalography was repeated on Nov. 12, 1959 with similar results but “showing some improvement.”

On Feb. 1, 1961, the right testis measured 1.8 cm. and the left, 1.9 cm. in the transverse diameters and approximately 3.5 cm. each along their longitudinal axes. The secondary sexual characteristics appeared grossly preserved, with perhaps a slight increase in the size of the breasts consistent with the slow addition of fat.

Repeated 24-hour urine specimens revealed the 17-ketosteroids ranging between 10.9 and 13.3 mg./gm. urinary creatinine. The normal range is between 6 and 10 mg./gm. The urinary gonadotrophins were found absent on one occasion but thereafter were present in aliquots of urine corresponding to 0.5 gm. creatinine. Normally, they are detectable in aliquots of 0.25 to 0.5 gm. creatinine.1-5,18,19

Comment. In this instance, myoclonus was abolished bilaterally. The role conceived to have been played by the surgical procedure in achieving this result necessarily is conditioned by the view adopted regarding the etiology of the disorder. If the disorder be conceived to have been “organic,” the operation may be supposed to have been “specific” in the sense that it interrupted a complicated neuro-anatomic circuit (the ansa lenticularis and/or closely neighboring structures), the integrity of which appears to have constituted a sine qua non of the perpetuation of the hyperkinesia; if, however, the disorder be conceived as “psychogenic,” the effect of operation must be regarded as “nonspecific,” i.e., as having been achieved through auto-suggestion or other placebo effect. Inasmuch as these issues are important for the present case, the other 2 cases in this series and myoclonic disorders in general, they merit some comment.

In summarizing the evidence bearing upon the etiology and pathology of myoclonus, Herz and Meyers5 listed the various loci and histopathologic types of lesion described in the literature. As no validated instance of a “psychogenic” case was encountered, no mention was made of a psychogenic etiology. Nevertheless, contentions favoring such have been raised from time to time, particularly in cases in which, like the present, the history discloses no evidence of a seemingly responsible antecedent and/or current “organic” disease. It is noteworthy that early in the present century parkinsonism, chorea and athetosis were regarded by many authorities as psychoneuroses.

Against a psychogenic interpretation of Case 1, the following appear to constitute cogent arguments: (1) the negative results of psychodynamic and psychometric inquiries and of psychotherapy employed in the patient’s behalf; (2) the abnormal preoperative electroencephalographic findings; (3) the failure of appearance of even transient placebo effects following several courses of physical and pharmaceutical treatment; (4) the failure of the first “sham” operation (which, in virtue of the “spontaneous” cessation of abnormal movements, might be supposed inadvertently to have been endowed with a high subjective potential for therapeutic suggestion) to induce alleviation of myoclonus for more than a few hours; (5) the incomplete character of the clinical improvement following the second operation (right ansotomy); (6) the promptly salutary and protracted effects of the second and third operations, at which bilateral interruption of the pallido-pallidofugal elements presumably was accomplished; and (7) the frequency with which other hyperkinesias (e.g., alternating tremors, dystonia, ballism etc.), currently regarded as unmistakably “organic” disorders, have been alleviated by pallidectomy, pallidofugal-fiber section, lesions in the tegmental fields of Forel and ventrolateral thalamotomy.

As to the alterations in libido and potency in Case 1, it would be difficult to interpret the data in any manner other than to infer that the operative lesions were responsible. A number of circumstances not ordinarily combined in patients subjected to ansotomy compelled perception of relationships that had up to this time gone unheeded. First, the patient was a male. Leaving aside for the moment the rather nebulous definitions of
libido, the criteria available in the male for evaluating sexual performance are more "operational," hence, more scientifically dependable, than those in the female. Second, the patient was a relatively young, married adult, well within the period of vigorous sexual activity. Third, reasonably dependable estimates of the patient's preoperative sexual performances (as judged in terms of frequency and the subjectively gratifying aspects of coitus) could be drawn and compared with reports of his postoperative status. Fourth, these estimates were verified from a second source—the patient's wife. Fifth, the partners overcame a culturally conditioned reluctance to discuss this "delicate" matter with their surgeon. Sixth, spontaneous penile erections were asserted by the patient to have occurred following the "sham" operation. Seventh, following right anotomy, sexual capacity was reduced perceptibly. Eighth, the deficiency was to a moderate degree offset by the use of testosterone. Lastly, promptly following bilateral homologous anotomy, the patient's sexual deficiencies became and have since remained complete.

Unfortunately, preoperative studies of urinary steroids were not made. However, the early and late postoperative urinary 17-ketosteroids (end-products of adrenocortical hormonal activity) were recorded as slightly above normal range, thus indicating that the production of ACTH by the pituitary gland was largely unimpaired. This observation warrants the inference that, whatever the sites of lesions produced incidental to bilateral pallido-pallidofugal section, the latter did not involve much, if any, of the posterior and/or anterior diencephalic neural mechanisms currently thought responsible for inciting the pituitary to secrete ACTH. The presence of urinary gonadotrophins warrants a similar inference. The gonadotrophins in Case 1 were absent on one occasion, but, inasmuch as experiences with repeated assays in any normal subject disclose occasional "negative" specimens, this one finding may be discounted. Previous animal experiments and studies of human eunuchs have indicated that the absence of urinary gonadotrophins does not per se induce libido or impotence in the male.

In brief, then, the tentative inference may be drawn that in the human male the neuroanatomic correlates of ACTH and gonadotrophins are located—at least in major part—at some remove from the relatively small septofornicohypothalamic region which neighbors upon the posterior margin of the more medial portions of the anterior commissure and the pallido-pallidofugal complex and which appears related to libido and penile potency. Pending acquisition of the brain for histologic study, one is obliged to be content with the surgically derived anatomic clues now at hand.

Case 2. T.M., a widowed stevedore aged 44 years, was first examined by the writer on Sept. 17, 1949. His chief complaint was of painless, irregular, vigorously executed but short-lived involuntary movements of the head, neck, trunk and all limbs. The disorder had begun some 4 years before and had slowly progressed in intensity and extent. Late in the third year, sporadic, involuntary vocalizations had appeared. These phenomena were absent during sleep and mild alcoholic intoxication and were intensified by emotional excitation. They impaired the patient's ability to carry heavy objects and interfered with his obtaining work.

Pharmacologic agents, including the barbiturates, bromides, vitamins and solanaceous alkaloids, had been employed without benefit. A psychiatric consultant gave the patient "clearance."

The patient had had an attack of influenza at the age of 31 years which had confined him to bed for 2 weeks under a physician's care. Delirium, headaches, hypersomnia and diplopia were denied. The patient had fathered two sons, the older of which was in good health and the younger of which was said to have died of "spinal meningitis" during preschool years.

Examination. Neurologically, the most conspicuous findings consisted of unpredictable, lightning-like twitches and jerkings of somatic musculature which resulted in grimaces, bizarre truncal movements and more or less extensive excursions of the various appendages. Motor power was unimpaired and no muscular atrophy was discernible. The special senses and the deep and superficial reflexes were considered within normal limits. The Romberg tests were negative, but tests of nonequilibrium coordination revealed impaired functions of limbs during the unexpected appearance of involuntary movements. Speech was normal, but from time to time vocalizations
sounding like "zzning-uh" were produced.

1st Operation—Sept., 20, 1955. During local procaine hydrochloride (1 per cent) anesthesia, the patient remained alert and cooperative. Scalp, cranial and dural flaps were reflected over the right posterior frontal regions of the brain. Right pallido-pallidofugal section was accomplished as described in Case 1, whereupon hyperkinesias of the limbs of the left side ceased abruptly. Those of the right side continued unaltered. No paresis or ataxia was observed. Truncal, m Unified and facial hyperkinesia persisted to a degree, but were perceptibly reduced in frequency, amplitude and vigor.

Postoperative period was uneventful and the patient was discharged on the 12th day following right ansotomy. Check-up studies at the 2nd and 3rd months after operation revealed a sustained reduction in the abnormal facial, meral and truncal movements and an all but complete alleviation of the myoclonic movements of the limbs of the left side. Involuntary vocalizations persisted and the hyperkinesia of the limbs of the right side appeared essentially unchanged.

2nd Operation—Dec. 14, 1955. A left verticofrontal craniotomy was accomplished during local procaine anesthesia. The patient became moderately drowsy soon after the anterior horn of the left lateral ventricle was entered, incidental to which his myoclonia subsided. Ansotomy nevertheless was executed. While the bone flap was being wired in place, the patient wakened and, to the considerable satisfaction of himself as well as the operating staff, abnormal movements were no longer in evidence.

Case. The patient became lethargic during the night following operation. Although usually arousable, he remained so for the next 3 days and thereafter was relatively apathetic for about a week, during which time he was incontinent. The vital signs remained near normal. Beginning at the 10th postoperative day, the patient went on to an uncomplicated convalescence.

By the 3rd postoperative month he had found occasional employment. Except for mild occult movements, felt, rather than seen, by the patient in the muscles of the lateral wall of the abdomen and between the shoulder blades, the myoclonus was abolished. Considerable satisfaction was expressed by the patient at this and the next 2 follow-up visits.

At the end of the 1st year the patient remarked that the one disappointing sequel to operation was that he no longer felt sexually stirred and that spontaneous erections had not occurred. Inquiry revealed that, prior to operation, he had sought the "services" of women on an average of once a month to 6 weeks; and that, since operation, no such activity had been sought because he felt that to initiate such was likely to prove un rewarding.

The patient has been regularly examined and/or in contact with the writer at intervals of from 6 to 12 months during the 5 years since operation. The hyperkinesia as yet remains in abeyance, but so do the patient’s libidinous impulses. Penile erection cannot be induced.

Comment. As in Case 1, bilateral ansotomy in Case 2 was followed by salutary results. The comments made following Case 1 bearing upon the etiology of the myoclonus apply similarly here except that in this instance the arguments against a psychogenic interpretation cannot be spelled out with the same confidence as in Case 1. However, in the absence of postoperative development of neurotic manifestations (which, had they developed, might have been regarded as a "displacement" of unresolved psychologic conflicts to other somatic modes of expression than hyperkinesia), the enduring alleviation of myoclonus provides presumptive evidence that the surgical procedure was definitive in a pathophysiologic sense, rather than merely autosuggestive.

As to the postoperative development of alibido and impotence, it is of interest that the patient did not advance information pertinent thereto in the form of a complaint; he appeared ready to laugh off the postoperative sequelae with remarks to the effect that he guessed he was "just getting old." Neither he nor his surgeon met the facts "head on" as they might have been prompted to do had the patient been younger, as was the circumstance in Case 1.

Case 3. K.T., an 18-year-old American-born farmhand of Japanese parentage, complaining of uncontrollable movements of the face, neck, trunk and all limbs, was first examined by the writer on Dec. 3, 1956. The disorder had first affected the muscles of the throat during the patient’s 6th year. Subsequently it had slowly increased in vigor and extent. The abnormal movements were absent during sleep and augmented by emotional changes. During the execution of voluntary acts they were somewhat suppressed. Nevertheless they interfered with eurypractic performances. Speech usually was normal, but, as the faciobuccolingual articulators and the diaphragm were hyperkinetic sporadically, speech at times was dysrhythmic and dysarthric and audible grunts, sounding like "us-s-s-s" or "jus-s-s-s," were produced frequently.
Numerous drug and physical therapies had proven ineffectual.

Past history revealed that the periods of gestation and infancy had been uneventful. During his 4th year the patient had sustained a head injury, following which he was unconscious for less than 5 minutes. For the 2 years prior to examination, vision had been blurry in the right eye.

On Nov. 30, 1956 "full drainage" pneumoencephalography (125 ml.) had been performed by the referring neurosurgeon, Dr. Jess T. Schwidde of Billings, Montana. Roentgenograms revealed no shift, enlargement or distortion of the ventricles and the distribution of subarachnoid gas appeared within normal limits. The spinal fluid was reported clear and contained 21 lymphocytes per c.mm. and 20 mg. per cent total protein. The Kahn and colloidal gold tests were negative.

The patient was admitted to the University Hospital on Jan. 11, 1957.

Examination revealed severe, nonpatterned, rapidly executed hyperkinesia of varying and unpredictable vigor and excursion involving the axial and appendicular musculature. An early posterior subcapsular cataract was found on the right side. Visual acuity was 20/50 O.D. and 20/40 O.S. without correction and proved correctable to 20/25 O.U. Kaiser-Fleischer rings were absent. No other neurological abnormalities were disclosed. The grips were recorded at 60 lbs. each.

Blood sugar, urea nitrogen, creatinine, cholesterol, plasma protein-bound iodine and alkaline phosphatase were recorded at normal levels. Electroencephalograms on Jan. 12, 1957, during wakeful and sleeping conditions, were interpreted as normal.

The preoperative Wechsler-Bellevue verbal-test score was 79 and was considered a valid index. Because of the severe hyperkinesia, performance tests could not be implemented.

1st Operation—Jan. 17, 1957. During general endotracheal anesthesia, right verticofrontal scalp, bone and dural flaps were reflected. The entire convexity of the exposed hemisphere revealed a dense, milky-appearing arachnoid. A biopsy specimen was taken. A presumed right pallido-pallidofugal interruption was accomplished, much as in Cases 1 and 2. The patient withstood the operation well.

Microscopic sections of the leptomeningeal neo-membrane and adjacent cortex revealed dense fibrous tissue in which were discernible many focal collections of chronic inflammatory cells. The membrane varied in thickness and vascularity from part to part. A single, small psammoma body was present.

Postoperative course was uneventful. An appreciable reduction in all abnormal movements, considerably more apparent on the left than on the right side, was observed throughout the remainder of the period of hospitalization. The patient was able to execute smoothly the safety-pin test of eupraxia with the unaided left hand—a performance of which he had been incapable prior to operation. He was discharged on the 8th postoperative day.

Rechecked at intervals of 2 to 3 months by Drs. Schwidde and Carl Baumann, further recession in the abnormal movements of the left side and a continued reduction of those of the axial and right appendicular structures were reported. At the end of the 1st year, no new neurologic or psychologic deficits had developed.

Because of extensive development and installation of a stereotactically controlled ultrasonic irradiator, the patient's admission for the 2nd stage of operation was delayed until March 3, 1959. At this time, the gains described by his local physicians were verified. The clinical and laboratory studies comparable to those implemented previously gave similar results. Except for the residual hyperkinesia, the neurological findings were essentially normal. Involuntary vocalization persisted. Electroencephalography was again reported as normal. The performance subtests of the Wechsler-Bellevue Intelligence Scale could now be taken in their entirety and, with the verbal subtests, revealed a full scale I.Q. of 72. The Archimedes spiral and Benton visual retention tests and the Minnesota Multiphasic Personality Inventory revealed no abnormalities.

2nd Operation—March 9, 1959. During local procaine (1 per cent) anesthesia, fixation of the head in the stereotactic head holder was accomplished; ventriculography (in which 4 cc. of 25 per cent thorium dioxide was employed as contrast medium) was performed; and the stereotactic coordinates pertinent to the cerebral target areas intended for irradiation with ultrasound were determined mathematically.

3rd Operation—March 16, 1959. During local anesthesia, the patient's head was re-engaged in the head holder and the position assumed on March 9 was duplicated precisely. A left central craniectomy was performed. Ultrasonic irradiation was directed at the following structures: on the right side, Forel's fields, H1 and H2, the tegmental tissues bordering on the lateral half of the dorsal (medial) aspect of the substantia nigra and the medial segment of the globus pallidus; on the left side, Forel's field H2 and the tegmental tissues bordering on the lateral half of dorsal (medial) aspect of the substantia nigra. As a result, a small but perceptible reduction in the abnormal movements was achieved, but the latter were in no sense abolished. Dural incisions revealed a left frontal leptomeningeal opacity similar in extent to that seen on the right side.

Postoperative course was characterized by 5 days of lethargy followed by slowly receding
periods of apathy, confusion and dysarthria. Before the patient's discharge on April 11, 1959, considerable improvement occurred and he was able to render effective "orderly" services to fellow patients. The abnormal movements of the face, neck, trunk and limbs of the right side were considered only slightly improved. Those of the left side were considered much improved.

Interim follow-up studies have been made by Drs. Schwidde and Baumann. At the most recent examinations (Dec. 1, 1960 and Jan. 6, 1961) the improvement in hyperkinesia described previously had been sustained. However, certain un-}

toward signs by now had appeared: lentine opacities had developed bilaterally to a degree that impaired vision and prompted recommendation for lentile surgery; and the patient, now aged 23 years, had worked briefly at 3 menial jobs but had shown no inclination to continue working. He had taken to going on "beer sprees" and had a few "scrapes" with the law. His ne'er-do-well behavior was ascribed to the "beatnik" influence.

On Oct. 7, 1959 a full-scale I.Q. score of 71 was obtained and projection tests revealed the patient to be infantile and somewhat depressed. However, no notable disorder of thought was disclosed. The psychologist contended that the patient could, if he would, perform satisfactorily at a routine type of job.

Current neurological findings include visual acuity 20/40 (corrected) bilaterally and normal visual fields. Speech is slightly slurred. The right grip measures 70 and the left 50 lbs. Alternate rates of motion are normal bilaterally. The tendon reflexes are more brisk on the right than the left, but "upper motor neuron signs" are absent. Hopping is done well on the right, but performed awkwardly on the left. The left lower limb is awkward. The hyperkinesia continues markedly reduced on the left, but elsewhere is present to a troublesome degree.

Inquiry on Jan. 6, 1961 indicated that the patient's sexual desires for the opposite sex were strong and unaltered as compared to his preoperative status. The patient acknowledged frequent spontaneous and self-induced erections and gratifying orgasms by masturbation (episodes of which occur on the average of once every 2 days) and successful coitus on 2 occasions.

Comment. In view of the early age of onset of the hyperkinesia in this case and the grossly abnormal leptomeninges disclosed at two operations, there appears little reason to doubt the existence of an "organic" disorder.

The patient's most conspicuous improvement followed the first transventricular operation, in which the leucotome was directed at the pallido-pallidofugal complex and necessarily penetrated the septoforni-cohypothalamic region. The degree of improvement that followed ultrasonic surgery was, by comparison, slight. The irradiation presumably effected lesions in pallidotegmental structures which were nonhomologous to those of the 1st operation, in that the septofornicohypothalamic structures were spared. In this important respect, the bilateral operation implemented in Case 3 differed from those in Cases 1 and 2.

DISCUSSION

The cerebral structures presumed to have been the anatomic sites of the topically homologous, surgically induced lesions on the right and left sides in Cases 1 and 2 are depicted in Figs. 3 and 4. All 4 procedures were implemented by the transventricular approach to the pallido-pallidofugal target devised in 1939 and used regularly by the writer\textsuperscript{11-13} until November 1957, when a high-frequency, ultrasonic irradiator focussed under stereotactic control\textsuperscript{7,11,16,17} was first employed. The surgical lesions unquestionably involved, in addition to the intended pallido-pallidofugal target, the dorsomedial hypothalamic nucleus, the adjacent fornix and perifornical gray matter, the posterior septal region and the posterior-inferior aspect of the anterior commissure.

In Case 3, the lesion on the right was accomplished similarly by transventricular approach and presumably involved the same structures. However, the 2nd procedure was implemented farther "downstream" by ultrasound and, presumably, involved the tegmental field of Forel and the dorsomedial margin of the substantia nigra.

Whereas in Cases 1 and 2 the myoclonus was abolished completely and so remains, now \(3 \frac{1}{2}\) and \(5 \frac{1}{2}\) years respectively since operation, in Case 3 only a reduction in the myoclonus, more notable in the left limbs than in the right, was accomplished. It appears plausible to suppose that the reasons for the incomplete alleviation of myoclonus and the preservation of libido and potency in Case 3 inhere in the nonhomologous character of his bilateral lesions.
The effectiveness of ansotomy in alleviating myoclonic movements appears comparable to that reported in other hyperkinesias, e.g., parkinsonian tremor, dystonia, intention tremor and ballism. In view of this circumstance, it should be recognized in regard to all surgical procedures employed for the relief of abnormal movements and/or rigidity that the lesions are not directed at the histopathologic processes responsible for the various hyperkinetic disorders (whatever such may be), but at interruption of an as yet incompletely comprehended neural circuit, the ongoing integrity of which, exerting its otherwise normal influence upon a deranged system of checks and balances, has the effect of augmenting the neuropathic dysequilibrium and thus rendering clinically overt that which might otherwise remain a latent hyperkinesia.

The development of postoperative alibido and impotence in male patients J. D. (Case 1) and T. M. (Case 2) and in at least 1 earlier male patient, a sufferer from parkinsonism, and the development of postoperative alibido in 1 earlier female patient, also a sufferer from parkinsonism—all 4 of whom had undergone bilateral homologous ansotomy—suggests that the neural mechanisms in the human corresponding to these two sexual functions are, at least in part, located at and/or traverse the relatively narrow neural region depicted in Figs. 2, 3 and 4.

Additional force is lent to this suggestion by the studies on the male squirrel monkey reported by MacLean et al. in 1959. During general anesthesia, the bare tips of fine, depth electrodes were implanted under stereotactic control (a) in the "medial preoptic region, close to the septum" and (b) "a few millimeters caudally." These sites were described as lying between Horsley-Clarke levels A10 and A6. During the wakeful state, electrical excitation of the former site regularly evoked penile erection after a latent period of between 6 and 7 seconds and stimulation of the latter site evoked erection after a latent period of 3 seconds. The turgidity of the erection could be increased by simultaneous summation of stimuli at both sites. Some evidence indicated the existence of synaptic connections between the two sites, e.g., hyperpolarization of the more caudal site rendered stimulation of the rostral site ineffective; and electrocoagulation of the caudal site abolished appearance of erections upon stimulation of the rostral site. The authors did not report whether or not the spontaneous appearance of erections and/or courting behavior was altered in consequence of such electrocoagulation.

The human observations appear to indicate that a region closely comparable to the more caudal of the two sites described by MacLean et al. was the seat of destructive lesions in those patients exhibiting postoperative deficits in libido and potency. The neurosurgical procedures unwittingly amounted to reverse counterparts of the studies on the monkey and the data reported constitute the first presumptive evidence pertinent to the physiology and pathophysiology of these aspects of the reproductive function in the human. In addition, there is strong indication that, wherever the central neural mechanisms may be that prompt the pituitary gland to release ACTH and gonadotrophins, they apparently are not located close to the mechanism concerned with penile erection.

Inquiry may properly be raised as to why more instances of compromise of sexual functions than the few cited above have not arisen during the 21 years since pallidopallidofugal section was first performed. In reply, it must be said that, under present circumstances, we are in no position to assert whether and how many additional patients have been thus affected. Until recently, the possibility had not forced itself to the writer's attention, hence, deliberate pre- and postoperative inquiries bearing upon the matter had not been made. However, it would be logically and empirically unsafe to assume that the failure of patients to explicate their observations is tantamount to absence of the sexual deficits here under discussion. For one thing, the age range of patients most commonly subjected to ansotomy happens to correspond to that in which physiological decrements in libido and potency may be expected to appear. This circumstance tends
to mask the phenomenon. Moreover, bilateral ansotomy at homologous sites has not been performed as frequently as unilateral ansotomy and/or the variety of noncorresponding bilateral procedures that have been used in recent years to relieve hyperkinesia. Finally, there is the reluctance on the part of patients, relatives and physicians to discuss such intimate matters. It is, however, clear that, if we are to acquire data in the near future capable of answering the question just raised, this reluctance must be overcome.

CONCLUSIONS

1. Presumptive evidence is cited to indicate that a small region of the adult human brain, namely, the dorsomedial hypothalamic nuclei, the fornix, perifornical gray matter, posterior-inferior septal region and pallido-pallidofugal complex, embraces at least in part central neural mechanisms essential to libido and penile potency.

2. The central neural mechanisms responsible for inciting pituitary excretion of ACTH and gonadotrophins in the human appear not to be located (at least not significantly so) at the site described.

3. The production of surgical lesions in this region, especially when implemented bilaterally and homologously, carries a serious risk of producing deficits in libido and potency.

4. In contemplating surgical procedures in the region referred to, a careful history bearing upon sexual capacities and propensities should be recorded routinely as a baseline against which subsequent developments, both positive and negative, may be measured and evaluated.

5. A candid preoperative discussion of the likelihood of the development of enduring postoperative deficits of these sexual functions should be initiated by the physician for the instruction of the patient and others concerned.

REFERENCES


