THE NEUROLOGICAL ASPECTS OF "MENTAL" SUBNORMALITY PART II[1]

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THE LIMBIC CHILDREN

The Limbic children constitute a clinically recognizable group. They have hyperphagia or omniphagia. Hence they are overweight. They have a disturbance of kinesis, usually hyperkinesis, less often hypokinesis. They explore by touch, even though visual activity is apparently normal. They have oral behaviour. They put everything into their mouths to test it out, to discover its nature. At times, they smell the object. They are the rockers, head or trunk rockers, the bangers, the rhythmic hitters, usually of themselves, and they are also the scratchers. Sometimes they pull out their hair. They have almost never acquired speech.

They frequently show microcephaly; and depression over the posterior fontanelle is often detectable.

Growth is stunted; secondary sexual characteristics are late in development; sexual drives are under-developed; dependence on routine is great; they cling to routine and are disturbed by change; fear is the basic reaction; placidity is present in some cases to a pathological degree. Attention is defective and therefore distractability great. Perseveration is common. They are stimulus-bound.

There is some resemblance to the Kluver and Bucy syndrome described in monkeys. After bilateral temporal lobectomy (including at least part of the hippocampal gyrus, the amygdala and the hippocampus) monkeys show (1) visual agnosia or "psychic blindness"; (2) strong oral tendencies, that is a tendency to examine everything with the mouth; (3) an over-attention and an over-reaction to various stimuli; (4) changes in food habits and increased appetite; (5) marked absence of emotional responses; (6) hypersexual behaviour. The over-attention to visual stimulation and the "oral" tendencies did not change, in some observations, during two years before the animal's death, but the agnosia decreased. The complete absence of emotional response became a marked placidity, and the greediness continued. The mixture of visual agnosia and increase in oral activity causes the animals repeatedly to pick up all objects in their environment. They manipulate each object in a compulsive way, mouth, lick and bite it, and then, unless it is edible, discard it. However, discarded objects are picked up again in a few minutes as if the animal had never seen them before and subjected to the same manipulation and oral exploration. In addition, the animals are easily distracted. They heed every stimulus, whether it is novel or not, and usually approach, explore, manipulate and if possible bite. Failure to ignore peripheral stimuli is termed hypermetamorphosis by Kluver. Terzian and De la Torre (1955), described the Kluver and Bucy syndrome in man after bilateral removal of the temporal lobe.

It is important to note that in this term limbic children, or children with the old brain, we do not envisage a pathological entity in which the phylogenetically

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old cortex is alone functioning (without the restraining or elaborating neo-cortex). Such a view would indeed be simplicist and an over simplification. This group of patients is comparable in a partial form with the Kluver and Bucy syndrome. There are important differences, however, in that rocking, head banging and other repetitive movements are apparently only seen in human pathology. Also the Kluver and Bucy syndrome is often only temporary after operation. In addition, it would be gross oversimplification to envisage the rest of the cortex as functioning normally or near normally in these severely subnormal patients. The term limbic children is introduced in order to focus attention on the functional activity of this region of the brain, and because a striking parallel of behaviour presented itself during observation of these children. We may in certain respects be witnessing the mixed effect of stimulation and ablation experiments, with an additional lack of connections with the neo-cortex.

We have only listed nine patients under the description of limbic children, since the syndrome was often only partial. For example, many patients were docile, hyperphagic and/or stimulus-bound without the other manifestations, and although behaviour in a subnormal population may often suggest visual agnosia special tests should be designed for its demonstration.

CLINICO-PATHOLOGICAL AND CLINICO-PHYSIOLOGICAL CORRELATIONS

In long-term (often life-long) problems of subnormality, clinico-pathological correlation is difficult to obtain. What is more important, it is often irrelevant, for structural lesions can only give partial explanation of neurological deficit. The reasons for implicating function or dysfunction of certain parts of the brain, in particular the limbic system, the amygdala and the hypothalamus, are neuro-physiological and neuro-chemical, and we shall discuss these here.

The discovery that the limbic system and the hypothalamus are intimately concerned not only with emotional expression but with the genesis of emotion has brought the fields of neurophysiology and psychology closer together than ever before.

The limbic lobe or rhinencephalon consists (Crosby, 1962), of the olfactory nerve, bulb and stalk; the anterior olfactory nucleus; the anterior olfactory nucleus; the anterior perforated space; the parolfactory area of Broca; the hippocampus and associated gray; the pre-pyriform cortex and various portions of the hippocampal gyrus; the isthmus of the fornicate gyrus; and the cingulate region. The important relations of the amygdala to various structures in the limbic lobe are well recognized, but anatomically it is not considered part of it, (Crosby, 1962). The limbic system connects with the anterior nucleus of the thalamus, the mammillary body, the interpeduncular nucleus, the medial forebrain bundle, the habenula and the septum.

The limbic cortex is phylogenetically the oldest part of the cerebral cortex. Histologically, it is made up of a primitive type of cortical tissue, the allocortex, surrounding the hilum of the hemisphere, and a second ring of a transitional type of cortex between the allocortex and the rest of the cerebral hemispheres.

The pre-pyriform cortex which in many mammals is a band of grey extending from the base of the olfactory crus towards the temporal part of the fornicate gyrus, in man is represented by small patches of cells. The pre-pyriform area and the peri-amygdaloid cortex are very intimately related with the adjoining area 28, which makes up the major part of the hippocampal gyrus. This area 28 (the entorhinal area) is relatively very much increased in primates and particularly in man. It is a six-layered region, which does not however have the typical layers seen in iso-cortex

but probably represents a transitional form of cortex. Histology indicates that the superficial layers are receptive in character and the fourth and sixth layers are for efferent discharges (Crosby, 1962). It may be that these areas of transitional cortex are more liable to mal-development than others.

A striking feature of the limbic system is its paucity of direct neocortical connections. There are a few fibers from the frontal lobe to adjacent limbic structures, and there are probably some indirect connections via the thalamus. However, the neocortex, to use Nauta's phrase, "sits astride the limbic system like a rider on a horse without reins". Another characteristic of limbic circuits is their prolonged after-discharge following stimulation. Reverberating circuits might be part-basis of perseveration of emotional responses that outlast the stimuli that initiate them, and perhaps also of those that are inappropriate.

The actual extent of the limbic system has changed little as mammals have evolved, but these regions have been overshadowed by the growth of the neocortex in man. The overall impression of behaviour in a subnormal population is a mixture of that of children and certain mammals. The "islands" of adult behaviour are those which in "normal" adults are considered the result of disinhibition. Thus, it is behaviour commonly thought to arise from the "old brain" acting without the restraining influence of the "new brain".

Emotional instability can also be considered a hallmark of mental subnormality; it is, at any rate, almost universal in these patients. A characteristic is the lightning change from expression of one emotion to another. This change is so rapid that it suggests a lack of the true emotion; the change merely involved the expression of the emotion.

The neurophysiological evidence of correlation is gathered from stimulation experiments with implanted electrodes, and to a lesser extent from ablation experiments. Stimulation of various parts of the amygdala in the cat have produced a large number of responses (MacLean and Delgado, 1953; Naquet, 1953; Gastaut et al. 1953). These responses include swallowing, chewing, licking lips, hissing, sniffing, salivation, gagging, biting, dilation of pupils, growling, acceleration or inhibition of respiration, panting, cardiac slowing, ipsilateral closure of eye, ipsilateral retraction of ear, and contralateral adversion of the head, contralateral pawing, adversion of the body, extension of the forelimb, flexion of the hindlimb; awakening, meowing, pricking up of ears, arrest of purring. In summary, actions related to feeding and oral behaviour, actions subserving the expressions of emotion, and movements of the limbs on both sides of the body.

The abnormal feeding habits and hunger or appetite drives of many of the subnormal patients have been described. The hypothesis has been made that rocking and self-injurious involuntary movements may be related to activity in the fornix, the septum, the hypothalamus and the medial forebrain bundle (the latter, however, may be vestigial in man). The special importance of the limbic system in certain neurological studies has been stressed by Pribram (1961) and Weiskrantz (1964).

Although stimulation or lesions of the hypothalamus can influence complex chains of behaviour, these experiments must not be taken to imply that the elements in such chains cannot be independently modified: direct behavioral evidence shows that they can.

While the neural mechanisms responsible are often referred to as situated in centres, it is clear that they are not anatomically localized but diffuse structures. Further the finding that the hypothalamus is important in a given type of behavior must not be taken to imply that responsiveness depends solely on the state of a hypothalamic mechanism.

Hyperphagia in rats can be produced by forebrain lesions as well as lesions in the hypothalamus (Richter and Hawkes, 1939). In sexual behaviour, Beach (1940, 1944) showed that removal of parts of the cerebral cortex resulted in a behaviour deficit which increased with the extent of the cortical damage.

The so-called rage response of cats can be evoked by stimulation in the hypothalamus, amygdala and midbrain, and can be abolished by hypothalamic lesions, and it can also be shown by cats lacking a telencephalon. The normal response, however, depends on complex interrelations between telencephalon and diencephalon. For example while removal of the neo-cortex may result in placidity, ablation of parts of the limbic lobe results in an increase in ferocity. The rage response of hypothalamic cats not only lacks direction, but also has a very low threshold and practically no after-response: unlike normal rage, it ceases almost as soon as the eliciting stimulus is removed (Bard and Mountcastle, 1964). In these aspects, the rage responses of subnormal patients are very similar. The higher levels seem to be concerned with the release and regulation of the response, while the mechanism of expression depends primarily on the hypothalamus (Akert, 1961). Even this, however, may be an oversimplification, because for example, after coagulation of the midbrain and certain parts of the anterior hypothalamus, threat behaviour caused by stimulation of the anterior hypothalamus is altered (Brown and Hunsperger, 1963).

Fear reaction can be produced in conscious animals by stimulation of the hypothalamus and the amygdaloid nuclei. Conversely, fear reaction is absent in situations in which they would normally be evoked when the amygdalae are destroyed. Monkeys are normally terrified by snakes. After bilateral temporal lobectomy, including the amygdalae, the monkeys approach snakes without fear, pick them up and even eat them. Maternal behavior is depressed by lesions of the septal nuclei in animals.

Chemical substances are secreted by neurones. Their concentration varies from place to place within the nervous system and it is logical to think that secretion and concentration levels are abnormal in anatomically abnormal brains. These known or suspected synaptic transmitter agents and "neural hormones" include acetylcholine thought to be secreted in the supraoptic and paraventricular nuclei and the mammillary bodies; norepinephrine, serotonin and histamine in the hypothalamus and anterior pituitary hormone-releasing factors in the median eminence of the hypothalamus. With increasing information of neuro-pharmacology and the site of drug action, correlation with subnormal behaviour will become more realistic.

Discussion

Study of the lack of intelligence, and observation of behaviour ungoverned by detectable purpose, may lead towards understanding the nature of intelligence and the nature of coherent, intelligible activity. Moving and dwelling among a community of the mentally subnormal is like attempting to steer one's ship through a crowded port in which every sailing ship has lost its rudder. All around is abnormal activity, with either hyperkinesis or pathological immobility. The inhabitants appear to be either in a hurry or stationary.

This confusion and apparent lack of directive acts as a challenge. To the presumed intact nervous system, the challenge is to comprehend the under- or over-responsiveness of the subnormal nervous system.

According to Julian Huxley, "the process of evolution involves the constant inter-adjustment of organisms with their environment". If we consider the individual as being in evolution from the embryo to the grave, we may describe certain

organisms as arrested at certain stages. We may also try to delineate levels of nervous system function, in the light of present neurophysiology. These levels of functioning do not seem linked to aetiology.

At the lowest level of evolution, organisms are not in contact with their environment. The first stimuli to which they respond are touch, food and warmth. The patients tend to be flexed, to assume the fetal position, to lie curled on the floor in a ball. They make constant movements of the arms and legs. There are two classes of movements. (1) Rhythmic, repetitive stereotyped, purposeless movements. These often inflict self-injuries, and include trunk rocking, head rocking, head banging, arm banging, flaying movements of the upper limbs, striking of the shins with the opposite heel. Many of these patients have bald patches on the head from friction. They frequently pace up and down the room or out of doors, covering the same territory, or they rock from one foot to the other, (this can also occur as a drug effect, with the thiazines). (2) Non-rhythmic, slower, writhing movements of the arms or legs. These movements appear semi-exploratory of the environment. In these patients, speech had never developed, and they are non-communicable in all spheres. Their behavior is oral; they suck fingers and clothes. Size and shape of the skull is variable, but microcephaly is common and there is very frequently a saucer-shaped depression over the posterior fontanelle. They are usually hyperkinetic, less often hypokinetic.

Next in the evolutionary scale are the stimulus-bound. These patients react to all incoming stimuli without discrimination. There is a poverty of central nervous system connections in that their reactions are predictable. They are unable to adapt, therefore "conservative" in their habits; they are scared of the unfamiliar because lacking in equipment necessary to deal with new situations.

There is no inner life, no inner speech, no consecutiveness of thought or activity because continually interrupted by incoming stimuli. Their behaviour appears as a series of habituated responses. These responses can be extinguished. It is as though the "switch-on and switch-off" mechanisms were lacking. This of course may be related to a faulty reticular activating system, and a widely dispersed property. The "switch-off" mechanism seems particularly deficient, perhaps because it is more sophisticated in its make-up. If learning from the environment consists in "closure" of neuronal connections so that all except the relevant ones are blocked, this may explain some aspects of subnormality.

These patients have difficulty in learning how to say "no". This distinction can easily be made on neurological testing. The I.Q. can be gauged according to whether the patient recognizes when the tuning fork does NOT vibrate. Other things being equal, rough assessment shows that with an I.Q. above 45 the patient can appreciate the "on" of vibration applied to the limbs, and with an I.Q. above 60 the "off" as well.

Next in the evolutionary scale are those with disturbance mainly of the motor system, suggesting maximal abnormality of the cerebral hemispheres and/or the cerebellum. Tonus is abnormal, with either hyper-or hypotonus. Movements may be either slow, spastic, involving the distal portions of the limbs, or lacking in co-ordination with cerebellar dysfunction. One can tell at a glance that these patients are not "rockers", and that they do not have hyperphagia. Their speech may be dysarthric or dysphasic, but it has at least developed. In some instances they may prefer gesture, but they are never non-communicative.

Higher in the evolutionary scale (because implying that the machinery for self-expression is relatively intact), we come to the study of behaviour. There are two methods of describing behaviour (Hinde, 1966). One involves reference to the

strength, degree and patterning of muscular contraction (or glandular or other physiological activity). The other refers not to these changes, but to their consequences.

The first method is often limited to patterns of limb or body movement, because a complete description would be too detailed and cumbersome, and some of these terms of behaviour are included in the previous section. Terms such as past-pointing, overshoot, tendon reflexes, salivation or sleeping posture are of this type.

The second method Hinde called "descriptive by consequence". "Picking-up work material", "dressing and undressing", or "approaching" do not refer to particular patterns of muscular contraction, but "cover all patterns which lead (or could lead, and there is room for ambiguity here) to the specified result. Descriptions such as these are apropriate when the behaviour involves orientation to objects in the environment, and when the motor patterns, although leading to a constant result, are themselves diverse. Thus they tend to be used especially for appetitive or searching behavior".

There are two advantages in this second kind of description. First, a brief description may cover a multitude of motor patterns each of which in itself is variable—for instance "approaching" covers walking, running, hopping, sidling, motor tics, involuntary movements, genuflexion, backward approach, avoidance of gaze, etc.; we thus have a shorthand.

The second advantage is that these descriptions often call attention to essential features of the behavior that may not appear in "physical" descriptions, such as orientation with respect to the environment, or responsiveness to external stimuli. Behaviour may change radically (e.g. from one type of appetitive activity to another) with little change in the motor pattern: or vice versa. a radical change in the patterns of muscular contraction (e.g. from walking to running) may be, for some purposes, of little significance.

Neurologists have in the past employed the term "apraxia" to describe inappropriately directed motor activity, a type of meaningless, insightless activity. Apraxia, however, does not seem to include what we frequently observe in a subnormal population. Here there is often a mixture of ungainliness amounting to gross motor inefficiency (this ungainliness arises from minor physical disabilities), associated with a lack of understanding of the act; and in addition an inability to compare the specific present action with past actions of a similar nature, due to failure of memory regulating activity pari passu with the carrying out of the act. Thus many subnormal patients start doing something correctly and in a meaningful, insightful way, so that one is not justified in terming this as apraxia. The activity then begins to peter out, the patient turns to the examiner as if for reinforcement and guidance having lost the train of action, and the subsequent behaviour or approach to the problem takes on the characteristics of apraxia. The patient handles the object as if he were seeing it for the first time.

This may be related to pathological lack of a "neuronal model".

When a novel stimulus is presented to a normal animal it shows an "orientation response": its responsiveness to the stimulus is enhanced and its readiness for action increased. The further elaboration of this response depends upon the consequences. If the stimulus is without correlates, the response wanes. The properties of this waning can be accounted for on the supposition that the animal forms a "neuronal model" of familiar sources of stimulation: non-coincidence between the stimulation received and the neuronal model leads to an orientation response.

As neurological knowledge increases, both further breakdown and further cohesive generalities in the study of behaviour will become possible.

THE NEUROLOGICAL ASPECTS OF "MENTAL" SUBNORMALITY

Summary

1. One hundred and eleven patients classified as "mentally" subnormal and requiring long-term institutional care have been studied from the neurological and behaviour point of view.

2. The inadequacy of our present state of knowledge and of our present methods of examination are revealed by current diagnoses such as "progressive cerebral deterioration" or "disturbed behaviour".

3. Since the functioning of the nervous system determines the behaviour of the organism, the neurological aspect cannot be divorced from the behavioural.

This is particularly important in these cases in which long-term observation is of value, in which pathology must often await a lifetime and autopsy studies do not necessarily reveal the diagnosis, let alone provide satisfactory clinico-pathological correlation.

4. A plea is therefore made for a new approach, beginning with clinical analysis, unprejudiced in that when the patient's mode of life, as a whole or in detail, does not fit into previously recognized categories, then new patterns are described.

5. The limbic children or children with the old brain is discussed as a neurophysiological concept and a working hypothesis, recognizing that this is a physiological or functional approach awaiting pathological basis.

6. At the resent time, comparative studies in animal behaviour are often revealing, not that results can be transferred from one species to another, but that methods in animal behaviour study could profitably be introduced into this field of specialized neurology.

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References

Akert, K. (1961). In D. E. Sheer, (Ed.). *Electrical Stimulation of the Brain.* Austin, Texas. University of Texas Press for the Hogg Foundation for Mental Health.

Bard, P. and Mountcastle, V. B. (1964). Some Forebrain Mechanisms Involved in Expression of Rage with Special Reference to Suppression of Angry Behaviour. *Res. Pub. Ass. Nerv. Ment. Dis.* 27: 362-404.

Barlow, H. B. (1963). In W. H. Thorpe and O. L. Zangwill. *Current Problems in Animal Behaviour.* Cambridge, England. Cambridge University Press, pp. 331.

Bastock, M. and Blest, A. D. (1958). An Analysis of Behaviour Sequences in Automeris Aurantiaca Weym (Lepidoptera). *Behaviour* 12: 243-284.

Beach, F. A. (1940). Effects of Cortical Lesions Upon the Capulatory Behaviour of Male Rats. *J. Comp. Psychol.* 29: 193-245.

Beach, F. A. (1944). Relative Effects of Androgen Upon the Mating Behaviour of Male Rats Subjected to Forebrain Injury or Castration. *J. Exp. Zool.* 97: 249-295.

Brown, J. L. and Hunsperger, R. W. (1963). Neurothology and the Motivation of Agonistic Behaviour. *Animal Behav.* 11: 439-448.

Bullock, T. H. (1961). The Origins of Patterned Nervous Discharge. *Behaviour.* 17: 48-59.

Crosby, E. (1962). In *Correlative Anatomy of the Nervous System.* New York. Macmillan.

Delgado, J. M. R. (1962). Lea and Febiger. *First Hahnemann Sym. on Psycholsom, Med.,* Philadelphia, Pennsylvania, pp. 221.

Fischer-Williams, M. and Bickford, R. G. (1965). A New Multipurpose Brain Depth Probe: Long-Term Implantation and Electroencephalographic Recording in Man, Monkey and Cat. *Mayo Clin. Proc.* 40: 791-804.

Gastaut, H., Naquet, R., Vigouroux, R., Roger, A. and Badier, M. (1953). Etude Electrographique chez l'homme et chez l'animal Des Décharges Epileptiques Dites "Psychomotrices". *Rev. Neurol.* 88: 310-354.

Graff, H. and Stellar, E. (1962). Hyperphagia, Obesity, and Finickiness. *J. Comp. Physiol. Psychol.* 55: 418-424.

Grossman, S. P. (1960). Eating or Drinking Elicited by Direct Adrenergic or Cholinergic Stimulation of Hypothalamus. *Science.* 132: 301-302.

Hebb, D. O. and Thompson, W. A. (1954). In G. Lindzey. *Handbook of Social Psychology.* Reading, Massachusetts. Addison-Welsey.

Hinde, R. A. (1966). In *Animal Behaviour. A Synthesis of Ethology and Comparative Psychology.* London. McGraw-Hill Book Company.

Hogan, J. A. (1965). An Experimental Study of Conflict and Fear: An Analysis of Behaviour of Young Chicks Toward a Mealworm. I. The Behaviour of Chicks which do not Eat the Mealworm. *Behaviour.* 25: 45-97.

Lind, H. (1959). The Activation of an Instinct Caused by a Transitional Action. *Behaviour.* 14: 123-135.

Lyon, G. Personal Communication.

MacLean, P. D. and Delgado, J. M. R. (1953). Electrical and Chemical Stimulation of Frontotemporal Portion of Limbic System in Waking Animal. *Electroencephalogy and Clin. Neurophysiology.* 5: 91-100.

Montgomery, K. C. (1955). The Relation Between Fear Induced by Novel Stimulation and Exploratory Drive. *J. Comp. Physiol. Psychol.* 48: 254-260.

Myers, R. D. (1964). *First Int. Symp. on Thirst. Florida State Univ.,* Oxford, England. Pergamon Press. pp. 533.

Naquet, R. (1953). Thesis, University d'Aix—Marseille, Marseille, France.

Noirot, E. (1965). Changes in Responsiveness to Young in the Adult Mouse: III. The Effect of Immediately Preceding Performances. *Behaviour.* 24: 318-325.

Olds, J. (1958). *Neurological Basis of Behaviour. Ciba Found. Symp.,* S. E. W. Wolstenholme and C. M. O'Connor. London, England: Churchill pp. 124-141.

Pribram, K. H. (1961). In D. E. Sheer. *Electrical Stimulation of the Brain,* Austin, Texas. Univ. of Texas Press for the Hogg Foundation for Mental Health.

Richter, C. P. and Hawkes, C. D. (1939). Increased Spontaneous Activity and Food Intake Produced in Rats by Removal of Frontal Poles of Brain. *J. Neurol. Psychiat.* 2: 231-242.

Sherrington, C. S. (1960). *The Integrative Action of the Nervous System.* Cambridge, England. Cambridge Univ. Press, (republished in 1947, pp. 252).

Sherrington, C. S. (1917). Reflexes Elicitable in the Cat from Pinna Vibrissae and Jaws. *J. Physiol.* 51: 404-431.

Stellar, E. (1954). The Physiology of Motivation. *Psychol. Rev.* 61: 5-22.

Stellar, E. (1960). In J. Field, H. W. Magoun and V. E. Hall (Ed.) Amer. *Handbook of Physiology,* Section 1, Vol. III, Physiol. Soc. Washington D.C. pp. 1501.

Terzian, H. and De la Torre, G. (1955). Syndrome of Kluver and Bucy. Reproduced in Man by Bilateral Removal of Temporal Lobes. *Neurology* (Minneap.). 5: 373-380.

Weiskrantz, L. (1964). Neurological Studies and Animal Behaviour. *Brit. Med. Bull.* 20: 49-53.