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MORBI NEURALES

AN ATTEMPT TO APPLY A KEY PRINCIPLE TO THE DIFFERENTIATION OF
THE MAJOR GROUPS *

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This paper is intended to be more modest than even its subtitle "attempt" denotes. The writers feel confident that a key principle, roughly reminding one of the principle of Gray's botany for the diagnosis of plants, is applicable to the diagnosis of nervous diseases. Central in the application of such a key principle is the idea of exclusion of diseases in diagnostic consideration by means of following a certain order. But granting the theoretical virtues of a key principle and of the method of orderly diagnosis by exclusion, we must remain uncertain whether the order adopted is the *one best way*, in the sense of the efficiency engineer who devised that term.

The attempt to apply a key principle to the differentiation of the major groups of nervous diseases was based on the comparative success of the application of an identical principle to the differentiation of the major groups of mental diseases. Since the publication of a key to the practical grouping of mental diseases, presented to the American Neurological Association in 1917, the principle of diagnosis per exclusionem in ordine has been presented to the Association of American Physicians in 1918, and in briefer form in a second paper on certain applications of the pragmatic method to psychiatry (1918). The idea of a possible extension of this principle of orderly exclusion to the problems of general medicine has met with the approval of Dr. Richard C. Cabot, who, at the June, 1919, meeting of the Association of American Physicians, displayed a tentative list (without definitive sequence) of the major groups of diseases in general. This list, as the sequel will show, may have some value for clinical neurology, and may even prove superior to the key to mental diseases formerly laid down.

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DIAGNOSIS BY EXCLUSION

Perhaps it is superfluous to argue here for the virtues of the key principle in the form of diagnosis per exclusionem in ordine. In a previous paper considerable argument has been devoted to the habitual underestimation of the values of the method of diagnosis by exclusion, of which DaCosta and his followers were guilty in the sixties and seventies of the last century. The method of diagnosis by exclusion was to be resorted to only in extremis, according to these authors, and especially according to the book on diagnosis by Da Costa, published in 1864, which was thought by some to have marked an epoch in the progress of internal medicine. The so-called indirect diagnosis or diagnosis by exclusion was, according to DaCosta, "not on ordinary occasions much employed, nor indeed," DaCosta continues, "is it to be recommended." DaCosta thought that to prove what a thing is by proving all that it is not is a very tedious process. He went on to say that it was "difficult to think of all the possibilities" and remarked that "pathology was in an imperfect state." But its imperfections seem to us to interfere with all other methods of diagnosis as much as with diagnosis by exclusion. As for the difficulty of thinking of all the possibilities, the device of placing these possibilities in manageable groups (after the manner of the diagnostic groups of animals, plants and minerals, that is, after the manner of Linnaeus), DaCosta and his colleagues in the single-volume textbooks of medicine did not conceive of such a process. Apparently, DaCosta felt that in the process of diagnosis by exclusion one would need to exclude theoretically every other disease than the one on which one would finally fix. As for the tediousness of this process of diagnosis, no doubt accurate diagnosis in difficult cases is and must remain extraordinarily tedious, but remains none the less interesting and important.

But diagnosis by exclusion is not diagnosis by exclusion in order. It is one thing to secure a list; it is another thing to make a sequence of the elements in the list. An aggregate does not forthwith throw itself into order, and especially not into that arithmetical arrangement called a "well-ordered" aggregate.

Now as a matter of fact, it seems to us that most workers in difficult cases do actually employ something like what has been described as diagnosis per exclusionem in ordine. In correspondence with some specialists, affirmative replies have been given to questions on this line. Ordinarily, diagnosticians are prouder, perhaps, of making a diagnosis of smallpox by the sense of smell at 100 yards (more or less) than to make a careful, logical diagnosis by exclusion in order. Yet the most expert diagnosticians are reduced to using diagnosis per exclusionem

from time to time. Even DaCosta admitted that the method had to be used on occasion. We hold that a method which has to be resorted to in difficult cases ought to be rationalized so that tyros and inexperienced workers may the earlier learn to make correct diagnoses. We hold that the field of mental and nervous diseases is a good field in which to begin, because the majority of cases in these fields are difficult of diagnosis. Especially in the field of mental diseases is it true that there is practically no single pathognomonic symptom in the whole branch. Starting with a slant toward some disease in which a given symptom is characteristic, the tyro can prove from the very best textbook that almost any mental case he has in hand is or might well be a case of the disease he starts to prove it to be. The tyro is only caught up short when the laboratory comes to the rescue with pathognomonic signs. But the tyro with a slant rushes in where the laboratory expert fears to tread. On this account, as there are practically no indicator symptoms in the field of mental disease, the method of diagnosis per exclusionem in ordine was found to have especial value for the mental diseases.

We shall not here rehearse the situation in the mental disease groups. We may persist in pointing out, however, that, if eleven groups of mental disease have been laid down as of pragmatic orderly value in diagnosis, there is no reason why some other worker may not prove that there are twelve or ten instead of eleven groups. Thus one acute observer believes that there should be added to the eleven-group system, as presented, a twelfth group of paranoic cases. Another acute observer suggests that there are really no senile diseases, that properly obtain the name, at least among the mental diseases, and that consequently the senescent-senile group should be deleted from the sequence and its diseases distributed otherwise; for example, among the arteriosclerotic brain diseases, the manic depressive group, etc. Let us insist, too, that not only may the groups be accordeonized, that is, interpolated and extrapolated without destroying the key principle involved, but the order of the groups may likewise be changed, possibly to advantage. Some considerations on this point have been given in the paper above mentioned on applications of the pragmatic method to psychiatry.

CLASSIFICATION OF DISEASES

For those who are willing to consider a rationalization of the method of diagnosis by exclusion in order (a method which we are convinced that every one dealing with complex cases must in some form employ) there is another bogey of greater difficulty. Pathology, as DaCosta might still remark, is in an imperfect state! Why classify

diseases in any definite manner until we know more about the etiology and genesis of diseases? The answer to this objection is that in practical life some account of stock must continually be taken. Next year the very basis of some well-recognized disease of today may be entirely overthrown; even today we may recognize that the etiology or genetic basis of that particular disease is very shaky or rickety. Nevertheless, it remains our duty to make diagnoses on the best basis available; otherwise there will be no common ground of discussion with colleagues and no common ground for practical measures of treatment. Accordingly, the imperfections of present day pathology should not deter us from practical classifications. We must cheerfully admit that these practical classifications may be changed from decade to decade. The pessimist or the worker weary in well-doing, may regret the long string of obsolete and obsolescent classifications that the world provides, but there is no genuine ground for such regret where the classifications have been made in good faith by careful and logical thinkers. Some authors appear to think that there will some day be a single classification of diseases, and every day we see classifications put forward by eager workers which they seem fondly to wish shall replace all other classifications. A glance into logical works on the topic of classification, even into the ancient work of John Stuart Mill called a *System of Logic*, would reveal to these workers the infinitude of classifications of the very same objects. A classification by causes may totally differ from a classification by genesis, and both may differ from a classification for the purposes of diagnosis, and in the end a classification by methods of treatment may be found to cut quite across all other classifications. Then, too, it is probably worth while to devote a sentence to the universal occurrence of the editorial imbecile (or shall we say idiot?) who makes fun of what he calls the classifying mania, apparently not being aware that almost the whole development of science and art depends on this so-called mania.

The etiologist may have in mind some particular factors which he would like to elevate into the so-called causes of disease. At the other end of the line some hospital administrator will be found who would like to classify all cases according to the amount of food, nursing and open plumbing which they may require. In between these two extremes may be found the practical diagnostician who has troubles of his own. The present effort deals with the troubles of the diagnostician rather than with the troubles of the etiologist or of the hospital administrator.

With more particular respect to the nervous diseases themselves is it not possible that the nervous diseases will very shortly fall into a common group with the mental diseases? Is there any point in producing an order of nervous diseases independently of an order of

mental diseases? Probably the new art of neuropsychiatry is on its way, and neuropsychiatrists with equally good training on both sides will become available. However, we believe that it is the pragmatic situation at the present time that psychiatrists and clinical neurologists are two separate and different kinds of specialists. No doubt the fundamental basis of both arts lies in the science of neuropathology, taken in its widest sense of including both structural and functional concepts—but that wide and deep science is still (to borrow a leaf out of DaCosta's vocabulary) in an imperfect state. The question is a practical one. It is something like the question whether we ought to separate the specialties of dermatology and syphilis or the specialties of surgery and gynecology. Now one opinion, now another, will prevail, and there will be a progress toward a final decision.

Classification of Nervous Diseases.—As we approach the topic of the nervous diseases taken in their general sense, we find that the distinction between "organic" and "functional" nervous diseases is the most fundamental one in many systematic works and in perhaps the majority of nonspecialistic medical minds. Is a case organic or is it functional?—this is about as far as the general practitioner likes to wander in the field of clinical neurology. He has an idea that by means of the "reflexes" the neurologist more or less effectively arrives at a diagnosis of organic nervous disease and is often somewhat astonished that the neurologist cannot, by doubling or trebling the number of "reflexes" taken, somehow arrive at a diagnosis doubly or trebly accurate. In our view, this no doubt perfectly true distinction between organic and functional diseases is of little diagnostic value in the individual case, and this is aside from any doubts we may have as to the definition of the term "functional."

We examined the nervous diseases in sundry systematic works and seemed to be able to put these diseases into five groups as follows:

Tentative sequence of major groups of nervous diseases due to

1. Infection.
2. Historrhesis.
3. Neuronatroph.
4. Imbalance.
5. Special.

Application of Classification in Diagnosis by Exclusion.—Another word as to its application by the tyro or by the general practitioner. Let us suppose the general practitioner confronted by a paralysis. Suppose in the course of his thinking he lights on the hypothesis that it is due to "neuritis." He may then consider or look up in the books

all the types of neuritis which he can find and look for signs or symptoms appropriate to one or the other of these groups. Now, of course, the diagnosis of neuritis may in itself be erroneous to start with, and the inquiry as initiated may become useless. According to our ideas, it would be much more to the advantage of the patient if the diagnostician proceed somewhat as follows: Here we have a case of paralysis of a limb! In the first place, is this paralysis at all related to infection? Are there any signs of (1) infection whether in temperature, in blood, or otherwise, in this patient? What tests should be employed to secure evidence on the point? In the second place, if the case shows no sign of infection let us turn to the question of injury or other lesion of whatever sort which may have affected the tissues from without. Even if we cannot absolutely exclude infection as a matter of fact, let us for the purposes of diagnosis exclude it as a matter of argument until the rest of our tests have become available. Excluding infection, is there any evidence in the situation of a condition to which we above gave the name (2) *historrhesis*? *Historrhesis* might include the severing of a nerve or the nerve tissues by a knife or a bullet. It would include the blocking of nerves and the by-effects of *historrhesis*. For diagnostic purposes it would include the effects of an apoplexy not only as directly due to the destruction of brain tissue, but also to the pressure on collateral tissues. Not only trauma and arteriosclerosis, but also tumors, would be included in this concept of *historrhesis*, from which for our purposes the orderly exclusion would have already excluded infection. The *historrhesis* inquiry sums up to an inquiry for trauma or other tissue-injuring lesions of noninfectious nature and of a focal character.

Thirdly, the diagnostician, having got rid of the hypothesis of (1) infection (including, of course, neurosyphilis) and of trauma and other forms of (2) *historrhesis*, would arrive, according to our scheme, to the group which we have termed (3) the *neuronatrophies*. Our first thought was to name these the "scleroses," since the group contains the various classic scleroses, like lateral sclerosis, multiple sclerosis, etc. But, on the whole, the primary fact appears to be that of *neuronatrophy*, a condition not of the general nature of *historrhesis*, but of a much finer and more differentiated type. With the infections and the common *historrhesis* out of the diagnostic way, the tyro would now come to the *neuronatrophies*, that is, to questions of combined system disease, syringomyelia, multiple sclerosis, etc. (The fact that, e. g., syringomyelia often results in *historrhesis* with secondary degeneration, should not bother the diagnostician a whit. He has already considered the *historrhesis* and eliminated perhaps a transverse myelitis from the field of diagnosis. He does not arrive primarily at

the question of syringomyelia, but only after having eliminated the effects of historrhexis [pressure myelitis] and infections [Pott's disease] which should be discriminated from syringomyelia.) (Again, some one might prove that multiple sclerosis is infectious. But the infectious group of nervous diseases, excluding some peripheral forms, is essentially a group of diseases in which pleocytosis of the spinal fluid is characteristic diagnostically. Multiple sclerosis appears to be in general outside this group. For the present pragmatic purposes multiple sclerosis should evidently fall among the classic neuron-atrophies rather than among the historrhexes or infections of the nervous system.)

Fourthly, if the diagnostician has been able to eliminate (1) infections, (2) rhexes of tissues, and (3) differential atrophies of neuron system, he will arrive at a somewhat nondescript group which we above termed (4) imbalance, under which the drug-neuritis cases, sundry metabolic disorders, the endocrinopathies and sympathetic system disorders fall. A great many of these conditions are conditions of what we might term tissue-imbalance. Imbalance is particularly well shown in the endocrine and in the various sympathetic disorders, and may also be shown in the metabolic group in some sense. Whether multiple neuritis should not sometimes be grouped among the infections and sometimes among metabolic conditions is a question which we do not care to take up. Our purpose is not to add to theories concerning causation or genesis, but merely to take the diseases as they stand in the systematic works and throw them into an effective diagnostic order for the purposes of orderly exclusion.

Fifthly, over and above the (1) infections, (2) historrhexes, (3) neuronatrophies and (4) imbalance groups we have (5) a special or miscellaneous group in the field of nervous diseases, including such conditions as the various aglias, migraines, tics, spasms and vertigo, to say nothing of myopathies, osteopathies, various tissue atrophies, unexplained hydrocephalus, etc. With respect to this last group, the modesty of the clinical neurologists must forbid their complaining about how little they know. The aim of the present classification of nervous diseases is not to display the ego of clinical neurologists, but to find a method for arriving at diagnoses in a more logical manner.

Comments on This System.—We conceive that a system like that of classification into these five groups will serve to clarify many problems in the tyro's mind. The expert may not need it. We suspect that the expert is already using some such method, perhaps one private to himself which he has never thought of betraying to ordinary man. In conversation with various diagnosticians we find that they habitually use such systematic works as Oppenheim in a manner

peculiar to themselves. Diseases in a book like Oppenheim's are worked over precisely as if they were in no important order whatever. We do not deny that Oppenheim's excellent work arranges the diseases in an order on a certain basis. We deny that the order adopted has much to do with practical diagnosis. Practical diagnosis in clinical neurology appears to have been chiefly a topical diagnosis devised to find, in the brain or spinal cord, the seat of lesions chiefly of the group we have above termed *historrhexes*. To adopt a more general medical attitude such as that defined in these five groups is a plan not found in textbooks of clinical neurology. We suspect, however, that the more mature the clinical neurologists become, the more they arrive at some such practical method for diagnosis by exclusion in order.

We have no idea of insisting that the order we here present is a final order, that it is an order which forbids interpolation or extrapolation, or that it is an order that cannot be changed to advantage. We would like to have the topic considered carefully by those who are concerned, and we are of the opinion that some uniformity of the scheme of elimination for practical diagnostic purposes for the various nervous diseases will greatly benefit the treatment of patients. If, by the naked eye, one has somehow guessed that a patient has a nervous disease, perhaps by means of the low power of the logical microscope, he will come to one of the five groups above mentioned as the logical site of the disease in question. Suppose now that he turns on the somewhat higher powers of the logical microscope. The five groups are subdivided somewhat as follows:

SUBDIVISION OF FIVE GROUPS OF NERVOUS DISEASES

1. Infections	
Treponema	Parasites
Bacteria	Unknown infections
2. <i>Historrhexes</i> , focal destruction (non-infectious)	
Trauma	Heightened intracranial
Vascular	pressure
3. Neuronatrophies, sclerosis ("classical degenerations")	
Systemic sclerosis	Hereditary sclerosis
Diffuse sclerosis	
4. Imbalance exo(neuro)genous	
Drug	Endocrine
Metabolic	Sympathetic
5. Miscellaneous	
Algas, migraine, vertigo	Tissue atrophies
Tics, spasms	Hydrocephalus
Myopathy	<i>Et al.</i>
Osteoarthropathy	

SYNOPSIS

1. INFECTION

(Diagnosis by signs of infection, pleocytosis of liquor, etc.)

<i>Treponema</i>	<i>Bacteria</i>	<i>Unknown</i>
Tabes	Epidemic meningitis	Poliomyelitis
Pachymeningitis cervicales	Tuberculous meningitis	Hydrophobia
hypertrophica	Streptococcus meningitis	Landry's disease
General paresis	Neuritis	Herpes zoster
	Tetanus	Chorea minor
	Myelitis	
	Encephalomyelitis	
	Abscess	

2. HISTORRHESIS (FOCAL DESTRUCTION)

(Diagnosis by reflex-signs, heightened intracranial pressure, etc.)

<i>Trauma</i>	<i>Vascular</i>	<i>Tumor</i>
Caisson	Little's disease	Recklinghausen's disease
	Hematomyelia	
	Anemia	
	Hyperemia	
	Hemorrhage	
	Softening	
	Hemorrhagic encephalitis	
	Polioencephalitis, acute	
	hemorrhagic, superior	
	Sinus thrombosis	
	Aneurysm	
	Pseudobulbar palsy?	

3. NEURONATROPHY

(Diagnosis by characteristic picture and history)

<i>Systemic Sclerosis</i>	<i>Diffuse Sclerosis</i>	<i>Hereditary Sclerosis</i>
Lateral sclerosis	Multiple sclerosis	Friedreich's ataxia
Combined system	Pseudosclerosis?	Amaurotic idiocy
Amyotrophic lateral sclerosis	Gliositis spinalis	Familial infantile paralysis
Primary muscular atrophy	Syringomyelia	Primary muscular atrophy, familial form (bulbar form)
Senile paraplegia	Paralysis agitans	Huntington's chorea

4. IMBALANCE

(Diagnosis by characteristic picture)

<i>Drug *</i>	<i>Metabolic Disorder</i>	<i>Endocrinopathy</i>	<i>Sympathetic</i>
Neuritis	Neuritis	Spasmophilia	Acroparesthesia
Multiple neuritis	Diabetic, senile, etc.	Tetany	Angioneurotic edema
			Myxedema intermittent
		Exophthalmic goiter	Hydrops articularum
		Cretinism	Raynaud's disease
		Infantilism	Erythromelalgia
		Acromegaly	Scleroderma

* It is possible that these cases should be placed under historrhesis, although many of the phenomena are reversible.

5. MISCELLANEOUS

<i>Algeas</i>	<i>Migraine</i>	<i>Vertigo</i>	<i>Tics—Spasms</i> Occupation-neuroses Tic convulsive
<i>Myopathy</i>	<i>Osteoarthropathy</i>	<i>Tissue Atrophies</i>	
Primary myopathy	Vertebral disease	Hemiatrophy facialis	
Dystrophy, muscular progressive	Luxation—fracture	Hemihypertrophy facialis	
Thomsen's disease	Caries		
Myasthenia gravis	Tremors		
Paramyoclonus	Syphilitic		
	Arthritis deformans		
	Osteomalaceous paralysis		
<i>Hydrocephalus</i>	<i>Undefined</i>		
Meningitis serosa	Caisson		
	Anomalies		
	Reflex paralysis		
	Exhaustion paralysis		
	Periodic paralysis		

SUMMARY

The method of diagnosis by orderly exclusion, already proposed for use in the diagnosis of mental diseases, is probably of equal value in the field of nervous diseases. The writers have endeavored to gather the main types of nervous disease into a comparatively small number of groups for successive consideration by the tyro, or even by the expert, in diagnostic elimination. Experts may prefer a different order from the one proposed; but it is unlikely that any neurologist fails to use, consciously or unconsciously, some form of orderly diagnosis.

Yet the student is quite likely to be taught that the best procedure is to pick out some striking symptom in a case under consideration, and to follow that symptom back to textbook models for a suggestion as to the entity involved. He then endeavors to match the data of the case in hand with the possibilities laid down in the textbook.

We think that it is much more desirable to take the general situation in the body at large into account, and to include, if possible, first, the hypothesis of an infectious origin for the symptoms. Secondly, we try to exclude, if possible, the effects of coarse and otherwise destructive lesions of the nervous system (historrhexas) namely, in general such conditions as show no signs of infection, but exhibit reflex disorders and signs of heightened intracranial pressure and the like, suggestive of focal lesion. Thirdly, we come to the hypothesis of the existence of one or other of those classic degenerations with which the neurologist is familiar. If infection, historrhaxis, and classic degenerations can be excluded, we then proceed, fourthly, to the

hypothesis of some kind of imbalance, perhaps metabolic or endocrine or sympathetic. If the diagnosis cannot be made on these lines, possibly the condition belongs in some miscellaneous and otherwise undefined or highly specialized group.

Even when the disease seems to be limited to a peripheral nerve, we consider that then the successive hypotheses of (1) infection, (2) historrhesis, (3) specialized neuronatrophy, (4) imbalance, can be preferably considered in that order. We think that by the pursuit of some such method as this the neurologist can bring his work better into line with that of general medicine. The method is, moreover, a very pragmatic method, since lines of treatment are specially indicated for the different great groups of disorders. But in so difficult a field we do not wish to dogmatize, and shall be content if our communication arouses interest in the application of the key or order principle to the diagnosis of nervous diseases.