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FURTHER OBSERVATIONS ON NEUROLOGICAL ABNORMALITIES IN MENTAL DEFECTIVES

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IN 1933 we published a survey of the neurological findings in 500 defectives, in which paper, as in the present one, the object was to discover possible variations from normal in reflexes and other neurological phenomena in those whose nervous systems might be developmentally imperfect. From both these studies it transpires that there is a considerable variation from normal in these findings, quite apart from the occurrence of clinically recognizable syndromes. In the present paper subjective factors such as observations on sensations have been omitted, since they are open to the criticism that the information obtained from defectives may not be dependable. The present investigation has therefore been confined to purely objective manifestations of the motor performance and reflex findings in mental defectives, with the object of determining the functional capacity of their motor apparatus.

The material consisted of 336 unselected mental defectives of both sexes and all ages. These were divided into the following grades: feeble-minded, 85 (25 per cent.); imbeciles, 177 (53 per cent.); and idiots 74 (22 per cent.).

In observing a group of mental defectives one of the most striking peculiarities to be noticed is the poor posture and apparent general slackness of muscular tone which these individuals present. That this may be rather an expression of poor general co-ordination than of any real alteration of muscle tone is suggested by the observation that when instructed in drill and deportment according to their capacities, many defectives assume a general bodily posture which is indistinguishable from that of the normal individual.

In order to test this general impression an attempt was made to determine if many of these persons showed a disturbance of muscular tone which could be described as pathological. For this purpose tone was classified as : (1) normal, (2) plastic, (3) spastic, (4) spastic with contractures, (5) contracture without spasticity, (6) catatonic, and (7) flaccid. Table I shows the results of this investigation.

TABLE I

	TOTALS	NORMAL	PLASTIC	SPASTIC	SPASTIC + CONTRAC- TURES	CONTRAC- TURES — SPASTICITY	CATA- TONIC	FLACCID
Feeble- minded	85	78 (92%)	1 (1%)	1 (1%)	1 (1%)	3 (3%)	1 (1%)	0
Imbeciles	177	141 (80%)	6 (3%)	11 (6%)	8 (5%)	5 (3%)	1 (1%)	5 (3%)
Idiots ..	74	48 (65%)	1 (1%)	14 (18%)	1 (1%)	0	0	10 (14%)
Total ..	336	267 (79%)	8 (2%)	26 (8%)	10 (3%)	8 (2%)	2 (1%)	15 (4%)

The number of cases presenting abnormal muscle tone is thus seen to be relatively small, viz. 21 per cent., and of these 12 per cent. of the whole group is made up of the spastic cases of hemiplegia, tetraplegia, diplegia, and paraplegia. Thus only 9 per cent. of those not presenting clinical syndromes show abnormalities of muscle tone.

The association of variation in tone with signs of pyramidal system dysfunction as exemplified by the following criteria was investigated : exaggeration of the biceps, wrist, or ankle-jerks; knee-jerks with positive Strumpell's sign (i.e. obtained off tibia) ; patellar or ankle clonus ; dorsiflexion of great toe in response to stroking of front of tibia; extensor plantar response.

TABLE II

	TOTALS	NORMAL	PLASTIC	SPASTIC	SPASTIC + CONTRAC- TURES	CONTRAC- TURES — SPASTICITY	CATA- TONIC	FLACCID
Feeble- minded	15	12 (80%)	0	0	1 (7%)	1 (7%)	1 (7%)	0
Imbeciles	72	49 (68%)	3 (4%)	9 (12%)	7 (10%)	4 (6%)	0	0
Idiots ..	30	17 (57%)	1 (3%)	10 (33%)	1 (3%)	0	0	1 (3%)

From these tables it is obvious that alterations in tone may occur without definite evidence of permanent recognizable lesions in the nervous system. It is of interest that in the group of the feeble-minded, abnormalities of tone are infrequent, while in the group of idiots such abnormalities are common. It is possible that such variations of tone in some cases depend more on a diffuse cortical inferiority with resultant defect of co-ordination, integration and inhibition than on localized lesions.

Disturbances of general co-ordination were obviously prevalent, and a study of these was next undertaken. In the previous study the prevalence of

choreiform movements amongst mental defectives had been noted in 25 per cent. of a total of 500. In the present series evidence of hyperkinesis was found as shown in Table III. This is clearly more prevalent than in normal individuals of the same age, and is a further indication that in mental defectives cortical control is likely to be poor. As to the choreiform movements, none were of a gross nature, resembling rather what is found in a mild or residual case of Sydenham's chorea. No correlation with reflex abnormalities was found. There was, however, a tendency for hyperkinesis to be associated more frequently with hypotonia than with hypertonia.

TABLE III

	TOTALS	TREMOR	CHOREA	ATHETOSIS	TICS	TOTAL HYPERKINESIS
Feeble-minded ..	85	10 (12%)	34 (40%)	0	0	44 (52%)
Imbeciles	177	17 (10%)	92 (52%)	15 (8%)	1 (0.5%)	125 (70.5%)
Idiots	74	4 (5%)	32 (43%)	3 (4%)	0	39 (52%)

The "cuisse-tronc" sign of Babinski, which depends upon faulty synergia of the back muscles, especially of the glutei, as shown when the subject lying on his back raises his trunk off the ground without the assistance of his hands, was also studied. A positive "cuisse-tronc" sign was indicated by the raising of the legs off the ground on sitting up. Babinski suggested that this occurred especially in the presence of lesions of the pyramidal tract. The results are shown in Table IV.

TABLE IV

	TOTAL	POSITIVE "CUISSÉ-TRONC" SIGN
Feeble-minded ..	85	19 (22%)
Imbeciles	177	55 (31%)
Idiots	74	47 (63%)

This type of inco-ordination was more manifest the lower the grade of defective, which may be explicable by the relatively low efficiency of their nervous system as a whole. Again, it was found that this sign of inco-ordination was by no means confined to cases in which the pyramidal tracts or cells were specially involved. It occurred in an appreciable number of cases whose reflex anomalies suggested a definite muscular hypotonia. The presence of this sign may therefore be taken as a manifestation of a general inco-ordination of the neuro-muscular apparatus rather than of any single localized lesion in the nervous system.

The following tests of cerebellar function (Tilney and Riley, 1928) were then made the subject of study: (1) disturbance of walking and turning; (2) Romberg's test; (3) past pointing test (hands and feet); (4) diadokokinesia; (5) rebound test; (6) leg raising. It must be realized that too much reliance could not be placed on results of such tests in defectives, as some

individuals had difficulty in understanding instructions. Taking the total errors of co-ordination found in relation to these tests Table V indicates the findings. It is clear that there is an inferiority on the part of low-grade subjects in their ability to perform such tests. When this is correlated with reflex variations results are obtained as shown in Table VI.

TABLE V

	TOTAL	CASES SHOWING SOME FAILURE IN CEREBELLAR TESTS
Feeble-minded ..	85	35 (41%)
Imbeciles	177	115 (65%)
Idiots	74	70 (95%)

TABLE VI

	TOTAL	WITH PYRAMIDAL SIGNS	WITH DIMINISHED REFLEXES	WITH NORMAL REFLEXES
Feeble-minded ..	35	7 (20%)	5 (14%)	23 (66%)
Imbeciles	115	50 (43%)	19 (17%)	46 (40%)
Idiots	70	29 (41%)	14 (20%)	27 (9%)

It will be seen, therefore, that more than half of the subjects with signs usually attributed to cerebellar dysfunction also show reflex anomalies indicative of disturbance of function in the nervous system either central or peripheral. It is difficult to be sure where the failure lies which determines the inco-ordination. It seems probable that a general irregular failure of the influences impinging on the final motor pathway is present, rather than specific cerebellar dysfunction. This is in accordance with the observed fact that cytoarchitectonic abnormalities in the defective are widespread; and that the most significant change is an irregular arrangement of cells. It is therefore improbable that the functional co-ordination of cortical effector cells, corpus striatum, red nucleus, cerebellum, vestibular apparatus, and spinal motor centres is really effectual in these cases. A gross defect in function on both sides of the arc is suggested by our previous observations of remarkably poor performance tests in spatial discrimination.

In the previous study we had included reflex anomalies in our general survey of neurological behaviour. In the present series the following reflexes were systematically examined: biceps-jerk, wrist-jerk, knee-jerk, ankle-jerk, plantar reflex, Oppenheim's reflex, and the presence of knee and ankle clonus.

Dividing the cases into (1) those who showed reflex changes suggesting anomalies in the pyramidal system, (2) those with definitely diminished reflexes, and (3) those with reflex responses within the normal range, the distribution is found in Table VII. As might be expected, the lower the grade the less likely were the reflexes to be normal. This was partly accounted for by those cases in the group showing definite clinical syndromes. It is clear, however, that many defectives show reflex anomalies which have no apparent relation to

established clinical syndromes. The question arose as to whether these reflex changes were or were not constant. Eighty unselected cases which had previously shown signs indicative of pyramidal system defect were therefore re-examined after an interval of at least 12 months. Twenty (25 per cent) showed definitely recognizable clinical syndromes of diplegia, hemiplegia, or paraplegia, while 60 (75 per cent.) manifested no such syndromes. In only three cases (3.75 per cent.) had all signs of such defect passed away. In 24 cases (30 per cent.) the findings were in all respects the same. In eight of these 24 there were recognizable syndromes, thus there was no evidence that these conditions had progressed. In the other 16 cases there was no evidence that any such syndromes were present. In 26 cases (32.5 per cent.) the manifestations of pyramidal defect had become more marked. Of these only eight showed definitely recognizable syndromes. It would appear to be somewhat rash to conclude that symptomatically the pathological condition, except in two of the syndrome cases, had definitely advanced. In the remaining 27 cases (33.25 per cent.) the pyramidal signs were altered in their distribution, but so far as could be observed not in any significant way. Of these only four suffered from a definitely recognizable syndrome, and 24 merely showed further variations in the already inconsistent state of their reflex record.

TABLE VII

	TOTAL	WITH PYRAMIDAL SIGNS	WITH DIMINISHED REFLEXES	WITH NORMAL REFLEXES
Feeble-minded	85	15 (18%)	17 (20%)	32 (62%)
Imbeciles	177	72 (41%)	28 (16%)	100 (43%)
Idiots	74	30 (41%)	16 (22%)	46 (33%)
All grades	336	117 (35%)	61 (18%)	178 (47%)

To amplify the neurological findings in mental defectives the organic reflexes were investigated in a similar group of low-grade patients. This group comprised 500 defectives who had a mental age between $1\frac{1}{2}$ and 6 years, as estimated by the Merrill-Palmer test. It will be seen that in the subjects constantly incontinent the chronological age was higher and the mental age lower than that for the whole of the series. Constant double incontinence is seen to be much more frequent in males than in females, and this difference is statistically highly significant, for it exceeds eight times the probable error. The association between a lower mental age and double incontinence is not absolute. Nevertheless on the average the mental age of the subjects with double incontinence is definitely lower. These figures suggest that, amongst defectives, the lower the mental age the greater the tendency to incontinence and that incontinence amongst defectives is commoner in males than females.

Though not established by the present enquiry, it would appear probable that voluntary control over the evacuation of urine and fæces is acquired later by defectives than by normal children. In the cases studied none of the defectives, with one exception, was below the age of 7 years. Above this age there was no reduction in the proportion of constantly incontinent cases as chronological age increased, thus showing that these patients do not "grow

out of it." The low mental age of these defectives, coupled with other available information, suggests that in them cortical development, though much below normal, has in the majority been at least sufficient to enable control to be established over unchecked spinal activity.

TABLE VIII

TOTAL	PERCENTAGE INCONTINENTS	PERCENTAGE CONSTANT DOUBLE INCONTINENCE	MEAN CHRONOLOGICAL AGE	AVERAGE MENTAL AGE (MERRILL PALMER-TEST)	CHRONOLOGICAL AGE OF 62. CONSTANTLY INCONTINENT 52 M. ; 10 F.	MENTAL AGE OF 62. CONSTANTLY INCONTINENT 52 M. ; 10 F.
Males .. 264	48.7	19.8	18.5 yrs.	3.6	19.2	2.8
Females 236	27.5	4.2	20.6 yrs.	3.8	21.5	2.8

In those with double incontinence it is significant that the mental age is low. It is possible that in defectives the sacral segments of the spinal cord may show structural defects. If this be so disturbances might be expected in both the somatic and autonomic reflexes of the parts concerned. To test this an examination of the bulbo-cavernosus and superficial anal somatic reflexes was carried out in 51 of the 52 doubly incontinent males. As a control 51 continent males of as far as possible equivalent mental ages were examined in the same way. The results of this examination are given in Table IX.

TABLE IX

	DOUBLE INCONTINENCE		CONTROLS	
	No.	%	No.	%
Bulbo-cavernosus and anal reflexes present	7	14	22	43
Bulbo-cavernous only	9	18	9	18
Anal only	0	—	4	8
Both reflexes absent	35	69	16	31
	51		51	

There are individuals in both groups with absent sacral somatic reflexes and also individuals in the two groups in whom the reflexes are present. Nevertheless the proportion of individuals in the control group with normal reflexes is significantly greater than the proportion in the incontinent group.

An attempt was made to determine the relation, if any, between the state of these somatic reflexes and the chronological and mental ages of the patients. In order to make this analysis it was necessary to know whether chronological and mental ages are correlated in these two groups. It was found that there was no such correlation, and therefore it was legitimate to examine separately the relation between the state of the reflexes and chronological and mental age respectively. It was found that the mental age of the control group exceeded that of the group with incontinence. This was unavoidable, as there were not enough individuals of low mental age in the former group to make the correspondence exact. In the group with incontinence there was no evidence of any association between the state of the reflexes and either chronological or mental age. In the control group, however, individuals with absent somatic reflexes had a rather lower mental age and a definitely lower chronological age.

The following deductions may be drawn from the results of this part of the examination : (1) The majority of the group with incontinence have some structural abnormality in the sacral segments of the cord which within the age limits investigated is of a permanent character. (2) A smaller proportion of the control group display a similar abnormality of somatic reflexes and the figures suggest that the reflexes may sometimes appear relatively late in life in individuals. (3) In the control group there appears to be some association between absence of reflexes and a specially low mental age. In view of these findings it would appear that in defectives who are incontinent there may be in addition to the cerebral defect a permanent abnormality in the proper development of the sacral segments and that this may stand in causal relationship to the incontinence.

In defectives in whom the sacral segments are efficiently developed so as to allow for proper organization of the reflexes, permanent incontinence is only met with in the presence of markedly imperfect cortical function, while in those cases in which the sacral development is interfered with, relatively higher grades of generalized cortical deficiency may still exhibit permanent complete incontinence.

Summary

An investigation of reflex abnormalities has been made in a group of 336 unselected institutional mental defectives.

Abnormal neurological signs were found more frequently in the lower-grade patients. In the total group abnormal variations in muscle tone were present in 21 per cent., hyperkinesis in 61 per cent., the "cuisse-tronc" sign of Babinski in 36 per cent., inco-ordination in 65 per cent., and signs of pyramidal tract dysfunction in 35 per cent. of cases. The association of these various abnormalities among themselves has been examined.

A re-examination of 80 unselected cases which had shown signs of pyramidal system defect was made after an interval of at least one year. In approximately one-third the signs had become more marked, in one-third the condition appeared stationary, in the remainder the abnormal neurological signs had altered in distribution while in only three cases had all evidence of defect passed away.

An investigation of some of the factors influencing incontinence of urine and faeces has been made in a group of 500 mental defectives. Constant incontinence was found to be associated with low mental age and to be much more frequent in males than in females. In defectives with incontinence the bulbo-cavernosus and superficial anal reflexes were more often absent than in a control group of equivalent mental age. It was concluded that in addition to poor cortical control a permanent abnormality of the sacral spinal segments is often a factor in the establishment of incontinence in mental defectives.

REFERENCES

- Gordon, R. G., Norman, R. M., and Berry, R. J. A. (1933). *J. Neurol. Psychopath.*, **14**, 97.
Gordon, R. G., and Norman, R. M. (1932). *Brit. J. Psychol.*, *I and II*.
Tilney, F., and Riley, H. A. (1923). "The Form and Function of the Central Nervous System," 2nd edition, p. 478.