

A CLINICAL CLASSIFICATION OF THE MENTALLY SUBNORMAL

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Today one sees less use of the terms mental defect, imbecile, idiot and severe subnormality, etc., as a diagnosis, but it still occurs too frequently for comfort. These terms are quasilegal terms that express the degree of the mental handicap and have no use from a diagnostic point of view. It seems that only by careful diagnosis can one hope to advance; only after Folling (5) had reached a clinical method of diagnosis in phenylketonuria were the biochemists able to get on with the job of sorting out the disease. There is little hope, therefore, that we can advance in the field of psychiatry generally and mental subnormality in particular until we can make accurate and exact diagnoses, and this paper is presented since it appears that we must have some sort of classification of disease before we are able to put a particular case in its correct category.

The classification here presented is only one of many possibilities and does not pretend to be complete in every detail but it has been used as a working classification for some time and has been found useful. It is not a complete aetiological classification but is essentially a clinical one. It will be found that many of the categories will have to be changed as our knowledge advances and some of the non-specific genetic diseases will have to be placed in a more exact metabolic group as understanding is reached. The accepted theory of 'one gene one enzyme' means that all genetic disease must be initially a metabolic one however far removed the symptoms may be from the primary gene action.

Some will no doubt complain that there are some exceptions from many of the accepted classifications of the past but this is intentional. Perhaps the most obvious one is the exclusion of epilepsy. I believe that epilepsy is only a symptom of disease and is not a disease per se, in the same way as anaemia is only a symptom of some underlying disorder. One must, however, admit that it may be possible for the treatment to lead to intellectual impairment, but this should then be ascribed to the drugs rather than the epilepsy. Epilepsy must be considered as the physical manifestation of a cerebral dysrhythmia which can be caused by many differing types of lesion, metabolic as in phenylketonuria, etc., or anatomical, to quote but two examples.

THE CLASSIFICATION

1. NON-PATHOLOGICAL MENTAL SUBNORMALITY.
2. PATHOLOGICAL MENTAL SUBNORMALITY.
 - (a) STRUCTURAL ABNORMALITIES OF THE NERVOUS SYSTEM.
 - (i) Abnormalities of the ventricular system.
 - (ii) Abnormalities of the vascular system.
 - (iii) Abnormalities of the brain substance.
 - (iv) Abnormalities due to infection.
 - (v) Abnormalities due to trauma.
 - (b) METABOLIC ABNORMALITIES:
INBORN ERRORS OF METABOLISM.
 - (i) Protein metabolism.
 - (ii) Carbohydrate metabolism.
 - (iii) Fat metabolism.

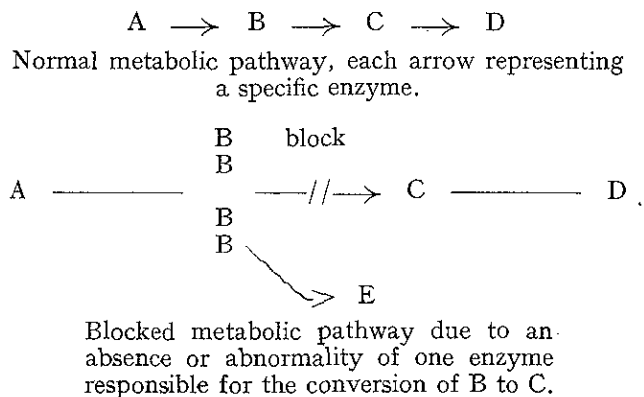
- (iv) Mineral metabolism.
- (v) Other metabolic abnormalities including endocrinopathy.
- (c) CHROMOSOME ABNORMALITIES.
 - (i) Abnormalities of the autosomes.
 - (ii) Abnormalities of the sex chromosomes.
- (d) OTHER GENETIC DISEASES.
 - (i) Dominant.
 - (ii) Recessive.
 - (iii) Sex-linked.
- (e) SECONDARY TO PSYCHIATRIC DISEASE.
 - (i) Psychotic illness.
 - (ii) Neurotic illness.
- (f) DEPRIVATION SYNDROMES.
 - (i) Special sense deprivation.
 - a) Deafness.
 - b) Blindness.
- (g) TOXIC CAUSES.
- (h) UNDIFFERENTIATED.

The first stumbling block in the above classification is the division between the pathological and the non-pathological types of mental subnormality. It is due to the muddling of these groups that the distribution curve of intelligence quotients is so skewed at the lower end of the scale. One would expect to find the low I.Q.s to occur with equal frequency as the high ones, but as is well known they are far more numerous. In this collection of the lower I.Q.s there must be those who have a low level of intelligence by chance, as well as those in whom the low I.Q. is a symptom of some underlying disease, the former being the non-pathological type of case and the latter the pathological. One would also expect that the non-pathological types would be much less frequent the lower the I.Q., and conversely, the nearer to I.Q. 70 the commoner they would be. Fraser Roberts (6) has considered this problem and thinks that the group here called non-pathological are probably genetic and of a multifactorial nature, but even so they appear to be the lower end or tail of the 'normal' part of the I.Q. distribution curve. These people have a low I.Q. as an isolated phenomenon and appear to be normal in other respects.

Stott (11) has made an appraisal of Lewis's classification of mental defect (8)—exception is taken to the use of the term "Subcultural" and all cases so classified were examined for environmental factors, etc. This is along similar lines to those put forward earlier by Bourne (1), who considered that pathological mothering or fathering would lead to mental defect. Stott, in his article, seemed to think that mental subnormality was always pathological and pointed out that adults were seldom so short as to render them biologically incompetent; the analogy between distributions of height and I.Q.s was not valid according to this author, but to me this seems erroneous reasoning—lack of height would certainly make a person biologically incompetent to enrol in the Guards. Clarke and Clarke (2) consider that Lewis's division is made on sound theoretical grounds, whereas other people were of the opinion that such a division was on descriptive grounds rather than on aetiological ones (Sarason 10). The theoretical basis for Lewis's classification seems sound on statistical grounds and is endorsed by clinical experience.

The sub-divisions of the pathological group need little explanation. It is the differentiation between the non-pathological and undifferentiated pathological groups that is difficult, and this will be elaborated later.

Sub-group B of the pathological group is perhaps the most interesting, for it is amongst these cases that we hope to find the reversible or preventable types of mental handicap. The group of inborn errors of metabolism is being closely studied today and a new disease is described about once a year. It seems they all follow the same theoretical plan, for in these diseases there is a genetically determined absence or abnormality of a single enzyme that is responsible for one particular chemical step. This is shown diagrammatically in the figure below.



In the blocked pathway there occurs an accumulation of substances to the left of the block, with a diminution of those to the right of the block. To reduce the excess of B an unusual pathway is often used which results in the appearance in the urine of an abnormal metabolite E.

Phenylketonuria is the best studied one of these diseases, and here protein is represented by A, phenylalanine by B, tyrosine by C and melanin and other substances by D. E is the abnormal metabolite that causes the typical urine test, phenylpyruvic acid, and appears because phenylalanine is metabolised in an unusual way leading to the production of this substance and others such as indolyl acetic acid and indolyl lactic acid, to mention the two best known. This theoretical mechanism also explains the rationale treatment which has been found successful in PKU, that is to withhold substances to the left of the block and feed those to the right. When this is done in phenylketonuria, as is well known, the hair darkens, the biochemistry returns to normal, the E.E.G. becomes normal, and the child develops normally if treatment is started early enough.

Abnormalities of protein metabolism that can cause, or are associated with mental subnormality, have been discussed in a review by Rundle (9). In this review the other metabolic errors have also been reviewed with reference to the most important literature, and need not detain us further. Woolf (13) has given a list of the genetically determined aminoacidurias, and of the 17 described eight are associated with mental subnormality.

The chromosome abnormalities are well documented and have also been reviewed with the literature by Rundle (9). It is apparent that abnormalities of chromosome number, especially an excess, have a high incidence of mental subnormality.

As stated earlier, the genetic diseases should correctly be placed with the metabolic errors if the 'one gene one enzyme' theory is accepted, but until such time as the biochemical lesion is known, they are best included in this group under the appropriate sub-heading.

Mental handicap in children arising from a psychiatric abnormality needs no further amplification except to say that one hopes that it will be possible to have a finer classification of the juvenile psychoses in the near future.

Amongst the group of deprivation syndromes the most likely to be missed is the high tone deafness, since these children often appear disturbed, with no useful speech and no obedience to simple commands. If they were spotted early and appropriate steps taken to correct the defect, they would not be regarded as mentally subnormal, and would not come into the present classification, yet one still sees the deaf child who has been missed and misdiagnosed as mentally subnormal.

The biggest problem that will arise is the division of the undifferentiated cases of mental subnormality into the pathological and non-pathological groups. This has been discussed by Dutton (4) and the simplest method of differentiation would seem to be by estimating the skeletal age of the individual, if a child. In the case of those first seen as adults it may not always be possible to differentiate these two groups, but the results of a longitudinal growth study may reveal that there is a prolonged growing period in the pathological group, since there is retardation of skeletal maturation.

The results of an anthropometric survey (cross sectional) are shown in Table I. (This table is essentially that produced by Dutton (3 and 4) with some additions.) In order that comparisons can be made between children of different ages, and for a comparison between the skeletal development and the height and weight, developmental quotients have been used, as described by Talbot et al (12), from whom the norms were taken. This is not perhaps the ideal method as ratios of this nature are liable to accentuate any errors of measurement. It has, however, been found to work in practice, and is a useful method for showing specific failures in growth. From a very limited study it appears that the dental development in the group labelled 'metabolic' follows closely the skeletal development. This metabolic group are so called for want of a more exact name. They are definitely pathological, and by comparison with the findings in the group of phenylketonurics, it seems likely that the underlying cause is a biochemical one, but as yet it has not been determined. It is also possible, even likely, that this group is not a homogeneous one. It should also be added that all in this group were examined thoroughly to exclude known metabolic errors and none of the boys had a sex reversal on genetic sexing. The method of selection is described in Dutton (3): the initial segregation being on a height basis — the clinical differentiation is more precisely done by a simple radiograph of the left hand and wrist.

The comparison between certain aspects of growth and development, together with biochemical findings, in two groups of boys matched for life age from the non-pathological and pathological undifferentiated or 'metabolic' group are given in Table 2 taken from Dutton (3).

Horstmann (7) reported dwarfism associated with sexual immaturity and skeletal retardation, and found that of 47 dwarfs with sexual infantilism 19 were retarded mentally, whilst of 12 dwarfs who were sexually mature there were none who were intellectually handicapped. It was also found that there was a positive response to growth hormone in the former group, and Rundle (9) points out that there are probably two distinct syndromes. In one there is a primary pituitary abnormality which does not cause mental retardation, and in the other there is a brain anomaly which causes the mental subnormality and affects the pituitary.

It does seem that there are two distinct groups of undifferentiated mental subnormality, and that it is possible to differentiate these in growing boys. So far no adequate study of girls has been made but the impression from a small study suggests that the same applies to them as to the boys.

TABLE I (G. Dutton, October 1960)

MEAN DEVELOPMENT QUOTIENTS IN CLINICAL GROUPS OF MENTALLY SUBNORMAL BOYS

GROUP	Number of Cases	$\frac{\text{Height age}}{\text{Life age}} \times 100$	$\frac{\text{Weight age}}{\text{Life age}} \times 100$	$\frac{\text{Bone age}}{\text{Life age}} \times 100$
Mongols	50	68	77	95
Organic	31	86	92	95
Psychotic	15	88	94	92
P.K.U.	5	79	81	75
'Metabolic'	22	72	73	79
Non-Path.	16	97	94	100
Normal range	—	80-120	80-120	80-120

TABLE 2 (Taken from Dutton (3))

META-BOLIC	NON-PATH	GROUP
13.0	13.2	MEAN AGE—YEARS.
2.7	0.1	HEIGHT DEFICIT IN YEARS.
2.0	0.2	WEIGHT DEFICIT IN YEARS.
1.8	0	SKELETAL DEFICIT IN YEARS.
6.6	7.3	TOTAL SERUM PROTEINS G%
127	141	TOTAL SERUM CHOLESTEROL mgm.%
10.8	10.7	TOTAL SERUM CALCIUM mgm.%
3.6	3.5	SERUM INORGANIC PHOSPHORUS mgm.%
20	20	SERUM ALKALINE PHOSPHATASE K.A. units%
6.6	10.1	URINE NON-PROTEIN NITROGEN G per 24 hrs.
0.5	1.2	URINE CREATININE G per 24 hrs.
72	238	URINE CREATINE mgm. per 24 hrs.
3.6	7.7	URINE 17 KETOSTEROIDS mgm. per 24 hrs.
7.7	11.2	URINE 17 KETOGENIC STEROIDS mgm. per 24 hrs.

An analysis of the biochemical findings in two matched groups of mentally subnormal boys (9 cases in each group).

SUMMARY

1. The need for an accurate diagnosis in mental subnormality is stressed and it is pointed out that the legal classification has little use in clinical classification.
2. A classification on clinical, aetiological and pathological grounds is given.
3. A method for segregating the undifferentiated into a pathological and non-pathological group is discussed.

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