

# Studies in Mental Handicap

## Part I: Prevalence and distribution by clinical type and severity of defect

C. M. DRILLIEN, S. JAMESON, and E. M. WILKINSON

*From the Department of Child Life and Health, University of Edinburgh*

An aetiological study of mentally handicapped children in Edinburgh has provided an opportunity for assessing the prevalence of mental handicap in this area and the distribution of handicapped children by severity of defect and clinical type.

A definite cause of mental handicap is found in only a minority of cases. Nevertheless, any aetiological investigation must begin with an attempt to classify patients by clinical condition, since associations of possible aetiological significance are likely to differ for those having subnormal intelligence only and those having different neurological or physical abnormalities in addition to subnormal intelligence.

In this study the term 'mentally handicapped' is applied to children who are considered unsuitable for education in ordinary schools primarily on account of subnormal intelligence (i.e. estimated intelligence quotients of less than 70), but not to dull and backward children who are educationally retarded and may, in certain circumstances, be ineducable in the normal school system.

### The Survey Sample

Children selected for study were all those with IQ test scores below 70, born in the years 1950-56, who were themselves resident in Edinburgh and/or whose mothers were resident in Edinburgh in 1962-64.

Names of children considered to be mentally handicapped were obtained from registers compiled by the public health department and school medical service. Registers of children classified as physically handicapped were also scrutinized, since some mentally handicapped children may be so classified for domestic reasons. All voluntary day centres for handicapped children in Edinburgh were contacted, as were residential institutions and schools (both voluntary and state) for mentally and physically handicapped children throughout Scotland.

A final scrutiny of the relevant registers was made in July 1964, to include children who had been ascertained

as mentally handicapped since the beginning of the study. The total number of children ascertained was 406, of whom 391 were living in the guardianship of their mothers (with or without their fathers) and 15 were in the care of the Local Authority or relatives other than the mother. Information about these 15 children is incomplete. In July 1964 the youngest children in the survey were aged 7½ years and the oldest children 14½ years.

### Classification by Severity of Defect

Most surveys of the prevalence of mental handicap by severity of defect make a division between those having IQ levels below 50 (the severely handicapped) and those in the IQ range 50-69 (mildly handicapped or educationally subnormal). In the present study 21 children were judged to have IQ levels between 50 and 54. Their distribution by clinical type, social grade, and later disposal approximated closely to that of children in the IQ range 44-49 and differed from that of children having IQ levels of 55 and over. It was, therefore, thought appropriate to classify these children as severely, rather than mildly, handicapped in the study of causative factors. However, in the later section on prevalence the usual convention has been adhered to, survey subjects being divided by severity of defect into those with IQ levels below 50, and those with levels between 50 and 69.

Apart from the most severely defective children where the severity of defect was not in doubt, two or more IQ test scores were available for most of the children included in the sample. All had been tested at least once. Children considered to be on the border-lines between ineducability, educability in special schools for the mentally handicapped, and educability in normal schools were assessed most frequently. All children capable of responding were tested on the Terman and Merrill Revision of the Stanford Binet Scale. Most children over the age of 8½ years had also been tested on the Wechsler Intelligence Scale for children.

Children with additional physical handicaps or marked retardation in speech were assessed on tests appropriate for these disabilities. However, it should be realized that in the presence of a severe physical handicap an assessment of intelligence, at best, can only be approximate.

The most severely handicapped children (with

estimated IQ levels below 30) were unable to respond to tests appropriate for less severely affected children. In these cases a judgement of intelligence level was based on locomotor and manipulative abilities, comprehension of speech, and the degree to which the child was capable of benefiting from habit training.

Survey children were classified by severity of defect into the following categories.

1. *IQ less than 20.* Some of these children had learned to walk; none had learned to talk; none had acquired any skills in self-help.

2. *IQ 20-29.* Apart from those with incapacitating physical disabilities and others resident in institutions, most of these children attended day centres since they were considered unsuitable for occupation (training) centres. They proved capable of learning to communicate with understanding adults, and of responding to toilet training and training in other areas of self-help.

3. *IQ 30-44.* These children were considered capable of benefiting from occupation centre training but not from education in formal school subjects.

4. *IQ 44-54.* About one-half of these border-line children were given a trial period in special schools, and about one-quarter were retained there; in many cases this was because of strong opposition of their parents to downgrading.

5. *IQ 54-69.* These children were educated in special schools apart from some with additional physical disabilities and some with severe behaviour problems.

A number of children, on successive IQ tests, scored just under or just over the levels given above. If the child had been tested on three or more occasions and over one-half of test scores came into a particular category, he was included in that category. If, on an equal number of testings, he scored just below or just above the border-line, the mean score was taken as indicating the most accurate assessment. These criteria did not apply to children with pathological conditions causing a steady deterioration in intellectual ability.

Of those pupils in Edinburgh special schools who were born between 1950-56, there were 73 (one-quarter of the total) who were considered to have IQ levels of 70 or over. These children are not included here as being mentally handicapped.

#### Classification by Clinical Types

Of 218 children in the IQ range 54 or less, all but 16 had been fully investigated in hospital. During the survey a further 10 children were brought to hospital for examination. Six children were not examined, though school medical reports were available.

The majority of children in the IQ range 55-69 were not ascertained as mentally handicapped until after school entrance, and many had never been examined by a paediatrician or a neurologist. Few mothers were willing to bring their children to hospital, though many were willing for examination at home. Of a total of 188 children in this group, 139 were examined by a paediatrician or neurologist, and school medical reports were available for 49.

On the basis of individual examinations and school medical reports, the children were classified in the following clinical type groups.

1. *No other abnormalities found*, apart from minor defects of common occurrence in the general population, such as squints, hernias, haemangiomas, and minor degrees of syndactyly. Number of cases, 155.

2. *Epilepsy*, with or without other defects causing little additional handicap. Most children in this group had recurrent major and/or minor fits for periods of 12 months or longer. Children having a history of not more than three convulsions or convulsive episodes in the first 18 months of life and/or one further episode between 18 months and 2 years were not classified as epileptic; nor were children who at later ages had an isolated convulsive episode associated with head injury or central nervous system infection. Number of cases, 39.

3. *Cerebral palsy*, with or without epilepsy and other defects. Number of cases, 63.

4. *Other major and multiple defects* (excluding cerebral palsy) with or without epilepsy. Most children in this group had multiple congenital abnormalities. Some had an isolated physical defect such as blindness or deafness. Number of cases, 43.

5. *Mongolism* (Down's syndrome). Number of cases, 73.

6(A). *Other well-recognized conditions* invariably, usually, or frequently associated with mental handicap (e.g. phenylketonuria, the cerebral lipoidoses, hydrocephalus). Number of cases, 24.

6(B). *Other well-recognized conditions* known to be genetic in origin but not characteristically associated with mental handicap (e.g. muscular dystrophy, amyotonia congenita, ectrodactyly). Number of cases, 9.

#### Classification by Social Grade of Parents

Mothers were visited at home; most of those having children classified as severely handicapped, by an experienced health visitor, and most of those having children classified as mentally handicapped but educable, by a medical social worker. No assessment of social grade was made for the 15 children whose mothers were not resident in Edinburgh.

Of the mothers of 211 severely handicapped children (IQ less than 55), 6 were not visited since they had no contact with their children who were illegitimate; family doctors advised against visiting 3 mothers with marked guilt feelings; 2 mothers were in hospital throughout the period of the survey; and 2 refused to co-operate. Of the mothers of 180 children with IQ levels of 55-69, one had no contact with her illegitimate child; it was thought inadvisable to visit 2 with psychiatric disorders; and 3 were unwilling to co-operate. Thus, an assessment of social grade was made after interview at home in 372 instances and in 19 was obtained from other sources.

Families were graded on criteria described elsewhere (Drillien, 1964) as: (1) 'middle class or superior working class'; (2) 'average'; and (3) 'poor working class'. A further category of (4) 'very poor working class' was included. These homes were described by interviewers as 'appalling'. Most parents were of very low intelli-

gence and many had received psychiatric treatment. Most fathers were usually or permanently unemployed; many had served prison sentences. Many mothers were in chronic ill-health. The illegitimacy rate was high. In many families parents were separated, and in others the mothers were known to have cohabited with a number of men, so that it was difficult or impossible to distinguish who, among the children, had been fathered by whom.

In a previous study (Drillien, 1964) social grading for the families of all infants born in Edinburgh 1953-56 (using the same criteria as employed here) was estimated as being 59% middle class and superior working class, 32% average, and 9% poor working class. Re-examination of the data indicated that not more than 1% of families would have been graded as very poor.

**Findings**

**Prevalence of mental handicap by severity of defect.** An attempt was made to assess the prevalence of mental handicap in the total population of children resident in Edinburgh and aged 7½-14½ years (i.e. born 1950-56), which, in July 1964, was estimated to be 39,498. The number of children in the survey sample known to have IQ levels below 70 was 406; 197 had IQ levels below 50; and 209 had IQ levels in the range 50-69.

Of the 197 children with IQ levels below 50, only 4 had been ascertained after the age of 7½ years. Of these, 2 with severe epilepsy were transferred from a school for the physically handicapped at 8½-9 years, because of steady deterioration in intellectual ability, and the other 2 who had rapidly progressive conditions were excluded from normal school at 9½ years. It seems likely that the ascertainment of these severely handicapped children is virtually complete.

It is more difficult to be confident of complete ascertainment of children in the IQ range 50-69. All children in local authority primary schools in Edinburgh are given group intelligence tests before transfer from infant to junior departments (normally at 7-7½ years); thus, ascertainment of children with IQ levels of 50-69 is likely to be made at a relatively early age. Nevertheless, in this IQ range, there was an obvious deficit in numbers born in the years 1953-56, which seemed likely to be due to late ascertainment.

Of those born 1950-52 (aged 12½-14½ years at the final scrutiny), none had been ascertained and transferred to special schools after the age of 12 years. 38% were ascertained before 7½ years, 52% between 7½ and 10½ years, and 8% between 10½ and 12 years. Assuming that the proportions of total children ascertained at different ages would be roughly the same for those born 1953-56 as those born 1950-52, it was calculated that in July 1964 40 mentally handicapped children would still be in normal school. This number has been added to that of children known to be mentally handicapped in the IQ range 50-69, making a total of 249 in this group.

TABLE I

*Prevalence of Mental Handicap by Severity of Defect in a Total Population of 394,498 Children*

| Severity of Defect IQ Range | No. of Cases | Mentally Handicapped (%) |
|-----------------------------|--------------|--------------------------|
| -19                         | 49           | 0.12                     |
| 20-49                       | 148          | 0.38                     |
| 50-69                       | 249*         | 0.63                     |
| Total                       | 446          | 1.13                     |

\* Including 40 cases calculated as not being ascertained in July 1964.

TABLE  
*Distribution of Survey Sample by Sex,*

| IQ   | Male |       |           |       |       |             | 55-69 | -19 | 20-29 |
|--|------|-------|-----------|-------|-------|-------------|-------|-----|-------|
|  | -19  | 20-29 | Total -29 | 30-44 | 45-54 | Total 30-54 |       |     |       |
| <i>Clinical Type</i>                       |      |       |           |       |       |             |       |     |       |
| (1) No other abnormalities .. .. .         | 1    | 2     | 3         | 9     | 12    | 21          | 66    | —   | 1     |
| (2) Epilepsy .. .. .                       | 5    | —     | 5         | 3     | 4     | 7           | 8     | 3   | —     |
| (3) Cerebral palsy .. .. .                 | 13   | 4     | 17        | 4     | 6     | 10          | 7     | 14  | 2     |
| (4) Major/multiple defects .. .. .         | 1    | 1     | 2         | 5     | 6     | 11          | 14    | 3   | —     |
| (5) Down's syndrome .. .. .                | 1    | 4     | 5         | 23    | 4     | 27          | 2     | 3   | 2     |
| (6A) Other recognizable conditions .. .. . | 2    | 5     | 7         | 2     | 5     | 7           | 3     | 3   | 1     |
| (6B) Other recognizable conditions .. .. . | —    | —     | —         | 1     | 2     | 3           | 5     | —   | —     |
| Total .. .. .                              | 23   | 16    | 39        | 47    | 39    | 86          | 105   | 26  | 6     |
| % .. .. .                                  |      |       | 100.0     |       |       | 100.0       | 100.0 |     |       |

The estimated prevalence of mental handicap by severity of defect is given in Table I.

**Distribution of the survey sample by severity of defect, clinical type, and sex.** Table II gives the numbers of boys and girls in the clinical type groups described above by severity of defect.

Among those with IQ levels less than 30, all but 6% had additional defects; nearly one-half suffered from cerebral palsy. In the IQ range 30-54 nearly one-half were mongols, over one-third had other defects, and less than one-quarter had no major disability other than subnormal intelligence. In the IQ range 55-69 two-thirds were of subnormal intelligence without other defects and belonged to no recognized clinical type.

Of the 73 special school pupils who were considered to be dull and backward rather than mentally handicapped, 2 suffered from epilepsy and were waiting other placement, and 71 were said in their school medical reports to have no other defects.

Whatever the degree of defect, there were more boys than girls. Similarly, in all clinical type groups (except Down's syndrome) there were more boys. Of the 73 special school pupils graded as dull and backward 48 (66%) were boys, a much higher proportion of boys than that found in any lower IQ group.

**Distribution of the survey sample by severity of defect, clinical type, and social grade.** In this and the following section, the 15 children whose mothers were not resident in Edinburgh have been excluded. Two had IQ levels below 30, 5 between 30 and 54, and 8 between 55 and 69. The social grade was not known of one other child with

cerebral palsy and an IQ of 55-69. This case has also been omitted from Table III which gives the distribution by severity of defect, clinical type, and social grade of the remaining 390 children; 69 of IQ less than 30, 142 of IQ 30-34, and 179 of IQ 55-69. The proportion of children coming from different social grades has been calculated for those having no abnormalities apart from subnormal intelligence, those in all other clinical type groups, by severity of defect.

Among those with IQ levels less than 30, and those having other abnormalities in the IQ range 30-54, the distribution by social grade was not much different from that of the general population. Of children with IQ levels of 30-54 and no other abnormalities, 29% came from poor homes (social grade 3) and 10% from very poor homes (social grade 4).

Differences in social grade as compared with the general population were even more marked in the IQ range 55-69, particularly if survey subjects had no additional abnormalities, in which case 36% came from poor homes and 20% from very poor homes. Of the 73 dull and backward children attending special schools 36% also came from poor homes, but in this category one-half came from homes graded as very poor.

**Distribution of survey sample by clinical type and major aetiological factor.** An attempt was made to subdivide cases of different clinical type by the most likely major aetiological factor causing mental handicap, taking into consideration pre-, peri-, and postnatal history, and the clinical condition of each individual child, as follows.

(a) *Mental handicap due to genetically determined*

II  
*Severity of Defect, and Clinical Condition*

| Female    |       |       |             | Total |     |       |           |       |       |             |       |
|-----------|-------|-------|-------------|-------|-----|-------|-----------|-------|-------|-------------|-------|
| Total -29 | 30-44 | 45-54 | Total 30-54 | 55-69 | -19 | 20-29 | Total -29 | 30-44 | 45-54 | Total 30-54 | 55-69 |
| 1         | 3     | 9     | 12          | 52    | 1   | 3     | 4         | 12    | 21    | 33          | 118   |
| 3.1       |       |       | 19.7        | 62.7  |     |       | 5.6       |       |       | 22.4        | 62.8  |
| 3         | 2     | 2     | 4           | 12    | 8   | —     | 8         | 5     | 6     | 11          | 20    |
| 9.4       |       |       | 6.6         | 14.5  |     |       | 11.3      |       |       | 7.5         | 10.6  |
| 16        | 3     | 4     | 7           | 6     | 27  | 6     | 33        | 7     | 10    | 17          | 13    |
| 50.0      |       |       | 11.5        | 7.2   |     |       | 46.5      |       |       | 11.6        | 6.9   |
| 3         | 2     | 2     | 4           | 9     | 4   | 1     | 5         | 7     | 8     | 15          | 23    |
| 9.4       |       |       | 6.6         | 10.8  |     |       | 7.0       |       |       | 10.2        | 12.2  |
| 5         | 27    | 6     | 33          | 1     | 4   | 6     | 10        | 50    | 10    | 60          | 3     |
| 15.6      |       |       | 54.0        | 1.2   |     |       | 14.1      |       |       | 40.9        | 1.6   |
| 4         | —     | 1     | 1           | 2     | 5   | 6     | 11        | 2     | 6     | 8           | 5     |
| 12.5      |       |       | 1.6         | 2.4   |     |       | 15.5      |       |       | 5.4         | 2.7   |
| —         | —     | —     | —           | 1     | —   | —     | —         | 1     | 2     | 3           | 6     |
|           |       |       |             | 1.2   |     |       |           |       |       | 2.0         | 3.2   |
| 32        | 37    | 24    | 61          | 83    | 49  | 22    | 71        | 84    | 63    | 147         | 188   |
| 100.0     |       |       | 100.0       | 100.0 |     |       | 100.0     |       |       | 100.0       | 100.0 |

*Distribution of Survey Sample by Severity of Defect, Clinical Condition, and Social Grade; and*

| Social Grade                               | Intelligence Quotient <29 |      |      |     |       | Intelligence Quotient 30-54 |      |      |     |
|--|---------------------------|------|------|-----|-------|-----------------------------|------|------|-----|
|  | 1                         | 2    | 3    | 4   | Total | 1                           | 2    | 3    | 4   |
| <i>Clinical Type</i>                       |                           |      |      |     |       |                             |      |      |     |
| (1) No other abnormalities ..              | 1                         | 2    | 1    | —   | 4     | 9                           | 10   | 9    | 3   |
| % .. .. .                                  | 25.0                      | 50.0 | 25.0 | —   | 100.0 | 29.0                        | 32.3 | 29.0 | 9.7 |
| (2) Epilepsy .. .. .                       | 6                         | 1    | 1    | —   | 8     | 5                           | 4    | 2    | —   |
| (3) Cerebral palsy .. .. .                 | 11                        | 14   | 6    | —   | 31    | 7                           | 8    | 1    | 1   |
| (4) Major/multiple defects ..              | 1                         | 3    | —    | 1   | 5     | 7                           | 7    | —    | 1   |
| (5) Down's syndrome .. .. .                | 7                         | 1    | 2    | —   | 10    | 31                          | 22   | 5    | —   |
| (6A) Other recognizable conditions .. .. . | 5                         | 1    | 5    | —   | 11    | 2                           | 4    | 1    | —   |
| (6B) Other recognizable conditions .. .. . | —                         | —    | —    | —   | —     | 1                           | —    | 2    | —   |
| Total (2)—(6B) .. .. .                     | 30                        | 20   | 14   | 1   | 65    | 53                          | 45   | 11   | 2   |
| % .. .. .                                  | 46.2                      | 30.8 | 21.5 | 1.5 | 100.0 | 47.8                        | 40.5 | 9.9  | 1.8 |

General population of Edinburgh births (%)

conditions. (i) *Definite*: This category included mongol children and those having well-defined genetically determined conditions invariably, usually, or frequently associated with mental handicap, such as phenylketonuria, tuberosc sclerosis, and hydrocephalus due to stenosis of the aqueduct.

(ii) *Possible*: Children in this category suffered from familial conditions (such as muscular dystrophy and cleido-cranial dysostosis) which are not characteristically associated with mental handicap, though the observed incidence of handicap may be significantly above that found in the general population. Presumably in some cases the mental handicap is also genetically determined and in others it occurs by chance.

(b) *Mental handicap due to other factors causing developmental malformation*. The term 'developmental' is used here to mean arising in early foetal life, during the period of organogenesis. Since the aetiology of most developmental malformations is obscure, this category might more accurately be classified as aetiology uncertain. However, indication of origin in early pregnancy narrows the area of search for causative factors, which may be expected to differ in this category from those operating in other cases of uncertain aetiology.

(i) *Probable*: This category included children who showed other anomalies (commonly occurring defects excepted) which must have arisen at an early stage of foetal life.

(ii) *Possible*: Mental handicap was considered to be possibly due to developmental malformation when parents and/or sibs suffered from developmental anomalies (excluding mental handicap and epilepsy); when the mother gave a history of habitual abortion and/or repeated threatened abor-

tion or hydramnios during the pregnancy of the survey child. These factors were commonly reported when the survey child himself showed other developmental anomalies, and have also been reported as significant aetiological associations in other studies of developmental malformations (Drillien, Ingram, and Wilkinson, 1966).

(c) *Mental handicap due to perinatal factors*.

(i) *Probable*: Birth injury (prenatal hypoxia and/or trauma at delivery) was considered to be the most likely cause of mental handicap in cases with a history of severe obstetric complications followed by immediate and continuing postnatal signs of cerebral damage.

Case 101 is an example of the sort of history given in cases classified as most probably due to birth injury.

The mother was admitted to hospital at 44 weeks' gestation in a collapsed state following a massive haemorrhage due to a central placenta praevia. A lower segment caesarean section was performed. The baby, a female, weighing 3600 g. (7 lb. 15 oz.), did not cry at birth. Shallow respirations commenced in 15 minutes. 7 hours later she showed marked head retraction and generalized twitching. By the ninth day there was some improvement; the baby was less irritable though generalized tremor persisted. She was lethargic and unable to suck. By 3 weeks she was much improved and was discharged home. She continued to be extremely difficult to feed. At 3 months she was having frequent myoclonic fits. She is now a grossly mentally handicapped child with cerebral diplegia, epilepsy, and cortical blindness.

Mental handicap following kernikterus, and associated with extreme prematurity (birthweight 1500 g. (3.3 lb.) or less) in the absence of any

III

Social Grade Distribution for All Singleton Legitimate Births in Edinburgh 1953-55 (Drillien, 1964)

| Total        | Intelligence Quotient 55-69 |            |            |            |              | All Cases With Intelligence Quotient < 70 |            |            |            |              |
|--------------|-----------------------------|------------|------------|------------|--------------|---|------------|------------|------------|--------------|
|              | 1                           | 2          | 3          | 4          | Total        | 1   | 2          | 3          | 4          | Total        |
| 31<br>100.0  | 7<br>6.3                    | 42<br>37.9 | 40<br>36.0 | 22<br>19.8 | 111<br>100.0 | 17<br>11.6                                | 54<br>37.0 | 50<br>34.2 | 25<br>17.2 | 146<br>100.0 |
| 11           | 3                           | 13         | 4          | —          | 20           | 14  | 18         | 7          | —          | 39           |
| 17           | 2                           | 5          | 5          | —          | 12           | 20  | 27         | 12         | 1          | 60           |
| 15           | 3                           | 9          | 10         | —          | 22           | 11  | 19         | 10         | 2          | 42           |
| 58           | 3                           | —          | —          | —          | 3            | 41  | 23         | 7          | —          | 71           |
| 7            | —                           | 3          | 1          | 1          | 5            | 7   | 8          | 7          | 1          | 23           |
| 3            | 1                           | 3          | 2          | —          | 6            | 2   | 3          | 4          | —          | 9            |
| 111<br>100.0 | 12<br>17.6                  | 33<br>48.6 | 22<br>32.3 | 1<br>1.5   | 68<br>100.0  | 95<br>38.9                                | 98<br>40.2 | 47<br>19.3 | 4<br>1.6   | 244<br>100.0 |
|              |                             |            |            |            |              | 58.5                                      | 32.2       | 9.3        |            | 100.0        |

indication that premature delivery was secondary to abnormality of the foetus (Drillien, 1965), has also been included as most probably due to perinatal factors.

(ii) *Possible*: Some infants who appeared to have suffered definite birth injury from their history of severe obstetric complications and postnatal signs suggestive of cerebral damage were classified as having handicap possibly due to birth injury, because of other features in the history suggestive of developmental origin of defect, e.g. Case 205.

At delivery the cord was found to be wound tightly round the neck. The baby, a female, was limp, apnoeic, and cyanosed, and responded slowly to resuscitation. From the first day she was noted to have a bulging fontanelle and to be very irritable with generalized twitching which persisted during the first week. At 9 days her condition was much improved, but her subsequent mental handicap was attributed to birth injury. Birthweight was 2465 g. (5 lb. 7 oz.) at term. Her 3 sibs (2 older brothers and a younger sister) were all over 3175 g. (7 lb.) at birth. The child herself has a bifid uvula and shortening of the left leg. The mother had Asian influenza during the first month of pregnancy.

When birthweight was between 1501 and 2000 g. it was considered that the premature delivery and/or the commonly associated complications might possibly be of aetiological significance.

(d) *Mental handicap due to postnatal factors.*

(i) *Definite or probable*: In 3 cases of mental handicap following severe head injury there was independent evidence that the children had been mentally normal before their accidents. In 2 cases of 'battered baby syndrome' it seemed highly probable that handicap resulted from cerebral injury; both

infants presented with multiple fractures and neurological signs of cerebral damage.

There was independent confirmation of previous normality in some cases of handicap following infective illness, such as tuberculous meningitis and encephalitis, and in some who were thought to have suffered from cerebral vascular accidents.

(ii) *Possible*: In other cases it was not possible to obtain evidence of previous normal development from anyone other than their relatives, and these cases were classified as being possibly postnatal in origin.

(e) *Aetiology uncertain.* This category included the large number of children who had no other anomalies, no maternal or family history suggestive of developmental origin of mental handicap, and no history of postnatal incidents or severe obstetric complications likely to have been causative. The majority of these children came from poor or very poor homes, and had parents and/or sibs who were also of low intelligence.

Five children were considered to be autistic in addition to there being certain mental handicap in 4 and probable handicap in one. All the children showed bizarre behaviour patterns, had very marked retardation of speech, and were described as being completely withdrawn. None had epilepsy or other clinical or pathological conditions. There is little agreement about the aetiology of the early psychoses and these cases have been classified as aetiology uncertain.

Table IV gives what appeared to be the major factor causing mental handicap by clinical type and severity of defect.

Table V gives numbers and proportions by clinical type and severity of defect separately for

TABLE  
Distribution of Survey Sample by Clinical Type,

| Major Aetiological Factor | Intelligence Quotient -54 |          |                |                        |                 |                                      | Total |       |
|---------------------------|---------------------------|----------|----------------|------------------------|-----------------|--------------------------------------|-------|-------|
|                           | 1                         | 2        | 3              | 4                      | 5               | 6                                    |       |       |
|                           | No Other Abnormalities    | Epilepsy | Cerebral Palsy | Major Multiple Defects | Down's Syndrome | Other Recognizable Conditions<br>A B |       |       |
| <i>Genetic</i>            |                           |          |                |                        |                 |                                      |       |       |
| (i) Definite .. ..        |                           |          |                |                        | 68              | 5                                    | 3     | 73    |
| (ii) Possible .. ..       |                           |          |                |                        |                 |                                      |       | 3     |
| Total .. ..               |                           |          |                |                        | 68              | 5                                    | 3     | 76    |
| % .. ..                   |                           |          |                |                        | 100.0           | 27.8                                 | 100.0 | 36    |
| <i>Developmental</i>      |                           |          |                |                        |                 |                                      |       |       |
| (i) Probable .. ..        |                           | 2        | 9              | 12                     |                 | 5                                    |       | 28    |
| (ii) Possible .. ..       | 3                         | 2        | 9              |                        |                 |                                      |       | 14    |
| Total .. ..               | 3                         | 4        | 18             | 12                     |                 | 5                                    |       | 42    |
| % .. ..                   | 8.6                       | 21.1     | 37.5           | 60.0                   |                 | 27.8                                 |       | 19.9  |
| <i>Perinatal</i>          |                           |          |                |                        |                 |                                      |       |       |
| (i) Probable .. ..        |                           | 1        | 9              | 1                      |                 | 2                                    |       | 13    |
| (ii) Possible .. ..       | 6                         |          | 2              | 5                      |                 | 1                                    |       | 14    |
| Total .. ..               | 6                         | 1        | 11             | 6                      |                 | 3                                    |       | 27    |
| % .. ..                   | 17.1                      | 5.3      | 22.9           | 30.0                   |                 | 16.7                                 |       | 12.8  |
| <i>Postnatal</i>          |                           |          |                |                        |                 |                                      |       |       |
| (i) Probable .. ..        |                           | 5        | 9              | 2                      |                 |                                      |       | 16    |
| (ii) Possible .. ..       |                           |          | 2              |                        |                 | 1                                    |       | 3     |
| Total .. ..               |                           | 5        | 11             | 2                      |                 | 1                                    |       | 19    |
| % .. ..                   |                           | 26.3     | 22.9           | 10.0                   |                 | 5.5                                  |       | 9.0   |
| Aetiology uncertain ..    | 26                        | 9        | 8              |                        |                 | 4                                    |       | 47    |
| % .. ..                   | 74.3                      | 47.4     | 16.7           |                        |                 | 22.2                                 |       | 22.3  |
| Total no. of cases ..     | 35                        | 19       | 48             | 23                     | 68              | 18                                   | 3     | 211   |
| % .. ..                   | 100.0                     | 100.0    | 100.0          | 100.0                  | 100.0           | 100.0                                | 100.0 | 100.0 |

TABLE V

Distribution of Survey Sample by Clinical Type, Severity of Defect, and Whether or Not a Possible or Probable Cause of Mental Handicap was Apparent. Figures in Parentheses Indicate Numbers and Proportions of Children from Social Grades 3 and 4 and/or Having Parents of Very Low Intelligence

|                                     | 1                      | 2            | 3              | 4                      | 5               | 6                                    |              | Total        |
|-------------------------------------|------------------------|--------------|----------------|------------------------|-----------------|--------------------------------------|--------------|--------------|
|                                     | No Other Abnormalities | Epilepsy     | Cerebral Palsy | Major/Multiple Defects | Down's Syndrome | Other Recognizable Conditions<br>A B |              |              |
| <i>Intelligence-Quotient -54</i>    |                        |              |                |                        |                 |                                      |              |              |
| Probable/possible cause apparent .. | 9 (3)                  | 10 (3)       | 40 (6)         | 20 (2)                 | 68 (7)          | 14 (6)                               | 3 (2)        | 164 (29)     |
| % .. ..                             | 25.7                   | 52.6         | 83.3           | 100.0                  | 100.0           | 77.8                                 | 100.0        | 77.7 (17.7)  |
| Aetiology uncertain ..              | 26 (10)                | 9 (—)        | 8 (2)          |                        |                 | 4 (—)                                |              | 47 (12)      |
| % .. ..                             | 74.3                   | 47.4         | 16.7           |                        |                 | 22.2                                 |              | 22.3 (25.6)  |
| Total .. ..                         | 35 (13)                | 19 (3)       | 48 (8)         | 20 (2)                 | 68 (7)          | 18 (6)                               | 3 (2)        | 211 (41)     |
| % .. ..                             | 100.0 (37.1)           | 100.0 (15.8) | 100.0 (16.7)   | 100.0 (10.0)           | 100.0 (10.3)    | 100.0 (33.3)                         | 100.0 (66.7) | 100.0 (19.4) |
| <i>Intelligence-Quotient 55-69</i>  |                        |              |                |                        |                 |                                      |              |              |
| Probable-possible cause apparent .. | 31 (12)                | 8 (—)        | 9 (4)          | 22 (10)                | 3 (—)           | 4 (1)                                | 6 (2)        | 83 (29)      |
| % .. ..                             | 27.9                   | 40.0         | 69.2           | 100.0                  | 100.0           | 80.0                                 | 100.0        | 46.1 (34.9)  |
| Aetiology uncertain ..              | 80 (53)                | 12 (4)       | 4 (1)          |                        |                 | 1 (1)                                |              | 97 (59)      |
| % .. ..                             | 72.1                   | 60.0         | 30.8           |                        |                 | 20.0                                 |              | 53.9 (60.8)  |
| Total .. ..                         | 111 (65)               | 20 (4)       | 13 (5)         | 22 (10)                | 3 (—)           | 5 (2)                                | 6 (2)        | 180 (88)     |
| % .. ..                             | 100.0 (58.6)           | 100.0 (20.0) | 100.0 (38.5)   | 100.0 (45.5)           | 100.0 (—)       | 100.0 (40.0)                         | 100.0 (33.3) | 100.0 (48.9) |

IV  
Severity of Defect, and Major Aetiological Factors

| Intelligence Quotient 55-69 |             |                |                        |                 |                               |            | Total        |
|-----------------------------|-------------|----------------|------------------------|-----------------|-------------------------------|------------|--------------|
| 1                           | 2           | 3              | 4                      | 5               | 6                             |            |              |
| No Other Abnormalities      | Epilepsy    | Cerebral Palsy | Major Multiple Defects | Down's Syndrome | Other Recognizable Conditions |            |              |
|                             |             |                |                        |                 | A                             | B          |              |
|                             |             |                |                        | 3               | 2                             | 6          | 3<br>8       |
|                             |             |                |                        | 3<br>100.0      | 2<br>40.0                     | 6<br>100.0 | 11<br>6.1    |
| 19                          | 1<br>3      | 1              | 21                     |                 | 1<br>1                        |            | 24<br>23     |
| 19<br>17.1                  | 4<br>20.0   | 1<br>7.7       | 21<br>95.5             |                 | 2<br>40.0                     |            | 47<br>26.1   |
| 11                          | 2           | 2<br>3         | 1                      |                 |                               |            | 4<br>15      |
| 11<br>9.9                   | 2<br>10.0   | 5<br>38.5      | 1<br>4.5               |                 |                               |            | 19<br>10.6   |
| 1                           | 2           | 3              |                        |                 |                               |            | 5<br>1       |
| 1<br>0.9                    | 2<br>10.0   | 3<br>23.0      |                        |                 |                               |            | 6<br>3.3     |
| 80<br>72.1                  | 12<br>60.0  | 4<br>30.8      |                        |                 | 1<br>20.0                     |            | 97<br>53.9   |
| 111<br>100.0                | 20<br>100.0 | 13<br>100.0    | 22<br>100.0            | 3<br>100.0      | 5<br>100.0                    | 6<br>100.0 | 180<br>100.0 |

those whose mental handicap was of uncertain aetiology and those where a probable or possible cause was apparent. In parentheses are given the numbers and proportions of children coming from social grades 3 and 4 and/or having one or both parents of very low intelligence (i.e. one or both parents attended special schools for the mentally handicapped, or attended normal school but admitted to 'never learning to read or write properly'; in most of these latter cases there was no special school available; in some cases the parents seldom or never attended school).

Among children with IQ levels below 55, mental handicap was thought to be genetically determined in 36% of cases, developmental in origin in a further 20%, due to perinatal factors in 13% and to postnatal infections or accidents in 9 per cent. In 22% aetiology was uncertain; one-quarter of these children came from social grade 3 and 4 homes and/or had parents of low intelligence, compared with one-sixth of those in other aetiological categories.

Among children in the IQ range 55-69 mental handicap appeared to be due to genetic factors in 6%, to be developmental in origin in a further 26%, to be

due to perinatal factors in 11%, and to postnatal factors in 3%. In 54% aetiology was uncertain; two-thirds of these children came from an impoverished environment compared with one-third in other aetiological categories.

Each clinical type group was considered separately in relation to presumed causation (Table IV).

1. *No other abnormality.* Of 146 children with no other abnormalities, the cause of mental handicap was uncertain in 106 (73%). Of these, 63 were reared in social grades 3 or 4 homes and/or had parents of very low intelligence. In 12% perinatal factors may have been causative. In only one case was mental handicap attributed to postnatal causes.

2. *Epilepsy.* In the group of 39 children with epilepsy and no other neurological or major physical defects, the cause of both mental handicap and epilepsy was probably or possibly due to postnatal factors in 18% and to adverse perinatal factors in 12%.

3. *Cerebral palsy.* Of 61 children with cerebral palsy, perinatal factors may have caused both mental handicap and neurological defect in 26% of cases, and postnatal factors in a further 22%.



4. *Other major or multiple defects.* 42 children had additional major or multiple defects (other than cerebral palsy) which were thought to be due to peri- or postnatal factors in 4. 2 blind children with retrolental fibroplasia had been nursed postnatally in high oxygen concentrations on account of very low birth weight. One child had high-tone deafness following tuberculous meningitis and another had cortical blindness which originated at 9 months from cerebral injury ('battered baby syndrome').

In the 39 other cases who had additional developmental anomalies, it seemed reasonable to suppose that mental handicap also originated in early foetal life. However, 5 of these had a history of adverse peri- or postnatal factors that might have been causatively associated with their mental handicap though not with their developmental defects.

5. *Down's syndrome.* Chromosomal investigation was carried out in only a few of the 71 cases. However, all showed the typical physical stigmata of this condition and all were assumed to be due to genetic factors.

6A. *Other well-recognized conditions usually associated with mental handicap.* The following clinical conditions were included in this group.

(a) *Hydrocephalus:* of 7 cases, 2 were associated with lumbar meningomyelocele, one was due to stenosis of the aqueduct, one other was presumed to be due to developmental malformation, and 3 probably resulted from birth injury.

(b) *Malformation of the brain:* this diagnosis was made following air encephalography in one case each of agenesis of the corpus callosum, cortical agenesis, and megalencephaly.

(c) *Metabolic disorders:* 2 children suffered from phenylketonuria, one from tuberose sclerosis, one from retinitis pigmentosa with obesity, and one was a cretin. One girl who died during the survey at 12 years was the second member of the family to die from Gaucher's disease. There was some difference of opinion as to the diagnosis in 2 children with rapid deterioration in intellectual functioning, but the consensus of opinion was that the one was a case of Niemann-Pick disease and the other of Van Bogaert's subacute sclerosing leucoencephalopathy.

(d) *Psychoses:* the 5 cases showing features of autism have already been described.

6B. *Other hereditary or familial conditions not usually characterized by mental handicap.* This group included 4 boys with very marked congenital laxity of ligaments. 3 were brothers who presented initially with difficulty in walking. The other boy was also referred initially to a surgeon who reported . . . 'I have seldom seen a child with such lax ligaments at ankles and knees' . . . ; an older brother who

is more severely mentally handicapped has laxity of knee ligaments and difficulty in walking.

One girl showed cleido-cranial dysostosis and a minor congenital heart lesion; 2 of her 4 sisters who had died of congenital heart defects also had similar bony abnormalities. A surviving brother has bony abnormalities, congenital atrophy of the left leg, left microphthalmia with cataract, and probable mental handicap.

Two boys suffered from muscular dystrophy; one had 3 healthy sisters and the other an unaffected elder brother. One other boy suffered from amyotonia congenita and one presented with ectrodactyly.

## Discussion

**Prevalence of mental handicap by severity of defect.** The incidence of mental handicap in school age children in this country is said to be between 2.0 and 2.5%, of whom about 1.5 to 2.0% are classified as educable in special schools and 0.3 to 0.5% as severely handicapped and ineducable (Scottish Education Department, 1958; Carter, 1962; Penrose, 1963). At best incidence figures can only provide a rough estimate, as it is virtually impossible to be confident of complete ascertainment. It is also difficult to compare figures from different regions, since the proportion of total school age children who are ascertained as mentally handicapped, and/or educated in special schools will depend largely on local facilities. It is even more difficult to compare figures from different countries, since the tolerance level for subnormal functioning will depend on prevailing social and cultural standards (Knobloch and Pasamanick, 1962).

In addition, it is well known that among special school pupils (as among adults in mental deficiency institutions) a proportion are not mentally handicapped, in the sense of having intelligence quotients of less than 70. Conversely, many children who would be considered mentally handicapped on this criterion are retained in the normal school system.

After a thorough investigation in the County of Middlesex, Goodman and Tizard (1962) estimated the prevalence of severe subnormality (IQ less than 50) as 0.35% of the total population aged 7 years and under 14 years. They stated that this prevalence was higher than that reported in most recent surveys. An even higher rate (0.49%) was found in the present survey, possibly as a result of the inclusion of children who were in institutions in other parts of Scotland but whose mothers were resident in Edinburgh, and of the good local

facilities for severely handicapped children, including active and publicity-minded branches of the associations for mentally handicapped and cerebral-palsied children.

The prevalence of less severely handicapped children with IQ levels of 50-69 (0·63%) was much lower than that generally accepted as indicating the true prevalence of handicap in this IQ range. This may be due in part to the exclusion from the survey sample of special school pupils considered to have IQ levels of 70 or over, who comprised one-quarter of total attenders in Edinburgh special schools.

Of the 73 children in this category, only 6 came from materially adequate homes and one-half came from homes described as very poor. It is likely that in a relatively prosperous city such as Edinburgh the proportion of children reared in very adverse circumstances would be low as compared with the country as a whole.

**Distribution by severity of defect, clinical type, and sex.** The Edinburgh study confirms the findings of other workers that a majority of severely handicapped subjects suffer from well-recognized syndromes and/or have other neurological and physical defects in addition to mental handicap (Lewis, 1929; Berg and Kirman, 1959). Crome (1960) reported on morphological findings in 282 brains obtained at necropsy from mentally handicapped persons, mostly of low grade. One-third were classified as recognized syndromes. Of the two-thirds who could not be so classified, all but 8 showed some morphological change.

There is also general agreement that a majority of higher grade defectives have no obvious pathological condition apart from subnormal intellectual functioning, which probably results in most cases from a combination of poor genetic endowment of intelligence and an adverse postnatal environment. Although these so-called 'subcultural defectives' (Lewis, 1929) are likely to have parents and sibs of low intelligence, there were few families, in the Edinburgh study, in which all or even most of the sibs were of subnormal intelligence. This suggests that additional causative factors may have been operating in the case of survey children.

That some cases of subnormal intelligence with no additional abnormalities and of uncertain aetiology, result from causes other than poor genetic endowment and material deprivation is shown by the Edinburgh children in this category (6%) who came from superior homes (six from Social Classes I and II). Four had normal sibs, all of whom attended Grammar schools. Of the other three, one resulted from the sole conception of an infertile

woman; one had two sibs of normal intelligence and one who suffered from mental handicap and associated congenital anomalies; and one (the son of a lawyer) had an older sister who attended normal school but was considered to be very dull and educationally retarded.

Most large surveys report a preponderance of males amongst the severely handicapped (Penrose, 1963), including mongols.

The sex ratio (M : F) in mongolism is said to be of the order 1·3 : 1 (Hug, 1951); in the Edinburgh survey sample the M : F ratio was 0·87 : 1. It was thought possible that the excess of females at ages 7½-14½ years might be due to an increased mortality in early childhood of male mongols. However, an even larger excess of female mongols was found on examination of the records of all mongol live births in two Edinburgh maternity hospitals during the years 1957-65 (M : F, 0·69 : 1). Penrose (1963) quotes figures that indicate that the preponderance of males amongst mongol children is most marked for those born to mothers aged 35-39 years, and that at maternal ages of 40 or over there is a slight excess of females. The difference in sex ratio of mongol children in Edinburgh from that reported from other regions was not due to any difference in age distribution of mothers, and there is no obvious explanation for the excess of females found.

It has been suggested that the larger number of severely handicapped males is liable to be counter-balanced by a converse excess of mildly subnormal females (Penrose, 1963, quoting figures of Dayton, 1939, for the number of male and female patients resident in the Massachusetts State Schools for the Mentally Defective). However, as Penrose points out, the distribution by sex and severity of defect of institutionalized patients may reflect social selection rather than genuine biological differences.

Similarly, differences in sex distribution of samples of defectives ascertained in childhood could be due to earlier ascertainment in one sex for social reasons.

In the Edinburgh survey 56% of children in the IQ range 55-69 were boys; the proportion of boys was the same whether or not the survey child showed other abnormalities. Among the two-thirds who had no other neurological or physical abnormalities, 52% of those born in 1950-52 were boys, compared with 59% of the later born (1953-56). Of the 73 dull and backward special school pupils (only 2 of whom had other abnormalities), 66% were boys. These findings suggest that the boys were likely to be ascertained at earlier ages than the girls, probably because their behaviour in school was more troublesome. However, the Edinburgh findings give no

support to the theory that females predominate amongst higher grade defectives.

### Summary

The following conclusions resulted from a survey of 406 mentally handicapped children (aged 7½-14½ years) in Edinburgh.

1. Prevalence of mental handicap was 1·13% of whom 0·50% were severely handicapped (IQ less than 50) and 0·63% were mildly handicapped (IQ 50-69). One-quarter of pupils in special schools were considered to be dull and backward (IQ 70 or over) rather than mentally handicapped.

2. Of 71 children with IQ levels below 30, 94% had other abnormalities in addition to mental handicap. Nearly one-half suffered from cerebral palsy. Of 147 children in the IQ range 30-54, over three-quarters had other abnormalities; 41% were mongols. Of 188 children in the IQ range 55-69, two-thirds had no other major defect apart from mental handicap. Of these over one-half came from poor or very poor homes and/or had parents of very low intelligence. Of 73 dull and backward children attending special schools, over 85% came from a similar environment.

3. In all clinical type groups (except Down's syndrome) and at all IQ levels there were more boys than girls.

4. Among the severely handicapped (IQ less than 55) genetic factors were thought to be causative in 36%; there was other evidence of origin in early foetal life in a further 20%; handicap was probably or possibly due to perinatal and postnatal factors in 13 and 9% and aetiology was uncertain in 22%.

Among higher-grade defectives (IQ 55-69) genetic factors were implicated in 6%; in a further 26% handicap may have originated in early foetal life; peri- and postnatal factors may have been

causative in 11 and 3%, and in 54% aetiology was uncertain.

A survey of this nature is dependent on the co-operation and assistance of many different persons in local authority medical and educational departments, and in hospital services.

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