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A Study of Clinical Diagnosis and Evaluation of Ability of Mentally Handicapped Children

Young Hyuk Lee and Chang Jun Coe

At The Institute of Handicapped Children, 1100 mentally handicapped children with the Griffiths Mental Development Scale (GQ) less than 80 were investigated during a 5 year period between January 1981 and December 1985 to determine the cause of their intellectual deficit, to diagnose their condition and to assess their ability. Of 1100 children, 776 (69.6%) were diagnosed clinically. Cerebral palsy was the most common cause (22.5%) followed by microcephaly (14.6%), seizure disorder (12.4%) and chromosomal anomaly (4.4%). Preventable disorders such as cerebral palsy, chromosomal anomaly, metabolic disorder and endocrinologic disorder were found in 314 cases (28.5%). It is emphasized that prenatal care, fetal monitoring, chromosomal study and a screening test for hypothyroidism and phenylketonuria are indeed required for prevention. When the Griffiths Mental Development Scale was used to assess the children's ability, 96.5% required medical and paramedical treatment such as education and training. In order to assess a mentally handicapped child completely, specialists from various fields are needed to work as a team in an assessment unit where knowledge from all fields can be concentrated. In addition, society as a whole should become more interested in these affairs.

Key Words: Mentally handicapped children, assessment unit

Opinions differ on the definition of mental retardation, but it is generally accepted that mental retardation is a condition of subnormal intellectual abilities or a deficit in learning and adaptation to society (Menkes 1985).

The prevalence rate is known to differ according to race, cultural and environmental status as well as cultural parameters for measuring intelligence. According to reports in the United States, the incidence of mental retardation in the preschool age group is 0.5%, while in the school age group it is about 1% and the total incidence is about 3% (Behrmann and Vaughan 1987; Swaiman and Wright 1982). There have been no reports on the incidence and epidemiology of mental retardation in Korea, but they are presumed to be similar to those of other countries.

Mentally handicapped children can provoke not only educational, social and economic problems but can also cause medical and psychological problems. As mental function is closely related to the function of the central nervous system, mental retardation has often been considered in connection with neurological abnormalities such as seizure, motor dysfunction or dysfunction of special sensory organs.

The purpose of this study was to define the causes of mental retardation, to evaluate the condition of mentally handicapped children, to help them by giving medical treatment or assistance such as hearing aids, braces or medication if needed, and finally, to offer a basis for better evaluation, further education and training.

MATERIALS AND METHODS

Of the children who visited The Institute of Handicapped Children between 1981 and 1985 for evaluation of delayed mental status, 1100 whose IQ was below 80 by Korea-WISC or Griffiths Mental Development Scale were selected for this study.

The parents of each child were interviewed in an attempt to obtain a detailed and reliable history of the family, pregnancy, delivery and early developmental status. Each child was physically examined for height, weight, head circumference and neurological

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Department of Pediatrics, Yonsei University College of Medicine, Institute of Handicapped Children
Address reprint requests to Dr. Y H Lee, Department of Pediatrics, Yonsei University College of Medicine, Seoul, Korea, 120-749

abnormalities by a pediatrician. Skull series, X-ray for bone age, brain computerized tomographic scan and electroencephalography were conducted when necessary. TORCH, hormonal (parathyroid, thyroid) and chromosomal studies were done when they were indicated. All children were screened for phenylketonuria by the urine ferric chloride test.

The children were divided into 5 groups according to their intelligence quotient as follows: borderline (70-80), mild retardation (55-70), moderate retardation (40-55), severe retardation (25-40) and profound retardation (less than 25). Children whose mental handicap was suspected to arise from psychiatric, sociopenvironmental and psychological problems were transferred to a psychiatrist, psychologist and social worker for better evaluation of their problems. Children who had problems in the fields of special sensory organs (speech, hearing and vision) were transferred to the specialists to evaluate to their problems.

After analyzing all data from various fields, we made plans for specific treatment, further evaluation and training. Periodic reassessment was done every 6 months for reevaluation of development of abilities and intelligence and effectiveness of education and treatment. We then made new educational and therapeutic plans according to the patients' individual handicapped status.

RESULTS

Among 1100 children, 723 were male and 337 were female and the male to female ratio was 1.9: 1. The age on admission was under 6 years in about 90% of the cases (Table 1).

Of 1100 children, 766 (69.6%) could be diagnosed clinically. Cerebral palsy was the most common diagnosis, for which the incidence was 22.5%. Other

Table 1. Age and sex distribution of mentally handicapped children

Age (year)	Male	Female	Total (%)
< 1	89	50	139 (12.6)
1 – 2	265	157	422 (38.4)
3 – 4	175	87	262 (23.8)
5 – 6	120	39	159 (14.4)
7 – 8	34	24	58 (5.3)
9 – 10	18	9	27 (2.5)
11 – 12	15	7	22 (2.0)
13 – 15	7	4	11 (1.0)
Total	723	377	1100 (100.0)

Table 2-a. Clinical diagnosis of mentally handicapped children

Clinical diagnosis	Number of cases (%)
Idiopathic	334 (30.4)
without delayed developmental milestone	232
with delayed developmental milestone	102
Cerebral palsy	248 (22.5)
without seizure	206
with seizure	42
Microcephaly	161 (14.6)
idiopathic	89
with cerebral palsy	55
with seizure disorder	10
with cerebral palsy & seizure disorder	7
Seizure related	136 (12.4)
infantile spasm	14
myoclonic seizure	40
dancing eye syndrome	4
HHE syndrome	4
other seizure	74
Chromosomal anomalies	49 (4.4)
Down syndrome	46
Turner syndrome	1
Klinefelter syndrome	1
XXX female	1
Endocrine & metabolic disorder	29 (2.6)
hypothyroidism	20
phenylketonuria	6
Wilson disease	2
gargoylism	1
CNS infection	19 (1.7)
meningitis	11
encephalitis	7
congenital syphilis	1
Phacomatosis	10 (0.9)
tuberous sclerosis	7
Sturge-Weber disease	2
neurofibromatosis	1

Table 2-b. Clinical diagnosis of mentally handicapped children

Clinical diagnosis	Number of cases (%)
CNS anomalies	9 (0.9)
agenesis of corpus callosum	5
meningomyelocele	2
moya moya disease	2
Funny fooking face syndrome	21 (1.9)
Prader-Willi syndrome	7
Crouzon's diséase	5
Treacher-Collins syndrome	2
Noonan syndrome	2
Pierre-Robin syndrome	1
de Lange syndrome	1
Aarskog syndrome	1
Larsen syndrome	1
Wolf syndrome	1
Post-Reye syndrome	8 (0.7)
Muscular disorder	5 (0.5)
progressive muscular disorder	3
myotonia dystrophica	1
familiar spastic paraplegia	1
Failure to thrive	4 (0.4)
Carbon monoxide poisoning	4 (0.4)
Hypoxic brain damage	2 (0.2)
Specific organ dysfunction	40 (3.6)
Developmental disorder & autism	21 (2.0)
Total	1100 (100.0)

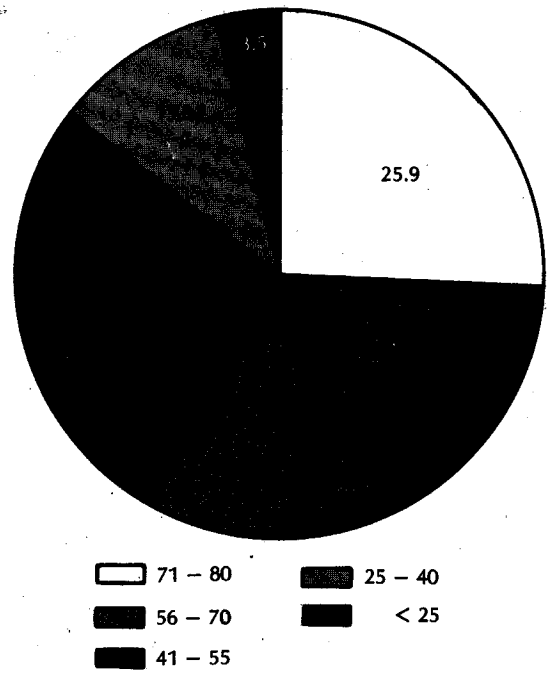

Fig. 1. GQ by Griffiths mental development scale.

causes were microcephaly (14.6%), hormonal diseases (2.6%), infection of the central nervous system (1.7%), phacomatosis (0.9%), anomalies of the central nervous system (0.9%), etc. Among 136 children with seizure disorder, 40 cases were myoclonic seizure and 14 were infantile spasm. Of 49 cases of chromosomal anomaly, the most common was Down's syndrome with 46 cases (Table 2).

We tested 1006 children by the Griffiths Mental Development Scale (GQ). The borderline group contained 25.9% of all cases, the mild, moderate, severe, profound groups contained 29.3%, 28.0%, 13.3%, and 3.5% respectively (Fig. 1).

The GQ test was divided into 6 items; locomotor, personal-social, hearing-speech, hand-eye coordination, performance and practical reasoning. A GQ scale below 80 comprised 83.2% of patients in locomotor, 94.0% in personalsocial, 93.4% in hearing-speech, 94.8% in hand-eye coordination, 93.3% in performance and 88.1% in practical reasoning (Fig. 2).

We performed brain C-T scanning to evaluate intracranial lesions associated with mental retardation in 441 cases. The C-T scan showed no gross abnormalities in 289 cases (65.5%), but in 152 cases (34.5%), we found abnormalities such as cortical atrophy, hydrocephalus, cerebral infarction, cerebromalacia, calcification, hemiatrophy, partial agenesis of corpus callosum, hydranencephaly, arachnoid cyst, porencephaly, and so on (Table 3).

All children were divided into 3 groups: preventable group, medical therapy group and special therapy group according to clinical diagnosis, neurologic assessment and therapeutic plans. Three hundred and forty one cases had preventable diseases such as cerebral palsy, chromosomal anomalies, hypopara-thyroidism, phenylketonuria, and failure to thrive. Four hundred and twenty-eight cases (38.9%) needed medical treatment such as anticonvulsants, muscle relaxants and hormonal replacement, etc. Physiotherapy was required in 501 cases (45.5%), speech therapy in 647 cases (58.8%), occupational

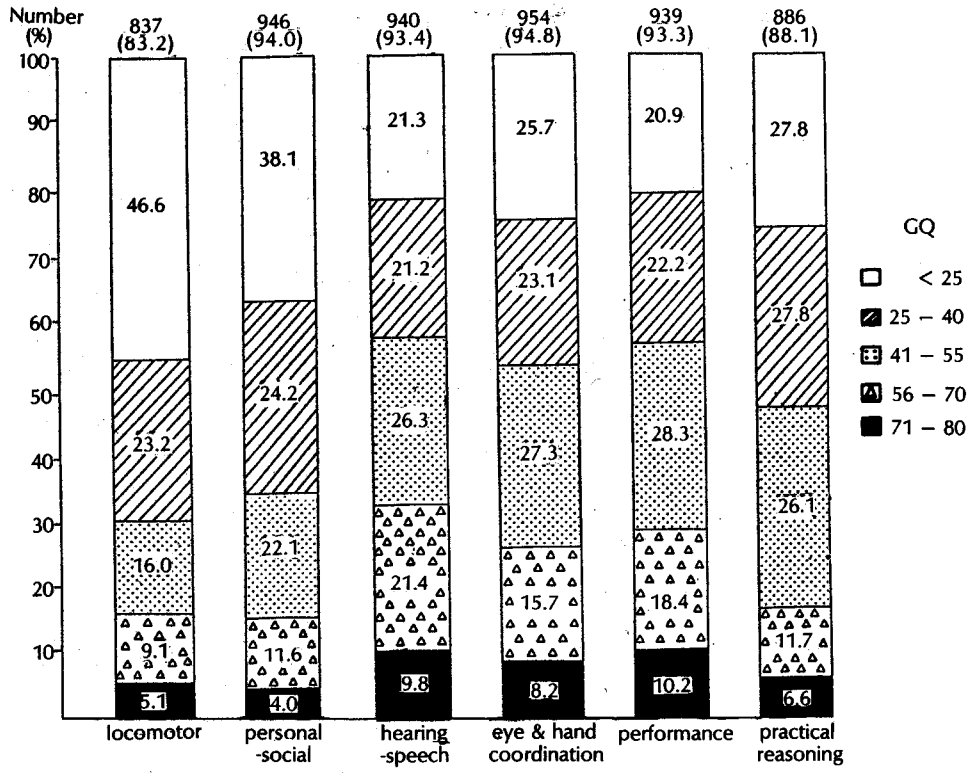

Fig. 2. Percentage of each scale in 1006 mentally handicapped children.

Table 3. Brain computerized tomographic findings in 441 mentally handicapped children

Findings	Numbers
Normal	289
Cortical atrophy	46
Hydrocephalus	31
Cortical atrophy & hydrocephalus	20
Infarction	16
Cerebromalacia	10
Calcification	7
Hemiatrophy	5
Agenesis of corpus callosum	5
Hydranencephaly	3
Arachnoid cyst	3
Porencephaly	3
Vascular malformation	2
Cerebellar atrophy	1
Total	441

Table 4. Therapies for mentally handicapped children

Therapy	%
Medical therapy	38.9
Special therapy	
Physiotherapy	45.5
Speech therapy	58.8
Occupational therapy	60.9
Special education	66.0
Hearing aids	2.4

therapy in 670 cases (60.9%), special education in 726 cases (66.0%) (Table 4.)

DISCUSSION

Mental retardation is difficult to define satisfactorily. Defined simply, a 'mentally handicapped person is one who does not grow up mentally to meet the average

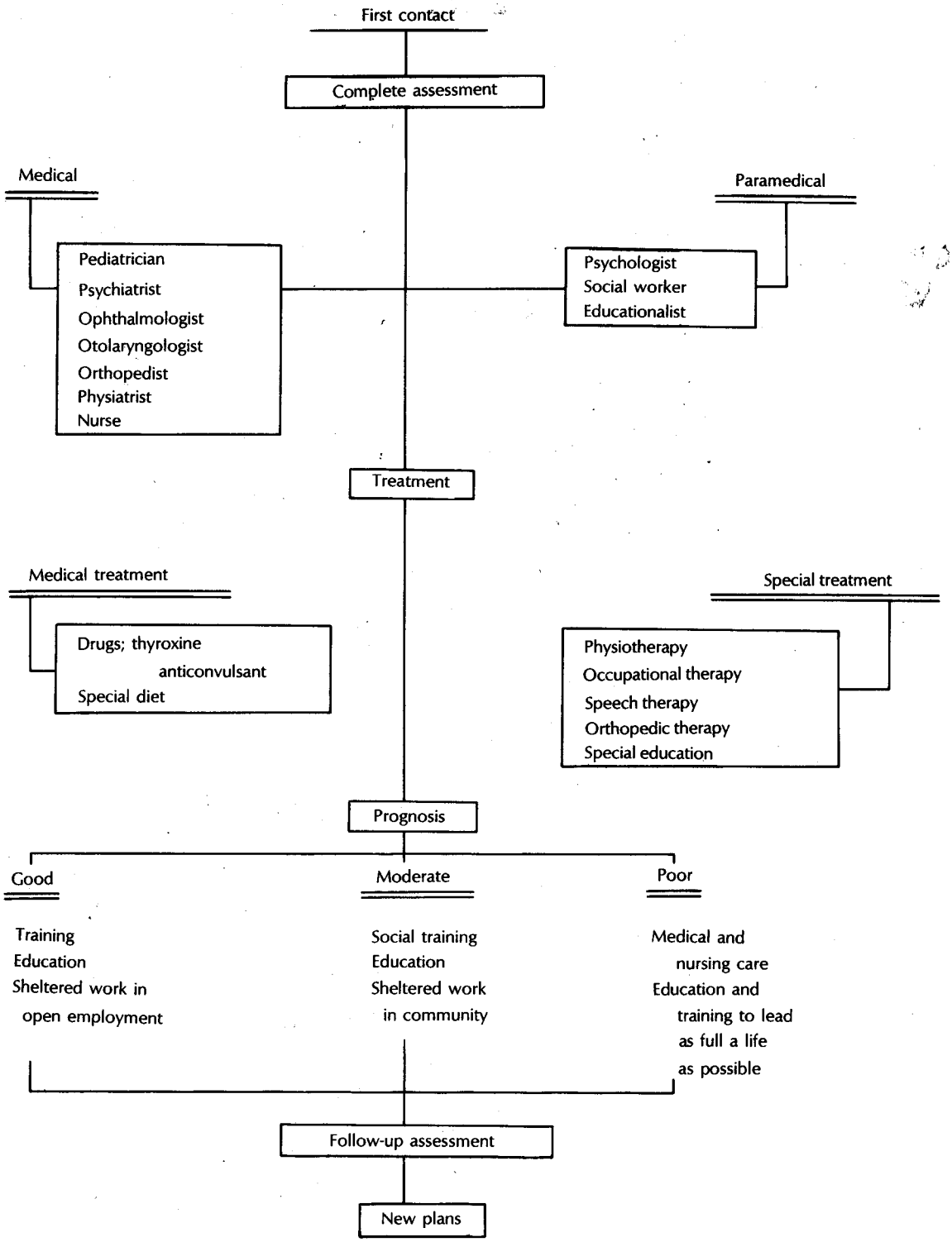

Fig. 3. *Organization of assessment unit for mentally handicapped children.*

standard' (Dutton 1975). But according to DSM-3 (Williams 1980) 'the essential features of mental retardation are significantly subaverage general intellectual functioning, resulting in or associated with deficits or impairments in adaptive behavior and with onset before the age of 18'. These definitions differ slightly, but the basic concepts are substantially the same.

Mental retardation may be defined in term of IQ when standardized tests for intelligence are used, but the measurement of intelligence has been regarded as defining general intelligence without regard to the separate processes of perception, analysis, memory interrelation and execution in the process of problem solving (Freeman, 1979). In terms of IQ, which is the ratio of mental age to chronological age expressed as a percentage, the usual tests have a mean of 100 with a standard deviation of 15 points so that the IQ range for normal person would be 70-130 IQ points. Those with an IQ of less than 70 are considered mildly mentally handicapped and less than 50 to be severely mentally retarded.

But there are some other opinions about mental retardation. Menkes (1985) asserted that an IQ of less than 75 indicates mental retardation because it is possible for an IQ to deviate ±5 points. Garber (1982) asserted that an IQ of 50-80 was cultural-familial, sociocultural or psychosocial retardation, so it must be included in mental retardation. Because of the difference in the definition of mental retardation by IQ, the borderline group (IQ=71-80) was included in this study.

In this study, 25.9% of all cases were in the borderline group, 29.3% were mildly retarded, 20.8% were moderately retarded, 13.3% were severely retarded and 3.5% were profoundly retarded. Of 1100 children, 96.5% required special education, training, physiotherapy, occupational therapy or medication, and 3.5% required institutional care.

Galton (1869) regarded mental retardation as a natural phenomenon parallel with variations in height or weight and as an essential feature in the nature of man. But Lewis (1933) investigated the causes of mental deficiency and recognized the fact that environmental effects played a role in the production of cerebral pathology. The conflict between these two approaches is still evident in scientific debates on the causes of mental retardation. Penrose (1972) pointed out that the numbers of idiots and imbeciles should be greater than Galton calculated, being the sum of natural variation and environmental effect.

The prevalence of mental retardation by Goodman and Tizard (1962) was 36 per 1000 in the age group 5-9 years, and 45.3 per 1000 in the age group 10-14 years. Abramowicz and Richardson (1975) said the prevalence rate of severe mental retardation is 0.54% of a population. There have been no reports on the prevalence and epidemiology of mental retardation in Korea, but it is presumed here that the prevalence here is probably similar to that of other countries.

The purpose of medical intervention in assessment is to define the causes of and to diagnose mental retardation. In our study, 69.6% of 1100 children could be given a clinical diagnosis. This is similar to the report of Crome and Stern (1973) who reported 65% and Turner (1975), 78%.

Among those diagnosed, cerebral palsy was the most common cause with an incidence of 22.5%. Other causes were microcephaly (14.6%), seizure disorders (12.4%), chromosomal abnormalities (4.4%), endocrine and metabolic disorders (2.6%), central nervous system infection (1.7%), and phacomatosis (0.9%). It is now widely known that certain disorders causing mental handicap can be recognized by prenatal tests and postnatal identification. These disorders are cerebral palsy, chromosomal abnormality, metabolic disorder and endocrinologic disorder. They comprised 341 cases in this study. Cerebral palsy and mental retardation overlap to some extent. About 50% of children with cerebral palsy have an IQ within the normal range. Of the remainder, about one half are severely retarded, and 10-20% of all severely retarded children have cerebral palsy. Eventually the prevention of cerebral palsy may reduce the incidence of mental retardation (Paneth and Stark 1983). Cerebral palsy rates increase directly with the severity of Apgar score of 3 or less is almost 300 times higher than that for infants with a 1 minute Apgar score of 7 or higher (Nelson and Ellenberg 1981). The relationship between low birth weight and premature delivery is very intimate. The risk of mental retardation for infants weighing less than 1500 gm at birth is about 9%, or 22 times that of infants of normal birth weight (Ellenberg and Nelson 1979; Gustavoson *et al.* 1977). Infants weighing 1500 to 2000 gm at birth have about a 3 or 4 times higher risk than that of larger infants. Asphyxia is also generally regarded as the major cause of disability. Quillingan (1976) has suggested that early recognition and elimination of fetal distress should reduce by one half the incidence of handicapping conditions or mental retardation. In this study, 27.5% were associated with cerebral palsy and it is one of the preventable disease (Opitz *et al.* 1977).

Prevention of mental retardation may be possible by treatment before the clinical manifestation of hypothyroidism. Arnold *et al.* (1981) reported that mental retardation was reduced to 3-4% among

hypothyroid patients detected by neonatal screening. Mental retardation from phenylketonuria also can be prevented by a low phenylalanine diet up to 8 weeks of age. In this study, of 304 mentally retarded children studied with the urine ferric chloride test, 6 were found to be afflicted by phenylketonuria. Thus, it might be necessary, in Korea as in other countries, to screen for early diagnosis of hypothyroidism, phenylketonuria, and other metabolic disorders in newborns.

We conducted chromosomal studies in 183 patients who had some morphologic abnormalities. We found chromosomal abnormalities in 49 cases out of 183 (26.8%). Down's syndrome was the most common; Turner's syndrome, Klinefelter's syndrome and XXX female syndrome were also found. Zellweger (1963) came to the conclusion that about 0.35% of the population had abnormalities of their chromosomes, either monosomy, trisomy or polysomy and that in at least two-thirds of such cases, mental retardation can also be found. He also calculated that one-fifteenth of all cases of mental retardation is due to chromosomal aberrations. Opitz (1979) reported that 154 among 1925 cases of mental retardation (8%) had a chromosomal abnormality in his study. In our study, the prevalence of chromosomal abnormality in mental retardation was 4.4%. But, if we had studied chromosomes more carefully, the incidence may have been higher. It is possible to detect chromosomal abnormality by amniocentesis during pregnancy. Thus, amniocentesis is necessary for the following criteria: maternal age above 35, positive family history of nervous system anomalies, parents who are carriers of a dominant syndrome or if one of the siblings has an abnormal chromosomal syndrome.

Mental retardation is closely related with neonatal sizure (Costeff *et al.* 1981). In general, fits occuring in the first few days of life carry a poor prognosis. Corbett *et al.* (1975) reported that 32% of children whose IQ was below 50 had a past history of neonatal seizure. Mental retardation is associated with infantile spasms and frequent myoclonic seizures. O'Donohoe (1976) reported that among 100 consecutive cases of infantile spasm, 21 were found to be normal physically and intellectually on follow-up, but 46% were severely abnormal intellectually, 27% had cerebral palsy and were usually severely mentally handicapped, and 19% were moderately or mildly subnormal. Aicardi (1973) reported that myoclonic epilepsy appearing before the age of 3 tends to have a higher rate and a more severe form of mental retardation than seizures with of onset after 3 years of age. Therefore, control of seizures with anticonvulsants is necessary, and in the case of infantile spasm, use of ACTH is necessary to prevent mental retardation. The authors used anticonvulsants in 178 cases with frequent seizues attacks to control seizures, and in this study, 14 cases were symptomatic infantile spasm patients who did not respond to treatment with ACTH.

The observation of speech and motor development in children with mental retardation of unknown origin is also important. Rutter and Martin (1972) reported that learning language expression was an important milestone in the developmental process and the delay suggested mental retardation. Hreidarsson *et al.* (1983) and Kaminer and Jedrysek (1983) reported that the start of walking in children with cognitive dysfunction was delayed when compared with the control group. With neural development, development of cognition and perception also occurs, and education and training before 4 years of age can lead to satisfactory results. Therefore when a child with delayed speech and motor skills is noted, it is important that the child be brought immediately to a specialist where he or she can be tested and early education and training can be initiated.

Computerized tomography was especially helpful in diagnosing intracranial lesions. In 34.5% of patients an abnormality was discovered by computerized tomography; cortical atrophy was noted in 46 cases, hydrocephalus in 31, cerebral infarction in 16, cerebromalacia in 10 and calcification in 7 cases. Cortical atrophy was mostly seen in patients with cerebral palsy (15 cases), seizure disorder (12 cases) and microcephaly (8 cases). Hydrocephalus was seen in mental retardation of unknown origin (8 cases), sequelae after meningitis (4 cases), and seizure disorder (5 cases). Among 20 patients with cortical atrophy and hydrocephalus, 14 were patients associated with cerebral palsy.

Four hundred and twenty eight patients needed specific medication, of whom 178 needed anticonvulsants, 159 muscle relaxants, 20 thyrotropic drugs, and 6 a low phenylalanine diet. At the time of diagnosis 649 patients (58.8%) needed speech therapy, 501 (45.5%) needed physiotherapy, 670 (64.9%) needed occupational therapy, and 726 (66.0%) needed special education. However, most of the patients could not receive proper treatment due to lack of therapists, lack of a facility for therapy, difficulty in bringing the patient to the hospital for therapy, economic problems and in particular, patients who lived in the countryside and those who lived far away from the hospital had difficulty in receiving proper treatment.

When assessing a mentally retarded child, the pediatrician, child psychiatrist, ophthalmologist, otolaryngologist, orthopedic surgeon, and physiatrist should work hand in hand with a psychologist, social worker and educationist in order to reach a proper diagnosis and to plan a therapy mode for each individual patient. Afterwards, medical therapy, diet control, physical therapy, speech therapy, occupational therapy and special education should be done by specialists in each field. Progress of the patient should be followed regularly for evaluation of development in ability, cognition and perception and effectiveness of education, and each patient should be analysed in order that a new education or treatment program may be set up.

At present, an assessment unit should be set up by centralizing the scattered treatment units, and the number of speech therapists, physiotherapists and occupational therapists as well as training centers should be increased.

Mentally handicapped children can not be diagnosed, assessed, educated by physician alone but requires a cooperative works with psychologist, teacher trained for special education as well as social workers as a team, it is because it is not the only educational problem but a social problem. The society as a whole should have better understanding for handicapped children.

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