

# An Epidemiological Study on the Cause of Mental Retardation in Yokohama City

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## **Abstract**

We investigated the cause of mental retardation in 337 individuals seeking healthcare services at the Yokohama City Social Welfare Center for Disabilities during a period of a year and a half, from October 1987 to March 1989.

The total number of participants in the study was 337 and consisted of 207 males and 130 females. Age ranged from 14 to 58 years with an average age of 22 years. IQ was evaluated using the Tanaka-Binet test. Individuals with an IQ under 50 were classified as having severe mental retardation and those with IQs between 51 and 70 were classified as having mild mental retardation, in accordance with international standards. The number of cases with severe mental retardation was 262 and 75 cases presented with mild mental retardation. Physical and neurological examination was performed in all cases in an attempt to ascertain the causes of mental retardation. Only when deemed necessary by the attending physician, and if parental consent was granted, electroencephalogram, brain CT scan, and chromosome analysis were carried out. In cases where the diagnosis had already been established, past medical records were consulted. The

results of the study were compared to results of a very similar study published by the University of Kuopio in eastern Finland.

The time of insult was categorized into four groups: prenatal, perinatal, postnatal and uncertain. In the severely mentally retarded group, the time of insult was considered to have occurred in the prenatal period in 25.6% of cases, in the perinatal period in 9.2% of cases, in the postnatal period in 8.0% of cases and was uncertain in 57.2% of cases. In the mildly mentally retarded group the insult occurred in the prenatal period in 14.7% of cases, in the perinatal period in 8.0% of cases, in the postnatal period in 12.0% of cases, and was uncertain in 65.3% of cases. In the mildly mentally retarded group, familial association was recognized in 27% of cases.

The Finland study revealed similar patterns. Both studies demonstrated that most individuals with severe mental retardation and a determinable cause have genetic and chromosomal abnormalities. The cause of mental retardation was harder to determine in individuals with mild mental retardation, and most of those with an uncertain cause had a positive family history. The above findings point out the need for

prenatal diagnosis and genetic counseling in certain cases, requiring careful attention to ethical issues.

Keywords: mental retardation, epidemiology, causes, Yokohama City, international, comparison

## **I. Background**

Mental retardation is a generic name for intellectual disorders with highly variable causes. Several studies are currently investigating the causes for these disorders. However, most are based on in-patient populations and very few study causes in a specific zone.<sup>1) 2) 3) 4)</sup>

We investigated the cause of mental retardation in 337 individuals in Yokohama City. Results are compared to those obtained by a similar study in another zone.

## **II. Participants and Methods**

Participants included 337 individuals (207 males and 130 females) with mental retardation seeking healthcare services at the Yokohama City Social Welfare Center for Disabilities during a period of a year and a half, from October 1987 to March 1989. Age ranged from 14 to 58 years with an average age of 22 years.

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between 51 and 70 were classified as having mild mental retardation, in accordance with international standards. The number of cases with severe mental retardation was 262 and 75 cases presented with mild mental retardation. Physical and neurological examination was performed in all cases in an attempt to ascertain the causes of mental retardation. Only when deemed necessary by the attending physician, and if parental consent was granted, electroencephalogram, brain CT scan, and chromosome analysis were carried out. In cases where the diagnosis had already been established, past medical records were consulted. The time of insult was categorized into four groups: prenatal, perinatal, postnatal and uncertain. These groups are further described below.

## **III. Results**

### **1. Prenatal causes (severe)**

The cause of severe mental retardation was determined in 67 cases, corresponding to 25.6%. Genetic or chromosomal anomalies were the most frequent cause, found in 46 individuals, 68.6%. Mental retardation associated to malformations, considered to be linked to the neurological problem, was present in 18 individuals, 26.9%. There were also 2 cases of fetal alcohol syndrome (Fig. 1).

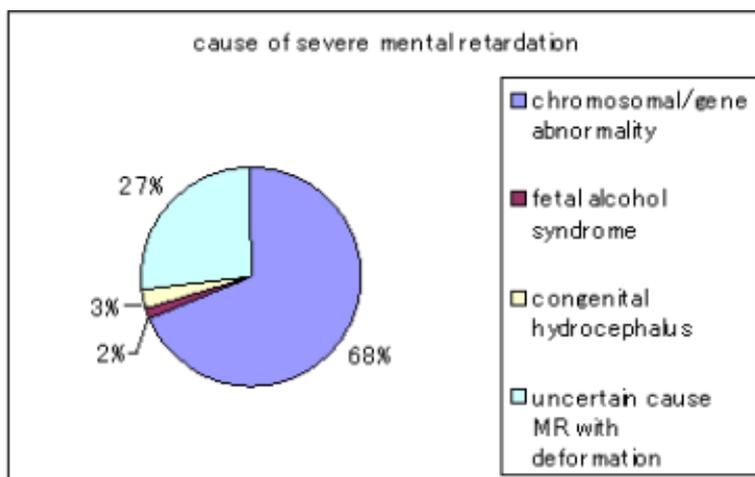
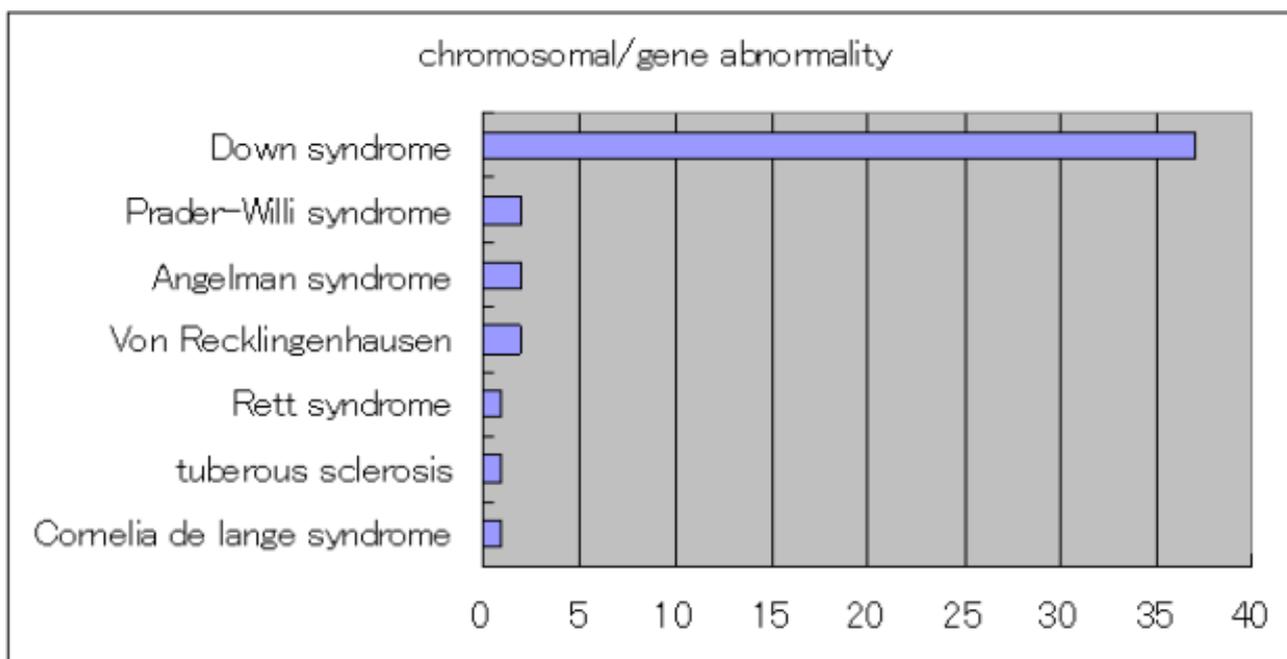


Fig. 2 lists the genetic and chromosomal anomalies found prenatally. Down syndrome stands out with 37 affected individuals, accounting for 80.4% of genetic and chromosomal anomalies. Two cases each of Prader Willi, Angelman and Von Recklinghausen syndrome were encountered. Most cases of the genetic and chromosomal anomalies shows severe mental

retardation. In particular, all cases of Down's syndrome show severe mental retardation. But in the cases of Down syndrome, the discrepancy between higher level of social skill and low IQ. In mild mental retardation cases only 2.6% presented with genetic or chromosomal anomalies

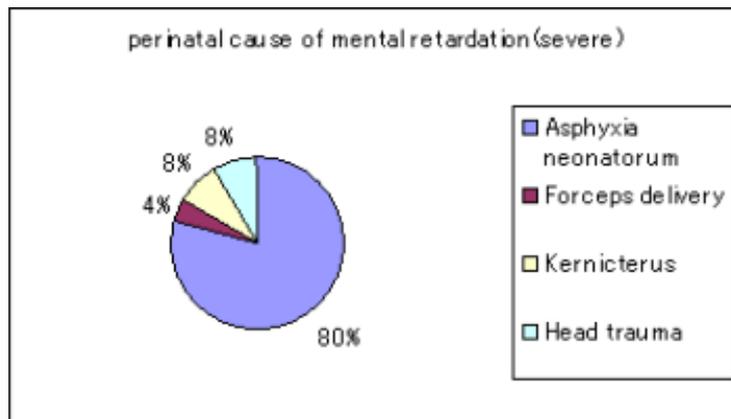


**2. Perinatal causes (severe)**

This group included patients with 1-minute and 5-minute Apgar scores under 6 and signs of mild cerebral palsy, and included 19 individuals, 79.2%. There is no agreement as to the degree in which

perinatal conditions cause mental retardation. For this reason, other 20 cases with perinatal conditions such as low birth weight, prolonged labor, premature rupture of membranes, and umbilical cord loops were not included in this group but under uncertain.(Fig.3)

Fig.3

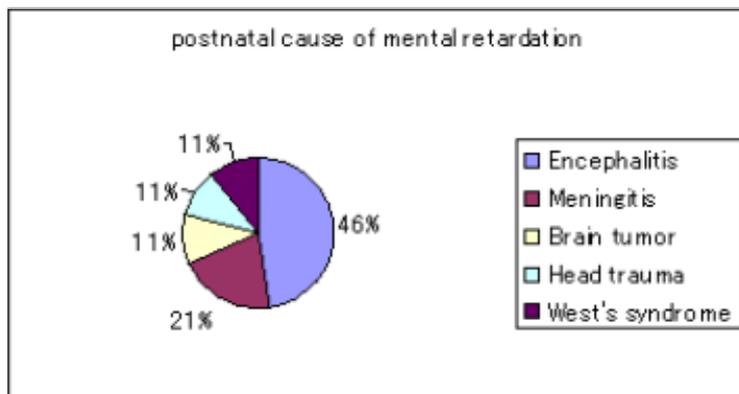


**3. Postnatal causes**

Encephalitis in 9 cases and meningitis in 4 cases were the most common causes, followed by blood group

incompatibilities, brain tumors and head trauma (Fig. 4).

Fig.4



#### 4. Summary

Table 1 summarizes the results of the study and the classification system for mild mental retardation causes. As mentioned earlier regarding prenatal causes, genetic and chromosomal anomalies are the most common

cause of severe mental retardation. Perinatal causes of mild and severe cases of mental retardation are led by asphyxia neonatorum. Encephalitis was the most common postnatal cause of mild and severe mental retardation.

Table 1. Causes of Mental Retardation (Yokohama)

Time of onset	Total MR (n=337)	SMR (n=262)	MMR (n=75)
<b>Prenatal</b>	<b>78</b>	<b>67</b>	<b>11</b>
Genetic or chromosomal anomalies	48	46	2
Fetal alcohol syndrome	2	1	1
Congenital hydrocephalus	2	2	0
Uncertain Cause MR with malformations	26	18	8
<b>Perinatal</b>	<b>30</b>	<b>24</b>	<b>6</b>
Asphyxia neonatorum	23	19	4
Forceps delivery	2	1	1
Kernicterus	2	2	0
Head trauma	3	2	1
<b>Postnatal</b>	<b>30</b>	<b>21</b>	<b>9</b>
Encephalitis	13	9	4
Meningitis	5	4	1
Blood group incompatibility	4	3	1
brain tumors	2	2	0
Head trauma	3	2	1
Postvaccinal encephalosis	2	0	2
West's syndrome	1	1	0
<b>Uncertain</b>	<b>199</b>	<b>150</b>	<b>49</b>
Gestosis	5	3	2
Low birth weight	7	5	2
Other perinatal conditions	13	9	4
others	174	133	41

#### IV. Discussion

A representative study of the causes of mental retardation centered on a specific zone is the study by Harberg et al. in Sweden <sup>2)</sup>. However, it was completed long ago and the study considered low birth weight and mild asphyxia as perinatal causes. Criteria for perinatal causes were unclear, resulting in perinatal causes being overvalued. For these reasons, the Harberg study wasn't used for comparison, using instead the study by Matilainen et al. <sup>6)</sup>. The Matilainen study in Finland detected 178 children with mental retardation in a group of 13.000 children between the ages of 8 and 9 years, using various test batteries. Of these 178 children, 151 agreed to be studied and were included. Those with an IQ under 55 were classified as having severe mental retardation and those with IQs between 56 and 70 were classified as having mild mental retardation. The severe mental retardation group included 46 males and 31 females, and the mild mental retardation group included 37 males and 37 females.

All cases in the Finland study underwent chromosomal analysis and electroencephalogram. Although brain CT was not available, similar exams were included. The classification of causes was also similar and was therefore considered appropriate for comparison. Table 5 shows the result of the comparison of the severe mental retardation group in the two studies. Both studies show very similar results for the perinatal and postnatal periods. However, our study determined prenatal causes in 25.6% of cases while the Finnish study determined them in 59.7% of cases.

The differing results may very well be due to the difference in age groups. In our study the average age was 22, while in the Finnish study it was between 8 and 9. This means the Finnish study, especially in the

severe mental retardation group, included such genetic or chromosomal anomalies as Trisomy 18, Smith-Lemli-Opitz syndrome, and others at high risk for young death such as ceroid lipofuscinosis.

Regarding mortality of mentally retarded individuals under the age of 20, those with an IQ under 35 have a 7-fold increased as compared to healthy individuals, and those with an IQ under 20 have a 31-fold increase <sup>7)</sup>. This means that individuals with a prenatal cause, who typically present with severe mental retardation, have a high mortality rate. The Finnish study includes individuals with severe mental retardation and a prenatal cause who would have died before they reached the average age of our study.

A second reason for discrepancy must be mentioned. The Finnish study found 4 cases of fragile X syndrome, accounting for 11% of genetic and chromosomal anomalies. Our study found no such cases. According to Nahba <sup>8)</sup>, fragile X syndrome frequency is race-dependant. Its frequency in Europe and America is 1 / 2500, while in Japan it is 1 / 10000. This might also explain the different results in these studies.

We will now compare the mild mental retardation groups in these studies. Both studies showed that the proportion of cases with an uncertain cause was higher than in the severe mental retardation group. Many of them suffer familial retardation, adding up to 27% in our study and 25% in the Finnish study. Many uncertain cause cases in the mild mental retardation group have strong hereditary factors as a possible cause. A study of an affected family by Farag <sup>9)</sup> showed that Mendelian inheritance was involved in 34% of cases. Therefore, if a complete study were performed including individuals with mild mental retardation, the familial inheritance rate should increase (Fig. 6).

Fig.6-1.Time of onset(severe mental retardation)

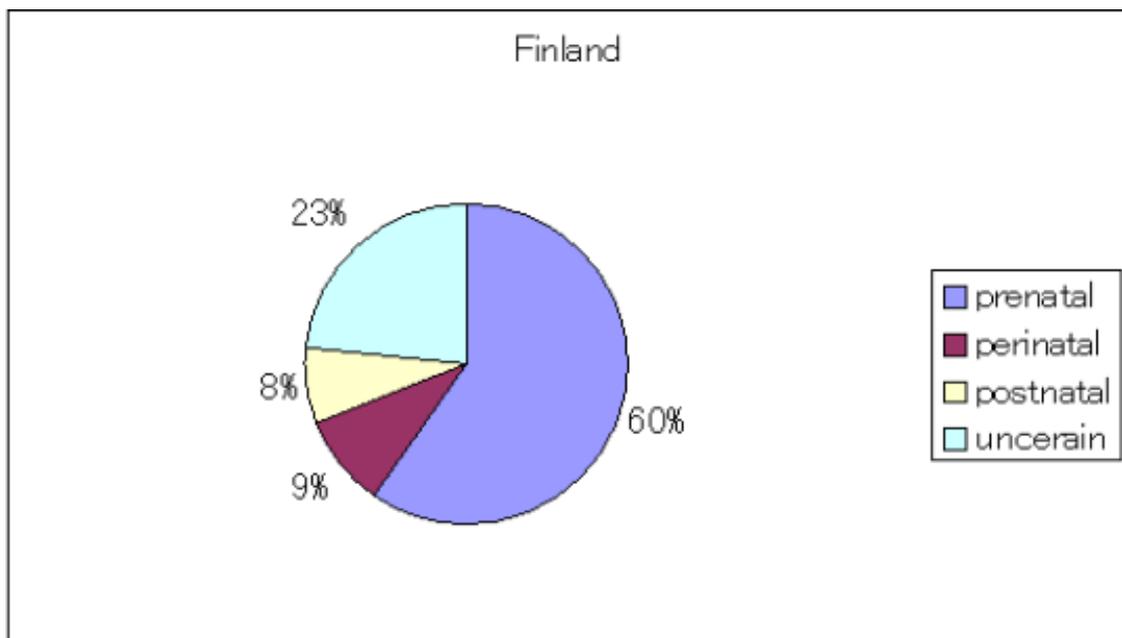
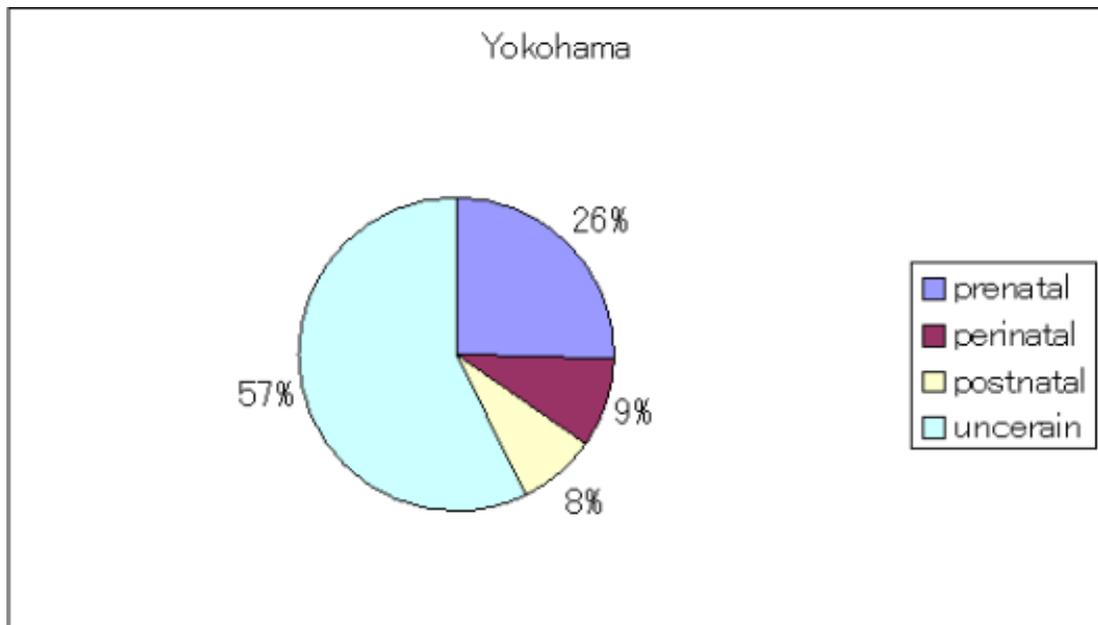
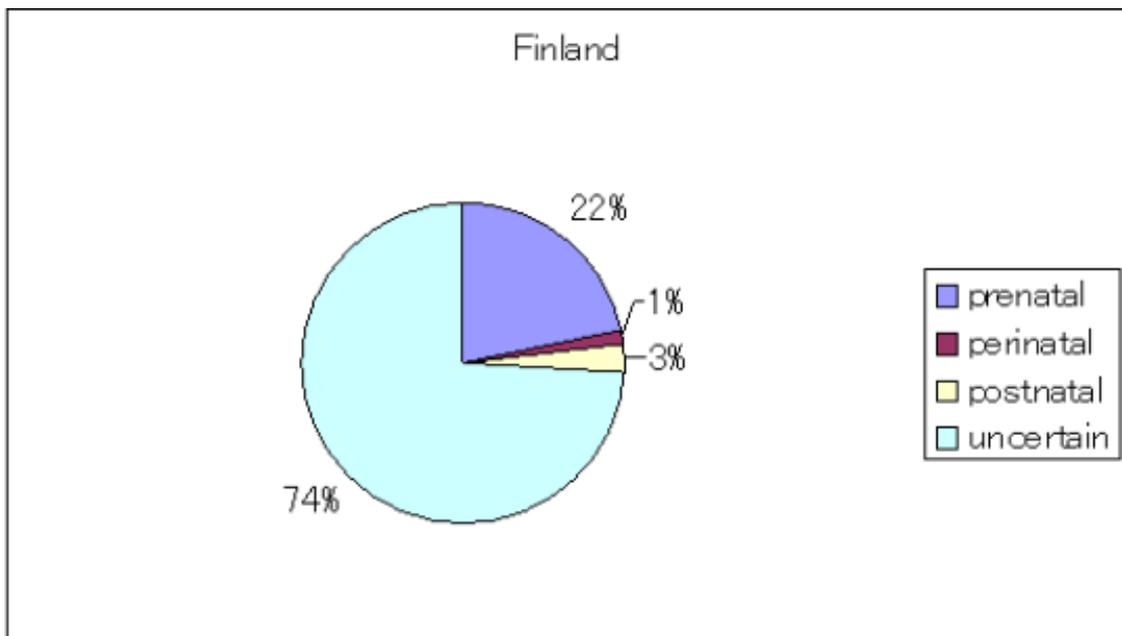
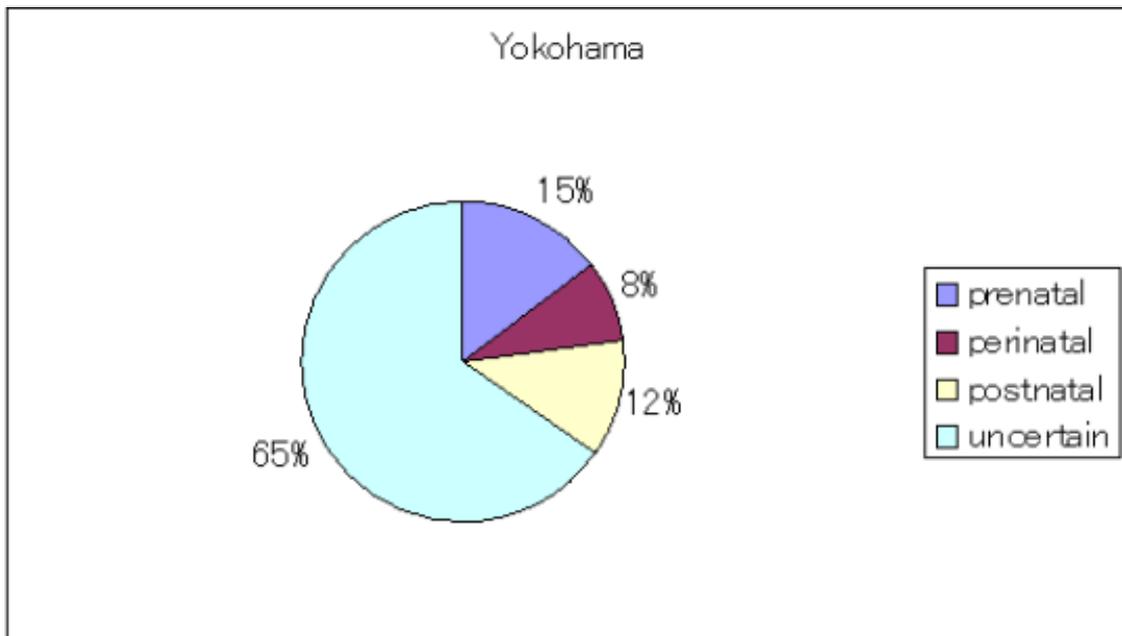


Fig.6-2.Time of onset(mild mental retardation)



We will now discuss the issue of the perinatal period criteria for cause classification. Low birth weight and asphyxia neonatorum are common conditions, but there is no agreement on whether low birth weight causes mental retardation or not. Low birth weight (under

2500g) wasn't included as a perinatal cause in either study.

According to Stewart (10), 4.6% of children with birth weight under 1000g will have an IQ under 70, 12% will have speech deficiencies, 14% will be borderline

mental retardation, 17% will have learning disabilities, and many will have a diminished hearing capacity.

Children with birth weight under 1000g who survive have a 10 to 35% risk of having a chronic disease and the possibility of this being a cause of mental retardation cannot be completely denied.

The development of neonatal medicine in recent years has enable studies to demonstrate that children with birth weight under 2500g usually recover from their initial delay and grow up healthy and normal.

Thus, considering low birth weight (under 2500g) is not currently considered a cause of mental retardation, nor a perinatal disease, it isn't included as a cause in this study.

## V. Conclusions

An epidemiological study of individuals with mental retardation in Yokohama City was carried out and compared to a similar Finnish study.

The difference between our study and the Finnish study is considered to arise from the dissimilar participant age average, and fragile X syndrome frequency. Individuals with severe mental retardation with a known cause were mainly affected by genetic or chromosomal anomalies. Individuals with mild mental retardation with an unclear cause showed a familial trend, also found in the Finnish study. The above findings point out the need for prenatal diagnosis and genetic counseling in certain cases, requiring careful attention to ethical issues.

Regarding mental retardation cause studies, an international classification system is lacking, and efforts should be made to create a detailed classification system.

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