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STUDY ABOUT THE INCIDENCE OF HEARING-SPEAKING DISORDERS IN A POPULATION WITH MENTAL DEFICIENCY

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ABSTRACT (online version)
This study is about the incidence of hearing-speaking disorders in a population with mental deficiency. We studied 596 children interned in Neurology and Psychiatry Clinical Hospital of Oradea during the 1999 - 2001 period. In 596 children, 393 presented different types of mental deficiency. The most frequent disorders observed are hearing loss or deafness, deaf-mutism, mutism and speaking retardation. Also, we related an increased frequency in rural area and in group of children with severe mental deficiency.
KEY WORDS: *Hearing-speaking disorders,hearing loss, mental deficiency.*

INTRODUCTION

Mental retardation is an idea, a condition, a syndrome, a symptom and a source of pain to many families. Its history dates back to the beginning of man's time on earth (Sheerenberger, 1983).

Mental retardation occurs in 2,5-3% of the general population (Teodorescu, 1996). About 6-7,5 million mentally retarded individuals live in the United States alone (American Association on Mental Retardation, 2002). This illness is defined as IQ score below 70-75. adaptive skills are the skills needed for daily life. Such skills include the ability to produce and understand language, home-living skills, use of community resources, health, safety, reading, writting and arithmetic skills and work skills (Bryant et. al., 1996; Lubetsky et. al.,1996).

Studies estimating the prevalence of mental health disorders among the individuals with mental retardation suggest that between 10 and 40% meet the criteria for dual diagnosis of mental retardation and mental health disorder. Nevertheless, individuals with mental retardation appear to display the full range of psychopathology evidenced in the general population. Individuals with mild cognitive limitations are more likely to be given a dual diagnosis that children with more significant disabilities. In diagnosing infant, it is important to distinguish between mental retardation and developmental delay.

Mental retardation is a developmental disability that first appears in children under the age of 18. It is defined as an intellectual functioning level (as measured by standard tests for intelligence quotient) that is well below average and significant limitations in daily living skills (or adaptative functioning). Many syndroms which are caused by the chromosomes, including Down, Patau and Turner syndromes, have associated deafness.

Deafness is determined in many cases by genetic causes. For example, it is estimated that one of 1000 children has hereditary deafness. In 50% of cases, is a genetic cause, but is important to mention that in different families can occur diverse types of inheritance.

Emery & Rimoin (1990) related that in 43,5% of cases the inheritance of deafness is autosomal recessive, in 6% is autosomal dominant and in 0,5% is X-linked recessive. They mention that environmental factors are responsable for 30% of congenital deafness and 20% of cases have idiopathic causes.

Deafness can be classified in many categories. In accordance to etiological criterion, the deafness possibly be hereditary and obtained. In accordance to the gravity, deafness or hearing loss has different degrees: mild, moderate, severe and profound. Also, in accordance to the localization, deafness is unilateral or bilateral. If we mention the affected segment, hearing loss possibly be conductive, sensoneural and mixed.

Hereditary hearing loss possibly be syndromic or non-syndromic. In a syndromic form of deafness, the diagnosis is simple to elaborate because of others congenital abnormalities associated. The Dysmorphology Laboratory from London related that more than 396 plurimalformative syndromes have associated deafness (Schaffer, 1995). Non-syndromic deafness occurs in two third part of cases and it isn't associated with congenital abnormalities.

Before showing the results of this study, it is probably worth mentioning that there is a group of syndromes that in themselves are rare disorders, but in which hearing loss is a consistent and/ or major feature and thus represent a significant group of conditions when one discusses hereditary hearing loss. A list of such conditions would include the oto-palatal-digital syndromes, the oral-facial-digital syndromes, skeletal dysplasias (in particular, osteogenesis imperfecta), metabolic storage disorders (especially mucopolysaccharidoses and Refsum's disease), Townes-Brock syndrome and Wildervanck syndrome.

MATERIAL AND METHOD

We investigated 596 children interned in Neurology and Psychiatry Clinical Hospital of Oradea between 1999 and 2001 period. The methods utilised were cytogenetic, clinical investigations, somatometrical, statistical, psychiatric investigation and hearing tests, too. There were realised family investigations and were constructed pedigrees.

RESULTS AND DISCUSSIONS

General aspects

We recorded 50 cases of hearing-speaking disorders in studied population, which means a frequency of 8,39%. Eight cases (16%) of all cases recorded, have no mental deficiency associated.

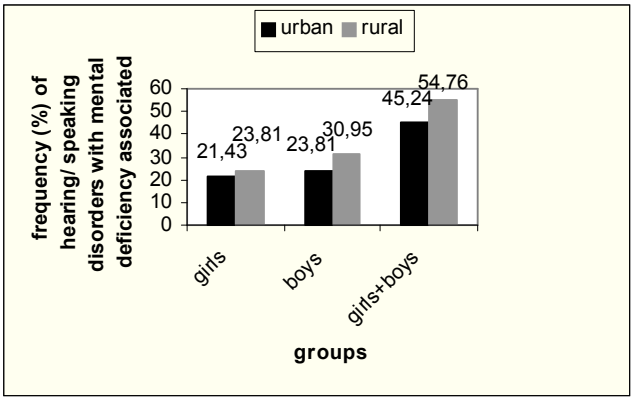

FIG. 1. Plotting of frequency of hearing-speaking disorders with mental deficiency associated in studied population.

There are 42 cases with mental deficiency associated; 16 (38,1%) of those associate mild mental retardation, 12 (28,57%) have medium mental retardation joined and 14 (33,33%) associate severe degree of mental deficiency.We can see in figure 2 the increase frequency of speaking retardation and hearing loss in children with mental deficiency group. The distribution of cases in accordance to the type of mental deficiency associated is related in tables 1, 2 and 3.

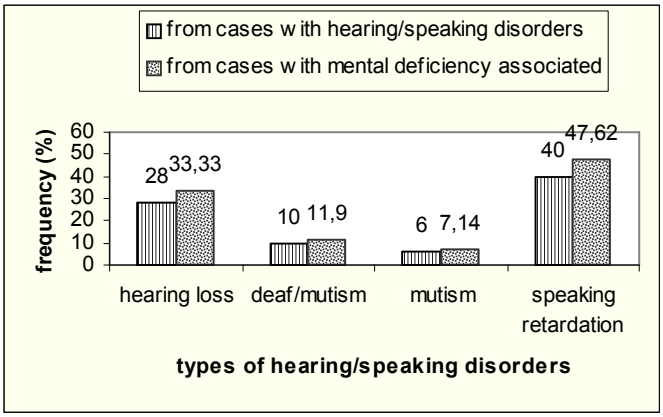

FIG. 2. Plotting the frequency of different types of hearing-speaking disorders associated with mental deficiency in studied population.

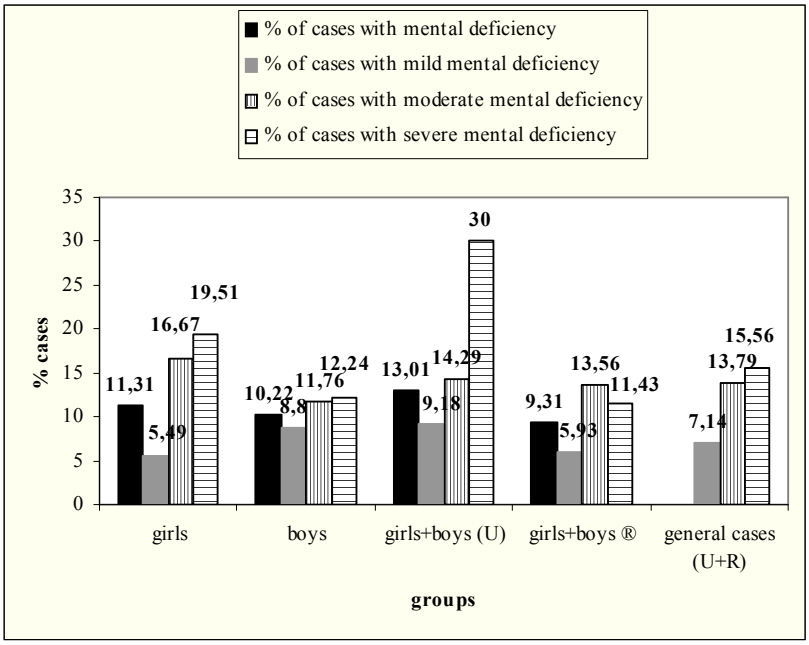

FIG. 3. Plotting the frequency of hearing-speaking disorders cases in studied groups of children with different types of mental deficiency associated.

TABLE 1. Representation of the frequency of hearing-speaking disorders cases with mild mental deficiency associated.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
4	25,0	1	6,25	5	31,25	6	37,5	16
U+R		%		U+R		%		Total %
5		31,25		11		68,75		100
U+U= 9 = 56,25% R+R= 7 = 43,75%								

TABLE 2. Representation of the frequency of hearing-speaking disorders cases with moderate mental deficiency associated.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
2	16,67	4	33,3	2	16,6	4	33,3	12
U+R		%		U+R		%		Total %
6		50,0		6		50,0		100
U+U= 4 = 33,33% R+R= 8 = 66,67%								

TABLE 3. Representation of the frequency of hearing-speaking disorders cases with severe mental deficiency associated.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
3	21,43	5	35,7	3	21,4	3	21,4	14
U+R		%		U+R		%		Total %
8		57,14		6		42,86		100
U+U= 6 = 42,86% R+R= 8 = 57,14%								

In figure 3 is represented the distribution of hearing/ speaking disorders cases who have associated mental deficiency. The results in this figure show an increase of frequency of hearing/ speaking disorders in groups with moderate and severe mental deficiency. In severe mental deficiency children group, we observed an important increase of cases frequency who proceed from urban area. Apparently, we could suppose the urban agglomeration for explain the cause, but if we follow the hearing loss

frequency in the same group, we can assume a genetic luggage. The inheritance can be autosomal dominant or recessive, or perhaps an X-linked inheritance.

Bridge (1997) calculated the recurrence risk. If there is in a family an individual affected by hearing loss, the risk of recurrence is 1/ 20. If the child of hearing loss affected individual is deaf too, we can consider an autosomal dominant inheritance and the risk will be about 50%. Hearing / speaking disorders are frequently observed in groups with mental deficiency. These disorders possibly be one of the mental deficiency traits or many occur isolated in population. Also, speaking retardation possibly occur because of mental deficiency and hearing loss, too.

Hearing loss

TABLE 4. Representation of the frequency of hearing loss cases in studied population

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
8	47,1	1	5,88	3	17,6	5	29,4	17
U+R		%		U+R		%		%
9		52,94		8		47,06		100
U+U= 11 = 60,70% R+R= 6 = 39,30%								

In studied population, we observed 17 cases with hearing loss. These 17 cases represent 34% of all cases of hearing/ speaking disorders. Of these 17 cases, 14 have associated different types of mental deficiency, which means 28% of all cases with hearing/ speaking disorders and 33,3% of cases with mental deficiency associated. Six cases associated mild mental retardation, 6 cases have severe mental deficiency and 2 cases associated moderate mental deficiency.

Is important to mention that girls associated mental deficiency more often than boys.

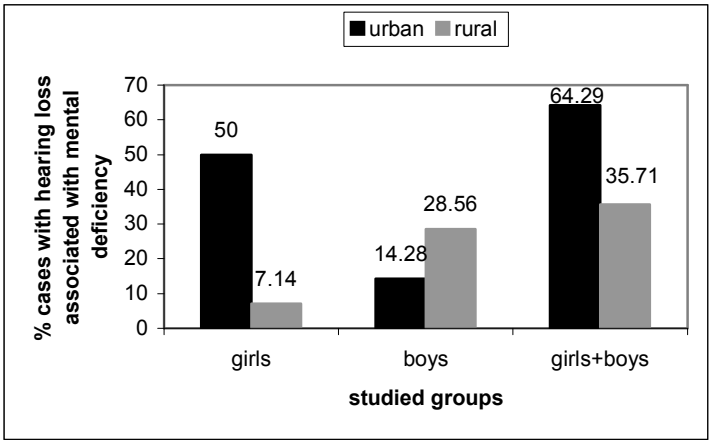

FIG. 4. Plotting the frequency of cases with hearing loss associated with mental deficiency in studied population.

TABLE 5. Representation of the frequency of hearing loss cases associated with mild mental deficiency.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
4	66,6	0	0	0	0	2	33,3	6
U+R		%		U+R		%		%
4		66,67		2		33,33		100
U+U= 4 = 66,67% R+R= 2 = 33,33%								

TABLE 6. Representation of the frequency of hearing loss cases associated with severe mental deficiency.

FETE				BĂIEȚI				Total
U	%	R	%	U	%	R	%	U+R
2	33,3	1	16,6	2	33,3	1	16,6	6
U+R		%		U+R		%		Total %
3		50,0		3		50,0		100
U+U= 4 = 66,67% R+R= 2 = 33,33%								

There were observed 2 cases of hearing loss who associate moderate mental deficiency.

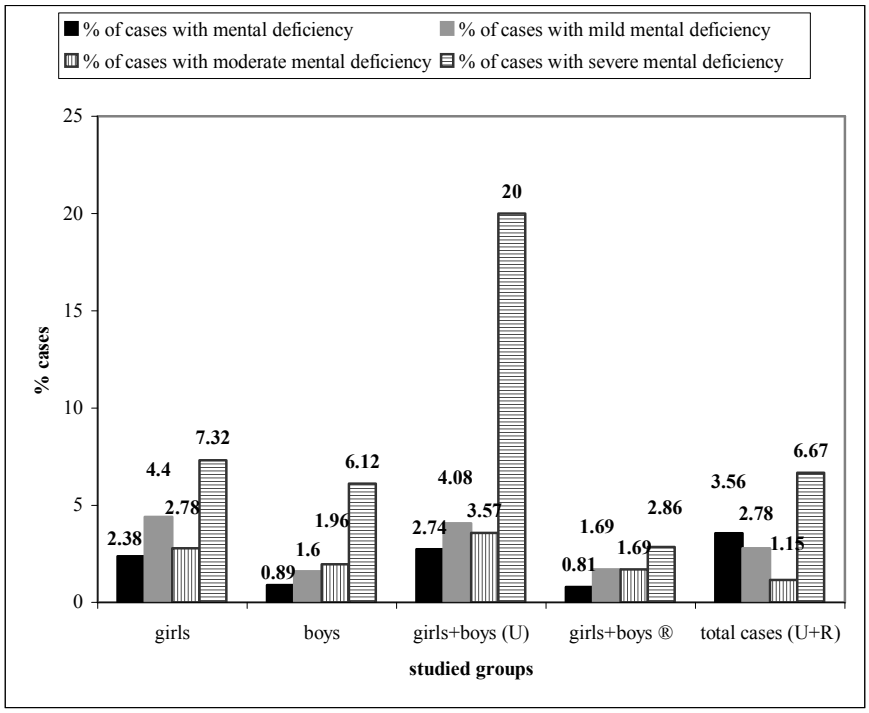

FIG. 5. Plotting the frequency of hearing loss cases with different types of mental deficiency associated in studied population.

In connection with the frequency of hearing loss associated with mental deficiency, we oserved an increase of cases who associate severe mental deficiency. The explanation of this result may be the chromosomal, non-chromosomal or gene disorders which caused the mental deficiency. Respective disorders determine hearing loss, too. The explanation may be also, that chrosomal or gene disorders which determine hearing loss cause the mental deficiency, too. Also, in some genetic disorders (such as Down syndrome, Turner syndrome) the pacient may has the propensity to ear infections, which possibly determine hearing loss. There are some syndromes who develop hearing loss, such as Alport syndrome, Waardenburg syndrome, treacher-Collins syndrome or Pendred syndrome.

In connection with affected sex, we didn't observe differents between the frequencies in girls and boys groups, which means that hearing loss most often is caused by an autosomal inheritance.

The results obtained are in concordance with those related by Amoaşii & Reaboi (2002), Diaconescu et al. (2002) and Marchian (2002, a, b). Marchian (2002, b) studied 2300 children affected by hearing loss who proceed from all areas from our country, and obtained a frequency of 26,8% for congenital hearing loss (which confirms the autosomal inheritance in most cases). It is important to mention the agglomeration of cases with congenital hearing loss in Pietroasa village from Bihor county. It's possibly to be a concentration of some genes responsable for a recessive inheritance.

Also, some authors demonstrated the hypothesis in concordance with palate cleft possibly determined hearing loss (March of Dimes, 2002).

Deaf-mutism. In studied population, we observed 5 cases (which means 0,84% of observed cases). These cases represent 1,27% of 393 children who associated mental deficiency. We also observed that all cases with deaf-mutism have associated mental deficiency. It seems that mutant gene determined either mental deficiency and deaf-mutism. Most cases (3 of 5) proceed from urban area. We observed one case who has associated mild mental deficiency, one case associated moderate mental deficiency and three cases associated severe mental deficiency.

Because of few number of deaf-mutism cases observed, we can't interpret with accuracy the results.

TABLE 7. Representation of the frequency of cases with deaf-mutism in studied population.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
1	20	1	20	2	40,0	1	20	5
U+R		%		U+R		%		%
2		40,0		3		60,0		100
U+U= 3 = 60,0% R+R= 2 = 40,0%								

Next figure represents the genealogical tree of a family with many cases of deaf.

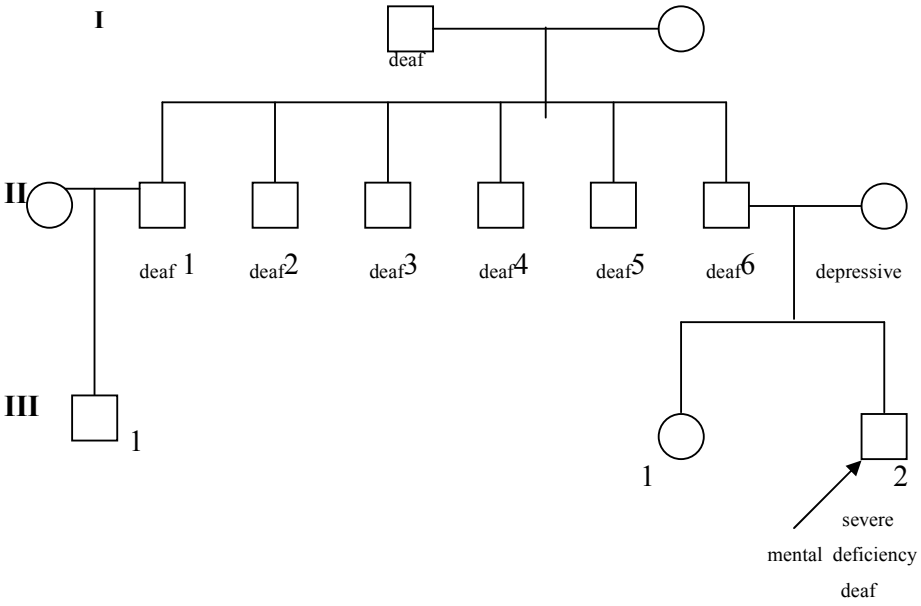

FIG. 6. Genealogical tree of D. family.

D. family is made of parents and two children. The princeps case is a boy, in age of 10, who has severe mental deficiency associated with cranial and facial abnormaties, dentition and jaw abnormalities and deaf-mutism. He has a healthy sister. It is important to mention that his mother was 39 years old when get birth this boy and she is depressive. Also, father of this child is deaf, such as his 5 brothers (boy's uncles) and father (boy's grandfather). It is interesting that his cousin (child of one of the uncles of sick boy) is healthy. We assume in this family an autosomal dominant inheritance. Because of absence of mental deficiency cases in this family, we we possibly explain the appearance of described case as a result of structural chromosomal disorder (including X fragil syndrome) or gene disorder. Either of gene disorder and chromosomal disorder could cause mental deficiency. Also, it's possibly another explanation for associated mental deficiency of this child: the antidepressive drugs administrated to mother during the pregnancy period.

Speaking retardation. In studied population we observed 25 cases of speaking retardation. Of course, there were more than these, but some of the children investigated are too young to cooperate when we examined them. The cases observed means 4,19% in studied population. In 25 cases, 20 associated mental deficiency. Most of cases proceed from rural area (tables 8 and 9).

TABLE 8. Representation of the frequency of cases with speaking retardation in studied population.

GIRLS				BOYS				Total
U	%	R	%	U	%	R	%	U+R
1	4	8	32	8	32	8	32	25
U+R		%		U+R		%		%
9		36,0		16		64,0		100
U+U= 9 = 36,0% R+R= 16 = 64,0%								

TABLE 9. Representation of the frequency of speaking retardation cases with associated mental deficiency.

FETE				BĂIEȚI				Total
U	%	R	%	U	%	R	%	U+R
1	5	7	35	6	30	6	30	20
U+R		%		U+R		%		%
8		40,0		12		60,0		100
U+U= 7 = 35,0% R+R= 13 = 65,0%								

CONCLUSIONS

- Over 10% of the cases with mental deficiency have hearing-speaking disorders.
- It were observed increased frequencies in groups with moderate and severe mental deficiency.
- The frequency of hearing-speaking disorders in group with severe mental deficiency could be more increased because of the young age of children, when they can't cooperate with the person who investigate them.
- Most of cases proceed from rural area, perhaps because of agglomeration of some mutant genes.

- We could assume an autosomal recessive or dominant inheritance. Also, we could assume an X-linked inheritance.

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