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Family Functionality in Mexican Children with Congenital and Non-Congenital Deafness

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***Abstract*—**A total of 100 primary caregivers (mothers, fathers, grandparents) with at least one child or grandchild with a diagnosis of congenital bilateral profound deafness were assessed in order to evaluate the functionality of families with a deaf member, who was evaluated by specialists in audiology, molecular biology, genetics and psychology. After confirmation of the clinical diagnosis, DNA from the patients and parents were analyzed in search of the 35delG deletion of the GJB2 gene to determine who possessed the mutation. All primary caregivers were provided psychological support, regardless of whether or not they had the mutation, and prior and subsequent, the family APGAR test was applied. All parents, grandparents were informed of the results of the genetic analysis during the psychological intervention. The family APGAR, after psychological and genetic counseling, showed that 14% perceived their families as functional, 62% moderately functional and 24% dysfunctional. This shows the importance of psychological support in family functionality that has a direct impact on the quality of life of these families.

***Keywords*—**Deafness, psychological support, family, adaptation to disability.

I. Introduction

PSYCHOLOGICAL support in families with members with deafness directly impacts psychosocial well-being, quality of life and family communication. In Mexico, similar to what happens in other countries, the family with a member with deafness requires accompaniment, particularly from the psychological aspect from the time of diagnosis and throughout the treatment, to collaborate in the process of adapting to the disability. In Yucatan, deafness is a public health problem with a higher incidence compared to the rest of Mexico [1], [2]. In the same sense, according to data from the World Health Organization, the numbers of people with this disability is increasing and in children under the age of five years.

The family is the basic biopsychosocial unit, which is constituted as a group whose members are united by kinship or mutual affinity, and one of its most important functions is to contribute to the health of all its members. The interaction of family members, especially parents towards their children in the first years of life, fosters the development, both psychological and affective that the child requires to interact with others [3]. At the time of the birth of a child, there are family changes, new roles for parents, new responsibilities, changes in family life habits and adaptation to a new family dynamics [4]. However, there is an imbalance with the birth of a child with a disability. Inadequate family functionality may influence the ability of parents to make good decisions in the rehabilitation of deaf children. Therefore, it is important to promote the understanding of the different aspects involved in adapting to disability, including etiology. A very important role is that of psychological counseling that helps to reduce difficulties in the process of adaptation and avoid family dysfunction [5], [6]. Several studies have analyzed this dynamic [7]-[9].

Different instruments have been developed to explore the functioning of the family. Among these are the family APGAR scale, which was designed in 1978 by Smilkstein [10]. The acronym APGAR is derived from the five components of the family function: Adaptability, defined as the use of intra and extrafamilial resources to solve problems when the balance of the family is threatened by a stress factor during a period of crisis; cooperation, such as participation in decision-making and responsibilities, which defines the degree of power of family members; development, such as the possibility of emotional and physical maturation, as well as self-realization of family members, through mutual support; affectivity, such as the relationship of love and care among family members, and the ability to resolve, such as the commitment or determination to dedicate time (space, money) to other members of the family.

In Mexico, the APGAR scale has been used to know the resources and limitations of the family system itself, including capacity to solve problems, communication, limits, affective involvement among its members and management difficulties, union and support. Deafness causes serious disorders in the acquisition of language, impairs cognitive development and interferes with school progress and socialization [11]. Research has shown that the earlier diagnosis and initiation of medical care and psychological intervention, the better results can be expected in the care and development of these children and their families [12], [13].

It is a fundamental requirement to assess the characteristics of the functionality of families with a member with deafness to identify the areas to work in the clinical setting to achieve family adaptation to disability. This can contribute to decrease family uncertainty in the diagnosis and prognosis that are the main stressors in families with members with disabilities [14]-[16]. With this intention, the objective of the study was to determine the level of functionality of families with a member

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with congenital and non-congenital deafness).

II. Methodology

This study involved 100 primary caregivers (mothers, fathers, grandmothers or grandfathers) with at least one child or grandchild with a diagnosis of bilateral profound deafness of presumably genetic origin. All of them attend the Yucatecan Pro Deficiente Auditivo Association (AYPRODA). Of these, 87 were mothers, four fathers and nine grandparents. The children were diagnosed in AYPRODA through audiological studies and after confirming the clinical diagnosis of bilateral profound deafness, the DNA of the patients and primary caregivers were analyzed in search of the 35delG deletion of the GJB2 gene to determine who possessed the mutation.

A sociodemographic questionnaire was used, consisting of 15 mixed-type replies to collect information on the diagnosis, the role of the relative, the primary caregiver's civil status, family type, region of origin, educational level of the primary caregiver, and number of family members with deafness.

The family functionality was classified based on the family Apgar test, which is an instrument designed to evaluate the perception of the systemic functioning of the family, and is useful in the identification of families at risk. The instrument has been validated in different North American, Asian and Hispanic communities, offering a high correlation with specialized tests. The criteria proposed by de Smilkestein were used for the assessment: 1. Adaptation: ability to use resources in search of the common good and mutual aid, and the use of resources to solve problems when the balance of the family is threatened, 2. Partnership/Participation: distribution of responsibilities among family members, sharing in solidarity the problems and decision-making, 3. Growth: achievement of emotional and physical maturity, self-realization of family members, through mutual support, 4. Affectivity: relationship of care and love that exists between family members, and 5. Resilience: ability to solve family group problems, sharing time, space, and money among family members.

The items on the scale are scored as always, sometimes and almost never, with values of 2, 1 and 0 points, respectively. Once the total score was obtained, we proceeded to the classification of family functionality: From 7 to 10 points: functional family, from 4 to 6 points moderate dysfunctionality, from 0 to 3 points severe dysfunctionality.

To carry out the study, it was approved by the institution AYPRODA, as well as the parents and with the assent of the children. They were informed of the objective and previous signing of informed consent and assent of the minor, confidentiality was guaranteed, and the primary parents and/or caregivers were included in the study. From March 2015 to July 2016, 100 primary caregivers (mothers, fathers, grandparents) were selected with at least one child or grandchild, diagnosed with congenital bilateral profound deafness and who were evaluated by specialists in audiology, genetics, psychology and molecular biology. Following the confirmation of the clinical diagnosis of non-syndromic deafness, the molecular analysis was performed to search for the 35delG deletion of the GJB2 gene in the affected patients and their first-degree relatives by means of the specific allele-PCR technique. Once the results of the molecular analyzes were obtained, the parents and/or grandparents were asked to apply the APGAR test and to provide psychological support during 20 sessions, in which the first psychological help was implemented through brief therapy with the objective of establishing a positive attitude to the adaptation to the disability of the primary caregivers. The primary caregivers were evaluated before and after the psychological support through the APGAR scale.

III. Results

From the 100 primary caregivers of children with deafness, 12 (12%) were positive for the 35delG mutation in the GJB2 gene, from five different families. The mean age of the positive cases with deafness of genetic origin was 12 years (range 3-16 years). The age of primary caregivers ranged from 25 years to 65 years. The sociodemographic characteristics are: Family role - Mother shows 87% of the sample. Marital status: Marrived 90%, separated 5%, widowed, 4% and single 1%. The type of family nuclear is 92% and extended is 8%. The region is 70% urban and 30% rural. The educational level of the primary caregiver is mostly primary level with 32%, secondary 32%, illiterate 8% and baccalaureate 6%, technical career 3% and none with a degree.

Regarding the perception of family functionality, prior to genetic counseling, 8% [16] perceived their families as highly functional, 65% moderately functional and 27% as severely dysfunctional. After the psychological support sessions, 35% described their family as highly functional, 57% as moderately functional and 8% as severely dysfunctional.

TABLE I
Results of the Pre-Post Evaluation APGAR

Dimension	PRE	POST				*p*
		N	S	A	Total	
Adaptation	Never	34	6	14	54	< 0.0001
	Some times	0	29	14	43	
	Always	0	0	3	3	
	Total	34	35	31	100	
Asociation/ Participation	Never	13	3	2	18	0.005
	Some times	0	59	8	67	
	Always	0	0	15	15	
	Total	13	62	25	100	
Increase	Never	6	6	1	13	0.007
	Some times	0	53	5	58	
	Always	0	0	29	29	
	Total	6	59	35	100	
Efectiveness	Never	11	17	5	33	< 0.0001
	Some times	0	54	3	57	
	Always	0	0	10	10	
	Total	11	71	18	100	
Resolution Ability	Never	4	5	3	12	0.003
	Some times	0	52	6	58	
	Always	0	0	30	30	
	Total	4	57	39	100	

The qualitative results of the APGAR dimensions are shown in Table I, which shows a statistical significance by means of the McNemar test in each of the dimensions.

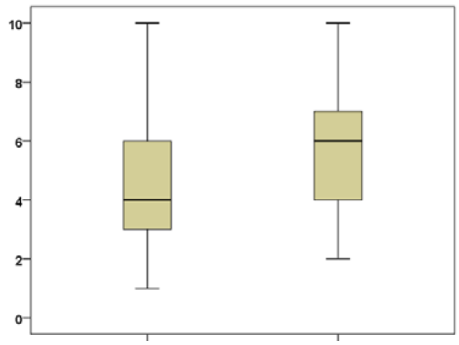

Fig. 1 APGAR results comparing the effectiveness on psychological orientation (pre-post evaluation)

Quantitatively, the mean APGAR score prior to psychological support was 4.57, and after intervention was 5.8 ($p < 0.001$), a difference of 1.23 (95% CI: 0.85-1.6) (Fig. 1).

IV. Conclusion

On the basis of the obtained research results, we can conclude that the majority of families with deaf children have certain problems in family functioning. Society, school and government should support these families through the designing a various programs for parents with deaf children so they can feel more competent in achieving their educational performance. It seems that parents do not have adequate education and in most of the cases, it is the mother who takes the lead in all the decisions about the progress and kind of education, and also the therapy that the child requires. In terms of the possibilities of setting certain requirements for their children; also, they do not have enough information about the options for their children so they are insecure about making decisions in the educational area. Lack of consistency can negatively affect the development of children with special needs. That is a constant problem in this kind of family. There is not enough dedication to the therapy or educative practices at home so the results could be show in a long time than normally does. The APGAR results help us to see the psychological orientation, so the parents are able to recognize what are the most important educative needs for their child. When the right educative choices are made, it can improve competence in children because they have the right treatment to improve their potential. Our findings have practical implications because they indicate the problems with which families with deaf children are faced, and point to the need of establishing support programs for these families.

It is important to adopt a multifactorial view regarding the family problems associated with deafness, and although the fact that one of its members has a disability tends to change the family dynamics, there are other factors that also modify it, such as poverty and discrimination, among others.

Generally, the type of disability is considered as the main cause between the differences in the behavior of the parents towards their children or the family dynamic; however, the disability itself is not the problem, rather it is related to the idea of "normality" and the vision of their expectations regarding their socialization. Thus, the behavior that parents adopt regarding the disability of their children is implicit in their own education and learning, as well as the influence exerted by society and its culture [15].

The incidence of cases in Yucatan is higher than that of Mexico, which makes the disease a public health problem for the country, and which, according to the WHO, is increasing, particularly in children under-5 years of age. So, we must think about specific public policies and not just those related to disability or sign language.

In this study, mutations in the GJB2 gene were identified in 12 of the 100 patients analyzed, corresponding to 12% of the cases. In the present study, it is estimated that up to 20% of cases of non-syndromic congenital deafness are due to mutations in this gene. This agrees that the GJB2 gene has been reported as the leading cause of autosomal recessive non-syndromic sensorineural hearing loss. Previous reports show that mutations in this gene vary according to the population studied. The 35delG homozygous mutation is commonly found in Italy, Spain, Portugal, France, England, Israel, Lebanon, Tunisia, Algeria, New Zealand, and Caucasian families in northern and southern Europe, with a prevalence ranging from 50% to 70%. It has been reported that in non-Caucasian populations this mutation has a low frequency [16].

The results of this study show that the birth of children with genetic deafness leads to social and psychological consequences that affect the ability of the family to adjust to the condition, according to what is referred to in the literature, as in the one conducted by Dueñas (2010) [16], who found that family dysfunction was reported with a low frequency; however, it behaved as a risk factor (OR = 1.44, 95% CI: 1.04-1.99, $p < 0.01$) for the appearance of diseases.

This study also reveals that psychological support favors this adaptation, since with 20 intervention sessions, a pre-post application of the family APGAR test showed a change from 8% to 35% of the family with highly functional perception. In other studies, with other types of childhood and motor disability, primary caregivers report moderate family functionality [17].

The study by Carla W. Jackson found that families were generally satisfied with the areas of family life, specifically in the emotional dimension and that the deafness of the children had the greatest impact on their well-being, as perceived in our study after the intervention of psychological support. On the other hand, other studies have found that the additional stress that is caused by caring for a child with some type of disability generates a high index of depression in the mothers in comparison to those who do not have a child with disability. However, providing psychological support to mothers with children with deafness has a positive impact and reduces the onset of depression, which translates into psychological well-being for the family, and therefore, a better functionality. This supports the benefits and the need for intervention of a

psychologist at the time of diagnosis, during the adaptation process and in the consolidation of optimal family functionality.

In this study, children with disabilities, compared with those who do not, have a lower quality of life, where the most affected areas referred to by the parents are health, welfare, employment and social life. Likewise, this is also reflected in parents, who reflect poor quality of life compared to parents who do not have children with disabilities, specifically in the physical and psychological domain [18]. Unlike girls, children aged between 11 and 12 years report high levels of physical, psychological, self-perception, autonomy and school environment than adolescents between 15 and 16 years old. While stress, depressive symptoms and concentration difficulties, affect the health and the quality of life in girls; this has to do with social and cultural factors due to adolescence, whereas sleep problems affect all children, with attention being given to the fact that sleep problems, depression and anxiety increase with age [19]. Other investigations have studied family functioning, through other instruments such as the FPQ (Fibromyalgia Participation Questionnaire), which also evaluates the therapeutic intervention in the participation and social functioning in patients, and have demonstrated good responsiveness, and are therefore recommended for evaluation studies and clinical trials [19].

For the purposes of this study, the family APGAR was used because it is a scale with international validity, easy access, but above all, because this instrument allows a greater interaction between the respondent and the one who applies it, which makes it possible to collect honest and detailed information regarding the family dynamics. It was also observed that it facilitates the self-reflexive process of who is interviewed, regarding their role and family relationship, and the interviewer allows him to identify specific conflicts to work with the family.

The birth of deaf children due to genetic causes implies social and psychological consequences that affect the family's ability to adjust to the condition. It is important to let parents know the cause of their children's deafness in order to understand the possibility of recurrence in other family members [20].

There has been a debate about when is the best moment to incorporate genetic testing into an auditory assessment. The emotional reaction of the parents to the diagnosis of hearing loss in the child is definitely a concern about deafness in general. Experience has shown that one of the hardest tests is the genetic one. Understanding parents' beliefs and knowledge throughout the genetic evaluation and testing process will be critical to better address their needs and interests, promote informed and collaborative decision making, and develop appropriate educational materials to assist parents with their children with hearing impairment [21].

We have been able to identify the contextualization and the competence of parents as the main themes of communication. Parents seemed to be trying to create a cognitive and emotional context for the genetic diagnosis of the child. Parents often ask, "Why did this happen to me?" and rationalize the diagnosis by eliminating risk factors that do not provide specific responses. In general, the stories showed that parents had limited the causes, by virtue of misunderstanding the genetics of their child's illness and contextualized the genetic diagnosis of the child by using family history as the adjustment to understand inherited etiologies. They were exploring the implications of the child's genetic diagnosis on the other family members, trying to understand the stages of their child's development and the risks of recurrence. These stories seem to highlight the attempts of parents to handle the deeper emotional issues surrounding the diagnosis so that psychological support ensures a better adaptation to the disability [22].

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