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Parental Consanguinity and Risk for Childhood Hearing Loss: A Retrospective Cohort Study

Consanguinidade Parental e Risco de Surdez Infantil: Estudo de Coorte Retrospetivo

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ABSTRACT

Introduction: Genetic causes are responsible for half of the cases of hearing loss, most of them being the result of non-syndromic genetic changes resulting from autosomal recessive inheritance. Parental consanguinity might be an indicator to consider in the diagnosis of these cases. The aim of this study was to assess its importance as a risk factor for childhood hearing loss.

Material and Methods: A retrospective cohort study conducted in a district hospital, between 2014 and 2018. We included all live births born during this period and excluded those with risk factors for childhood hearing loss other than parental consanguinity and those without hearing screening. We formed two study groups: newborns with parental consanguinity and newborns without risk factors. All the participants underwent hearing screening with the primary outcome of this study being the result of the screening. Those with a not normal result or with parental consanguinity also underwent diagnostic audiological evaluation.

Results: Among 8513 live births, we studied 96 newborns with first-degree parental consanguinity and 96 newborns without risk factors. We found a statistically significant difference (p = 0.007) between the groups, with a 'refer' screening result rate of 24% in the group with parental consanguinity and 9.4% in the group without risk factors. We diagnosed one case of sensorineural hearing loss and another of mixed hearing loss in the first group and none of these cases in the second.

Conclusion: Parental consanguinity was associated with a higher risk of a refer screening result in newborns, which suggests the need to consider this as a risk factor for childhood hearing loss.

Keywords: Consanguinity; Deafness/congenital; Hearing Loss/etiology; Hearing Loss, Sensorineural/etiology; Infant, Newborn; Neonatal Screening; Parents

RESUMO

Introdução: A etiologia genética é responsável por metade dos casos de surdez, a maioria fruto de alterações genéticas não-sindrómicas decorrentes de herança autossómica recessiva. A consanguinidade parental constitui um possível indicador a considerar para o diagnóstico destes casos, pelo que este estudo pretende avaliá-la como fator de risco para a surdez infantil.

Material e Métodos: Estudo de coorte retrospetivo realizado de 2014 a 2018 num hospital distrital. Incluímos todos os nados-vivos nascidos neste período, sendo excluídos aqueles com outros fatores de risco para surdez infantil (que não a consanguinidade parental) e aqueles sem rastreio auditivo. Formámos dois grupos de estudo: recém-nascidos com consanguinidade parental e recém-nascidos sem fatores de risco. Todos os participantes realizaram rastreio auditivo, sendo o seu resultado o *outcome* primário do estudo. Aqueles com resultado anormal ou com consanguinidade parental efetuaram ainda avaliação audiológica diagnóstica.

Resultados: Entre os 8513 nados-vivos, estudámos 96 recém-nascidos com consanguinidade parental em primeiro grau e 96 recém-nascidos sem fatores de risco. Verificámos uma diferença estatisticamente significativa (p = 0,007) entre os grupos relativamente aos resultados do rastreio auditivo, tendo-se detetado uma taxa de *refer* de 24% no grupo com consanguinidade parental e de 9,4% naquele sem fatores de risco. Diagnosticámos um caso de surdez sensorioneural e outro de surdez mista no primeiro grupo e zero destes casos no segundo.

Conclusão: A consanguinidade parental associou-se a um risco significativamente superior de resultado *refer* no rastreio auditivo de recém-nascidos com consanguinidade parental e sugere a necessidade de considerar este critério como um fator de risco para surdez infantil.

Palavras-chave: Consanguinidade; Pais; Perda Auditiva/etiologia; Perda Auditiva Neurossensoria/etiologia; Rastreio Neonatal; Recém-Nascido; Surdez/congénita

INTRODUCTION

Hearing plays a crucial role in an individual's ability to communicate and interact socially. As a consequence, hearing loss can have a decisive impact on quality of life.[1-3] It is currently recognised that around 50%[1] of all cases of hearing loss have a genetic cause. Moreover, this percentage increases to 70% if we consider only the congenital cases and may become even higher due to the decreased prevalence of infectious diseases as a result of vaccination.[1]

Genetic hearing loss may present early in life or have a late onset and may be syndromic (30%) or non-syndromic (70%).[1,2] Of the non-syndromic cases, 80% are the result of autosomal recessive inheritance and in these situations, there is no parental history of the disease.[1,3] Because

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consanguineous parents are more likely to be homozygous for the same trait, parental consanguinity is an important clue to the possibility of recessive inheritance of a condition.[3] Several studies have reported a higher incidence of autosomal recessive diseases in consanguineous families, and there is also data suggesting that profound hearing loss is more prevalent in countries where consanguineous marriages are common.[1-5] These types of marriages are common practice in many Asian, African and South American communities. In Portugal, the Romani communities also present important consanguineous relationships.[2,6] Consanguinity can be defined by marriages between second or closer cousins.[2] Its effect depends on the degree of kinship between the parents: first cousins have a higher risk of disease than second cousins and more distant kinship relationships have a risk of genetic defects close to that of the general population.[4]

Hearing loss affects one to three per 1000 newborn babies (NB) with no known risk factors and 20 to 40 per 1000 NB with risk factors.[7] Currently, the risk factors specified in Portugal's *Rastreio Auditivo Neonatal Universal* (RANU - the Infant Hearing Loss Screening and Intervention Group) and international guidelines (the Joint Committee on Infant Hearing) that may be suggestive of genetic inheritance are family history of hearing loss in childhood, the presence of craniofacial anomalies or genetic syndromes associated with hearing loss.[7,8] It is therefore inferred that children with no family history of hearing loss or syndromic stigma are classified as not being at risk when they may, in fact, have an increased susceptibility for hearing loss if their parents are consanguineous. As such, given the current absence of genetic screening tests, it becomes important to identify other conditions (in addition to those currently recognised) that may effectively be risk factors for a genetic cause.

Following the empirical notion that a significant number of diagnosed cases of hearing loss were associated with a history of parental consanguinity, the coordinating team of RANU at Centro Hospitalar do Baixo Vouga (CHBV), a level II public hospital in Aveiro, Portugal, started to include first-degree parental consanguinity as a risk factor for hearing loss since 2013. In addition, we also conducted a parallel study between 2014 and 2018 regarding the RANU results at the CHBV that suggested first-degree parental consanguinity was the most frequently observed risk factor in children diagnosed with sensorineural hearing loss.

As we are unaware of the existence of national studies in this context, the aim of this study was to assess whether the history of first-degree parental consanguinity was associated with a higher risk of hearing loss in the population of the Aveiro region in Portugal.

MATERIAL AND METHODS

We conducted a retrospective cohort study at the CHBV, the main public hospital in the Aveiro region, in Portugal, between 2014 and 2018. We included all live births born in this hospital between the 1st January 2014 and the 31st December 2018. To avoid biasing the results, we excluded all those who presented risk factors[9] for hearing loss (Table 1) other than first-degree parental consanguinity as well as those who did not undergo any hearing screening test.

For the purposes of this study, and taking into account our clinical experience, we defined exposure as the existence of a history of first-degree parental consanguinity, i.e., parents in a relationship with first cousins or closer. We consecutively selected all NB with this condition in order to establish the group of exposed NBs. For each NB included in this group a NB with no known risk factors was randomly selected to establish the non-exposed group. Information regarding the existence or not of first-degree parental consanguinity and the remaining risk factors was obtained through the systematic survey of all mothers, carried out by the paediatrician responsible for each NB.

All NBs underwent neonatal hearing screening. The result of each screening test was defined as 'pass' (no changes in both ears) or 'refer' (with changes in one or both ears). The screening method used was evoked acoustic

Table 1 – Risk factors for hearing loss incorporated in the exclusion criteria

Risk factors for hearing loss
Family history of hearing loss in childhood
Prematurity ≤ 32 weeks gestation
Birth weight < 1500 g
Apgar score of 0 – 4 at the first minute or 0 – 6 at the fifth minute of life
Craniofacial malformations or stigma associated with hearing loss
Congenital infection (toxoplasmosis, rubella, cytomegalovirus, herpes and syphilis)
Neonatal sepsis/meningitis and/or taking ototoxic medicines for five or more days
Hyperbilirubinemia (serum levels indicating the need for exsanguineous transfusion)
Intracranial haemorrhage
Hospitalisation for more than 48 hours and mechanical ventilation in Intensive Care Unit

otoemissions using a Natus® MADSEN AccuScreen device. NBs with a first 'refer' screening result were sent to a second screening evaluation using the same technique and device. All children that repeated screening were, as recommended, tested in both ears. Apart from the screening test and regardless of the results, NBs belonging to the exposed group were referred for diagnostic audiological assessment. This evaluation included a RANU consultation which comprised both an otorhinolaryngological medical assessment and a diagnostic test using the auditory brainstem potentials method (performed with an Interacoustics Eclipse EP25 device from Interacoustics®). Furthermore, NBs in the non-exposed group (without risk factors) were only referred for this assessment in case of a 'refer' in two screening tests. All screening and diagnostic tests were carried out by one of two audiologists with experience of pediatrics, in a dedicated room and in spontaneous sleep. All devices were calibrated annually by the representative companies.

We defined as primary outcome the result of the first screening test of each NB and as secondary outcomes the results of the second screening and the diagnostic tests. In order to detect a relative risk (RR) of three for a 'refer' screening result, assuming a frequency of 10% in the non-exposed population, with a confidence interval of 95% and a test power of 80%, we calculated that we would need a sample size of 59 participants in each group. This calculation was performed using the EpiTool online for sample size calculation for descriptive and analytical studies.

Data were collected through consultation and retrospective review of the electronic medical records in the database of the *Plataforma Online de Rastreio Auditivo Neonatal Universal* (Universal Newborn Auditory Screening Online Platform) created at the CHBV, which includes clinical information and data from all hearing assessments (screening and diagnostic) of all NBs born in this hospital. This platform was authorized by the *Comissão Nacional de Proteção de Dados* (National Data Protection Commission) and won a *Boas Práticas em Saúde* (Best Practices in Health) award. The study protocol received clearance from the Ethics Committee and the Data Protection Officer of the CHBV with the reference number 26-01-2022/CES. Collected data included demographic and clinical data such as gender, gestational age, type of delivery, birth weight, risk factors, audiological screening results, diagnostic audiological evaluation results and the age of the NB on the date of each evaluation. Each participant was monitored from birth to the date of the last screening assessment or, whenever necessary, diagnostic assessment in a RANU consultation. Considering the period of the study, NBs were followed for a maximum of five years for the development of hearing loss.

In the descriptive analysis we used the mean and standard deviation (SD) to characterise normally distributed quantitative variables and the median and the interquartile range (IQR) to describe quantitative variables without normal distribution. To assess the normality of distributions we resorted to the analysis of Kolmogorov-Smirnov test. Qualitative variables were expressed as absolute number and percentage. For the inferential analysis of continuous variables, we used the *t* test for independent samples or the Mann-Whitney U test, as applicable. For the analysis of categorical variables, we used the chi-square test. We defined as statistically significant a *p* value less than 0.05 for all tests performed. The data analysed was entered into Microsoft Excel® and statistical analysis was performed using IBM® SPSS® Statistics, version 27.0, for Mac®.

RESULTS

During the study period, we found 115 cases of first-degree parental consanguinity, among 8513 live births, which translates into a prevalence rate of 1.4% during the five-year period under analysis. Nineteen NBs were excluded from the study: 14 due to a family history of hearing loss; one due to an Apgar score of six at the fifth minute of life associated with craniofacial malformation; two due to the administration of ototoxic medicines for more than five days; one due to intracranial haemorrhage; and one due to the absence of screening tests. Therefore, we included a total 192 children: 96 NBs with first degree parental consanguinity, constituting the exposed group, and 96 NBs with no risk factors, constituting the non-exposed group. The results of the assessment of each group are summarised in Table 2. The birth weight variable had a normal distribution ($p = 0.200$). The gestational age, age at the date of the first screening and the age at the date of the second screening did not follow a normal distribution ($p < 0.001$).

In the group of NBs without risk factors there was a predominance of male children (57.3%) while in the exposed group there was a majority of female children (51%). The mean birth weight was 3163.9 g (SD 425.2) in the non-exposed group and 3139.2 g (SD 493.0) in the exposed group. The median gestational age was 39 weeks (IQR 38 – 40) in those without risk factors and 38 weeks (IQR 37 – 39) in the group with parental consanguinity. In both groups, there was a predominance of eutocic deliveries (63.5% in the non-exposed group and 50% in the exposed group) and vaginal delivery using forceps was more frequent (11.7%) than delivery using suction (7.4%) in the exposed group.

In the group of NBs without risk factors there was a 'refer' rate of 9.4% in the first screening while in the exposed group this percentage was 24%. In the non-exposed group, there was a predominance of right ear 'refer' results (3.1% in the left ear *vs* 7.3% in the right ear) while in the exposed group this percentage was overlapping (18.8% in the left

Table 2 – Demographic and clinical characteristics of the study groups (n = 192)

	Non-exposed Group (no risk factors)	n	Exposed Group (first-degree parental consanguinity)	n	*p*-value
Gender		96		96	0.247[a]
Female	42.7%	41	51.0%	49	
Male	57.3%	55	49.0%	47	
GA (weeks)	39 (IQR, 38 – 40)	94	38 (IQR, 37 – 39)	94	0.107[c]
Type of birth		96		94	0.017[a]
Eutocic	63.5%	61	50.0%	47	
Caesarean section	14.6%	14	30.9%	29	
Forceps	7.3%	7	11.7%	11	
Suction cup	14.6%	14	7.4%	7	
Weight (grams)	3163.9 (BW, 425.2)	94	3139.2 (BW, 493.0)	94	0.712[b]
First screening		96		96	0.007[a]
'Pass'	90.6%	87	76.0%	73	
'Refer'	9.4%	9	24.0%	23	
First screening LE		96		96	< 0.001[a]
'Pass'	96.9%	93	81.3%	78	
'Refer'	3.1%	3	18.8%	18	
First screening RE		96		96	0.029[a]
'Pass'	92.7%	89	82.3%	79	
'Refer'	7.3%	7	17.7%	17	
Age at first screening (days)	2 (IQR, 2 – 3)	96	2 (IQR, 2 – 4)	96	0.322[c]
Second screening		9		22	[d]
'Pass'	88.9%	8	81.8%	18	
'Refer'	11.1%	1	18.2%	4	
Age at second screening (days)	19 (IQR, 16 – 33)	8	33 (IQR, 24 – 54)	22	[d]
Diagnostic assessment		1		36	[d]
Normal	0%	0	58.3%	21	
Conductive HL	100%	1	36.1%	13	
Sensorineural HL	0%	0	2.8%	1	
Mixed HL	0%	0	2.8%	1	

SD: standard deviation; IQR: interquartile range; LE: left ear; RE: right ear; **a**: chi-square test; **b**: *t*-test for independent samples; **c**: Mann-Whitney U test; **d**: variables with an insufficient number of cases for a reliable inferential analysis to be carried out; HL: hearing loss

ear *vs* 17.7% in the right ear). In both groups the median age at first screening was two days. Of the 'refer' NBs in the first screening, all those belonging to the group without risk factors underwent a second screening in which a 'refer' result rate of 11.1% was found. In the group of NBs with first degree parental consanguinity there was one NB who missed this assessment and a 'refer' rate of 18.2% was found. The median age at the date of the second screening was 19 days for the group of non-exposed and 33 days for the group of exposed NBs.

The diagnostic assessment in the group of NBs without risk factors was only carried out in the one child who presented 'refer' in two screening tests and in whom conductive hearing loss was detected. In the group of NBs with first degree parental consanguinity, there was a 62.5% rate of absenteeism in the diagnostic assessment and this analysis was only carried out in 36 children. Most of these children presented normal results (58.3%), with one case of sensorineural hearing loss and one case of mixed hearing loss.

There were no statistically significant differences regarding gender, birth weight, gestational age or age at the date of the first screening. On the contrary, regarding the type of delivery, the difference between the groups was statistically significant (p = 0.017). A statistically significant difference was also found in the results of the first screening, both when considering the overall screening result (p = 0.007)

and when analysing the result separately for each ear ($p < 0.001$ for the left ear; $p = 0.029$ for the right ear). As for the results of the second screening, the age of each child at the time of the screening and the diagnostic assessment, it was not possible to carry out an inferential analysis, as we did not have a minimum sample size to allow for reliable statistical tests.

As for measures of association and impact, we calculated a relative risk of 2.6 (95% CI, 1.2 – 5.2) for a 'refer' result at first screening in the group of NB with first-degree parental consanguinity, an attributable risk of 61% and a number required to cause harm of 9.

DISCUSSION

This study found that having first degree parental consanguinity was associated with a three times higher risk of having a 'refer' hearing screening result compared to children without risk factors. When we analysed the proportion of screenings attributable to consanguinity, we found that first degree parental consanguinity accounted for 61% of the risk of a 'refer' result, meaning that if this factor did not exist, this risk would decrease by 61%. We also determined that, for every nine NBs with first degree parental consanguinity, there was an additional case of a 'refer' result in the screening. No other studies were found with a similar design analysing the results of screening between consanguineous children and those without risk factors.

While hearing screening does not guarantee the diagnosis of hearing loss and, therefore, does not establish a direct relationship between its result and diagnosis, we estimate that a higher number of 'refer' screening results corresponded to a higher risk of hearing loss. The fact that the CHBV is a district hospital and, as such, serves a small population, limits the number of diagnosed cases of hearing loss. This reality made it impossible for us to carry out a cohort study with the results of a diagnostic audiological assessment instead of screening results as the outcome. A national multi-centre study could have overcome this limitation. Still, several reports in the literature suggest that first-degree parental consanguinity is associated with a higher risk of sensorineural hearing loss. Almazroua *et al* found a 3.5 times higher risk of sensorioneural hearing loss in consanguineous marriages than that non-consanguineous.[3] Some authors investigated the cause of this association and Kavitha *et al* performed a prospective MRI study that suggested that genetic defects resulted in a cochlea with normal morphology but abnormal function.[10] A study carried out in Qatar, a country with one of the highest rates of consanguinity in the world, showed a strong correlation between parental consanguinity and hearing loss.[4] In a study from Pakistan, a country with a high rate of consanguinity, there was a positive association between consanguinity and profound sensorineural hearing loss.[2] A study in Oman, a Middle Eastern country, showed an association between the incidence of severe hearing loss and consanguineous marriages.[11] These reports appear not only in Middle Eastern countries but also in European countries. Although consanguineous marriages are not culturally frequent in the West, globalisation and migration give rise to small communities in which consanguinity is a frequent practice.[11,12]

In this context, a study has shown that the prevalence of hearing loss in British children of Bangladeshi origin is at least 2.3 times higher than the British average, with consanguinity contributing towards the increase in prevalence along with other environmental factors.[12] The results presented in different studies are so relevant that a Belgian guideline developed by consensus of several experts suggests not only screening but also diagnostic audiological assessment during the neonatal period for this group of children.[13] In Portugal, parental consanguinity is still an existing practice, in particular within the Romani community.[6]

In our study, first-degree parental consanguinity showed a prevalence rate of 1.4% in the studied population. In another study carried out in parallel at the CHBV, first-degree parental consanguinity was the third most common risk factor among children with risk factors for hearing loss, the second most frequent risk factor in children referred at the first hearing screening and the one most that was found more often in the group of children diagnosed with sensorineural and mixed hearing loss (two had parental consanguinity, two had family history and two had both). We thus perceive that although it is not a common practice in Portugal, parental consanguinity has a significant impact on the population of the Aveiro region.

Our study used a non-random sampling for the establishment of the group of NBs with parental consanguinity. However, what could be seen as a limitation turned out to be an advantage, since the use of a consecutive sample ensured the selection of all cases of consanguinity throughout the study period, which meant a representative sample of the population being studied and contributing to the internal validity of the study. The comparison of consanguineous children with others without risk factors also minimised the risk of bias. Collectively, the performance of screening and diagnostic tests by an experienced paediatric audiologist helped to limit the number of false-positive screening results but we can not rule out the risk of bias because they knew which NBs had risk factors for hearing loss. The homogeneity between the groups in terms of gestational age, birth weight and age at the time of screening reinforces their comparability. As for the statistically significant differences between the groups regarding the type of delivery, some authors suggest that caesarean deliveries are associated with a higher risk of referral for screening due to greater

fluid retention in the ear, although others suggest that these differences are related to the timing of the screening.[15,16] In our study the median age at first screening was the same between the two groups so the differences do not correlate with this fact. As far as variation in screening results and diagnosis according to the use of forceps or suction is concerned, we found no data in the literature to suggest differences in outcomes.

As for the rate of absenteeism regarding diagnostic audiological assessment found in the group of NBs with parental consanguinity, we noticed that it is higher than the already significant absenteeism rate detected in our hospital (62.5% *vs* 44.56%). This fact may be due to a reduced perception by parents of the harmful effects of consanguinity, and the association between parental consanguinity and illiteracy. This knowledge highlights the importance of parental education on this issue.[1,14,17]

CONCLUSION

Children with first degree parental consanguinity had a three times higher risk of having a 'refer' hearing screening result which probably corresponds to a higher risk of hearing loss. Knowing this, we intend to draw attention to the evidence suggesting a significant association between parental consanguinity and the prevalence of childhood hearing loss. Failure to consider this criterion as a risk factor may lead us to include children with increased risk of hearing loss in a non-risk group, limiting not only the type of assessment performed but also the observation of these children. Further national studies are necessary in order to confirm the cost-effectiveness of considering this criterion as a risk factor in Portugal. Regarding the practice at our hospital center, given the data collected, it seems prudent to maintain first-degree parental consanguinity as a risk factor for hearing loss.

AUTHOR CONTRIBUTIONS

BL: Literature research; study design; data collection; statistical analysis; draft of the manuscript.

ACL: Literature research; data collection and interpretation; draft of the manuscript.

DP, LC: Data collection; draft of the manuscript.

MMA, MAB: Study design; critical review.

JV: Data collection and processing.

MLA: Critical review.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

COMPETING INTERESTS

MLA has received consulting fees, payment or honoraria for lectures, presentations, speakers' bureaus, manuscript writing or educational events, support for attending meetings and/or travel from Sanofi, and participated on a Data Safety Monitoring Board or Advisory Board for Sanofi.

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REFERENCES

1. Flint PW, Francis HW, Haughey BH, Lesperance MM, Lund VJ, Robbins HT, et al. Cummings otolaryngology head and neck surgery. 7th ed. Amsterdam: Elsevier; 2010.
2. Sajjad M, Khattak AA, Bunn JG, Mackenzie I. Causes of childhood deafness in Pukhtoonkhwa province of Pakistan and the role of consanguinity. J Laryngol Otol. 2008;122:1057–63.
3. Almazroua AM, Alsughayer L, Ababtain R, Al-shawi Y, Hagr AA. The association between consanguineous marriage and offspring with congenital hearing loss. Ann Saudi Med. 2020;40:456-61.
4. Benera A, ElHakeemb AA, Abdulhadib K. Is there any association between consanguinity and hearing loss. Int J Pediatr Otorhinolaryngol. 2005;69:327-33.
5. Zakzouk S. Consanguinity and hearing impairment in developing countries: a custom to be discouraged. J Laryngol Otol. 2002;116:811–6.
6. Mendes M, Magano O, Candeias P. Estudo Nacional sobre as Comunidades Ciganas. Lisboa: Alto Comissariado para as Migrações; 2014.
7. Grupo de Rastreio e Intervenção da Surdez Infantil. Recomendações para o Rastreio Auditivo Neonatal Universal (RANU). Acta Pediatr Port. 2007;38:209-14.
8. The Joint Committee on Infant Hearing. Year 2019 position statement: principles and guidelines for early hearing detection and intervention programs. J Early Hear Detect Interv. 2019;4:1–44.
9. Direção-Geral da Saúde. Rastreio e tratamento da surdez com implantes cocleares em idade pediátrica. Lisboa: DGS; 2017.
10. Kavitha Y, Sabarigirish K, Joish UK, Saxena S, Dutta A. A study of effect of consanguinity on cochlear morphology in patients with congenital bilateral profound sensorineural hearing loss. Indian J Otolaryngol Head Neck Surg. 2017;69:483-7.
11. Khabori MA, Patton MA. Consanguinity and deafness in Omani children. Int J Audiol. 2008;47:30-3.
12. Bajaj Y, Sirimanna T, Albert DM, Qadir P, Jenkins L, Cortina-Borja M, et al. Causes of deafness in British Bangladeshi children: a prevalence twice that of the UK population cannot be accounted for by consanguinity alone. Clin Otolaryngol. 2009;34:113–9.
13. Vos B, Senterre C, Lagasse R, SurdiScreen Group, Levêque A. Newborn hearing screening programme in Belgium: a consensus recommendation on risk factors. BMC Pediatr. 2015;15:160.
14. Dudda R, Muniyappa HP, Puttaraju S, Lakshmi MS. A qualitative study on knowledge and attitude towards risk factors, early identification and intervention of infant hearing loss among puerperal mothers - a short

survey. J Clin Diagn Res. 2017;11:MC01-5.
15. Smolkin T, Mick O, Dabbah M, Blazer S, Grakovsky G, Gabay N, et al. Birth by cesarean delivery and failure on first otoacoustic emissions hearing test. Pediatrics. 2012;130:e95-e100.
16. Güven SG. The effect of mode of delivery on newborn hearing screening results. Turk Arch Otorhinolaryngol. 2019;57:19-23.
17. Ajallouyan M, Radfar S, Nouhi S, Tavallaie SA, Amirsalari S, Yousefi J, et al. Consanguinity among parents of Iranian deaf children. Iran Red Crescent Med J. 2016;18:e22038.