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ORIGINAL ARTICLE

Delayed Diagnosis of Childhood Deafness: The Value of False Negatives in the Programme for Early Detection of Neonatal Hearing Loss☆

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Hearing loss;
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Abstract
Introduction: Despite its importance, the existence of false negatives (patients who are told they hear well, but they have some degree of hipacusia) is rarely evaluated in programmes for early detection of hearing loss. The aim of this study is to determine the variables that can lead to a delayed diagnosis, especially the existence of false negatives and the lack of registration of risk factors.
Methods: A retrospective study of prevalence has been carried out, in which the medical records of children diagnosed with sensorineural hearing loss born within 2005 and 2012 in the health centres of study have been analysed.
Results: Of the 32 children with sensorineural hearing loss, 16 passed the OAE, 12 did not passed the OAE, and in four they were not carried out. Of the children who passed the OAE, 57% have severe hearing loss. 66% of children with hearing loss presented a risk factor for hearing loss at birth, being the most frecuent family history of hearing loss, but only 7% of those with family history of hearing loss were included in the risk group.
Conclusions: The results of the study indicate that the late diagnosis of hearing loss is related to the presence of false negatives to the OAE and the non-registration of risk factors.

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PALABRAS CLAVE
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Retraso en el diagnóstico de sordera infantil: el valour de los falsos negativos en el Programa de Detección Precoz de Hipoacusias Neonatales

Resumen
Introducción: A pesar de su importancia, la existencia de falsos negativos (pacientes a los que se les dice que oyen bien, pero en realidad son hipoacúsicos) no suele ser evaluada en los programas de detección precoz de hipoacusia. El objetivo de este estudio es determinar las variables que pueden a llevar a un retraso diagnóstico, en especial la existencia de falsos negativos así como la falta de registro de factores de riesgo.
Métodos: Se ha realizado un estudio observacional retrospectivo de prevalencia, analizando las historias clínicas de los pacientes con hipoacusia neurosensorial nacidos entre 2005 y 2012 en las áreas de salud del estudio.
Resultados: De 32 niños con hipoacusia neurosensorial, 16 pasaron las OEA, 12 no pasaron las OEA y a 4 no se les realizaron. De los pacientes con hipoacusia pero que pasaron las OEA, el 57% tiene una hipoacusia severa y/o profunda. El 66% de los niños con hipoacusia presentaban algún factor de riesgo de hipoacusia, siendo los antecedentes familiares de hipoacusia el más frecuente, pero solo el 7% de los que tenían antecedentes familiares fueron incluidos en el grupo de riesgo. La tasa de pacientes falsos negativos en el estudio es muy elevada.
Conclusiones: Los resultados del estudio indican que el diagnóstico tardío de las hipoacusias infantiles en las áreas estudiadas está relacionado con la presencia de falsos negativos a las OEA y con la falta de registro de los factores de riesgo.

Introduction

Hearing loss in children has implications for their emotional, academic and social development, as well as their capacity for acquiring language and speech. The debilitating potential of this disease is largely mitigated if it is diagnosed early and the appropriate treatment and rehabilitation is started promptly.[1]

Newborns with risk factors have an increased incidence of hearing loss, between 10 and 20 times higher than those with no risk factors. Similarly, between 50% and 75% of children with bilateral hearing loss equal to or above 40 dBHL have one or more risk factors.[2] Up to 80% of hearing loss in the paediatric population already presents at birth or in the neonatal phase and around 95% occurs in children born to families with normal hearing.

Hearing loss screening was officially implemented in our community in 2004 and from then on every hospital with a mother and baby unit, whether public or private, has screened all newborns.

The recommendations of the *Comisión para la Detección Precoz de la Sordera* (Committee for the Early Detection of Deafness), are detailed in Table 1.[3] Based on these recommendations, the objectives of the *Programa de Detección Precoz de las Hipoacusias Neonatales en la Comunidad Autónoma* (Programme for the Early Detection of Neonatal Hearing Loss in Autonomous Communities) are[4]:

1. To detect hearing disorders in all newborns in the Region of Murcia in the first month of life.
2. To diagnose any hearing disorder before three months of life.
3. To establish treatment for all children who have been diagnosed with hearing loss before six months of life.

Detecting patients with risk factors is essential in the programme, because there is a greater prevalence of hearing loss in these patients. They should be easy to register and the main risk factors are well known. The fact that a patient has a hearing loss risk factor makes it necessary to use another test in addition to otoaccoustic emissions.

Furthermore, the existence of false negatives (patients that pass otoaccoustic emission tests but in fact have hearing loss) in the screening programme might result in a significant delay in diagnosis, due to a false sense of security that the patient has passed the first test successfully.

Numerous assessments have been made of the programmes for the early detection of hearing loss in the different autonomous regions of Spain. In the great majority, the main indicators analysed are coverage, false positives and follow-up in the different phases of the screening programme until diagnosis and in the treatment and rehabilitation stages.[5–9] However, we have found no study that analyses the rate of false negatives in the Spanish programme, although one mentions their possible existence,[10] confirming that half the diagnostic centres acknowledge that they have encountered late cases of hearing loss that had achieved a ''PASS'' in the initial hearing screening.

We found one article that precisely studies a series of cases of children with sensorineural hearing loss despite having passed the hearing screening test. The study undertaken in Pensylvania (U.S.A.)[11] analysed the clinical histories of 923 children with sensorineural hearing loss diagnosed between 2001 and 2011, amongst whom 78 (8.5%) had passed