



(RESEARCH ARTICLE)



## Ocular involvement in hearing-impaired children: A retrospective study

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### Abstract

Hearing impairment in children is a major public health issue that may be associated with ophthalmological abnormalities, particularly in syndromic conditions, leading to a dual sensory deficit with significant functional consequences.

This study aimed to evaluate the prevalence and characteristics of ocular involvement in hearing-impaired children and to highlight the importance of systematic ophthalmological screening.

A retrospective descriptive study was conducted in the Department of Otorhinolaryngology at Mohammed VI University Hospital in Marrakech between January 2014 and January 2023. A total of 688 children with hearing impairment were reviewed, and those under 15 years of age with at least moderate hearing loss were included. Ocular involvement was identified in 42 patients, corresponding to a prevalence of 6.1%. The mean age was 40 months, with a predominance of females. Hearing loss was bilateral in 78.5% of cases and predominantly sensorineural, with a mean threshold of 73 dB. Ocular involvement was bilateral in 75% of cases, with retinal abnormalities being the most frequent, followed by adnexal and iris involvement. Syndromic etiologies were identified in the majority of cases, particularly Waardenburg and Usher syndromes.

Cochlear implantation was performed in selected patients, especially those with progressive visual impairment.

Ocular involvement in hearing-impaired children is not uncommon and may have significant implications for functional development. Systematic ophthalmological screening is essential for early detection, identification of syndromic conditions, and optimization of management within a multidisciplinary approach.

**Keywords:** Hearing impairment; Ocular involvement; Oculo-auditory syndromes; Cochlear implantation; Multidisciplinary management

### 1. Introduction

Hearing impairment in children represents a major public health concern due to its significant impact on language acquisition, cognitive development, academic performance, and social integration [1]. Early diagnosis and appropriate management are essential to reduce long-term functional and psychosocial consequences [1].

Ophthalmological abnormalities are frequently associated with pediatric hearing loss, particularly in syndromic conditions [2]. This association may be explained by shared embryological origins and molecular mechanisms between the inner ear and ocular structures, especially the retina [3]. Several studies have shown that children with hearing impairment are at increased risk of visual disorders, with a higher prevalence in syndromic forms [2].

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The coexistence of auditory and visual impairments results in a dual sensory deficit that may significantly affect communication skills, psychomotor development, and overall quality of life [3]. In addition, some ocular conditions associated with hearing loss are progressive, such as retinitis pigmentosa in Usher syndrome, which may lead to progressive visual loss or blindness [4]. Early identification of these conditions is therefore crucial to optimizing management strategies, including timely auditory rehabilitation.

Despite this well-established association, ophthalmological evaluation is not systematically performed in all hearing-impaired children, potentially leading to delayed diagnosis and suboptimal care [2].

This study aimed to evaluate the prevalence and characteristics of ocular involvement in hearing-impaired children and to highlight the importance of routine ophthalmological screening as part of a multidisciplinary approach.

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## 2. Materials and Methods:

We conducted a retrospective descriptive study in the Department of Otorhinolaryngology and Head and Neck Surgery at Mohammed VI University Hospital in Marrakech, Morocco, over a nine-year period from January 2014 to January 2023. A total of 688 children with hearing impairment were reviewed. Among them, patients presenting with ophthalmological abnormalities were identified and included in the study.

Children were eligible for inclusion if they were younger than 15 years and had at least moderate hearing loss. Patients with post-traumatic hearing loss, chronic otitis media, or cerebral palsy were excluded.

Data were collected retrospectively from medical records and included demographic characteristics (age and sex), hearing impairment characteristics (laterality, degree, and type), ophthalmological findings (type and laterality of ocular involvement), etiological profile (syndromic and non-syndromic forms), and therapeutic management (auditory rehabilitation and ophthalmological treatment).

All patients underwent a complete otorhinolaryngological examination, including pure-tone audiometry and auditory brainstem response testing when indicated. The degree of hearing loss was classified according to the average hearing threshold.

All patients also underwent a comprehensive ophthalmological evaluation, including visual acuity assessment, anterior segment examination, and fundus examination. Additional investigations were performed when necessary based on clinical findings.

Data analysis was descriptive. Quantitative variables were expressed as means with ranges, and qualitative variables as frequencies and percentages. Patient confidentiality was strictly respected, and all data were anonymized prior to analysis.

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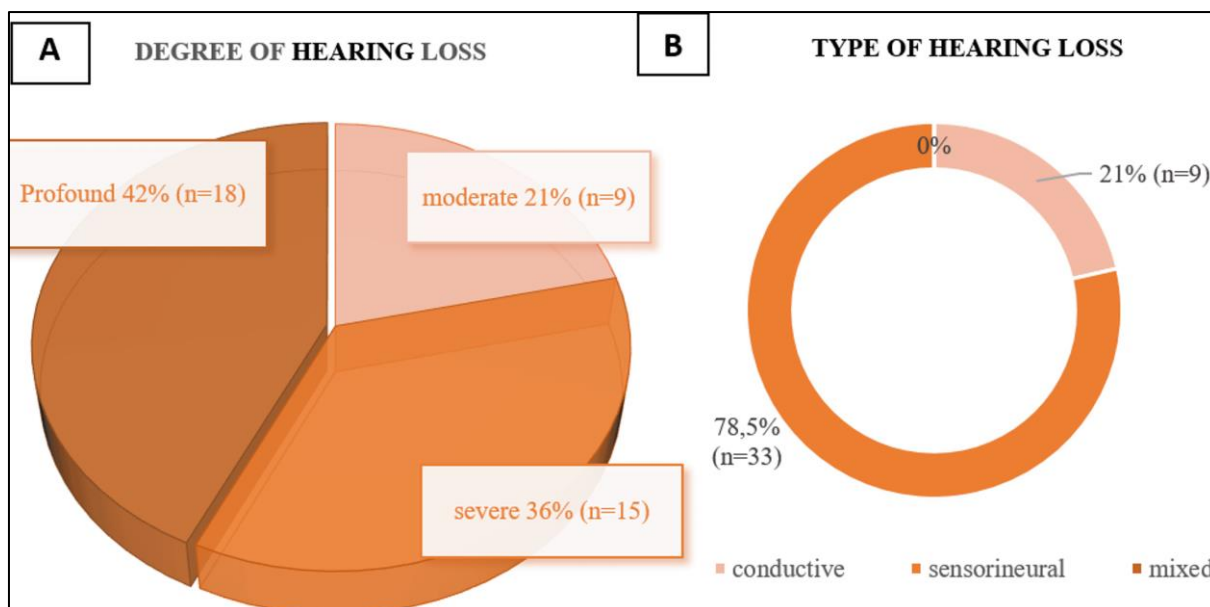
## 3. Results

During the study period, a total of 688 children were followed for hearing impairment. Among them, 42 patients presented with ophthalmological abnormalities, corresponding to a prevalence of 6.1%.

The mean age of patients was 40 months (range: 16 months to 15 years), with a female predominance (25 females and 17 males).

Regarding hearing impairment, the majority of patients had bilateral involvement in 33 patients (78.5%), while 9 patients (21.5%) had unilateral hearing loss. The mean hearing threshold was 73 dB (range: 40-120 dB).

In terms of severity, profound hearing loss was the most frequent, followed by severe and moderate forms (Figure 1A). Sensorineural hearing loss was predominant, whereas conductive hearing loss was less common (Figure 1B). No cases of mixed hearing loss were identified.



**Figure 1** Distribution of Hearing Loss Severity (A) and Type of Hearing Loss (B)

Ophthalmological involvement was bilateral in 31 patients (75%) and unilateral in 11 patients (25%).

Retinal abnormalities were the most frequent findings (20 cases, 47.6%), followed by adnexal involvement (14 cases, 33.3%). Other lesions included iris abnormalities (4 cases, 9.5%), optic nerve involvement (2 cases, 4.8%), and isolated corneal and lens involvement (1 case each, 2.4%) (Table 1).

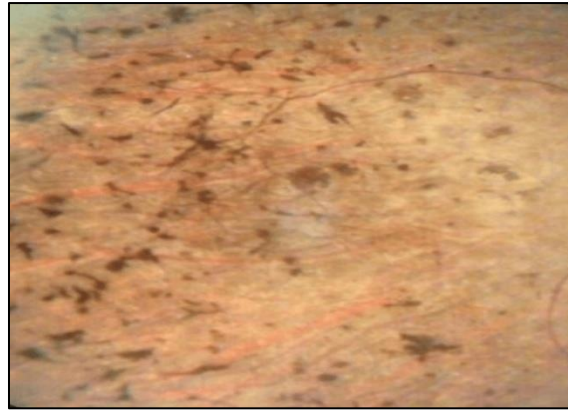
**Table 1** Distribution of ocular involvement in hearing-impaired children

| Type of ocular involvement | Number of cases (n) | Percentage (%) |
|----------------------------|---------------------|----------------|
| Retinal involvement        | 20                  | 47.6           |
| Adnexal involvement        | 14                  | 33.3           |
| Iris abnormalities         | 4                   | 9.5            |
| Optic nerve involvement    | 2                   | 4.8            |
| Corneal involvement        | 1                   | 2.4            |
| Lens involvement           | 1                   | 2.4            |
| Total                      | 42                  | 100            |

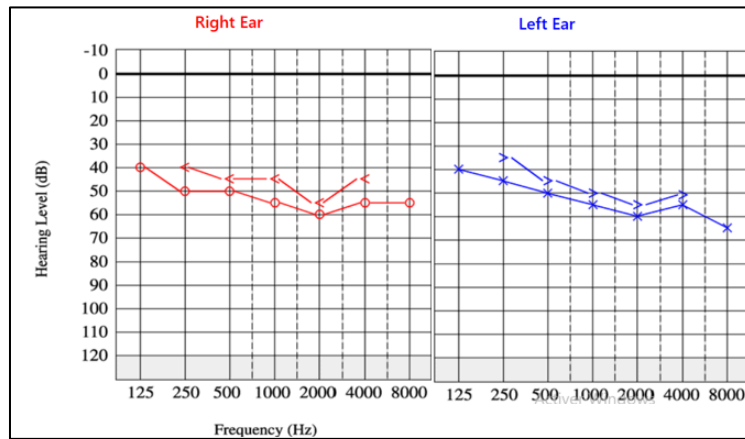
Syndromic etiologies were identified in 31 patients (73.8%), representing the majority of cases. The most frequent syndromes were Waardenburg syndrome (Figure2) (7 cases, 16.7%), Usher syndrome (5 cases, 11.9%) (Figure 3), Goldenhar syndrome (4 cases, 9.5%), and otomandibular syndrome (5 cases, 11.9%). Other syndromes included Wolfram, Alström (Figure 4), CHARGE, Stickler, KID syndrome, Franceschetti syndrome, and Refsum syndrome.



**Figure 2** Iris heterochromia in a patient with Waardenburg syndrome

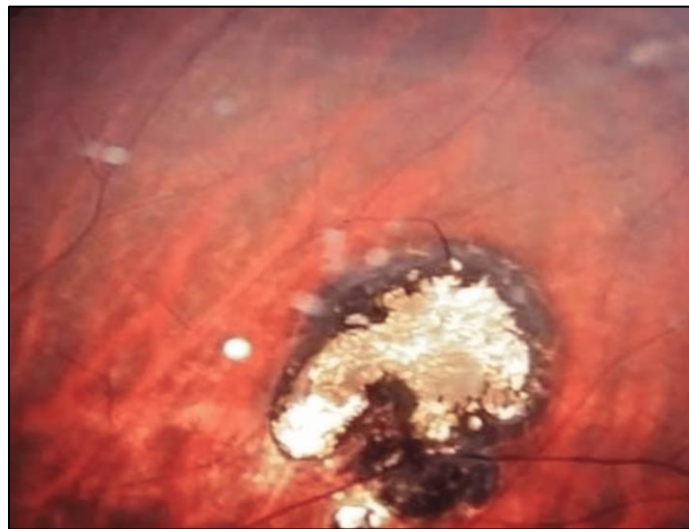


**Figure 3** Fundus photograph showing retinitis pigmentosa in a patient with Usher syndrome



**Figure 4** Clinical features and pure-tone audiometry in a patient with Alström syndrome, revealing bilateral moderate sensorineural hearing loss

Non-syndromic ocular abnormalities were observed in 11 patients (26.2%), including strabismus (5 cases, 11.9%), amblyopia (4 cases, 9.5%), macular coloboma (1 case, 2.4%) (Figure 5), and posterior uveitis (1 case, 2.4%).



**Figure 5** Chorioretinal coloboma of the left eye in an 11-year-old child during an ophthalmological examination as part of a pre-implantation assessment

Regarding auditory management, 22 patients (52.4%) were fitted with conventional hearing aids, while 14 patients (33.3%) underwent cochlear implantation. Cochlear implantation was primarily indicated for severe-to-profound bilateral sensorineural hearing loss, particularly in children with limited benefit from conventional amplification.

Among implanted patients, several presented with syndromic conditions, notably Usher syndrome. In these cases, cochlear implantation was performed at an early stage, especially when associated with progressive visual impairment, in order to ensure rapid auditory rehabilitation and to compensate for the anticipated visual decline.

However, some patients who met the criteria for cochlear implantation could not benefit from this intervention due to limited financial resources and lack of medical coverage.

All patients received speech and language therapy as part of a comprehensive rehabilitation program. This intervention aimed to promote auditory perception, speech development, and cortical integration of auditory input.

Early auditory rehabilitation was prioritized, particularly in children at risk of dual sensory impairment, to optimize communication outcomes and support neurocognitive development.

Overall management strategies are summarized in Table 2

**Table 2** Distribution of auditory management modalities in hearing-impaired children with ocular involvement.

| Type of management    | Number of cases (n) | Percentage (%) |
|-----------------------|---------------------|----------------|
| Hearing aids          | 22                  | 52.4           |
| Cochlear implantation | 14                  | 33.3           |
| Speech therapy        | 42                  | 100            |

Ophthalmological management included optical correction in 31 cases (73.8%) and surgical treatment in selected patients, including cataract surgery (2 cases, 4.8%), corneal transplantation (2 cases, 4.8%), dermoid excision (3 cases, 7.1%), and ectropion repair (2 cases, 4.8%).

Orthoptic rehabilitation was prescribed for all patients.

#### 4. Discussion

In our study, ocular involvement was identified in 6.1% of hearing-impaired children. While this prevalence appears lower than that reported in the literature, where rates ranging from 20% to 60% have been described [5,6], this difference should not be interpreted solely as a lower occurrence. It may rather reflect variations in screening practices, differences in study populations, and the potential underdiagnosis of subtle or early-stage ophthalmological abnormalities, particularly in young children (Table 3).

**Table 3** Comparison of ocular involvement prevalence across studies

| Study                | Population             | Prevalence |
|----------------------|------------------------|------------|
| Mafong et al. (6)    | Pediatric hearing loss | ~40%       |
| Boumendil et al. (2) | Syndromic deafness     | Up to 80%  |
| Present study        | 688 children           | 6.1%       |

More importantly, the clinical significance of ocular involvement goes beyond its prevalence. Even a relatively low frequency remains relevant given the functional consequences of combined auditory and visual impairment. The coexistence of these deficits exposes children to a dual sensory handicap, which may profoundly affect language acquisition, cognitive development, and social integration [3,7]. This highlights the importance of systematic screening regardless of the apparent severity or etiology of hearing loss. In addition, the reported prevalence is strongly influenced by the proportion of syndromic forms and the extent of systematic ophthalmological evaluation.

The association between auditory and visual disorders can be explained by shared embryological and genetic mechanisms involving the inner ear and ocular structures, particularly the retina [2,3]. This common origin accounts for the frequent coexistence of these impairments, especially in syndromic conditions.

In our series, retinal involvement was the most common ocular abnormality, followed by adnexal and iris involvement. This distribution aligns with previous studies, particularly in syndromic conditions such as Usher and Alström syndromes, where retinal degeneration is a major feature [4,8]. The predominance of retinal lesions is clinically significant because these conditions are often progressive and may lead to severe visual impairment.

The high proportion of bilateral ocular involvement (75%) observed in our study is also consistent with findings reported by Mafong et al. [5], who emphasized the frequent association of bilateral and progressive ocular lesions in children with sensorineural hearing loss. Such findings strongly suggest an underlying genetic or syndromic origin, even in the absence of obvious dysmorphic features. While the diagnosis of oculo-auditory syndromes may be straightforward in the presence of craniofacial dysmorphism, it remains challenging when only neurosensory deficits are present.

In addition to syndromic conditions, non-syndromic ocular abnormalities such as strabismus and amblyopia were identified in our cohort. Although generally less severe, these conditions remain clinically relevant, as they may lead to permanent visual impairment if not detected early. Similar observations have been reported in the literature [2,3], highlighting that even non-syndromic hearing loss may be associated with visual disorders requiring early detection and management.

The identification of ocular involvement has direct implications for therapeutic strategy. In our study, cochlear implantation was preferentially performed in children with progressive visual impairment, particularly in syndromic conditions such as Usher syndrome. This approach is supported by several studies emphasizing that early cochlear implantation is crucial in children at risk of visual loss to ensure adequate language development before the onset of severe visual impairment [9,10]. Early auditory rehabilitation promotes optimal cortical integration of auditory stimuli and improves long-term communication outcomes.

However, our study also highlights disparities in access to cochlear implantation. Some patients who met the criteria for implantation could not benefit from this intervention due to financial constraints and lack of medical coverage. Similar limitations have been reported in low- and middle-income countries, where access to advanced hearing rehabilitation remains unequal [11]. This aspect should be considered when interpreting functional outcomes.

Finally, our findings confirm the central role of systematic ophthalmological screening in hearing-impaired children. Early identification of ocular involvement not only facilitates the diagnosis of syndromic conditions but also allows timely adaptation of therapeutic strategies, particularly regarding the timing of auditory rehabilitation. Multidisciplinary management involving otorhinolaryngologists, ophthalmologists, and rehabilitation specialists is essential to optimize functional outcomes in children with dual sensory impairment [3,7,12-15].

However, this study has some limitations, including its retrospective design and the potential underestimation of subtle ophthalmological abnormalities. In addition, the single-center nature of the study may limit the generalizability of the findings.

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## 5. Conclusion

Ocular involvement in hearing-impaired children is relatively frequent, particularly in syndromic conditions, with a predominance of retinal abnormalities and bilateral presentation suggesting a possible genetic origin. These findings emphasize the need for systematic ophthalmological evaluation in all children with hearing loss to ensure early detection and appropriate management, especially in those at risk of dual sensory impairment. A multidisciplinary approach combining auditory rehabilitation and ophthalmological care is essential to optimize outcomes, while improving access to advanced treatments such as cochlear implantation remains a major challenge in resource-limited settings.

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## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

### *Statement of ethical approval*

This retrospective study was conducted in accordance with institutional guidelines and the principles of the Declaration of Helsinki.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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