

Research Article

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Risk Factors for Severe-To-Profound Sensorineural Hearing Loss in Cochlear Implant Recipients: Consanguinity, Bacterial Meningitis and The Perinatal Cluster in 369 Patients

I Ghoulzani*, R Chelly, S Bensimimou, M Lahjaoui, M Loudghiri, W Bijou, Y Oukessou, S Rouadi, R L Abada, M Roubal and M Mahtar

Department of Otorhinolaryngology and Head and Neck Surgery, Hôpital 20 Août 1953, CHU Ibn Rochd, Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco

ABSTRACT

Background and Objective: The risk factor profile of severe-to-profound sensorineural hearing loss (SNHL) differs markedly between populations, and it determines which preventive measures are worth deploying. In settings where consanguineous marriage is common and bacterial meningitis is still frequent, that profile has rarely been characterised in a large cohort. We analysed the prevalence of recognised risk factors in 369 cochlear implant recipients and their association with the clinical phenotype of the hearing loss.

Methods: All patients implanted in a single tertiary referral department between January 2010 and December 2024 were reviewed. Sixteen predefined risk factors - familial, perinatal and acquired were abstracted from a standardized institutional registry, together with the type of hearing loss (congenital, genetic/familial, acquired), its period of onset relative to language acquisition, and its laterality. Prevalences were compared between phenotypic groups using the chi-square or Fisher exact test; odds ratios (OR) with 95% confidence intervals (CI) were estimated with the Haldane Anscombe correction.

Results: Parental consanguinity was the single most prevalent risk factor (123 patients; 33.3%), followed by a family history of deafness (108; 29.3%) and bacterial meningitis (43; 11.7%). The two familial factors were tightly linked: a family history of deafness was present in 69.1% of consanguineous families versus 9.0% of non-consanguineous ones (OR 22.6, 95% CI 12.6–40.4; $p < 0.001$). Consanguinity and family history were both associated with congenital or genetic hearing loss (OR 3.69 and 4.42; $p < 0.001$) and with prelingual onset (OR 2.73 and 3.31; $p < 0.001$), whereas meningitis, neonatal distress, prematurity, neonatal hospitalisation, recurrent chronic otitis media and head trauma were all associated with acquired, later-onset disease. Perinatal factors clustered in 62 patients (16.8%). No identifiable risk factor was found in 137 patients (37.1%). Maternal rubella and cytomegalovirus infection were recorded in no patient.

Conclusions: The risk factor profile of this cohort is dominated by a familial axis, driven by consanguinity, and by a residual burden of bacterial meningitis that has largely disappeared from high-income series. These two axes call for different interventions: genetic counselling and targeted molecular diagnosis for the first, and completion of pneumococcal and Haemophilus influenzae type b vaccination coverage with systematic post-meningitis audiological assessment for the second. The absence of any documented congenital cytomegalovirus infection, in a cohort where 37.1% of patients had no identifiable risk factor, almost certainly reflects under-testing rather than true absence.

*Corresponding author

I Ghoulzani, Department of Otorhinolaryngology and Head and Neck Surgery, Hôpital 20 Août 1953, CHU Ibn Rochd, Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco.

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Introduction

Severe-to-profound sensorineural hearing loss is the end point of a heterogeneous set of causes: monogenic disorders, perinatal insults, infections of the inner ear and the central nervous system, ototoxicity and trauma. The relative weight of these causes is not universal. It varies with the local rate of consanguineous marriage, with vaccination coverage, with the quality of perinatal care and with access to antenatal screening, and it shifts over time as those determinants change [1,2].

That variation matters clinically because it dictates which preventive and diagnostic measures are worth deploying. Where hereditary deafness predominates, genetic counselling and targeted molecular diagnosis carry the highest yield; where post-meningitic deafness persists, vaccination coverage and systematic audiological assessment after meningitis are the priority; where perinatal insults dominate, targeted neonatal hearing screening of at-risk newborns is the most efficient strategy. In Morocco, consanguineous marriage remains frequent and bacterial meningitis has not disappeared as a cause of acquired deafness, yet the risk factor profile of the severely deaf population has been described mainly through the lens of molecular genetics rather than through the full spectrum of familial, perinatal and acquired factors [3].

Cochlear implant recipients constitute a useful, if selected, window on this population: they are systematically and uniformly investigated, and their records capture familial, perinatal and acquired exposures in a standardized manner. We therefore analysed the prevalence of sixteen predefined risk factors in 369 consecutive implant recipients, examined how these factors cluster, and tested their association with the type of hearing loss, its period of onset and its laterality.

Materials and Methods

Design and Population

This was a retrospective, single-centre, cross-sectional analysis of all patients who underwent cochlear implantation in the Department of Otorhinolaryngology and Head and Neck Surgery of Hôpital 20 Août 1953, CHU Ibn Rochd, Casablanca, between January 2010 and December 2024. Records containing fewer than 20% of the registry items were excluded as non-analysable, leaving 369 consecutive patients. All had bilateral or unilateral severe-to-profound sensorineural hearing loss meeting national implantation criteria.

Risk Factors and Definitions

Sixteen risk factors were defined a priori and recorded as present or absent at the time of candidacy assessment, on the basis of a structured parental or patient interview supplemented by available medical documents. Familial factors were parental consanguinity (first- or second-degree cousin marriage) and a family history of deafness in a first- or second-degree relative. Antenatal and perinatal factors were maternal rubella, cytomegalovirus infection, a pregnancy reported as not well conducted, a complicated delivery, prematurity, neonatal distress, neonatal jaundice and neonatal hospitalisation. Acquired factors were bacterial meningitis, mumps, recurrent chronic otitis media, head trauma, exposure to ototoxic drugs and exposure to loud noise. A congenital malformation of any system was recorded separately. The perinatal cluster was defined as the presence of at least one of prematurity, neonatal distress, neonatal jaundice or neonatal hospitalisation.

The hearing loss was classified by the referring team as congenital, genetic or familial, or acquired; by its period of onset relative to language acquisition as prelingual, perilingual or postlingual; and by its laterality. All patients underwent a standardized audiological workup, high-resolution computed tomography of the temporal bones and magnetic resonance imaging of the cerebellopontine angle and brain. Systematic molecular genetic testing and congenital cytomegalovirus screening were not available during the study period.

Statistical Analysis

Analyses were performed with Python 3.12 (pandas, SciPy). Prevalences are given as counts and percentages of the number of patients with available data. Comparisons between phenotypic groups used the Pearson chi-square test, or the Fisher exact test when expected counts were below 5. Odds ratios with 95% confidence intervals were estimated using the Haldane–Anscombe continuity correction, since several strata contained zero cells. A two-sided p value below 0.05 was considered significant. Because the aetiological label (congenital, genetic or acquired) was assigned partly on the basis of the same exposures, no analysis was performed of the association between an individual risk factor and the presence of an “identified aetiology”, which would have been circular by construction. Given the exploratory nature of the analysis, no correction for multiple comparisons was applied and no multivariable model was fitted.

Ethical Considerations

The study was approved by the institutional ethics committee and conducted in accordance with the Declaration of Helsinki. All data were anonymised before analysis, and informed consent for the anonymous use of clinical data was obtained from all patients or their legal guardians.

Results

Prevalence of Risk Factors

Among the 369 patients, parental consanguinity was the most prevalent risk factor, reported in 123 families (33.3%), followed by a family history of deafness in 108 (29.3%). Bacterial meningitis, the leading acquired factor, was reported in 43 patients (11.7%). Neonatal hospitalisation (40; 10.8%), neonatal distress (37; 10.0%) and recurrent chronic otitis media (35; 9.5%) came next, followed by prematurity (19; 5.1%), congenital malformation (13; 3.5%), neonatal jaundice (10; 2.7%), head trauma (9; 2.4%) and ototoxic drug exposure (8; 2.2%). Loud noise exposure (4), complicated delivery (3) and mumps (2) were rare. Notably, maternal rubella and cytomegalovirus infection were documented in no patient, and the pregnancy was reported as well conducted in 347 of 369 women (94.0%). The full prevalence profile is shown in Figure 1 and Table 1.

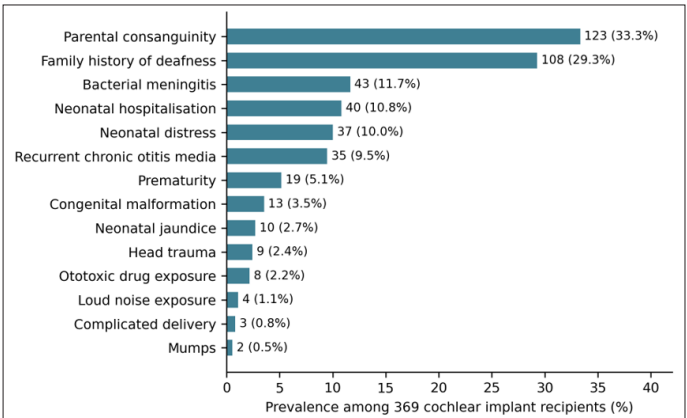


Figure 1: Prevalence of the Sixteen Predefined Risk Factors Among 369 Cochlear Implant Recipients. Maternal Rubella and Cytomegalovirus Infection were Documented in no Patient and are not Displayed

Table 1: Prevalence of Risk Factors (N = 369)

Risk factor	n	%
Parental consanguinity	123	33.3
Family history of deafness	108	29.3
Bacterial meningitis	43	11.7
Neonatal hospitalisation	40	10.8
Neonatal distress	37	10.0
Recurrent chronic otitis media	35	9.5
Prematurity	19	5.1
Congenital malformation	13	3.5
Neonatal jaundice	10	2.7
Head trauma	9	2.4
Ototoxic drug exposure	8	2.2
Loud noise exposure	4	1.1
Complicated delivery	3	0.8
Mumps	2	0.5

Maternal rubella	0	0
Cytomegalovirus infection	0	0
Perinatal cluster (≥ 1 perinatal factor)	62	16.8

Accumulation and Clustering of Risk Factors

One hundred thirty-seven patients (37.1%) had no identifiable risk factor at all, 153 (41.5%) had a single factor, 52 (14.1%) had two and 27 (7.3%) had three or more (Figure 2). Perinatal factors clustered: at least one of prematurity, neonatal distress, neonatal jaundice or neonatal hospitalisation was present in 62 patients (16.8%), and these factors co-occurred far more often than expected by chance, reflecting the shared underlying situation of a compromised newborn requiring intensive care.

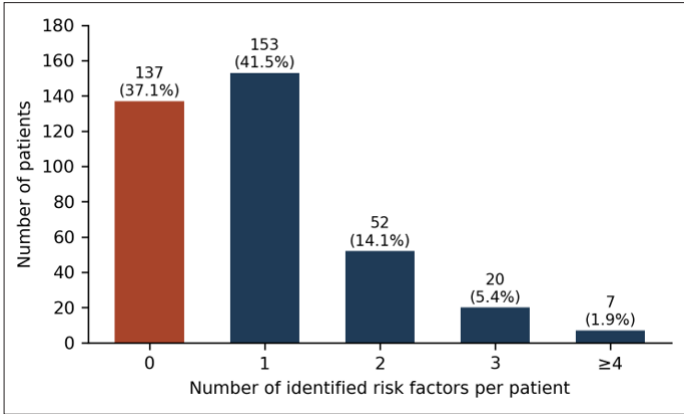


Figure 2: Number of Identified Risk Factors Per Patient (N = 369)

The two familial factors were the most tightly linked pair in the dataset. A family history of deafness was reported in 85 of 123 consanguineous families (69.1%) versus 22 of 244 non-consanguineous families (9.0%), an odds ratio of 22.6 (95% CI 12.6–40.4; $p < 0.001$) (Figure 7). Consanguinity was somewhat more frequent among patients of rural origin (38.0% versus 30.8%), but the difference was not significant ($p = 0.171$), and it did not differ by socioeconomic stratum ($p = 0.905$).

Risk Factors and Type of Hearing Loss

The hearing loss was classified as acquired in 152 patients (41.2%), congenital in 150 (40.7%) and genetic or clearly familial in 67 (18.2%). The risk factor profile separated these groups sharply (Figure 3). Parental consanguinity was reported in 71.2% of the genetic group, 32.9% of the congenital group and 17.8% of the acquired group ($p < 0.001$), and a family history of deafness in 83.6%, 21.3% and 13.2% respectively ($p < 0.001$). The pattern reversed for every acquired exposure: bacterial meningitis was reported in 27.6% of the acquired group versus 0.7% and 0% of the congenital and genetic groups ($p < 0.001$), and the same gradient was seen for neonatal hospitalisation (21.7% vs 1.3% and 7.5%; $p < 0.001$), neonatal distress (19.7% vs 2.0% and 6.0%; $p < 0.001$), recurrent chronic otitis media (20.0% vs 2.0% and 3.0%; $p < 0.001$), prematurity (10.5% vs 1.3% and 1.5%; $p < 0.001$) and head trauma (5.9% vs 0% and 0%; $p = 0.001$).

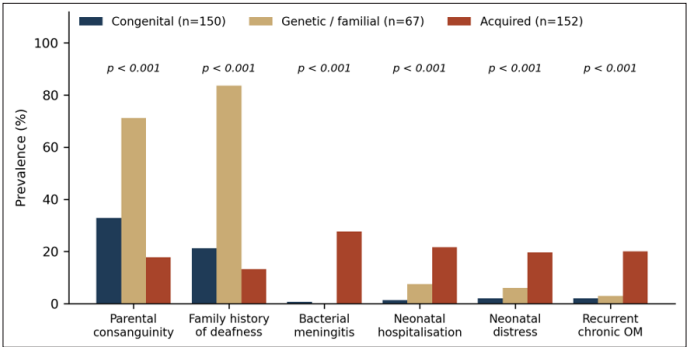


Figure 3: Prevalence of the Six Commonest Risk Factors According to the Type of Hearing Loss. OM: Otitis Media

Expressed as odds ratios for congenital or genetic (as opposed to acquired) hearing loss, a family history of deafness carried an OR of 4.42 (95% CI 2.58–7.56; $p < 0.001$) and parental consanguinity an OR of 3.69 (95% CI 2.25–6.03; $p < 0.001$), while bacterial meningitis (OR 0.02), head trauma (OR 0.03), recurrent chronic otitis media (OR 0.10), prematurity (OR 0.13), neonatal hospitalisation (OR 0.13), neonatal distress (OR 0.14) and ototoxic drug exposure (OR 0.13) were all inversely associated. A congenital malformation was not associated with either group (OR 1.50; $p = 0.571$) (Figure 4).

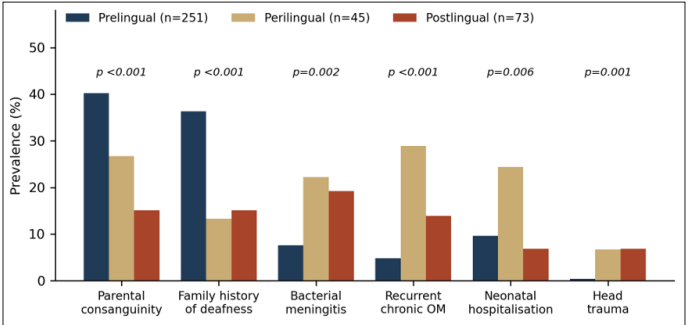


Figure 4: Risk Factors Associated with Congenital or Genetic Hearing loss, as Opposed to Acquired hearing Loss. Odds Ratios with 95% Confidence Intervals, Logarithmic Scale. Significant Associations are Shown in Red

Risk Factors and Period of Onset

Onset was prelingual in 251 patients (68.0%), perilingual in 45 (12.2%) and postlingual in 73 (19.8%). The familial axis again predicted early onset: consanguinity was present in 40.2% of prelingual, 26.7% of perilingual and 15.1% of postlingual patients ($p < 0.001$), and a family history of deafness in 36.3%, 13.3% and 15.1% respectively ($p < 0.001$). Conversely, bacterial meningitis was reported in 7.6% of prelingual patients but 22.2% of perilingual and 19.2% of postlingual patients ($p = 0.002$), recurrent chronic otitis media in 4.8%, 28.9% and 13.9% ($p < 0.001$), and head trauma in 0.4%, 6.7% and 6.8% ($p = 0.001$). The perilingual group — small but clinically distinctive — carried the heaviest burden of acquired and perinatal exposures, consistent with an insult occurring during the period of language acquisition (Figures 5 and 6).

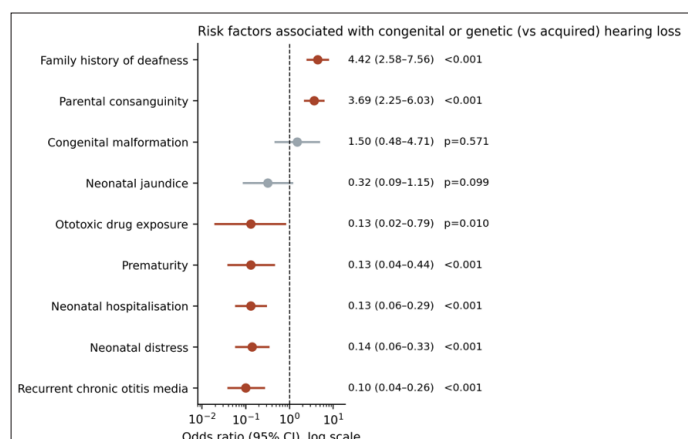


Figure 5: Prevalence of Risk Factors According to the Period of Onset Relative to LANGUAGE Acquisition. OM: Otitis Media

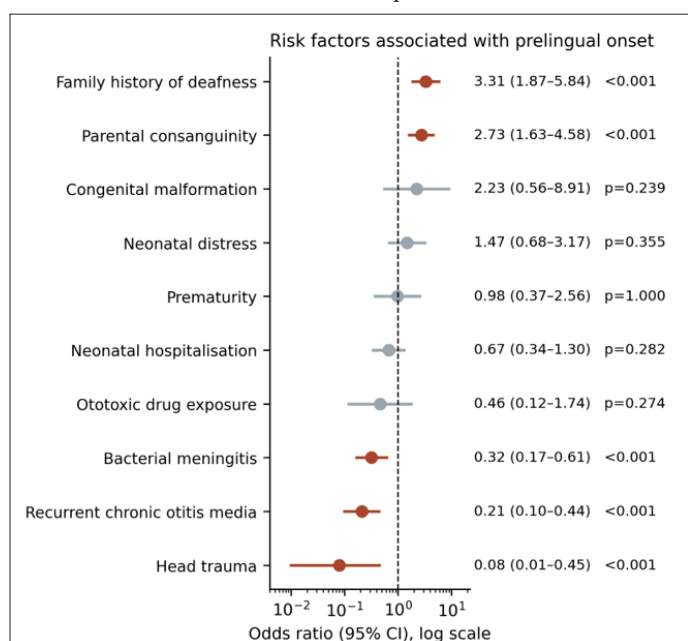


Figure 6: Risk Factors Associated with Prelingual Onset. Odds Ratios with 95% Confidence intervals, Logarithmic Scale

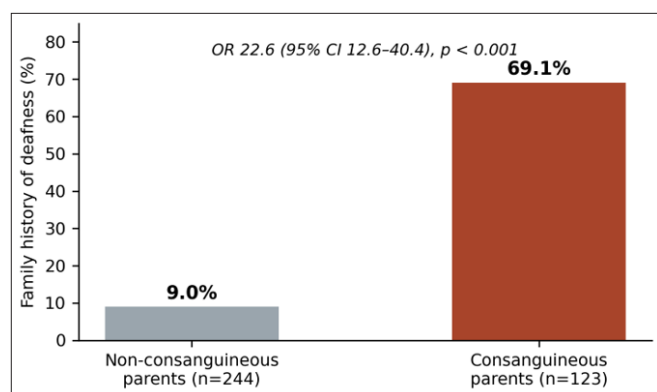


Figure 7: Family History of Deafness According to Parental Consanguinity

Hearing Loss was Bilateral in 359 patients (97.3%). Bilaterality was Near-Universal in Both Consanguineous (97.6%) and Non-Consanguineous Families (97.1%), so no Association Could be Tested Meaningfully.

Table 2: Prevalence of the Principal Risk Factors by Type of Hearing Loss and by Period of Onset

Risk factor	Congenital	Genetic	Acquired	p
Parental consanguinity	32.9%	71.2%	17.8%	<0.001
Family history of deafness	21.3%	83.6%	13.2%	<0.001
Bacterial meningitis	0.7%	0%	27.6%	<0.001
Neonatal hospitalisation	1.3%	7.5%	21.7%	<0.001
Neonatal distress	2.0%	6.0%	19.7%	<0.001
Recurrent chronic otitis media	2.0%	3.0%	20.0%	<0.001
Prematurity	1.3%	1.5%	10.5%	<0.001
Head trauma	0%	0%	5.9%	0.001
Risk factor	Prelingual	Perilingual	Postlingual	p
Parental consanguinity	40.2%	26.7%	15.1%	<0.001
Family history of deafness	36.3%	13.3%	15.1%	<0.001
Bacterial meningitis	7.6%	22.2%	19.2%	0.002
Recurrent chronic otitis media	4.8%	28.9%	13.9%	<0.001
Neonatal hospitalisation	9.6%	24.4%	6.8%	0.006
Head trauma	0.4%	6.7%	6.8%	0.001

Discussion

Two axes structure the risk factor profile of this cohort. The first is familial and is driven by consanguinity; the second is acquired and is dominated by bacterial meningitis and by a cluster of perinatal insults. They point to entirely different preventive strategies, and the balance between them differs substantially from that of contemporary European and North American series, where hereditary deafness is identified mainly through molecular testing and where post-meningitic deafness has become rare.

The Familial Axis

Parental consanguinity, present in a third of our families, was the most prevalent risk factor and the strongest correlate of the congenital and genetic phenotypes. Its effect is mediated almost entirely through familial recurrence: a family history of deafness was more than seven times as frequent in consanguineous families, with an odds ratio above 22. This is the expected signature of autosomal recessive non-syndromic hearing loss, in which cousin marriage raises the probability that both parents carry the same pathogenic variant.

Our figures are consistent with those recently reported in a Moroccan implanted paediatric population, in which 55.7% of families were consanguineous and next-generation sequencing identified a causative variant in 65.9% of cases, GJB2 being the commonest gene involved [3]. Molecular testing was not available to us during the study period, and this is the most consequential gap in the present analysis: 137 patients (37.1%) had no identifiable risk factor, a group in which recessive genetic causes are very likely to be over-represented. Introducing targeted genetic testing into the candidacy workup would convert a substantial share of this idiopathic group into a diagnosed one, with direct benefits for family counselling, recurrence risk estimation and, increasingly, eligibility for emerging gene-directed therapies.

The practical implication of the consanguinity family history association is that the family, not only the patient, is the appropriate

unit of assessment. When a deaf child of consanguineous parents presents for implantation, systematic audiological screening of siblings and genetic counselling for the couple are justified on the strength of the recurrence risk alone, irrespective of whether molecular testing is available.

The Acquired Axis

Bacterial meningitis accounted for 11.7% of our cohort and for more than a quarter of the acquired group a proportion that has largely disappeared from high-income series following the introduction of conjugate pneumococcal and *Haemophilus influenzae* type b vaccines [4,5]. Sixteen per cent of our patients had not completed pneumococcal or *Haemophilus influenzae* type b vaccination at the time of candidacy, and completing coverage is the single most effective preventive measure available for this axis.

The consequences of post-meningitic deafness extend beyond its frequency. Labyrinthitis ossificans develops within weeks of the infection and begins at the round window and the basal turn, precisely where the electrode array must enter, so that a delayed referral converts a straightforward implantation into a technically difficult one [6,7]. Every child surviving bacterial meningitis should therefore undergo audiological assessment before hospital discharge, and confirmed profound hearing loss should be referred for urgent rather than routine implantation.

Recurrent chronic otitis media, present in 9.5% of the cohort and in 20% of the acquired group, deserves separate mention. Its association with the perilingual group (28.9%) suggests that a proportion of these children experienced progressive hearing loss during the very period of language acquisition, when middle ear disease is most prevalent and most likely to be dismissed as a transient conductive problem.

The Perinatal Cluster and the Absence of Documented Congenital Infection

Perinatal factors prematurity, neonatal distress, jaundice and neonatal hospitalisation were present in 16.8% of patients and co-occurred systematically, as expected given that they largely describe one clinical situation: the newborn requiring intensive care. This cluster is precisely the population targeted by risk-based neonatal hearing screening in countries without universal screening, and it represents the most easily identified group in which such a programme could be piloted at low cost.

The most striking negative finding of this study is the complete absence of documented maternal rubella and congenital cytomegalovirus infection. Congenital cytomegalovirus is recognised as the leading non-genetic cause of congenital SNHL worldwide, accounting for a substantial share of cases in every population in which it has been systematically sought [8,9]. Its apparent absence here almost certainly reflects the fact that it was never tested for: diagnosis requires viral detection within the first three weeks of life or retrospective testing of the neonatal dried blood spot, neither of which was routinely available. Combined with the 37.1% of patients without any identifiable risk factor, this suggests that a meaningful fraction of the “idiopathic” group in our cohort is in fact congenital cytomegalovirus disease. Adding targeted cytomegalovirus testing to the workup of infants who fail hearing screening would be a low-cost, high-yield addition to current practice.

Limitations

Several limitations should temper the interpretation of these results. First, this is a study of prevalence and of internal

associations within an implanted cohort, not a case-control study: without a hearing control group, no true relative risk of deafness can be estimated, and the odds ratios reported describe associations between exposures and clinical phenotype within the deaf population only. Second, cochlear implant recipients are a selected subset of the deaf population those meeting audiological criteria and reaching a tertiary centre so the profile described may not represent all severe-to-profound hearing loss in Morocco. Third, exposures were recorded retrospectively from parental interview and available documents, with the recall bias inherent to that method; perinatal factors in particular are likely to be under-reported for older patients. Fourth, the aetiological classification was clinical and radiological, without molecular or virological confirmation, and the analysis of the “identified aetiology” endpoint was deliberately omitted as circular. Finally, no correction for multiple comparisons was applied and no multivariable model was fitted, so the reported associations should be read as effect directions rather than adjusted estimates.

Conclusions

In 369 cochlear implant recipients, the risk factor profile of severe-to-profound sensorineural hearing loss was dominated by two distinct axes. A familial axis, driven by parental consanguinity in a third of families and strongly associated with a family history of deafness, with congenital or genetic disease and with prelingual onset, calls for genetic counselling, sibling screening and the introduction of targeted molecular diagnosis. An acquired axis, dominated by bacterial meningitis and by a perinatal cluster present in one patient in six, calls for completion of conjugate vaccination coverage, systematic audiological assessment after meningitis with expedited implantation, and risk-based neonatal hearing screening of newborns requiring intensive care. The absence of any documented congenital cytomegalovirus infection, alongside 37.1% of patients with no identifiable risk factor, identifies the most immediately actionable gap in the current diagnostic pathway [10-15].

Declarations

Ethics Approval: Approved by the institutional ethics committee of CHU Ibn Rochd, Casablanca. Informed consent was obtained from all patients or their legal guardians.

Conflicts of Interest: The authors declare no conflicts of interest.

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Author Contributions: All authors contributed to the conception of the study, data collection, analysis and critical revision of the manuscript, and approved the final version.

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