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Genetic Landscape of Hearing Loss in the Caribbean: A Narrative Review

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The Caribbean region has a diverse population of about 40 million people, spread over 13 sovereign states. This review aims to describe the existing studies on hereditary hearing loss (HL) in the Caribbean population. We systematically reviewed scientific articles on HL prevalence, genetic causes, technology use, and environmental effects in Caribbean nations and the Caribbean diaspora in the United States. Key findings show that HL rates, with diverse genetic variables, vary across Puerto Rico, Cuba, and the Dominican Republic. Local resources and technology have been used to diagnose HL, particularly in rural areas. Environmental factors tend to affect HL prevalence in various regions. This literature review of Caribbean-focused studies helps guide future research and healthcare strategies, particularly concerning genetic drift caused by migration to the United States. Understanding these factors can help diagnose and treat HL in America's diverse population.

The Caribbean, which includes 13 sovereign states and a population of more than 44 million people, is a mosaic of ethnicities comprising African, European, Indian, and Indigenous communities (Figure 1, Table 1).[1,2] Despite several genetic studies on various disorders in the region, hearing loss (HL) research remains limited.[3] HL in at least 50% of the affected people with congenital or prelingual-onset HL is thought to be genetic in origin.[4,5] Genetic HL can be syndromic (30% of inherited HL) or non-syndromic (70% of inherited HL). The most common form of inheritance is autosomal recessive (AR), which accounts for up to 80% of all individuals.[6-8] Autosomal dominant inheritance accounts for approximately 20% of cases, with the remaining 5% belonging to X-linked and mitochondrial inheritance forms.[9]

Online Mendelian Inheritance in Man includes around 600 deafness diseases as phenotypic findings, either isolated or as part of a syndrome.[10] Moreover, the Hereditary Hearing Loss Homepage lists over 150 genes associated with non-syndromic HL.[11] Previous studies have shown that the etiological diagnostic rate of HL after genetic testing of genes and variants differs significantly among ethnic and racial groups, with some of these studies including a Caribbean population.[12,13] *GJB2* variants underlying AR non-syndromic HL have been identified as the most common cause in people of European and Asian ancestry.[14,15] In contrast, *GJB2* variants are rare in Black people.[16-18] Moreover, *GJB2* is not considered a prominent cause of HL among Caribbean Hispanic populations.[19] A previous study of a highly mixed Caribbean population found increased *HL* gene heterogeneity in the racial and ethnically diverse cohort.[20]

While much less common than *GJB2* variants, multi-ethnic cohort studies from throughout the world have found that *STRC*, *MYO15A*, *SLC26A4*, *USH2A*, *MYO7A*, *OTOF*, *CDH23*, *TMC1*, *TECTA*, *MYO6*, and *TRIOBP* variants are relatively common compared with other gene variants.[12,13]

The growing area of genetics and genomics is transforming our approach to molecular diagnosis and enabling the development of gene-based treatments for disorders such as HL.[21-23] The efficacy of such treatments and the equitable benefit from these exciting developments depend on our understanding of the genes involved and their global distribution.

Caribbean immigrants have contributed significantly to the genetic fabric of the United States, notably in Florida, New York, New Jersey, and Massachusetts. This vigorous diaspora has brought novel genetic variants, increasing America's genetic diversity and expanding the possibilities for scientific research and health resilience. There is also evidence of a relationship between self-reported HL and frailty among Afro-Caribbeans, which is not shown in other studied cohorts such as AAs, Hispanics, and European Americans.[24]

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Despite growing interest and advancements in HL research, the Caribbean's genetic contributions to HL remain unexplored. This review aims to close this gap by examining studies that investigate the genetic etiologies of HL in the Caribbean and to guide future research into the genetics of HL in this culturally and genetically diverse region.

METHODS

Literature search strategy: A systematic literature review was conducted to identify relevant studies on HL in the Caribbean islands. A comprehensive search of two electronic databases, PubMed and Google Scholar, was conducted from their establishment until August 21, 2023. The search strategy used a combination of relevant

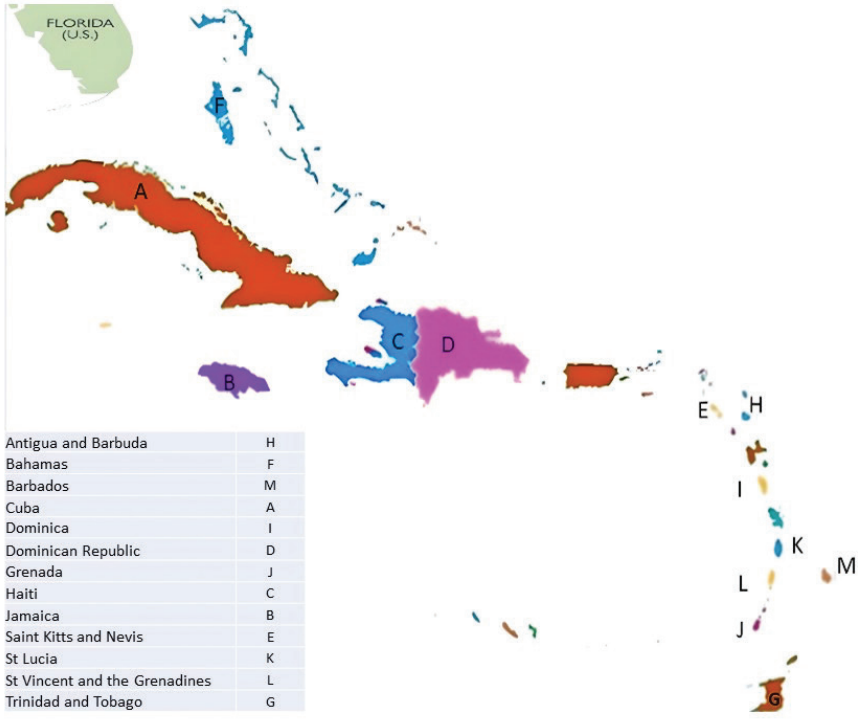

FIG. 1. Map of the Caribbean Islands.

TABLE 1. Ethnic Groups in Caribbean Countries.

Caribbean country	Population size *	African descent (%)	European descent (%)	Indigen (%)	Mixed (%)	East Indian (%)	Other (%)
Antigua and Barbuda	101,489	87.1-95.1	0.1-1.7	N/A	2.9-3.9	0.1-1.1	0.6-3.7
Bahamas	358,508	90.6	4.7	N/A	2.1	N/A	1.9
Barbados	303,431	92.4	2.7	N/A	3.1	1.3	0.3
Cuba	10,985,974	9.3	64.1	N/A	26.6	N/A	N/A
Dominica	74,656	75	0.8	4	19	N/A	N/A
Dominican Republic	10,790,744	7.8	17.8	N/A	73.9	N/A	3.2
Grenada	114,299	82.4	N/A	N/A	13.3	2.2	1.3
Haiti	11,470,261	95	N/A	N/A	5	N/A	N/A
Jamaica	2,820,982	67.1	N/A	N/A	31.1	0.8	0.4
Saint Kitts and Nevis	54,817	92.5	2.1	N/A	3	1.5	0.6
St. Lucia	167,591	85.3	N/A	N/A	10.9	2.2	1.6
St. Vincent and the Grenadines	100,804	66	4	2	19	6	3
Trinidad and Tobago	1,407,460	34.2	N/A	N/A	22.8	35.4	N/A

*Population size data from the World Fact Book (2023).

keywords and controlled vocabulary terms. The search keywords included "HL," "deafness," "sensorineural hearing loss or SNHL," and "Caribbean." To ensure comprehensive coverage, the search strategy included specific names for Caribbean countries and territories such as Antigua and Barbuda, the Bahamas, Barbados, Cuba, Dominica, Dominican Republic, Grenada, Haiti, Jamaica, St. Kitts and Nevis, St. Lucia, St. Vincent and the Grenadines, and Trinidad and Tobago.

Eligibility criteria: The review includes studies that met the following criteria: Case reports, reviews, prospective and retrospective observational studies, and research articles were eligible for inclusion. Studies on HL in the Caribbean population were included. The review excluded review articles and studies that did not focus on the Caribbean population or HL disorders. Furthermore, articles that did not have a comprehensive abstract when the full text was unavailable were excluded.

To ensure the review was comprehensive, the reference lists of the included studies were manually examined for any other studies that met the eligibility criteria but were not captured by the electronic database search.

Data extraction and synthesis: Data were retrieved from the selected studies using a standardized data extraction form. The following information was collected: (a) study characteristics: author(s), publication year, study design, and geographical focus; (b) population characteristics: sample size, age range, and inclusion/exclusion criteria; (c) HL assessment: methods used for HL assessment, severity classification, and diagnostic criteria; (d) key findings: HL prevalence, etiological factors, local HL evaluation techniques, and genetic expansion.

Given the predicted heterogeneity in study designs, populations, and outcomes, the results were summarized using a narrative synthesis approach. The results were categorized based on the geographical origins of the studies and the main themes that emerged, such as prevalence rates, risk factors, genetic findings, and HL therapies in the Caribbean population.

Ethical considerations: This review exclusively utilized publicly available published data and did not involve human subjects; therefore, ethical approval was unnecessary.

Limitations: The review may be limited by publication bias because it only included published studies available in the specified databases. Furthermore, the quality of different research may vary, affecting the overall robustness of the findings.

This comprehensive literature review aimed to investigate the existing knowledge about HL in the Caribbean islands. The review approach included a thorough search strategy, study selection, and data extraction to provide a clear and unbiased synthesis of the available information.

SUMMARY OF EVIDENCES

Between 1986 and 2022, 16 studies on HL in the Caribbean were identified, revealing five common themes: HL prevalence (Table 2), genotypic and phenotypic expansions (Figure 2), genetic drift and founder effect, utilization of local resources and technology, and environmental factors influencing HL.

HL prevalence in the Caribbean population

Research on diverse Caribbean communities, particularly Dominicans and Puerto Ricans in the United States, has revealed slight differences in the prevalence of HL (Table 2). A detailed population-based study of three U.S. areas with significant Caribbean Hispanic representation found that HL prevalence varied by ethnicity, age, and gender, with Puerto Rican males and older people having higher rates.[25] Further research on childhood-onset HL in Puerto Rico suggested a 3.6% prevalence, with slightly varying rates reported in the Dominican Republic and Jamaica, reflecting the region's heterogeneity.[26-31]

Genotypic and phenotypic expansions

Studies on the genetic causes of HL in regions such as Puerto Rico have revealed a mix of syndromic and non-syndromic forms of HL. One study from Puerto Rico yielded results from an annual national survey of 336 participants without molecular tests.[26] Of the recorded etiologies of HL, 15% were related to genetic or hereditary factors. Within this group, 2%, 2%, 4%, and 4% indicated Down, Usher, Treacher Collins, and Goldenhar syndromes, respectively; 16% indicated a familial history of HL, and 71% referred to "other"

TABLE 2. HL Prevalence in Caribbean Communities in the U.S. and in the Caribbean Islands

Age group		Cuban/U.S. (%)	Dominican/ U.S. (%)	Puerto Rican/U.S. (%)	Dominican/Island (%)	Jamaican/Island (%)	Puerto Rican/ Island (%)
< 18 y		N/A	N/A	N/A	4.95	4.9	3.6
18-44 y	Male	6.5	6.3	7.1	N/A	N/A	N/A
	Female	4.7	7.1	8.1	N/A	N/A	N/A
45 y+	Male	39.7	29.3	41.2	N/A	N/A	N/A
	Female	26.6	23.6	31.4	N/A	N/A	N/A
Total		20.6	14.1	21.2	4.95	2.7	3.6
References		Cruickshanks et al.,[25] 2015			Levy et al.,[27] 2018 and Urban et al.,[31] 2022	Inclusive education, 2021 and Lyn et al.,[28] 1998	Albertorio et al.,[26] 1999

HL, hearing loss; U.S., United States.

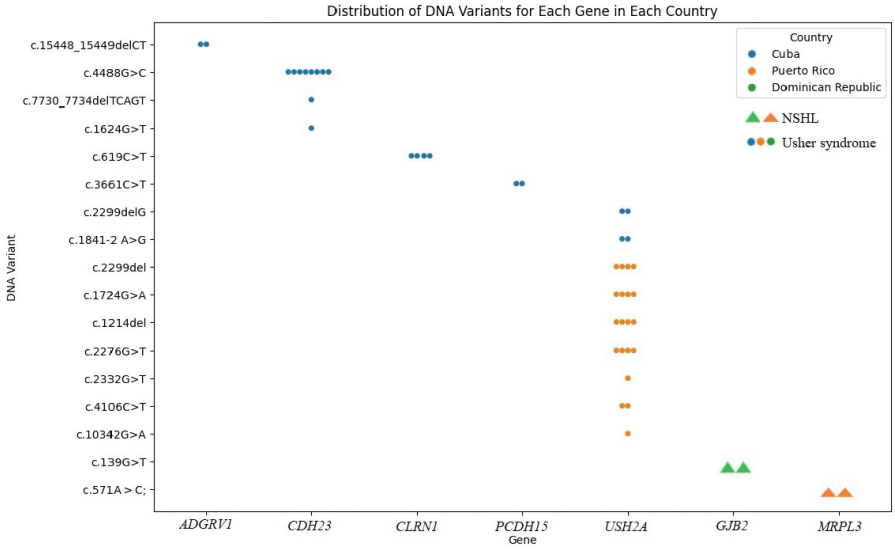

FIG. 2. The allele counts are represented. Each dot and triangle represent an allele.

unknown factors, which were closely associated with genetic components.

Another study focused on the presence of rare mutations in *GJB2*, *GJB6*, and mitochondrial DNA (mtDNA) in the African American (AA)/Black and Caribbean Hispanic populations. The study investigated the genetics of 109 mostly simplex AA/Blacks and Caribbean Hispanics from the Dominican Republic and Puerto Rico and 187 control participants with the same ethnic background.[3] The researchers could not find the three specific deafness-associated point mutations in mtDNA (m.1555A>G, m.3243A>G, and m.7445A>G). The study found that only 1% of the population had biallelic *GJB2* pathogenic variants (*GJB2* c.139G>T; p.Glu47*), while there were no hemizygous *GJB6* deletions.[3] These findings indicate that *GJB2* pathogenic variants, *GJB6* deletions, and the three common mtDNA variants were not crucial in these minority groups.[3]

A study of 21 patients (unrelated probands) with Usher syndrome in Cuba and Puerto Rico found a few recurrent variants (Figure 2).[32,33] It was found that the p.Cys759Phe variant in the *USH2A* gene resulted in a severe ophthalmological phenotype despite previously being associated with a mild phenotype.[32,34] A case report in Puerto Rico revealed further phenotypic expansion, with the finding of a rare phenotype of macular edema in two patients with Usher syndrome.[35]

A young Puerto Rican girl developed combined oxidative phosphorylation deficiency-9 (COXPD9) due to a pathogenic homozygous variant c.571A > C; p.Thr191Pro in the *MRPL3* gene. This report described previously unreported features such as Leigh syndrome, cataracts, hypotonia, scoliosis, myopathy, exercise intolerance, childhood-onset cardiomyopathy, and microcephaly. This patient was the eldest recorded case of COXPD9 at 11 years old.[36]

Genetic drift and founder effect

A study published in 1986 demonstrated genetic drift and the founder effect in the Caribbean, as seen by the high prevalence of HL on the small island of Saint Barthélemy in the French West Indies. The island's historical population included a Swedish population and a Black population from neighboring British islands, which combined contributed to a distinctive genetic environment affected by genetic drift. The study's segregation analysis of audiometric indices across 165 Saint Barthélemy families revealed strong evidence for a significant genetic effect. The familial solid aggregation was linked to a single putative recessive gene with a relatively high frequency (0.40). Individuals homozygous for this putative gene were more susceptible to ototoxicity, revealing the rationale for the increased prevalence of HL in the population. This notable observation of a rare gene with a high frequency in a relatively isolated population indicates a founder effect. The concept that a small group of original settlers brought the gene and then increased in popularity due to their limited genetic diversity supports the idea of genetic drift and the founder effect shaping the genetic landscape of Saint Barthélemy.[37]

Using local resources and technology to improve HL detection

In 2005, a study conducted in Cuba revealed the long-term benefits of implementing innovative technology within their means. The study used the Cuban-developed AUDIX system, a novel technique for recording multiple auditory steady-state potentials. This innovative approach successfully supported early diagnosis and treatments for HL, resulting in improved patient outcomes.[38]

In 2018, researchers in Port-au-Prince, Haiti, used smartphone technology to improve treatment pathways for HL in resource-limited populations. The initiative of otoscopic images of the tympanic membrane and audiometric evaluations significantly

improved the diagnostic process. Using locally available resources, such as smartphones, enabled the early detection and management of hearing impairment. Among 122 people initially tested using the smartphone hearing assessment and otoscopy, 31 individuals were marked for further evaluation due to failed tests, indicating a significant 25.4% prevalence of potential HL cases.[39]

A related study in remote regions surrounding La Romana, Dominican Republic, emphasized the impact of technology even more. The study focused on 423 participants aged 5 to 17 yr and used tablet audiometry. The initial screening, which included otoscopy and audiogram tests, revealed 44 individuals (10.4%) who did not meet the screening criteria. A subsequent evaluation with a semiautomated tablet audiometer found that 5.9% of individuals had suspected HL. These studies, conducted in Cuba, Haiti, and the Dominican Republic, demonstrate how strategically deploying local resources and technology within Caribbean communities can effectively facilitate early detection of HL in geographically isolated areas, reducing the need for individuals to travel to urban centers for audiological evaluations.[27]

Environmental factors contributing to HL

Various studies have shown that cultural and environmental influences impact HL. Unique factors inherent in each ethnicity can influence the prevalence of HL in a community. A study conducted in Trinidad and Tobago, known for hosting the lively Caribbean Carnival festival, investigated the realm of steelpan musicians. The steelpan originated in Trinidad, emerged from reused oil drums, and became widely recognized as the only truly new musical instrument of the 20th century. Steel bands spend several months extensively rehearsing for Carnival, with practice sessions lasting 6 to 8 h daily. These sessions expose musicians to high sound intensity levels, typically produced by steelpan orchestras. The ambient noise within the steel bands can reach levels exceeding 100 dB, similar to the intensity experienced near the center stage of a rock concert.

The study investigates the impact of prolonged exposure to high-intensity sound levels on the hearing of steelpan players. A controlled cross-sectional pilot study was conducted with 29 steelpan players and 30 control participants. Steelpan players had a significantly higher prevalence of HL than the control group, particularly at frequencies of 3000 Hz, 4000 Hz, and 6000 Hz ($p < 0.01$). Moreover, a correlation was observed between the duration of playing the instrument and the severity of HL, indicating that prolonged exposure resulted in greater auditory damage. Long-term exposure to high-intensity sound levels during steel band rehearsals and performances significantly contributes to HL among steelpan musicians in Trinidad.[40]

Furthermore, a study in Cuba investigated an epidemic that occurred between 1992 and 1993, which included SNHL. In this outbreak, the affected individuals were often young adults with severe SNHL. Pure tone audiometry revealed bilateral and symmetrical high frequency (4-8 kHz) HL in patients. During the epidemic, many Cuban patients had abnormal brainstem auditory evoked responses, including those with abnormal audiometry results.

Potential causative factors were investigated, and a micronutrient deficiency was identified as the underlying cause of HL. The shortage of basic food supplies caused by economic problems and the economic embargo imposed on Cuba exacerbated the effects of dietary restrictions, resulting in B-vitamin deficiencies. In the absence of malnutrition, a lack of essential micronutrients is identified as a primary factor in the epidemic. Thiamine, cobalamin, folate, sulfur, and amino acid deficiencies were found as contributing factors due to dietary restrictions imposed in Cuba during the epidemic.[41] These findings underscore the role of cultural and environmental factors in determining hearing health among diverse ethnicities.

In conclusion, these themes collectively depict a complex and multifaceted image of HL in the Caribbean, highlighting various genetic and environmental factors, the importance of localized technological solutions, and the distinct genetic characteristics of isolated populations. This comprehensive understanding is crucial in developing targeted interventions and future research areas.

The Caribbean has a complex genetic tapestry formed by diverse ethnic heritages. Countries like Cuba, with its blend of European, Indigenous, and African ancestry, and Dominica, with its solid African origins and Kalinago people, highlight the region's genetic diversity. The findings from this review are important for improving genetic and genomic medicine applications, particularly for hereditary conditions such as HL.

Significantly, the frequent migration from the Caribbean to the United States increases the possibility of genetic drift, which might lead to rare *HL* gene variants in the American population. By learning more about these genes, we can improve the diagnosis and early treatment of genetic HL in the diverse population of the United States, eventually improving outcomes and understanding in this critical area of healthcare.

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Conflict of Interest: The authors declare that they have no conflict of interest.

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