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Cognitive and Behavioural Associations of Visual and Hearing Impairments Across the Lifespan in People With Down Syndrome, a Scoping Review

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ABSTRACT

Background: Hearing and visual impairments are common in individuals with Down syndrome (DS), yet it remains unclear whether sensory impairments are associated with cognitive and behavioural functioning such as language, memory and socialisation. This literature review examines those associations.

Methods: A scoping review of four databases was conducted to examine associations between sensory impairments and cognitive or behavioural outcomes in individuals with DS. Study evidence was assessed based on design, sample size, assessment modalities and statistical significance.

Results: Of 1471 studies screened, 38 met inclusion criteria. Hearing impairments were associated with delays in language and motor development during infancy, lower overall cognitive ability and social communication difficulties in childhood and reduced adaptive functioning and intellectual decline in adulthood. Visual impairments were linked to learning difficulties in childhood and to declines in adaptive behaviour, visual-motor integration and cognitive performance in adulthood. Subdomains of cognitive functioning such as memory, reasoning or processing speed were not reported for adults.

Conclusion: Uncorrected hearing and visual impairments in individuals with DS are associated with a range of cognitive and behavioural outcomes across the lifespan. The strongest associations were observed between hearing impairments and language delays in infancy and childhood, and between visual impairments and adaptive behaviour challenges in childhood and adulthood. Sensory impairments were associated with challenges in similar domains—particularly language, adaptive functioning and learning—across the lifespan. This suggests a potential continuity of these associations over time and that long-standing uncorrected impairments may be associated with reduced cognitive reserve and increased vulnerability to decline—highlighting the need for early identification and intervention.

1 | Introduction

Down syndrome (DS) is caused by a trisomy of the 21st chromosome, and the prevalence is estimated to be one in 1499 people in

the United States, with worldwide prevalences varying by country depending in part on the frequency of elective terminations (de Graaf et al. 2017, 2022). Individuals with DS can present with a variety of co-occurring conditions, defined as the ‘concurrence

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of multiple diseases in one person' (de Groot et al. 2003). Hearing and visual impairments are two co-occurring conditions that are common in this population, with both having a high lifetime prevalence of up to 85% (Krinsky-McHale et al. 2014; Santoro et al. 2021). Some studies have suggested associations between uncorrected sensory impairments in the DS population and specific cognitive (e.g., intellectual functioning, language and reading skills) and behavioural (e.g., social engagement, activities of daily living and motor skills) domains (Bull et al. 2022; Hendrix et al. 2021; Krinsky-McHale et al. 2014; Laws and Hall 2014; Santoro et al. 2021; Startin et al. 2020).

Sixty percent of children with DS are reported to have visual impairments (Bull et al. 2022). This prevalence increases to 85% in adults with DS above the age of 60, compared to just 20% in age-matched individuals with non-DS intellectual disabilities (Krinsky-McHale et al. 2014; Startin et al. 2020). Visual impairments in adulthood have been shown to be associated with reduced opportunities for independence along with challenges with communication and social engagement (Krinsky-McHale et al. 2014). Specific types of visual impairments in adults with DS include reduced sensitivity across spatial frequencies, reduced stereopsis and challenges with colour discrimination (Krinsky-McHale et al. 2012). Spatial frequencies are important for the recognition of objects and faces (Lacroix et al. 2022); difficulties in stereopsis have been associated with falls (Mehta et al. 2022), and colour discrimination is important for activities of daily living such as matching clothes (Mary et al. 2016).

Between 47% and 85% of people with DS are reported to have some form of hearing impairment (Santoro et al. 2021). Children with DS often have conductive hearing loss (e.g., otitis media), while 53% of adults have sensorineural hearing loss, which is in part due to the increased prevalence of presbycusis (Chen et al. 2021). It is estimated that age-related hearing loss occurs 30 years earlier in people with DS than in the general population (Meuwese-Jongejeugd et al. 2006). Potential reasons for early onset hearing loss include childhood pathologies, such as otitis media, that have lifelong effects on hearing, or other co-occurring conditions that may impact hearing, such as dementia (Nittrover and Lowenstein 2024; McCarron et al. 2005). Associated symptoms for adults with DS and hearing loss have included dizziness, vertigo and balance issues as well as language problems (Hendrix et al. 2021; Laws and Hall 2014).

Despite the myriad of studies examining sensory impairments in individuals with DS, many examined only one type of sensory impairment during a specific life stage, and few investigated the associations between uncorrected sensory impairments with cognition and behaviour. This review aims to bridge this gap by compiling and analysing findings from existing studies on how uncorrected visual and hearing impairments relate to cognitive and behavioural functions in individuals with DS at different life stages. Additionally, this review seeks to assess the certainty of evidence for these associations in the current literature and highlight areas that require further investigation. The aim is to provide a comprehensive resource that can guide future studies and inform best practices for clinical management and intervention strategies.

2 | Methods

This scoping review was conducted following the Preferred Reporting Items for Systematic Reviews, Meta-Analysis and Scoping Review (PRISMA-ScR) guidelines based on the Joanna Briggs Institute (JBI) Manual for Evidence Synthesis (Peters et al. 2020). Four electronic databases were searched (PubMed, Scopus, Cochrane CENTRAL and Web of Science) using a comprehensive search strategy with no date or language restrictions.

The inclusion criteria were: (1) target population of individuals with DS; (2) mentioned uncorrected hearing and/or visual impairments and (3) association studied between hearing or visual impairments and a cognitive or behavioural change, delay or decline. In this review, 'uncorrected' refers to sensory impairments identified at the time of assessment, regardless of whether they had been previously treated or addressed since that information may not always be available. Meta-analyses, letters, editorials, comments and book chapters were excluded. The full search terms and strategies can be seen in Table 1. The protocol of this scoping review is based on the protocol registered by Betoni et al. (2023) on Open Science Framework and can be viewed at https://osf.io/rqszw/?view_only=31e1e324a8ef40299306e9bca50b326a.

Screening was conducted using Covidence in two phases by two independent reviewers (N.B. and C.O.), with conflicts resolved by a third reviewer (I.L.). Phase 1 involved screening titles and abstracts to identify potentially relevant articles. Phase 2 consisted of full-text reviews to confirm final study inclusion. If articles were written in another language, they were translated to English using an online neural machine translation website. Following study selection, two reviewers (N.B. and C.O.) extracted pertinent data on study characteristics, sensory impairments assessed, cognitive or behavioural outcomes examined and key findings.

The primary outcomes were any associations found between hearing or visual impairments and areas of cognition (e.g., attention, language, memory and overall cognition) or behaviour (e.g., activities of daily living, social engagement and motor skills). Given the scoping nature of this review, no additional statistical analyses were performed beyond those presented in the original included studies.

A modified Grading of Recommendations, Assessment, Development and Evaluation (GRADE) approach was applied to assess the certainty of evidence included in this scoping review (Guyatt et al. 2008). 'Certainty' refers to the degree of confidence that the reported associations reflect true relationships between sensory impairments and cognitive or behavioural outcomes, based on study design, sample size, statistical significance, effect size and the quality of assessment tools used. Study designs were categorised according to the Joanna Briggs Institute (JBI) Levels of Evidence and Grades of Recommendation (2014), where randomised controlled trials (JBI Level 1) were ranked highest. As seen in Table 2, the modified GRADE criteria defined 'High' certainty studies as randomised controlled trials (JBI Level 1) with strong statistical significance ($p < 0.01$) and well-designed or standardised

TABLE 1 | Electronic database search strategies.

PUBMED	Search terms	Results
#1	"Down Syndrome"[mh] OR "Down Syndrome" OR Mongolism OR "47,XY,+21" OR "Trisomy G" OR "47,XX,+21" OR "Down's Syndrome" OR "Trisomy 21"	45634
#2	"Vision Disorders"[mh] OR "Vision disorder*" OR "Visual disorder*" OR "Macropsia*" OR "Macropsia*" OR "Visual Deficit*" OR "Vision impairment*" OR "impaired vision" OR "Visual impairment*" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR Hemeralopia* OR "Day Blindness" OR Metamorphopsia* OR Deaf-Blind Disorders	108104
#3	"Hearing disorders"[mh] OR "Hearing disorder*" OR "Hearing loss"[mh] OR "hearing loss*" OR Deafness* OR Deaf OR Hypoacusis* OR "Hearing impaired" OR "Hearing impairment*" OR Dysacusis OR Paracusis OR "Distorted Hearing" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit"	141592
#4	#1 AND #2	293
#5	#1 AND #3	606
#6	#4 OR #5	822
#7	((("Down Syndrome"[mh] OR "Down Syndrome" OR Mongolism OR "47,XY,+21" OR "Trisomy G" OR "47,XX,+21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND ("Vision Disorders"[mh] OR "Vision disorder*" OR "Visual disorder*" OR "Macropsia*" OR "Macropsia*" OR "Visual Deficit*" OR "Vision impairment*" OR "impaired vision" OR "Visual impairment*" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR Hemeralopia* OR "Day Blindness" OR Metamorphopsia* OR Deaf-Blind Disorders)) OR ("Down Syndrome"[mh] OR "Down Syndrome" OR Mongolism OR "47,XY,+21" OR "Trisomy G" OR "47,XX,+21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND ("Hearing disorders"[mh] OR "Hearing disorder*" OR "Hearing loss"[mh] OR "hearing loss*" OR Deafness* OR Deaf OR Hypoacusis* OR "Hearing impaired" OR "Hearing impairment*" OR Dysacusis OR Paracusis OR "Distorted Hearing" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit")))) NOT (Letter[pt] OR editorial[pt] OR Comment[pt]) Sort by: Most Recent	812
SCOPUS	Search terms	Results
#1	((TITLE-ABS-KEY ("Down Syndrome" OR mongolism OR "47,XY, 21" OR "TrisomyG" OR "47,XX, 21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND TITLE-ABS-KEY ("Vision Disorders" [mh] OR "Vision disorder*" OR "Visualdisorder*" OR macropsia* OR micropsia* OR "Visual Deficit*" OR "Visionimpairment*" OR "impaired vision" OR "Visual impairment*" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visualabnormality*" OR hemeralopia* OR "Day Blindness" OR metamorphopsia* OR "Deaf-Blind Disorder*")) OR ((TITLE-ABS-KEY ("Down Syndrome" OR mongolism OR "47,XY, 21" OR "TrisomyG" OR "47,XX, 21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND TITLE-ABS-KEY ("Hearing disorder*" OR "Hearing loss" OR "hearingloss*" OR deafness* OR deaf OR hypoacusis* OR "Hearing impaired" OR "Hearingimpairment*" OR dysacusis OR paracusis OR "Distorted Hearing" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit*"))))	1003

(Continues)

TABLE 1 | (Continued)

SCOPUS		Search terms	Results
#2		((TITLE-ABS-KEY ("Down Syndrome" OR mongolism OR "47,XY, 21" OR "Trisomy G" OR "47,XX, 21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND TITLE-ABS-KEY ("Vision Disorders" [mh] OR "Vision disorder*" OR "Visual disorder*" OR "Visual impairment*" OR "Visual deficit*" OR "Vision impairment*" OR "impaired vision" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR hemeralopia* OR "Day Blindness" OR metamorphopsia* OR "Deaf-Blind Disorder*")) OR ((TITLE-ABS-KEY ("Down Syndrome" OR mongolism OR "47,XY, 21" OR "TrisomyG" OR "47,XX, 21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND TITLE-ABS-KEY ("Hearing disorder*" OR "Hearing loss" OR "hearingloss*" OR deafness* OR deaf OR hypoacus* OR "Hearing impaired" OR "Hearing impairment*" OR dysacusis OR paracusis OR "Distorted Hearing" OR "Auditory impairment*" OR "Auditory difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit*")) AND (EXCLUDE (DOCTYPE, "ch") OR EXCLUDE (DOCTYPE, "ed") OR EXCLUDE (DOCTYPE, "le") OR EXCLUDE (DOCTYPE, "no") OR EXCLUDE (DOCTYPE, "bk"))))	926
WEB OF SCIENCE		Search terms	Results
#1		("Down Syndrome" OR Mongolism OR "47,XY,+21" OR "Trisomy G" OR "47,XX,+21" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") AND ("Vision disorder*" OR "Visual disorder*" OR Macropsia* OR Micropsia* OR "Visual Deficit*" OR "Vision impairment*" OR "impaired vision" OR "Visual impairment*" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR Hemeralopia* OR "Day Blindness" OR Metamorphopsia* OR "Deaf-Blind Disorders" OR "Hearing disorder*" OR "hearing loss*" OR Deafness* OR Deaf OR Hypoacus* OR "Hearing impaired" OR "Hearing impairment*" OR Dysacusis OR Paracusis OR Paracusis OR "Distorted Hearing" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit*") EXCLUDE: Letter, Editorial, Book Chapter	482
CENTRAL		Search terms	Results
#1		MeSH descriptor: [Down Syndrome] explode all trees	509
#2		MeSH descriptor: [Hearing Disorders] explode all trees	2613
#3		MeSH descriptor: [Vision Disorders] explode all trees	2205
#4		("Down Syndrome" OR Mongolism OR "Trisomy G" OR "Down's Syndrome" OR "Downs Syndrome" OR "Trisomy 21") OR #1	1130
#5		("Vision disorder*" OR "Visual disorder*" OR Macropsia* OR Micropsia* OR "Visual Deficit*" OR "Vision impairment*" OR "impaired vision" OR "Visual impairment*" OR "Visual loss" OR "Visual disturbance*" OR "Ocular impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR Hemeralopia* OR "Day Blindness" OR Metamorphopsia* OR "Deaf-Blind Disorders" OR "Hearing disorder*" OR "hearing loss*" OR Deafness* OR Deaf OR Hypoacus* OR "Hearing impaired" OR "Hearing impairment*" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit*") OR "impaired vision" OR "Vision impairment*" OR "Vision Disability" OR "Visual disability*" OR "Visual challenge*" OR "Vision problem*" OR "Visual problem*" OR "Visual abnormality*" OR Hemeralopia* OR "Day Blindness" OR Metamorphopsia* OR "Deaf-Blind Disorders" OR "Hearing disorder*" OR "hearing loss*" OR Deafness* OR Deaf OR Hypoacus* OR "Hearing impaired" OR "Hearing impairment*" OR "Auditory impairment*" OR "Hearing difficult*" OR "Hearing challenge*" OR "Hearing problem*" OR "Hearing disability*" OR "Auditory deficit*" OR "Hearing Deficit*") OR #2 OR #3	12004
#6		#4 AND #5	21

Note: Electronic database search strategies across four databases yielded a total of 2241 records—Scopus (n = 812), Web of Science (n = 482) and CENTRAL (n = 21).

TABLE 2 | Certainty of evidence criteria using modified GRADE framework.

GRADE category	Determining criteria
High certainty (5)	- Randomised controlled trials (JBI Level 1), well-designed standardised testing used to assess cognition and behavioural outcomes, and strong statistical significance ($p < 0.01$)
Higher certainty (4)	- Quasi-experimental studies (JBI Level 2), well-designed standardised testing used to assess cognition and behavioural outcomes, with $p < 0.01$ or $d > 0.8$ if association is made OR - Observational studies (JBI Level 3) with large sample size (≥ 100 participants), well-designed standardised testing used to assess cognition and behavioural outcomes, and strong statistical significance ($p < 0.01$) or effect size ($d > 0.8$ or $\eta^2 > 0.14$) if association is made
Moderate certainty (3)	- Observational studies (JBI Level 3) with medium sample size (20–99 participants), well-designed standardised testing used to assess cognition and behavioural outcomes, $p < 0.05$ or effect size of $d > 0.5$ or $\eta^2 > 0.06$ if association is made OR - Observational studies (JBI Level 3) small size (< 20), well-designed standardised testing used to assess cognition and behavioural outcomes, and strong statistical significance ($p < 0.01$) or effect size ($d > 0.8$ or $\eta^2 > 0.14$) if association is made OR - Quasi-experimental studies (JBI Level 2), well-designed standardised testing used to assess cognition and behavioural outcomes, with $p < 0.05$ or effect size of $d > 0.5$ or $\eta^2 > 0.06$ if association is made

(Continues)

TABLE 2 | (Continued)

GRADE category	Determining criteria
Low certainty (2)	- Observational studies (JBI Level 3) with poorly designed measures used to assess cognition and behaviour, and/or weaker statistical significance ($p \geq 0.05$) or effect size ($d < 0.5$ or $\eta^2 < 0.06$) when association is made OR - Case series/case reports (JBI Level 4) with moderate sample size (≥ 20 participants) with poorly designed measures used to assess cognition and behaviour
Very low certainty (1)	- Case series/case reports (JBI Level 4) with small sample size (< 20 participants) with poorly designed measures used to assess cognition and behaviour OR - Expert opinion/theoretical papers (JBI Level 5)

Note: Certainty of evidence criteria using modified GRADE framework for each study that met inclusion criteria for the scoping review. This framework categorises studies into five certainty levels (high, higher, moderate, low and very low) based on study design, sample size, strength of assessments, statistical significance (p -values) and effect size (Cohen's d or η^2). Abbreviations: GRADE—Grading of Recommendations Assessment, Development, and Evaluation; JBI—Joanna Briggs Institute level of evidence.

cognitive and behavioural assessments used. ‘Very Low’ certainty evidence included small-sample case reports (< 20 participants) with poorly designed cognitive and behavioural assessments, as well as expert opinion or theoretical papers (JBI Level 5). By incorporating both study design (using JBI Levels of Evidence) and study strength indicators such as statistical significance, effect size and evaluation of tools used for assessment, the modified approach allowed for a more refined appraisal that recognises meaningful contributions from lower-tier evidence while still highlighting higher-quality findings.

3 | Results

3.1 | Overview

This scoping review initially identified 2241 studies from databases including Scopus (926), PubMed (812), Web of Science (482) and CENTRAL (21). After the removal of duplicates manually (43) and through Covidence (727), 1471 studies were screened. Of these, 1374 were excluded during the abstract and title screening. Subsequently, 97 full-text studies were assessed for eligibility and 59 were excluded for the following reasons: 28 studies did not test an association between hearing or visual

impairments and cognitive or behavioural outcomes; 17 studies did not focus on individuals with DS or failed to disaggregate results specific to this population; 7 were review articles, 6 were expert opinion pieces and 1 was a book chapter, all of which did not include original data or statistical analysis relevant to the review question (Figure 1).

Within the included 38 studies, 23 examined hearing impairments, 7 examined visual impairments and 8 examined both hearing and visual impairments in separate analyses (Figure 1). Studies utilised various designs such as cross-sectional (23), cohort (6), nonrandomised experimental (4), qualitative research (2), case reports (2) and case-control (1). A substantial portion of the included studies (27 of 38; 71%)

were cross-sectional, case report or qualitative in nature, assessing participants' current sensory, cognitive or behavioural status without a clear timeline of impairment onset or prior intervention history. Thirteen studies included a control or comparison group, most commonly comparing individuals with DS to typically developing peers, individuals with other developmental disabilities or matched DS subgroups based on hearing or vision status. Twelve studies used some form of matching—typically for age, IQ or language ability—while Pappas et al. (1994) did not report any matching procedures or adjustments for potential confounders. Only two studies, Chapman et al. (2006) and Laws and Hall (2014), explicitly excluded participants with autism or other neurodevelopmental conditions.

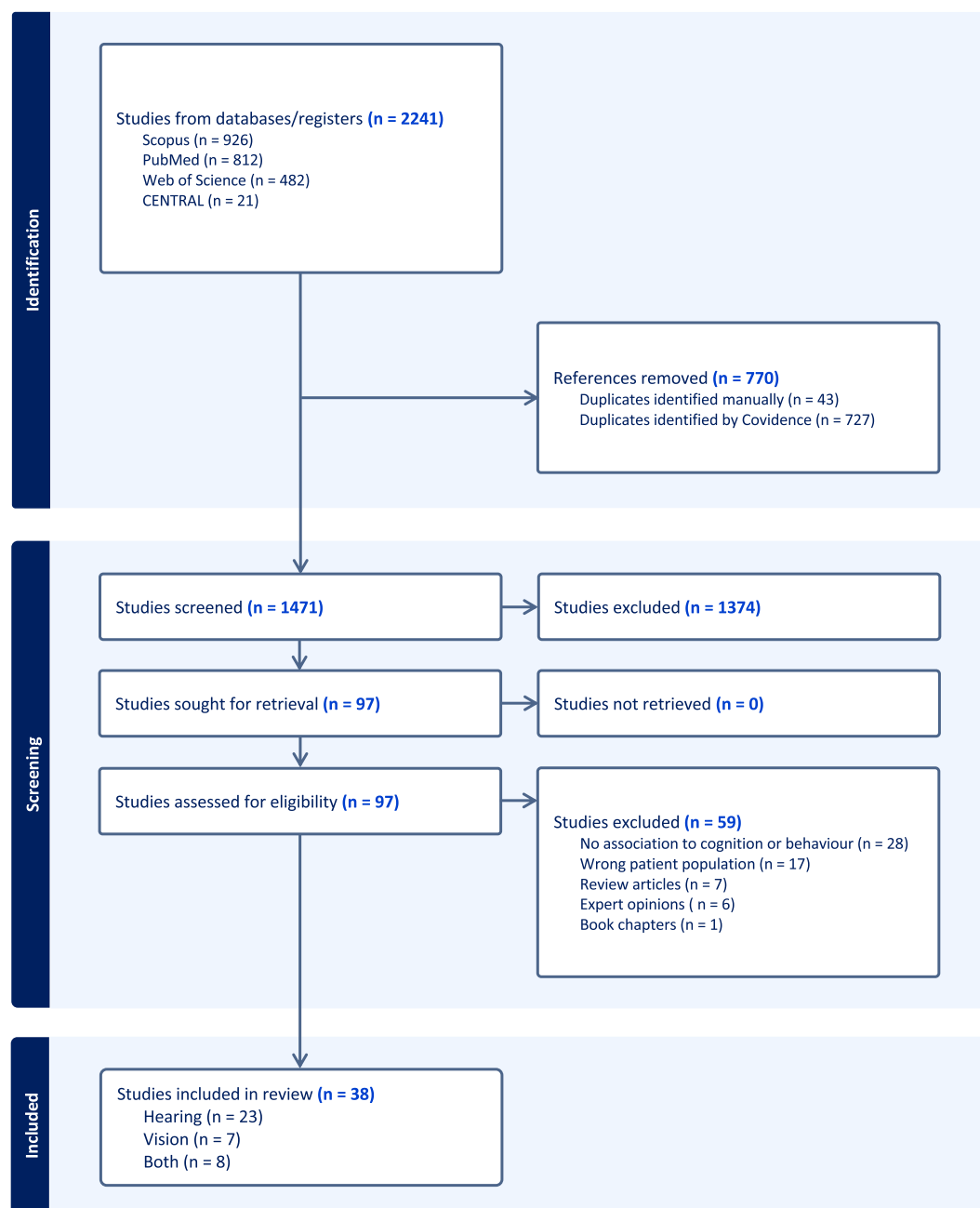


FIGURE 1 | PRISMA flow diagram illustrating the study selection process. The diagram shows the number of records identified from four databases (Scopus, PubMed, Web of Science and CENTRAL), duplicates removed, records screened, full-text articles assessed for eligibility and studies included in the final review. A total of 38 studies were included: 23 related to hearing, 7 to vision and 8 to both domains.

The studies were published between 1985 and 2023. Of the 38 included studies, over half (57.9%) were published more than 15 years ago, prior to 2010. The median year of publication was 2005. The average age parameters for the studies ranged from less than 1 year of age to 54 years of age. The total number of participants across all studies and control groups was 3073. The geographic location of these studies spanned multiple countries including the United Kingdom (11), United States (10), Canada (3), Italy (3), Netherlands (3), Japan (2), Australia (1), Bangladesh (1), Colombia (1), Finland (1), Israel (1) and the United Arab Emirates (1).

3.2 | Certainty of Evidence for Hearing Impairment Studies

Using the modified GRADE framework, the studies exploring associations between hearing impairments and cognitive or behavioural outcomes in individuals with DS ranged from 'Very Low' to 'Moderate' certainty and received an average score of 2.4 on a scale from 1 ('Very Low' certainty) to 5 ('High' certainty), with the strongest associations being related to the language challenges in infancy and childhood (Table 3). For instance, the Chapman et al. (1991) study, rated as 'Moderate' certainty, found that greater degrees of hearing loss were significantly associated with reduced expressive language abilities in childhood ($p < 0.01$). Similarly, Alzyoudi et al.'s study (2022), also rated 'Moderate', found that even mild conductive hearing loss was correlated with lower language comprehension scores ($p < 0.05$) in childhood, reinforcing the established link between hearing impairments and language development. In contrast, studies exploring hearing and cognitive outcomes showed more variability. The Chapman et al. (2006) study, rated 'Low' certainty, reported associations between mild hearing loss and reduced nonverbal communication, but limitations in study design and sample size reduced confidence in these findings. The weakest evidence emerged in the behavioural domain. The Downs et al. (2013) study, rated 'Very Low' certainty, qualitatively identified ear problems such as otitis media as barriers to social communication but lacked quantitative data or statistical rigour.

3.3 | Uncorrected Hearing Impairment Findings Across the Lifespan

There were consistent findings of problems associated with uncorrected hearing impairment across the lifespan (Figure 2). In infancy, hearing impairments are associated with greater delays in the acquisition of motor milestones and increased emotional challenges (Harigai 1994), as well as with greater delays in expressive and receptive language skills (Pappas et al. 1994) compared to DS infants without hearing impairments. In childhood, hearing impairments are associated with impairments in overall intellectual functioning (Libb et al. 1985; Namatame et al. 2019) and balance (Downs et al. 2013), social (Porter et al. 2023; Yunker et al. 2009) and language challenges (Alzyoudi et al. 2022; Chapman et al. 2006; Groen et al. 2008; Laws 2004; Laws and Hall 2014; Saksida et al. 2021; Tieri et al. 1996). In adulthood, hearing impairments are associated with poorer motor functioning (Jeffery and Whiteside 2020), greater language and communication difficulties (Marcell and Cohen 1992; Marcell et al. 1995; Määttä et al. 2006),

barriers to employment opportunities (Pikora et al. 2014) and intellectual deterioration (Hewitt et al. 1985) compared to adults with DS without hearing loss.

3.4 | Certainty of Evidence for Visual Impairment Studies

The studies exploring associations between visual impairments and cognitive or behavioural outcomes in individuals with DS were generally rated as 'Low' to 'Moderate' certainty and received an average score of 2.7 on a scale from 1 ('Very Low' certainty) to 5 ('High' certainty), with the strongest associations being related to the adaptive behaviour in childhood and adulthood (Table 4). One study, de Weger et al. (2021), received a 'Higher' certainty rating and reported statistically significant associations between visual acuity and adaptive functioning in childhood ($p < 0.01$). Other studies, such as those by Krinsky-McHale et al. (2012) and Hewitt et al. (1985), also found statistically significant associations between ophthalmic disorders and greater adaptive functioning ($p < 0.05$) and intellectual functioning challenges ($p < 0.01$) in adulthood. However, these studies were rated 'Moderate' in certainty due to less statistical significance or smaller sample sizes. Määttä et al.'s study (2006) also found a relationship between visual impairments and difficulties with verbal communication in adulthood, but their conclusion was rated 'Low' in certainty due to limitations in study design and the assessments used to evaluate language and cognition.

3.5 | Uncorrected Visual Impairment Findings Across the Lifespan

There were consistent findings of outcomes related to uncorrected visual impairments in childhood and adulthood, but no studies examined these relationships in infancy (0 to 2 years of age) (Figure 3). In childhood, children with DS who have visual impairments have greater delays in literacy skills (Nandakumar and Leat 2010; Nandakumar et al. 2011) and learning differences (Merrick and Koslowe 2001), along with increased challenges in adaptive behaviours and physical navigation (de Weger et al. 2021; Wilton et al. 2021) compared to children with DS without visual impairments. In adulthood, those with DS who have visual impairments have more communication-related challenges (Määttä et al. 2006), decreased overall cognitive performance (Hewitt et al. 1985) and greater challenges with adaptive behaviours, including daily living skills (Krinsky-McHale et al. 2012; Roeden and Zitman 1995, 1997) compared to those with DS without visual impairments.

3.6 | No or Mixed Associations

Eight studies did not find an association between hearing or visual impairments and any cognitive or behavioural outcomes (Dressler et al. 2015; Islam et al. 2022; Jarrold and Baddeley 1997; Jarrold et al. 2002; Pradilla et al. 2020; Roizen et al. 1993; Schworer et al. 2022; Turner et al. 1990) (Table 5). Additionally, three studies revealed mixed results. Marcell and Cohen (1992) found individuals with DS who had poor hearing tended to perceive and process acoustic signals more slowly

TABLE 3 | Summary of studies showing uncorrected hearing impairments associated with cognition or behaviour in individuals with Down syndrome.

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean	n	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
				age (years)							
(Alzyoudi et al. 2022)	Phonological process in Arabic-speaking children with Down syndrome: A psycholinguistic investigation	EAB	CS	9–12	30	Age matched generally developing children; <i>n</i> = 30	Tympanometry, acoustic reflexes, impedance audiometry and previous visit records	The Arabic standardised articulation test	Mild conductive hearing loss is associated with more errors in articulation (<i>p</i> < 0.02)	3	Moderate
(Chapman et al. 1991)	Language skills of children and adolescents with Down syndrome: I. comprehension	US	CS	12.54	48	Mental age matched; normally developing children; <i>n</i> = 48	Audiometry	PPVT-R	Degree of hearing loss is correlated with variations in comprehension skills (<i>p</i> < 0.01)	3	Moderate
(Chapman et al. 2006)	Effect of memory support and elicited production on fast mapping of new words by adolescents with Down syndrome	US	CS	15.94	19	Non-DS syntax comprehension matched group; <i>n</i> = 18	Hearing screening by licensed audiologist	TACL-3 and PPVT-III	Mild hearing loss is associated with reduced number of new words being learned (<i>p</i> < 0.05)	3	Low
(Downs et al. 2013)	Exploring opportunities available and perceived barriers to physical activity engagement in children and young people with Down syndrome	UK	Q	16.9	8	None	Parent reporting hearing issues	The Youth Physical Activity Promotion model during semistructured interviews	Ear problems, like otitis media, are barriers to physical activity and could be associated with balance issues	4	Very low

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean age (years)	n	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
(Groen et al. 2008)	Weak hand preference in children with Down syndrome is associated with language deficits	UK	CS	9.92	29	Age matched; normal development; n = 43	Audiometry	GFTA and TROG-2	Hearing deficits are associated with worse articulation and expressive grammatical ability (<i>p</i> < 0.043)	3	Moderate
(Harigai 1994)	Longitudinal studies in hearing-impaired children with Down's syndrome	JAP	C	0.25–15	110	None	Tympanometry, auditory brainstem response and behavioural audiometry	Tsumori-Image Infant Developmental Questionnaire and parent questionnaires on hearing and language development	Hearing delays are associated with delays in walking steadily, emotional disturbances and language delays	3	Low
(Hewitt et al. 1985)	Ageing in Down's syndrome	UK	C	54.3	23	None	Serial recordings of hearing, not clarified further	SBIS	Hearing loss is associated with significant intellectual deterioration (lower mental age) (<i>p</i> < 0.001)	3	Moderate

(Continues)

TABLE 3 | (Continued)

Study info		Demographics			Methods and results			Evidence ratings			
Author(s)/year	Title	Country	Study	Mean age (years)	n	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
(Jeffery and Whiteside 2020)	Templates and temporality: An investigation of rhythmic motor production in a young man with Down syndrome and hearing impairment	UK	CR	23.8	1	None	Audiogram	Gross motor movements assessed visually against a checklist developed by first author	Hearing issues partly contribute to gross motor movement difficulties	4	Very low
(Laws and Hall 2014)	Early hearing loss and language abilities in children with Down syndrome	UK	CC	6.19	16	DS diagnosis group; normal hearing; mean age 6.62; n = 25	Retrospective audiology clinic records	Leiter-R, BPVS-2, RDLS, GFTA-2 and MLU-W	Hearing loss is significantly associated with challenges with comprehension, expression, speech accuracy and receptive vocabulary (p<0.002–0.038)	3	Moderate

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean age (years)		Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
				n							
(Laws 2004)	Contributions of phonological memory, language comprehension and hearing to the expressive language of adolescents and young adults with Down syndrome	UK	CS	16.5	23	None	Audiometry	MLU, CELF-R, CNRep and TROG	Hearing loss is significantly associated with lower language comprehension ($p < 0.001$, $\eta^2 = 0.311$), expressive language ($p = 0.009$, $\eta^2 = 0.172$), vocabulary ($p = 0.004$, $\eta^2 = 0.202$) and speech accuracy ($p = 0.014$, $\eta^2 = 0.196$).	3	Moderate
(Libb et al. 1985)	Hearing disorder and cognitive function of individuals with Down syndrome	US	CS	14.5	28	None	Audiometry and tympanograms	WISC-R, WAIS-R and SBIS	Abnormal tympanograms are significantly associated with lower IQ ($p < 0.01$)	3	Moderate
(Määttä et al. 2006)	Sensory impairments and health concerns related to the degree of intellectual disability in people with Down syndrome	FIN	CS	M: 29; F: 35	129	None	Audiometry	IQ-based classification of intellectual disability (mild-profound); speech ability categorised from medical records	Hearing loss is significantly associated with the severity of intellectual functioning and speech impairment ($p < 0.05$)	3	Low

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean age (years)	n	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
(Marcell and Cohen 1992)	Hearing abilities of Down syndrome and other mentally handicapped adolescents	US	CS	18.8	26	Mentally handicapped non-DS; matched IQ and chronological age; <i>n</i> = 26	Audiometry and tympanograms	TOLD-2P, Miller-Yoder Comprehension Test, backward masking, auditory digit span and gating task	Hearing loss is correlated with poorer word recognition in noise (<i>p</i> < 0.05), though hearing difficulty did not predict performance in complex cognitive tasks (such as sentence imitation and sentence comprehension)	3	Moderate
(Marcell et al. 1995)	Sentence imitation by adolescents and young adults with Down's syndrome and other intellectual disabilities	US	C	18.8	26	IQ-matched individuals without DS with other intellectual disabilities; <i>n</i> = 26	Audiometry, acoustic reflex testing and tympanograms	TOLD-2P, Miller-Yoder Comprehension Test, backward masking and auditory digit span	Middle ear dysfunction, such as that caused by recurrent otitis media, is associated with sentence imitation challenges (<i>p</i> < 0.05)	3	Moderate

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
		Country	Study	Mean age (years)	n	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
Author(s)/year	Title										
(Namatame et al. 2019)	Psychomotor development and rehabilitation in Down syndrome: investigations with complications	JAP	CS	9.8	58	None	Retrieved from medical records	Enjoji Infant Analysis, Tsumori Infant Mental Development Test, K-Test, Development Test, Tanaka Binet, Suzuki Binet, WISC-III and WISC-IV	Hearing loss is associated with lower IQ values and a significant delay in sitting alone ($p = 0.015$), lying on stomach ($p = 0.024$), and standing up ($p = 0.037$)	3	Moderate
(Pappas et al. 1994)	Otological and rehabilitative management of children with Down syndrome	US	CS	0.92	6	Individuals with DS with no referral or treatment; mean age 1.74; $n = 6$	Otoscopy, tympanometry and audiometry	Standardised speech-language assessments (unspecified)	Children with otitis media with effusion exhibit delays in expressive language and receptive understanding	3	Low
(Pikora et al. 2014)	Health conditions and their impact among adolescents and young adults with Down syndrome	AUS	CS	23.6	197	None	Parent-reported hearing impairments	Index of Social Competence, parental reports of mental health	Hearing impairments are associated with increased safety concerns and restrict opportunities to participate in employment and community leisure activities	3	Low

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean age		Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE
				(years)	n						
(Porter et al. 2023)	'I think it impacts all areas of his life': perspectives on hearing from mothers of individuals with Down syndrome.	US	Q	13.9	16	None	Parent-reported hearing impairments	Parent-reported observations through semistructured interviews	Hearing difficulties are associated with challenges with speech, social situations, academics and interpersonal relationships	4	Very low
(Prasher 1995)	Screening of hearing impairment and associated effects on adaptive behaviour in adults with Down syndrome	UK	CS	42.2	201	None	Audiometry and tympanometry	ABS	Hearing impairments are associated with greater challenges with independent adaptive behaviour	3	Moderate
(Saksida et al. 2021)	The influence of hearing impairment on mental age in Down syndrome: preliminary results	IT	C	5.84	17	None	Audiometry	BALC	Hearing impairments may crucially be associated with auditory and language abilities (d = 1.01, p = 0.069)	3	Moderate
(Tieri et al. 1996)	Hearing disorders in Down's syndrome	IT	NRE	6-14	15	DS diagnosis group; normal hearing; age 7-17; n = 15	Audiometry and tympanogram	Brunet-Lézine and Leiter psychomotor development tests	Children with DS and hearing loss have more language delays than children with DS and normal hearing	2	Low

(Continues)

TABLE 3 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/year	Title	Country	Study	Mean age (years)	Comparison Group(s)	Hearing assessment	Cognition/behaviour assessment type	Findings	JBI	GRADE	
(Yunker et al. 2009)	Vestibular pathology presenting as behavioural problems in a child with Down syndrome: a case report.	CAN	CR	17	1	None	Audiometry	Parent-reported observations	Hearing impairment severity is associated with increased agitation in social settings	4	Very low

Note: Summary of studies showing uncorrected hearing impairments associated with cognition or behaviour in individuals with Down syndrome. This table includes study information (author(s), year, title, country and study type), participant demographics (mean age, sample size and presence of comparison groups), hearing assessment methods, cognition and behaviour assessment tools, key findings and evidence ratings. Abbreviations: Study Types: C—Cohort study, CC—Case-control study, CR—Case-report, CS—Cross-sectional study, NRE—Nonrandomized experimental study, Q—Qualitative research; Demographics: DS—Down syndrome; Cognition/Behaviour Assessments: ABS—Adaptive Behaviour Scale, AAMR ABS—American Association on Mental Retardation Adaptive Behaviour Scale, BALC—Battery for the Assessment of Language in Children, BPVS-2—British Picture Vocabulary Scale-Second Edition, CELF-R—Clinical Evaluation of Language Fundamentals—Revised, CNRep—Children's Nonword Repetition Test, GFTA—Goldman-Fristoe Test of Articulation, GFTA-2—Goldman-Fristoe Test of Articulation-Second Edition, Leiter-R—Leiter International Performance Scale-Revised, MLU—Mean Length of Utterance in Words, PPVT-III—Peabody Picture Vocabulary Test—Third Edition, PPVT-R—Peabody Picture Vocabulary Test, Revised, RDLS—Reynell Developmental Language Scales, SBIS—Stanford-Binet Intelligence Scale, TACL-3—Test for Auditory Comprehension of Language—Third Edition, TOLD-2P—Test of Language Development-2 Primary, TROG—Test for Reception of Grammar—Version 2, WAIS-R—Wechsler Adult Intelligence Scale-Revised, WISC-III—Wechsler Intelligence Scale for Children-Third Edition, WISC-IV—Wechsler Intelligence Scale for Children-Fourth Edition, WISC-R—Wechsler Intelligence Scale for Children-Revised; Evidence Ratings: GRADE—Grading of Recommendations Assessment, Development, and Evaluation, JBI—Joanna Briggs Institute level of evidence.

INFANCY (0 - 2)	CHILDHOOD (2 - 18)	ADULTHOOD (18+)
<p>Delays in expressive language and receptive understanding (Pappas et al. 1994)</p> <p>Delay in walking steadily (Harigai 1994)</p> <p>Emotional challenges (Harigai 1994)</p> <p>Significant delays in sitting alone, lying on stomach, and standing (Namatame et al. 2019)</p>	<p>Barriers to sports, physical activities, and balance issues (Downs et al. 2013*)</p> <p>Challenges with speech comprehension, expression, accuracy, and/or receptive vocabulary (Alzyoudi et al. 2023; Chapman et al. 1991; Groen et al. 2008; Laws et al. 2004; Laws & Hall 2014; Saksida et al. 2021; Tieri et al. 1996)</p> <p>Delayed word processing speed (Chapman et al. 2006)</p> <p>Impairment in overall intellectual functioning (Libb et al. 1985; Namatame et al. 2019)</p> <p>Impaired social and interpersonal relationships (Yunker et al. 2009*; Porter et al. 2023*)</p>	<p>Adaptive behaviour skills challenges (Prasher 1995)</p> <p>Barriers to employment opportunities (Pikora et al. 2014)</p> <p>Communication and language skills challenges (Määttä et al. 2006; Marcell & Cohen 1992; Marcell et al. 1995)</p> <p>Intellectual deterioration (Hewitt et al. 1985)</p> <p>Lower overall cognitive ability (Määttä et al. 2006)</p> <p>Motor function challenges (Jeffery & Whiteside 2020*)</p>

FIGURE 2 | Associations between uncorrected hearing impairment and behaviour or cognition across lifetime in individuals with Down syndrome; * = studies with ‘Very Low’ certainty of evidence as determined by the modified GRADE framework.

compared to age and IQ-matched controls; however, there was no such association found with more complex cognitive tasks such as sentence repetition. Roeden and Zitman (1995, 1997) found a statistically significant association between visual impairment and increased challenges with daily living skills; however, no such association was seen between hearing impairment and daily living skills. The eleven studies that found no or mixed associations were generally rated from ‘Low’ to ‘Moderate’ certainty with an average score of 2.8. One study, Jarrold and Baddeley (1997), was rated ‘Higher’ certainty and reported that hearing difficulties had no significant association with verbal short-term memory performance in childhood, suggesting that cognitive impairments at this age may occur independently of sensory impairments. A study by Islam et al. (2022), rated ‘Low’ certainty, also found no statistically significant association between either hearing or vision loss and behavioural outcomes in childhood or adulthood ($p=0.32$), although the study was limited by sample size and lacked detailed sensory assessment data. Similarly, studies by Dressler et al. (2015) and Jarrold et al. (2002), both rated ‘Moderate’ certainty, found that visual anomalies in adulthood and hearing difficulties in childhood were not significantly linked to impairments in daily living skills or short-term memory.

3.7 | Assessment Quality

Approximately two-thirds of the included studies used standardised, psychometrically validated assessments to measure cognitive or behavioural outcomes, which enhances confidence in the reported associations. Among these, the Stanford-Binet Intelligence Scale (SBIS) and Wechsler Intelligence Scale for Children (WISC) were used in studies such as Hewitt et al. (1985) and Namatame et al. (2019) to assess intellectual functioning. These tools are considered gold standards, as they evaluate key cognitive domains including reasoning, memory and

processing speed. Similarly, the Vineland Adaptive Behaviour Scales (VABS)—as used in studies by Dressler et al. (2015) and Roeden and Zitman (1995, 1997)—is a widely accepted instrument for assessing adaptive functioning across communication, socialisation and daily living domains in individuals with intellectual disabilities. Another frequently used assessment in studies was the Test for Reception of Grammar (TROG), which provided domain-specific assessments of language in neurodevelopmental populations (e.g., Namatame et al. 2019; Laws and Hall 2014). However, some studies, such as by Porter et al. (2023) and Yunker et al. (2009), relied on caregiver reports, clinical observations or retrospective chart reviews without the use of standardised behavioural assessments. While these approaches can provide useful contextual information, they lack the precision and reproducibility of formal testing, and rarely offer domain-level detail.

3.8 | Sensory Impairment and Cognitive Decline

Two studies examined the association between sensory impairments and cognitive decline in adults with DS. Hewitt et al. (1985) found that both hearing and visual impairments were significantly associated with reduced intellectual functioning, as evidenced by a lower mental age ($p<0.0001$) measured by the SBIS. While domain-level scores were not reported, the SBIS evaluates multiple facets of cognitive ability—including verbal and nonverbal reasoning, short-term memory and processing speed—suggesting that the observed decline in mental age may reflect broader impairments across these domains. Similarly, Määttä et al. (2006) reported that individuals with sensory impairments exhibited significantly greater severity of intellectual and speech impairments ($p<0.05$) compared to adults with DS without a sensory impairment, though the specific cognitive domains affected were not clearly delineated.

TABLE 4 | Summary of studies showing uncorrected visual impairments associated with cognition or behaviour in individuals with Down syndrome.

Study info		Demographics				Methods and results		Evidence ratings			
Author(s)/ year	Title	Country	Study	Mean age (years)	Comparison		Vision assessment	Cognition/ behaviour assessment type	Findings	JBI	GRADE
					n	group(s)					
(de Weger et al. 2021)	Differences between children with Down syndrome and typically developing children in adaptive behaviour, executive functions and visual acuity	NET	CS	8.8	104	None	Visual acuity charts	Vineland-S, MEFS and BRIEF	Severity of visual impairment is related to adaptive behaviour challenges of children ($r = -0.396$, $p = 0.001$)	3	Higher
(Hewitt et al. 1985)	Ageing in Down's syndrome	UK	C	54.3	23	None	Visual acuity charts	SBIS	Vision loss is associated with significant intellectual deterioration (lower mental age) ($p < 0.0001$)	3	Moderate
(Krinsky- McHale et al. 2012)	Ophthalmic disorders in adults with Down syndrome	US	CS	50.93	455	None	Medical records review of ophthalmic diagnose	AAMR ABS, WISC-R, and BVMI	Ophthalmic disorders are negatively associated with adaptive behaviour ($p = 0.048$, $d = 0.193$), visuospatial organisation ($p = 0.005$, $d = 0.312$), and visual- motor integration ($p = 0.048$, $d = 0.216$)	3	Moderate
(Määttä et al. 2006)	Sensory impairments and health concerns related to the degree of intellectual disability in people with Down syndrome	FIN	CS	M: 29; F: 35	129	None	Visual acuity charts	IQ-based classification of intellectual disability (mild- profound); speech ability categorised from medical records	Vision loss is significantly associated with the severity of intellectual and speech impairment ($p < 0.05$)	3	Low
(Merrick and Koslowe 2001)	Refractive errors and visual anomalies in Down syndrome	ISR	CS	12.5	86	None	Cycloplegic and ophthalmologic examination	No specifications on methodology of separating into mild, moderate and severe learning difficulty groups	Greater degrees of strabismus correlate with greater learning difficulties ($p < 0.01$)	3	Low

(Continues)

TABLE 4 | (Continued)

Study info			Demographics			Methods and results			Evidence ratings	
Author(s)/ year	Title	Country	Study	Mean age (years)	<i>n</i>	Comparison group(s)	Vision assessment	Cognition/ behaviour assessment type	Findings	JBI GRADE
(Nandakumar and Leat 2010)	Bifocals in children with Down syndrome (BIDS) —visual acuity, accommodation and early literacy skills	CAN	NRE	8–18	14	None	Visual acuity charts, cycloplegic and refraction	WRMT-G, TVPS-R and BVMI	Improvement in visual acuity results in improved scores in early literacy skills ($p = 0.047$)	2 Moderate
(Nandakumar et al. 2011)	Bifocals in Down syndrome study (BIDS): analysis of video recorded sessions of literacy and visual perceptual skills	CAN	NRE	8–18	14	None	Distance and near visual acuity, cycloplegic and refraction	WRMT-G, TVPS-R, and BVMI	Bifocals lead to faster and improved performance on literacy tests including word identification ($p = 0.008$) and reading speed ($p = 0.0015$)	2 Moderate
(Pikora et al. 2014)	Health conditions and their impact among adolescents and young adults with Down syndrome	AUS	CS	23.6	197	None	Parent-reported visual deficits	Index of Social Competence (measures communication, self- care and community skills); parental reports of mental health (anxiety and depression)	Visual impairments are associated with increased safety concerns and restrict opportunities to participate in employment and community leisure activities	3 Low
(Roeden and Zitman 1995)	Ageing in adults with Down's syndrome in institutionally- and community- based residences	NET	CS	DSi: 53.8; DSg: 40.5	71	Learning- impaired controls with age, living conditions and IQ matched; $n = 46$	Stycar Vision Test and medical records review	VABS, SONIT 2.5–7	Vision loss is significantly related to increased challenges with daily living skills ($p < 0.05$)	3 Moderate

(Continues)

TABLE 4 | (Continued)

Study info			Demographics			Methods and results			Evidence ratings		
Author(s)/ year	Title	Country	Study	Mean age (years)	n	Comparison group(s)	Vision assessment	Cognition/ behaviour assessment type	Findings	JBI	GRADE
(Roeden and Zitman 1997)	A longitudinal comparison of cognitive and adaptive changes in subjects with Down's syndrome and an intellectually disabled control group	NET	C	46.3	67	Non-DS with intellectual disabilities; age and living conditions similar to the DS group; <i>n</i> = 48	Stycar Vision Test and medical records review	VABS, SONIT 2.5–7	Visual deficits are significantly associated with impaired personal daily living skills (<i>p</i> < 0.05).	3	Moderate
(Wilton et al. 2021)	Behavioural features of cerebral visual impairment are common in children with Down syndrome	UK	CS	9.9	8	None	Visual acuity and retinoscopy	51-question cerebral visual impairment screening inventory	Refractive error severity is related to severity of behavioural challenges (bumping into doors, unable to finish food on plate) (<i>p</i> = 0.010)	3	Low

Note: Summary of studies showing uncorrected visual impairments associated with cognition or behaviour in individuals with Down syndrome. This table includes study information (author(s), year, title, country and study type), participant demographics (mean age, sample size and presence of comparison groups), vision assessment methods, cognition and behaviour assessment tools, key findings and evidence ratings.

Abbreviations: Countries: AUS—Australia, CAN—Canada, FIN—Finland, ISR—Israel, NET—Netherlands, UK—United Kingdom, US—United States; Study Types: C—Cohort study, CS—Cross-sectional study, NRE—Nonrandomized experimental study; Demographics: DS—Down syndrome, DSj—those with Down syndrome who were institutionalised, DSG—those with Down syndrome living in a group home; Cognition/Behaviour Assessments: AAMR ABS—American Association on Mental Retardation Adaptive Behaviour Scale, BRIEF—Behavioural Rating Inventory of Executive Function, BVMI—Beery-Buktenica Developmental Test of Visual-Motor Integration, MEFS—Minnesota Executive Function Scale, SBIS—Stanford-Binet Intelligence Scale, SONIT—Snijders-Oomen Nonverbal Intelligence Test, TVPS-R—Test of Visual-Perceptual Skills—Revised, VABS—Vineland Adaptive Behaviour Scales, Vineland-S—Vineland Adaptive Behaviour Screening, WISC-R—Wechsler Intelligence Scale for Children-Revised, WRMT-G—Woodcock Reading Mastery Tests—Grade level; Evidence Ratings: GRADE—Grading of Recommendations Assessment, Development, and Evaluation, JBI—Joanna Briggs Institute level of evidence.

INFANCY (0 - 2)	CHILDHOOD (2 - 18)	ADULTHOOD (18+)
No studies found in this age range	<p>Bumping into doors and not seeing food on plate (Wilton et al. 2021)</p> <p>Lower literacy skills (Nandakumar & Leat 2010; Nandakumar et al. 2011)</p> <p>Greater learning challenges correlated with the severity of visual impairment (Merrick & Koslowe 2001)</p> <p>Lag in adaptive behaviours correlated with the severity of visual impairment (De Weger et al. 2021)</p>	<p>Adaptive behaviour challenges (Krinsky-McHale et al. 2012)</p> <p>Barriers to employment opportunities (Pikora et al. 2014)</p> <p>Greater challenges with performance on daily living skills (Roeden & Zitman 1995; Roeden et al. 1997)</p> <p>Intellectual deterioration (Hewitt et al. 1985)</p> <p>Lower overall cognitive ability (Määttä 2006)</p> <p>Speech impairment (Määttä et al. 2006)</p> <p>Visual-motor integration challenges (Krinsky-McHale et al. 2012)</p>

FIGURE 3 | Associations between uncorrected visual impairment and behaviour or cognition across lifetime in individuals with Down syndrome.

3.9 | The Effects of Interventions

Four studies demonstrated the direct benefits of treating sensory impairments in terms of language development, academic achievement and social skills. Intensive treatments, such as speech and language pathology referrals early in childhood (before the age of 1), alongside routine otological management, were shown to lead to age-appropriate oral language development for the first 2 years of life compared to those who did not receive adequate treatment (Pappas et al. 1994). For those with auditory sensitivities, hearing protection and noise avoidance positively affected behaviour and improved social interactions (Yunker et al. 2009) (Table 3). As for vision, the provision of bifocals to improve near acuity improved literacy skills and overall educational achievement at school in children with DS (Nandakumar and Leat 2010; Nandakumar et al. 2011). Additionally, eight studies emphasised the value of continued routine screening for hearing and visual impairments and a ‘health maintenance’ approach to monitor infections and illnesses throughout life (de Weger et al. 2021; Merrick and Koslowe 2001; Pappas et al. 1994; Pikora et al. 2014; Pradilla et al. 2020; Prasher 1995; Roizen et al. 1993; Turner et al. 1990). Referrals for speech, language and physical therapy are also recommended as early as possible if any cognitive or behavioural challenges do develop (Laws and Hall 2014; Namatame et al. 2019; Yunker et al. 2009).

4 | Discussion

This review highlights the associations between sensory impairments and cognition or behaviour in individuals with DS throughout different life stages. Thirty studies out of 38 in this review found at least one association between sensory impairments and language, motor ability, intellect or activities of daily living. The majority of studies were graded as ‘Moderate’ certainty of evidence (23 studies) and examined childhood (18 studies). The most common findings related hearing impairments in infancy and childhood to language delays and visual

impairments in childhood and adulthood to adaptive behaviour challenges; however, high quality longitudinal studies are needed to better establish directionality and strengthen the evidence base.

While the absence of statistical significant relationships in some studies may reflect true null effects, it is also possible that methodological limitations—such as small sample sizes, lack of longitudinal data or insensitive measurement tools—obscured existing relationships. The large number of ‘Low’ certainty studies highlights the need for more rigorous research designs capable of detecting subtle or domain-specific associations of sensory impairments on child and adult outcomes in this population.

Notably, many of the same domains associated with sensory loss in childhood—such as language, adaptive behaviour and learning capacity—were also present in adulthood, suggesting a possible continuity of associations across the lifespan. While direct comparisons between early and late onset sensory impairments were not made in the included studies, the consistent findings of multidomain cognitive and behavioural associations in childhood suggest that sensory impairments during critical developmental periods may be linked to more persistent challenges than those identified later in life. For example, uncorrected hearing loss in infancy and early childhood was frequently associated with delays in language, motor development and verbal reasoning—skills foundational to later cognitive development. Similarly, childhood visual impairments were linked to literacy difficulties and reduced adaptive functioning, indicating potential disruption of early learning trajectories. However, it is important to note that these studies often did not report the age of onset or duration of the sensory impairments, making it unclear whether the observed associations reflected recent onset or the long-term consequences of impairments present since childhood. These patterns underscore the importance of regular hearing and vision screening—not only to address immediate concerns but also to support long-term developmental outcomes. By synthesising findings across the lifespan, this review offers a

TABLE 5 | Summary of studies showing no or mixed associations between uncorrected hearing or visual impairments and cognition or behaviour in individuals with Down syndrome.

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/ year	Title	Country	Study	Mean age (years)	<i>n</i>	Comparison group(s)	Vision/ hearing assessment	Cognition/ behaviour assessment type	Findings	JBI	GRADE
(Dressler et al. 2015)	Vision problems in Down syndrome adults do not hamper communication, daily living skills or socialisation	IT	C	28.7	49	None	Visual assessments for refractive errors, strabismus and cataracts	VABS and CPM	Visual anomalies are not associated with impaired communication, daily living skills or socialisation	3	Moderate
(Islam et al. 2022	Age and gender- related differences in quality of life of Bangladeshi patients with Down syndrome: a cross-sectional study	BAN	CS	6.2, 13.8 and 34.6	468	None	Self-reported or family-reported deficits in survey	The level of disability was determined by the consulting physician and IQ test	Hearing loss (<i>p</i> = 0.32) and vision loss (<i>p</i> = 0.49) were not significantly associated with severity of intellectual functioning challenges	3	Low
(Jarroll and Baddeley 1997)	Short-term memory for verbal and visuospatial information in Down's syndrome	UK	NRE	13.3	15	Children with moderate learning difficulties (<i>n</i> = 15) and normal developing groups (<i>n</i> = 15), matched for vocabulary age however DS group significantly older than both control groups (<i>p</i> < 0.01)	Word discrimination ability assessed at different sound levels	MTDT	Hearing difficulties have minimal effect on ability to discriminate words at relatively high sound intensity levels	2	Higher

(Continues)

TABLE 5 | (Continued)

Study info		Demographics				Methods and results			Evidence ratings		
Author(s)/ year	Title	Country	Study	Mean age (years)	<i>n</i>	Comparison group(s)	Vision/ hearing assessment	Cognition/ behaviour assessment type	Findings	JB1	GRADE
(Jarrod et al. 2002)	Verbal short-term memory in Down syndrome: A problem of memory, audition or speech?	UK	CS	14.3	19	Mild developmental delay (<i>n</i> = 19) and normal development (<i>n</i> = 19) groups, matched for verbal mental age, visual and hearing deficits excluded in control groups	Tests of digit memory with and without the addition of visual stimulus support	DST and CBCL	Hearing difficulties are not associated with challenges in verbal short-term memory	3	Moderate
(Marcell and Cohen 1992)	Hearing abilities of Down syndrome and other mentally handicapped adolescents	US	CS	18.8	26	Mentally handicapped non-DS; matched IQ and chronological age; <i>n</i> = 26	Pure-tone audiometry and tympanograms	TOLD-2P, DST, Miller-Yoder Comprehension Test, backward masking, auditory and gating task	Hearing loss correlated with poorer word recognition in noise (<i>p</i> < 0.05), though hearing difficulty did not predict performance in complex cognitive tasks (such as sentence imitation and sentence comprehension)	3	Moderate
(Pradilla et al. 2020)	Prevalence of hearing loss in a population of schoolchildren with Down syndrome from Bogotá, Colombia	COL	CS	11.23	40	None	Audiometry	Parent-reported language developmental milestones	Hearing loss is not associated with delays in language developmental milestones	3	Low

(Continues)

TABLE 5 | (Continued)

Study info		Demographics					Methods and results			Evidence ratings	
Author(s)/ year	Title	Country	Study	Mean age (years)	n	Comparison group(s)	Vision/ hearing assessment	Cognition/ behaviour assessment type	Findings	JBI	GRADE
(Roeden and Zitman 1995)	Ageing in adults with Down's syndrome in institutionally- and community- based residences	NET	CS	DSi: 53.8; DSg: 40.5	71	Learning- impaired controls with age, living conditions and IQ matched; <i>n</i> = 46	Stycar Hearing Test and audiometry	VABS and SON 2-7	Hearing loss is not related to worse performance on daily living skills	3	Moderate
(Roeden and Zitman 1997)	A longitudinal comparison of cognitive and adaptive changes in subjects with Down's syndrome and an intellectually disabled control group	NET	C	46.3	67	Non-DS with intellectual disabilities; age and living conditions similar to the DS group; <i>n</i> = 48	Stycar Hearing Test and audiometry	VABS and SON 2-7	Hearing impairments are not significantly associated with functional impairments	3	Moderate
(Roizen et al. 1993)	Hearing loss in children with Down syndrome	US	CS	0.17-3.5	47	None	Auditory brainstem response, pure-tone audiometry, tympanometry and otoscopy	REEL	Hearing loss is not associated with language quotients	3	Moderate

(Continues)

TABLE 5 | (Continued)

Study info			Demographics			Methods and results			Evidence ratings	
Author(s)/ year	Title	Country	Study	Mean age (years)	n	Comparison group(s)	Vision/ hearing assessment	Cognition/ behaviour assessment type	Findings	JB1 GRADE
(Schworer et al. 2022)	Associations among co- occurring medical conditions and cognition, language and behaviour in Down syndrome	US	CS	12.7	73	None	Parent-reported hearing or visual deficits	SBIS-5, EVT, PPVT, SRS- 2, CBCL, and BRIEF-2	Hearing and visual deficits are not significantly associated with cognition, language, social behaviour, adaptive behaviours or executive functioning	3 Moderate
(Turner et al. 1990)	Health problems in children with Down's syndrome	UK	CS	9.2	117	None	Parent-reported along with audiometry and formal eye testing	Parental and teacher questionnaire on behavioural issues, school attendance, and adaptive functioning based on Parental Child Health Form, National Study for Child Health and Education	Hearing and visual deficits are not linked to behaviour	3 Low

Note: Summary of studies showing no or mixed associations between uncorrected hearing or visual impairments and cognition or behaviour in individuals with Down syndrome. This table includes study information (author(s), year, title, country and study type), participant demographics (mean age, sample size and presence of comparison groups), vision and hearing assessment methods, cognition and behaviour assessment tools, key findings and evidence ratings.

Abbreviations: Study Types: C—Cohort study, CS—Cross-sectional study, NRE—Nonrandomized experimental study; Demographics: DS—Down syndrome; Cognition/Behaviour Assessments: BRIEF-2—Behavioural Rating Inventory of Executive Function-2, CBCL—Child Behaviour Checklist, CPM—Coloured Progressive Matrices, DST—Digit Span Task, EVT—Expressive Vocabulary Test, MTDT—McCormick Toy Discrimination Test, PPVT—Peabody Picture Vocabulary Test, REEL—Receptive and Expressive Emergent Language Test, SBIS-5—Stanford-Binet Intelligence Scales, Fifth Edition, SON 2-7—Snijders-Oomen Nonverbal Intelligence Test for Children Aged 2.5-7, SRS-2—Social Responsiveness Scale-2, TOLD-2P—Test of Language Development-2 Primary, VABS—Vineland Adaptive Behaviour Scales; Evidence Ratings: GRADE—Grading of Recommendations Assessment, Development, and Evaluation, JBI—Joanna Briggs Institute level of evidence.

novel framework for considering how the timing and duration of sensory impairments may relate to the severity and scope of cognitive and behavioural differences in individuals with DS.

4.1 | Implications for Practice

The potential interplay between sensory impairments and behavioural and cognitive outcomes in individuals with DS is an important consideration for providers. Behavioural features, such as social or communication challenges, have been shown through this review to be associated with sensory impairments, yet these behaviours may be misattributed solely to psychological or developmental differences. As such, treatment plans could mistakenly target symptoms or sequelae rather than an underlying cause that is treatable. Cognitive features—such as difficulties with attention, verbal memory or problem-solving—may also be associated with an unrecognised sensory impairment. Overlooking the potential consequences of untreated hearing or vision loss may lead to misdiagnosis and management approaches that do not fully address the individual's needs—curtailing developmental potential or the preservation of cognition in adulthood.

4.2 | Strengths and Limitations

The strength of this review lies in its comprehensive approach, encapsulating a wide range of studies without date or language restrictions and including individuals across the lifespan to provide a broad understanding of how sensory impairments may be associated with developmental and functional outcomes. Another strength of this review is the incorporation of a structured assessment of evidence across all included studies using a modified GRADE framework. This approach provided a nuanced evaluation of study rigour based on design, sample size, statistical significance and assessment quality. This allowed for a more informed interpretation of the strength of evidence and helped identify areas where future research could be improved methodologically.

However, the large range of studies found across the world also introduced several limitations. Of the 1471 studies initially screened, only 38 met the inclusion criteria, highlighting the inherent challenge of developing search strategies that adequately capture the diverse terminology, assessments and outcomes related to sensory and neurodevelopmental domains. Geographical variation in healthcare infrastructure, screening protocols and cultural context may also influence both the identification and treatment availability of sensory impairments. Study designs and sample sizes varied widely, and inconsistencies in the definitions and diagnostic criteria for hearing and visual impairments further complicated synthesis and limited comparability across studies, as demonstrated by the large range of GRADE ratings ('Very Low' to 'Higher' certainty). Not all studies with control groups adjusted for potential confounders, such as co-occurring conditions like autism—which is reported in as high as 39% of people with DS—also limiting the strength of the conclusions (Spinazzi et al. 2023). Additionally, many included studies were retrospective rather than prospective, often assessing participants' current cognitive, behavioural or sensory

status without a clear timeline of when impairments or associated outcomes first emerged, or if previous treatments such as hearing aids or glasses were used before the study. This limits the ability to infer directionality or developmental timing of associations. Notably, 57.9% of studies were published 15 or more years ago, during periods when screening practices, diagnostic tools, and access to services may have differed significantly from current standards.

4.3 | Gaps in Literature and Future Directions

With the low number of relevant studies (38 out of 1471 screened) and only 11 being experimental, cohort or case-control studies, this review highlights the need for more rigorous and targeted research to better understand the associations between cognitive and behavioural domains with visual and hearing impairments in individuals with DS. Additionally, longitudinal studies are needed to understand the progression and long-term effects of sensory impairments to help provide directionality to the associations identified in this review.

No studies were found assessing associations between visual impairments and cognitive or behavioural issues in infancy (0 to 2 years of age). This may be in part due to the challenges of assessing vision at this young age, though the current guidelines from the American Academy of Pediatrics recommend visual assessments as early as 6 months of age for this population given the high rates of visual impairments in childhood, such as cataracts and nasolacrimal duct obstruction (Bull et al. 2022). Therefore, more studies are needed to examine the prevalence of visual impairments in infancy and their associations with cognition and behaviour and to develop effective treatments or early interventions.

One area of research that was not fully captured through this scoping review was the association between sensory impairments and Alzheimer's disease (AD) in people with DS. While two studies in this scoping review identified an association between visual or hearing impairments and cognitive challenges in adulthood (Hewitt et al. 1985; Määttä et al. 2006), other studies outside of this scoping review provide additional evidence for this relationship with AD (Hwang et al. 2020; Benejam et al. 2022; Lad et al. 2024; Lin et al. 2011; Liu et al. 2016; McCarron et al. 2005; Pache et al. 2003). In the general population, dual sensory impairments are associated with a 112% increased risk of developing AD dementia (Hwang et al. 2020; Lad et al. 2024). Non-specific loss of colour vision, seen in the DS population, has been linked to AD pathological changes in the visual association cortex (Pache et al. 2003). AD-related degeneration in the posterior parietal and inferior temporal cortices affects depth perception (Benejam et al. 2022). Similarly, hearing loss in both the general population and animal models has been associated with increased incidence of cognitive decline in those with AD dementia (Lin et al. 2011; Liu et al. 2016). In adults with DS, a longitudinal cohort in the UK found that individuals with DS and AD had higher prevalences of both vision and hearing loss compared to those with DS and no dementia (McCarron et al. 2005). Given the high prevalence of co-occurring sensory conditions in adults with DS, additional studies are needed to better understand the causal relationship between sensory loss

and AD and whether treatment of sensory loss could mitigate risk for dementia.

5 | Conclusion

This scoping review highlights a growing body of evidence suggesting that uncorrected hearing and visual impairments in individuals with DS are associated with a range of cognitive and behavioural differences across the lifespan. The strongest associations were observed between hearing impairments and language development delays in infancy and childhood, and between visual impairments and adaptive behaviour challenges in childhood and adulthood. Several domains found to be associated with sensory loss in childhood—such as language development, adaptive functioning and learning capacity—were also reported in adult populations, suggesting a possible continuity of associations over time. The consistent presence of cognitive associations in both developmental and later-life periods raises the possibility that uncorrected sensory loss may be associated with reduced cognitive reserve and increased vulnerability to decline over time.

The current literature is limited by methodological variability, underrepresentation of early childhood (particularly infancy) and a lack of longitudinal data needed to clarify the timing and directionality of these associations. The majority of studies were conducted over 15 years ago and may not reflect current screening and intervention standards. Future research should prioritise longitudinal, mechanistically informed designs and report sensory histories across the lifespan to better understand how the timing and duration of hearing and vision loss relate to developmental and cognitive outcomes in this population.

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Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

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