education sciences

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Review

Children Who Are Deaf/Hard of Hearing with Disabilities: Paths to Language and Literacy

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Received: 3 May 2019; Accepted: 8 June 2019; Published: 14 June 2019

Abstract: Students who are Deaf with Disabilities (DWD) comprise an extremely heterogeneous population. Similar to students who are d/Deaf or hard of hearing (DHH), students who are DWD vary in terms of degree, type, and age at onset of hearing loss, amplification, and preferred communication method. However, students who are DWD are also diverse in terms of type, etiology, and number and severity of disability(ies). Presented in this article is an overview of DWD followed by foci on Deaf with learning disabilities, Deaf with intellectual disabilities, Deaf with autism spectrum disorder, and deafblindness. Particular attention is given to communication, language, and literacy development.

Keywords: deaf with disabilities 1; deafblind 2; autism spectrum disorder 3; learning disabilities 4; intellectual disabilities 5

1. Introduction

The population of students who are dDeaf/hard of hearing with disabilities (DWD) is difficult to characterize and quantify, yet is thought to comprise between 40% to 50% of the population of students who are d/Deaf or hard of hearing (DHH) in the United States [1–4]. Students who are DWD are extremely diverse and reflect both diversity of the DHH population as well as variation in type and severity of the various accompanying disabilities (e.g., variation in expression of autism spectrum disorder) [5]. Further, etiologies of deafness are considered to be possible risk indicators for concomitant disabilities [2,6,7]. Such etiologies include hereditary syndromes (e.g., Usher, CHARGE, Goldenhar, and Down syndromes), maternal infections (e.g., congenital rubella, cytomegalovirus, toxoplasmosis), prematurity, meningitis, anoxia, and trauma [2,6–9]. Identified concomitant morbidities include intellectual or developmental disabilities, autism spectrum disorder (ASD), specific learning disabilities, attention deficit disorder (ADD), attention deficit hyperactive disorder (ADHD), orthopedic impairments, emotional disabilities, speech and language impairments, traumatic brain injury, health impairments, low vision, legal blindness, and deafblindness [4–6]. The disabilities may include one or several of those listed and each of the disabilities vary in presentation and degree. However, it is important to note that the disabilities are multiplicative rather than additive as each interacts with the hearing loss to impact communication, cognition, social development, behavior, and physical development [4,6,10,11]. Moreover, the presence of disabilities makes compensation for loss of hearing much more difficult [9,12]. In recent years, there has been an increase in attention and research focused on the area of DWD [13,14]; however, there remains much to be known about prevalence, accurate assessment methodology, and effective intervention [3,10,12,15]. In addition, there is a shortage of teachers prepared to teach this unique student population and few teacher preparation programs provide coursework related to working with students who are DWD [5,6,10,14]. Included in this article is an overview of the

population of children and youth who are DWD including estimated prevalence, difficulties encountered in accurate assessment, and education, language, and literacy considerations. Attention is then turned to four of the most prevalent concomitant disabilities—learning disabilities (LD), intellectual disabilities (ID), ASD, and deafblindness. The extant research base in each area is presented with particular emphasis on communication and literacy development.

2. Prevalence and Identification

Estimated prevalence of students who are DWD varies in the literature with a reported range of 25% to 51% of students who are DHH [2,3,6,7,10,15]. In a 2010 study of 100 children with severe to profound sensorineural hearing loss, Chilosi et al. [2] found that 48% of the sample had one or more additional disabilities. The majority of other studies reporting DWD prevalence cite the Annual Survey of Deaf and Hard of Hearing Children and Youth (Annual Survey) conducted by the Gallaudet Research Institute (GRI) as their source. Since the survey began including data on students who are DWD in 1999, prevalence has remained relatively constant with a mean of 42% across the years from 1999 to 2013 [3]. The latest available statistic from the GRI count is 2013 which reports that 59% of the DHH population had no accompanying disabilities and therefore, 40% had identified concomitant disabilities [4]. However, the population of students who are DHH in this count is much lower than in previous counts. Researchers in the field posit that the GRI count is likely under representative of the population because not all respondents complete the section on disabilities [3,15]. In addition, there are differences among the child count surveys reporting student disability in the United States. For example, in the 2009–2010 school year, The United States Department of Education Child Count reported 79,431 children receiving services under the category of deaf-hard of hearing. In the same year, the GRI reported 37,608 students as deaf-hard of hearing. In looking at the population of students who are deafblind in 2009–2010, the Child Count [16] reported 1575 students as deafblind, the GRI, 1778 [17], and the Deaf-Blind census conducted by the federally funded National Consortium on Deaf-Blindness (NCDB) reported 9195 children and youth as deafblind [18]. This discrepancy is likely because the Department of Education Child Count only counts children by primary disability (e.g., deaf/hard of hearing or deaf-blind), and NCDB has focused national attention on identifying students as deaf-blind. However, given the relatively high deaf-blind count by NCDB, it could be argued that other accompanying disabilities might also see higher prevalence numbers if attention was focused on their identification. Importantly, in examining the variance across data sources and the lack of concrete prevalence figures, it is likely that the population is underserved as well as under-identified [19].

Early intervention is critical to communication, language, and social-emotional development in children who are DHH [20,21]. Further, early intervention has been demonstrated to positively impact outcomes across domains of children with disabilities [22,23]. However, identification of disabilities in children who are DHH is challenging, and hence, diagnosis and subsequent early intervention frequently occurs at an older age than children who are DHH alone [8,24–26]. Therefore, opportunities for early, comprehensible language input are missed [5,24,27]. Challenges in identification of children who are DWD include: (a) Diagnostic overshadowing [27,28], (b) difficulties obtaining accurate auditory assessment information [29,30], and (c) limitations inherent in existing assessment instruments [10,12,15]. Diagnostic overshadowing occurs when two disabilities share common indicators and the first identified or the most prominent "overshadows" identification of the second. For example, because communication and social skills are affected in both ASD and DHH, ASD might be missed in a child who is deaf or vice versa [8,25,27,28,31]. Further, auditory assessment is difficult when a child, because of physical, communication, or cognitive difficulties, is unable to reliably indicate responses to auditory stimuli. Therefore, determination of degree and type of hearing loss can be inaccurate [29,32,33]. Finally, available assessment tools for disabilities are often not designed to accommodate for the complex needs of children who are DWD [10,12,13,15,28,34] and in fact, some assessment instruments specifically advise that instruments not be used to evaluate children with a hearing loss (e.g., the Autism Diagnostic Observation Schedule (ADOS)) [35]. Given such challenges, there is inherent danger that children will be misdiagnosed and receive education

that is inadequate and inappropriate in meeting all of the needs that result from the disabilities [12,14,19,36]. Further, because the diagnosis of DWD is often delayed, research on effective intervention strategies has tended to focus on older children and thus, evidence-based strategies for early intervention for children who are DWD are scarce [5]. However, even among the school-age population, there is a distinct lack of knowledge regarding effective curriculum methods and teaching strategies [10].

3. Educational, Language, and Literacy Considerations

3.1. Educational Considerations

According to the Individuals with Disabilities Education Act of 2004 (IDEA, 2004) schools must provide special education and related services that are designed to meet the needs of children with disabilities as adequately as the needs of students without disabilities are met [37]. IDEA further requires that the Individualized Education Program (IEP) address all education needs that result from the child's disability(ies), but while IDEA provides definitions of educational disability labels, it is silent on primary and secondary disability [38]. Therefore, a child can be classified on the IEP as DHH, but if other disabilities are present, IDEA requires that the full range of needs be addressed regardless of assigned label. Further, IDEA 2004 mandates that students with disabilities be educated in the least restrictive environment (LRE) and that a continuum of placements be available. IDEA 2004 also specifically delineates that for students who are DHH, teams must consider a student's language and communication in developing the IEP (Title 1,B,614,(d),(3)(B)(iv)). Increasingly, students who are DWD are educated in inclusive settings with typically developing peers and as of 2013, the majority of students who are DWD were included in general education settings (ASD and ID, to a lesser extent) [3,4]. Given the range and intensity of needs, including communication, it is imperative that teachers collaboratively team with professionals across disciplines if individual needs, including communication and literacy needs, resulting from the hearing loss as well as accompanying disabilities are to be met in any of the settings along the placement continuum [6,10,14,38,39].

3.2. Communication Considerations

Children who are DHH vary widely in degree of hearing loss, age at onset of hearing loss, presence of assistive listening devices including hearing aids and cochlear implants, home language, and chosen communication modalities [39]. However, across the range, language development is often delayed due to reduced access to language [1,20]. Such delays are likely to be magnified in children who are DWD who might struggle with receptive and expressive language as well as both visual and auditory language [1,11]. Children who are DWD with physical disabilities may have difficulty with fine motor control that can inhibit both sign and spoken language [11]. Children with ASD may struggle with visual attention to faces and gestures and both communication and social delays are known hallmarks of ASD [5,27,40]. Finally, children with intellectual disabilities may have difficulties with joint attention and might need additional processing time to receive and express language [1,26]. Studies examining auditory receptive and expressive language development of children who were DWD and received cochlear implants or hearing aids prior to 3 years of age found that as a group, at age 3 and 5, language scores on standardized tests improved; however, when examined by type of disability, language scores of children with ASD, developmental disabilities, and cerebral palsy in addition to hearing loss, declined relative to typically developing peers. Better outcomes were significantly associated with higher maternal education [13,41]. Such findings suggest that more research in the field is needed in order to enhance early intervention with families in these particular disability areas in order to improve language outcomes. Moreover, although still relatively uncharted territory [15], research is emerging across deafness and disabilities in terms of effective strategies for enhancing visual language, cochlear implantation habilitation, and use of augmentative and alternative communication (AAC).

3.3. Literacy Considerations

Language is a critical foundation for reading, whether it is expressed in signed language or spoken language [42–44]. Traditional or conventional literacy is often defined as reading and writing. Emergent literacy is a term applied to a stage of literacy development that occurs before conventional reading and writing and involves experiences with literacy and a growing knowledge of the components of early literacy, including identification of letters, learning letter sounds, and writing one's name [45]. Children then develop increasingly complex reading and writing skills in a variety of genres. In some fields the definition of literacy has expanded to include participation in literacy activities and communication [46]. Digital literacy refers to competencies in the use of online resources to accomplish literacy tasks [47]. Whether signed or spoken, language is the foundation of literacy [42–44]. If we are to prepare children for the literacy demands of this century, there is a dire need to increase the attention paid to their experiences, concept development, vocabulary, and deeper level comprehension [45].

Literacy challenges in the DHH population may stem from a lack of early identification, reduced exposure and engagement in language and literacy, inappropriate educational supports for the development of communication and language, and additional disabilities [48,49]. Moreover, disabilities compound the literacy challenges faced by DWD. While the professional literature addresses literacy development for children with each of the disabilities addressed in this paper, there is very little information available about teaching literacy to students who are DWD. One is left to apply the literacy research in the disability area with what is known about teaching literacy to DHH.

4. d/Deaf/Hard of Hearing with Learning Disabilities

The Individuals with Disabilities Education Improvement Act defines a specific learning disability as a disorder in one or more of the basic psychological processes involved in understanding or in using language, spoken or written, that may manifest itself in the imperfect ability to listen, think, speak, read, write, spell, or do mathematical calculations (Sec. 602 (30)(a)) [37].

The literacy delays that are often experienced by children who are DHH make identification of a learning disability difficult. Additionally, the disability and English proficiency exclusion criteria in the Individuals with Disabilities Education Act may erroneously result in some professional teams attributing literacy delays solely to DHH or to English learning instead of considering an additional contributing disability [48,50]. In previous authorizations of IDEA, the discrepancy model was followed, meaning that a student must demonstrate a discrepancy between their ability and academic performance prior to identification of a specific learning disability. With the reauthorization in 2004, professional teams may now identify a learning disability based on the child's response to a research-based intervention [51].

The Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5) [52] includes a diagnostic criteria for a "specific learning disorder" (p. 66) that involves the child exhibiting at least one of six characteristics for six or more months and persisting after interventions have been implemented, with substantially delayed academic performance, and onset of learning difficulties in the school years. The six characteristics involve difficulties in reading words, reading comprehension, spelling, writing, numbers and calculations, and mathematical reasoning. The diagnostic criteria include an exclusion principle stating that the characteristics cannot be explained better by a different diagnosis.

In addition to the learning implications stated by IDEA and the DSM-V, The Learning Disabilities Association of America includes implications of specific learning disabilities that include "reasoning, attention, memory, coordination, social skills, and emotional maturity"[53]. Memory challenges may include both visual and auditory information [51]. Some social-emotional challenges may occur due to repeated experiences with academic failure. Professional teams must also be mindful of comorbidity with attention deficit/hyperactivity disorder.

Deafness is an auditory perceptual problem, whereas a learning disability is a processing issue, that may include "visual-perceptual problems, attention deficits, perceptual-motor difficulties,

severe inability to learn vocabulary and English-language structures, consistent retention and memory problems, or consistent distractive behaviors or emotional factors" [50] (Identification, assessment p. 57). In 1989, Laughton suggested the following description of learners who are DHH-LD: Individuals who are DHH with learning disabilities have significant difficulties with acquisition, integration, and use of language and/or nonlinguistic abilities. These disorders are presumed to be caused by the coexisting conditions of central nervous system dysfunction and peripheral sensorineural hearing impairment, and not by either condition exclusively [50] (Identification, assessment p. 57).

Specific learning disability is the most common disability in children and youth who are DHH, with dyslexia (involving difficulties in learning to read) being the most common type of learning disability [48]. Other learning disabilities include dysgraphia (involving difficulties in writing, including fine motor skills) and dyscalculia (difficulties in understanding mathematics). Estimates of prevalence vary. It has been estimated that about 7–8% of DHH children have a learning disability [54]; and that 10% of the deaf population is thought to specifically have dyslexia [55].

4.1. Assessment and Identification

Much of the professional literature on DHH-LD addresses assessment and identification. Teachers of DHH children are critical to the referral process, assessment, and potential identification of a learning disability. In a survey study of 91 teachers, Soukup and Feinstein [50] found that teachers most offered referred children based on observations of visual perceptual issues (which are not associated with DHH) and behavior. These teachers suggested neuropsychological screening as well as assessment of communication and language as being important to identifying a learning disability.

When considering potential referrals, Sheetz [56] suggests observing for learning characteristics that are associated with learning disabilities, but not with DHH, such as difficulties with social skills, metacognition, visual-perception, severe difficulty in learning vocabulary, and inattentiveness (the latter being due to the co-occurrence of LD with attention deficit disorder). Further, Sheetz suggests comparing literacy achievements with DHH peers. Additional potential cues to a learning disability could be serial memory deficits, weak signing and conversational skills, poor comprehension of questions and pronouns, weak spelling skills, and academic achievement and reading scores that are lower than DHH peers [50,57].

When assessing DHH children for a potential learning disability, the following types of data are needed: Case history (including onset of hearing loss), educational history, measures of intelligence, achievement tests, neuropsychological screening (including visual-motor screening), testing for adaptive behavior functioning, audiological and visual screening, and assessment of communication and language (including a measure of mean length of utterance) [44,48,50,57,58]. Additionally, relatively slow visual perceptual speed has been suggested as a potential biomarker of a learning disability in both individuals who are hearing and individuals who are DHH [55]. Caemmerer et al. [48] suggest considering the child's experiences with English and if the parent and child share communication forms, information that should be included within the child's case history. When assessing for dyslexia in DHH learners, instruments used to measure intelligence and to identify dyslexia should be appropriate for DHH learners [50,55].

Information on appropriate assessment instruments and processes is available through the Center on Literacy and Deafness (CLAD). This site (https://clad.education.gsu.edu) [59] includes a checklist "Documentation Guidelines for Learning Disabilities". Additional information on appropriate literacy tests for use with learners who are DHH can be found in Morere and Allan's book, Assessing Literacy in Deaf Individuals: Neurocognitive Measurement and Predictors [60]. This text provides guidance on how to assess academic knowledge, executive functioning, literacy, visual-spatial functioning, among many other areas of relevance when identifying a potential learning disability.

4.2. Literacy

The identification of a learning disability in a DHH child will occur once that child is school-age and has already acquired language. Thus, this section will address reading and writing.

4.2.1. Reading

"Reading is matching speech sounds with print and involves a complex set of skills involving perception (looking at the text); cognition (i.e., logical reasoning, background knowledge, knowledge of concepts, memory); social skills (i.e., theory of mind); and language skills (i.e., phonology, semantics, syntax, and pragmatics or discourse)" [61] (Deaf and hard of hearing students p. 346). The National Institute of Child Health and Human Development identified the following six factors as being the most important to literacy development in hearing children: "Phonological awareness, alphabetics, vocabulary, fluency, text comprehension, and motivation"[42] (Literacy and deaf and hard of hearing p. 151). Each of these factors has become subject to research and debate in the field of deafness, both to evaluate their relative importance to the development of literacy in the DHH child, and to identify how deafness impacts the development of each factor, including the impact of communication modalities on each of the six factors.

Mohammed, Campbell, Macsweeney, Barry, and Coleman [62] describe two camps of thought to explain reading difficulties as being either being phonological in nature (which is the dominant viewpoint and in accordance with the National Reading Panel), or due to perceptual difficulties, either visual or auditory. Dyslexia was thought to be caused by a visual-spatial deficit and now more commonly is thought to be caused by an auditory phonetic processing problem or central linguistic deficit [63]. While Lomas et al. [61] acknowledge that phonological knowledge supports the decoding of new words and recommend direct phonological instruction, they assert that Deaf children learn to read visually as a substitute for applying phonological knowledge, with some applying sign language, fingerspelling, and visual phonics to support reading [42,49,61]. Even with cochlear implantation and an associated increase in speech, this does not necessarily carry over to improved reading [61]. Miller's [64] study suggests that reading comprehension is a perceptual issue (rather than being due to phonological deficits) for both individuals who are DHH and individuals who have LD.

There are three major curricular approaches to teaching literacy to DHH students that are also referenced in the LD literature. The text-based approach (also known as the bottom-up approach) starts with the building blocks of reading, such as morphemes and phonemes, with phonics taught early. While this approach is effective with most hearing children, it is not effective with Deaf children due to their lack of knowledge of the English phonological system [65], which would also be a challenge for children who are Deaf with LD.

The second approach is the subject-based approach (also known as the top-down approach), which recognizes the importance of context and prior knowledge). DHH children may not have the prior knowledge or contextual information to make this approach meaningful [65]. This same limitation may be experienced by children who are DHH-LD.

The third approach is the interactive or compensatory-interactive approach, combining elements of the text-based and subject-based approaches. The interactive approach recognizes that all forms of language (reading, writing, spelling, speaking, and listening) are inter-related [65]. Attention to context and use of the child's language are foundational to this approach. Paul [44] suggests that a sight word approach is limited and other approaches are needed for higher literacy achievements. Strategies associated with the interactive approach include writing to support reading and reading to support writing (such as reading and writing journals), use of high interest readings (such as trade books), and thematic units [65]. In their longitudinal study of children at risk of developmental dyslexia, ages 5–8 years, Helland, Tjus, Hovden, Ofte, and Heimann [66] found that the bottom-up approach was most effective to making gains in phonological awareness and working memory; whereas the top-down approach was most effective in verbal learning, knowledge of letters, and literacy. More research is needed to determine the application of the interactive approach to children who are DHH-LD.

The general approach to teaching children with specific learning disabilities is to offer explicit and systematic instruction with ample opportunities to practice. "Explicit instruction refers to instruction that is clear and direct and in which expected outcomes are conspicuous to students" [51] (Specific learning disabilities, p. 296). Systematic instruction builds on the child's knowledge and applies behavioral principles, such prompting and reinforcement. Reading approaches and programs for children with dyslexia are based on auditory input, but this approach is not helpful to most Deaf learners, including those with learning disabilities, because of the reliance on linking speech to print.

Shared reading has been recognized as important to the reading development of both children who are DHH and children with LD. When parents engage their child in shared reading, they can support active involvement of the child, expand the child's utterances, and gradually expect more of the child's participation over time. During shared reading the child will learn about handling books, that books are read top to bottom and left to right, how language in books is different from speech or sign language, prediction, drawing inferences, gaining vocabulary, while improving listening comprehension and other literacy skills [67]. Parents can support literacy by providing a literacy rich environment with plenty of books in the home, although this is linked to the parents' economic situation. Robertson recommends that shared readings occur more than once daily, that the parent follows the child's interests, and that preschool children with hearing loss need even more hours with literacy materials than hearing children. Parental involvement is also important when the child enters school. Marschark and Knoors state: "We know that parents' involvement in their children's language development, learning, and education is perhaps the best predictor of their educational outcomes" [43] (Educating deaf learners, p. 233).

Pakulski and Kaderavek [68] suggest beginning with stories about the child's actual experiences, called language experience stories, moving to storybooks later. Such books may be shared in the home or at school. Language experience stories are highly engaging to the child. Parents and others should modify their language to the correct level for the child, and provide connections between objects, pictures, print, and sign. Adults must provide a great deal of practice with concepts and vocabulary. When sharing experience stories with a signing child, the adult must consider where to locate signs while referencing objects and pictures in the book. The sign may occur next to or on top of pictures in books, with connections supported by pointing to pictures. When moving on to storybooks, it is helpful to select texts that are predictable (to reduce vocabulary demands), such as those with repeated phrases or refrains [65]. Easterbrooks and Stephenson [69] present the following ten evidence-based literacy practices for students who are DHH, many of which will also apply to learners who are DHH/LD "independent reading, use of technology, phonemic awareness and phonics, metacognitive reading strategies, writing to promote reading, reading in the content areas, shared reading and writing, semantic (meaning-based) approach to vocabulary, morphographemic approach (learning roots, suffixes and prefixes) to vocabulary and fluency" (Examination of twenty approaches, pp. 386–391).

4.2.2. Writing

There is very little research and professional literature about how to teach writing to students who are DHH-LD. Teachers of the DHH should be aware of the writing struggles experienced by students who are DHH and students with learning disabilities. Deaf students tend to exhibit rigid writing, write in shorter sentences, use a limited vocabulary, exhibit issues with clauses, may need support in sequencing of words, and they have difficulty in writing in a passive voice [44,70]. Students with learning disabilities also tend to produce writings that are shorter, with partial or fragmented coverage of topics. They require support with all phases of writing, from planning to revision and across all genres [71]. Background knowledge, memory, and executive functions impact the writing of students with LD [71]. Graham, Collins, and Rigby-Wills [72] conducted a meta-analysis of 53 studies, comparing the writing of students with LD and peers without disabilities. In addition to needing support with all phases of writing, they found that writing "required the orchestration of handwriting, typing, spelling, and sentence construction skills" (Writing characteristics, p. 199) combined with the role of motivation.

Moores [70] suggests that the three approaches in reading have a parallel in teaching writing to students who are DHH and that neither the bottom-up or top-down has resulted in writing performance on par with hearing children. The interactive-compensatory approach includes, "instruction on sign to print, fingerspelling to print, sound to print, and morphological awareness as well as on functional pragmatic aspects" (Specific learning disabilities, p. 51). Performance in written language is similar to reading performance and reading and writing instruction should be integrated [44,67]. Given the varied forms of communication used by students who are DHH-LD, an interactive approach seems worthy of additional research.

Writing strategies from the field of LD, such as the use of graphic organizers, will support some learners who are DHH-LD. Dexter and Hughes [73] conducted a meta-analysis of 16 articles about the effects of graphic organizers across content areas. They found that graphic organizers are effective in improving recall, vocabulary, and higher thinking skills, such as inference.

Teachers will find the Innovation Configuration on Writing, published by the CEEDAR Center to include evidence-based practices in writing that are application to DHH/LD learners, including information about vocabulary instruction, teaching prewriting, and metacognitive reflection. In this document, Troia [74] presents a detailed table of evidence-based writing practices derived from a review of 16 meta-analyses. While these studies are not specific to students who are DHH-LD, this document may suggest potentially effective strategies as well as potential topics for future research. Further it suggests that technology will support the writing performance of students who are DHH-LD. "Technology runs the gamut from basic word processing with grammar and spell checkers to more sophisticated applications such as digital stylus for transcribing notes on a tablet device and then using software to convert the handwritten text to typewritten text, automated scoring of writing samples with feedback and collaborative writing platforms" [74] (Evidence-based practices, writing p. 11). Multi-media instruction will also support students who are DHH-LD to learn to write.

5. d/Deaf/Hard of Hearing with Intellectual Disabilities

According to the American Association on Intellectual and Developmental Disabilities, intellectual disabilities (ID) originate before the age of 18 and are characterized by limitations in (a) adaptive behavior including conceptual skills, social skills and practical skills, and (b) intellectual functioning. An IQ score of 70 to 75 indicates limitations in intellectual functioning [75]. However, the population of individuals with ID is broad, with some individuals requiring limited, intermittent support, and others, more intensive, consistent support [9,76]. In addition, some students with ID have multiple disabilities who, because of the combination of disabilities, often have intensive support needs [2,12,76]. Prematurity is the single most common cause of DHH-ID, however, there are multiple other etiologies including genetic (either syndromic or non-syndromic), causes. One syndrome commonly implicated in DHH-ID is Down syndrome or trisomy 21 [1,9,12]. It is estimated that about two thirds of children with Down syndrome have either a sensorineural or conductive hearing loss [77].

5.1. Assessment and Identification

Identification of ID in children who are DHH is particularly difficult and intellectual disabilities may be under- or over-identified. For example, delays in language and reading might be inappropriately attributed to ID rather than deafness or conversely, in children, with ID, hearing loss might be missed because language delays are attributed to ID rather than possible hearing loss [1,8,12]. It is therefore important that assessments be carried out by an interdisciplinary team comprised of individuals with knowledge of both deafness and ID [1,12]. Further, when using standardized, norm-referenced testing, it is critical that children with DHH be compared to other children who are DHH rather than children who are hearing [1,12].

If undetected and untreated, hearing loss can severely interfere with cognitive, communication, and social development of individuals with intellectual development [1,57,78]. In 2012, Herer [78] published a study in which the hearing of 9,961 Special Olympic athletes with ID across seven sports venues was evaluated [78]. The audiological protocol followed involved ear canal inspection

followed by otoacoustic emission (OAE) screening. The hearing of each athlete not passing the OAE was then further tested using pure tone audiometry followed by tympanometry screening if pure tone testing was failed. Results revealed that 24% of the athletes had previously undetected hearing loss; a rate that is much higher than would be expected in the general population. Of these, 12.8% had sensorineural hearing loss and 10.9% had conductive/mixed loss. Such results suggest that regular hearing evaluations could significantly improve the quality of life of individuals with ID. Moreover, a team approach to audiological evaluation might be necessary to ensure accurate results. In addition, children with ID might need advanced training in play audiometry in order to successfully participate in auditory assessment [1,12,33].

While there is a clear relationship between some etiologies of deafness and the presence of comorbid disabilities, identification of the accompanying disabilities might be delayed [2,25,28]. Chilosi et al. [2] studied 100 children with severe/profound sensorineural hearing loss using a diagnostic protocol that included neurodevelopmental, genetic, neurometabolic, and brain magnetic resonance imaging (MRI) assessment. Forty-eight percent of the sample were found to have one or more additional disabilities. Further, of the 80 children who had MRIs, 37 showed signal abnormalities. The frequency and type of disability were positively correlated with etiology of the hearing loss. The researchers concluded that for children with particular etiologies, assessment that includes brain MRI can detect cortical abnormalities and thereby, contribute to therapeutic intervention.

The link between assessment and intervention is critical to achievement of successful outcomes and improved quality of life of individuals with DHH-ID [1,12] and intervention based on appropriate assessment can contribute to the development of cognition, communication, and literacy skills; skills that are inextricably bound to one another and necessary for high quality of life [13,19,79,80]. However, although research into effective intervention for students who are DWD is low across the disability areas, it is the lowest in the area of DHH-ID [3].

5.2. Communication and Language

Children who are DHH with ID receive and express communication through various means. They may perceive language visually or through the use of assistive hearing devices such as hearing aids and cochlear implants [5,13,26,31]. Expressive communication might be nonsymbolic through facial expressions, gestures, and body movements or symbolic through various sign language systems, spoken language, pictures, and/or AAC [1,5,9,12].

5.2.1. Cochlear Implants

The use of cochlear implants by individuals who are DWD is increasing and concomitantly, an increasing number of research studies examining its effectiveness with the population have been conducted. In a 2008 study, Wily et al. [26] performed a retrospective study of 42 children who received cochlear implants at 36 months of age or younger. Outcomes were measured through an auditory skills checklist that used a combination of parent interview and clinician observation. Findings of the study concluded that regardless of disability type, the children who were DWD made measurable progress in auditory skill development. However, amount of progress was significantly affected by the developmental quotient (DQ) of the children. Children with a DQ of at least 80 progressed at a similar rate to typically developing deaf children. Children with a DQ of less than 80 had approximately half the rate of progress seen in the typically developing deaf children although the rate of progress was commensurate with their developmental delay level [26]. These results are similar to the studies mentioned earlier that found variation in benefit from implantation among disability types [13,41]. A further study by Meinzen-Derr et al. [31] examined post-implant language gains between children with development delays who had cochlear implants and a cognitively matched control group of hearing children. Results indicated that the children with cochlear implants had significantly lower levels of language development than the control group of hearing peers with ID; suggesting that delays were significantly lower than their cognitive potential [31]. Berrettini et al. [81] examined the impact of cochlear implants on speech perception skills using pre-post testing and

parent survey in 23 children with neuropsychiatric disorders. Additionally, the results from ten children with intellectual disabilities were disaggregated and analyzed. The results of all participants were variable but gains were seen in speech perception, communication abilities, and quality of life. The children with intellectual disabilities saw similar gains when compared to other study participants. Of note is that 100% of families reported gains with 74% reporting increases in child speech, 96% in child interaction skills, and 100% in both attention and social skills with classmates [81]. An additional 2014 study by Zaidman-Zait et al. [82] exclusively examined parental perception of cochlear implants in children with a variety of additional disabilities. Parents reported increases in communication skills, enjoyment of music, and safety. Challenges reported were managing the devices, getting children to use them, funding, and multiple appointments [82]. Consistent themes across DHH-ID cochlear implant studies are (a) variation in type of disability, (b) need for knowledge in how to maximize post-implant gains in children with DWD including ID, and (c) recognition of diverse types of benefit that can be attained [13,26,31,41,81,82].

5.2.2. Sign Language and Augmentative and Alternative Communication (AAC)

Augmentative and Alternative Communication or AAC can be broadly divided into unaided and aided types. Unaided types included gestures and signs and aided types utilize an external device. Aided types can be simple devices not dependent on technology (e.g., line drawings, objects) or more high-technology devices that include speech generating devices (SGD) or voice outcome communication devices (VOCA) [1,11,83]. Sign language frequently plays an important role in communication in children with DHH-ID, however, important considerations when teaching sign language are degree of ID, short-term memory skills, ability to physically form signs, and the environment. Therefore, signs may need to be adapted and other communication methods may need to be available to augment sign [9,80]. Such other communication methods might include aided AAC devices. Lee et al. [83] examined the use of a VOCA device with five deaf children with multiple disabilities who had received cochlear implantation and found that use of the VOCA improved speech perception, speech production, receptive vocabulary kills, and communicative behaviors. At preintervention, the children were at a pre-symbolic level of communication and by the end of intervention, all five were able to use the VOCA to express communicative intents [83]. Davis et al. [11] reviewed 14 experimental studies of AAC use among individuals with hearing loss and one or more additional disabilities. Half of the reviewed studies used a non-electronic communication device including photographs, line drawings, or printed words. Of the seven that used an electronic device, six involved an SGD. Most of the participants were teens or young adults which runs contrary to research findings that AAC is more effective when introduced in early intervention before other communication methods fail [84]. An additional finding in the review was that although SGD was used in the studies, only one referenced the issue that the children with DWD might not have access to the speech output. Without such access, the user might not to know if the device was properly working or be able to repair communication breakdown if unable to monitor the communication message [11]. In general, findings of the various studies in the Davis et al., review and the other studies in this section suggest that AAC use does not hamper either speech or sign use but rather increases communication initiation and use. However, there is a need for continued research into AAC use with the population of individuals who are DHH with ID [11,80,83].

5.3. Literacy

The extant research-base on literacy development in children who are DHH-ID is limited to the extreme. However, four studies are of note. The first study blended communication and literacy. Using a low-technology picture dictionary, Highnote and Allgood et al. [80] found students with DHH-ID at a community vocational center were successfully able to use the dictionary to initiate communications. In addition, improvements were seen in spelling of target vocabulary words. The participants also used American Sign Language but since co-workers did not know sign language, the addition of the picture dictionary allowed for increased communications The second study is a pilot study that examined a curriculum developed at St. Joseph's School for the Deaf that used a

reading and writing workshop approach [79]. Participating students ranged in age from seven to twelve and were determined to have significant support needs. The curriculum integrated cognition and literacy development. Lessons in the curriculum were scaffolded and students collaborated with each other throughout. Reading abilities were assessed using a curriculum-based reading assessment and at the end of the study, all students accurately answered reading comprehension questions and had increased ability to illustrate story points through drawing [79]. The third study, a single-case design study by Evmenova and Behrmann [85] included two postsecondary students with Down syndrome who were also hard of hearing. The intervention studied involved the use of video narration, two types of captions (highlighted text and picture/word-based captions, and interactive video searching for answers to questions. Factual and inferential comprehension of non-fiction videos increased through the use of adapted and interactive video clips [85]. The final study examined statewide alternate reading assessment and students who were DHH with cognitive disability [86]. Participants (N = 214) were measured on an alternative assessment that involved three literacy levels. Level A included concrete tasks related to personal experiences, matching, orienting, sorting, and receptive labeling. Level B included picture representation, limited content area vocabulary, and receptive and expressive labeling, sorting, classifying, and identifying function. Level C included symbolic representation using complex pictures, extensive content area vocabulary, and production of responses to open-ended questions. Participant data over a 6-year period were then analyzed. Findings indicated that even though depth of content knowledge increased and test items were, by design, increasingly complex across grade levels, students had a stable proficiency level. However, reading performance of the students with DWD was lower than that of other students with significant disabilities who took the alternative test. The authors posit that this finding supports a multidisciplinary approach to reading instruction that includes special educators, teachers of the deaf, and speech and language pathologists [86]. In summary, from the limited quantity of research specifically examining literacy development and children and youth who are DHH-ID, it is apparent that much more targeted research is needed if teachers are to effectively support the students in attaining crucial literacy skills including those that are early emergent.

6. d/Deaf/Hard of Hearing with Autism Spectrum Disorder

The Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5) defines autism spectrum disorder (ASD) as (a) persistent deficits in social communication and social interaction across multiple contexts including deficits in reciprocity, non-verbal communication, and deficits in maintaining and understanding relationships, (b) restricted repetitive patterns of behavior, interests, or activity including stereotyped behavior, insistence on sameness, highly restricted, fixated interests, and hyper-or hyporeactivity to sensory input, (c) symptoms must be present in early developmental period, (d) symptoms must cause clinically significant impairment in social, occupational or other areas of importance to current functioning, and (e) disturbances are not better explained by intellectual disability or global developmental delay [52]. In 2018, the Centers for Disease Control and Prevention (CDC) estimated that 1 in 59 children have ASD (1.7%) and it is four times more common in boys than girls [87]. The 2013 GRI count reported an incidence of 3% of ASD in the DHH student population [4] and similarly, other researchers have reported a higher incidence of ASD in the DHH population than is found in the population of hearing students [8,25,88] but there is not consensus in the literature of whether prevalence is actually higher [30]. However, it is also thought that the diagnosis of ASD is often overlooked in the population of students who are DHH [25,88] and diagnosis is complicated by the lack of standardized assessment instruments for ASD that are appropriate for the DHH population, difficulties in audiological assessment, overlapping characteristics of ASD and DHH, and diagnostic overshadowing [25,27,28,30,40,88]. The literature base in DHH-ASD is limited but includes identification and assessment [28,30,31,89], social interaction and behavior [90–93], speech development [13,32,41], sign language [94–96], AAC [97], family supports [98,99], and teacher perceptions [34,100]. A search of the research revealed no research specific to literature skills and DHH-ASD, therefore, what is included in the literacy section below is extrapolated from the literature on ASD.

6.1. Assessment and Identification

Early and intensive intervention is known to be beneficial in ameliorating the impacts of ASD on language, cognitive, and behavioral skills [101] and the average age of ASD diagnosis in hearing children is 56 months. However, in a retrospective review of children diagnosed with permanent hearing loss and ASD, Meinzen-Derr et al. [31] found an average of 66.5 months of age for ASD diagnosis in the DHH population even though many of the children in the study displayed ASD symptoms prior to diagnosis. The authors theorize that speech, language and social development differences were fully attributed to the hearing loss rather than as possible indicators of ASD (diagnostic overshadowing). Diagnosis is further complicated by the lack of diagnostic and screening instruments for ASD that are appropriate for the population of children who are DHH. In a review of 14 ASD assessment instruments, de Vaan et al. [28] found that most ASD assessment instruments did not have norms for children who are DHH and at least a quarter of the test items on the instruments were invalid for the population Therefore, much caution is needed in interpreting such existing instruments and there is an urgent need for instruments that are valid and reliable for use with the DHH population [28,40]. In addition, there is a need for assessors who have training and experience with both individuals with DHH and ASD and if possible, the assessment should be conducted by an evaluator who can communicate in the child's preferred communication modality [40]. Because it is uncommon for professionals to have expertise in both ASD and DHH, professional collaboration in conducting and interpreting the assessment results is necessary [28,40].

Although universal hearing screening has allowed for early identification of hearing loss in many infants, many children with ASD may be delayed in receiving a diagnosis of hearing impairment, and therefore, intervention is delayed [30,88]. In order to maximize auditory assessment results, Beers et al. [30] recommend that the audiological test setting and procedures be modified as children with ASD might become upset in the sound booth. Such suggested modifications include reading social stories about the assessment children prior to assessment, providing picture schedules outlining steps in the assessment, allowing parents or siblings to come into the booth, and allowing breaks as necessary. In addition, parents can introduce headphones at home to accustom the child to wearing them [30,32]. Because behavioral testing results in children with ASD tend to have low reliability, test batteries should include objective measures such as otoacoustic emissions testing and tone-burst auditory brainstem testing [30].

Although many of the symptoms of ASD overlap with DHH including delayed language, pragmatic language difficulties, delayed joint attention, and delayed theory of mind, there are subtle differences that can distinguish the two [25,40,89]. Children who are DHH and receive appropriate early intervention will have a language development trajectory that is more typical than is seen in children with ASD. In addition, delay of theory of mind is not as pronounced in children with DHH as it is in children with ASD. Further, children who are DHH alone also do not exhibit the difficulties maintaining social relationships that are seen in children with ASD. Patterns of stereotyped movements and rigidity in schedules are also not characteristic of children who are DHH without ASD [40,96]. Finally, in addition, to language and communication differences noted in the language section below, Kellogg et al. [89] found that children with ASD had delays in symbolic play not typically seen in children DHH without ASD and two of the three participating children in their study of children with DHH-ASD, lost skills over time.

6.2. Communication and Language Development

As detailed below, recent research on receptive access to communicion for children who DHH-ASD has primarily focused on effectiveness of cochlear implantion. A relatively large body of research on expressive communication has centered around the use of sign language and AAC.

6.2.1. Cochlear Implants

Historically, DHH with associated ASD was considered a contraindication for receiving a cochlear implant because of additional difficulties in language and communication. However,

increasing numbers of children who are DHH-ASD are now receiving implants. Results on its effectiveness in improving speech and language have been mixed as might be expected in a disorder as broad as ASD [31,102,103]. In a 2004 study, Donaldson et al. [103] found that children who could complete standardized receptive and expressive vocabulary tests did see improved scores, and children who could not complete the tests, improved in raw scores when compared with themselves post-cochlear implantation. Researchers have therefore concluded that open set speech may not be a primary goal but rather, becoming more engaged with the environment through sound, recognizing parental voices, and having increased safety and quality of life [31,103]. In addition, more knowledge of rehabilitation [102] and multi-system communication including speech, sign, and AAC post-implantation is needed [31].

6.2.2. Sign Language and AAC

Several recent studies have examined sign language and children who are DHH with ASD. The participating children in the studies were from families of Deaf signers and exposed to sign language from birth. Shield and colleagues conducted three studies looking at palm reversal, pronouns, and sign language echolalia in children who are DHH with ASD [96,104,105]. In the first study that examined palm reversal, Shield and Meier [105] found that the three participating children with DHH-ASD demonstrated errors across several sign parameters including location, handshape, movement, and palm orientation at a higher rate than would be expected in children who are DHH. More than half of the errors were in palm orientation including palm reversal of finger spelled words which was not seen in a comparison group of DHH students. Such reversals suggest that the children with DHH-ASD were producing the sign from their own perspective rather that reversing the palm. In the second study, Shield, Meier, and Tager-Flusberg [104] looked at the use of sign language pronouns by children who were DHH-ASD. Conclusions drawn from the study were that the children were able to point but avoided using pronouns in favor of names. Children with higher language skills were more likely to point. This finding is surprising giving that sign pronouns are often transparent and involve indexical point to self or others. The third study by Shield et al. [106] examined sign language echolalia in children who are DHH-ASD [96]. Echolalia or repeating the utterances of others has been shown to occur in the majority of verbal hearing children with ASD in early childhood. As language skills increase, echolalia decreases. Although the participating children in the Shield et al. study were older, 7 of the 17 children with DHH-ASD showed evidence of manual echolalia and the children who produced echolalic signs had lower receptive language than those who did not. Age, intelligence, and severity of ASD were not related to the echolalia, but rather the relationship appeared related to language comprehension. Results of this study indicated that the children were repeating signs with little comprehension as evidenced by frequency in errors of directionality (e.g., child imitated outward movement of adult's sign as outward movement from the child), reduplication (e.g., several repetitions of the sign for more), and timing (e.g., children repeated signs at the same time the adult was signing). Similar to children who are DHH alone, echolalia was seen less frequently in children with higher linguistic skills. Across the studies, significant differences between DHH children and DHH-ASD children suggest that palm reversals, paucity of pronoun use, and echolalia are characteristics of ASD rather than deafness.

Deaf individuals use their hands, face, eyes, and body to convey a wide range of emotions, but the hands and the face are particularly important [95]. In 2014, Denmark et al. [95] studied facial emotional comprehension of 12 individuals who were DHH and 13 individuals with DHH-ASD using videotape of an individual signing. In one videoed sequence, only the hands of the signer were visible, in the second, both hands and face were visible. They found that the DHH only group identified more emotions when both hands and face were visible, and in both conditions, the DHH only group correctly identified more emotions than the DHH-ASD group. A follow-up study by Denmark et al. [107] found that participating children with DHH-ASD produced fewer facial expressions when retelling a video story using sign language than did a peer group who were DHH only. Salient and common expressions such as surprise and disgust were shown by the children with DHH-ASD more than other expressions that involved more complex theory of mind. The findings of

these studies suggest that children with DHH-ASD might need particular intervention on how to interpret and convey emotional information [107]. In addition, support in developing theory of mind might be needed.

A final study by Bhat et al. [94] looked at differences between praxis performance and receptive language of native signing Deaf children with and without ASD. Praxis performance was observed in finger spelling using American Sign Language. The participants with DHH-ASD had lower receptive language scores than their peers who were Deaf alone. They also made more praxis errors and were slower in finger spelling. Praxis errors included errors in sequencing, body part use, orientation of fingers, and movement space. Errors involving proximal joints were also more frequent. The authors suggest that findings indicate a need for motor interventions and the use of complementary communication strategies such as visual pictures.

It is estimated that up to 30% of children with ASD have very limited expressive language [108] and the Shield et al. study found a similar percentage of non-verbal children with DHH-ASD [104]. Therefore, as suggested above, an additional communication modality is often necessary for many children with the dual diagnoses. Malandraki and Okalidou [97] described the usage of the Picture Exchange Communication System (PECS) [109] in a case study of a child with DHH-ASD. In the study, PECS was modified through replacement of verbal reinforcement, prompts, and trainer responses with multi-modal communication, gradual replacement of pictures with written words, and greatly increased intervention time. Preintervention, the ten-year-old participant had limited and non-functional communication. By the end of the four-month training period, the child was using the PECS communication book spontaneously. In addition, he demonstrated increases in play and interaction with peers, increased vocalizations, and decreased stereotypical movements. The use of such visual communication systems is supported by the research of Maljaars et al. [110] suggesting that individuals who are DHH with both ID and ASD have enhanced visual perception when compared to hearing individuals with ID. Further, individuals with low adaptive functioning and ASD, with and without hearing loss, have enhanced visual perception.

6.3. Literacy Development

Unfortunately, there is a dearth of research on children who are DHH-ASD and therefore, educators must look to research on children who are DHH, children who are d/Deaf with disabilities other than ASD, and the field of ASD. As noted previously, the population of children with DHH-ASD is very broad and there is not a one size fits all instructional model for the population. It is plausible, that depending on severity of ASD, strategies suggested in the DHH-LD section of the article could be helpful.

In looking at emerging literacy in particular, it is also likely that many of the evidence-based visual strategies used with hearing children with ASD for communication, behavior, and social development such as picture symbol systems [97,109], picture schedule systems [111–113], social stories [114–116], and video modeling [92,117,118] might have the concomitant effect of helping children with DHH-ASD along the path to literacy as they promote attention to pictures, tracking print and pictures from left to right, and story comprehension. However, the take-away point in this section is that an evidence-base is needed in order to teach effectively teach literacy and emergent literacy skills to children who are DHH-ASD.

7. Deafblindness

Deafblindness is defined in the Individuals with Disabilities Education Act (IDEA 2004) as "concomitant hearing and visual impairments, the combination of which causes such severe communication and other developmental and educational needs that they cannot be accommodated in special education programs solely for children with deafness or children with blindness" (IDEA 2004, p. 46756) [37].

Like deafness, deafblindness may be congenital or adventitious. Deafbindness reduces access to sensory information and incidental learning, while also impacting motor development, engagement in daily activities, concept development, communication, and language [39,119,120]. Vision and

hearing are the distance senses for learning; thus, the impact of having reduced functioning in both distance senses cannot be understood by adding up the impact of the hearing loss and vision loss; rather the impact is sometimes described as multiplicative [121].

According to the 2017 National Child Count of Children and Youth who are Deaf-Blind [122] that presents the census data from state deafblind technical assistance projects, there are 10,000 children and youth, ages 0–21 years, identified as being deafblind in the United States. Of these, 4590 have either a moderate-severe, severe, or profound hearing loss and 1098 have cochlear implants. Low vision is the most common category for visual impairment, with 3204 identified, and another 2449 being identified as legally blind. An additional 1050 children have either light perception only or total blindness. The vast majority of children identified have some functional hearing and/or vision to support the development of language and literacy. The most commonly identified cause of deafblindness is complications of prematurity, followed by hereditary syndromes. Additional disabilities complicate learning with 87% of the children and youth having at least one additional disability and 43% having four or more additional disabilities. According to this census, the most common additional disabilities are speech-language impairments, cognitive impairments, and orthopedic or physical impairments. The racial and ethnic diversity mirrors that of the U.S. population. In short, this is a small population of great heterogeneity.

7.1. Assessment and Identification

Identification of deafblindness is dependent on medical and school professionals understanding the IDEA definition of deafblindness. Hearing loss is usually identified early through newborn screening [1]; however, early identification of deafblindness continues to be a problem, with half of the children on the national census identified after the age of five years [122]. Children who are deafblind may not respond to behaviorally based vision and hearing testing. In many cases, they will require an electrophysiological test, such as the auditory brain stem response test, to identify auditory thresholds [120]. They will require an extensive set of vision tests to identify their acuity, visual field, and the potential presence of cortical visual impairment. Additionally, they require conduction of both functional hearing and vision assessments across familiar daily activities and environments for the purpose of determining how they use their hearing and vision in daily life. Children who are deafblind need to be identified as early as possible to avoid more severe developmental consequences. They must then be served by professionals who understand the interactive effects of their complex disabilities. These children will benefit from early intervention programs that emphasize the use of residual vision and hearing, and the development of tactual skills to support the development of concepts, communication, language, and literacy.

7.2. Communication and Language

The field of deafblindness generally follows a developmental and child-guided approach to communication intervention. The van Dijk approach to assessment and communication intervention has been adopted internationally and is intended to support the development of presymbolic to early linguistic communicators. Van Dijk's approach emphasizes the establishment of trust, responsiveness to the child's communicative attempts, sharing of emotional states, communicating using the child's expressive forms, building different types of conversations based on the child's interests, coactive techniques (including coactive movement routines), sequential memory strategies, the use of drawings, and the achievement of symbolic understanding and expression [123–126]. Early conversations are sensitive to the child's emotional state and need for safety, focus on the child's preferred topics, integrate the use of movement and objects, and establish turn taking [125,127].

Communication and language intervention are based on thorough assessment. The Communication Matrix [128] measures seven developmental levels, from preintentional behavior to early language, including the growth from unconventional to conventional communication. The assessment results in a profile that supports the educational team to plan individualized interventions [129]. Comprehensive communication programming will address expressive and

receptive forms, intents and functions, content or vocabulary, and context (including the physical environment, activities and routines, communication partner skills, and pragmatics [46].

In calculating the level of evidence for research-based practices in visual impairments, DHH, and deafblindness, Ferrell, Bruce, and Luckner [130] found that research on communication was an area of relative strength in deafblindness. The reader is referred to Ferrell et al. for a complete explanation of how each evidence level was defined. Research evidence for the child-guided approach and its role in promoting conversation is at the limited level, with the exception of its application to improving communication partner skills, which is now at the moderate level due to studies by Janssen and colleagues [46]. There is a moderate level of evidence for applying principles of systematic instruction for increasing rate and the variety of communicative intents/functions, and vocabulary [130]. There is also a moderate level of evidence for the role of tactile approaches and strategies (see [131,132]), including the use of touch cues, tactual materials, attending to the use of the hands in learning, tangible representations or symbols, in improving communication [133].

7.2.1. Cochlear Implants

Auditory perception, communication, and speech are valued outcomes of cochlear implantation [134]. There are differences in the outcomes valued by parents of children who are DHH and parents of children who are deafblind. Across studies, parents of children who are deafblind report that they value non-speech outcomes, such as evidence of "improvements in attention; interactions with objects; listening, which may break down isolation and enhance engagement; responsiveness; increased awareness of environmental sounds, which may improve safety; and increased vocalizations" [130] (Evidence-based practices, p.68). The outcomes of cochlear implantation in children who are deafblind vary greatly; however, there are etiological outcomes patterns and other predictor variables that should be shared with parents when they are considering the possibility of cochlear implantation [130]. Parents need direct instruction about the potential benefits of cochlear implants, how to conduct daily checks on their functioning, and instructional strategies to support their use [135].

7.2.2. Augmentative and Alternative Communication (AAC)

Children and young adults who are deafblind use a range of expressive and receptive communication forms, including objects, partial objects, textures, photographs, line drawings, gestures (invented and conventional), signs, sign language, spoken language, and speech generating devices. The educational team will consider the learner's characteristics, including vision, hearing, tactual skills, and motor skills when selecting receptive and expressive forms. Many children who are deafblind use tangible representations or symbols to communicate. Tangible representations are iconic, meaning they share a physical resemblance to the referent (what they represent). This shared resemblance may be visual or tactual. Tangible representations include photographs, pictures, objects, and partial objects. Rowland and Schweigert [136,137] developed interventions that demonstrated how to use tangible representations in daily schedules and within daily routines. Object cues are object representations that are used receptively; whereas touch cues are tactual hints about something that is about to occur (such as gently touching the face with a washcloth, just prior to washing).

Sign or sign language is another frequently used form of communication by individuals who are deafblind. Child characteristics, such as vision, size of hands, and learning style impact how signs are introduced. The adult may model the sign or present the sign (for the child to see and touch) and then shape the sign on the child's hands. The adult may be positioned face-to-face or behind the child (in which case the child may be able to perceive how the signs are formed more easily [138]). Children with Usher syndrome may change how they respond to receptive sign language, requiring tracking of the hands or differences in signing space. Use of signs involves understanding their handshape, orientation of the sign to the body, movement of the sign, sense of touch if using tactual sign, and balance [139].

Language and culture are intertwined. Souria [140] explained that the cultural sign language of individuals who are deafblind is not static, but is rather a "language in the making" (p. 21) because it is not learned through sharing language with others who are deafblind. Instead it borrows from other languages, such as modifying signs used by the Deaf. Sign language for individuals who are deafblind is co-constructed with their communication partners and includes signs that were invented by the individual who is deafblind.

7.3. Literacy

The field of deafblindness has adopted an expansive definition of literacy to include communication and the application of low and high technologies to support interactions and conversation [46]. "A new, more inclusive view of literacy includes all learners [141,142], begins at birth [143], and recognizes that the materials and media of literacy differ across learners" [130] (Evidence-based practices, p. 76). Emerson and Bishop [144] referred to literacy that is experienced though technology as the new literacy. Miles [142] discussed the social aspects of literacy, and the benefits to both the child who is deafblind and to society which will benefit from knowing more about the thoughts and experiences of individuals who are deafblind. The state of evidence for literacy practices in the field of deafblindness is at the emerging level, relying on professional literature with few intervention studies to inform practice [130].

Literacy is grounded in a rich array of experiences involving many opportunities for hands-on learning [142]. All children benefit from literacy rich environments in the home and school. For children who are deafblind, this would include books in print and braille, auditory books, tactile books, accessible labeling of literacy materials, interactive software, and adapted commercial books [145]). In a 2019 study of emergent literacy materials and strategies in classrooms with students who are deafblind or with multiple disabilities and visual impairment, McKenzie found that teachers most often provided lessons on the daily news, morning circle, read alouds, and schedules. Supported writing activities, shared reading, and choice-making were offered less frequently. She suggested the need for literacy centers, accessible labeling, increased read alouds, opportunities to scribble, increased IEP objectives on literacy, and a learning media assessment to determine the best medium for presenting and producing literacy to each child [145].

Literacy approaches in deafblindness are both individualized and personalized [146]. Individualization includes the selection of materials, approaches, and technologies that match the strengths and needs of each child. Personalization includes readings and writings about the child's preferences and his experiences, thus greatly reducing memory load. Literacy lessons in the field of deafblindness include daily schedules and calendar systems, story boxes, experience stories, journals, choice-making opportunities [133], and shared reading. Teacher-made materials will be important to personalizing the literacy program and to ensure that text is at the correct level [147].

The daily schedule, which may be expressed in multiple communication forms, provides opportunities for the child to learn about sequence, left to right displays, representations of daily events, locations, and people [148,149] while providing enhanced predictability to the child's life. The daily schedule is far more than a tool to support transition between activities. Before and after each represented activity, the child should engage with the schedule and share a conversation about each activity. In this way the child practices the representations for common daily activities, learns key vocabulary, and shares conversations rooted in daily events. Often representations in the daily schedule appear in two or more communication or literacy forms, such as objects with print and braille labels.

Story boxes are collections of objects that relate to the content of a personalized or commercially produced book. They allow the child an opportunity for additional tactual information about the text. Closely related, experience books which are also known as memory books, are texts about something the child has experienced, such as a trip to the zoo. The representations must be salient, e.g., capture what was most significant to the child about that particular experience (as opposed to what hearing and sighted people found to be salient). These personalized forms of literacy are co-constructed with

the child, often include labeling in print and braille, and are enjoyed with others through with repeated shared readings [133,150].

Interactive journals, also known as home-school books, allow an adult at home or school to engage in shared reading with the child who is deafblind about an event that occurred in the other environment and associated vocabulary [147]. This is important to building distancing across time and space, a necessary milestone to the development of language [151]. Bruce and Conlon [152] described a school-home journal with pages that included an object representation, an auditory device with a simple message the child could repeat, and print and braille labels. Like experience books, interactive journals are a form of personalized literacy that reduce cognitive load.

Most classrooms provide daily opportunities for children to make choices. The challenge is to ensure that choice-making is authentic, e.g., the child actually selects a preferred option. This requires that the child has preferences, has experiences that are referenced by the representations, is presented with accessible options, comprehends the specific representations for the presented options, and has a clear indicating response [133,153].

Children who are deafblind have far fewer opportunities to observe why people engage in writing due to reduced observation. Therefore, they need adults who will communicate with them about the purposes of daily writing activities and include them in creating drawings and writings. Early expressive writing experiences will include writing one's name and the names of a few other people important in the child's life, words on the daily schedule, and words that represent preferences [147]. Later, the child may engage in writing about personal experiences and may co-construct journals and other literacy materials.

Some children who are deafblind will become braille readers and writers. Steinman, LeJeune, and Kimbrough [154] presented Chall's [155] reader-based, or top-down, developmental approach to print and braille literacy. Braille reading is more cognitively demanding than print reading because braille is cognitively processed one cell at a time; whereas multiple print characters can be processed at the same time [154]. Thus, it takes a significant number of years to completely master the braille literacy code, and longer for the Nemeth code for mathematics. The American Printing House for the Blind is a resource for pre-braille curricula and materials, as well as specialized curricula for the instruction of braille literacy. Additional literacy resources include the Paths to Literacy project (www.pathstoliteracy) and Project Salute (www.projectsalute.net).

8. Recommendations for Practice and Research

8.1. Recommendations for Practice

Despite the heterogeneity of the population of students who are DWD, there are several implications for practice across the disabilities. Due to the importance of early identification of both hearing loss and disabilities, practitioners must be aware of current assessment practices and be knowledgeable about learner characteristics that should evoke a referral to initiate an evaluation for a disability. Communication and language development are frequently delayed in children with DWD and may take varied forms depending on the needs of the children. However, it is essential to quality of life and learning. Therefore, teachers should be prepared to support communication development across forms and in children with a variety of learning and physical needs. Further, assistive technologies must be selected based on the unique needs of each DWD learner and a plan developed to support their implementation across environments. Depending on severity of disabilities, literacy development may need to be seen in a broad context with the understanding that text reading and literacy may not always be synonymous. Teachers must be skillful in the implementation of both child-guided and systematic instructional approaches, and of common literacy lessons such as calendar systems, shared reading, and experience books. In addition, practitioners should be knowledgeable about major approaches to traditional reading and writing (text-based, subject-based, and compensatory-interactive), the relative merits of each, and the nuances of implementing each approach with a child who is deaf with an identified disability. This is especially true given that most approaches used with children who are deaf are visually-based, but

vision may not be functional or a preferred learning modality for some learners who are DWD. Finally, it is critical that teachers appreciate that the effects of being deaf with a disability are best understood as being complex and interactive. The complex needs of learners who are DWD call on teams to engage in interprofessional collaborative practices.

8.2. Recommendations for Research

Although there is a growing body of literature about assessment for identification of a disability, there is a need for research on how to assess learners who are DHH with specific disabilities to determine individualized instructional programming and provide ongoing measurement of learning progress. While the research evidence is relatively strong in the area of communication for learners who are DHH with ASD or who are deafblind, there is an ongoing need for additional research on communication and language in learners who are DHH with ID or LD. There is a dire need for research on teaching traditional literacies (reading, writing, and spelling) to children with each of the disabilities that may co-occur with deafness. This research is critical to the development of literacy curricula. Further, there is a continued need to understand how technologies can support learners who are DWD in receptive and expressive communication across communication forms, and in traditional literacy. Finally, research teams will need to continue to document how outcomes associated with the implementation of assistive technologies vary across learners who are DWD and are different from the outcomes experienced by learners who are DHH.

9. Conclusions

Students who DWD are diverse, but because of the complex and interactive nature of their disabilities, share a need for interventions that specifically target communication and literacy. Evidence-based interventions from the different disability areas may be applicable and effective, but their use must be tempered by consideration of sensory and learning characteristics of individuals who are DWD. As seen in the sections above, the research base on DWD is increasing, but there is much to be known in terms of prevalence, identification, assessment, and evidence-based practices in communication and literacy across the disabilities. Moreover, there is a strong need for teachers of the DHH to be prepared to teach DWD learners in order for students who are DWD to achieve maximization of potential.

Author Contributions: Catherine Nelson and Susan M. Bruce contribute to review of research and writing of article

Funding: This research received no external funding.

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

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