Memory Profiles of Down, Williams, and Fragile X Syndromes: Implications for Reading Development

Frances A. Conners, PhD, Marie S. Moore, MA, Susan J. Loveall, MA, Edward C. Merrill, PhD

ABSTRACT: The purpose of this review was to understand the types of memory impairments that are associated with intellectual disability (ID, formerly called mental retardation) and the implications of these impairments for reading development. Specifically, studies on working memory, delayed memory and learning, and semantic/conceptual memory in Down syndrome, Williams syndrome, and fragile X syndrome were examined. A distinct memory profile emerged for each of the 3 etiologies of ID. Memory profiles are discussed in relation to strengths and weaknesses in reading skills in these three etiologies. We suggest that reading instruction be designed to capitalize on relatively stronger memory skills while providing extra support for especially challenging aspects of reading.

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Memory is one of the most important aspects of cognition affected by intellectual disability (ID, formerly called mental retardation). It is important not only because of its role in everyday activity but also because of its role in the development of higher-level cognitive and academic functions, such as reasoning, mathematic computation, and reading. Although debate continues on whether memory is multifactorial or unitary, there is a wide variety of memory measures that reflect different sets of memory processes (e.g., storage, retrieval, updating, rehearsal, and activation). In ID, various memory processes may be differentially impaired, especially across specific etiology groups. Further, various memory processes are differentially important to reading development at different stages. The purpose of this article is to review the research on memory in ID in relation to the known involvement of memory in reading development, and to consider how and when children with ID may experience the greatest difficulties with reading. As memory profiles differ by etiology, relative strengths and weaknesses in reading may differ by etiology as well; thus, in this article, we focus on 3 etiologies of ID—Down syndrome (DS), Williams syndrome (WS), and fragile X syndrome (FXS).

MEMORY: MULTIFACTORIAL OR UNITARY?

Perhaps, the best known model of human memory is the Atkinson-Shiffrin[1,2] information processing model, a multifactorial model. This model, introduced in 1968, emphasized separate memory stores, differentiated in terms of how long each held information. These were sensory memory (SM), short-term memory (STM), and long-term memory (LTM). According to the model, these stores work together as an information processing system. Information from the environment flows into the system to be encoded first into SM for analysis of sensory properties. From there, attended information is forwarded to STM. In STM, a small amount of information can be held temporarily and then selected information can be relayed to LTM for more permanent storage. STM and LTM work together to retrieve information from LTM when needed. Although not the first multifactorial model of human memory, Atkinson and Shiffrin's has been highly influential, both in promoting the general concept of a multipart memory system and in delineating a distinction between STM and LTM.

In the Atkinson-Shiffrin model, STM is very limited both in time (i.e., less than a minute) and in capacity (holding only $\sim 7 \pm 2$ items at a time),[3] although these limitations can be stretched by using chunking and rehearsal strategies. Information held in STM may be coded in terms of its verbal or visuospatial features.[4,5] In contrast to STM, LTM can hold vast quantities of information over long periods of time, possibly indefinitely. Some of this information is experiential or episodic and can be verbal or visuospatial (e.g., a scene or a conversation that one has experienced). Other information in LTM is conceptual or semantic (e.g., word meanings or complex ideas that one has learned or acquired over time). LTM is conceptualized as an extensive network of connections among bits of stored information. Within this network, there are large and small structures or sets of connections that allow a person to retrieve sets of related information together.

From the Department of Psychology, University of Alabama, Tuscaloosa, AL.
Received February 2010; accepted February 2011.
Address for reprints: Frances A. Conners, PhD, Box 870348, Tuscaloosa, AL 35487-0348; e-mail fconners@bama.ua.edu.

In addition to STM and LTM, working memory (WM) is an important construct to multifactorial memory models. WM is the part of the memory system that holds and actively processes information.[6,7] This includes comparing, judging, computing, relating, finding, or otherwise managing information. A well-known model of WM is Baddeley's multicomponent model of WM,[7–9] which includes a central executive component that guides the system and 3 subsidiary components that have specialized roles. The phonological loop and visuospatial sketchpad are specialized for processing phonological/verbal and visuospatial information, respectively, and the episodic buffer binds phonological and visuospatial information into a single episodic representation.[10] Although there is some evidence that STM and WM are distinct memory constructs,[11] growing evidence suggests that traditional STM and WM tasks draw from the same set of underlying processes[12–15] and do not represent separate memory constructs.

Several recent memory models have challenged the multifactorial standard, reinterpreting well-known empirical findings without the need for separate memory components.[16–21] For example, Cowan's[17] embedded processing model suggests that what is referred to as WM is simply the part of memory that is currently activated. The most highly activated information is in the focus of attention, which is limited in capacity and can only accommodate a small amount of information. Thus, it is the limitation of the focus of attention rather than a particular memory store that results in limited immediate memory performance. Nairne's[20] feature model suggests that to remember, we activate a set of cues from previous processing records to reconstruct the earlier experience or information. Following a choice rule, the cues are used to select candidate information that is stored in memory. It matters little how recently the information being retrieved was encountered (i.e., "short-term" or "long-term"). What is important is how well the set of cues uniquely corresponds to the earlier experience or information. Memory limitations arise because new information constantly overwrites (or interferes with) similar aspects of earlier information, making the unique matching of retrieval cues with memory records more difficult. In our analysis of strengths and weaknesses in memory associated with reading skills in children with ID, we do not assume that memory is multifactorial or that it is unitary. We organize the empirical findings in terms of related memory skills.

In the following section, we discuss the memory skills that are known to be related to early reading skills. Later in this article, we ask whether these memory skills are relative strengths or weaknesses in DS, WS, and FXS, and how reading development might be affected by memory profiles associated with these 3 syndromes.

READING AND ITS RELATION TO MEMORY

The ultimate goal of reading is comprehension of print, which involves forming a meaningful mental representation of text.[22,23] Successful reading comprehension depends on the development and integration of a wide variety of skills that allow the reader to accurately identify each word, access the meaning of the words, process combinations of words as they occur in phrases and sentences, and use background knowledge and context to interpret the sentences.

The Simple View of Reading

To understand the most basic component processes of reading, Gough and Tunmer[24] proposed the Simple View of reading, a model that is widely accepted by reading researchers.[25–27] According to the Simple View, reading comprehension has 2 main components—word recognition and language comprehension. Word recognition refers to word-level reading by recognizing whole or parts of words or by sounding out; language comprehension refers to understanding the meaning of words, phrases, sentences, and longer communications. Without word recognition, one would not be able to read the words on the page, and without language comprehension, one would not be able to understand their meaning. Thus, a combination of the 2 skills is necessary to achieve reading comprehension.

According to the Simple View, the relative importance of word recognition and language comprehension to skilled reading behavior changes with development.[28–30] Early in reading development, word recognition is more important than language comprehension because the primary goal is to accurately identify words. As reading skills develop and words are read more rapidly, language comprehension becomes the more important component. Memory is important in the Simple View of reading because it is required to succeed both in word recognition and comprehension, as well as in integrating these 2 skills to produce reading comprehension.

Word Recognition

Early in the acquisition of word recognition skills, children learn the visual patterns of common words to which they have repeated exposure (e.g., stop, exit, and cat).[31] Without using an alphabetic code, they begin to recognize certain words by their visual features. Ehri[31] has called this the prealphabetic phase of reading and this is related to visual memory. For example, Stuart et al[32] found that among prealphabetic 5 year olds, immediate and 10-second delayed visual memory correlated strongly with sight word learning; however, among 5 year olds with some alphabetic skills, there was no correlation. Thus, visual memory may be particularly important to word recognition before children know letter-sound associations. In addition to visual memory, associative learning is important to word recognition in early reading development. Nilsen and Bourassa[33] found that for children who had alphabetic knowledge but could not yet read alphabetically, the ability to learn associations between black-and-white designs and spoken nonwords was very closely related to the rate of learning new reading words.

Eventually, children learn to read new unfamiliar words by matching letters with sounds in sequence or sounding out.[34,35] Alphabetic, or phonological, reading requires an awareness that words are made up of phonological segments such as syllables and phonemes. It also requires adequate memory to hold each sound temporarily while decoding the entire word. Strattman and Hodson[36] found that phonological awareness and verbal WM both contributed uniquely to nonword reading in second graders, after the influence of nonverbal ability was removed.

As children gain experience in reading words, they begin to associate common letter combinations (or orthographic features) with more complex phonological mappings. They no longer have to rely as heavily on phonological decoding; instead, they can quickly read words using their orthographic features.[31,37] As readers become more skilled, they build up their orthographic knowledge and are able to access and name more and more words very quickly. Phonological awareness and associative learning are still important in supporting word reading,[38] as is verbal WM.[11] Windfuhr and Snowling[38] found that for children aged 7 to 11 years, in addition to phonological awareness, the ability to learn associations between abstract shapes and spoken nonwords contributed significantly to both word and nonword reading after age and IQ were controlled. Kail and Hall[11] found that for children aged 8 to 13 years, a verbal WM composite was significantly related to word recognition, with measures of simultaneous processing and storage more closely related than measures of immediate memory. Rapid automatic naming (e.g., of letters, digits, colors, or objects), believed to reflect speed of retrieval of phonological information from memory, is also related to word reading, especially the pause time between names.[39–42] Although the exact reason for this relation is under debate, one possibility is that rapid naming speed reflects the strength of association between visual and phonological forms, which is important to word reading.

Reading Comprehension

As children become skilled at word recognition, they begin to focus more on comprehension in reading. Reading comprehension requires building and maintaining a mental representation of the text while concurrently decoding additional words of the text.[22–23] Research suggests that strength in reading comprehension is related to WM, particularly verbal WM.[12,43–48] One possible reason for this link is that good WM skills allow one to suppress irrelevant information from being processed, while allowing relevant information to be processed. The ability to suppress irrelevant information may help in the building of a mental representation that is coherent.[49–53] In addition to WM, semantic or conceptual memory aids reading comprehension. This is the repository of knowledge that is needed for the interpretation and disambiguation of text.[22,23] Many studies have shown that especially for expository text, prior knowledge related to the text enhances text comprehension.[54–56] This is presumably because prior knowledge allows for generation of accurate inferences.[57,58] Thus, the quality and quantity of information in semantic memory—and its accessibility—is important to reading comprehension.

In sum, word recognition is typically supported by visual memory (in the prealphabetic phase), associative learning, verbal WM, and rapid phonological retrieval. Reading comprehension is typically supported by verbal WM and semantic/conceptual knowledge. Next, we consider the memory profiles associated with DS, WS, and FXS to determine whether there are clear strengths or weaknesses in these aspects of memory. Also, for each syndrome, we discuss implications for reading development.

ID, MEMORY, AND READING: 3 SYNDROMES

Although memory difficulties are common to all individuals with ID, there are clear memory profiles associated with specific etiologies. In this section, we discuss memory and reading profiles associated with DS, WS, and FXS. The memory findings are organized into 3 sections—aspects of WM, delayed recall and learning, and semantic/conceptual memory. Although we review these aspects of memory broadly to characterize a memory profile for each of the syndromes, we are particularly interested in the memory processes that are most important to reading development (visual memory, associative learning, verbal WM, rapid phonological retrieval, and semantic/conceptual knowledge). Thus, toward the end of each section, we summarize these aspects of the memory profile and discuss how they relate to reading skills. The most researched of the 3 etiologies is DS, followed by WS, and then FXS. As we are concerned with relative strengths and weaknesses, we include only studies that benchmarked memory abilities in some way to participants' developmental level. Most commonly, this has been done by comparing performance of the target syndrome group with performance of a typically developing group matched on a mental age or ability measure (e.g., receptive vocabulary age, overall mental age, and nonverbal raw score). As we are concerned with reading development, our review focuses on school-age children and adolescents. Finally, because we are interested in how memory profiles relate to etiology, we highlight cross-etiology comparisons to address the issue of etiology specificity.

Down Syndrome

DS is the most common genetic syndrome resulting in ID, occurring in 1 of 733 births.[59] People with DS usually have a triplication of chromosome 21 instead of the usual 2. Common physical symptoms include short stature, hearing impairment, congenital heart disease, and distinct craniofacial features.[60] Among the craniofacial features are epicanthal folds, flat nasal bridge, and small jaw and oral cavity.[61–66] The degree of cognitive impairment associated with DS ranges from mild to severe, although moderate impairment is most common. Language production and grammar comprehension are typically extremely limited as is immediate verbal memory, and visual skills are relatively stronger. Brain regions that are affected in DS include the medial temporal lobe (including the hippocampus), the frontal cortex, and the cerebellum.[67–70]

Aspects of Working Memory

Several reviews have already established very clearly that young people with DS are poor in tasks measuring immediate memory for verbal material (e.g., digits and words) relative to typically developing children of similar developmental level.[71–76] Most commonly, DS and typically developing groups are matched on general cognitive ability, nonverbal ability, or receptive vocabulary, avoiding those language areas that are severely impaired in DS, such as syntax and expressive language. This pattern is also usually present for nonword repetition[77–80] and sentence repetition.[77,81] As these reviews note, poor performance on these tasks is not attributable to hearing or speech articulation difficulties that often co-occur with DS.[82–85] Further, this impairment is clearly etiology specific. Participants with DS perform more poorly on immediate verbal memory tasks when compared with mixed-etiology ID controls[83,84,86–90] or controls with other specific etiologies.[77,90–92] One exception is FXS, in which performance can be just as poor as in DS.[80,93–95] Researchers have examined possible underlying problems in rehearsal,[81,87,96–99] phonological sensitivity,[99–101] and linguistic support,[100,101] but none of these has fully explained poor immediate verbal memory performance of individuals with DS.[71]

Strikingly, immediate memory is much better in the visuospatial domain than the verbal domain in DS and is usually consistent with developmental level.[75] This is true for the classic Corsi block task, which measures memory for spatial sequences,[82–84,102–107] and for a variety of other immediate visuospatial memory tasks.[93,102,108] Researchers making etiology group comparisons have found that participants with DS perform similarly to or better than participants with ID, WS, and FXS.[89–92,95,106] One exception is that participants with DS seem to perform below their developmental level (although similar to participants with WS or FXS) on immediate recall of spatial locations simultaneously presented, at least when matched on receptive vocabulary.[80,93,103] These tasks require participants to look at an array of objects or squares for 5 to 10 seconds and then point to where in the display each object or square had appeared.

In addition to having problems with verbal immediate memory, young people with DS have problems on tasks requiring simultaneous processing and storage, such as backward digit span and backward word span.[80,104,106] These tasks require keeping some information in memory while manipulating other information. In addition, Lanfranchi et al[104] found that when matched on logical operations, participants with DS performed more poorly than controls on 2 other tasks that require simultaneous processing and storage—a selective word recall task (remember the first word of each list) and a dual request word recall task (remember the first word of each list and tap when a target word occurs). These researchers showed that as the control component of the task increased, so did the magnitude of the group difference. Participants with DS may perform below IQ-matched participants with general ID,[106] but their difficulty in simultaneous processing and storage is not as extreme as those with FXS.[80,95] A similar degree of impairment in simultaneous processing and storage is present in DS in the visuospatial and verbal domains.[103–107]

Delayed Memory and Learning

Young people with DS also have very clear and consistently documented difficulties on delayed memory and learning tasks, in both verbal and visuospatial domains. Several studies showed that when asked to remember words they studied 10 to 15 minutes earlier, participants with DS performed below their general cognitive ability level.[109–112] Other studies showed that when given repeated exposure to learn word lists, young participants with DS also performed below their general cognitive ability level.[105,109,110,112,113]

In the visuospatial domain, the pattern is similar. Compared to controls matched on general cognitive ability, young people with DS have significant difficulty with delayed memory for abstract patterns,[105,107,109,114] object pictures,[115] locations of abstract patterns and object pictures,[105,107] previously fragmented pictures,[110–112] and grid patterns.[107] Visuospatial learning is below general cognitive ability as well. Research has shown that young people with DS have great difficulty learning to use landmarks to navigate. a virtual water maze,[105] learning block location sequences,[112] and learning to identify object pictures.[115] Although the available data on associative learning suggest an impairment in DS,[105,107,116] these data are based on a visual-spatial association task, and the impairment could be linked to problems in delayed visuospatial memory rather than associative learning per se. There is little evidence yet to judge whether difficulties in delayed memory and learning seen in DS are etiology specific, but available data suggest that this is a possibility.[109–111,115] For example, 1 study showed that delayed verbal memory impairments were present for participants with DS but not WS.[110,111]

Semantic/Conceptual Memory

In DS some aspects of semantic memory are particularly poor, while others are relatively stronger. One way of assessing semantic memory is to ask children to free recall a story that was read or told to them earlier. This measure reflects children's ability to understand new information, store it accurately, and recall it in an organized way. Children and adolescents with DS recall fewer details than peers of the same receptive vocabulary level,[80] and they recall fewer gist units,[117] propositions,[118] and informative units[119] than peers of the same general cognitive level. However, they seem to remember content words[119] and novel words[118] at a rate consistent with their general cognitive level. The difficulty in recalling meaningful units may be etiology specific, because participants with DS have performed worse than participants with ID when matched on general cognitive ability level[109] and marginally worse than participants with FXS when matched on receptive vocabulary level.[80] It is difficult to say, however, whether these results reflect memory problems above and beyond language limitations; these studies did not control for group differences in grammatical or expressive language skills.

To measure efficiency in accessing information in semantic memory without the influence of language comprehension, several researchers have used speeded picture naming tasks or semantic fluency tasks. Speeded picture naming tasks require accessing the meaning of a picture, associating it with its name, and producing the name. In 2 studies,[120,121] participants with DS were as fast at picture naming as controls matched on general cognitive level, although in one of these studies, participants with DS made more visual-semantic errors (e.g., producing the name of a visually similar item). The picture naming findings are similar to findings on rapid naming of digits and letters,[120] which presumably depend less on semantic memory and more on well-learned visual-phonological associations.

In semantic fluency tasks, participants call out as many category exemplars as possible in a designated time period (e.g., 60 seconds). This task requires fast access to semantically similar concepts in memory and reflects the richness of one's category knowledge. Most studies showed that young people with DS produce as many category exemplars in semantic fluency tasks as their typically developing controls, whether matched on general cognitive ability level[90,105,122] or on receptive vocabulary level.[108,123] Thus, at this time, there is no clear evidence of specific impairment in retrieval of information from semantic memory.

Summary of DS Memory Profile

Children with DS have many severe memory limitations. The memory profile in DS includes weaknesses relative to developmental level in immediate verbal memory, simultaneous processing and storage, delayed memory and learning, and gist recall. It includes relative strengths in immediate visuospatial memory and rapid retrieval of semantic and phonological information. Critical for reading development, children with DS are poor at verbal WM and use of semantic/conceptual knowledge for story recall. They are relatively better at immediate visual memory, retrieval of phonological information, and retrieval of semantic/conceptual information.

Implications for Reading Development

Due to impairments in WM, delayed memory, and learning, as well as in some aspects of semantic/conceptual memory, it might be expected that children with DS would be poor in word recognition and in reading comprehension. Surprisingly, several studies have shown that word recognition can be at or even above nonverbal ability level or receptive vocabulary level in DS.[124–129] This is surprising given that verbal WM and phonological awareness[130] are so poor in DS. Further, their ability to phonologically recode (sound out) words is clearly not to the level of their word recognition.[128,131–133] Thus, it is plausible that children with DS use their relatively strong immediate visual memory and phonological retrieval skills to compensate for their weaknesses in verbal WM and phonological awareness skills, as several researchers have suggested.[126,134–136] Two studies have shown that when matched on reading level to typically developing children, young people with DS perform similarly on irregular word reading (which relies on visual-orthographic skills) but more poorly on nonword reading (which relies on phonological recoding skills).[128,133] In other words, visual-orthographic skills were consistent with word recognition level, whereas phonological recoding skills were comparatively weak.

As expected from the poor verbal WM and poor story recall in DS, reading comprehension is extremely poor. When matched with typically developing children or slow readers who are reading at the same level, children with DS perform more poorly in reading comprehension[133,137] and make less progress over time.[138] A recent eye-tracking pilot study[137] suggested that compared with controls, children with DS may tend toward more regressive saccades (backward eye movements) relative to forward saccades during text reading, indicating more difficulty with sentence processing. These participants needed to read complex sentences 2 times before showing the usual "wrap-up" effect (long fixation on the last word of a sentence), which was shown by typically developing readings after a single reading.

Williams Syndrome

WS is caused by a microdeletion on the long arm of chromosome 7 affecting >20 genes. Although the literature often cites 1 in 20,000 as the prevalence of WS, a recent population-based study suggests 1 in 7,500.[139] Characteristic features of WS include facial dysmorphology (wide mouth, upturned nose, and puffy eyes), heart and blood vessel abnormalities, and mild to moderate ID.[140] Also, WS is associated with severe difficulty with visuospatial processing, overly friendly personal style, and interest in music.[141–143] On the surface, people with WS seem to have good language skills due to their verbosity. However, language is generally in line with nonverbal ability, with relative strength in concrete vocabulary and relative weakness in abstract vocabulary and spatial language.[144,145] Brain regions affected in WS include the midbrain, thalamus and basal ganglia, occipital lobe and temporal lobe.[146]

Aspects of Working Memory

Although in DS immediate memory is much worse in the verbal than the visuospatial domain, the reverse is true for WS. Immediate verbal memory is generally similar or only slightly worse than what would be expected based on developmental level. This is true whether matched on receptive vocabulary,[147–149] nonverbal ability,[150–153] general mental age,[77,90] or when compared with one's own general ability level.[154,155] In addition, youth with WS perform similarly to ID controls[91,113,147] except for those with DS, whom they outperform.[77,90,92,112,156] Furthermore, phonological similarity, word length, frequency, lexicality, concreteness, primacy, and recency have the same effects on immediate verbal memory in young people with WS as in typically developing peers or peers who have ID.[147,152,157,158] In contrast, young people with WS perform much more poorly than typically developing children on visuospatial memory tasks such as the Corsi span task, whether matched on receptive vocabulary, nonverbal ability, or general mental age.[91,150,153,159–163] However, this impairment seems to be related primarily to spatial as opposed to visual memory.[161,162,164] It is not accounted for entirely by general difficulty in visual perception or spatial ability that are characteristic of WS[162,165] and seems to be etiology specific.[91,156,159,162]

Very few studies have examined simultaneous processing and storage in relation to developmental level in WS. However, a pattern of poor performance on spatial tasks of this type and relatively better performance on verbal tasks of this type is beginning to emerge. In 1 study,[160] researchers gave participants with WS and their general mental age controls a radial arm maze task that involved keeping a running mental record of which arms had been visited, while visiting others. On this visuospatial processing and storage task, participants with WS were slower and made more errors. Similar impairments were reported for a computer-presented spatial keeping-track task[163] and a backward Corsi task[153] relative to controls matched on general mental age and nonverbal ability, respectively. In contrast to the visuospatial domain, simultaneous processing and storage in the verbal domain seems less impaired. On reverse digit recall tasks, participants with WS scored consistently with their own general cognitive ability,[155] similarly to typically developing controls matched on grammatical understanding,[166] and nonsignificantly higher than participants with DS matched on age and IQ.[156] A recent study using a slightly older sample (mean chronological age = 19–11) showed reverse digit recall performance below that of typically developing participants matched on nonverbal mental age.[153]

Delayed Memory and Learning

Unlike immediate memory, youth with WS are poor in delayed memory and learning in both verbal and visuospatial domains. When compared with typically developing controls at the same receptive vocabulary level or to their own general cognitive ability level, young people with WS perform more poorly on word list or name learning tasks.[114,150,167] Two studies using these tasks also tested recall of learned words after 10 or 15 minute delay, one finding that participants with WS performed more poorly than TD participants[150] and the other finding that participants with WS performed similarly to TD participants.[167] Possibly, participants with WS did better in the latter study because they had picture supports during the learning trials,[167] discrepancy has to do with different matching procedures. In addition to word list learning, young people with WS also perform poorly relative to their own general cognitive level on auditory-visual associative learning and on delayed retention of learned associations.[155] Difficulty in delayed memory may not extend to recognition tasks, however,[114,167] and when engaging in rehearsal, youth with WS show the expected recall advantage for early list items (primacy effect), which can be considered an instance of delayed verbal memory.[150,157] Even so, their performance on early list items is lower than would be expected based on their receptive vocabulary level.[150,157]

In the visuospatial domain, youth with WS also have clear difficulties on delayed memory and learning tasks. For example, in one study,[150] children and adolescents with WS performed below receptive vocabulary-level controls in drawing abstract figures from memory after a 10-minute delay, even when their memory scores were corrected for their ability to draw figures initially. Similar to immediate recall, the difficulty seems to be in the spatial rather than visual aspects of the tasks. Most studies show that young people with WS have no particular difficulty with delayed picture recognition.[110,111,114,155,163,167] Also, in a study directly contrasting learning of objects versus object locations, participants with WS performed similarly to developmental controls on the objects but more poorly on the locations.[115] Unlike immediate spatial memory, however, the difficulty with delayed spatial memory may be accounted for by more basic impairment in spatial processing. On a delayed visuospatial recall task, participants with WS performed below controls when matched on vocabulary level, but similarly when matched on spatial reasoning ability.[114] Thus, children with WS have difficulty with delayed spatial memory, but the impairment may not be in memory per se. This pattern of difficulty on delayed spatial memory tasks and the link to problems in spatial processing in general appear specific to WS. Participants with DS did not show the same pattern.[114,115]

Semantic/Conceptual Memory

In spontaneous language and on fluency tasks, individuals with WS reportedly tend to use unusual vocabulary words. This observation has led researchers to examine storage, organization, and access to semantic representations in memory in WS. Studies measuring speeded naming of pictures (or objects) suggest some difficulty in access to semantic representations for young people with WS relative to their developmental level. Compared with controls matched on general cognitive ability, nonverbal ability, or receptive vocabulary, participants with WS were slower[168] or less accurate[120,169,170] or both.[171,172] In contrast, alphanumeric naming was similar to that of controls in both speed and accuracy.[120,149] Thus, access to semantic representations in memory seems affected more so than access to phonological representations.

Access to semantic representations, however, seems affected only for retrieval of the precise names of pictures and not for retrieval of any of a number of possible category exemplars. In semantic fluency tasks, youth with WS show no difficulty at all relative to their developmental level. In several studies, young people with WS generated as many category exemplars as typically developing controls matched on general cognitive ability,[90,122,170] nonverbal mental age,[169,172] or receptive vocabulary,[173] as well as ID controls matched on age and IQ.[174,175] Temple et al[170(p463)] suggested that there is a "looser criterion for target identification" in WS, and this could explain the discrepancy between the speeded naming findings and the semantic fluency findings. Thomas et al[171] suggested that the use of unusual vocabulary words in spontaneous speech of individuals with WS is best explained by extra-lexicon factors such as desire for social engagement.

Despite speculations to the contrary, evidence suggests that the organization of semantic memory in WS is typical. Tyler et al[176] examined relatedness of concepts in semantic memory by measuring semantic priming effects—the speed advantage in recognizing a word after being "primed" by a semantically related versus an unrelated word. They found the same semantic priming effects for functional relations and for category relations in young people with WS that were reported in typical same-age controls. Also, in speeded naming and fluency tasks, the effects of frequency, semantic category, and typicality seem to be similar in young people with WS as in controls matched on general cognitive ability[122] or receptive vocabulary.[171,174]

Summary of WS Memory Profile

The memory profile for WS includes weaknesses in immediate spatial recall, verbal and spatial delayed memory and learning, and semantic retrieval involving precise names. It includes relative strengths in immediate verbal and visual recall, visual delayed memory and learning, and phonological retrieval. Most relevant to reading are weaknesses in visual-auditory learning and semantic retrieval as well as relative strengths in immediate recall (both visual and verbal) and rapid phonological retrieval.

Implications for Reading Development

Despite impairments in spatial memory and associative learning, several relative strengths in the WS memory profile should facilitate reading development. Relative strength in visual memory should help children with WS develop a prealphabetic sight vocabulary. Also, relative strength in verbal WM and phonological retrieval could offset weakness in associative learning to facilitate phonological recoding and word recognition. The very small literature on reading skills of children with WS says little about prealphabetic sight-word reading.[177] However, most studies suggest that both word and nonword reading are on par with nonverbal or general cognitive level.[178–180] Laing and coworkers compared youth with WS to typically developing children of the same word recognition level and found that although the WS group was slower in learning contrived sight words (e.g., LTR for letter), they were just as good at using phonetic cues.[106] Possibly, their difficulty learning the contrived sight words was related to poor visual-auditory associative learning.[155]

It would be reasonable to predict relatively good reading comprehension in WS based on relative strengths in verbal WM and word recognition. Also, despite some subtle impairments in semantic processing, most aspects of semantic memory function at the expected level. However, at least 3 studies have shown that reading comprehension is far lower than word recognition.[149,178,181] The weakness in reading comprehension may be due to those aspects of semantic memory that are specifically impaired in WS. However, in 1 study, listening comprehension was as good as word recognition and much better than reading comprehension.[149] According to the Simple View of reading, good word recognition and good language comprehension should produce good reading comprehension. Thus, there may be a factor such as attentional control[26] or general processing speed[182] that contributes to poor reading comprehension in WS. Menghini et al[153] showed that young people with WS were worse than controls matched on nonverbal mental age at tasks measuring attentional control with visual materials, such as the trail making test. Further research is needed to understand the underlying reasons for unusually poor reading comprehension in WS.

Fragile X Syndrome

FXS is the most common known inherited form of ID, occurring in ~1 in 4000 males and 1 in 8000 females.[183,184] It is caused by an expansion of the CGG trinucleotide sequence of the *FMR1* gene found on the X chromosome. This expansion reduces the gene's production of the fragile X mental retardation protein (FMRP) protein, which plays an important role in normal brain functioning. Those who have a larger expansion (a full mutation) are more severely affected than those with a moderate expansion (a premutation). Furthermore, females are less likely to be severely affected compared with males because they have a second X chromosome that is likely to be unaffected. Nearly all males with the full mutation have moderate to severe ID, whereas only 30% to 50% of females with the full mutation have intellectual or learning impairments. Physical symptoms of males include a large head circumference in infancy, long ears, a large lower jaw, and postpubertal testicular enlargement.[185,186] Brain areas known to be affected include the cerebellar vermis and superior temporal gyrus.[187–189] FXS is strongly associated with executive functioning impairments, mathematics difficulties, and autistic-like behaviors.[190–192] As most of the memory literature on females with FXS focuses on those who do not have ID, this article covers only males with FXS.

Aspects of Working Memory

Very poor verbal immediate memory in males with FXS is apparent on digit recall tasks[80,93,193–195] and nonword repetition tasks[80] across a variety of matching measures including receptive vocabulary, logical operations, general cognitive ability, and nonverbal ability level. In addition, FMRP level correlates with digit span.[196,197] The weakness in verbal immediate memory in males with FXS seems to be of similar magnitude as in DS.[80,93–95] However, unlike DS, there are also visuospatial immediate memory impairments in FXS. Males with FXS consistently perform below their developmental level on visual[93,195,198] and spatial[80,93,193,194,198,199] immediate memory tasks. This is true whether matched on receptive vocabulary, logical operations, general cognitive ability, or nonverbal ability level. Difficulties in visuospatial immediate memory can be more severe in FXS than in other etiologies of ID,[80,94,95] although not every study has shown this.[93,200]

Males with FXS also perform very poorly on tasks measuring simultaneous processing and storage. This is consistent with the well-known problems in executive functioning. With only a few exceptions,[195,199] males with FXS perform worse than typically developing controls[80,193,195,198] and controls with DS[80,95] of similar receptive vocabulary, logical operations, or nonverbal ability. This includes reverse digit recall, grouped digit/word recall, and reverse Corsi tasks. Lanfranchi et al[193] found that the more cognitive control a task required, the more clear the impairment was, regardless of whether the task was verbal or visuospatial.

Delayed Memory and Learning

Some aspects of delayed memory and learning also seem problematic relative to developmental level in FXS. For example, Kogan et al[201] found that boys with FXS performed below typically developing controls matched on receptive vocabulary at basic object discrimination learning and generally lower than controls with DS. Other research suggests that males with FXS perform at their receptive vocabulary or nonverbal ability level in recall and recognition of objects[93,198] and similarly to matched males with DS,[95] although they might be at a disadvantage when not expecting a test.[93] None of these studies addressed delayed memory or learning of verbal material. However, one study showed particularly poor performance on visual-auditory learning.[199] In this study, boys with FXS made significantly more errors in immediate and delayed recall after practicing associations between rebus symbols and word names than typical children of the same nonverbal age.

Semantic/Conceptual Memory

There is very little research on semantic memory in individuals with FXS. However, one study showed males with FXS performed below receptive vocabulary level controls in story retelling.[80]

Summary of FXS Memory Profile

Males with FXS have severe impairments in most aspects of WM, in both the verbal and visuospatial domains. They are also poor at learning tasks and story retelling. All of these are important for reading development. Delayed memory may not be quite as severely affected in boys with FXS. Little is known about rapid phonological retrieval or semantic/conceptual memory in boys with FXS; these are also very important to reading development, and further research is needed.

Implications for Reading Development

For boys with FXS, impairments in visual and verbal WM and in associative learning would predict great difficulty in prealphabetic sight-word reading as well as in phonological recoding and word recognition. Unfortunately, little research has been done on any of these aspects of reading in this population. It is fairly clear that word recognition skills are not as poor as arithmetic skills,[94,194,202] but this is probably due to exceptionally poor arithmetic skills rather than especially good word recognition skills. Buckley and Johnson-Glenberg[203] reported that young males with FXS did better on word recognition than typically developing children matched on nonverbal ability. However, this difference may be inflated because 1 of 2 measures in the matching variable was immediate memory. As immediate memory is severely impaired in FXS, most other skills would be higher, even word recognition. It also should be noted that nothing is known yet about phonological retrieval in FXS, and if this is relatively good, it could offset the effect of WM impairments on word recognition. In a separate article, Johnson-Glenberg reported that as expected based on the FXS memory profile, boys with FXS were worse on nonword reading than expected for their reading level.[199] A recent large-sample parent survey indicated that by 6 to 10 years old, ~80% of boys with FXS could recognize letters, ~60% knew letter sounds, and ~60% could read words by sight.[204] These percentages were fairly stable through the over 20 years age group, suggesting little progress through the adolescent and young adult years, although these are not longitudinal data.

There is almost no research at all on reading comprehension in boys with FXS. Although little is known about semantic memory in FXS, based on severe impairments in verbal WM, great difficulties in reading comprehension would be expected. The recent parent survey[204] indicated that only ~40% of boys in the age range 6 to 10 years could read basic picture books, and ~20% could read books that contained new words or concepts. Again, these rates increased very little through adulthood. Clearly, more research is needed on reading skills of boys with FXS.

CONCLUSION

Although memory difficulties are common in ID regardless of etiology, the present review indicates that the exact type and magnitude of difficulty varies with the etiology of ID. Cross-etiology variability in memory difficulties leads to somewhat different expectations and outcomes for reading development. In DS, relatively good immediate visual memory and rapid phonological retrieval presumably provide for relatively good word recognition skills. However, very poor verbal WM skills probably contribute to difficulties in phonological recoding and reading comprehension. In WS, relatively good visual and verbal WM and rapid phonological retrieval may allow for relatively good word and nonword reading skills. Reading comprehension is poor, however, and this might be attributable to specific impairments in semantic retrieval and/or a factor such as attentional control. In FXS, severe impairments in visual and verbal WM, learning, and story telling predict difficulty in word recognition and reading comprehension. Little is known about rapid phonological retrieval or semantic memory, which might also affect these reading skills. Research on reading skills in FXS has not yet produced firm findings.

The focus of this article is on memory and its relation to reading in ID. However, language skills are also extremely important to reading development, and they also may be differentially strong or weak in particular ID syndromes. For example, in DS, poor phonological awareness and receptive grammar probably contribute to phonological recoding and reading comprehension difficulties. WS and FXS also have distinct language profiles that probably contribute to strengths and weaknesses in reading. Research is needed to examine how memory and language profiles work together to influence reading outcomes in youth with ID.

Although the present article focused on cross-syndrome variability, there are also obvious cross-syndrome similarities in reading skills profiles. For example, in both DS and WS, there is a pattern of relatively strong word recognition and very weak reading comprehension. This pattern is similar, despite very different memory profiles and very different language profiles in the 2 syndromes. More research is needed to determine the complex of memory, linguistic, and other factors that create this particular pattern of reading skills.

Finally, it is important to remember that substantial within-syndrome variability exists, and it would be a mistake to assume that the same memory or reading profile would apply to every individual in a syndrome group. Instructional approaches should be tailored to the individual, providing extra support in weak areas and finding ways to capitalize on relative strengths. However, a basic understanding of the general patterns of performance associated with genetic syndromes of ID may provide a starting point for educational planning.

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