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Williams Syndrome and Music: A Systematic Integrative Review

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WILLIAMS SYNDROME AND MUSIC: A SYSTEMATIC INTEGRATIVE REVIEW

by

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A thesis submitted to the Graduate College
in partial fulfillment of the requirements
for the degree of Masters of Music
School of Music
Western Michigan University
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WILLIAMS SYNDROME AND MUSIC: A SYSTEMATIC INTEGRATIVE REVIEW

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Researchers and clinicians have often cited the curious relationship between individuals with Williams syndrome and music. This review aimed to systematically identify, analyze, and synthesize research and findings related to Williams syndrome and music. Thirty-one articles were identified that examined this relationship and were divided into seven areas by their findings. This process covered a diverse array of methodologies, with aims to: 1) report current findings; 2) assess methodological quality; and 3) discuss the potential implications and considerations for the clinical use of music with this population. Collecting and analyzing available research on the relationship between Williams syndrome and music could serve as a platform for future research on the clinical potential for music with this population and guide future inquiry in all areas.
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INTRODUCTION

Review of Literature

Some of the earliest accounts of Williams syndrome (WS) were identified in descriptions of case studies with two seemingly unrelated sets of characteristics (see Berdon, Clarkson, & Teele, 2011- for review). In the 1950s, cases were reported of infantile hypercalcemia, further characterized by failure to thrive, a distinctive facial appearance, and developmental delays (Bongiovanni, Eberlein, & Jones, 1957; Schlesinger, Butler, & Black, 1956). In the early 1960s, cases identified by New Zealand cardiologist John C. P. Williams and German physician Alois Beuren were characterized by supravalvular aortic stenosis (SVAS) along with a similar presentation of persistent growth failure, distinctive facial appearances, developmental delays, and an overly friendly personality (Beuren, Apitz, & Harmjanz, 1962; Williams, Barratt-Boyes, & Lowe, 1961). By the mid-1960s, case reports describing individuals with both hypercalcemia and SVAS, along with other features common to both phenotypes, suggested that these seemingly unrelated presentations were variations of the same phenotype (Berdon et al., 2011).

Williams syndrome (WS; also referred to as Williams-Beuren syndrome) is a rare neurodevelopmental disorder caused by a hemizygous microdeletion on chromosome 7 (7q11.23), composed of approximately 26-28 genes and commonly including one allele of the elastin gene, with an estimated prevalence of 1 in 7,500-10,000 live births (Ewart et al., 1993; Pober, 2010; Strømme, Bjørnstad, & Ramstad, 2002). Prior to the early 1990s, the diagnosis of WS was primarily based on clinical phenotype (Martens, Wilson, & Reutens, 2008). However, more contemporary methods of diagnosis include laboratory tests such as fluorescence in situ hybridization (FISH) and array comparative genomic hybridization (Pober, 2010).

Individuals with WS commonly present with a unique array of medical, developmental, and social-emotional qualities. Distinctive facial features are often described as “elfin” or “pixielike,” including almond-shaped eyes, a stellate pattern in the iris, a broad forehead, high and prominent cheekbones, a flat nasal bridge with upturned nose, full lips, a broad mouth, and abnormal dentition (Levitin and Bellugi, 1998). Medically, individuals with WS may experience
cardiovascular abnormalities (approximately 70% have supravalvular aortic stenosis—narrowing of the large blood vessel that carries blood from the heart to the rest of the body), hypertension (approx. 50%), hypercalcemia (5-50%), subclinical hyperthyroidism (15-30%), precocious puberty, poor weight gain, low muscle tone, brisk reflexes, curvatures of the spine, and sleep irregularities (Pober, 2010). A higher incidence of hearing sensitivities has also been reported, including: hyperacusis (lowered threshold and higher detectability for hearing sounds), odynacusis (lowered pain threshold for loudness of sounds), auditory aversions (fear of certain types of sounds at normal volume) and auditory fascinations (strong attraction to certain types of sounds) (Levitin, Cole, Lincoln, & Bellugi, 2005).

Individuals with WS have been anecdotally described as unusually social, friendly, polite, highly empathetic, irresistibly drawn to strangers, and driven by social interaction (Doyle, Bellugi, Korenberg, & Graham, 2004; Plesa Skwerer & Tager-Flusberg, 2016). Compared to peers, individuals with WS are generally described as more hyperactive (approx. 65% meet DSM-IV criteria for ADHD), distractible, anxious, and more prone to develop specific phobias (Dykens, 2003; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006). Järvinen et al. (2012) remarked on “intriguing dissociations” apparent in WS which include overly friendly behavior with a difficulty making friends, social fearlessness coupled with anxiety, and abundant positive affect with maladaptive behaviors. Individuals with WS fall in the mild to moderate range of intellectual disability, with mean IQ scores falling between 50-60 with a range of 40-100, and IQ appears to be relatively stable with age (Martens et al., 2008). Many individuals with WS struggle with deficits in global versus local processing of visual stimuli (Bihrle, Bellugi, Delis, & Marks, 1989), visuospatial and visual motor skills (Farran, 2005), and pragmatics (Brock, 2007). However, these individuals also possess relative strengths in expressive language, receptive vocabulary, phonological memory, facial perception and processing, and music (Brock, 2007; Martens et al., 2008).

The relationship between individuals with WS and music has been recognized even from early case reports. These reports described children with WS as having good singing skills (von
Arnim & Engel, 1964) and an ability to learn songs with ease (Udwin, Yule, & Martin, 1987). Individuals with WS show a high affinity for music: demonstrating high engagement in musical activities, expressing an interest in music at an early age, and exhibiting a heightened emotional responsiveness to music (Don, Schellenberg, & Rourke, 1999; Dykens, Rosner, Ly, & Sagun, 2005; Levitin et al., 2004). Investigation into this unique relationship has been conducted in a variety of areas, revealing a higher incidence of absolute pitch (Lenhoff, Perales, & Hickok, 2001), an extreme tendency toward fundamental/holistic pitch processing (Wengenroth, Blatow, Bendszus, & Schneider, 2010), and greater activation of emotion-related areas of the brain in response to music in WS (Levitin et al., 2003; Thornton-Wells et al., 2010).

Early investigation into the musical skills of individuals with WS remarked on apparently enhanced or preserved abilities in music, including: a keen sense of pitch in reproducing songs, an enhanced skill for producing rhythms, and greater musical creativity through the ability to improvise, compose, and show increased expressiveness in musical interactions (Lenhoff, Wang, Greenberg, & Bellugi, 1997; Levitin & Bellugi, 1998). Later studies have yielded more precise and mixed results, suggesting that musical abilities in WS are more likely to be areas of relative strength rather than preserved functioning (Hopyan, Dennis, Weksberg, & Cytrynbaum, 2001; Martens, Reutens, & Wilson, 2010; Martínez-Castilla & Sotillo, 2008; Martínez-Castilla, Sotillo, & Campos, 2011).

Rationale for Research

Since its discovery, researchers and clinicians have often cited the curious relationship between individuals with Williams syndrome and music. This phenotypic curiosity has been the subject of multiple articles and empirical studies over the past 20 years. Research examining this relationship covers a diverse array of topics. Currently, no research or formal review has been conducted to examine the relationship between Williams syndrome and music through a comprehensive research lens. Notably, there are also no published articles examining the clinical use of music with individuals with WS. Collecting and analyzing available research on the
relationship between Williams syndrome and music could guide future inquiry in all areas and serve as a platform for future research on the clinical potential for music with this population.

**Purpose**

The purpose of this review is to systematically identify, analyze, and synthesize research findings related to Williams syndrome and music. This process will cover a diverse array of methodologies, with aims to: 1) report current findings; 2) assess methodological quality; and 3) discuss the potential implications and considerations for the clinical use of music with this population.

**Research Questions**

1. What does the current literature reveal about the relationship between WS and music?
2. What are the methodological strengths and weaknesses of the included studies?
3. What are potential implications and considerations for the clinical use of music with this population?
METHODS

Design

This thesis borrowed from methodologies of systematic and integrative reviews.

A systematic review is a methodologically rigorous and comprehensive review of literature intended to identify, select, and critically appraise relevant research. A systematic review attempts to answer a specific research question by gathering and analyzing all empirical evidence that meets predetermined criteria. It follows explicit and systematic methods to reduce bias and enhance the quality of the findings. The studies included in these reviews tend to follow similar methodologies and/or report similar outcomes. (The Cochrane Collaboration, 2011; Moher, Liberati, Tetzlaff, Altman, & The PRISMA Group, 2009).

An integrative review is a broader approach that includes studies which utilize different methodologies, including both theoretical and empirical literature, in order to gain a greater perspective of a particular phenomenon. Integrative reviews incorporate a wide range of purposes: to define concepts, to review theories, to review evidence, and to analyze methodological issues of a particular topic. However, analyzing and synthesizing varied primary sources is a major challenge in undertaking an integrative review and leaves room for bias and error. The studies included in the integrative review often need to be divided into subgroups according to some logical system to facilitate analysis. (Souza, Silva, & Carvalho, 2010; Whittemore & Knafl, 2005).

This review drew from and follows the framework of an integrative review with respect to the inclusion and analysis of research from a diverse set of methodologies and covers a range of topics related to Williams syndrome and music. In order to enhance the rigor and reduce the risk for bias or error, the formal search for literature more closely mirrored that of a systematic review in an attempt to identify, select, and compile a comprehensive database of primary sources related to this topic.
Search Strategy

An electronic search was conducted from the following databases: PubMed, PubMed Central, EMBASE, SCOPUS, Web of Science, EBSCO, and ProQuest. These databases were searched using the terms: “Williams syndrome” and “music.” Additionally, reference lists from included articles were reviewed for other relevant articles.

Eligibility Criteria

The following inclusion and exclusion were used to determine eligibility.

Criteria for Inclusion:

- Date Range: published before 2017 (through December 2016)
- Subject: Williams syndrome
- Subject: music
- Participants (where applicable): Williams syndrome
- Published in English
- Published in peer-reviewed journal

Criteria for Exclusion:

- Participants (where applicable) include a mixture of individuals with WS and individuals with other diagnoses and outcomes are not differentiated by the authors
- Participants (where applicable) have comorbid diagnoses of WS and another diagnosis
- Article subject is hyperacusis without reference to music
- Review article

Search Procedure

Search results were managed using Endnote X8. After all database searches were complete and duplicates removed, all remaining search results were first screened for inclusion based on the title and abstract. Finally, the remaining articles were screened using the full-text of the article. (For a full list of exclusions based on full-text, see Appendix A). All screening was duplicated by a secondary independent reviewer trained in following the aforementioned criteria in order to prevent the omission of potentially relevant articles. All discrepancies were settled between the two reviewers.
Data Extraction

The primary author extracted data from the included articles using a Data Extraction Form created for this study (Appendix B). Two secondary independent reviewers, trained in the use of the extraction form, each reviewed a random selection of 25% of the articles using sections: 4: Participants and Groups; 5: Tasks and Outcomes; and 7: Other Information. Findings from each of the secondary reviewers were reviewed with the primary author; all discrepancies were settled between the two reviewers.
RESULTS

Search Results

The formal search identified 31 articles that met the criteria for inclusion (Figure 1). These articles were allocated into seven groups according to their outcomes and findings (Table 1). Some of the articles were included in multiple groups. Overall, the 31 articles included 38 studies as some of the articles included more than one report. Two of these articles (Lense & Dykens, 2013a; Järvinen et al., 2012) included only one report that met the criteria for inclusion. Only data from that report was extracted for analysis.

*Figure 1. PRISMA Flow Diagram*
Figure 2 shows the cumulative number of publications by year, demonstrating a fairly gradual and consistent growth in available literature over time. Table 2 shows the number of articles published by journal. Table 3 shows which databases in which the articles were indexed. Of note, 100% of the included articles were indexed within the ProQuest database.

Table 1

| Topic                                                      | # of articles* |
|------------------------------------------------------------|----------------|
| Musicality: Interest, Experience, and Engagement            | 10             |
| Musical Skill: Tonal and Rhythmic Skills                   | 9              |
| Emotional Responsiveness                                   | 12             |
| Musical Processing: Absolute Pitch, Amusia, and Auditory Processing | 4              |
| Brain Imaging and Morphology                               | 5              |
| Cognitive Processes: Memory and Math                       | 3              |
| Fears, Anxieties, and Problem Behaviors                    | 3              |

* 9 articles were included in 2 topic areas and 3 articles were included in 3 topic areas.

Figure 2. Cumulative Number of Publications by Year
Table 2
Number of Inclusions by Journal

| Journal                                                      | # of articles: |
|--------------------------------------------------------------|----------------|
| Child Neuropsychology                                        | 4              |
| Music Perception                                             | 4              |
| Frontiers in Psychology                                      | 3              |
| Neuropsychologia                                             | 3              |
| Research in Developmental Disabilities                       | 3              |
| American Journal on Intellectual and Developmental Disabilities | 2              |
| Brain Sciences                                               | 2              |
| American Journal of Mental Retardation                       | 1              |
| Autism Research                                              | 1              |
| Developmental Psychobiology                                  | 1              |
| Exceptional Children                                         | 1              |
| Journal of Intellectual Disability Research                  | 1              |
| Journal of Mental Health Research in Intellectual Disabilities| 1              |
| Neuroimage                                                   | 1              |
| Neuroreport                                                  | 1              |
| PLoS One                                                     | 1              |
| Social Cognitive and Affective Neuroscience                  | 1              |

Table 3
Number of Inclusions by Database

| Database                  | # of inclusions | % of articles indexed in database |
|---------------------------|-----------------|-----------------------------------|
| ProQuest                  | 31              | 100.00%                           |
| SCOPUS                    | 28              | 90.32%                            |
| PubMed                    | 25              | 80.65%                            |
| Web of Science            | 23              | 74.19%                            |
| EMBASE                    | 20              | 64.52%                            |
| EBSCO                     | 12              | 38.71%                            |
| PubMed Central            | 11              | 35.48%                            |
FINDINGS

Thirty-one articles that met the criteria for inclusion were allocated into seven groups according to their outcomes and findings. Some of the articles were included in multiple groups.

Musicality: Interest, Experience, and Engagement

Ten of the included articles reported findings related to musicality (Table 4). The affinity for music and overall musicality among individuals with WS has been anecdotally cited since early case reports (Udwin et al., 1987; von Arnim & Engel, 1964). Yet, the concept of ‘musicality’ is difficult to define or quantify. For the purposes of this paper, interest, experience, and engagement were included as aspects of musicality, aside from musical skill. Other facets may include creativity, expressivity, sensitivity, and emotionality, which fall under the category of ‘engagement’ in this paper (for findings related to emotional responsiveness, see ‘Emotional Responsiveness’ section, pg. 25). Given the lack of formalized assessment measures in these areas, most of the studies included in this section utilized parent report via various questionnaires to examine these aspects of musicality. The most utilized tools among the included studies were the Musicality Interest Scale (MIS; Blomberg, Rosander, & Andersson, 2006) and the Salk/McGill Music Inventory (SAMMI; Levitin et al., 2004). Many other studies in this paper utilized questionnaires to examine musicality indirectly, however did not report findings as a focus of their study (Blomberg et al., 2006; Dai et al., 2012; Lense & Dykens, 2013b; Lense, Gordon, Key, & Dykens, 2014; Lense, Shivers, & Dykens, 2013).

Studies in this section continue to shed light on this aspect of the WS phenotype with an overall high level of agreement. Levitin et al. (2004) conducted a comprehensive survey with the largest sample of individuals with WS included in this study (n=118), as well as typically developing (TD) controls and two comparison groups of individuals with other neurodevelopmental disorders, Autism Spectrum Disorder (ASD) and Down Syndrome (DS). The WS group significantly differed from the other comparison groups in multiple areas, many of which are consistent with findings from other studies. Specifically, in comparison to all groups, individuals with WS manifest interest in music at an earlier age, spend more hours per
12

week listening to music, and demonstrate higher emotional responses to music (also Don et al., 1999), the effects of which were reported to last longer in individuals with WS.

Levitin et al. (2004) further report that, compared to TD peers, individuals with WS are similar in overall musical involvement; however individuals with WS show a higher interest in music-related activities (also Don et al., 1999; Levitin & Bellugi, 1998); are as likely to play instruments, however individuals with WS play instruments more often; and show similar frequency of spontaneously generating music. When compared to comparison groups of others with neurodevelopmental disorders, individuals with WS: exhibit higher interest and engagement with musical activity, are more likely to play a musical instrument (also Dykens et al., 2005), spend more hours per week playing a musical instrument, play or create original music and rhythms more frequently, are more accurate in their reproductions of songs, and are rated higher in musical skill and achievement (also Dykens et al., 2005). Also, individuals with WS tend to express a higher degree of enjoyment with music compared to siblings or peers (Dunning, Martens, & Jungers, 2015; Martens, Jungers, & Steele, 2011) and individuals with greater linguistic capacity tend to be more interested in music (Ng, Lai, Levitin, & Bellugi, 2013).

Using factor and discriminant function analyses, Levitin et al. (2004) also established seven principal components that predicted group membership for 70% of the cases among participants with WS and all comparison groups: age of onset of musical behaviors, listening habits, sensitivity to music, spontaneity, reproduction, negative reactions to music, and exposure to music theory.

Reflecting on the musical engagement or involvement in individuals with WS, between 50-90% of individuals with WS engage in some type of musical training (Dunning et al., 2015; Lense et al., 2013; Martens et al., 2011). However, information about the musical training of participants is inconsistently reported across studies and may be subject to bias when participants are recruited from music camps. Many individuals with WS elect to participate in choir or band throughout school. In terms of private lessons, considering the physical, cognitive, and visuospatial limitations of those with WS, many take lessons in piano, voice, or drums, with
fewer taking lessons on other instruments such as violin or guitar. Other musical activities may include dance or theater. Anecdotally, individuals with WS tend to express a higher degree of interest in musical toys (such as toy instruments or cause and effect toys that make music), are more likely to make ‘instruments’ out of household items, and begin to play instruments an average of 5 years before formal training (Levitin et al., 2004).

Lense & Dykens (2013b) taught 46 children and adults with WS to play a novel instrument, the Appalachian dulcimer, in a single, 35min, semi-structured, adaptive lesson. Gained skill on the novel instrument was associated with number of types of formal lessons, number of instruments played formally and informally, hours per day playing instruments or singing, parent report of musical skill, and better visual-motor integration ability. However, self-reported use of auditory learning strategies predicted greater skill on the dulcimer beyond musical skill (as rated by a solo musical performance) and visual-motor integration. Similarly, Lense & Dykens (2013a) reported that greater musical skill on a prepared solo music performance, as judged by trained judges, was associated with starting lessons at a younger age, exposure to multiple types of lessons, and amount of time currently spent playing music. These studies suggest that the number of types of musical training and not duration of individual lessons may be a more reliable indicator of musical achievement in individuals with WS (see also, Reis, Schader, Milne, & Stephens, 2003). Lense et al. (2013) suggest that this may be related to the fact that musical training in WS is often different and more inconsistent in individuals with WS than TD populations. Families of individuals with WS often experience difficulty in finding suitable teachers and individuals with WS may experience discouragement related to the fine motor skills required to play certain instruments. These families may be forced to seek out various teachers or multiple avenues for musical enrichment, with the only constant being the continual involvement in some type of musical activity as opposed to sustained duration in a particular musical activity.

Many of the included studies offered various considerations for the musical education of individuals with WS. To promote participant success, Lense & Dykens (2013b) selected an
instrument that was challenging but didn’t require intensive fine motor skills, placed numbered stickers along the frets, did not use written music- instead focused on repeated rhythms and patterns, and utilized a curriculum with a mixture of familiar song and improvisation. Don et al. (1999) discussed adaptations concerning physical and cognitive limitations and suggested that training could incorporate shorter sessions with a predictable routine, include simple tasks such as imitation and repetition, and provide opportunities for creative and emotional expression. Reis et al. (2003) also point out a preference for social, auditory, and group learning in individuals with WS. Taken together, these considerations suggest that musical training for individuals with WS may be better suited toward a focus on strengths, learning preferences, and the development of musical expressiveness through play and improvisation, as opposed to learning specific skills such as reading music and learning scales or notes (Hopyan et al., 2001).

Lense et al. (2013b) is the only included study to directly examine musical training, which was evaluated by implementing a single semi-structured lesson. Longitudinal studies of musical development and studies examining the effects of repeated musical training over time would help to increase understanding of learning strategies and supports necessary for individuals with WS. Future studies are needed to examine the impact of these proposed supports on musical training and may have implications for other educational or clinical interventions.

One overall limitation shared among articles in this section is the reliance on parent report to interpret facets of musicality. The subjectivity inherent in this type of assessment leaves room for under- or over-reporting responses to music. Despite the difficulty in operationalizing the concept of ‘musicality,’ future studies would benefit from operationalizing individual aspects of musicality to examine their role in the WS phenotype more closely.

Overall, the long-cited claims of an affinity for music within individuals with WS appear to be expressed through the concept of musicality or the manner in which individuals with WS interact with music.
| Author/Year | N     | Age\(^1\) (yrs) | Task(s)                                                                 | Finding(s)                                                                 |
|------------|-------|-----------------|-------------------------------------------------------------------------|----------------------------------------------------------------------------|
| Dunning, Martens, & Jungers (2015) | WS) 44 | WS) 8-48        | Musicality questionnaire                                                 | Increased enjoyment and frequency of music listening compared to peers and siblings |
| Lense & Dykens (2013a) (study #2 only) | WS) 13 | WS) 29.47 +/- 6.35 | Prepared solo music performance in front of mixed familiar/unfamiliar audience. | Greater musical skill associated with starting lessons at younger age, exposure to multiple types of lessons, and amount of time currently spent playing music |
| Lense & Dykens (2013b) | WS) 47 | WS) 7-49        | Single semi-structured lesson on a novel instrument (Appalachian dulcimer); Musicality Interest Survey (MIS)\(^2\) | Self-reported use of auditory learning strategies predicted greater skill on a novel instrument beyond previous musical skill and visual-motor integration (provides considerations for musical education) |
| Lense, Shivers, & Dykens (2013) | WS) 73 | WS) 10-51       | Musicality Interest Survey (MIS)\(^2\) Amusia battery                  | High percentage of WS involved in musical training; Exposure to various types of musical training was a better predictor of musical skill than cumulative duration |
| Ng et al. (2013) | WS) 55 TD) 19 | WS) 16-52 TD) 18-41 | Compared measures of musicality, sociability, and language comprehension; Salk McGill Music Inventory (SAMMI)\(^3\) | Greater interest associated with greater linguistic capacity |
| Martens, Jungers, & Steele (2011) (2 studies) | WS) 38 | WS) 6-59        | Musicality questionnaire                                                 | Increased enjoyment and frequency of music listening compared to peers and sibling |
|           | WS) 38 | WS) 7-50        |                                                                         |                                                                           |
Table 4 cont.
Musicality: Interest, Experience, and Engagement

| Author/Year | N   | Age¹ (yrs) | Task(s) | Finding(s) |
|-------------|-----|------------|---------|------------|
| Dykens et al. (2005) | WS) 31 | WS) 10.22 (4.86) | Musicality questionnaire | WS more likely to take music lessons, play an instrument, and have higher ratings of musical skill |
|             | PW) 26 | PW) 10.26 (4.86) |         |            |
|             | DS) 32 | DS) 11.5 (4.49) |         |            |
|             |       | All) Range 4-21 |         |            |
| Levitin et al. (2004) | WS) 118 | WS) 20.88 (11.48) | Salk McGill Music Inventory (SAMMI)³ | Greater emotional responses to music, manifest interest in music at an earlier age, more hours per week listening to music than all other groups; Higher musical accomplishment, engagement, and interest than ASD/DS |
|             | TD) 118 | PW) 19.38 (6.70) |         | 7 musical factors predict group membership |
|             | ASD) 30 | DS) 18.83 (7.11) |         |            |
|             | DS) 40 | All) Range 8-47 |         |            |
|             |       |       |         |            |
| Reis et al. (2003) | WS) 16 | WS) 20.4 (10.4) | 10-day intensive music program | Use of a talent development approach improved musical skill and engagement (provides considerations for musical education) |
|             |       | Range: 5-50 |         |            |
|             |       | TD) 20.9 (7.4) |         |            |
|             |       | Range: 5-44 |         |            |
| Don, Schellenberg, & Rourke (1999) | WS) 19 | WS) 8-13 | Parent interview and questionnaire | Greater interest and range of emotional responses to music (provides considerations for musical education) |
|             | TD) 19 | TD) 5-12 |         |            |

ASD = Autism Spectrum Disorder; DS = Down Syndrome; PW = Prader-Willi Syndrome; TD = Typically Developing Control; WS = Williams Syndrome

¹Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years

²Musicality Interest Scale (MIS): Questionnaire with subscales in: 1) interest in and liking of music, 2) emotional reactions to music, and 3) musical skills.

³Salk McGill Music Inventory (SAMMI): Comprehensive questionnaire with subscales in: 1) demographic information, 2) interest in music, 3) emotional responsiveness to music, 4) music creativity and reproduction, 5) musical training, 6) age of onset of musical behavior.
Musical Skill: Tonal and Rhythmic Skills

Nine articles assessed a wide variety of tonal and rhythmic skills (Table 5). Given the complexities in measuring these skills and their component parts, these articles utilized a wide variety of tasks and outcome tools, with very little overlap in standardized measures. However, assessment tasks used within these studies tended to follow similar structures.

Earlier studies reported similar performance on tonal and rhythmic tasks when compared to peers matched for mental age (MA) (pitch discrimination: Don et al., 1999; rhythm production: Levitin & Bellugi, 1998). However, when compared to TD peers matched for chronological age (CA), subsequent studies more consistently reported that individuals with WS tend to perform below their TD peers on both tonal and rhythm skills (pitch discrimination: Hopyan et al., 2001; Martens et al., 2010; Martínez-Castilla & Sotillo, 2014; rhythm discrimination: Hopyan et al., 2001; Martens et al., 2010; Martínez-Castilla et al., 2011; pitch production/singing: Martens et al., 2010; Martínez-Castilla & Sotillo, 2008; rhythm production: Martens et al., 2010; Martínez-Castilla et al., 2011; beat and meter perception: Lense & Dykens, 2016) and equivalent to TD peers on perception of musical expressiveness (Hopyan et al., 2001).

Tonal Skills. The tonal skills assessed in these studies encompass a variety of skills such as being able to match pitches, sing melodies accurately, distinguish if pairs of notes, chords, or melodies are the same or different, identify the number of notes in a chord, sing the final note of a melodic phrase, perceive dissonance, and detect errors in phrasing and melodic contour. (Claims about the prevalence of absolute pitch (AP) in WS have yielded mixed results and are discussed in a later section, see ‘Absolute Pitch’ section, pg. 30).

Martínez-Castilla et al. (2008) utilized both acoustical analysis and perceptual judgments from both musicians and non-musicians to assess singing skills by having participants sing two well-known songs. Individuals with WS were determined to have significantly worse singing skills than TD individuals matched for CA. Acoustical analysis revealed that individuals with WS produced significantly more tuning and interval errors (e.g., a wrong pitch), marginally more contour errors (e.g., a pitch error that falls in the opposite direction than that of the target pitch,
changing the overall contour of the melody), and had poorer key stability. However, all participants with musical training performed better than those without musical training with respect to all of these variables and tended to sing at a slower tempo, which is likely to have reduced pitch errors. Individuals with WS also made more time errors, independent of musical training. Perceptual judgments by musicians rated individuals with WS as having worse intonation, yet those with musical training were perceived to have improved intonation over those without musical training. These findings appear to bring to light the benefits of musical training. However, despite poorer performance, the musically trained individuals with WS in this study had more training than the TD controls, which might suggest that they benefit from training to a lesser degree than TD peers or that individuals with WS may need more training to reach similar levels of accomplishment. (Lense & Dykens, 2013b; Lense et al., 2013; and Martens et al., 2010 also share effects of musical training). Another study analyzing singing skills of a familiar song using different assessment measures found that ratings of singing skill were predicted by levels of amusia, duration of training, and time spent playing (Lense et al., 2013).

Martínez-Castilla, Rodriguez, & Campos (2016) examined and compared the development of pitch-related skills in individuals with WS and TD peers matched for CA. Performance on four pitch-related tasks was compared to chronological age and a battery of cognitive skills. Findings revealed that the development of pitch-related skills is atypical in WS. For the TD group, performance on all tasks improved with CA and standardized measures of cognitive development. The TD group also demonstrated a linear developmental progression of skills: pitch discrimination developed first, chord discrimination and dissonance perception developed later, followed by tonal closure. This progression is logical in that the discrimination of pitch involves processing of individual notes; discrimination of chords and perception of dissonance involves processing of multiple notes and early harmonic structure; and finally, tonal closure involves decisions based on established tonal relationships (i.e., individual pitches combine to form chords and melodies, which combine to establish key and tonality).
Individuals with WS demonstrated a less clear pattern: development of pitch discrimination preceded chord discrimination and tonal closure, yet no other developmental relationships were found, despite performance above chance on all tasks. For the individuals with WS, chronological age predicted chord discrimination; matrix reasoning predicted chord discrimination and tonal closure; and backward digit-span predicted pitch discrimination, chord discrimination, and tonal closure. These findings are also logical in that chord discrimination and tonal closure involve perception of cadential and harmonic relationships, similar to the skills necessary to perceive patterns in matrix reasoning. Also, the skills predicted by backward digit-span require auditory working memory to store and compare stimuli. Thus, although the atypical development of pitch-related skills in WS may not develop linearly or in synchrony, this may be a reflection of the atypical cognitive development in WS as the development of specific pitch skills was predicted by the requisite cognitive skills.

Given the heightened language abilities of individuals with WS and that processing of auditory language and music involves the processing of similar components (such as pitch, volume, and duration), two studies examined the relationship between musical skills and various language skills. Don et al. (1999) compared performance on a language skills battery with performance on standardized tests for tonal and rhythmic discrimination. All language measures were moderately correlated with both tonal and rhythmic musical skill. However, this was also true for the control group of TD matched by MA based on receptive vocabulary, which may indicate a developmental relationship between these domains. Musical abilities of participants with WS were also significantly associated with intelligence, but not with visuospatial abilities.

Martínez-Castilla & Sotillo (2014) examined the ability to discriminate pitch in both music and prosody. Although WS performed worse than TD on all tasks, pitch discrimination in music was significantly correlated to and predicted discrimination of prosody based on pitch only. Furthermore, pitch discrimination in prosody was not impacted or predicted by chronological age, vocabulary, or auditory memory. This suggests pitch processing in both music
and prosody utilizes similar mechanisms. When using a more complex task involving combined pitch, volume, and length of prosody, no relationship was found for either WS or TD individuals.

Although tonal skills in WS appear to more consistently fall below that of TD peers of similar CA, this may be related to a variety of factors, such as: atypical development of pitch skills, general cognitive deficits, or differences in the benefits received from formal musical training. Future studies are needed to examine the extent to which these factors influence tonal skills in individuals with WS.

**Rhythmic Skills.** The rhythm skills assessed in these studies also encompassed a variety of skills, such as being able to distinguish if pairs of rhythmic patterns are the same or different, repeating rhythmic patterns accurately, clapping in time to the beat of a musical passage, detecting changes in tempo over time, and maintaining stable tempo over a musical performance.

Earlier studies of rhythmic skills focused more on discrimination than production tasks and only assessed one of these skills at a time. Martínez-Castilla et al. (2011) presented the first study to assess both rhythmic production and discrimination skills in the same sample of adolescents and adults with WS. Given the discrepant results of previous studies, Martínez-Castilla and colleagues also sought to remediate methodological limitations of previous rhythm studies related to sample size, heterogeneity of sample, cognitive measurement, matching of control groups, and recording artifacts. Overall, individuals with WS performed significantly worse than TD controls matched for CA on both the discrimination and production tasks. Rhythm skills were also affected by cognitive level, as the difference between WS and TD individuals lost significance when controlled for IQ. This stands in contrast to Don et al. (1999), who found that individuals with WS performed at a level below their MA on a rhythm discrimination task.

Lense & Dykens (2016) extended their assessment of rhythm skills beyond discrimination and production tasks, instead examining beat and meter perception skill. Consistent with other rhythm studies in WS, both beat and meter perception skills in WS fell below that of TD matched for CA with similar levels of musical training, although the authors
reported a high degree of individual variability in both groups. Performance on beat and meter perception tasks were correlated, which makes logical sense as the perception of beat is a precursor to perceiving meter. Both of these skills were also predicted by IQ and a tendency toward a fundamental-processing style in the WS group (see ‘Auditory Processing’ section, pg. 33). Greater performance was associated with cumulative years and the number of different types of musical training. Beat and meter perception have also been implicated in various adaptive and social skills. Comparing performance on these tasks to measures of adaptive function, beat perception was significantly associated with communication (e.g., expressive, receptive, and written skills) and socialization (e.g., interpersonal relationships, play, leisure, and coping skills); meter perception was significantly associated with socialization.

Fewer studies have examined rhythm production skills than perceptual abilities for rhythm. Individuals with WS perform worse than CA-matched typically developing peers on tasks requiring them to repeat rhythmic patterns, either by singing or clapping (Martens et al., 2010; Martínez-Castilla et al., 2011). One study reported equivalent abilities in both WS and TD individuals in the ability to clap in time to the beat of a musical passage (Martens et al., 2010).

Qualitatively, Levitin & Bellugi (1998) reported a few observations following their study requiring participants to repeat various rhythmic patterns by clapping. First, individuals with WS and peers matched for MA demonstrated good temporal conservation. In other words, if a participant made an error by producing more or fewer notes than the target pattern, they also generally altered the length of the notes to match the overall length of the target. Second, when individuals with WS made an error in reproducing a rhythm, their errors were more likely to remain rhythmically consonant or compatible with the target rhythm. The authors viewed these errors as creative completions or extensions of the target pattern. This was not observed in the control group. However, Martínez-Castilla et al. (2011) reported the opposite relationship when employing a similar task using a TD control group matched for CA, whereby age-matched TD controls produced more creative completions than their WS counterparts. One explanation for this difference could be related to age, as participants with WS in Levitin and Bellugi’s study
were older/matched by MA, perhaps allowing time for more musical experiences, while in Martínez-Castilla et al.’s study the participants were matched for CA.

Overall, individuals with WS do not appear to present with preserved function in musical skills. Rather, they appear to present with ‘relatively’ good skills in relationship to their cognitive level. Interestingly, many of the authors in this section also reported anecdotal findings related to the participants with WS in their studies. Tonally, Martínez-Castilla & Sotillo (2008) reported that participants with WS sang with more ‘decoration,’ personality, expressiveness, and creativity. Rhythmically, Levitin & Bellugi (1998) reported that participants with WS demonstrated a strong sense of phrasing and meter, sensitivity to rhythmic changes, and musical creativity in rhythm (i.e., creative completions). Therefore, musical strengths in this population may fall more within their expressiveness or creativity in music than within specific formal skills.

Similar to tonal skills, rhythm skills in WS also appear to more consistently fall below that of TD peers of similar CA. However, given the discrepant findings reported here, particularly within production skills, future studies assessing both perceptual and production skills for rhythm using the same sample are needed. Also, an examination of the development of rhythmic skills in individuals with WS, similar to the developmental trajectory approach used by Martínez-Castilla et al. (2016), is needed to continue to shed light on the relationship between musical skills and other cognitive processes.

An overall limitation of the studies included in this section is the lack of control groups with other developmental delays. Given the discrepant findings between articles related to matching for CA versus MA and the relationships between musical skill and cognitive function, study of various musical skills in WS in comparison to other developmentally delayed populations may shed light on these relationships and the musical skills profile of individuals with WS.
| Author/Year                        | N   | Age \(^1\) (yrs) | Skill(s)                                      | Task(s)/Tool(s)                                                                                     | Finding(s)                                                                                     |
|-----------------------------------|-----|------------------|-----------------------------------------------|---------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------|
| **Lense & Dykens**<br>(2016)<br>(2 studies) |     |                  |                                               |                                                                                                   |                                                                                                 |
|                           | WS) 74 | WS) 26.4 ± 9.6   | Beat Perception                               | Beat Alignment Test (BAT)\(^2\): determine if a metronome matched the beat of a musical passage; Montreal Battery of Evaluation of Amusia- meter subtest (MBEA-m)\(^3\); determine if a melody was in duple or triple meter | WS < TD CA on both beat and meter perception; High degree of individual variability in both groups |
|                           | TD) 52 | TD) 24.3 ± 9.4   | Meter Perception                               |                                                                                                   |                                                                                                 |
|                           | WS) 50 | WS-BAT\(^2\)<br>26.2 ± 8.4<br>WS-MBEA\(^3\)<br>26.8 ± 8.3 | Beat Perception Meter Perception | Compared measures of beat and meter perception to measures of cognitive and adaptive function (Vineland-II) |                                                                                                 |
| **Martínez-Castilla, Rodríguez, & Campos**<br>(2016) |     |                  |                                               |                                                                                                   |                                                                                                 |
|                           | WS) 20 | WS) 6-17         | Pitch discrimination                           | Compared performance pitch related skills to CA and standardized measures of cognitive development |                                                                                                 |
|                           | TD) 54 | TD) 4-17         | Chord discrimination Dissonance perception Tonal closure |                                                                                                   |                                                                                                 |
| **Martínez-Castilla & Sotillo**<br>(2014) |     |                  |                                               |                                                                                                   |                                                                                                 |
|                           | WS) 14 | WS) 8-17         | Pitch discrimination                           | Compared pitch processing in both music and prosody discrimination                               |                                                                                                 |
|                           | TD) 26 | TD) 8-17         |                                               |                                                                                                   |                                                                                                 |
| **Martínez-Castilla, Sotillo, & Campos**<br>(2011) |     |                  |                                               |                                                                                                   |                                                                                                 |
|                           | WS) 20 | WS) 20.10 (5.87)<br>Range: 12-32<br>TD) 20.03 (6.20) | Rhythm discrimination Rhythm production | Same/different discrimination task Echo clapping task                                           |                                                                                                 |
| **Martens, Reutens, & Wilson**<br>(2010) |     |                  |                                               |                                                                                                   |                                                                                                 |
|                           | WS) 25 | WS) 8-41         | Various tonal and rhythm skills in discrimination and production | Specimen Aural Test (SAT)\(^4\) Bentley Measures of Musical Abilities\(^5\) |                                                                                                 |
|                           | TD) 25 | TD) 8-41         |                                               |                                                                                                   |                                                                                                 |
| Author/Year | N | Age<sup>1</sup> (yrs) | Skill(s) | Task(s)/Tool(s) | Finding(s) |
|-------------|---|-----------------------|----------|-----------------|------------|
| Martínez-Castilla & Sotillo (2008) (2 studies) | WS) 7 TD 7 | WS) 10-30 TD 10-30 | Singing | Singing a familiar song, measured using both acoustical analysis and perceptual judgments by musicians/non-musicians | WS < TD CA singing skill; Those with musical training performed better; WS may benefit less from musical training than TD |
| | WS) 15 TD 15 | WS) 17-32 TD 17-32 | Pitch Matching Singing | Pitch matching task; Singing a familiar song, measured using both acoustical analysis and perceptual judgments by musicians/non-musicians | |
| Hopyan et al. (2001) | WS) 14 TD 14 | WS) 12 TD 12 (3) | Pitch discrimination Rhythm discrimination Melodic imagery Phrasing perception | Primary Measures of Music Audiation (PMMA)<sup>6</sup>; Musical Aptitude Profile (MAP)<sup>7</sup> | WS < TD CA on pitch and rhythm discrimination; WS = TD CA on perception of musical expressiveness |
| Don, Schellenberg, & Rourke (1999) | WS) 19 TD 19 | WS) 8-13 TD 5-12 | Pitch discrimination Rhythm discrimination | Primary Measures of Music Audiation (PMMA)<sup>6</sup> | WS = TD MA on pitch discrimination WS < TD MA on rhythm discrimination; Music and language skills moderately correlated |
| Levitin & Bellugi (1998) | WS) 8 TD 8 | WS) 9-20 TD 5-7 | Rhythm production | Echo clapping task | WS = TD MA |

CA = Chronological Age; MA = Mental Age; TD = Typically Developing Control; WS = Williams Syndrome

<sup>1</sup> Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years

<sup>2</sup> BAT = Beat Alignment Test (Iversen and Patel, 2008);
<sup>3</sup> MBEA-m = Montreal Battery of Evaluation of Amusia- meter subtest (Peretz, Champod, & Hyde, 2003);
<sup>4</sup> SAT = Specimen Aural Test (Nickson & Black, 1962);
<sup>5</sup> Bentley = Bentley Measures of Musical Abilities (Bentley, 1985);
<sup>6</sup> PMMA = Primary Measures of Music Audiation (Gordon, 1985);
<sup>7</sup> MAP = Musical Aptitude Profile (Gordon, 1995)
Emotional Responsiveness

Twelve articles reported findings related to emotional responsiveness to music (Table 6). These findings were collected using a variety of measures, including: parent report via questionnaires and interviews, various affect identification tasks, and measures of autonomic reactivity to musical and emotional stimuli.

Of the studies that relied on parent report through various questionnaires or interviews (such as the MIS or SAMMI), the most frequently cited findings included an overall heightened emotional responsiveness to music, such as more intense and longer lasting emotional reactions, when compared to both TD peers and others with neurodevelopmental disorders (Don et al., 1999; Levitin et al., 2004). Other findings included a greater range of emotional responses to music (Don et al., 1999, Dykens et al., 2005); a significant correlation between emotional responsivity to music and social-emotionality (i.e., identification of another’s emotions, desire to please, empathy, etc.) (Ng et al., 2013); and emotional responsiveness was most predicted by auditory sensitivities (Lense et al., 2013).

On tasks involving the identification of the emotional valence of various auditory or visual stimuli, individuals with WS tended to perform with mixed results. Some studies documented that individuals with WS tended to perform comparatively to TD matched for CA (auditory: Bhatara et al., 2010; Järvinen et al., 2016; visual: Lense, Gordon, et al., 2014, Järvinen-Pasley et al., 2010) while others reported poorer performance compared to TD (auditory: Hopyan et al., 2001; Järvinen et al., 2012; visual: Bhatara et al., 2010). However, individuals with WS tended to perform more accurately than individuals with ASD or other intellectual disabilities (Bhatara et al., 2010; Järvinen-Pasley et al., 2010). Within both the visual and auditory domain, WS exhibit a bias toward increased competence in identifying social (i.e., faces and voices) over non-social (i.e., images and music) affective stimuli (Järvinen et al., 2012; Järvinen et al., 2016; Järvinen-Pasley et al., 2010).

Lense, Gordon, et al. (2014) examined the influence of a brief musical excerpt on the identification of visual affect. Although the emotional valence of the musical prime didn’t have
an effect on the accuracy in identifying the valence of the facial stimuli, individuals with WS demonstrated significantly faster reaction time when the valence of the target face matched the preceding music. Similarly, Järvinen-Pasley et al. (2010) also showed a similar lack of improvement in accuracy for emotionally congruent over incongruent auditory/visual stimuli; however this could possibly be due to the exaggerated interest in faces in WS.

As expected, individuals with WS reported positive emotional states in response to positively valenced music. However, the same individuals reported personally experiencing both positive and negative emotional states in response to negatively valenced music (Dykens et al., 2005). This resonates with a consistent finding across studies that individuals with WS are more accurate in identifying the affective valence of positive (happy) over negative (sad or fear) stimuli (Hopyan et al., 2001; Järvinen et al., 2012; Järvinen et al., 2016) and rate intensity of positive affect as more intense (Järvinen-Pasley et al., 2010). These findings suggest a possible lack of awareness or differentiation between more complex emotions such as fear, worry, sadness, and loneliness in individuals with WS. Future studies are needed to examine reports of these more complex emotions and the perceptions or effects of more nuanced emotional states in music with individuals with WS.

A few studies examined responses of various autonomic systems to musical and auditory stimuli. Järvinen et al. (2012) found greater heart rate variability in response to vocal affect compared to musical affect, suggesting increased arousal to social auditory stimuli. Using a similar paradigm, Järvinen et al. (2016) found similar variability in heart rate along with significantly increased electrodermal activity in response to musical stimuli in the WS group, a pattern not found in the TD or ASD groups, suggesting further autonomic arousal in response to music. The WS and ASD groups also exhibited diminished habituation to both vocalizations and music over time, indicating both differential and sustained arousal compared to TD. In another autonomic area, Dai et al. (2012) found an increase in levels of oxytocin and arginine vasopressin in response to music in individuals with WS, despite elevated basal levels of these neuropeptides. However, these responses were also seen, although to a lesser extent for
vasopressin, in response to a cold pressor test (placing a hand in ice cold water). Taken together, these findings suggest that disruptions or dysregulation in autonomic reactivity to music may relate to the enhanced emotional responsiveness to music found in WS.

A strength of the articles included in this section is the inclusion of multiple types of control groups. However, one limitation in comparing reports of emotional responsiveness in WS to others with neurodevelopmental disorders is the potential for responses to be impacted by the social and expressive nature of individuals with WS. Individuals with WS are more verbally expressive compared to others with neurodevelopmental disorders, which could lead parents of individuals with WS to be more aware of their internal experiences and reactions to music.

It is possible that some of the responses to music reported in this section are related to the presence of auditory stimuli in general, rather than specifically to the presence of music. Studies examining the role of auditory processing in emotional responses to music are needed.
| Author/Year | N | Age (yrs) | Task(s) | Finding(s) |
|-------------|---|----------|--------|------------|
| Järvinen et al. (2016) | WS) 12 | WS) 10-14 | Affect identification task (auditory) | WS = TD CA = ASD identifying auditory affect; WS Social > Non-social affect; WS increased arousal (HR variability and EDA) to vocalizations and music; WS diminished habituation to both vocalizations and music |
| Lense, Gordon, et al. (2014) | WS) 13 | WS) 27.1 +/- 7.1 | Affect identification task (visual) With auditory prime | WS = TD CA; WS faster reaction time when visual and auditory stimuli congruent > incongruent |
| Lense, Shivers, & Dykens (2013) | WS) 73 | WS) 10-51 | Musicality Interest Scale (MIS)^2 | Emotional responsiveness was predicted by auditory sensitivities |
| Ng et al. (2013) | WS) 55 | WS) 16-52 | Compared measures of musicality, sociability, and language comprehension. | Significant correlation between emotional responsivity to music and social-emotionality |
| Dai et al. (2012) | WS) 13 | WS) 19-42 | OT and AVP measured during music and cold pressor test | WS higher baseline OT; Increased OT and AVP in response to music and cold pressor |
| Järvinen et al. (2012) | WS) 20 | WS) 13-46 | Affect identification task (auditory) | WS < TD CA identifying auditory affect; WS Social > Non-social affect; WS increased arousal (HR variability to vocal affect > music affect |
| Bhatara et al. (2010) | WS) 11 | WS) 13-43 | Affect identification task (auditory- expressivity in musical performance) | WS = TD CA; WS > ASD Recognizing emotion in musical performance |
| Järvinen-Pasley et al. (2010) | WS) 21 | WS) 12-40 | Affect identification task (visual) With musical prime | WS = TD CA (social) WS < TD CA / WS > DD WS Social > Non-social affect; No difference when visual and auditory stimuli congruent > incongruent |
### Table 6 cont.

**Emotional Responsiveness**

| Author/Year | N     | Age\(^1\) (yrs) | Task(s)                                                                 | Finding(s)                                                                 |
|-------------|-------|------------------|-------------------------------------------------------------------------|----------------------------------------------------------------------------|
| Dykens et al. (2005) (2 studies) | WS) 31 | WS: 10.22 (4.86)  | Compared measures of problem behaviors and musicality                   | n/a (study 2 only for this area)                                           |
|             | PW) 26 | PW: 10.26 (4.86)  |                                                                          |                                                                            |
|             | DS) 32 | DS: 11.5 (4.49)   |                                                                          |                                                                            |
|             |       | All) Range 4-21   |                                                                          |                                                                            |
|             | WS) 26 | WS: 20.88 (11.48) | Compared measures of fears, anxieties, problem behaviors, and musicality | Greater range of emotional responses to music; WS reported experiencing both positive and negative emotions in response to negatively valenced music |
|             | PW) 16 | PW: 19.38 (6.70)  |                                                                          |                                                                            |
|             | DS) 25 | DS: 18.83 (7.11)  |                                                                          |                                                                            |
|             |       | All) Range 8-47   |                                                                          |                                                                            |
| Levitin et al. (2004)          | WS) 118| WS: 20.4 (10.4)   | Salk McGill Music Inventory (SAMMI)\(^3\)                              | High levels of emotional responsiveness; Emotional effects of music listening last longer in WS |
|             | TD) 118| TD: 20.9 (7.4)    |                                                                          |                                                                            |
|             | ASD) 30| ASD: 18.2 (7.7)   |                                                                          |                                                                            |
|             | DS) 40 | DS: 17.2 (9.2)    |                                                                          |                                                                            |
|             |       | Range: 5-51       |                                                                          |                                                                            |
| Hopyan et al. (2001)            | WS) 14 | WS: 12 (3)        | Affect identification task (auditory)                                   | WS < TD CA identifying auditory affect;                                    |
|             | TD) 14 | TD: 12 (3)        |                                                                          |                                                                            |
| Don, Schellenberg, & Rourke (1999) | WS) 19 | WS: 8-13          | Parent Questionnaire/Interview                                          | High levels of emotional responsiveness; Greater range of emotional responses to music |
|             | TD) 19 | TD: 5-12          |                                                                          |                                                                            |

ASD = Autism Spectrum Disorder; AVP = Arginine Vasopressin; CA = Chronological Age; DD = Developmentally Delayed Control; DS = Down Syndrome; EDA = Electrodermal Activity; HR = Heart Rate; OT = Oxytocin; PW = Prader-Willi Syndrome; TD = Typically Developing Control; WS = Williams Syndrome

\(^1\) Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years

\(^2\) Musicality Interest Scale (MIS): Questionnaire with subscales in: 1) interest in and liking of music, 2) emotional reactions to music, and 3) musical skills.

\(^3\) Salk McGill Music Inventory (SAMMI): Comprehensive questionnaire with subscales in: 1) demographic information, 2) interest in music, 3) emotional responsiveness to music, 4) music creativity and reproduction, 5) musical training, 6) age of onset of musical behavior.
Musical Processing: Absolute Pitch, Amusia, and Auditory Processing

Four articles included findings related to various aspects of musical processing (Table 7).

**Absolute Pitch.** Absolute pitch (AP) is the rare ability to identify or produce the pitch of a sound without use of a reference tone. This particular skill occurs in 1 in 10,000 people, more commonly in cultures with tonal languages, and its acquisition is often associated with early musical exposure and training between the ages of 3-6 years (Takeuchi & Hulse, 1993).

Two of the included studies examined absolute pitch, presenting contradictory results. Lenhoff et al. (2001) tested five musically trained individuals with WS for abilities of AP. These individuals performed near ceiling levels on traditional measures for AP, leading authors to conclude that the prevalence of AP is higher among individuals with WS than the general population. Martínez-Castilla, Sotillo, & Campos (2013) calculated that, given the prevalence of WS in the US/Canada, this could indicate that the incidence/prevalence could be 10 times greater than typically developing populations! Lenhoff et al. also suggested that the critical period for developing AP may be extended in WS, given that all of the individuals in the study began their musical training after the proposed critical period for the general population.

The study of AP in individuals with WS presents some difficulty with respect to their cognitive limitations. Traditional measures of assessing AP require participants to be able to label pitches using traditional musical nomenclature (i.e., note names). Most musically trained individuals with WS do not learn music formally or learn how to read sheet music, making it difficult to conduct accurate assessments with a suitable sample size. Results from Lenhoff et al. (2001) may need to be interpreted with caution as the small sample of 5 participants all had musical training, were able to label musical notes, and therefore may not be representative of the larger WS population.

Martínez-Castilla et al. (2013) attempted to overcome this limitation by utilizing both traditional measures for AP and a novel paradigm examining participants’ long-term memory for target stimuli. The latter does not require the ability to label musical notes and allowed for the study of a much larger sample, including participants with and without musical training.
Performance on both measures was near chance for both the WS and control groups, contrasting the near ceiling level performance on both measures by 2 self-reported AP possessors. These results indicated that the incidence/prevalence of AP in WS is not remarkable. Additionally, performance on both measures was not associated with cognitive ability or musical training. Since all of the participants with WS began their musical training late and did not develop AP, Martínez-Castilla and colleagues refuted the findings of Lenhoff et al. (2001) related to the extended critical period for the development of AP.

Martínez-Castilla et al. (2013) pointed to minor differences between their study and that conducted by Lenhoff et al. (2001), including use of different pitch registers and differences in the number of items presented, however they argued these are unlikely to have had an effect on the results as modifications made were likely to make the task easier. Other possible explanations for the variation between the two studies could be explained by differences in musical training and cognitive level between the two samples. However, detailed information on the musical training was not offered by Lenhoff et al. Also, although overall IQ was reported in both studies, Lenhoff et al. did not report their instrument used to assess cognitive functioning and the variability in cognitive functions in WS is widely known (see Martens et al. 2008, for review).

Some additional neuroanatomical evidence is also often cited when discussing AP in WS. In typically developing populations, musicians with AP have shown a stronger leftward asymmetry in the planum temporale when compared to musicians without AP or non-musicians (Schlaug, Jäncke, Huang, & Steinmetz, 1995). This anatomical correlate was reported in individuals with WS by Hickok, Bellugi, & Jones (1995). However, subsequent reports have not replicated this finding (Galaburda & Belugi, 2000) while others have found the opposite pattern (Chiang et al., 2007; Eckert et al., 2006).

The variability in behavioral and neuroanatomical findings within WS speaks to the need for continued inquiry. Future studies should investigate AP using non-traditional tasks that do not require participants to label musical notes, such as the one outlined in Martínez-Castilla et al.
(2011) or through the use of musical instruments. These studies should also examine left planum temporale volumes and their associations to performance on AP tests.

**Amusia.** On the opposite end of the spectrum, amusia is the inability to recognize or reproduce musical tones, despite typical cognitive abilities or exposure to music. Individuals with amusia have difficulty with basic music tasks such as recognizing and discriminating melodies, singing, distinguishing between meters, tapping along with a beat, and have poor musical memory (Sloboda et al., 2005). Amusia can be acquired through neurological damage; however congenital amusia is prevalent in 1.5-4% of the population (Kalmus & Fry, 1980; Peretz & Vuvan, 2017).

Lense et al. (2013) assessed a large sample (n = 73) of individuals with WS for characteristics of amusia using a battery of assessments including the Distorted Tunes Test (DTT; Drayna, Manichaikul, de Lange, Snieder, & Spector, 2001), a test for pitch amusia whereby participants heard a series of well-known songs, some of which were altered to have note errors (the contour and rhythm was preserved), and were asked if the song was played correctly. Eleven percent of participants met the criteria for amusia. Performance on the DTT was not associated with age, sensitivity to sound, family musical environment, and time spent playing/listening to music. It was moderately associated with IQ, number of types of musical training, cumulative duration of training, singing skill, and musical interest. DTT performance was negatively associated with a measure of spectral/fundamental processing (i.e., poorer DTT = generally mixed or somewhat spectral processing, see next section). Only musical training was a predictor of DTT performance and those who scored better on the DTT made fewer interval deviations, fewer contour errors, and sang at a slower tempo during a solo musical performance.

Although the DTT is the only outcome tool for amusia reported in this section, the Montreal Battery of Amusia (MBEA; Peretz, Champod, & Hyde, 2003), another tool commonly used in the assessment of amusia, was utilized in another study (Lense, Dankner, Pryweller, Thornton-Wells, & Dykens, 2014). Lense and colleagues argue that the DTT is a better task for WS given its highly engaging stimuli and the fact that the MBEA uses same/different tasks,
which requires working memory, which is impaired in WS. Future studies are needed to
determine the most suitable assessment for amusia for individuals with WS. Future studies are
also needed to examine more of the temporal qualities of music as they relate to amusia.

**Auditory Processing (Global/Local and Spectral/Fundamental Perception).** Musical
sound perception is an incredibly complex phenomenon, with a high degree of individual
variability. In the general population, there is an even distribution between those who perceive
sound by decomposing it into its harmonics (spectral processing) and those who perceive sound
based on the fundamental frequency (fundamental processing). Wengenroth et al. (2010)
reported an extreme and nearly-uniform fundamental or holistic processing bias in individuals
with WS, which is a marked deviation from the even distribution found in TD individuals in the
control group.

Two studies (Lense & Dykens, 2016; Lense et al., 2013) have repeated this Spectral
Fundamental Processing task (SPF; Schneider et al., 2005; Wengenroth et al., 2010). Lense et al.
(2013) reported a range from extreme fundamental processing to somewhat spectral, with a mean
of somewhat fundamental, in a sample of 73 individuals with WS. Lense & Dykens (2016) did
not report the results of the SPF with their sample of 74 individuals with WS. However, beat and
meter perception skills were predicted by a fundamental-processing style. Also, all of
participants with WS identified as amusic (n = 8) within the study scored between the two
processing styles, suggesting that a lack of a distinct processing style might contribute to
perceptual deficits. Future studies are needed to examine the relationships between processing
style and perceptual skills.

Deruelle, Schön, Rondan, & Mancini (2005) examined tendencies for global and local
perception of musical elements in children with WS by presenting pairs of melodies and asking
participants if the two melodies were the same or slightly different. Some of the melodies in the
global condition included a single pitch change that violated the overall contour of the first
melody, representing a disruption in the global property of the melody. Melodies in the local
condition included a similar pitch change; however this change was consistent with the overall
contour of the melody and represented a change to the local properties of the melody. Individuals with WS performed similarly on both tasks: their performance was consistent with the control group for the local condition but far below the control group for the global condition. This deficit in global processing stands in contrast to the general advantage for global over local processing of musical stimuli in TD children (Ouimet, Foster, & Hyde, 2012) and is consistent with perceptual deficits for visuospatial stimuli in individuals with WS (Bihrlé et al., 1989; Farran, 2005). These results have been replicated with adults with WS (Elsabbagh, Cohen, & Karmiloff-Smith, 2010).

Although the studies in this section reported findings in different areas of auditory and musical perception, a consistent theme across these studies is the pervasiveness of atypical auditory processing in individuals with WS. Although these divergent auditory processes may present with a high level of variability within WS, the pervasiveness with which they are reported may account for some of the unique affinity for music noted in the population.
### Table 7
**Musical Processing: Absolute Pitch, Amusia, and Auditory Processing**

| Author/Year | N  | Age\(^1\) (yrs) | Topic(s) | Task(s) | Finding(s) |
|-------------|----|-----------------|----------|---------|------------|
| Lense, Shivers, & Dykens (2013) | WS) 73 | WS) 10-51 | Amusia; Auditory Processing | Distorted Tunes Test (DTT); Spectral/Fundamental Processing Task (SPF) | Higher incidence of amusia in WS compared to general population; Amusia strongly predicted musical skill; Overall tendency toward fundamental processing in WS |
| Martínez-Castilla, Sotillo, & Campos (2013) (2 studies) | WS) 7 | Range: 15-32 | Pitch identification task: Label pitches without use of a reference tone | Prevalence of AP in WS is not higher than the general population; Both WS and TD performed equally and near chance |
|            | TD) 14 | WS Trained) 21.96 (6.8) |        | Pitch memory task: Discriminate if two tones were same/different following a retention interval filled with a distracting melody | Discriminate if two melodies were same/different; ‘Different’ melodies had errors that either violated or preserved the overall contour of the previous melody |
|            |     | Range: 15-32 |        |        | Deficits in global rather than local perception of auditory stimuli in WS |
|            | AP) 2 | TD Untrained) 21.96 (5.7) |        |        | Battery of tasks for absolute and relative pitch: identifying single notes; identifying natural notes in harmonic dyads/triads; pitch production and transposition |
|            |     | Range: 14 & 16.4 |        |        | Higher prevalence of AP in WS than the general population; The critical period for acquisition of AP may be extended in WS |
| Deruelle et al. (2005) | WS) 16 | Range: 12y7m (4y) |             |        |        |
|            | TD) 16 | Range: 8y7m - 19y3m |             |        |        |
|      |     | TD) 13y5m (3y7m) |             |        |        |
| Lenhoff, Perales, & Hickok (2001) | WS) 5 | WS) 13-43 |             |        |        |

\(^1\)Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years

\(^2\)DTT = Distorted Tunes Test (Drayna, Manichaikul, de Lange, Snieder, & Spector, 2001)

\(^3\)SPF = Spectral/Fundamental Processing Task (Schneider et al., 2005; Wengroth et al., 2010)
Brain Imaging and Morphology

Five of the included articles employed various neuroimaging tests (Table 8), including magnetic resonance imaging (MRI), functional magnetic resonance imaging (fMRI), diffusion tensor imaging (DTI), and electroencephalogram (EEG).

Using structural MRI, Martens et al. (2010) found similar planum temporale and primary auditory cortex volumes between individuals with WS and typically developing controls, despite significantly smaller overall brain volume in WS. Bilateral planum temporale volumes were significantly larger in individuals with WS and no significant difference in asymmetry was found. Left planum temporale volume was greater in a subset of WS participants who performed better on various musical production tasks (SAT, Nickson & Black, 1962) and was positively correlated with the ability to sing the final note of a melodic phrase. Although larger, the difference in primary auditory cortex volume was not significant.

Lense, Dankner, et al. (2014) examined neural correlates of amusia in a sample of individuals with WS, some of which met the criteria for amusia. Diffusion tensor imaging (DTI) revealed that individuals with amusia displayed decreased connectivity along the superior longitudinal fasciculus (SLF), a fiber pathway that connects the temporal and frontal lobes. This remained significant after controlling for musical training, suggesting that amusia is not a result of a lack of musical experiences. Lense, Dankner, et al. confirmed that these findings are highly consistent with previous research on amusia in TD populations and suggest amusia is related to poor connectivity rather than dysfunction in the primary auditory cortex in both populations.

Two functional magnetic resonance imaging (fMRI) studies examined individuals with WS while undergoing various listening tasks. Levitin et al. (2003) examined auditory and emotional areas in a small sample of individuals with WS and TD controls while they listened to various excerpts of classical music, noise, and silence. Results revealed remarkable differences between the two groups, indicating more variable and widely diffuse activations in WS participants in contrast to the well-defined activations of TD controls. Individuals with WS demonstrated significantly reduced activation in the temporal lobes (superior temporal gyrus,
medial temporal gyrus, superior temporal sulcus); whereby all of these areas were found to be areas of activation for TD. Significant bilateral temporal lobe activation was evident for both groups, although to a lesser degree for WS, while listening to noise, demonstrating the ability to distinguish between music and noise processing on a neuroanatomic level. A notable finding in the WS group was significantly greater activation in the right amygdala, suggesting a possible neural correlate for the heightened emotional reactions to music in WS. Other findings included consistent activation of the cerebellum, pons, and brainstem in the WS group.

In a similar task, Thornton-Wells et al. (2010) found 19 significant clusters of activation that were different between the WS and TD groups. Similar increased activations as Levitin et al. (2003) were found in bilateral cerebellum and emotional areas (insula, parahippocampal gyrus, posterior cingulate). Amygdala activation was found, however it did not reach significance. In contrast, increased activations in the bilateral superior temporal gyrus were also found. However, findings revealed activation of occipital and early visual areas in response to music and other auditory stimuli, most notably to simple notes and chords. These findings were consistent across two follow-up studies using subsets of participants. This activation of visual areas in response to auditory stimuli demonstrates a cross-modal processing of auditory information somewhat similar to synesthesia, although to a lesser degree.

Lense, Gordon, et al. (2014) measured EEG oscillatory activity during an affective priming task, whereby participants heard brief emotionally valenced musical excerpts and were asked to make judgments about the emotional valence of facial stimuli. Individuals with WS demonstrated greater evoked alpha power in response to happy musical excerpts than compared to sad, which reflects sensory processing and attentional control. This activity was positively associated with parental ratings of emotional reactions to music on the Musical Interest Scale (MIS; Blomberg et al., 2006). Individuals with WS also demonstrated significantly greater evoked gamma activation in response to facial stimuli that matched the valence of the musical prime, which is believed to be associated with integration across sensory modalities.
Conducting neuroimaging tests such as fMRI present unique challenges given the incidence of anxiety and hearing sensitivities in individuals with WS. Both Levitin et al. (2003) and Thornton-Wells et al. (2010) sought to mitigate these issues and thus optimize their results by employing similar strategies to systematically orient their WS participants to the fMRI procedure. This involved various supports such as sending participants audio and video recordings of the fMRI machine to introduce them to the sounds of the machine, use of an fMRI simulator, interacting with the machine and staff prior to the scan, and speaking to an individual with WS who had previously completed multiple scans. Such supports may be beneficial in future neuroimaging studies.

At face value, many of these findings suggest a neurological basis for the unique affinity for music found in individuals with WS, including the effects of more diffuse neurological activation, activation of emotional centers, and cross-modal activation of auditory and visual/affective areas in response to music on the enhanced attraction to sounds and music, greater interest in music, and heightened emotional responsiveness to music. At a fundamental level, these studies also provide further support for the pervasiveness of differential or atypical processing of auditory information in individuals with WS.

Limitations of articles included in this section speak to the need for controls with other developmental disabilities. Also, given the small number of studies included in this section, the need for replication using various methods is evident. Given the pervasiveness of atypical auditory processing in individuals with WS, future neuroimaging studies would benefit from a broader consideration of a priori regions to include regions involved in the entire network of auditory processing.
| Author/Year                  | N    | Age\(^1\) (yrs) | Imaging | Areas Examined                                      | Findings(s)                                                                 |
|-----------------------------|------|-----------------|---------|---------------------------------------------------|-----------------------------------------------------------------------------|
| Lense, Dankner, et al. (2014) | WS) 17 | WS) 16-48       | MRI     | Primary auditory cortex (STG, TTG); Pars orbitalis of IFG and SLF | Decreased connectivity along the superior longitudinal fasciculus        |
|                             | TD) 13 | TD) 16-48       | DTI     |                                                   |                                                                             |
| Lense, Gordon, et al. (2014) | WS) 13 | WS) 27.1 +/- 7.1 | EEG     | n/a                                              | Increased evoked alpha in response to happy vs sad; Increased evoked gamma in response to congruent affective stimuli |
|                             | TD) 13 | TD) 27.7 +/- 6.0 |         |                                                   |                                                                             |
| Martens, Reutens, & Wilson (2010) | WS) 25 | WS) 8-41        | MRI     | Primary auditory cortex Planum temporale         | Larger bilateral planum temporale, no difference in asymmetry             |
|                             | TD) 25 | TD) 8-41        |         |                                                   |                                                                             |
| Thornton-Wells et al. (2010) | WS) 13 | WS) 16-33       | MRI     | Not stated, appears to be entire brain, no a priori | Activation of occipital and early visual areas in response to music       |
|                             | TD) 13 | TD) 17-27       | fMRI    |                                                   |                                                                             |
| Levitin et al. (2003)       | WS) 5 | WS) 28.8 (14.6) | fMRI    | STG, MFG, SFG, cerebellum, amygdala, cingulate gyrus, pons | More diffuse activation; Decreased temporal lobe activation; Increased right amygdala activation |

DTI = Diffusion Tensor Imaging; EEG = Electroencephalogram; fMRI = Functional Magnetic Resonance Imaging; MFG = medial frontal gyrus; MRI = Magnetic Resonance Imaging; IFG = inferior frontal gyrus; SFG = superior frontal gyrus SLF = superior longitudinal fasciculus; STG = superior temporal gyrus; TD = Typically Developing Control; TTG = transverse temporal gyrus; WS = Williams Syndrome

\(^1\)Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years
Cognitive Processes: Memory and Math

Three of the included articles examined the relationships between music and various cognitive processes (Table 9).

Memory. Two of the included articles examined the effects of melody on word learning as a facet of verbal memory. Martens et al. (2011) presented two sets of participants with WS with a memory task requiring them to recall a list of sentences that were either spoken or sung to a familiar melody. Participants with prior musical training, in the form of private lessons, demonstrated significantly better long-term recall of the sentences when they were sung compared to when they were spoken. This was not observed for participants without musical training and was not correlated to musical enjoyment, time spent listening to music as a child, or heightened emotional reactions to music. A follow-up study examining both short and long-term verbal memory found identical results for long-term recall and no effect on short-term recall. Authors also noted that participants exhibited greater attention and were more still during conditions in which the sentences were sung, regardless of musical training.

Dunning et al. (2015) found nearly identical results when the sentences were sung to a novel melody. In this study, those with formal musical training performed significantly better with sung or spoken sentences than those without musical training. Although not significant, participants without musical training also performed better when the information was presented through song. This study also examined the musical training of their sample in greater detail. Performance on the memory task was not impacted by age, verbal IQ, musical enjoyment, length of time listening to music per day, or length of participation in music lessons, choir, or music therapy.

Math. As part of the Music & Minds program, a 10-day intensive music program for individuals with WS, Reis et al. (2003) reported a positive influence of music on acquisition of math skills. Participants attended two daily math sessions focused on proficiency with fractions, including practical applications to time, money, measurement, musical notes, and objects. This program incorporated music as both a learning tool and instructional methodology throughout
the duration of the program. Authors reported that a majority of the participants increased their understanding of the covered math concepts and expressed increased confidence related to fractions following the program. Unfortunately, details on the instructional and assessment methods used were not reported.

Although available literature in this section is sparse, together these findings indicate a potential for clinical intervention addressing cognitive processes using music. Also, anecdotal reports of increased attention during musical tasks and confidence following a musical program speak to the merit of the inclusion of music to support the acquisition of cognitive skills in WS.
| Author/Year                          | N   | Age\(^1\) (yrs) | Topic(s)     | Task(s)                                                                 | Finding(s)                                                                 |
|-------------------------------------|-----|-----------------|--------------|--------------------------------------------------------------------------|---------------------------------------------------------------------------|
| Dunning, Martens, & Jungers (2015)  | WS) 44 | WS) 8-48        | Verbal Memory| Recall a list of sentences that were either spoken or sung to a novel melody | WS with musical training demonstrated improved verbal memory on both sung and spoken conditions |
| Martens, Jungers, & Steele (2011)   | WS) 38 | WS) 6-59        | Verbal Memory| Recall a list of sentences that were either spoken or sung to a familiar melody | WS with musical training demonstrated improved verbal memory when sentences were sung > spoken |
| (2 studies)                         |     |                 |              |                                                                          |                                                                           |
|                                     | WS) 38 | WS) 7-50        |              |                                                                          |                                                                           |
| Reis et al. (2003)                  | WS) 16 | Unclear; Only DOB given | Math Skills | 10-day music and math curriculum focused on understanding fractions | Majority of participants increased their understanding of math concepts; Expressed increased confidence in math |

WS = Williams Syndrome

\(^1\) Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years
Fears, Anxieties, and Problem Behaviors

Three of the included articles examined relationships between music and fears, anxieties, and problem behaviors in individuals with WS (Table 10), which are known to be phenotypic characteristics of the population. Two of the studies relied mostly on correlational or associative measures using various questionnaires and assessments. Blomberg et al. (2006) collected questionnaire data from two separate familiar respondents of 38 subjects with WS. Many significant correlations were found between measures of hyperacusis and fears. Data collected on fears using the Fear Survey Schedule for Children-Revised (FSSC-R; Ollendick, 1983) revealed high levels of fears, which was consistent in level and types of fears with previous research (Dykens, 2003). Thirteen percent of the participants scored above the suggested cutoff for hyperacusis, which is five times greater than the percentage found in the general population using the same outcome measure (Hyperacusis Questionnaire (HQ); Khalfa et al., 2002). However this is much lower than previous reports of prevalence of hyperacusis in WS (Klein et al., 1990; Gothelf et al., 2006) and consistent with studies using narrower definitions of hyperacusis (Levitin et al., 2005).

Few associations were found between musicality and hyperacusis or fears. A weak significant negative correlation was found between musical skill and the ‘failure and criticism’ subscale of the FSSC-R, indicating that musically-accomplished individuals with WS are slightly more resilient to perceptions of criticism. A weak significant positive correlation was found between emotional reactions to music and measures of hyperacusis. This finding is contrary to the early hypothesis concerning the protective factors of musicality in preventing or managing anxiety. However, it is also consistent with findings from Lense et al. (2013) that emotional responsiveness was predicted by auditory sensitivities in individuals with WS.

Using similar measures Dykens et al. (2005) found that lower levels of fears and anxiety were associated with increased frequency, duration, and skill in producing music. Results using the Child Behavior Checklist (CBCL; Achenbach, 1991) indicated that, for individuals with WS, externalizing symptoms (such as aggressive behaviors or noncompliance) were negatively
correlated with frequency of listening to music, and internalizing symptoms (such as anxiety, depression, withdrawal, somatic complaints, etc.) were negatively correlated with producing music. Independent of musical skill, engaging in producing or listening to music may have positive effects on maladaptive behaviors. Also, although music may not be effective in preventing anxiety, it may assist in managing its symptoms.

Overall, these two studies share similar limitations in that they are reliant on parent report, which poses possible bias and the risk for underestimation related to mental health information. The results of these studies are also correlational and do not indicate if these associations are the product or cause of fears and anxiety. Finally, it is worth noting that fear and anxiety are not the same, as fear is more of an immediate response and anxiety is a more future-oriented process. Conclusions about music’s relationship to these may be different in mitigating the future-oriented aspects of anxiety as compared to the immediate experience of fear.

Lense & Dykens (2013a) examined cortisol reactivity during a solo musical performance in individuals with WS as a measure of psychological stress. Increases in cortisol are often seen in typically developing populations during times of social stress and evaluation, such as public performances or presentations (Beck, Cesario, Yousefi, & Enamoto, 2000; Taylor et al., 2010). Cortisol measures, taken prior to and 20 minutes following a prepared solo musical performance in front of a live audience of familiar and unfamiliar individuals, remained stable and showed no significant change. Although this could be interpreted as a lack of psychosocial stress related to a musical performance, it is also possible that the participants in the study could have experienced significant anticipatory anxiety in preparation for the performance and thus exhibited higher baseline cortisol, which could account for the lack of reactivity following the performance. Baseline cortisol was found to be strongly associated with musical skill, as rated by judges during the performance, but not with music listening or anxiety. As it is hard to draw conclusions from a lack of an identified effect, future studies are needed to clarify this finding using more frequent measures of cortisol surrounding musical performance.
| Author/Year                              | N         | Age¹ (yrs) | Topic(s)          | Task(s)                                                                 | Finding(s)                                                                 |
|-----------------------------------------|-----------|------------|-------------------|--------------------------------------------------------------------------|----------------------------------------------------------------------------|
| Lense & Dykens (2013a)                  | WS) 13    | WS) 7-49   | Fears             | Measured salivary cortisol before and after a prepared solo music         | No significant change in cortisol in response to musical performance;      |
| (study #2 only)                         |           |            |                   | performance in front of mixed familiar/unfamiliar audience.              | Baseline cortisol significantly correlated with rated musical skill        |
| Blomberg, Rosander, & Andersson (2006)  | WS) 38    | WS) 10-50  | Fears             | Compared measures of fears, hyperacusis, and musicality                 | Fears and anxieties could be associated with hyperacusis                   |
| Dykens et al. (2005)                    | WS) 31    | All) 4-21  | Problem Behaviors | Compared measures of problem behaviors and musicality                    | Externalizing symptoms negatively correlated with listening to music;      |
| (2 studies)                             | PW) 26    |            |                   |                                                                          | Internalizing symptoms negatively correlated with producing music;       |
|                                         | DS) 32    |            |                   |                                                                          | Lower levels of fear and anxiety associated with increased frequency,     |
|                                         |           |            |                   |                                                                          | skill, and duration in producing music                                   |
|                                         | WS) 26    | All) 8-47  | Fears Anxiety     | Compared measures of fears, anxieties, problem behaviors, and musicality.|                                                                            |
|                                         | PW) 16    |            | Problem Behaviors |                                                                          |                                                                            |
|                                         | DS) 25    |            |                   |                                                                          |                                                                            |

DS = Down Syndrome; PW = Prader-Willi Syndrome; WS = Williams Syndrome

¹ Age in years, expressed in one of the following formats based on information available: a) range in years, b) mean (standard deviation), or c) mean ± years
Methodological Critique: A Critical Review

Of the 31 articles included, all but one that included more than one study either utilized entirely new samples or added new participants to a subset of participants from the first sample. For this reason, when analyzing various methodological aspects of the included articles it was decided to interpret these results through the number of studies as opposed to the number of articles, as the samples for these studies were largely different.

Method of Diagnosis. Over the past 30 years, there have been many developments in methods for diagnosing WS. This was reflected in the variability of the reporting of diagnostic methods in the included studies. Early cases of WS relied on the medical and clinical phenotype to receive a diagnosis of WS. Since the 1990s, DNA testing using fluorescent in situ hybridization (FISH) has become the most widely used test of genetic confirmation for WS (Lowery et al., 1995). Of the included studies, 56% reported their participants’ diagnoses were genetically confirmed using the FISH test; 31% reported diagnoses were “genetically confirmed” but did not specify the method; 11% reported the use of a phenotype index (American Academy of Pediatrics, Committee on Genetics, 2001; Pérez-Jurado, 1997; Preus, 1984); 8% were unclear, as diagnostic information was either included in a demographics questionnaire but not reported, reported for some participants and unknown for others, or reported the age of diagnosis but not the method; and 14% did not mention confirmation of diagnosis. Many of these studies also reported that their participants exhibited characteristics of the clinical phenotype in addition to their reported methods of diagnosis. Although there has not been any indication that having confirmatory genetic testing has a significant impact on the findings of previous studies, the scientific rigor of WS research would be enhanced by emphasizing the importance of utilizing a genetic and phenotypic confirmation of WS when recruiting participants for future studies (Martens et al., 2008).

Method of Recruitment. The low prevalence of WS presents a challenge when recruiting participants. Of the included studies: 31% recruited participants from a music camp; 25% recruited from a convention or conference; 31% recruited from a national WS association
(from the USA, Canada, Spain, or Sweden); 22% recruited from an established research program or genetics clinic; 8% recruited from a parent support group; and 6% were either unclear or did not report their method of recruitment. Many of these studies recruited from more than one source.

**Matching of Control Groups.** The appropriate method for matching TD control groups has been the subject of debate. Individuals with WS have been compared to typically developing controls based on chronological age as well as those of similar mental age or cognitive level. Only two studies (6%: Levitin & Bellugi, 1998; Don et al., 1999) matched for mental age, both of which were published before the year 2000. Since then, the remainder of studies utilizing typically developing controls (61%) matched for chronological age. Twenty-two percent matched for sex/gender and 11% matched for musical training. Although it was possible to infer based on available information, 19% of the studies did not explicitly state their matching criteria. One study utilized participants with WS as a control group and thus does not fit into any of the above categories.

**Types of Control Groups.** Various types of control groups were recruited for comparison purposes throughout the included studies: 64% TD control groups; 8% Autism Spectrum Disorder (ASD); 11% Down Syndrome (DS); and 6% Prader-Willi Syndrome (PW). Some of the studies included multiple control groups. Nearly one-third of the studies (31%) did not utilize a control group, including only participants with WS. Future studies should consider including both typically developing controls and controls with other developmental disabilities or cognitive deficits in order to better characterize the diverse range of musicality in individuals with WS.

**Reporting IQ.** Nine different tests of cognitive function were utilized across the included studies. Given the time-span covered in this study, some of these were revised versions of the same test. Overall, 39% of the studies used the Kaufman Brief Intelligence Test (KBIT, Kaufman & Kaufman, 1990; KBIT-2, Kaufman & Kaufman, 2004) and 33% used various versions of the Wechsler Intelligence Scale (WISC-R, Wechsler, 1974; WPPSI-R, Wechsler,
1989; WISC-III, Wechsler, 1991; WAIS-III, Wechsler, 1997; WASI, Wechsler, 1999; WPPSI-III, Wechsler, 2002; WISC-IV, Wechsler, 2003). Some of the studies, particularly those with samples of a wide age range, employed multiple measures depending on the age of the participants. 14% of the studies reported IQ values but did not list which test was used and 6% reported using a test but did not include the resulting values. 14% did not assess for IQ.

Reporting Hearing Loss and Sensitivities. Screening for both hearing loss and sensitivities for participants with WS is particularly relevant to studies of music given that musical stimuli fall under the auditory domain. Of the included studies: 44% did not screen for hearing loss or sensitivities; 31% relied on parent report and did not specify the method of collection; 17% relied on parent report through a mentioned questionnaire (8% Sensitivities to Sounds Questionnaire, Lense & Dykens, 2013b; 6% Salk/McGill Music Inventory (SAMMI), Levitin et al., 2004; 3% Hyperacusis Questionnaire (HQ), Khalfa et al., 2002); and 8% utilized a threshold audiometry test.

Testing Limitations. Future studies may benefit from consideration of methodological challenges in evaluating individuals with WS:

- Discrimination tasks involving determinations of ‘same vs. different’ require verbal memory, a skill that may be impaired in individuals with WS (Don et al., 1999). Hopyan et al. (2001) utilized 3 different pre-tests to confirm understanding of the concepts of same/different. Future studies may benefit from such screening measures or the assessment of verbal memory as a covariate.

- Several studies reported near ceiling effects for TD participants (Järvinen-Pasley et al., 2010; Martens et al., 2010; Martínez-Castilla & Sotillo, 2014). The modifications to tasks that may be required to accurately evaluate individuals with WS may make tasks too easy for TD, limiting the comparisons made between groups.

- Many studies utilized perceptual ratings by trained judges (Musicality: Lense & Dykens, 2013a; Lense & Dykens, 2013b; Lense et al., 2013; Tonal/Rhythm: Levitin & Bellugi, 1998; Martínez-Castilla & Sotillo, 2008; Martínez-Castilla et al., 2011). Although these judges
were trained and inter-rater reliability was established, these perceptual judgments are subject to bias and may be impacted by each judge’s musical background. Martínez-Castilla & Sotillo (2008) found that acoustical analysis of a musical performance produced more accurate and reliable results than ratings by either musicians or non-musicians.

- When utilizing multiple test measures, consideration should be given to task length, complexity, and order. Don et al. (1999) and Hopyan et al. (2001) both assessed pitch and rhythm discrimination skills. Participants in both of these studies scored worse on the rhythm test, which always followed the pitch test. Thus, test order was a possible confound for these studies. Don et al. also reported attentional difficulties during testing, which could have differentially affected performance on these tasks.

- Similarly, 23% of the included studies reported excluding participants based on the inability to understand or follow directions, inability to attend to task, refusal to participate in structured task, or presence of recording artifacts during neuroimaging. Future studies may benefit from improved screening measures to assess for these issues.

- Considering the disposition toward social interaction among individuals with WS, presentation of live versus recorded musical stimuli may impact results as individuals with WS may interact differently with socially presented stimuli.

- Other known methodological issues include small sample sizes (excluding survey data from Levitin et al. (2004), the mean WS sample size among the included studies was 23), large age ranges in samples, methods of matching controls, and potential bias related to methods of recruitment.

**Risk of Biases.** Two common biases were found throughout the included studies that are known biases of many studies of individuals with WS. The first of these is related to the recruitment of participants. Nearly half (47%) of the included studies recruited participants from either a summer music camp or a national convention. Both of these methods are predisposed toward those who attend such events and are potentially not representative of the general WS population. This may be of particular relevance in the study of the relationship between music
and WS. Although previous involvement in music or heightened musical skill are generally not requirements to attend such camps, individuals who attend summer music camps may be predisposed toward greater musical involvement or interest.

The second bias is related to the reliance on parent report. Roughly 75% of the included studies utilized parent report for various outcome tools, questionnaires, and to report hearing sensitivities and musical training. Although the demographic information gathered related to diagnosis and musical training leaves little room for error, relying on parent report to determine prevalence of hyperacusis and levels of anxiety, emotional responsiveness to music, musical interest or engagement, musical skill, etc. leaves room for under or over reporting of this information. However, Fisher, Mello, & Dykens (2014) attest to the accuracy of parent-report over self-report with individuals with WS. Thus, use of parent report may be the most accurate method of assessment in some cases.

**Proposed Reporting Guidelines.** Considering the methodological and reporting limitations outlined above, future studies should:

- Assess and report the following, including instruments used and results:
  - IQ
  - Hearing sensitivities and impairment
  - Musical training (both duration and types)
- Additionally identify and report:
  - Method of diagnosis or genetic confirmation
  - Method of recruitment of participants
- Note whether stimuli are presented via live or recorded audio, consider its effects on task performance (engagement, desire to please, salience)
DISCUSSION

This review identified 31 articles examining the relationship between WS and music. These articles were divided into seven categories, many of which align with general phenotypic characteristics of the syndrome.

Williams Syndrome and Music: Perspective on the Phenotype

Overall, the musical profile of individuals with WS appears to be deeply rooted in their affinity for music. This is expressed through consistent reports of increased interest in music, a greater propensity toward musical activities, and heightened emotional responsiveness to music. Although many individuals with WS share a strong affinity for music, a smaller percentage demonstrate strong musical skills. Many mixed results found in this review point to a high degree of variability in skill and engagement in music, also suggesting that overall musicality may not predict musical skill. Individuals with WS appear to present with relatively good musical skills that are more in line with their cognitive abilities than chronological age. Musical strengths for this population seem to be based more in expressivity and musicality over formal musical skills.

Atypical auditory processing, autonomic irregularities, and differential neurobiology might underlie this affinity for music and other aspects of the phenotype. It is unlikely that the deletion of genetic material found in individuals with WS directly predisposes these same individuals toward a greater interest in music. Instead, it is highly plausible that the resulting differences in structural and functional anatomy uniquely affect the manner in which music is perceived and processed within this population. However, it is important to remember that the phenotype in neurodevelopmental disorders does not fully emerge from the outset, but develops gradually over time (Martínez-Castilla et al., 2016). Thus, it is difficult to tease apart the biological and sociological influences that relate to this unique relationship between WS and music.
Music and IQ

When examining the effects of cognitive ability (IQ) on performance during musical tasks, multiple studies identified a correlation between task performance and measures of IQ (Lense & Dykens, 2016; Martínez-Castilla et al., 2016; Martínez-Castilla et al., 2011). This is also supported by the numerous reports of WS performance falling below TD peers matched for CA, yet fairly similar to TD peers matched for MA. However, other studies found no such relationships (Hopyan et al., 2001; Martínez-Castilla & Sotillo, 2008; Martínez-Castilla & Sotillo, 2014). Considering these contradictory findings in light of the atypical cognitive profile of WS, it is possible that performance on these tasks is more predicted by specific cognitive processes than an overall measure of intellectual ability. Future studies should continue to examine specific cognitive skills and cognitive developmental milestones as potential covariates when comparing performance on tasks to TD peers.

Perceptual Abilities

The ‘creative completions’ noted by Levitin and Bellugi (1998) may explain some of the variance in skill for perceptual or discrimination tasks reported in this review and also points to an interesting disparity in the perceptual abilities of individuals with WS. On a task requiring participants to accurately reproduce a given rhythm by clapping, errors produced by participants with WS tended to be more rhythmically compatible with the target rhythm than the comparison group, thus preserving the overall structure of the original rhythm. Given this, it may also be true that on tasks requiring participants to judge whether two rhythms are the same or different, participants with WS may judge two items as being the same if they share similar overall rhythmic structures instead of comparing them on the basis of individual beats (Hopyan et al., 2001). Similarly, acoustical analysis of song-singing revealed that individuals with WS make many more errors in accuracy for individual pitches than those that alter the overall contour of the melodic phrase (Martínez-Castilla & Sotillo, 2008). In summary, individuals with WS seem to conserve the overall structure, contour, or idea of a musical phrase better than they can discriminate or reproduce it exactly.
On one hand, individuals with WS may be better at conserving global musical structures, such as melody and meter, than their component parts, such as individual pitches, beats, or rhythms. Yet, on the other hand, this stands in contrast to visuospatial deficits that have been reported in individuals with WS, which show remarkable deficits in the processing of global over local aspects of visual stimuli (Bihrlle et al., 1989). In the auditory domain, Deruelle et al. (2005) reported similar deficits in the perception of global contour in melody compared to local pitch elements. Possible explanations for this striking disparity could be the inherent difference between production and discrimination tasks or it may be that these tasks may require different cognitive skills. Basic pattern perception skills are stronger in individuals with WS than auditory rote-learning or working memory, which are necessary for various same/different discrimination tasks (Don et al., 1999, Martínez-Castilla et al., 2016). Future studies should continue to examine this relationship and should consider examining perceptual skills in both the auditory and visual domains within the same sample.

**Neuromodularity**

Many articles reviewed in this study interpreted their findings either in support of or against claims of cognitive modularity, specifically whether or not music constitutes an innate neurological module independent of general cognition (support for: Lenhoff et al., 2001; Levitin & Bellugi, 1998; Levitin et al., 2004; Wengenroth et al., 2010; support against: Don et al., 1999; Martínez-Castilla & Sotillo, 2008; Martínez-Castilla & Sotillo, 2014; Martínez-Castilla et al., 2011; Martínez-Castilla et al., 2013; Martínez-Castilla et al., 2016). Although these claims are outside the scope of this review, these discrepant findings indicate the need for further inquiry into this debate.

**Clinical considerations**

Many articles proposed the potential for including music in both clinical and educational interventions with individuals with WS. However, to date, no published studies have examined the clinical use of music with this population. Articles point to the heightened interest in and motivation for musical activity (Don et al., 1999; Levitin et al., 2004) as well as greater attention
during musical versus non-musical activity (Martens et al., 2011) within the WS population as rationale for incorporating music into clinical intervention to assist in modulating and maintaining arousal, attention, and engagement with clinical tasks. Proposed target domains of intervention among the included studies were diverse, covering: social and communication skills (Lense & Dykens, 2016); language and prosody skills (Martínez-Castilla & Sotillo, 2014); auditory-motor connections (Lense & Dykens, 2013b); emotional understanding and sensitivity (Ng et al., 2013); management of anxiety (Dykens et al., 2005); attention and concentration (Lense & Dykens, 2013b); and educational outcomes (Dunning et al., 2015; Martens et al., 2011). More studies are needed to explore how this affinity for music can be harnessed in clinical and educational interventions.

Existing research on the effects of music on non-musical function is promising for many of the above areas. Active use of instruments in synchrony with a musical beat supports optimal kinematics through the coupling of auditory and motor processes (Thaut, 2005) and the provision of temporal limits that cue and constrain movement (Lim, Miller, & Fabian, 2011; Thaut, Schleiffers, & Davis, 1991). Playing musical instruments also provides the opportunity to practice fine and gross motor skills, which may be a more preferred and motivating activity for many individuals with WS. Pairing novel information to music has been shown to be an effective tool for enhancing recall of target information (Gfeller, 1983; Knott, 2017; Wolfe & Horn, 1993). Furthermore, the structure and predictability provided by the rhythmic aspects of music supports and guides attention (Geist & Geist, 2012; Thaut, 2005).

Considering the proposed potential and existing clinical literature outside of WS, future studies are needed to explore musical intervention with individuals with WS. Music therapy may benefit individuals with WS by:

- Composing songs using targeted academic content to aid in retrieval of academic information
- Using songs to structure procedural information to promote adherence to routines or enhance acquisition of academic concepts, such as math skills
- Playing instruments to target various fine and gross motor skills
Simulating social exchanges during group music making to support development of pragmatic communication skills

Playing, composing, or discussing music as a means of self-expression to improve frustration tolerance (e.g., dealing with setbacks in learning academic material) and positive coping skills in the management of anxiety

Incorporating music into treatment in collaboration with other allied health providers to enhance engagement, motivation, and attention to task

Although unpublished literature was not included within the scope of this study, multiple theses and dissertations were identified during the formal search process that have begun to examine the effects of music on non-musical function in individuals with WS. Appendix C lists these identified unpublished theses and dissertations. However, this list may not be exhaustive as unpublished literature was not within the scope of this study.

Conclusions

The affinity for music observed in this population since the earliest of case reports has been given considerable attention. This review contributes to the existing literature by examining the unique relationship between WS and music through a systematic and comprehensive research lens. Following an exhaustive search for literature, this review: 1) reported current findings related to WS and music; 2) assessed the methodological quality of the included articles; and 3) discussed the potential implications and considerations for the clinical use of music with this population. A better understanding of the musical profile of individuals with WS is one step closer in understanding the role of music within the WS phenotype.
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## Appendix A: Exclusions Based on Full-Text and Reasons for Exclusion

| Author(s)                  | Year | Title                                                                 | Publication                                                                 | Reason for Exclusion               |
|----------------------------|------|-----------------------------------------------------------------------|----------------------------------------------------------------------------|-----------------------------------|
| Carrico, A. H.             | 2015 | Constructing a two-way street: An argument for interdisciplinary collaboration through an ethnomusicological examination of music therapy, medical ethnomusicology, and Williams syndrome | *Voices: A World Forum for Music Therapy, 15(3), 1-16*                     | Positional Paper                  |
| Carrico, A. H.             | 2014 | Discovering "diffability": Musical experiences and perspectives of individuals with Williams syndrome at Whispering Trails | Thesis: Florida State University                                            | Unpublished Thesis                |
| Don, A. J.                 | 1999 | Auditory pattern perception in children with Williams syndrome        | Dissertation: University of Windsor                                         | Unpublished Dissertation           |
| Elsabbagh, M. Cohen, H.    | 2010 | Discovering structure in auditory input: Evidence from Williams syndrome | *American Journal on Intellectual and Developmental Disabilities, 115(2), 128-139* | Music not focus of article; Auditory Perception |
| Fidler, D. J. Lawson, J. E. Hodapp, R. M. | 2003 | What do parents want?: An analysis of education-related comments made by parents of children with different genetic syndromes | *Journal of Intellectual and Developmental Disability, 28(2), 196-204*     | Music minimal focus of article    |
| Heaton, P. Allen, R.       | 2009 | "With concord of sweet sounds...": new perspectives on the diversity of musical experience in autism and other neurodevelopmental conditions | *Annals of the New York Academy of Sciences, 1169, 318-325*               | WS minimal focus of article; Book chapter, published in journal |
| Kwak, E. E.                | 2009 | An exploratory study of the use of music therapy in teaching mathematical skills to individuals with Williams Syndrome | Dissertation: Michigan State University                                    | Unpublished Dissertation           |
| Lenhoff, H. M.             | 2009 | Soaking in the music                                                  | *Exceptional Parent, 39(3), 40-45*                                        | Magazine Article                  |
| Lenhoff, H. M.             | 1998 | Information sharing: Insights into the musical potential of cognitively impaired people diagnosed with Williams syndrome | *Music Therapy Perspectives, 16(1), 33-36*                               | Informational Article             |
| Lenhoff, H. M. Wang, P. P. Greenberg, F. Bellugi, U. | 1997 | Williams syndrome and the brain                                      | *Scientific American, 277(6), 68-73*                                      | Magazine Article                  |
# Appendix A cont.: Exclusions Based on Full-Text and Reasons for Exclusion cont.

| Author(s)          | Year | Title                                                                 | Publication                                                                                     | Reason for Exclusion                  |
|--------------------|------|----------------------------------------------------------------------|-------------------------------------------------------------------------------------------------|----------------------------------------|
| Lense, M. D.       | 2011 | Musical interests and abilities in individuals with developmental    | *International Review of Research in Developmental Disabilities, 41, 265-312*                  | Book Series/Review                     |
| Dykens, E. M.      |      | disabilities                                                        | *Annals of the New York Academy of Sciences, 1060, 325-334*                                   |                                        |
| Levitin, D. J.     | 2005 | Musical behavior in a neurogenetic developmental disorder: Evidence | *Review Article*                                                                             |                                        |
|                    |      | from Williams Syndrome                                              |                                                                                                 |                                        |
| Mackenzie, L. B.   | 2005 | ‘When words fail, music speaks’: How communication transforms       | *Dissertation: University of Massachusetts Amherst*                                           | Music not focus of article;            |
|                    |      | identity through performance at the Berkshire Hills Music Academy   |                                                                                                 | Unpublished Dissertation               |
| Maher, B. A.       | 2001 | Music, the brain, and Williams syndrome: Rare disorder offers insight| *Scientist, 15(23), 20-21*                                                                    | Partially Music Magazine Article      |
|                    |      | into the genetic basis of cognition                                 |                                                                                                 |                                        |
| Milne, H.          | 2004 | The development of talent in young adults with Williams syndrome:    | *Australasian Journal of Special Education, 28(2), 79-101*                                     | Data re-published in Reis et al. (2003)|                                        |
|                    |      | An exploratory study of ecological influences                       |                                                                                                 |                                        |
| Milne, H.          | 2002 | A comparative case study of persons with Williams syndrome and      | *Dissertation: University of Connecticut Thesis: Middle Tennessee State University*           | Unpublished Dissertation               |
|                    |      | musical interests                                                    |                                                                                                 |                                        |
| Pridmore, M. D.    | 2014 | Affective priming effect of music on emotional prosody in Williams   | *Unpublished Thesis*                                                                         |                                        |
|                    |      | syndrome                                                            |                                                                                                 |                                        |
| Stambaugh, L.      | 1996 | Special learners with special abilities                              | *Music Educators Journal, 83(3), 19-23*                                                       | Music minimal focus of article;        |
|                    |      |                                                                     |                                                                                                 | Magazine Article                      |
| Sacks, O.          | 1995 | Musical ability                                                      | *Science, 268(5211), 621-622*                                                                 | Letter                                |
| Wengenroth, M.     | 2010 | Individual MRI segmentation reveals enlarged auditory cortex in     | *Child Neuroradiology, 20(3), 196-197*                                                       | Conference Abstract/Unable to Retrieve|
| Blatow, M.         |      | Williams syndrome as a neural substrate of training-independent     |                                                                                                 |                                        |
| Bendszus, M.       |      | musicality                                                           |                                                                                                 |                                        |
| Schneider, P.      |      |                                                                     |                                                                                                 |                                        |
| Wengenroth, M.     | 2010 | Leftward lateralization of auditory cortex underlies holistic sound | *PLoS One, 5(8), 1-10*                                                                     | Not Music; Auditory Perception      |
| Blatow, M.         |      | perception in Williams syndrome                                      |                                                                                                 |                                        |
| Bendszus, M.       |      |                                                                     |                                                                                                 |                                        |
| Schneider, P.      |      |                                                                     |                                                                                                 |                                        |
Appendix B: Data Extraction Form

| Thesis ID | 1-3 authors: Author, Author, & Author (yyyy) 4+ authors: Author et al. (yyyy) |
|-----------|--------------------------------------------------------------------------------|
| Report ID # (from endnote) | |
| Name/ID of Individual Performing Data Extraction | |
| Date Form Completed (dd/mm/yyyy) | |

### 1. General Information

1. **Author(s)**
2. **Year**
3. **Title of Publication**
4. **Journal** (journal, volume(issue), pages)
5. **Keywords**
6. **Database(s) accessed**
7. **Notes:**

### 2. Eligibility

| Study Characteristics | Inclusion Criteria (exclude if no) | Yes/No/Unclear | Location in text (pg & ¶/fig/table) |
|-----------------------|-----------------------------------|----------------|-------------------------------------|
| 7. Date Range         | Published before 2017 (through Dec 2016) |                |                                    |
| 8. Subject(s) of study | Williams syndrome                |                |                                    |
| 9. Language           | English                           |                |                                    |
| 10. Publication       | Published in a peer-reviewed journal |                |                                    |

| Study Characteristics | Exclusion Criteria (exclude if yes) | Yes/No/Unclear | Location in text (pg & ¶/fig/table) |
|-----------------------|------------------------------------|----------------|-------------------------------------|
| 11. Participants      | Participants include a mixture of individuals with WS and individuals with other diagnoses and outcomes are not differentiated by the authors |                |                                    |
| 12. Subject(s) of study | Participants have comorbid diagnoses of WS and another diagnosis |                |                                    |
| 13. Design of study   | Hyperacusis without reference to music |                |                                    |
| 14. Decision:         | Review/Magazine/Positional/etc.    |                |                                    |

15. **Reason for exclusion**

Notes:
Notes on using a data extraction form:

- Be consistent in the order and style you use to describe the information for each included study. Record any missing information as unclear, not described, or N/A, to make it clear that the information was not found in the study report(s), not that you forgot to extract it.
### 3. Methods

| Description as stated in report/paper | Location in text (pg & ¶/fig/table) |
|--------------------------------------|-------------------------------------|
| 16. Aim of study                     |                                     |
| 17. # of Reports (if more than one)  |                                     |

Notes:

### 4. Participants and Groups

*Label each group within each box (where applicable), using consistent labeling for each row. Copy and paste this table for multiple reports/experiments within the same article.*

| Participants | Description as stated in report/paper | Location in text (pg & ¶/fig/table) |
|--------------|---------------------------------------|-------------------------------------|
| 18. Total N  |                                       |                                     |
| 19. Group name(s) |                                  |                                     |
| 20. Number in group (n) |                                |                                     |
| 21. Control Group(s) matched by: |                                 |                                     |
| 22. Age (range, mean, etc.) |                                   |                                     |
| 23. Sex (M = n; F = n) |                                     |                                     |
| 24. Race/Ethnicity |                                      |                                     |
| 25. Method of Diagnosis |                                    |                                     |
| 26. Method(s) of recruitment of participants |                            |                                     |
| 27. Inclusion/Exclusion criteria (used in study) |                          |                                     |
| 28. IQ Values |                                       |                                     |
| IQ Method of Collection |                                 |                                     |
| 29. Hearing Screening |                                  |                                     |
| Hearing Screening Method of Collection |                          |                                     |
| 30. Musical Training |                                   |                                     |
| Musical Training Method of Collection |                            |                                     |
| 31. Other Screening (aside from IQ/Hearing) |                   |                                     |
| 32. Other treatment(s) received (in addition to study intervention) |                   |                                     |

Notes:
5. Tasks and Outcomes

Number each outcome within each box, using consistent numbering for each row. Copy and paste this table for multiple reports/experiments within the same article.

|   | Description as stated in report/paper | Location in text (pg & ¶/fig/table) |
|---|--------------------------------------|-------------------------------------|
| 33. **Description of task(s)** *(general description of each task or tool)* | | |
| 34. **Setting** *(including location and social context)* | | |
| 35. **Outcome name(s)** | | |
| 36. **Outcome tool(s)** | | |
| 37. **Gathered from Participants, parent report, etc.** | | |
| 38. **Outcome definition(s)** *(operational definition)* | | |

Notes:

6. Results

List each outcome appropriately under Quantitative/Qualitative Copy and paste table for multiple reports/experiments within the same study.

|   | Description as stated in report/paper | Location in text (pg & ¶/fig/table) |
|---|--------------------------------------|-------------------------------------|
| 39. **For Quantitative Data** | | |
| Outcome | | |
| Outcome | | |
| Outcome | | |
| Notes: | | |

|   | Description as stated in report/paper | Location in text (pg & ¶/fig/table) |
|---|--------------------------------------|-------------------------------------|
| 40. **For Qualitative Data:** | | |
| Outcome | | |
| Outcome | | |
| Outcome | | |
| Notes: | | |
### 7. Other information

| 41. | Key conclusions of study authors |
| 42. | Suggested areas for further research |
| 43. | References to other relevant studies |

Notes:

### 8. Risk of Bias assessment

| Domain | Risk of bias | Support for judgment | Location in text |
|--------|--------------|----------------------|------------------|
| 44. Sampling bias (self-selection: predisposition toward involvement) | Low/High/Unclear | | |
| 45. Self/Parent Report biases (reporting on behalf of another; social desirability bias) | | | |
| 46. Other bias | | | |

Notes:

### 9. Applicability

| 47. Does the study directly relate to the review question? | Yes/No/Unclear | Any issues of partial or indirect applicability?: |
|--------------------------------------------------------|---------------|-----------------------------------------------|

| 48. Which area(s) of the review does this publication relate to? | |

Notes:
# Appendix C: Unpublished Theses and Dissertations

| Author          | Year | Title                                                                                   | Type/University                  |
|-----------------|------|-----------------------------------------------------------------------------------------|----------------------------------|
| Carrico, A. H.  | 2014 | Discovering "diffability": Musical experiences and perspectives of individuals with      | Thesis Florida State University   |
|                 |      | Williams syndrome at Whispering Trails                                                  |                                  |
| Pridmore, M. D. | 2014 | Affective priming effect of music on emotional prosody in Williams syndrome              | Thesis Middle Tennessee State University |
| Barnett, B. A.  | 2013 | Does novel music improve verbal memory in individuals with Williams syndrome?            | Thesis The Ohio State University  |
| Ross, G.        | 2011 | Perception of emotional expression in musical performance by individuals with Williams   | Thesis Western Michigan University|
|                 |      | syndrome                                                                                 |                                  |
| Woitulewicz, L. M. | 2011 | The effects of background music, rhythm, and noise on a sustained attention task in     | Thesis Western Michigan University|
|                 |      | adults with Williams syndrome                                                            |                                  |
| Kwak, E. E.     | 2009 | An exploratory study of the use of music therapy in teaching mathematical skills to      | Dissertation Michigan State University |
|                 |      | individuals with Williams Syndrome                                                       | Thesis Western Michigan University|
| Hata, M.        | 2006 | A survey of music therapists regarding the efficacy of music therapy in the treatment of | Dissertation University of Massachusetts Amherst |
|                 |      | children and adolescents with Williams syndrome                                          | Thesis University of Connecticut  |
| Mackenzie, L. B. | 2005 | 'When words fail, music speaks': How communication transforms identity through          | Dissertation University of Windsor|
|                 |      | performance at the Berkshire Hills Music Academy                                          | Thesis Western Michigan University|
| Milne, H.       | 2002 | A comparative case study of persons with Williams syndrome and musical interests         | Dissertation University of Wyoming|
| Don, A. J.      | 1999 | Auditory pattern perception in children with Williams syndrome                           | Thesis Western Michigan University|
| Pawuk, L. G.    | 1999 | A comparison of rhythmical abilities and behaviors between typical children and children|                                  |
|                 |      | with Williams Syndrome                                                                    |                                  |