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João Pedro Jacinto Pina Barreto

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À minha avó Aninhas

Aos meus pais, Clotilde e Fernando

À minha irmã, Ana Maria

A todos aqueles que me apoiaram e inspiraram, em Portugal e na Eslovénia

Williams Syndrome: a review on its musical phenotype

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Abstract

Williams syndrome is a neurodevelopmental disorder characterized not only by cognitive impairment but also by sociable personalities, preserved verbal abilities and a particular fondness for music. Given the importance that musical and non-musical auditory stimulation has in the lives of people with Williams syndrome, this disease has been regarded as an worthy model to address how the human brain processes music and how music affects emotional development. Moreover, given their large engagement in musical activities, it is believed that music is crucial to help these patients to achieve their full potential. Thus, several studies in recent years have addressed the musical phenotype in Williams syndrome, focusing on areas such as audiology, behavioural sciences, neuroanatomy or neurobiology. The aim of this review is to give a broad perspective on those studies, as well as some considerations about their limitations and possible future directions for this topic.

Key-words: Williams syndrome; music

Introduction

Williams syndrome (WS, also known as Williams-Beuren syndrome) is a multisystem neurodevelopmental disorder affecting approximately 1 in 7500 persons (Stromme, Bjornstad, & Ramstad, 2002). It is a genetic condition caused by the usually sporadic hemizigous deletion of 1.5 to 1.8 million base pairs in the long arm of chromosome 7 (position 7q11.23). This corresponds to approximately 28 transcribed genes, out of an estimated total number of 1000 genes in this chromosome (Schubert, 2009).

WS patients are characterized by mild to moderate intellectual disability, but the full WS phenotype comprises a constellation of neurologic, cardiovascular, endocrine, and facial features whose extent ranges from subtle to severe. The facial features are highly variable: children often have a flat nasal bridge, periorbital puffiness, long philtrum, wide smile, short upturned nose, and delicate chin, while older patients have slightly coarser features (Pober, 2010). The cardiovascular hallmark of the syndrome consists of stenoses of medium- and large-sized arteries, most commonly in the supravalvular aortic region. These stenoses are the most well-established genotype-phenotype correlation of the syndrome and are produced by the loss of the gene for elastin (ELN) (Pober, Johnson, & Urban, 2008). Endocrine-related problems include diabetes mellitus, subclinical hypothyroidism, hypercalciuria and hypercalcemia, as well as decreased growth spurt (Waxler, Levine, & Pober, 2009).

Diagnosing the syndrome is primarily based on the clinical recognition of the characteristic phenotype and subsequent visualization of the microdeletion using FISH (fluorescent *in-situ* hybridization) as a rapid and accurate laboratory test to confirm the disease. However, given the wide variability in WS phenotypes, it is difficult to make genotype-phenotype correlations in WS (Tassabehji, 2003) and there is still no genetic test

that can predict its severity in an individual (Pober, 2010). Moreover, the number of recently created transgenic animal models of WS has clearly demonstrated the complexity of the combinatorial power that result from haploinsufficient gene deletions (Osborne, 2010; Schubert, 2009).

The neuropsychological phenotype of WS is equally complex. IQ across the entire spectrum of affected individuals ranges from 40 to 100 - with an average IQ of 50-60 in older patients – which corresponds to mild/moderate intellectual disability (Martens, Wilson, & Reutens, 2008). Moreover, this disorder is characterized by seemingly paradoxical differences in high-level cortical functioning: some faculties are relatively preserved (music, language, facial processing and social drive), while others show severe deficits (conceptual reasoning, spatial ability, motor coordination, problem solving, arithmetic processing) (Hopyan, Dennis, Weksberg, & Cytrynbaum, 2001; Levitin et al., 2003).

These patients have, as adults, high levels of behavioural disturbances such as anticipatory but not social anxiety, obsessions, irritability and distractibility. Despite their friendly personality, many are socially isolated, and only a few manage to live independently. Psychiatric comorbidities, if present, are difficult to manage and contribute to lower quality of life (Pober, 2010).

Musicality in WS

The aim of this review is to give a broad perspective on recent studies about the musical phenotype in Williams Syndrome. We will address studies that focus on audiological, neuroanatomical, neurobiological and behavioural/emotional aspects of WS, as well as studies that approach musicality as a whole and how musicality is affected in WS patients (summarized in Table 1). Given the well-known genetic cause and the proeminent cognitive and behavioural manifestations concerning music in WS, music has been the subject of intense study in the search for phenotype-genotype clues.

Within the WS cognitive profile, music appears to be an area of relative strength that defies the otherwise decrease in cognitive ability rather than a preserved function (M. Lense & Dykens, 2013; M. D. Lense, Shivers, & Dykens, 2013). In fact, individuals with WS are reported to have greater musical creativity, spend more time listening to music and show stronger reactions to music. Even if not being skillful musicians, their ability to play musical instruments is to be noted, especially in face of other motor and general cognitive impairments. On the other hand, a sensorineural hearing loss for higher-tones has also been reported as these individuals grow older (Pober, 2010), and they usually have lowered hearing thresholds and fear certain sounds generally regarded as normal: hyperacusis and auditory allodynia, respectively (Levitin et al., 2003).

Although musicality is not easy to define, it may be used as a summary term that comprises interest in music, emotional reaction to music, musical expressivity and musical abilities (Blomberg, Rosander, & Andersson, 2006; Ng, Lai, Levitin, & Bellugi, 2013). WS patients are commonly described as frequently listening to music and even producing it, as well as having above-average musical memory and emotional engagement with music (Levitin, 2005). More than analytic skills concerning pitch and rhythm, musical strengths in

WS consist of a strong engagement with music as a means of expression (Hopyan et al., 2001). As an example of the importance that music has in the WS phenotype, is the fact that music camps and academies were specifically created to cater to the WS population (Ng et al., 2013).

In a study by Levitin and colleagues, questionnaires to parents were used to get information on WS patients' interest in music, general emotional responsiveness to music, musical creativity, music reproduction and musical training, as well as on the age of onset of several musical behaviours. They found that WS patients rated higher than Down syndrome and autism for musical accomplishment, engagement and interest in music and with equivalent rates when compared to normal controls (Levitin et al., 2004). In a separate study it was shown that WS patients, when submitted to an echo clapping task, showed not only similar rhythmic skills to those of normal controls, but also tended to provide, when making errors, musically compatible rhythms (Levitin et al., 2004). This was interpreted as a marker of rhythmic ability or creativity (Levitin, 2005). As to melodic production, WS were better than people with Down syndrome, but not as good as normal controls, suggesting that rhythmic capabilities override melody capabilities in WS (Levitin, 2005). Finally, an unusually extreme predominance of holistic sound perception in WS has been functionally demonstrated by increased amplitudes of left auditory evoked fields, and this could provide a theoretical basis for their high rhythmic creativity, as well as for the particular fondness of WS patients for percussive instruments (Wengenroth, Blatow, Bendszus, & Schneider, 2010). As a note, holistic (or synthetic) listeners perceive sounds as a whole, with emphasis on the fundamental tone, and prefer rhythmic beats; on the other hand, spectral (or analytical) listeners decompose sounds into their single harmonic constituents and prefer instruments rich in overtones (Schneider & Wengenroth, 2009; Wengenroth et al., 2010).

Audiological features and reactivity to sound in WS

In spite of their unquestionable affinity for music, WS patients also display a variety of auditory abnormalities. In particular, there have been claims of hyperacusis in WS. These should be taken cautiously, however, given the different uses the term has had. Medically, hyperacusis should be defined as lowered hearing thresholds, i.e. the ability to hear soft sounds other people just cannot hear (Levitin, Cole, Lincoln, & Bellugi, 2005). Two other conditions are usually grouped under the erroneous umbrella term of "hyperacusis": *odynacusis*, a lowered pain threshold for loud sounds; and *auditory allodynia*, an aversion/fear of sounds that would not be normally perceived as painful. In a questionnaire-based study of auditory disorders in WS compared to autism, Down syndrome and normal controls, Levitin and his team found that true hyperacusis, although only present in the WS group, had a rate of 4.7% (much lower than in studies that had used the term indiscriminately) (Levitin et al., 2005). It has also been suggested that other features commonly found in WS such as enhanced fear and anxiety, can be related to hyperacusis, since a clear association has been found between social avoidance based on hyperacusis and fear of animals (Blomberg et al., 2006).

On the other hand, WS patients also display *auditory fascinations*, an attraction for certain sounds (Levitin et al., 2005; Levitin et al., 2003). A rate of 9% for auditory fascinations in WS patients is also contrasted with almost no cases in the Down syndrome, autism or normal groups. It is extremely interesting, though, to notice that such fascinations always begun as aversions and were usually connected with broad-band sounds (eg. vacuum cleaners, lawnmowers, vehicle engines). These early auditory aversions (with a striking rate of 80%) also occur to unusual sounds such as those produced by cows, clearing one's throat, a champagne cork or a coffee maker. The behaviour of children with

WS, though, make it obvious that the sounds are aversive because they are intrinsically frightening and are not aversive because they are simply loud (Levitin et al., 2005). Over time, these aversions eventually decrease or turn into fascinations, and it is even known from anecdotal reports that the relation to the object of the fascination goes beyond the auditory stimulus by itself: WS patients collect the sound-producing objects themselves, recognize their brand and model by the sound they produce, and collect its pictures from magazines (Levitin, 2005; Levitin et al., 2005). An even higher rate of 91% for odynacusis in WS is to be noted, especially by its significant association with auditory allodynia.

A study by Gallo and colleagues examined behavioural reactions to sound in young children with WS, including acoustic startle eye blinks, a widely used test of anxiety and fear. The focus was put on sound reactivity as a whole, without trying to specify the underlying audiological defect. When exposed to conventional toys emitting mild intensity sounds, WS patients would seek their parents or place hands over hears, while there was no typical behavior in the control group. However, these aversive behaviours could also be triggered by non-auditory stimulus that had been followed by auditory ones, suggesting a pattern of anticipatory anxiety (Gallo, Klein-Tasman, Gaffrey, & Curran, 2008).

Altogether, the above mentioned studies suggest that the aversive or fascinating responses to particular classes of sounds are likely also the result of behavioral and psychological characteristics of the WS phenotype rather than simply a reflection of audiological pathologies. These different auditory abnormalities also reveal alterations in distinct neuronal mechanisms: while hyperacusis, odynacusis and auditory allodynia may be related with hyperexcitability of the auditory cortex (or even to a widespread cortical enhancement), auditory fascinations are probably related with recruitment of emotional centers of the brain after particular auditory stimuli (Levitin et al., 2005).

In addition to the emotional responses to particular sounds, it is also interesting to look at auditory features more music-related, such as absolute pitch and amusia, when trying to fully understand the variability of the musical phenotype in WS. Although claims had been made that WS would be connected with high prevalence of absolute pitch (the ability to produce or identify pitch without reference tones), recent studies with larger samples show it to be a rare ability, with the same prevalence of 1 in 10.000 people as in normal population (Martinez-Castilla, Sotillo, & Campos, 2013). Given the musical fondness of WS patients it could be thought that the rate of amusia (inability to differentiate musical pitch or tone-deafness) would be lower than in the normal population, but it reached 11% in a sample of 73 adolescents and adults with WS, in contrast with the rate of 4% of amusia in normal population (M. D. Lense, Shivers, et al., 2013). Musical training was negatively related with amusia levels. There was no association, though, with auditory sensitivity, which instead predicted emotional responses to music (M. Lense & Dykens, 2013).

Another relevant aspect as far as audiological phenotype is concerned is hearing loss of cochlear origin. Mainly affecting the high frequencies, this is more common in WS (ranging from 13% to 90% of patients) than it is in the general population and it is sensorineural in at least half of the cases (Barozzi et al., 2013). Hearing loss in WS has an early onset and is probably progressive (Barozzi et al., 2012). This fact may appear to be counterintuitive when trying to understand hypersensitivity to sound in WS; however, cochlear hearing loss is commonly associated with a process called recruitment (a quick increase in loudness perception, in response to small intensity changes) (Gallo et al., 2008; Levitin et al., 2005). Possible causes for this cochlear fragility (related with outer hair cells dysfunction and high-frequency sensorineural hearing loss) include a reduced level of elastin, abnormalities of the olivocochlear efferent system and maturation deficits of the

brain stem auditory pathways (Barozzi et al., 2012; Barozzi et al., 2013). It is important to monitor the patients' auditory thresholds periodically, making it even possible to identify cochlear dysfunction before a possible onset of hearing loss, which will benefit from an audiological follow up (Barozzi et al., 2012).

Neuroanatomical and neurobiological correlations in WS musical phenotype

Despite an overall reduction in brain volume, WS individuals have been described to have a superior temporal gyrus similar in size to that of normal controls (Martens, Reutens, & Wilson, 2010). This neuroanatomical relative enlargement is compatible with a profile of musical strengths: the temporal gyrus comprises both the primary auditory cortex and the planum temporale (association auditory cortex). It is also in close proximity with the middle temporal gyrus that is also an important brain area for music processing (Levitin et al., 2003). It is interesting to notice that a subgroup of WS patients with particular good performance in musical tasks in a laboratory setting, also presented an enlarged left planum temporale, suggesting a neuroanatomical correlation of musical variability within the syndrome (Martens et al., 2010) Other important aspect is the preserved size of the temporal lobe, the superior temporal sulcus and the amygdala, when adjusted for the overall brain reduction. Such data suggest a neurodevelopmental etiology for the prominence of affective strategies in perception, cognition and communication in WS (Levitin et al., 2003). Wengenroth and colleagues give yet another proposal of structural/phenotype correlation, considering WS as unique genetic model to study training-independent auditory properties. In fact, the increased left auditory cortex volume found in WS, even without musical education, is comparable to the enlargement seen in professional musicians (Wengenroth et al., 2010).

When testing the brain processing of music by fMRI (functional magnetic resonance imaging), WS participants show a relatively dispersed activation in both the neo- and paleocortex (including greater activation of the right amygdala, cerebellum and brainstem), contrasting with the more well-defined neocortical pattern in normal controls (with focal activation of the superior and middle temporal giri and superior temporal sulcus) (Levitin et

al., 2003). The greater activation of the right amygdala may be of importance in interpreting the attraction of WS patients towards music, given the role of the amygdala in mediating the emotional aspects of musical cognition (Levitin et al., 2003). Emotional reactions to noises could also be influenced by the same brain areas, since music and noise were found to be more similarly processed in WS patients than in control participants (Levitin et al., 2005; Levitin et al., 2003).

There are also reports of synesthesia-like activations in the brain of WS patients: specific visual areas are activated by musical and non-musical stimuli. These findings may provide an explanation for the vivid visual imagery described by patients with WS when listening to music; however, these features may not be properly considered real synesthesia, since no consistent and repeatable responses occur to the same stimulation. Musical cues may be used in WS in such different contexts as refocusing attention or identifying and managing emotions (Thornton-Wells et al., 2010).

Musical training is a model of auditory-motor interaction, compatible with a mirror-neuron system (MNS); this means learners benefit more from auditory models than from verbal instructions (Lahav, Saltzman, & Schlaug, 2007; M. Lense & Dykens, 2013). Music making has been used as intervention in neurological and neurodevelopmental disorders, since it enhances auditory-motor connections, and could provide an enjoyable means to improve such abilities as attention, memory or motor skills (M. Lense & Dykens, 2013). In an experimental study involving training on how to play a musical instrument, the end-performance of WS patients was correlated with their prior musical experience and with their visual-motor skills; moreover, the end-performance could be predicted by self-reported use of auditory learning strategies, contrasting with no effect by visual or instructional learning (M. Lense & Dykens, 2013).

Another important observation is the maintenance in WS patients of the common effect of music-related improvement in long-term verbal memory. This improvement consists of better recall of sung sentences in comparison to spoken sentences. This effect occurs only in those individuals with WS who had taken formal music lessons, although the emotional reaction to music did not influence the performance on memory tasks. One possible explanation for these findings is that musical learning increases synchronization of neural pathways that support verbal learning and memory (Martens, Jungers, & Steele, 2011).

Music, emotions and sociability in WS

Williams syndrome is a good model for studying the connections between music and mood, not only for the high prevalence of anxiety, but also because WS patients are more likely to have higher ratings of musical skills and more musical training than in other types of mental retardation (Dykens, Rosner, Ly, & Sagun, 2005).

When compared to Prader-Willi syndrome or Down syndrome, WS patients showed less aggressiveness when increasing the frequency of listening to music and lower levels of anxiety/fears when producing music (Dykens et al., 2005). However, WS patients with less symptoms could be more attracted to musical activities in the first place, thus producing a bias in this conclusion; further studies should be performed to assess the possible effects of music therapies in WS anxious profile. In face of the sociability and empathy evidenced by WS patients, it has been suggested that musical activities in a band or chorus, instead of playing alone, could be particularly efficient as cognitive therapy (Dykens et al., 2005).

The reports of parents of WS individuals vividly describe higher levels of emotional engagement with music and sound – namely being consumed by the affective reactions to music. These phenomenological observations are validated by statistical analysis comparing WS with Down syndrome, autism and normal controls: WS show more and earlier interest in music and its effects last longer on these individuals (Levitin et al., 2004). The emotional response to sound in WS can be exemplified by anecdotal reports of sitting for hours enchanted by certain sounds or learning to name cars or vacuum cleaners by their model based only on acoustic information (Levitin et al., 2005). Although emotional connection is the reason why people enjoy music, the way the latter modulates emotional processing is not clear. Thus, WS is a very good model to study the neural mechanisms of emotional connection to music (M. D. Lense, Gordon, Key, & Dykens, 2013).

An enhanced relationship between music and socio-emotional processing in WS is suggested by studies using electroencephalogram oscillatory activity. WS individuals, when compared to controls, show greater gamma band activity when submitted to emotionally congruous stimuli (eg. happy music and a happy face) vs. incongruous ones. These findings are compatible with multisensory integration of music and face processing (M. D. Lense, Gordon, Key, & Dykens, 2013). It is also possible to relate autonomic nervous system activity with the heigtened emotionality to face expressions and music: these stimuli lead to an arousal state, with increased heart rate and changes in electrodermal activity (Jarvinen et al., 2012).

A more complex level of analysis deals with the suggestion by some authors of a possible genetic link between musicality and sociability, given the evolutionary role that music has played in social bonding, and the effect that music has in social interaction for patients with WS, either used as a conversation topic or as a means to create bonds (Levitin et al., 2004). This effect is supported by the empathic nature of WS, and by the double dissociations verified between WS and autism: high vs. low sociability and empathy, high vs low musical engagement; large vs. small neocerebellar volume in the context of a small vs. normal brain volume (Levitin, 2005).

In a questionnaire-based study, Ng and colleagues found that expression of emotions through music may be, in WS, linked to the sensitivity and response to emotions of other people (Ng et al., 2013). Given the social phenotype of WS, namely the gregarious personality (even in the presence of high levels of nonsocial anxiety), links may be found between musicality and social behaviour in WS (Jarvinen, Korenberg, & Bellugi, 2013; Ng et al., 2013). There are multiple ways in which music may be beneficial and even therapeutic in WS. Using music as an aid for raising the levels of emotional expression and affective

awareness seems possible in face of the more entwined correlation between musicality and sociability in WS (Ng et al., 2013). Engaging in musical activities, as already pointed out, decreases anxiety levels and the corresponding maladaptive behaviors (Dykens et al., 2005). And besides social and affective domains, music has been used as a teaching tool in cognitive areas such as mathematics (Ng et al., 2013).

Although much more studies in this field are needed , music appears to be an useful tool to help WS individuals in reaching their full potential. Music therapy, in particular, has been used for social and emotional disturbances in neurodevelopmental disorders; thus, future research will probably emphasize its role in WS (Ng et al., 2013). To sum up, and although patients with neurodevelopmental disorders such as WS, when compared to typically developing people, undoubtedly experience music differently, they persist in enjoying it and can benefit from it (Heaton & Allen, 2009).

The relatively preserved musical skills, contrasting with other impaired higher cortical functions, have raised the possibility that cognitive processes such as music might be independent modules of mind or even an independent type of intelligence (Hopyan et al., 2001; Levitin et al., 2003). In recent years, though, this issue has been treated by neuroscientists more as the existence of specificity domain rather than a strong modularity, which would oppose a developed musicality to a lower general cognition (Levitin, 2005).

Recent studies about WS and music: what has been done and its limitations

In recent years, many studies about WS and music have been conducted (Table 1). However, it has to be noted that there are many limitations when it comes to studies with WS patients.

The first limitation is the difficulty to obtain a reasonable number of participants with WS in studies, given the frequency of the syndrome (Barozzi et al., 2012).

In addition, those based in questionnaires answered by parents are obviously limited by their recall and possible over-/underestimations, as well as by the higher verbal and emotional expressiveness in WS, when compared to autism or Down syndrome (Dykens et al., 2005; Levitin et al., 2005). The use of interviews with WS patients themselves has shown that reports from relatives tend to underestimate their problems (Blomberg et al., 2006). However, it is not a perfect method, given both the cognitive deficits and the urge to please others that are present in WS (M. Lense & Dykens, 2013).

One limitation frequently present in studies about WS is the absence of comparison groups of other developmentally disabled people. The inclusion of Autism and Down syndrome, as well as age-matched controls, can test the null hypothesis that any observations are due to neurodevelopmental impairment or delay in general and not specifically connected to WS (Levitin et al., 2005).

In the context of studies using fMRI, the hyperacusis and auditory aversions that characterize the syndrome, prevented WS patients to cooperate with the procedures (Levitin et al., 2003). Levitin and his team developed a systematic orientation program that overcame those difficulties by reducing the patient's fear of the scanners. Another problem that can arise in this context is the existence of certain contraindications to MRI, such as aortic valve prostheses (Wengenroth et al., 2010).

In studies of sound reactivity, one important aspect is that objective audiological examinations need to be performed in a standard laboratory setting, in an attempt to differentiate between pure anxiety and the auditory abnormalities seen in the context of WS (Barozzi et al., 2012; Gallo et al., 2008).

The characteristic attentional deficits in WS patients make it difficult to use tests that require paying attention. This difficulty can be managed, for instance, by administering tests personally instead of using computerized versions (Levitin et al., 2004). In studies with music, differences in the musical domains tested, as well as the difficulties of some WS individuals in understanding the concept of same/different, are limitations to be taken into account (Hopyan et al., 2001). Besides, musical knowledge and training is sometimes required to some studies and it is quite uncommon to get to contact with WS patients in these conditions (Martinez-Castilla et al., 2013).

Conclusion

Williams syndrome is a unique disorder in many aspects, and its musical phenotype is certainly one of them. As this review made clear, the wide range of factors that modulate WS patients' response to sounds and music make it an extremely interesting field of research for the future. Even if it is difficult to fully understand the role played by music in WS, all these aspects, from neurobiology and audiology to social and behavioural sciences, suggest that such a role is not a small one. Future studies in WS will not only contribute to better understanding music cognition as a whole, but they will also provide useful data on the multiple ways that these individuals can benefit from musical therapy.

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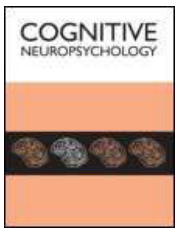
Table 1: Recent studies on Williams Syndrome and Music Abbreviations: WS- Williams Syndrome; TD- typically developing

Authors	**Year**	**Sample (and control groups)**	**Main methods used**
Ng, R. *et al.*	2013	55 adults with WS; 19 age-matched TD controls	Questionnaires; verbal assessment
Lense, M; Dykens, E.	2013	46 children and adults with WS	Questionnaires; cognitive and musical skills assessment; musical instrument (dulcimer) lessons, followed by an interview
Lense, M; Shivers, CM; Dykens, E.	2013	73 adolescents and adults with WS	Questionnaires; cognitive, sound perception and singing assessment
Barozzi, S *et al.*	2013	24 children with WS	Pure-tone audiometry; acoustic immittance measurements; evoked otoacoustic emissions
Lense, M; Gordon, R *et al.*	2013	13 young adults with WS; 13 age-matched TD controls	Observation of reaction to auditory stimuli and facial expressions; electroencephalogram (EEG)
Martinez-Castilla, P *et al.*	2013	Study 1: 7 musically trained adolescents and adults with WS; 14 musically trained TD controls; 2 musicians with absolute pitch Study 2: 27 adolescents and adults with WS; 54 TD controls; 2 musicians with absolute pitch	Study 1: pitch-identification task; Study 2: pitch memory test

Jarvinen, A *et al.*	2012	Experiment 1: 20 individuals with WS; 27 TD controls Experiment 2: 20 individuals with WS; 26 TD controls	Experiment 1: cognitive assessment; visual affect identification task; electrodermal activity and electrocardiogram measurements Experiment 2: cognitive assessment; audiometry; auditory affect identification task; electrodermal activity and electrocardiogram measurements
Barozzi, S *et al.*	2012	69 children and young adults with WS	Otoscopic examination; comprehensive audiological assessment: pure-tone audiometry, speech audiometry, timpanometry and measurement of the acoustic reflex, evoked otoacoustic emissions, brainstem auditory evoked response
Martens, MA *et al.*	2011	2 groups, each one with 38 children and adults with WS	Questionnaires; memory tasks
Wengenroth, M *et al.*	2010	36 children and adolescentes with WS; 20 age-matched TD controls	Sound perception test; magnetoencephalography; magnetic resonance imaging
Martens, MA *et al.*	2010	25 children and adults with WS; 25 age-matched TD controls	Musical tasks; magnetic resonance imaging

Thornton-Wells, TA *et al.*	2010	Study 1: 13 adolescents and young adults with WS; 13 age-matched TD controls Study 2: 6 WS individuals from study 1 who showed activations to musical stimuli in occipital lobe Study 3: 4 WS individuals from study 2	Study 1: musical stimuli and functional magnetic resonance imaging Study 2: musical stimuli and functional magnetic resonance imaging; retinotopy and color localizer experiments Study 3: musical and non-musical stimuli and functional magnetic resonance imaging
Gallo, FJ *et al.*	2008	21 children with WS; 20 children with other developmental disabilities of mixed etiology	Assessment of developmental functioning and social interaction with exposure to mild intensity sounds
Blomberg, S *et al.*	2006	38 children and adults with WS	Questionnaires
Dykens, E *et al.*	2005	Study 1: a total 89 children and adults; with WS (n=31), Prader-Willi syndrome (n=26) and Down syndrome (n=32) Study 2: a total 67 children and adults; with WS (n=26), Prader-Willi syndrome (n=16) and Down syndrome (n=25)	Study 1: questionnaires Study 2: anxiety and musical measurements
Levitin, DJ *et al.*	2005	Parents of people with WS (n=118), Down syndrome (n=40), autism (n=30) and TD controls (n=118)	Questionnaires

Levitin, DJ *et al.*	2004	Parents of people with WS (n=118), Down syndrome (n=40), autism (n=30) and TD controls (n=118)	Questionnaires
Levitin, DJ *et al.*	2003	5 adults with WS	Musical stimuli and functional magnetic resonance imaging
Hopyan, T *et al.*	2001	14 children and adolescentes with WS; 14 age-matched TD controls	Musical tasks



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