In Press: Research in Development Disabilities

20th April 2020

Heaton, P., Ridley, E., Makhmood, S. & Riby, D. M.

Hearing the feeling: Auditory emotion perception in Williams Syndrome

Corresponding Author. Pamela Heaton.

P.Heaton@gold.ac.uk

Psychology, Goldsmiths University of London,

New Cross, London, SE14 6NW.

Ellen Ridley. ellen.ridley@durham.ac.uk

Department of Psychology, Durham University,

Upper Mountjoy, South Road, Durham DH1 3LE.

Sonya Makhmood. S.Makhmood@gold.ac.uk

Psychology, Goldsmiths University of London,

New Cross, London, SE14 6NW.

Deborah Riby. deborah.riby@durham.ac.uk

Centre for Developmental Disorders,

Department of Psychology, Durham University,

Upper Mountjoy, South Road, Durham DH1 3LE.

Abstract.

Background. Studies investigating recognition of facial expressions of emotions in Williams syndrome (WS) have reported difficulties in recognising negative expressions of emotion and a reliance on atypically developing underlying processes during task performance.

Aim. The aim of the study was to extend these findings to the recognition of emotions in auditory domains.

Method and Procedures. Children and adolescents with WS, together with chronological (CA) and verbal mental age matched (VMA) typically developing (TD) comparison groups, were asked to judge expressions of happiness, sadness, anger, and fear in vocal and musical conditions.

Outcomes and Results. Total emotion recognition scores did not differ between WS and VMA matched groups but profiles of discrimination across emotion categories were markedly different. For all groups, the accessibility of emotion category cues differed across music and speech domains. The results suggested that emotion discrimination is more strongly linked with cognitive ability in WS than in TD.

Conclusions and implications. Although WS and TD groups showed a significantly different profile of discrimination across emotion categories, similarities in the pattern of discrimination across domains and in the correlates of auditory emotion processing were observed. The results are discussed in the context of typical and atypical developmental trajectories and compensatory mechanisms in WS.

What this paper adds. This paper contributes to work on the social/emotional and cognitive phenotype in WS. It extends studies investigating discrimination of emotions from faces by exploring the pattern and cognitive correlates of emotion recognition within the auditory domain. Consistent with face processing studies we observed difficulties discriminating negative emotions from vocalisations and music in children and adolescents with WS. However, we analysed the structure and correlates of emotion recognition across domains and groups and this suggested important similarities in the architecture of auditory emotion recognition in WS and TD. Whilst atypical development of the amygdala and other neural structures places constraints on emotion recognition in WS, our results revealed considerable variability and positive correlations between emotion recognition, age, intelligence and musical experience. We propose that cognitive skills and musical experience may function as compensatory mechanisms in WS.

1. Introduction. Williams syndrome (WS) is a relatively rare neurodevelopmental disorder with a reported prevalence between 1 in 7,500 and 1 in 20,000 live births (Morris, Demsey, Leonard, Dilts & Blackburn, 1988; Strømme, Bjørnstad & Ramstad, 2002). It is caused by a hemizygous deletion of approximately 28 genes on chromosome 7q11.23 (Tassebehji, 2003) that results in mild to moderate intellectual disability and a highly uneven profile of cognitive skills. Within the cognitive domain, and in relation to their overall intellectual ability, individuals with WS often show markedly stronger performance on verbal compared with non-verbal tasks, particularly where the latter have a visual-spatial component (Bellugi, Wang, & Jernigan, 1994; Jarrold, Baddeley & Hewes, 1999; Donnai & Karmiloff-Smith, 2000). The co-occurrence of skills presumed to be relatively more intact or impaired across different cognitive domains, was taken as early evidence in support of a modular

account of brain organisation (Fodor, 1983; 1985; Pinker, 1991). However, many of the more recent studies of WS have adopted the developmental approach advocated by Karmiloff-Smith and colleagues, and have revealed fine-grained impairments within cognitive domains previously believed to be relatively intact (Karmiloff-Smith, Grant, Berthoud, Davies, Howlin & Udwin, 1997), and a reliance on atypically developing underlying processes during task performance (Karmiloff-Smith, 2008; 2011; Westerman, Mareschal, Johnson, Siois, Spratling & Thomas, 2007; Johnson, 2011; Thomas, Purser & Richardson, 2013).

A highly salient characteristic of the WS social phenotype is an increased propensity for social engagement (e.g. Doyle, Bellugi, Korenverg & Graham, 2004) and a greater interest in social than non-social stimuli (Järvinen, Korenberg & Bellugi, 2013; -Järvinen -Pasley et al., 2008a; Martens, Wilson & Reutens, 2008, Riby & Hancock, 2008; 2009). Järvinen, Ng, Crivelli, Arnold, Woo-Von Hoogenstyn and Bellugi (2015) investigated associations between responses to social stimuli, social functioning and -autonomic reactivity in WS, and showed that elevated autonomic arousal to faces was positively associated with levels of social functioning in this group. Atypically increased attention to faces (Riby & Hancock, 2008) is evident early in development (Mervis, Morris, Klein-Tasman, Bertrand, Kwitny, Appelbaum & Rice, 2003) and some aspects of face recognition in WS are commensurate with chronological age (CA) (Bellugi, Wang & Jernigan, 1995; Plesa-Skewere, Faja, Schofield, Verbalis & Tager-Flusberg, 2006; Annaz, Karmiloff-Smith, Johnson & Thomas, 2009). However, identification of emotional expressions from faces in WS is most frequently in line with mental age (MA) (Gagliardi et al., 2003;Lacroix, Guidetti, Toge & Reilly, 2009; Plesa-Skwerer, Faja, Schofield, Verbalis & Tager-Flusberg, 2006a; Plesa-Skwerer, Verbalis, Schofield, Faja & Tager-Flusberg, 2006b;

Porter, Coltheart & Langdon, 2007; Porter, Shaw & Marsh, 2010) with declines in performance when judging negative expressions of emotion (Plesa-Skwerer et al., 2006 a,b; Porter et al., 2007; Porter, Shaw & Marsh, 2010). The study of developmental trajectories has provided important insights into cognitive skills in WS (Paterson, Brown, Gsodl, Johnson & Karmiloff-Smith, 1999; Karmiloff-Smith, Thomas, Annaz, Humphreys, Ewing, Brace, et al., 2004) and linked to this method for studying emotion recognition, the results from two studies have reported an absence of age-related gains in emotion recognition in this group (Gagliardi et al., 2003; Martinex-Castilla, Burt, Borgatti & Gagliardi, 2015). In these studies recognition performance increased in line with age in TD and in line with intelligence in WS.

Consistent with results from face perception studies, research investigating the recognition of vocal emotions has revealed developmental delays that are more marked when emotions are negatively valenced (Plesa-Skwerer, Faja, Schofield, Verbalis & Tager-Flusberg, 2006; Järvinen-Pasley, Pollak, Yam, Hill, Grichanik, Mill & Bellugi, 2010b). In one study, Plesa-Skwerer and colleagues (2006a) administered the paralanguage subtests of the Diagnostic Analysis of Nonverbal Accuracy Scale (Norwicki & Duke, 2001) to test recognition of vocal expressions of emotions in participants with WS, intellectual disability and TD. The results showed that participants with WS recognised happy vocal emotions as well as CA-matched TD participants, whilst recognition of sad, angry and fearful vocal emotions was less accurate than that of CA-matched TD controls, and similar to that of participants with comparable intellectual ability. In a more recent study, Järvinen, Ng, Crivelli, Neumann, Arnold, Woo-Von Hoogenstyn, Lai, Trauner & Bellugi and colleagues (2016) showed that discrimination of happy, sad and fearful, vocal and musical stimuli did not differ across groups with WS, Autism Spectrum Disorder (ASD) and

TD once differences in intellectual ability were taken into consideration. However, autonomic nervous system (ANS) reactivity to auditory stimuli was also measured and revealed marked differences across groups. In comparison with TD children, children with WS showed a less systematic pattern of autonomic responsivity to the different emotion stimuli and also failed to show a habituation effect. Both clinical groups showed increased arousal to vocal stimuli compared with TD, and the WS group also showed increased arousal to music.

Studies investigating auditory processing across language and music domains are important to debates on modularity, and may increase our understanding of development in WS. Although musical impairments in acquired brain injury have been discussed in the context of modularity theory (Peretz & Coltheart, 2003), music psychologists have become increasingly interested in the shared evolutionary origins of music and language and the processes involved in speech and music perception (Patel, 2008). Brown (2000) has proposed that music and language developed in tandem from an early and highly expressive form of vocal communication, termed musilanguage. Consistent with this account are results from neuroimaging studies (Knösche, Neuhaus, Haueisen, Alter, Maess, Witte & Friederici, 2005; Patel, 2004; Maess, Koelsch, Gunter & Friederici, 2001; Tillman, Janata, & Bharucha, 2003; Koelsch, Gunter, Cramon, Zysset, Lohmann & Friederici, 2002) showing that many of the same cognitive and neural resources are recruited during music and speech processing. In addition to investigating cognitive and neural processes involved in speech and language perception, commonalities in the types of informational content within these domains has been investigated. Juslin and Lauukka (2003) reviewed studies of vocal emotions and musical performance and showed that emotions in music and speech were signalled by the same patterns of psychoacoustic cues. Vocal

and musical expressions of different emotions (anger, fear, happiness, sadness and tenderness) are communicated by the same, unique patterns of intensity, energy, pitch level, variability, contour and microstructural irregularity. Studies investigating perception of vocal and musical emotions have reported significant correlations in identification scores across conditions for both TD adults (Laukka & Juslin, 2007) and children (Allgood & Heaton, 2015) and are consistent with a shared resources model, for components of music and speech processing (Patel, 2008).

To our knowledge, only one study has studied auditory processing in WS in the context of modularity theory. Motivated by prior work showing that pitch in music and prosody rely on common processing mechanisms in TD (Dankovicova, House, Crooks & Jones, 2007; Magne, Schon & Besson, 2003), Martinex-Castilla and Sotillo (2014) tested pitch discrimination in musical and prosodic stimuli in children and adolescents with WS and TD. The results revealed a significant correlation between scores on the musical and prosodic pitch tasks for both groups. As the authors concluded, these results challenge modular accounts of music and language processing in WS (Levitin & Bellugi, 1998; Pinker, 1991) and suggest similarities in the architecture of pitch processing across WS and TD groups.

Experimental studies of music perception in WS require careful consideration in terms of research design. Thomas, Annaz, Ansari, Scerif, Jarrold and Karmiloff-Smith, (2009) have provided a strong case for the use of a developmental trajectory approach in studies of neurodevelopmental disorders. However, the main aim of the current study was to extend work on recognition of facial emotions in WS, and these studies have typically matched comparison groups on the basis of verbal mental age (VMA) and chronological age (CA). Furthermore, studies investigating identification

of emotions in music and vocalisations have reported increases in line with CA in TD children (Heaton, Allen, Williams, Cummins, & Happe, 2008; Sauter, Panatonni & Happe, 2013) and in line with VMA in Autism Spectrum Disorder (Heaton et al., 2008; Quintin, Bhatara, Poissant, Fombonne & Levitin, 2011) and Down Syndrome (Heaton et al., 2008). We therefore included CA and VMA matched TD groups for comparison and investigated the processes involved in emotion recognition in TD and WS. A second important consideration in the design of the study concerns the effects of day-to-day musical experience on auditory processing skills in childhood. In a recent review article, Thakur, Martens, Smith and Roth (2018), reported that 47% of studies investigating musical skills in WS recruited participants at a music summer camp or a national convention. The potential for bias is obvious. The importance of controlling for musical experience across comparison groups in experimental studies is highlighted by work on musical enrichment in TD children. Schon, Magne and Besson (2004) showed that musical training in childhood improves pitch acuity for both music and language and there is evidence showing that relatively short periods of musical training during childhood influence the development of the brain. For example, Schlaug, Norton, Overy and Winner (2005) reported enhanced activation of the bilateral temporal lobes and superior temporal gyri during rhythmic and melodic discrimination tasks in five to seven year old children after just 12 months of musical training. In our study no participants were recruited via a specialist music provision and we measured day-to-day musical experiences in both WS and TD participants.

The overarching aim of the current study was to investigate recognition of emotions in music and vocalisations in WS and TD. VMA has been shown to predict overall levels of facial emotion recognition in WS (e.g. Plesa Skwerer et al., 2006), and is significantly correlated with recognition of musical and vocal emotions in ASD

(Heaton et al., 2008; Quintin, Bhatara, Poissant, Fombonne & Levitin, 2011) and DS (Heaton et al., 2008). Therefore our first hypothesis was that overall levels of discrimination would be commensurate with VMA in the WS group. Motivated by studies showing an atypical trajectory of emotional face processing skills in WS (Gagliardi et al., 2003; Martinex-Castilla, et al., 2015), our second hypothesis was that the cognitive correlates of emotion recognition and the pattern of discrimination performance would distinguish WS and TD groups.

2. Methods.

2.1 Participants.

15 participants with WS were recruited via a local research database and through collaboration with the Williams Syndrome Foundation UK. All participants had previously had their diagnosis confirmed with genetic fluorescent in situ hybridisation testing. Two groups of typically developing children (TD) were recruited from mainstream state schools in the North East and South East of England and through local research databases for families and children. The first TD group (n = 18) was matched to the WS group for chronological age and the second TD group (n = 19) was matched to the WS group for verbal mental age, using age equivalence scores from the British Picture Vocabulary Scale (BPVS II; Dunn, Whetton, & Burley, 1997). Non-verbal intelligence was assessed using the Raven's Coloured Progressive Matrices test (RCPM; Raven, Court & Raven, 1990) with a maximum possible score of 36. Full sample demographics are provided in Table 1. All participants were screened for their day-to-day musical experience. Parents and carers were asked whether their child participated in (a) individual music lessons (b) class music lessons (c) music therapy (d) dance/movement, on a weekly basis. For each positive response they were asked whether this activity took half an hour (score = 1),

one hour (score = 2), one and a half hours (score = 3) or two plus hours (scored = 4) each day (max score = 16). Participant data regarding musical experience are also shown in table 1.

Table 1. Description characteristics of participants with Williams Syndrome and typically developing comparison participants

Measures	Mean (SD) range		
	Williams Syndrome	CA matched group	VMA matched group
N	15	18	19
CA	11.60 (3.08)[1] 6-16	10.73 (3.13)[1] 6-16	6.44 (1.72) 4-10
BPVS AQ score	6.53 (2.58)[2]	11.37 (3.96)	7.06 (2.16)[2]
RPCM Raw score	15.13 (8.25)	30.83 (4.73)	24.37 (7.35)
Weekly musical engagement	2.63 (2.13) 0–7	2.37 (2.36) 0-8	2.21 (2-4) 0-9

1 No significant difference between

WS and CA groups on	
chronological age t = .80, p=.43	
2 No significant difference between	
WS and VMA groups on verbal	
ability t = -.66, p=.52	

2.2 Procedure and stimuli

Participants began testing by completing the BPVS II and the RPCM tests to evaluate verbal and non-verbal abilities. Participants were then asked to listen to vocalisations and musical excerpts evoking happy, sad, fearful and angry emotions.

The experiment included 64 trials, organised in 2 blocks of 32 musical excerpts and two blocks of 32 vocalisations. Each block included 4 happy, 4 sad, 4 fearful and 4 angry stimuli randomised across emotion type. The vocal stimuli were developed by Sauter (2006). Adult female and male actors expressed happy, sad, fearful and angry emotions non-verbally (e.g. crying/laughter). The fearful, sad and happy musical stimuli were taken from a set developed by Quintin, Bhatara, Poissant, Fombonne & Levitin, (2011) and the angry musical stimuli were sourced from a set developed by Eerola and Vuoskoski (2010).

The presentation of the auditory stimuli adopted the method used in a previous study investigating auditory emotion recognition in 5 – 10 year old TD children (Algood &

Heaton, 2015). The music clips were 30s long and the vocalisations were repeated 3 times in a 10s time frame at 0, 3 & 6s. This ensured equal exposure to emotion cues across the two conditions. Vocal and musical blocks were counterbalanced across participants.

As an introduction to the task, participants were presented with four cartoon faces depicting the four emotions (happy, sad, fearful, and angry) and the researcher probed their understanding of the emotions (e.g. "tell me about a time when you felt very happy?"). In order to proceed to the experimental trials, participants had to correctly label the emotions expressed by the cartoon faces. Throughout the task participants indicated their response verbally to the researcher or by pointing to the corresponding cartoon face. Responses were recorded for accuracy.

All testing sessions were completed in a quiet setting, either at home, in the local University research facilities, or in school depending on the needs of the participant. Participants received a certificate of participation. Ethical approval for the study was granted by Goldsmiths, University of London, and Durham University.

3. Results

The raw data (32 music trials and 32 vocal trials) are shown as % scores in table 2.

Table 2. Experimental data (shown as % scores) for WS and TD participants

Condition	Mean (SD)	Range

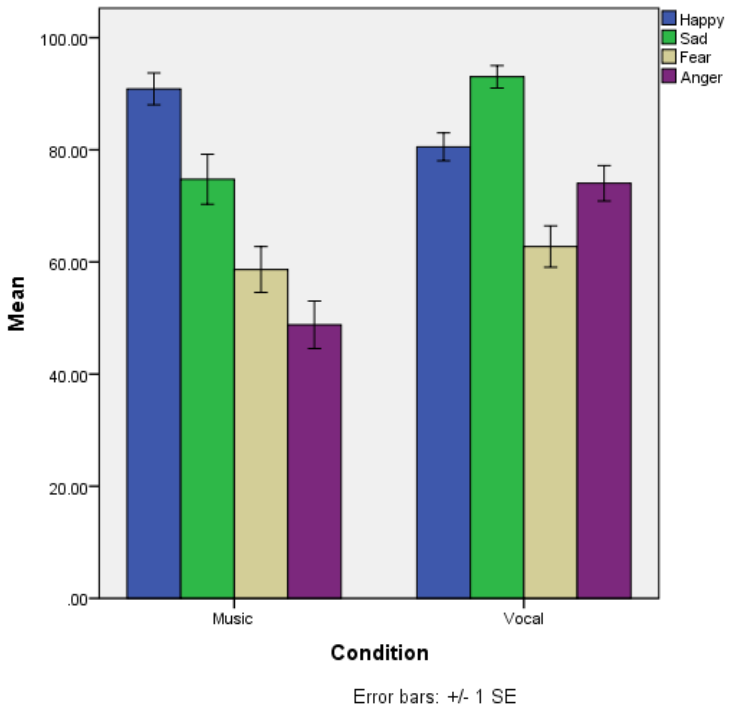
Group			
	WS	CA group	VMA
Vocal Condition			
Total	76.46 (15.60) 47–97	83.16 (7.74) 69–97	73.52 (11.47) 47-91
Happy	85.00 (17.80) 50-100	85.42 (15.61) 50-100	72.37 (17.96) 38-100
Sad	90.00 (16.50) 50-100	97.92 (4.80) 88-100	90.79 (18.09) 25-100
Fear	59.17 (30.42) 13-100	70.84 (20.02) 38-100	72.37 (23.42) 25-100
Anger	70.83 (25.73) 13-100	78.47 (20.02) 38-100	72.30 (23.41) 25-100
Music Condition			
Total	60.42 (20.78) 22-91	80.39 (13.63) 59-97	63.00 (21.27) 47-91
Happy	87.50 (22.16) 0-100	97.22 (9.15) 63-100	87.50 (25.60) 13-100
Sad	59.17 (33.22) 0-100	91.67 (14.22)50-100	71.05 (37.05) 0-100
Fear	65.00 (31.05) 0-100	62.50 (30.62) 00-100	50.00 (26.35) 0-88
Anger	30.00 (18.17) 0-75	70.14 (25.77) 25-100	43.42 (31.28) 0-100

A 3 x 2 x 4 analysis of variance with group (WS, CA, VMA) as the between subjects variable with condition (vocal, music) and emotion category (happy, sad, fear and anger) as the within subjects variables were carried out on the data. Levene's homogeneity of variance test carried on the group variable showed a non-significant result (p = .071, n.s.). Mauchly's sphericity test on the emotion variable was not

significant (p = .202). Mauchly's test of sphericity for the condition by emotion interaction was significant, so lower bound estimates of significance were used for testing effects involving this interaction.

There was a significant main effect of condition ($F(1, 49) = 19.7$, $p < .001$, partial $\eta^2 = .286$), and the condition by group interaction was not significant ($F(2, 49) = 2.9$, $p = .063$, n.s.). All groups scored higher in the vocal than in the musical condition. The main effect of emotion category was significant ($F(3, 49) = 45.6$, $p < .001$), partial $\eta^2 = .48$) and the condition by emotion interaction was significant: $F(1, 49) = 15.1$, $p < .001$, partial $\eta^2 = .235$. The three way group by condition by emotion interaction was not significant ($F(2, 49) = 2.24$, $p = .117$, n.s.). The condition by emotion interaction is shown in figure 1.

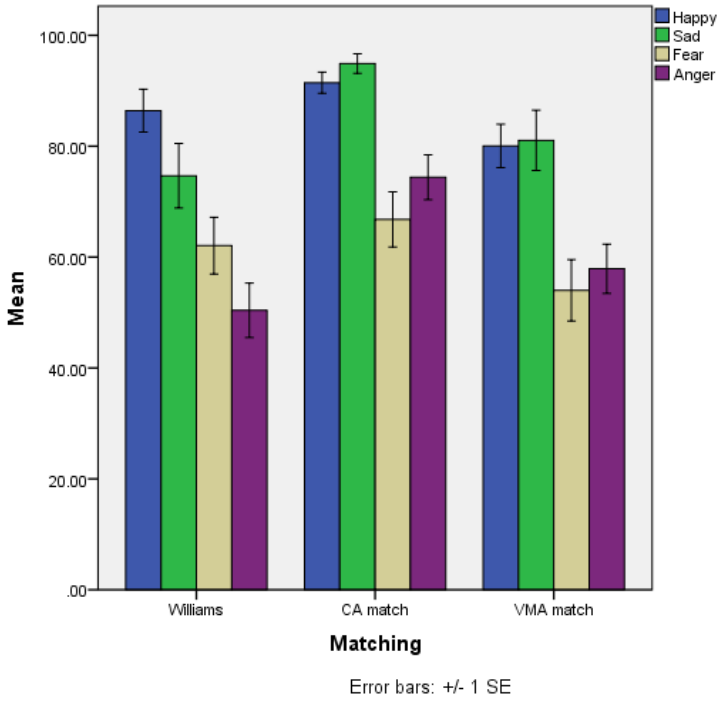
Figure 1: Condition by emotion category interaction.



Paired comparisons for the music vs vocal conditions for each emotion showed that discrimination of happy stimuli was significantly higher in the music condition (Percentage mean difference = 10.8, $t(51) = 2.9$, $p = .005$), and discrimination of sad (mean difference = 18.3%, $t(51) = 4.7$, $p < .001$) and angry (mean difference = 25.2%, $t(51) = 5.25$, $p < .001$) was significantly higher in the vocal condition. Discrimination of fearful stimuli did not differ across conditions (mean difference = 4.1%, $t(51) = .86$, n.s.).

The main effect of group was significant: ($F(2, 49) = 5.8$, $p = .005$, partial $\eta^2 = .191$. Post hoc analysis using Tukey's test revealed higher scores in the CA group than the VMA group (by 13.6%, p = .011) and the WS (by 13.4%, p = .019). Scores for the VMA and WS were not significantly different (0.16% p = .999, n.s.). The group by emotion interaction was significant $F(6, 147) = 2.17$, $p = .049$, partial $\eta^2 = .081$ and is shown in figure 2.

Figure 2: Group by emotion category interaction



The CA and VMA groups appeared to show the same response profile across emotions and a repeated measures analysis carried out on the two TD groups showed that the group by emotion interaction was not significant ($F(3, 105) = .172$, $p = .915$). A second repeated measures, comparing a combined TD group (VMA & CA) with the WS group showed a highly significant group by emotion interaction ($F(3, 150) = 4.2$, $p = .007$, partial $\eta^2 = .077$). One-way ANOVAs carried out on the different emotions showed a significant difference between TD and WS for sad, ($F(1, 51) = 4.61$, $p = .037$, $\eta^2 = .084$) and anger ($F(1, 51) = 6.7$, $p = .013$, $\eta^2 = .118$). The group difference was not significant for fear ($F(1, 51) = .073$, $p = .788$, $\eta^2 = .001$) or for happy ($F(1, 51) = .032$, $p = .858$, $\eta^2 = .001$).

Correlations carried out on the total scores for the music and vocalisation conditions were highly significant for the VMA group ($r=.70$, $p=.01$) and the WS group ($r=.59$, $p=.02$) but not for the CA group ($r=.17$, $p=.50$). Correlations between total vocal and musical scores and background data are shown in table 3.

Table 3. Correlations between background variables and vocal and musical discrimination scores.

Measures	Williams Syndrome		CA matched group		VMA matched group		Combined TD group	
	Music	Vocal	Music	Vocal	Music	Vocal	Music	Vocal
CA	.47	.23	.56*	.23	.75**	.58**	.68**	.53**
BPVS	.85***	.41	.71**	.11	.56*	.40	.66**	.42*
AQ Score								
RPCM	.82***	.32	.84**	.18	.59**	.53*	.73**	.55*
Raw score								
Musical	.44	.48	.35	.12	.51*	.61**	.41*	.40*
Engagement								

*p < .05. **p < .01. ***p < .001

Correlations between specific emotions and background data are shown in table 4.

Table 4. Correlations between background variables and discrimination scores for emotion categories.

Measures		CA	BPVS	RPCM	MusExp
WS	Happy	.43	.49	.53*	.30
	Sad	.52*	.69**	.77**	.51
	Angry	.54*	.66**	.66**	.63*
	Fear	.24	.48	.54*	.29
CA	Happy	.16	.13	.23	-.06
	Sad	.32	.36	.63**	.21
	Angry	.30	.25	.36	.26
	Fear	.50*	.62**	.61**	.26
VMA	Happy	.64**	.52*	.55*	.53*
	Sad	.59**	.40	.70**	.53*
	Angry	.43	.22	.14	.52*
	Fear	.50*	.44	.35	.17
Comb	Happy	.50**	.44**	.57**	.33
TD	Sad	.50**	.43**	.72**	.40*
	Angry	.50**	.40*	.37*	.39*
	Fear	.52**	.56**	.50**	.21

*p < .05. **p < .01. ***p < .001

4. Discussion

In the study of music and vocal emotion recognition, participants with WS performed at a level that was broadly in line with their mental, but not their chronological age. This finding is consistent with studies investigating the discrimination of facial expressions of emotion in WS (Gagliardi et al., 2003; Lacroix, et al., 2009; Plesa-Skwerer et al., 2006a; Plesa-Skwerer et al., 2006b; Porter, et al. 2007; 2010). However, Thomas and Karmiloff-Smith (2002) have discussed how 'residual normality', or broadly comparable task performance across typical and atypically developing groups can mask differences in the underlying cognitive processes involved in task performance. An aim of the study was therefore to investigate patterns of performance within as well as across the groups.

One similarity between the groups was that vocal and musical identification scores were highly correlated for WS and VMA groups. Martinex-Castilla and Sotillo (2014) reported a significant correlation between scores on musical and linguistic pitch processing tasks in participants with WS and TD and our results also suggest similarities in the architecture of auditory processing in WS and younger TD children. In our study scores did not correlate across conditions for the older CA matched group and this may reflect higher and less widely distributed scores in this group.

The pattern of accuracy across auditory domains did not differ across groups. Total recognition scores were higher in the vocal than the musical condition for CA (2.8%), VMA (10.5%) and WS (16%) groups. However correct identification of happy was higher in the music condition and did not differ across conditions for fear. The most salient difference across WS and TD groups was seen in the pattern of responses to the different emotion categories. For both TD groups scores were highest for sad, with a small decrease for happy, and larger decreases for anger and fear categories. The WS group scored highest on the happy condition and their pattern of discrimination for sad, angry and fearful emotions was very different to that of controls. For example, there was a sharp decrease in identification of fear compared with sad stimuli for the VMA (27.3%) and CA (27.8%) groups, whilst this difference was small (12.6%) for the WS group. The group comparisons showed that WS and TD group differed significantly on sad and anger but not on fear and this is likely to result from difficulties in distinguishing negative emotions in the WS group. A similar pattern of auditory emotion discrimination has been reported in earlier behavioural studies (e.g. Plesa-Skwerer et al., 2006 a,b), and has been linked with atypical brain development in this group (Haas & Reiss, 2012). For example, increased attention and heightened responses to happy faces (Haas, Mills, Yam, Hoeft, Bellugi & Reiss, 2009; Dodd & Porter, 2010) and reduced arousal in response to fearful and angry faces (Meyer-Lindenberg, Hariri, Munoz, Mervis, Mattay, Morris & Berman 2005; Haas et al., 2009; Plesa-Skwerer et al., 2009) have been associated with altered amygdala volume (Reiss, Eckert, Rose, Karchemskiy, Kesler, Chang, Reynolds, Kwon, & Galaburda 2004; Martens, Wilson, Dudgeon, & Reutens, 2009; Capitao, Sampaio, Sampaio, Vasconcelos, Fernandez, Garayzabal, Shenton, & Goncalves, 2011b) and function (Meyer-Lindenberg et al., 2005; Haas et al., 2009). The

amygdala is involved in music perception (Blood & Zattore, 2001), and patient data shows that amygdala damage impairs recognition of musical expressions of fear (Gosselin, Peretz, Nulhaine, Hasboun, Beckett, Baulac & Samson, 2005). Consistent with studies showing atypical development of the amygdala and associated neural structures, emotion identification scores for the WS group were not CA equivalent. However, it is important to note that emotion discrimination was not uniformly low in this group, and one participant with WS achieved exceptionally high levels of identification of sad (100%), angry (88%) and fear (94%) stimuli. Juslin and Lauukka (2003) have shown that recognition of specific vocal and musical emotions relies on the identification of different configurations of psychoacoustic cues. Good auditory emotion recognition may then reflect strengths in the cognitive abilities recruited during task performance and the extent of the individual's levels of exposure to emotional auditory stimuli.

The correlations carried out on the combined TD data (4 – 16 yrs) provided insights into factors associated with developmental increases in emotion recognition within vocal and musical domains. For the combined sample of TD participants, recognition of musical and vocal emotions was positively associated with CA, VMA, non-verbal intelligence and musical engagement. For WS, musical emotion recognition was highly correlated with VMA and non-verbal intelligence, and showed moderate to large effect sizes for CA and musical engagement. Correlations carried out on the verbal condition data for the WS group, showed moderate to large effects sizes for VMA, non-verbal intelligence and musical engagement but were not statistically significant.

As profiles of emotion category identification sharply distinguished WS and TD groups, these scores also correlated with background data. For TD participants identification scores for all emotion categories were significantly correlated with CA, VMA and non-verbal intelligence and sad and angry also correlated with musical experience. Previous studies investigating recognition of facial expressions of emotion showed that scores increased in line with age in TD and in line with intelligence in WS (Gagliardi et al., 2003; Martinex-Castilla, Burt, Borgatti & Gagliardi, 2015). In our study recognition scores for sad and angry emotions were positively correlated with age. However levels of VMA and non-verbal intelligence were more strongly associated with correct identification of negative emotions in WS than in the TD groups. This suggests that intellectual strengths may enable a degree of compensation during emotion recognition in WS. It was interesting to note that scores on the emotion categories were either significantly correlated with the measure of musical engagement or showed large to medium effect sizes for the WS group. This finding supports and extends prior work highlighting the value of musical engagement for individuals with WS (Dykens, Rosner, Ly & Sagun, 2005).

The results from our study show that emotion recognition in WS should be studied from a developmental perspective. WS is a relatively rare disorder, and in common with many other studies, our interpretation of the results is constrained by group size. Our decision to use a group-matched designed was informed by studies showing VMA levels of emotional face recognition in WS and findings showing that vocal and musical emotion recognition increases in line with VMA in developmentally atypical groups. Consistent with criticisms of group matching in studies of neurodevelopmental disorders (Thomas et al., 2009) the between group comparisons were less informative than the within group analyses. The comparison of group means

suggested that auditory emotion recognition is broadly commensurate with VMA in WS, despite a marked difference in the pattern of discrimination across WS and TD groups and significant within group heterogeneity. Impairments in recognising negative emotions have been linked with abnormalities in the form and function of the amygdala in WS (Haas & Reiss, 2012). However, our study provides clear evidence for age, ability and experience related increases in auditory emotion recognition during childhood and adolescence in this group. Karmiloff-Smith (1998) proposed that development is the key to understanding developmental disorders, and our results fully endorse this insight. The identification of factors associated with gains in auditory emotion recognition in WS has implications for our understanding of this disorder, and may also help in the formulation of future educational and therapeutic approaches.

Acknowledgements. We thank all the individuals who participated in our study. We also thank Maurice Douglas and Sasha Matock for help with stimulus preparation. Funding: This work was supported by the Baily Thomas Charitable Fund.

Allgood, J. & Heaton, P. (2015). Developmental change and cross-domain links in vocal and musical emotion recognition performance in childhood. *British Journal of Developmental Psychology*, **33**, (2), 398-403.

Annaz, D., Karmiloff-Smith, A., Johnson, M. H., and Thomas, M. S. (2009). A cross-syndrome study of the development of holistic face recognition in children with autism, Down syndrome, and Williams syndrome. *J. Exp. Child Psychol.* **102**, 456–486.

Bellugi, U., Wang, P. P. & Jernigan, T. L. in *Atypical Cognitive Deficits in Developmental Disorders: Implications for Brain Function* (eds Broman, S. H. & Grafman, J.) 23–56 (Erlbaum, Hillsdale, New Jersey, 1994).

Blood, A.J. & Zattore, R.J. (2001). Intensely pleasurable responses to music correlate with activity in brain regions implicated in reward and emotion. *Proceedings National Academy of Science, U S A.* **98**(20): 11818–11823.

Brown, S. (2000). "The 'Musilanguage' model of language evolution," in *The Origins of Music*, eds S. Brown, B. Merker, and N. L. Wallin (Cambridge, MA: MIT Press), 271–300.

Capitao, L., Sampaio, A., Sampaio, C., Vasconcelos, C., Fernandez, M., Garayzabal, E., Shenton, M. E., and Goncalves, O. F. (2011b). MRI amygdala volume in Williams Syndrome. *Resarch in Developmental Disability,* **32**, 2767–2772.

Dankovicova, J., House, J., Crooks, A. & Jones, K. (2007). The Relationship between Musical Skills, Music Training, and Intonation Analysis Skills. *Language and Speech*, **50** (Pt 2):177-225

Dodd, H. F., and Porter, M. A. (2010). I see happy people: attention bias towards happy but not angry facial expressions in Williams syndrome. *Cognitive Neuropsychiatry,* 15, 549–567.

Donnai, D., & Karmiloff-Smith, A. (2000). Williams syndrome: from genotype through to the cognitive phenotype. *American Journal of Medical Genetics*, **97**(2):164-71

Doyle, T., Bellugi, U., Korenberg, J.R., & Graham, J. (2004). 'Everybody in the world is my friend': Hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics,* **124A**, 263– 273

Deruelle C., Mancini J., Livet M. O., Casse-Perrot C., De Schonen S. (1999). Configural and local processing of faces in children with Williams syndrome. *Brain Cogn.* **41**, 276–298;

Dykens, E.M., Rosner, B.A. Ly, T. & Sagun, J. (2005). Music and anxiety in Williams syndrome: a harmonious or discordant relationship? *American Journal Mental Retardation.* 110 (5) 346-58.

Dunn, L. M. & Dunn, L. M. *Peabody Picture Vocabulary Test* (American Guidance Service, 1997).

Eerola, T. & Vuoskoski, J.K. (2010). A comparison of the discrete and dimensional models of emotion in music. *Psychology of* ***Music***, **39**(1), 18-49.

Fodor, J. (1983). *The modularity of mind.* Cambridge, MA: MIT Press.

Fodor, J.A. 1985. 'Précis of the Modularity of Mind', *Behavioral and Brain Sciences* **8:1**, 1-5

Gagliardi, C., Frigerio, E., Burt, D. M., Cazzaniga, I., Perrett, D. I., & Borgatti, R. (2003). Facial expression recognition in Williams syndrome. *Neuropsychologia* 41, 733–738.

Gosselin, N., Peretz, I., Noulhiane, M., Hasboun, D., Beckett, C., Baulac, M. & Samson, S. (2005). Impaired recognition of scary music following unilateral temporal lobe excision. *Brain,* 128 (3), 628-640.

Grice,S.J., Spratling,M.W., Karmiloff-Smith, A, Halit, H., Csibra, G., de Haan G. & Johnson, M.A. (2001). Disordered visual processing and oscillatory brain activity in autism and Williams Syndrome. *Neuroreport*, **12** (12):2697-700

Haas, B. W., Mills, D., Yam, A., Hoeft, F., Bellugi, U., and Reiss, A. (2009). Genetic influences on sociability: heightened amygdala reactivity and event-related responses to positive social stimuli in Williams syndrome. *Journal of Neuroscience*, 29, 1132–1139.

Heaton, P., Allen, R., Williams, K. & Cummins, O. & Happé, F., (2008). Do social and cognitive deficits curtail musical understanding? Evidence from Autism and Down syndrome. *British Journal of Developmental Psychology, **26**,* 171 – 182.

Jarrold, C, Baddeley, A.D., & Hewes, A.K. (1999). Genetically dissociated components of working memory: evidence from Down's and Williams syndrome. *Neuropsychologia.* 37 (6):637-51.

Järvinen-Pasley, A, Bellugi U, Reilly J, Mills, D.L, Galaburda A, Reiss A.L, Korenberg J.R. (2008a). Defining the social phenotype in Williams syndrome: a model for linking gene, the brain, and behavior. *Developmental Psychopathology*, **20** (1) :1-35.

Järvinen-Pasley, A., Vines, B.W., Hill, K.J., Yam, A., Grichanik,M., Mills, D., Reiss, A.L., Korenberg, J.R. & Bellugi, U. (2010).Cross-modal influences of affect across social and non-social domains in individuals with Williams Syndrome. *Neuropsychologia*, 48, (2), 456-466.

Järvinen A, Korenberg J.R, Bellugi U. (2013). The social phenotype of Williams syndrome. *Current Opinion in Neurobiology*, **23** (3):414-22

Järvinen, A., Ng, R., Crivelli, D., Arnold, A. J., Woo-VonHoogenstyn, N., & Bellugi, U. (2015). Relations between social-perceptual ability in multi- and unisensory contexts, autonomic reactivity, and social functioning in individuals with Williams syndrome. *Neuropsychologia*, *73*, 127–140.

Järvinen, A., Ng, R., Crivelli, D., Neumann, D., Arnold, A. J., Woo-VonHoogenstyn, N., Lai, P.. Trauner, D., & Bellugi, U. (2016). **Social functioning and autonomic nervous system sensitivity across vocal and musical emotion in Williams syndrome and autism spectrum disorder.** Devevelopmental Psychobiology, 58 (1),17-26.

Johnson, M. (2011). Interactive Specialization: A domain-general framework for human Functional brain development? *Developmental Cognitive Neuroscience*, **1,** (1), 7-21

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). II. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience, **12, (***Suppl1), 30-46.

Juslin, P. N. & Laukka, P. (2003) Communication of emotions in vocal expression and music performance: Different channels, same code? *Psychological Bulletin* 129:770 – 814.

Juslin, P.N. & Västfjäll, D. (2008). Emotional responses to music: The need to consider underlying mechanisms. Behavioural and Brain Sciences, 31 (5) 559-575.

Karmiloff-Smith, (1994). Beyond modularity: a developmental perspective on cognitive science, Cam Karmiloff-Smith, A. (1998). Cambridge, MA: MIT Press.

(Karmiloff-Smith, A. (1998). Development itself is the key to understanding developmental Disorders. Trends in Cognitive Science, 2, (10) 389-398.

Karmiloff-Smith, A. 2011. 'Brain: the neuroconstructivist approach', in E.K. Farran and A. Karmiloff-Smith (eds.), Neurodevelopmental disorders across the lifespan: a neuroconstructivist approach, Oxford: Oxford University Press, 37-58.

Karmiloff-Smith, A, Grant J, Berthoud I, Davies M, Howlin P, Udwin O. (1997). Language and Williams syndrome: how intact is "intact"? *Child Development.* 68 (2):246-62.

Karmiloff-Smith A., Thomas M., Annaz D., Humphreys K., Ewing S., Brace N., et al. (2004). Exploring the Williams syndrome face-processing debate: the importance of building developmental trajectories. *J. Child Psychol. Psychiatry* **45,** 1258–1274

Knösche, T.R., Neuhaus, C., Haueisen, J., Alter, K., Maess, B., Witte, O.W., Friederici, A.D., 2005. The perception of phrase structure in music. Hum. Brain Mapp. 24, 259–273

Koelsch, S., Gunter, T.C., Cramon, D.Y., Zysset, S., Lohmann, G. & Friederici, A.D. (2002). *Bach speaks: a cortical "language-network" serves the processing of music*. *Neuroimage* **17**, 956–966.

Lacroix A[1], Guidetti M, Rogé B, Reilly J (2009). Recognition of emotional and nonemotional facial expressions: a comparison between Williams syndrome and autism. *Research in Developmental Disability,* **30** (5) 976-85

Laukka, P. & Juslin, P.N. (2007). Similar patterns of age-related differences in emotion recognition from speech and music. *Motivation and Emotion,* 31 (3), 182-191.

Levitin, J. & **Bellugi, U. (1998). Musical Abilities in Individuals with Williams Syndrome.** *Music Perception*, **15** (4), 357-389.

Maess, B., Koelsch, S., Gunter, T. & Friederici, A.D. (2001) *Musical syntax is processed in Broca's area: an MEG study*. *Nature Neuroscience*, **4**, 540–545.

Martens M.A, Wilson S.J, Reutens D.C. (2008). Research Review: Williams syndrome: a critical review of the cognitive, behavioral, and neuroanatomical phenotype. *Journal of Child Psychology and Psychiatry*. **49** (6):576-608.

Martens, M. A., Wilson, S. J., Dudgeon, P., and Reutens, D. C. (2009). Approachability and the amygdala: insights from Williams syndrome. *Neuropsychologia* 47, 2446–2453.

Martínez-Castilla,P. & Sotillo, M. (2014). Pitch processing in children with Williams syndrome: Relationships between music and prosody skills. *Brain sciences* **4** (2), 376-395

Martínez-Castilla P., Burt, M., Borgatti, R. & Gagliardi C. (2015). Facial emotion recognition in Williams syndrome and Down syndrome: A matching and developmental study. *Child Neuropsychology*, **21** (5), 668 – 92.

Meyer-Lindenberg, A., Hariri, A. R., Munoz, K. E., Mervis, C. B., Mattay, V. S., Morris, C. A., and Berman, K. F. (2005). Neural correlates of genetically abnormal social cognition in Williams syndrome. *Nature Neuroscience*, 8, 991–993

Mervis C.B, Morris C.A, Klein-Tasman B.P, Bertrand J, Kwitny S, Appelbaum L.G & Rice C.E. (2003). Attentional characteristics of infants and toddlers with Williams syndrome during triadic interactions. *Developmental Neuropsychology*. **23**, (1-2): 243-68.

Mervis, C.B. & John, A,E. (2010). Cognitive and behavioral characteristics of children with Williams syndrome: Implications for intervention approaches. *American Journal Medical Genetics.* **154C**:229–248.

Morris CA, Demsey SA, Leonard CO, Dilts C, Blackburn BL. (1988). Natural history of Williams syndrome: physical characteristics. Journal of Pediatrics, 113(2):318–326

Nowicki, S., & Duke, M. P. (2001). Nonverbal receptivity: The Diagnostic Analysis of Nonverbal Accuracy (DANVA). In J. A. Hall & F. J. Bernieri (Eds.), *The LEA series in personality and clinical psychology. Interpersonal sensitivity: Theory and measurement* (pp. 183-198). Mahwah, NJ, US: Lawrence Erlbaum Associates Publishers.

Peretz, I. & Coltheart, M. (2003). Modularity of music. *Nature Neuroscience*, **6** (7) 688-91.

Patel, A.D. (2008). Music, Language, and the Brain. New York: Oxford Univ. Press.

Paterson, S.J, Brown J.H, Gsödl M.K, Johnson M.H,& Karmiloff-Smith A. (1999). Cognitive modularity and genetic disorders. *Science;* **286** (5448):2355-8

Pinker, S. (1991). Rules of language. *Science, 253*(5019), 530-535.

Plesa-Skwerer, D., Faja, S., Schofield, C., Verbalis, A., & Tager-Flusberg, H. (2006a). Perceiving facial and vocal expressions of emotion in individuals with Williams syndrome. *American Journal on Mental Retardation,* **111**, 15–26.

Plesa-Skwerer, D., Verbalis, A., Schofield, C., Faja, S., & Tager-Flusberg, H. (2006b). Social-perceptual abilities in adolescents and adults with Williams syndrome. *Cognitive Neuropsychology*, **23**, 338–349.

Plesa-Skwerer, D., Borum, L., Verbalis, A., Schofield, C., Crawford, N., Ciciolla, L., and Tager-Flusberg, H. (2009). Autonomic responses to dynamic displays of facial expressions in adolescents and adults with Williams syndrome. *Social Cognitive Affective Neuroscience,* 4, 93–100.

Porter, M., Coltheart, M., & Langdon, R. (2007). The Neuropsychological Basis of Hyper-sociability in Williams and Down Syndrome. *Neuropsychologia,* **45**, 2839-2849.

Porter MA, Shaw T, & Marsh PJ. (2010). An unusual attraction to the eyes in Williams-Beuren syndrome: a manipulation of facial affect while measuring face scanpaths. Cognitive *Neuropsychiatry.* 2010;15(6):505–530

Quintin, E.M., Bhatara A, Poissant H, Fombonne E, & Levitin DJ. (2011). Emotion perception in music in high-functioning adolescents with autism. *Journal of Autism and Developmental Disorders*, **41** (9), 1240-55.

Reiss, A. L., Eckert, M. A., Rose, F. E., Karchemskiy, A., Kesler, S., Chang, M., Reynolds, M. F., Kwon, H., and Galaburda, A. (2004). An experiment of nature: brain anatomy parallels cognition and behavior in Williams syndrome. *Journal of Neuroscience,* 24, 5009–5015.

Riby., D.M., & Hancock, P.J. (2008). Viewing it differently: Social scene perception in Williams syndrome and Autism. *Neuropsychologia* 46(11):2855-60

Riby., D.M., & Hancock, P.J. (2009). Do faces capture the attention of individuals with Williams syndrome or autism? Evidence from tracking eye movements. Journal of Autism and Developmental Disorders, 39 (3):421-31

Sauter, D,A., Panattoni, C & Happe, F. (2013). Children's recognition of emotions from vocal cues. *British Journal of Developmental Psychology,* **31** (1), 97-113.

Schlaug, G., Norton, A., Overy, K., & Winner, E. (2005). Effects of Music Training on the Child's Brain and Cognitive Development. *Annals of the New York Academy of Sciences*, 1060, 219-30.

Schon, D., Magne, C. & Besson, M. (2*004). The music of speech: Music training facilitates pitch processing in both music and language. Psychophysiology*, 41, 341–349

Strømme, P., Bjørnstad, P. G. & Ramstad, K. *Prevalence estimation of Williams syndrome*. *J. Child Neurol.* **17**, 269–271 (2002).

Tassabehji M. Williams-Beuren syndrome: a challenge for genotype-phenotype correlations (2003). Human *Molecular Genetics*, **12**(2):229–237.

Thakur, D., Martens, M.A., Smith, D.S. & Roth, E. (2018). Williams Syndrome and Music: A Systematic Integrative Review. Front. Psychol., 14 November 2018. https://doi.org/10.3389/fpsyg.2018.02203

Tillmann, B., Janata, P. & Bharucha, J.J. (2003) *Activation of the inferior frontal cortex in musical priming*. *Cognitive Brain Research*. **16**, 145–161.

Thomas, M.S. & Karmiloff-Smith, A. (2002). Are developmental disorders like cases of adult brain damage? Implications from connectionist modelling. *Behavioral and Brain Sciences* 25 (6):727-750

Thomas, M.S, Annaz, D; Ansari, D; Scerif, G; Jarrold, C; Karmiloff-Smith, A. (2009). Using developmental trajectories to understand developmental disorders. *Journal of Speech, Language, and Hearing Research*, 52, 336-358.

Thomas, M. S, Purser, H. R. M. & Richardson, F. M. (2013). Modularity and developmental disorders. In: P. D. Zelazo (Ed.), *Oxford Handbook of Developmental Psychology*. Oxford: Oxford University Press.

Quintin, E.M., Bhatara, A., Poissant, H., Fombonne, E. & Levitin, D.J. (2011). Emotion Perception in Music in High-Functioning Adolescents with Autism Spectrum Disorders. Journal of Autism and Developmental Disorders, 1 (9), 1240-1255.

Westermann, G., Mareschal, D., Johnson, M. H., Sirois, S., Spratling, M. W., & Thomas, M. S. C. (2007). Neuroconstructivism. Developmental Science, 10(1), 75-83

Zarchi, O, Attias J, Gothelf D.(2010). Auditory and visual processing in Williams syndrome. Israel Journal of Psychiatry Related Sciences ;47(2):125-31.