Facial expression recognition in Williams syndrome

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Abstract

Individuals with Williams syndrome (WS) excel in face recognition and show both a remarkable concern for social stimuli and a linguistic capacity for, in particular, emotionally referenced language. The animated full facial expression comprehension test (AFFECT), a new test of emotional expression perception, was used to compare participants with WS with both chronological and mental age-matched controls. It was found that expression recognition in WS was worse than that of chronologically age-matched controls but indistinguishable from that of mental age controls. Different processing strategies are thought to underlie the similar performance of individuals with WS and mental age controls. The expression recognition performance of individuals with WS did not correlate with age, but was instead found to correlate with IQ. This is compared to earlier findings, replicated here, that face recognition performance on the Benton test correlates with age and not IQ. The results of the Benton test have been explained in terms of individuals with WS being good at face recognition; since a piecemeal strategy can be used, this strategy is improved with practice which would explain the correlation with age. We propose that poor expression recognition of the individuals with WS is due to a lack of configural ability since changes in the configuration of the face are an important part of expressions. Furthermore, these reduced configural abilities may be due to abnormal neuronal development and are thus fixed from an early age.
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1. Introduction

Williams syndrome (WS) is a genetic disorder resulting from the deletion of a section of chromosome 7 whose function normally includes the production of the protein elastin [14]. Individuals with WS are characteristically highly social [18], enjoy using varied and emotionally referenced language and linguistic tools [22] and appear good at facial recognition [3]. Because of these characteristics, people interacting with an individual with Williams syndrome for the first time may be surprised to learn that they suffer from a developmental disorder that is also characterised by low intelligence and poor spatial abilities. In the study presented here, we investigate facial expression recognition ability. This ability lies at the intersection of two fields in which individuals with WS characteristically excel: linguistic description of affect and facial recognition.

Individuals with WS are classically thought to exhibit a hypersociability, commonly referred to as "cocktail party" style, that is manifested as a friendly, but often exaggeratedly friendly, drive for social interaction with other people [18]. Jones et al. [18] have shown that the positive social disposition of individuals with WS is present from infancy. For example, they found that, during experiments, infants with WS showed a remarkable interest in engaging the experimenter using eye contact, smiling and cooing, and when unable to complete a task rather than becoming upset as a normal child would, instead played with the table or waved to their parents. The overtly friendly outlook of those with WS tends to extend to unfamiliar people, leading parents of individuals with WS to worry about their children's abnormal tendency to seek out and engage strangers. This tendency to approach strangers is mirrored in the ratings given by individuals with WS of the approachability of photographs of unfamiliar individuals in comparison to both chronological (CA) and mental (MA) age-matched controls. The positive social bias of individuals with WS may also affect their perception of facial expressions of emotions leading to more

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positive interpretations of facial expressions than would be made by other participants. Comparisons have been made between individuals with focal bilateral damage to the amygdala and individuals with WS who have been noted to have a smaller amygdala [2]. Thus participants with WS may mirror the particularly poor abilities of amygdala-damaged individuals for evaluation of fear [1] and disgust [8], while being proficient in recognising identity from faces.

Studies suggest that individuals with WS are at, or close to, normal levels of performance for their age on standardized tests of facial recognition, such as the Benton test [6]. The Benton test is a face-matching task used to assess ability to recognize unfamiliar faces. Participants are presented with a photograph of a target individual and have to select the second photograph of the same individual from six pictures that depict faces viewed from a variety of angles and lighting conditions. Participants with WS typically perform significantly better than would be expected, given their mental age (MA), but tend to be poorer than adults or chronologically age-matched controls [3,4,27].

Individuals with WS, like individuals with other forms of cognitive impairment, may not show their true abilities at an experimental task because of their distractibility, short attention span and poor understanding of complex tasks. To maximise participant performance, the animated full facial expression comprehension test (AFFECT), a new test of facial expression recognition performance, was devised with participant capacities in mind: minimal information apart from a single test image was presented on screen. To avoid participant distraction the test was blocked to allow testing over a number of sessions should the subject become tired. Instructions were simple and responses of participants were recorded during the test by the experimenter so motor skills were not required. Animated facial expressions were used in the expression test to help engage participants' attention.

In the study presented here, the abilities of participants with WS were compared with those of CA and MA matched control groups on emotional expression labelling in dynamically presented facial expressions. The use of emotional language and face recognition have both been reported to be strengths of individuals with WS and so it might be hypothesised that their performance would be better than expected for their mental age. There is ERP evidence though that processing of faces is abnormal in WS [23]. Furthermore, participants with WS have been noted to make spontaneous comments suggesting that they use a feature-matching strategy [27] and their face recognition abilities may not be as good in face recognition tasks that do not lend themselves to

a piecemeal processing strategy [12]. As facial expressions change the configuration of the face, face expression recognition may lend itself to configurational processing and the performance of individuals with WS may suffer as a result. Additionally, individuals with WS appear to have a positive outlook that might result in a bias to perceive facial expressions as being positive, while neurological abnormalities (e.g. as noted earlier to the amygdala) might result in individuals with WS being more sensitive in the recognition of some facial expressions than others.

To allow comparison with previous work, subjects performed the Benton task. It was expected that, as in previous studies [3,4], individuals with WS would perform on the Benton task at a level commensurate with their chronological rather than their mental age. Previous studies have found that performance on the Benton task improves with age [2], possibly because of improved ability in the use of a piecemeal strategy. We expected to replicate this correlation.

2. Methods

2.1. Participants

A total of 26 participants with Williams syndrome (14 females, 12 males), each of whom manifested the typical deletion as shown by fluorescent in situ hybridization (FISH), participated in this study. The participants with WS were matched for sex and chronological age with 26 chronological age controls (CA) and for sex and mental age with 26 mental age controls (MA). All control participants were healthy, with no past history of psychiatric or neurological illness, and scored at least 85 in the Stanford–Binet test [7]. The characteristics of the samples are summarized in Table 1.

Informed consent was given by the parents of participants or where appropriate, the participant themselves, for participation in the study.

2.2. Procedure

One day prior to testing participants completed the Stanford–Binet intelligence scale [7] to establish cognitive ability. Participants with WS then performed the Benton test of facial recognition [6] to assess face recognition.

Immediately prior to testing, to accustom participants to the task of labelling emotional expressions and to assess emotional expression familiarity, all participants took part in a task presented as a 'game'. During this game, participants

Table 1
Participants with WS matched with controls for mental age (MA) and chronological age (CA)

Group	N	MA (years): mean (S.D.)	Range (years)	CA (years): mean (S.D.)	Range (years)	IQ: mean (S.D.)	Range
WS	26	5.94 (1.48)	4.3–11.5	14.35 (6.64)	5.6–32.3	50 (14.0)	33–81
MA	26	6.07 (1.46)	4.1–11.3	5.5 (1.47)	4.3–11.3	109 (10.4)	95–129
CA	26	15.1 (6.36)	6.1–29.6	14.49 (6.71)	5.8–29.7	112 (9.6)	89–133