Facial expression recognition in Williams syndrome and Down syndrome: A matching and developmental study

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Abstract

In this study both the matching and developmental trajectories approaches were used to clarify the development of facial expression recognition in Williams syndrome (WS) and Down syndrome (DS). The matching approach showed that neither individuals with WS nor DS exhibit a specific proficiency for the expression of happiness or a specific impairments for negative emotions but presented the same pattern of emotion recognition as typically developing (TD) individuals. Thus, the better performance on the recognition of positive compared to negative emotions often reported in WS and DS is not specific to these populations but represents a typical pattern. Prior studies based on the matching approach suggested that the development of facial expression recognition is delayed in WS and atypical in DS. Nevertheless, and even though performance levels were lower in DS than in WS, the developmental trajectories approach used in this study found that not only individuals with DS but also those with WS present atypical developmental trajectories. Unlike in the TD participants where developmental changes were observed along with age, in the WS and DS development was static. Thus, both individuals with WS and those with DS reach early maximum levels of facial expression recognition due to cognitive constraints.

Key words: Williams syndrome; Down syndrome; facial expression recognition; matching; developmental trajectories

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Typical development of emotional facial recognition starts early in infancy and continues through childhood, adolescence and adulthood (Herba & Philipps, 2004) playing an important role in social cognition, facilitating social communication and interaction (Haxby, Hoffman & Gobbini, 2000). Consequently, problems recognizing facial expressions may have detrimental effects upon social behavior (Riby & Back, 2010). Such problems have been noted in individuals with Williams syndrome (WS) and Down syndrome (DS) (e.g., Porter, Coltheart, & Langdon, 2007; Santos, Rosset, & Deruelle, 2009; Wishart, 2007). Here we assessed facial expression recognition skills in WS and DS compared the developmental pattern of these skills in both syndromes with typical development.

Recognition of emotional facial expressions in Williams syndrome

WS is a neurodevelopmental disorder caused by a hemizygous deletion in 7q11.23 (Ewart et al., 1993). It has a prevalence of 1: 7500 to 20000 live births (Morris, Demsey, Leonard, Dilts, & Blackburn, 1988; Strømme, Bjørnstad, & Ramstad, 2002) and is characterized by mild to severe intellectual disability and a defined cognitive profile (Mervis et al., 2000). Individuals with WS are often described as very sociable, empathic and friendly (Dykens & Rosner, 1999; Gosh & Pankau, 1994, but also present high levels of anxiety and fears (Dykens, 2003; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006), difficulties when interacting with others (Frigerio et al., 2006), poor social relationships (Davies, Udwin, & Howlin, 1998) and indiscriminate approach to strangers (Jones et al., 2000).

Problems in social understanding and social behavior in individuals with WS have been related to their difficulties in facial expression recognition (Porter et al., 2007;

Santos et al., 2009). Despite a relative strength for recognizing facial identity (e.g., Bellugi, Wang, & Jernigan, 1994), individuals with WS present impairments across different facial expression recognition tasks (Riby, Doherty-Sneddon, & Bruce, 2008). In labeling, identification and matching tasks, both with static and dynamic stimuli, it is generally reported that individuals with WS perform more poorly than typically developing (TD) peers matched on chronological age (CA) but at the same level as TD children and individuals with developmental disabilities of similar mental age (MA) (Gagliardi et al., 2003; Lacroix, Guidetti, Rogé, & Reilly, 2009; Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006; Plesa-Skwerer, Verbalis, Schofield, Faja, & Tager-Flusberg, 2006; Porter et al., 2007; Porter, Shaw, & Marsh, 2010).

Nevertheless, for the emotion of happiness, several studies (Plesa-Skwerer, Faja et al, 2006; Plesa-Skwerer, Verbalis et al, 2006; Santos et al., 2009) have found a lack of significant differences between individuals with WS and TD controls matched on CA. This lack of significant effect has been interpreted as showing that individuals with WS perform better on positive than negative emotions (Plesa-Skwerer, Faja et al., 2006), but may arise from response biases label expression as happy or from ceiling effects as happy facial expression tend to be better recognized (Plesa-Skwerer, Faja et al., 2006; Santos et al., 2009).

Recognition of emotional facial expressions in Down syndrome

DS is caused typically by the presence of an extra chromosome 21 and is the most common cause of intellectual disability of genetic origin, with a prevalence of 1:700 to 1000 live births (Roizen & Patterson, 2003). Despite individuals with DS usually being referred to as sociable, friendly and affectionate (Fidler, Most, & Philofsky, 2009), they also present problems in social functioning, showing little accommodation to social partners, stubbornness, social withdrawn (Coe et al., 1999;

Wishart & Pitcairn, 2000) and a tendency to approach strangers inappropriately (Porter et al., 2007).

Impairments of facial expression recognition in individuals with DS have also been linked to their social behavior (Hippolyte, Barisnikov, Van der Linden, & Detraux, 2009; Wishart, 2007; Wishart, Cebula, Willis, & Pitcairn, 2007). As in WS but to a lesser extent, skills for identity recognition are better than those for recognizing facial expressions in DS (Williams, Wishart, Pitcarin, & Willis, 2005; Wishart & Pitcarin, 2000). Across labeling, matching and identification tasks with static stimuli, individuals with DS consistently perform significantly lower than TD peers of the same MA (Kasari, Freeman, & Hughes, 2001; Hippolyte et al., 2009; Hippolyte, Barisnikov, Van der Linden, 2008; Porter et al., 2007; Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000). However, these results seem to be observed only from a mental age of 4 years and above (Kasari et al., 2001). At 3 years of MA, children with DS perform as well as TD MA-matched peers (Kasari et al., 2001). Performance in DS is better with dynamic than static stimuli, but even with dynamic stimuli adults with DS present impairments in facial expression recognition (Virji-Babul, Watt, Nathoo, & Johnson, 2012).

Individuals with DS seem to have greater difficulty recognizing negative emotions compared to a relative strength for happiness (Hippolyte et al., 2009). Studies have also variously reported specific impairments in DS of fear (Kasari et al., 2001; Williams et al., 2005; Wishart & Pitcairn, 2000), surprise (Hippolyte et al., 2008; Hippolyte et al., 2009; Wishart & Pitcairn, 2000), anger (Kasari et al., 2001) and sadness (Porter et al., 2007). This variability may relate to the relatively small number of trials per emotion that are typical in these studies making solid conclusions difficult (Williams et al., 2005). Therefore it is still an open question whether individuals with

DS present specific impairments for any emotion.

Facial expression recognition skills of individuals with DS have also been compared to those of MA-matched peers with other developmental disabilities. No significant differences have been found between individuals with DS and those with nonspecific intellectual disabilities or individuals with Fragile-X syndrome (Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000). Yet, despite the lack of significant differences between groups, in the same studies, only participants with DS performed significantly lower than TD children matched on MA (Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000). This has been interpreted as suggesting that specific difficulties for facial expression recognition in DS are subtle and may only be detectable after comparisons with TD controls (Wishart et al., 2007). Nevertheless, researchers have emphasized the need of conducting more cross-syndrome comparisons between individuals with DS and those with developmental disabilities of known genetic origin in order to study the possible specificity of their emotion recognition impairments (Carvajal, Fernández-Alcaraz, Rueda, & Sarrión, 2012; Williams et al., 2005).

Comparisons between facial expression recognition skills of individuals with Williams syndrome and Down syndrome

To our knowledge, only one study by Porter et al. (2007) has compared the skills for recognizing emotional facial expressions between both populations. This study used a labeling task with static stimuli to assess the facial expression recognition skills of individuals with WS and DS compared to TD individuals. Both individuals with WS and DS were significantly poorer than similarly aged TD peers, but while individuals with WS performed at the same level as typical MA peers individuals with DS performed significantly worse on the negative emotions (sad, fear and anger) than either

individuals with WS or TD peers match on MA. Although no significant differences were found between the two disorder groups for happiness, Porter et al. (2007) suggested that the better performance of participants with DS on the positive compared to the negative emotions could be due to response biases for happiness. Therefore, although, as previously mentioned, individuals with DS perform at the same level as individuals with intellectual disability of different etiologies on facial expression recognition tasks (Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000), when compared to participants with WS, cross-syndrome differences seem to emerge.

Studying the development of facial expression recognition in Williams syndrome and Down syndrome

The research on facial expression recognition in WS and DS discussed above has mainly used a matching approach in which the facial expression recognition skills of participants with WS or DS have typically been compared to those of two groups of TD children matched either on MA or CA. Individuals with WS are typically found to exhibit developmental delay: performing more poorly than similarly aged TD peers but at the same level as TD children matched on MA (Gagliardi et al., 2003; Porter et al., 2007; Riby, 2012). In comparison, children with DS perform more poorly than either TD participants matched on CA or MA and thus seem to present a developmental deviance or atypicality (Kasari et al., 2001; Williams et al., 2005; Wishart & Pitcairn, 2000). Results from the matching approach therefore suggest a dissociation between WS and DS.

However, in the matching approach, age is not specifically assessed and, therefore, despite the fact that development should be the key for understanding developmental disorders (Karmiloff-Smith, 1998), the role of change over time is not taken into account (Thomas et al., 2009). Consequently, the differential role of

development itself cannot be fully evaluated in the aforementioned cross-syndrome comparisons, meaning that little is known about the development of facial expression recognition in WS and DS. Both individuals with WS and DS show atypically increased attention to human faces (Gunn, Berry, & Andrews, 1982; Jones et al., 2000; Kasari, Sigman, Mundy, & Yirmiya, 1990; Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2008). However, this attention to faces does not lead them typical levels of proficiency in deciphering facial expressions (Campos, Martínez-Castilla, & Sotillo, 2013; Kasari et al., 2001). Studies have also shown that, while emotion recognition skills correlate with both CA and MA in typical children, no significant correlations are observed between the same variables in individuals with WS or DS (Gagliardi et al., 2003; Santos et al., 2009; Williams et al., 2005; Wishart & Pitcairn, 2000). These results of cross-sectional studies suggest that the developmental pathways for facial expression recognition in both syndromes are different from those observed in typical children (Riby, 2012; Santos et al., 2009; Williams et al., 2005).

Rather than cross-sectional studies longitudinal studies, are a better method to understand development. In terms of facial expression recognition the only relevant study followed children with DS for two years but found no improvement in facial expression recognition (Kasari et al., 2001). Although longitudinal designs are important for understanding development, they are costly and difficult to carry out (Thomas et al., 2009) often resulting in their being under-powered particularly for the interpretation of null results. For this reason, the cross-sectional developmental trajectories approach (which for simplicity will be referred to as the developmental trajectories approach) has been proposed as an alternative method to study development (see Thomas et al., 2009 for a full and detailed description of this approach). Like the matching approach, this approach uses data collected at single time points, but linear

regression is used to model the relationship between age (CA or MA) and the measure of ability (in our case facial expression recognition) separately for the typical and disorder groups, and the resulting trajectories of these models are then compared (Thomas et al., 2009). Thus, this approach allows for developmental change to be compared across typically and atypically developing groups using cross-sectional data (Thomas, Purser, & Van Herwegen, 2012). Developmental trajectories with CA as the predictor provide a theory-neutral comparison to assess whether the disorder group deviate from the typical developmental trajectory on the experimental task (Annaz, Karmiloff-Smith, Johnson, & Thomas, 2009; Thomas et al., 2009). In turn, developmental trajectories linking performance to MA constitute a means to evaluate whether the deficits in the disorder group are in line with the developmental state of cognitive system more generally (Annaz et al., 2009). Thus, in comparison to a matching approach this methodological approach provides a richer description of developmental delay in terms such as delayed onset, slowed rate of development, or both delayed onset and slowed rate (Thomas et al., 2009). It also allows atypical development to be described in terms which distinguish between no systematic relationship (a random distribution between performance scores and age) and a zero trajectory (a trajectory whose gradient is zero, showing a system that has reached an early limit due to cognitive constraints on development) (Thomas et al., 2009). This latter distinction is particularly important. As mentioned above, previous studies of both DS and WS have shown a lack of relationship between age and performance on facial expression recognition tasks. However, they have not been able to distinguish between a lack of systematic relationship and a zero trajectory despite their very different implications. These descriptors may allow for revealing ways in which similarities can occur between the disorder groups studied (Thomas et al., 2009).

The developmental trajectories approach has been successfully used to other aspects of the cognitive profile in WS and DS (e.g., Thomas et al., 2006; Thomas et al., 2010) including facial identity recognition in both populations (Annaz et al., 2009; Karmiloff-Smith et al., 2004). For example, although the scores of individuals with WS usually fall within the normal range on face recognition tasks such as the Benton test (Benton, Hamsher, Varney, & Spreen, 1983), the developmental trajectories approach has shown that the development of face recognition skills is delayed compared to that of TD individuals (Karmiloff-Smith et al., 2004). Therefore, this approach can offer great insight on the development of the area under study. To our knowledge, no previous research has employed this approach to compare facial expression recognition in WS and DS in order to study possible differences or similarities in developmental delay or atypicality.

Scope and aims of the study

In this study, children with WS, DS and TD peers were administered a labeling task with dynamic stimuli: facial expressions of happiness, sadness, fear, anger and disgust. Both the matching and developmental trajectories approaches were used.

The matching approach was used to ascertain whether, in comparison with TD peers, individuals with WS or DS present better performance for positive than negative emotions; whether individuals with WS and DS present specific deficits for any emotions; and whether there are cross-syndrome differences in expression recognition impairments. As previously mentioned, small numbers of trials per emotion, response biases for the label happy, and ceiling effects on the tasks used in prior studies leave these issues unresolved (Plesa-Skwerer, Faja et al., 2006; Porter et al., 2007; Santos et al., 2009; Williams et al., 2005). A number of steps were taken to overcome these issues: a relatively large number of trials for each emotion were used; an unbiased

response index (Wagner, 1993) to compensate for response bias; and to avoid both floor and ceiling effects a task designed to be sensitive across the ability range of the participants. We hypothesized that, by overcoming the aforementioned issues, individuals with WS and DS would not show specific proficiency or impairment for any of the emotions.

The developmental trajectories approach was used to test whether skills for recognizing different expressions develop in WS and DS as they do in typical development, and thus to assess different possible types of developmental delay or atypicality. Cross-syndrome comparisons provide information on the specificity of the developmental patterns in WS and DS. Unlike conclusions which can be reached based on the matching approach we were able to test the hypothesis that the developmental trajectories in this domain would be similar and atypical in both WS and DS. In addition, the use of developmental trajectories allowed us to distinguish a zero developmental trajectory from a lack of systematic relationship between facial expression recognition and participant age.

Method

Participants

Our sample was composed of 20 participants with WS[1], 20 participants with DS and 40 TD controls. Participants with WS presented both the clinical phenotype of the syndrome (Preus, 1984) and the classic-length deletions of the WS critical region confirmed by FISH (Fluorescence In-Situ Hybridisation) analysis. Participants with DS had confirmed trisomy 21 without mosaicism. TD control participants were healthy

[1] Participants with WS were a subset of those reported in Gagliardi et al. (2003) selected to have the same CA and MA as participants with DS. However, it should be noted that the scope and aims of the current research are different from those of Gagliardi et al. (2003)'s study.

children with no prior history of learning disabilities, psychiatric or neurological illness. Individuals with WS and those with DS were recruited during a program for genetic syndromes at the Scientific Institute 'Eugenio Medea' (Bosisio Parini, Lecco, Italy). TD children were recruited from mainstream schools and showed average school performance in language, arts and reading. Written informed consent, approved by the Ethics Committee of the Institute, was obtained from participants' parents or from participants themselves when appropriate.

For the matching approach, groups were matched as follows. Participants with WS were matched on CA and MA to participants with DS (p = .98, p = .37, respectively). A group of 20 TD participants were also matched on CA to both the WS and DS groups (p = .95, p = .99, respectively) and a different group of another 20 TD controls was matched on MA to participants with WS and those with DS (p = .75, p = .80, respectively). The Italian version of the Stanford–Binet test (Bozzo & Mansueto, 1993) was used to measure all participants' MA. Descriptive characteristics of the groups (CA and MA means and ranges, and gender distribution) are shown in Table 1. For the developmental trajectories approach TD participants were collapsed in a single group, as done in previous research (Thomas et al., 2001; Thomas et al., 2006).

INSERT TABLE 1 ABOUT HERE

Materials and Procedure

The animated full facial expression comprehension test (AFFECT) (Gagliardi et al., 2003) was administered to all participants. In this computerized test, participants are required to label emotional expressions of happiness, sadness, fear, anger and disgust which are presented by means of animated faces. The use of these animated stimuli ensures that the test is ecological and engaging for participants (e.g., Gagliardi et al., 2003; Recio, Schacht, & Sommer, 2013) while also being considered a realistic method

for assessing the development of facial expression recognition (Herba & Philipps, 2004). The faces are presented animated from neutral to four different intensities (25, 50, 75 and 100% of the full-blown facial expression). The inclusion of different levels of intensity helps to avoid ceiling and floor effects and thus extends the sensitivity of the test (Gagliardi et al., 2003). The test comprised of a total of 80 trials (5 emotions x 4 identities x 4 intensities) and, therefore, includes 16 trials for each emotion. Participants are asked to label each of the emotional facial expressions presented. In the current study, the procedure for administering the test was the same as that reported in Gagliardi et al. (2003). Thus, participants were asked to label each of the emotional facial expressions presented.

In the current study, the procedure for administering the test was the same as that reported in Gagliardi et al. (2003). Thus, participants were asked to label each of the emotional facial expressions presented (happiness, sadness, fear, anger and disgust). Participants' responses were coded by the experimenter according to the five possible emotional labels. Prior to testing, to ensure that participants were familiar with the emotional facial expressions included in the study and to get them used to the labeling test, they were engaged in a game task. In this task, participants were asked to label the emotional facial expressions of the experimenter and to produce their own facial expressions of the five emotions. No participant had difficulty in this initial task. Immediately afterwards, the AFFECT test was administered. The test was divided into four blocks of 20 trials each. Each block included the five emotions animated to the four intensities presented by a different individual. The testing session began with five practice trials which were not used elsewhere during the experiment. Then, the first two blocks were administered, followed by a break of 1 hour to avoid tiredness and attentional problems. Finally, participants were presented with the remaining two

blocks.

Accuracy was measured by an index that controls for response biases, the unbiased hit rate proposed by Wagner (1993, 1997). The unbiased hit rate takes response biases into account by estimating the 'joint probability both that a stimulus is correctly identified (given that it is presented) and that a response is correctly used (given that it is used)' (Wagner, 1993, p. 16). It is calculated by multiplying the hit rate for a particular emotion (the number of times the emotion is correctly identified divided by the number of trials where the emotion has been presented) by the differential accuracy (the number of times the emotion is correctly identified divided by the total number of trials where that emotion has been given as a response) (Montirosso, Peverelli, Frigerio, Crespo, & Borgatti, 2010; Wagner, 1993, 1997). This accuracy measurement is easy to interpret because the resulting values are proportions ranging from 0 to 1.

As recommended in the literature on the topic (e.g., Wagner, 1997), chance performance was also calculated. Chance performance refers to the 'joint probability of the co-occurrence by chance of the stimulus and response of a particular class' (Wagner, 1997, p. 51) and is calculated by multiplying the number of times an emotion is presented divided by the total number of trials, by the number of times an emotion is used divided by the total number of trials (Wagner, 1997).

Statistical analyses were performed with SPSS 15.0. To correct for the increased likelihood of obtaining results by chance caused by performing multiple comparisons values of $p \leq .01$ were declared significant.

Results

As previously explained, to fulfill the aims of the study the analysis based on matching is presented followed by those for the developmental trajectories approach.

Results from the matching approach

Accuracy measurements and chance performance per emotion in each of the four matched groups included in the study are presented in Table 2. No floor or ceiling effects were observed in the WS, DS or MA-matched TD groups. In the CA-matched TD group, performance was near ceiling only for the emotion of happiness. In each group (WS, DS, MA-matched TD, CA-matched TD), unbiased hit rates were first compared to chance performance by means of dependent *t*-tests in order to assess whether participants were answering at chance. For each emotion, accuracy was significantly higher than chance performance in all groups ($p \leq .001$ for all cases).

INSERT TABLE 2 ABOUT HERE

In order to compare accuracy between groups, a mixed factorial ANOVA was run with group as a between-subjects factor, emotion as a within-subjects factor and the unbiased hit rates as dependent variable. The main effects of group, $F(3, 76) = 25.88$, $p < .001$, $\eta_p^2 = .51$, and emotion, $F(3.54, 269.33) = 67.61$, $p < .001$, $\eta_p^2 = .47$, were significant. However, no significant effect was found for the interaction between the two factors. Pair-wise comparisons with Bonferroni correction showed that the DS group performed significantly lower than the WS, MA-matched TD and CA-matched TD groups ($p \leq .001$ for all comparisons). No significant differences were found between participants with WS and their MA-matched TD peers but participants with WS performed significantly lower than the CA-matched TD group ($p < .001$). In turn, for all the groups, accuracy was significantly higher for happiness than for sadness, fear, anger or disgust ($p < .001$), and no significant differences were found when comparing performance on any of the negative emotions.

Results from the developmental trajectories approach

Cross-sectional developmental trajectories were built for the TD, WS and DS

groups. Performance on the labeling task for each emotion was modeled by running separate linear regression analyses for each group where either CA or MA were entered as predictors for accuracy. The developmental trajectories of the TD, WS and DS groups are shown in Figures 1, 2, and 3, respectively. For the sake of clarity, individual data points are not shown.

Developmental trajectories for the typically developing group:

Accuracy on the task improved significantly both with CA and with MA for every emotion (CA: happiness, $F(1, 38) = 32.29$, $p < .001$, $R^2 = .46$, sadness, $F(1, 38) = 11.84$, $p < .01$, $R^2 = .24$, fear, $F(1, 38) = 22.32$, $p < .001$, $R^2 = .37$, anger, $F(1, 38) = 13.27$, $p < .01$, $R^2 = .26$, and disgust, $F(1, 38) = 10.05$, $p < .01$, $R^2 = .21$; MA: happiness, $F(1, 38) = 34.32$, $p < .001$, $R^2 = .47$, sadness, $F(1, 38) = 8.94$, $p < .01$, $R^2 = .19$, fear, $F(1, 38) = 22.41$, $p < .001$, $R^2 = .37$, anger, $F(1, 38) = 12.89$, $p < .01$, $R^2 = .26$, and disgust, $F(1, 38) = 7.3$, $p = .01$, $R^2 = .16$).

INSERT FIGURE 1 ABOUT HERE

Developmental trajectories of emotions were compared by means of repeated-measures ANCOVAs with emotion as the within-subjects factor. To assess for differences between trajectories at their onset, CA and MA were re-scaled to account for the youngest age included in the group (Thomas et al., 2009). These re-scaled measurements were entered as covariates in separate analyses. The analyses showed that accuracy was higher for happiness compared to any of the negative emotions (p <.001 for all comparisons). No other significant differences were found regarding the main effect of emotion. ANCOVAs run with CA as a covariate showed a significant effect of CA across comparisons between emotions (p <.001). The interaction between emotion and CA was non-significant for all comparisons. The same pattern of results was found when MA was included as a covariate in the analyses. Thus, the effect of MA was

significant for all the comparisons between emotions ($p \leq .001$) but no significant effect was found for the interaction between emotion and MA.

Developmental trajectories for the Williams syndrome group:

In contrast to what was observed in the TD group, for participants with WS, CA did not significantly predict accuracy on the task for any of the emotions (happiness, $F(1, 38) = 0.73$, $p = .41$, $R^2 = .04$, sadness, $F(1, 38) = 0.79$, $p = .39$, $R^2 = .04$, fear, $F(1, 38) = 0.66$, $p = .43$, $R^2 = .04$, anger, $F(1, 38) = 0.07$, $p = .79$, $R^2 = .00$, and disgust, $F(1, 38) = 0.001$, $p = .97$, $R^2 = .00$). Neither was MA reliably related to accuracy for any of the emotions included in the study (happiness, $F(1, 38) = 1.18$, $p = .29$, $R^2 = .06$, sadness, $F(1, 38) = 0.79$, $p = .39$, $R^2 = .04$, fear, $F(1, 38) = 2.38$, $p = .14$, $R^2 = .12$, anger, $F(1, 38) = 2.23$, $p = .15$, $R^2 = .11$, and disgust, $F(1, 38) = 1.11$, $p = .31$, $R^2 = .06$). Using curve estimation it was checked whether a non-linear trajectory (i.e., logarithmic, inverse, exponential, growth) fitted the data better. The non-linear functions were not significant either. Since neither linear nor non-linear functions gave a significant fit to the data in the WS group, comparisons of developmental trajectories for the different emotions could not be carried out.

INSERT FIGURE 2 ABOUT HERE

As previously mentioned, a lack of significant relationships between accuracy and CA or MA can be due to two different possibilities. First, it may be that performance is random with respect to CA or MA, which has been referred to as no systematic relationship (Thomas et al., 2009). Second, a zero trajectory may fit the data, i.e., it may be that accuracy scores are distributed horizontally so that performance is not changing with age (Thomas et al., 2009). To distinguish between these two possibilities, Thomas et al. (2009) proposed the use of a method by which data are transformed by a 45º anti-clockwise rotation. After the rotation, the R^2 value of

randomly distributed data will non-significant and close to zero if performance is random with respect to age. Aternatively, if rotation makes the R^2 value and become highly significant then it suggests a zero trajectory increase (see Thomas et al., 2009 for full details on this method). The rotation method was applied to the accuracy data for each emotion in the WS group. For trajectories built against CA, the rotation method produced near zero R^2 values in all the emotions included in the study (happiness, $F(1, 38) = 0.088$, $p = .77$, $R^2 = .005$, sadness, $F(1, 38) = 0.002$, $p = .97$, $R^2 = .000$, fear, $F(1, 38) = 0.015$, $p = .90$, $R^2 = .001$, anger, $F(1, 38) = 0.003$, $p = .95$, $R^2 = .000$, and disgust, $F(1, 38) = 0.20$, $p = .66$, $R^2 = .011$). However, when MA was taken as predictor, all the R^2 values increased and were significant after rotation (happiness, $F(1, 38) = 160.23$, $p < .001$, $R^2 = .90$, sadness, $F(1, 38) = 132.89$, $p < .001$, $R^2 = .88$, fear, $F(1, 38) = 138.71$, $p < .001$, $R^2 = .89$, anger, $F(1, 38) = 141.66$, $p < .001$, $R^2 = .89$, and disgust, $F(1, 38) = 110.09$, $p < .001$, $R^2 = .86$). Therefore, CA showed no systematic relationship with accuracy whilst the developmental trajectories built against MA showed a gradient of zero (i.e., performance did not improve along with MA but remained static).

Developmental trajectories for the Down syndrome group:

Similarly to what was found for participants with WS, in the DS group, CA did not significantly predict accuracy for any of the emotions, although there was a tendency for anger accuracy to decrease with CA (happiness, $F(1, 38) = 0.14$, $p = .71$, $R^2 = .01$, sadness, $F(1, 38) = 0.85$, $p = .37$, $R^2 = .05$, fear, $F(1, 38) = 0.002$, $p = .97$, $R^2 = .00$, anger, $F(1, 38) = 4.23$, $p = .054$, $R^2 = .19$, and disgust, $F(1, 38) = 0.10$, $p = .75$, $R^2 = .01$). MA was not a reliable predictor either (happiness, $F(1, 38) = 0.96$, $p = .34$, $R^2 = .05$, sadness, $F(1, 38) = 2.50$, $p = .13$, $R^2 = .12$, fear, $F(1, 38) = 1.58$, $p = .23$, $R^2 = .08$, anger, $F(1, 38) = 0.41$, $p = .53$, $R^2 = .02$, and disgust, $F(1, 38) = 0.05$, $p = .33$, $R^2 = .05$). Non-linear functions did not provide any better fit to the data. Consequently,

comparisons of developmental trajectories for the different emotions could not be carried out in the DS group either.

INSERT FIGURE 3 ABOUT HERE

In order to distinguish between no systematic relationships and zero trajectories, the rotation method was also applied to the data of the DS group. For trajectories built against CA, the rotation method produced non-significant R^2 values for all the emotions except for fear, where (after correction for multiple comparisons) a tendency towards significance was observed (happiness, $F(1, 38) = 2.65$, $p = .12$, $R^2 = .13$, sadness, $F(1, 38) = 0.45$, $p = .51$, $R^2 = .02$, fear, $F(1, 38) = 7.37$, $p = .014$, $R^2 = .29$, anger, $F(1, 38) = 0.64$, $p = .43$, $R^2 = .03$, and disgust, $F(1, 38) = 1.14$, $p = .72$, $R^2 = .01$). However, all trajectories built with MA as a predictor became highly significant after the rotation method (happiness, $F(1, 38) = 92.66$, $p < .001$, $R^2 = .84$, sadness, $F(1, 38) = 61.30$, $p < .001$, $R^2 = .77$, fear, $F(1, 38) = 157.09$, $p < .001$, $R^2 = .90$, anger, $F(1, 38) = 24.92$, $p < .001$, $R^2 = .58$, and disgust, $F(1, 38) = 31.90$, $p < .001$, $R^2 = .64$). Therefore, the distribution of accuracy scores was random with respect to the predictor of CA while MA produced zero trajectories.

Discussion

The aims of the study were organized according to the different methodological approaches adopted in this research. Results are therefore first discussed by following the same structure. Then, results are further discussed and lines of future work are outlined. Finally, conclusions are drawn.

Discussion of results obtained from the matching approach

Comparisons between the results of all groups (WS, DS, TD MA-matched and TD CA-matched) on the facial expression recognition task showed that while individuals with WS performed at the level of their TD peers matched on MA (but

below the level of the TD group matched on CA), the performance of the DS group was significantly lower than that of the TD MA-matched group. In addition, participants with WS obtained significantly higher results than participants with DS. These results are consistent with those previously found in the literature in this respect regarding both the performance of individuals with WS or DS compared to TD peers (Gagliardi et al., 2003; Kasari et al., 2001; Hippolyte et al., 2008; Hippolyte et al., 2009; Porter et al., 2007; Porter et al., 2010; Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000) and the cross-syndrome comparison (Porter et al., 2007).

Importantly, these between-group differences were found for all the emotions assessed in this research including happiness. Therefore, as hypothesized, this result would not support the idea suggested in prior research that individuals with WS or DS present a relative sparing for happiness together with more severe impairments for the recognition of negative emotions (e.g., Plesa-Skwerer, Faja et al., 2006; Porter et al., 2007). The finding that unlike some previous studies participants in the current research with WS or DS did not show proficiency for the emotion of happiness may be a result of the lack of floor and ceiling effects across the ability range of the disorder groups in combination with the use of an index to control for response biases. Thus, the methodological problems that introduced confounds in previous studies (Plesa-Skwerer, Faja et al., 2006; Porter et al., 2007; Santos et al., 2009) were overcome in this research.

Even so, we found higher performance on the positive compared to the negative emotions in the WS and DS groups. However, importantly, the same pattern of emotion recognition was found both for the CA and the MA-matched TD groups. Hence, unlike what has been previously suggested (Plesa-Skwerer, Faja et al., 2006), this pattern of results was not specific to participants with WS or DS. On the contrary, as already noted in the literature (Adolphs, 2002), better recognition of positive facial expressions

represents the typical pattern. Thus, the differences observed between positive and negative expressions in the four groups included in the study can be accounted for by the hierarchical organization of the basic emotional facial expressions (Adolphs, 2002; Plesa-Skwerer, Faja et al., 2006) in which individuals first categorize these expressions into the superordinate categories of happy and unhappy, and then distinguish the subordinate categories (Adolphs, 2002). Consequently, when happiness is compared with sadness, fear, anger and disgust, the negative expressions result more confusable (Adolphs, 2002). The fact that there was only one positive emotion in face of four negative emotions might lead to artificial inflation of the accuracy of identifying happiness. Thus, it could be argued that the inclusion of other positive emotions may lead to different results. However, it is important to note that TD children accurately recognize the basic emotion of happiness before the other basic negative emotions (Herba & Phillips, 2004), even when children are presented with only two emotional facial expressions (e.g., happiness vs. anger) (Labarbera, Izard, Vietze, & Parisi, 1976). Similar results have been found in WS and DS. When presented with happy and fearful facial expressions, both children with WS and those with DS show sensitivity to happy expressions but have difficulties understanding the communicative significance of fear in faces (Thurman & Mervis, 2013). Therefore, it seems unlikely that the higher performance on happiness in comparison with the negative emotions is only the result of a possible artifact in the design of the study.

Apart from studying the possible differences for recognizing positive vs. negative emotions in WS and DS, with the matching approach we also aimed to clarify whether individuals with WS or with DS present specific impairments for any of the emotions included in the study and whether cross-syndrome differences exist . Prior studies have been inconsistent as to specific expression deficit . As previously

mentioned, studies have usually included a small number of trials per emotion and this has made it difficult to reach solid conclusions on the matter (Williams et al., 2005). Compared to other studies, the current research included a considerably larger number of trials per emotion. Under this circumstance and, as hypothesized, neither participants with DS nor those with WS showed specific deficits for any of the emotions studied and thus no cross-syndrome differences were found in this respect. Furthermore, as aforementioned, the pattern of emotion recognition was the same for the WS, DS and TD groups.

Discussion of results obtained from the developmental trajectories approach

In the TD participants, accuracy on the facial expression recognition task improved both along with CA and MA. Therefore, we found that TD individuals exhibit developmental changes in facial expression recognition. Moreover, as previously described (e.g., Herba & Philipps, 2004), happiness was recognized earlier than the negative emotions. Results also showed that the rate of development was the same for all the emotions included in the study.

Unlike TD participants, expression recognition in WS and DS was not predicted either by CA or MA. Therefore, the two disorder groups showed atypical facial expression recognition development. The lack of significant relationships between CA or MA and accuracy on facial expression recognition in WS and DS is consistent with prior findings in the literature (Gagliardi et al., 2003; Santos et al., 2009; Williams et al., 2005; Wishart & Pitcairn, 2000).

In the current research, this atypicality was further elucidated by using the rotation method (Thomas et al., 2009). For both WS and DS while performance was randomly related to CA, using MA as predictor for accuracy produced zero trajectories. The null relationship between accuracy and CA can be explained by taking into account

the cross-sectional design of the methodological approach followed. Given the usual large variability of severity in individuals with developmental disorders, when working with cross-sectional data, performance and participants' CA will not be necessarily related (Thomas et al., 2009). Instead, severity is factored out by using MA a significant relationship between MA and accuracy may be expected. Yet, as aforementioned, we found trajectories with a gradient of zero when MA was entered as predictor.

Zero trajectories are interpreted as showing a system that has reached its maximum developmental level due to cognitive constraints (Thomas et al., 2009). However, before drawing this conclusion, when zero trajectories are observed it is important to rule out different possible artifacts (i.e., chance performance, ceiling or floor effects, and non-linear trajectories) that may better explain the data (Thomas et al., 2009). In our study, participants with WS and those with DS performed significantly above chance level and no floor or ceiling effects were found for these groups. In addition, non-linear functions did not represent a good fit for the data. Therefore, we would conclude that the zero trajectories found when MA was taken as predictor show that development on facial expression recognition is indeed static across MA both in WS and DS. Thus, individuals with WS and those with DS progress as much as they can and reach an early maximum developmental level due to the constraints of their cognitive systems. Here lies the atypicality.

As previously discussed, prior studies of facial expression recognition in WS and DS have only used the matching approach and this has led to conclude that facial expression recognition is delayed in WS and atypical in DS (e.g., Kasari et al., 2001; Porter et al., 2007; Riby, 2012; Williams et al., 2005; Wishart & Pitcairn, 2000). Instead, by using the developmental trajectories approach we have found that not only individuals with DS but also those with WS exhibit developmental atypicality, as

expected. Thus, whilst the use of a matching approach would suggest development dissociation between WS and DS, the inclusion of the developmental trajectories approach in this study has revealed similarities in the developmental pathways for recognizing emotional facial expressions both in WS and DS.

Further discussion, lines of future work and limitations of the study

One of the questions now arising would be why participants with DS performed significantly lower than participants with WS despite the fact that on facial expression recognition tasks no significant differences are usually found between individuals with DS and those with other developmental disorders (Williams et al., 2005; Wishart et al., 2007; Wishart & Pitcairn, 2000). It should be noticed that in the current research, facial expression recognition was assessed through a labeling task. Labeling tasks are often used in the literature (e.g., Adolphs, 2002) and the same type of task was used in the only prior study comparing facial expression recognition in WS and DS (Porter et al., 2007). However, labeling tasks require not only facial perceptual skills but also verbal skills for labeling. Therefore, these tasks lie at the intersection of facial emotion perception and linguistic description of affect (Gagliardi et al., 2003). Individuals with DS typically present lower verbal abilities than individuals with WS (e.g., Bellugi et al., 1994) so this may partially explain the lower results obtained by participants with DS compared to their peers with WS. Nevertheless, it should be considered that the impairments for facial expression recognition in DS have been found across different tasks and not only labeling (Kasari et al., 2001; Wishart & Pitcairn, 2000). Therefore, although the verbal requirements of the task may have contributed to the lower results of participants with DS, it seems unlikely that this factor can account on its own for the differences observed between the DS and WS groups.

As suggested by Kasari et al. (2001), the difficulties of children with DS in

recognizing emotional facial expressions, especially when verbal skills are also required, may be related to the environmental input to which they are exposed. Thus, compared to maternal speech to TD children, mothers' speech to children with DS includes fewer emotion terms (Tingley, Gleason, & Hooshyar, 1994). Therefore, children with DS could be less exposed to emotion labels, which could affect their emotional knowledge and, in consequence, also their skills for facial expression recognition (Kasari et al., 2001). To our knowledge, no such research has been performed in WS and maternal speech has not yet been examined in WS. Considering the impact that a different environment can have on the development of children with developmental disorders (Karmiloff-Smith, 2009), this is an issue that warrants further research.

Another factor that may also contribute to explaining the results found in the current study is the atypical character of face processing in both WS and DS. In WS, holistic and configural facial processing has been found to be atypical across development so that, unlike TD individuals, those with WS seem to specialize in featural processing (e.g., Annaz et al., 2009; Karmiloff-Smith et al., 2004; but see Isaac & Lincoln, 2011, for contradictory results on configural processing). The development of a featural-based strategy for face processing in individuals with WS may arise from sticky fixation and difficulties from infancy to plan rapid saccadic eye movements (Brown et al., 2003; Karmiloff-Smith, 2009). This, in turn, may contribute to the explanation of why individuals with WS are fascinated with faces from early in development (Karmiloff-Smith, 2012). In DS, face processing has been scarcely investigated but there is also evidence of atypical development whereby individuals with DS seem to be poor at featural processing and rely more on holistic strategies (Annaz et al., 2009). Although individuals with WS and those with DS present different

face encoding strategies (Annaz et al., 2009), in both cases atypical configural processing may account for their impairments for recognizing emotional facial expressions (Carvajal et al., 2012; Gagliardi et al., 2003). Further research should study the processes underlying the development of facial expression recognition in individuals with WS and those with DS.

Atypicalities have also been found on the neural mechanisms underpinning facial expression recognition in WS and DS. In WS, greater amygdala volumes relative to TD controls have been reported (Martens, Wilson, Dudgeon, & Reutens, 2009; Reiss et al., 2004). In addition, studies conducted with adults with WS have shown reduced activation of the amygdala to negative facial expressions (Haas et al., 2009; Meyer-Lindenberg et al., 2005; Mimura et al., 2010) but heightened amygdala response to positive faces (Haas et al., 2009). These structural and functional abnormalities have been linked to the social profile of individuals with WS (Haas et al., 2010; Martens et al., 2009). In DS, few structural or functional imaging studies have been carried out (Wishart et al., 2007). Even so, amygdala volumes have been found to be reduced in children and adults with DS (Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Krasuski, Alexander, Horwitz, Rapoport, & Shapiro, 2002; Śmigielska-Kuzia et al., 2011). Nevertheless, in DS results are not clear for children (Pinter et al., 2001) and amygdala reduction may be associated to dementia in adulthood (Aylward et al., 1999). Yet, in order to better understand facial expression recognition in WS and DS, future studies should compare patterns of amygdala activation to emotional facial expressions in both syndromes.

The fusiform gyrus, a critical structure for face recognition, is also atypical in WS and may be associated with the static development in facial expression recognition skills seen here. Within the fusiform gyrus, a functionally defined area, the Fusiform

Face Area (FFA) is responsive to faces with areas nearby and overlapping the FFA having been found to be activated in discriminations of objects which an individual is an expert on (McGugin, Gatenby, Gore, & Gauthier, 2012) as well as by body parts (Schwarzlose, Baker, & Kanwisher, 2005). In WS the fusiform gyrus itself appears to be enlarged (Reiss et al., 2004) and also better connected with other structures (greater fractional anisotropy) compared to TD controls (Hass et al., 2012; Haas et al., 2013). Furthermore, the FFA enlargement in WS is due to an increased overlap with object selective areas of the fusiform gyrus and the enlargement seems functional at least in facial identity recognition ability (Golarai et al., 2010). The location of the FFA within the fusiform gyrus appears to be determined by the structural connectivity of the fusiform gyrus in TD individuals (Saygin et al., 2012). Thus, it may be that individuals with WS, through recruiting larger areas of the fusiform gyrus are making use of areas which are better adapted (possibly by virtue of their connections to other areas of the brain) for more general object processing rather than having the abilities and connectivity which would enable very specialized face processing. This may limit face processing ability contributing to explaining both the early maximum developmental level seen here and the featural-based strategies somewhere else reported (Annaz et al., 2009; Karmiloff-Smith et al., 2004).

An important finding of the current study is that both individuals with WS and those with DS reach an early maximum developmental level for facial expression recognition and that, once this level is reached, performance is static across age. This has implications for the ability of individuals with the two syndromes to advance in their knowledge of the social cues which are expressions. A zero trajectory implies that further learning is constrained by the underlying neurocognitive abnormalities and thus further understanding of other people's emotional expressions would only be achieved

through the use of very different strategies which recruit other neural processes. Not improving in their skills for recognizing emotional facial expressions along with age may, in turn, trigger a cascade of negative effects along the years. Thus, atypical rates of social approachability in WS and DS seem to be somehow linked to problems for labeling emotional facial expressions (Porter et al., 2007). In turn, inappropriate evaluation of social approachability from emotional facial expressions leads to approaching strangers indiscriminately (Jones et al., 2000; Porter et al., 2007) and this places individuals with WS and those with DS in high risk of exploitation. In fact, individuals with WS or DS suffer from high rates of victimization and social vulnerability (Fisher, Moskowitz, & Hodapp, 2013) and this is one of the most common concerns expressed by their parents. Intervention programs based on teaching the social meaning of the emotional facial expressions could help to ameliorate this problem. Future research should not only design these programs but also assess their efficacy. Difficulties for recognizing emotions in the faces of others have also been associated with problems in social behavior and psychiatric disorders such as anxiety (e.g., Demenescu, Kortekaas, den Boer, & Aleman, 2010; Izard et al., 2001), problems often found in individuals with WS or DS (Dykens, 2000). Further research should therefore assess whether such problems are indeed related to the difficulties individuals with WS or DS present for facial emotion recognition. Future research should thus study the impact of these difficulties in their lives.

Conclusions

We studied facial expression recognition in WS and DS by using both the matching and developmental trajectories approaches. By using the matching approach and taking steps to avoid artifacts that may have introduced confounds in prior research we have ascertained that individuals with WS or DS do not present specific impairments

for any of the negative emotions included in the study and that they do not exhibit proficiency in the recognition of happiness. Unlike previously studies (e.g., Hippolyte et al., 2009; Plesa-Skwerer, Faja et al., 2006), we found that the better performance on positive than negative emotions usually reported in WS and DS is not specific of these populations but reflects a typical pattern. Thus, individuals with WS and those with DS present the same pattern of emotion recognition as TD individuals. Prior results from the matching approach have led to the suggestion that while individuals with WS present a developmental delay in facial expression recognition, individuals with DS present atypical development in the same domain (e.g., Kasari et al., 2001; Porter et al., 2007; Riby, 2012; Williams et al., 2005; Wishart & Pitcairn, 2000). However, by using the developmental trajectories approach, we have shown that both groups exhibit atypical development in facial expression recognition even though performance levels are lower in DS than in WS. In both individuals with WS and those with DS, constraints linked to their cognitive systems prevent age related facial expression development.

Acknowledgments

This study was partially supported by Italian National Institute of Health research funding (Ministero della Sanità) grant to RB (RC/01/02/2013) and partially by the Department of Developmental and Education Psychology, Universidad Nacional de Educación a Distancia, Spain. We also thank Elisa Frigerio and Ilaria Cazzaniga for their help in data collection. We are especially grateful to all the parents and participants who collaborated in this research.

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Table 1. *Descriptive Characteristics of the WS, DS and TD Groups.*

	WS group	DS group	CA-matched TD group	MA-matched TD group
N	20	20	20	20
Gender (M/F)	10/10	11/9	9/11	9/11
CA Mean	147.35 (56.38)	150.45 (36.62)	151.90 (40.54)	59.50 (9.98)
CA Range	68-283	90-216	90-262	49-82
MA Mean	67.10 (9.15)	62.85 (11.79)	170.30 (42.89)	64.83 (8.55)
MA Range	51-82	43-79	102-262	51-78

Note: CA and MA are expressed in months. Standard deviations are presented in brackets.

Table 2. *Unbiased Hit Rates and Chance Performance for each Emotion and Group.*

Emotion	Measurement	WS group	DS group	CA-matched TD group	MA-matched TD group
Happiness	Unbiased hit rate	0.74 (0.16)	0.54 (0.24)	0.96 (0.05)	0.75 (0.13)
	Chance performance	0.05 (0.01)	0.06 (0.02)	0.04 (0.002)	0.05 (0.01)
Sadness	Unbiased hit rate	0.39 (0.22)	0.22 (0.19)	0.60 (0.24)	0.43 (0.21)
	Chance performance	0.03 (0.02)	0.03 (0.02)	0.03 (0.01)	0.03 (0.01)
Fear	Unbiased hit rate	0.46 (0.23)	0.26 (0.31)	0.71 (0.23)	0.43 (0.24)
	Chance performance	0.03 (0.01)	0.02 (0.02)	0.04 (0.007)	0.04 (0.02)
Anger	Unbiased hit rate	0.47 (0.19)	0.21 (0.14)	0.62 (0.19)	0.46 (0.17)
	Chance performance	0.05 (0.02)	0.05 (0.02)	0.04 (0.01)	0.04 (0.01)
Disgust	Unbiased hit rate	0.33 (0.22)	0.19 (0.16)	0.63 (0.23)	0.49 (0.23)
	Chance performance	0.03 (0.02)	0.04 (0.03)	0.04 (0.009)	0.04 (0.02)

Note: Standard deviations are presented in brackets.

Figure Captions

Figure 1. Developmental trajectories for the typically developing group.

Figure 2. Developmental trajectories for the Williams syndrome group.

Figure 3. Developmental trajectories for the Down syndrome group.