

# Is There a Social Module? Language, Face Processing, and Theory of Mind in Individuals with Williams Syndrome

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## Abstract

■ Many species can respond to the *behavior* of their conspecifics. Human children, and perhaps some nonhuman primates, also have the capacity to respond to the *mental states* of their conspecifics, i.e., they have a "theory of mind." On the basis of previous research on the theory-of-mind impairment in people with autism, together with animal models of intentionality, Brothers and Ring (1992) postulated a broad cognitive module whose function is to build representations of other individuals. We evaluate the details of this hypothesis through a series of

experiments on language, face processing, and theory of mind carried out with subjects with Williams syndrome, a rare genetic neurodevelopmental disorder resulting in an uneven linguistic-cognitive profile. The results are discussed in terms of how the comparison of different phenotypes (e.g., Williams syndrome, Down syndrome, autism, and hydrocephaly with associated myelomeningocele) can contribute both to understanding the neuropsychology of social cognition and to current thinking about the purported modularity of the brain. ■

## INTRODUCTION

Sometimes nature provides the scientist with experiments of its own, in the form of various abnormal phenotypes. Down syndrome, autism, hydrocephaly with associated myelomeningocele, and Williams syndrome are among such examples. Because of their different patterns of abilities and impairments, these phenotypes can shed light on theories of normal development. Taking Down syndrome as an example, one might conclude from the seeming across-the-board retardation in language and all cognitive domains that a theory of development calling on domain-general mechanisms of learning would suffice to account for both normal development and its abnormalities. However, as soon as we consider syndromes with uneven cognitive profiles, i.e., with particular islets of ability or impairment, such a conclusion becomes questionable.

The value of comparing different abnormal phenotypes can be illustrated from the domains of language

and face processing. For example, the majority of adolescent subjects with Williams syndrome perform well on language and face processing tasks, despite serious deficits in spatial cognition, number, and problem solving (Bellugi, Bihrlé, Neville, Jernigan, & Doherty, 1992; Bellugi, Bihrlé, Jernigan, Trauner, & Doherty, 1990; Bellugi, Marks, Bihrlé, & Sabo, 1988; Bellugi, Wang, & Jernigan, 1993; Bihrlé, Bellugi, Delis, & Marks, 1989; Karmiloff-Smith, 1992a,b; Karmiloff-Smith, Grant, & Berthoud, 1994; Reilly, Klima, & Bellugi, 1991; Udwin, 1990; Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987; Wang & Bellugi, 1993, 1994; Wang, Doherty, Hesselink, & Bellugi, 1992). Why do seemingly unrelated skills like language and face processing cluster in the development of Williams syndrome, whereas more ostensibly related skills such as facial and visuospatial processing do not? Nativists would tend to use these facts to support a strictly modular, genetically specified view of development, in which certain modules can be spared and others damaged.

Connectionist theorists would tend to seek an explanation in terms of domain-general learning algorithms and of the massive superimposed input of speech and faces available to young infants. However, on the basis of the comparison of different phenotypical patterns, a far more epigenetic view is called for, in which skeletal architectural, computational, and/or representational predispositions interact with proprietary inputs, resulting in a gradual process of modularization as an emergent product of development (Elman, Bates, Johnson, Karmiloff-Smith, Parisi, & Plunkett, in press; Johnson & Karmiloff-Smith, 1992; Karmiloff-Smith, 1986, 1992a,b,c; Kuhl, 1991).

In this paper, we compare two phenotypes, Williams syndrome (WS) and autism, in respect of the capacity that normal children display to make inferences on the basis of others' mental states. Many theorists, particularly of a Piagetian or connectionist persuasion, might assert that such a capacity derives solely from domain-general learning mechanisms applied to representations of sociocultural input. Yet a rather specific deficit in intentionality and reasoning about other minds (henceforth "theory of mind") has been identified in individuals with autism, with other aspects of their cognition relatively spared (Baron-Cohen, Leslie, & Frith, 1985, 1986; Frith, 1989; Happé, 1993; Leekam, Baron-Cohen, Perrett, Milders, & Brown, 1993; Sigman, Ungerer, Mundy, & Sherman, 1987; see Baron-Cohen, Tager-Flusberg, & Cohen, 1993, for a review). This suggests that theory-of-mind computations cannot be explained by domain-general mechanisms alone.

A crucial distinction must be drawn between the capacity to respond to another's *behavior*, on the one hand, and to respond to another's *mental state*, on the other. Most species possess mechanisms that allow them to respond to conspecific behavior. Human children, and perhaps some nonhuman primates (Premack, 1988; Premack & Woodruff, 1978), also have the capacity to evaluate and respond to another's mental state (Astington, Harris, & Olson, 1988; Karmiloff-Smith & Russell, 1994; Leslie, 1987; Perner, 1988, 1991; Perner & Wimmer, 1985; Wellman, 1990; Wimmer & Perner, 1983). The capacity to respond to another's mental state is more complex than responding to another's behavior for two reasons. First, it involves nonobservables (making inferences about another's mind). Second, it entails the capacity to represent what in philosophy of mind is called "propositional attitudes," and to differentiate these from the "propositional contents" on which they operate (Dennett, 1971). A statement such as "*There is a pencil on the table*" may be a true or false proposition about the state of the world. By contrast, "I BELIEVE (PRETEND, DESIRE, KNOW, THINK, GUESS, HOPE, CLAIM, etc.) that '*there is a pencil on the table*'" involves a mental attitude toward that proposition and does not entail the truth or falsehood of the content.

The earliest manifestation of the capacity for computations of the propositional-attitude type seems to ap-

pear in toddlers around 18 months, in their pretend play (Leslie, 1987). The capacity is progressively generalized to understanding mental state verbs such as BELIEVE, THINK, and KNOW, and from about 4 years of age is extended to understanding false belief (Perner, 1991; Wellman, 1990; Wimmer & Perner, 1983). As mentioned above, individuals with autism, even with IQs within the normal range, have difficulty with tasks involving propositional attitudes and the attribution of mental states to others, where normal subjects as young as 3 to 4 years old succeed (Baron-Cohen, 1989a, 1991; Baron-Cohen et al., 1985, 1986, 1993; Frith, 1989; Happé, 1993; Leslie & Frith, 1988). Individuals with autism also show deficits in using direction of eye gaze as an index of another person's mental state of interest, attention, or intention (Baron-Cohen, Campbell, Karmiloff-Smith, Grant, & Walker, in press; Leekam et al., in press), and in interpreting nonliteral intentional language such as jokes, sarcasm, and irony (Happé, 1991, 1993; Leekam, 1991).

The theory of mind deficit in autism, as well as animal models of intentionality, have been invoked to support the interesting claim that socially-relevant representations are processed within what constitutes a broad cognitive module (Brothers & Ring, 1992). A module, according to Fodor (1983), is hard-wired (not assembled from more primitive processes), of fixed neural architecture, domain specific, fast, autonomous, mandatory, automatic, stimulus-driven, and insensitive to central cognitive goals. A further characteristic of a module is that it is informationally encapsulated from knowledge elsewhere in the system.

The module proposed by Brothers and Ring encodes high-level psychological representations of other individuals, including general biological stimuli such as faces, voices, and animate movement. The processing of social information depends on intact limbic structures, i.e., the amygdala and orbital frontal cortex with which the amygdala is interconnected (Price, Russchen, & Amaral, 1987). Damage to the limbic frontal lobes gives rise to an inability to generate appropriate automatic responses to real-life social stimuli, despite preserved social problem solving when presented in abstract verbal form (Eslinger & Damasio, 1985). This fits with the fact that the few high-functioning people with autism who succeed on theory-of-mind tasks continue to fail to make use of mental state inferences in their daily lives (Happé, 1991).

In this paper we evaluate the Brothers-Ring hypothesis of a broad cognitive module for representing and processing stimuli relevant to other individuals, including face processing, language, and theory of mind. We focus in particular on the theory-of-mind capacities of a group of subjects with Williams syndrome, for whom it has already been established that language and face processing are rather well preserved compared to other serious cognitive deficits (Bellugi, Bihrlé, Neville, Jernigan, & Doherty, 1992; Bellugi, Bihrlé, Jernigan, Trauner, & Do-

herty, 1990; Bellugi & Morris, in press; Bellugi & Wang, 1992; Bellugi, Wang, & Jernigan, 1994; Bihrlé, Bellugi, Delis, & Marks, 1989; Karmiloff-Smith, 1992a,b; Karmiloff-Smith, Grant, & Berthoud, 1993; Reilly, Klima, & Bellugi, 1991; Udwin, 1990; Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987; Wang & Bellugi, 1993, 1994; Wang, Doherty, Hesselink, & Bellugi, 1992). Since the relative preservation of language and face processing in WS can be related to species-specific representations of other individuals, we used the Brothers-Ring hypothesis to predict that WS subjects will also perform well on a variety of theory-of-mind tasks.

It is of course essential to keep in mind distinctions between different aspects of language and face processing, some of which relate to representing other individuals in general, others that relate more directly to theory-of-mind computations, and yet others that seem to be unrelated to theory of mind. With respect to face processing, computing the direction of eye gaze (a general capacity) is not the same as using direction of eye gaze to infer another's intention. Categorizing facial expressions as happy or sad may involve different capacities for interpreting a facial expression as conveying thinking, surprise, or curiosity. Likewise in language, processing syntactic form involves different capacities for understanding the pragmatics of another's intended meaning, e.g., in the case of sarcasm and irony. Some of these capacities may cluster with theory-of-mind abilities, whereas others may be quite distinct from them. In comparing WS and autism, we can thus predict different patterns of abilities and deficits across the theory-of-mind-related aspects of language and face processing.

In the next two sections, we provide an overview of the biological, neurological, and cognitive profile of Williams syndrome. We then present a summary of the studies we carried out, which support our prediction of a relatively preserved theory-of-mind capacity in WS subjects. We go on to discuss the results in terms of the purported modular structure of the mind and, by comparing different abnormal phenotypes, we evaluate the Brothers-Ring hypothesis of the existence of a broad cognitive module for processing social stimuli relevant to the representation of other individuals. In the final section, we give details of the methodology.

## **WILLIAMS SYNDROME: BIOLOGICAL/NEUROLOGICAL PROFILE<sup>1</sup>**

Williams syndrome [or, in continental Europe, known as Beuren's syndrome (Beuren, 1972; Beuren et al., 1984) and, in the United Kingdom, also as infantile hypercalcemia] affects approximately 1 in 20,000 to 50,000 live births (Greenberg, 1990). It was initially described in the 1950s and 1960s (Williams et al., 1961; Beuren et al., 1964; Fanconi et al., 1952; Lightwood, 1952; Creery, 1953). The first WS Parents' Association was started in the United Kingdom in 1980 (C. Cooper, personal communi-

cation), and there rapidly followed the establishment of WS associations in the United States, France, Belgium, and elsewhere. The syndrome has recently received renewed attention. For instance, there are now some 10 WS clinics in the United States, as opposed to none a few years ago. In particular, geneticists and cognitive psychologists have focused on the particularities of the syndrome, leading to better diagnostic tools. WS can now be diagnosed early in infancy by genetic or metabolic markers, but until very recently it often went undiagnosed until middle childhood, adolescence, or even adulthood. With the new diagnostic tools, the incidence of WS may actually turn out to be higher than the above estimate.

WS is typified by a number of common physical characteristics (Jones & Smith, 1975; Morris, Demsey, Leonard, Dilts, & Blackburn, 1988). First, the syndrome gives rise to a typical facial dysmorphology: an elfin-like face, with heavy orbital ridge, temporal dimples, full cheeks, retroussé nose, flat nasal bridge, flared nostrils, stellate iris pattern, wide mouth, and irregular dentition (McKusick, 1988). WS frequently results in hypertension, as well as supravalvular aortic stenosis and peripheral pulmonary artery stenosis, a vascular disease causing a narrowing of major arteries. WS infants are below average birth weight and often show no spontaneous sucking, with frequent vomiting during the first few months. They manifest unusual sacral creases and poor muscle tone, as well as premature aging of the skin. WS children also suffer from hyperacusis, an abnormal sensitivity to sound, although this tends to become less acute with development (Marriage, 1994).

WS is genetic in origin (although of course the phenotype is epigenetic). Autism has also been shown to have a genetic basis (Frith, 1989; Rutter, 1991). WS always occurs in both members of monozygotic twins, but not in the case of dizygotic twins (Murphy et al., 1990). Several hypotheses have been put forward with respect to the precise cause of WS. It has been argued that the mode of transmission can be autosomal dominant (Morris & Moore, 1991), but most cases are thought to result from new mutations. One hypothesis about the cause of WS relates to abnormal calcium metabolism, which may result from a defect in the gene used to produce calcitonin (a calcium-regulating hormone) and/or in the gene used to produce a modified version of calcitonin gene-related peptide (CGRP), a chemical messenger in the brain (Culler, Jones, & Deftos, 1985; see discussions in Bellugi, Bihrlé, Neville, Jernigan, & Doherty, 1992; Simmons, Culler, Bellugi, & Greenberg, 1990). Several studies in Britain, the United States, and France have now ruled out uniparental disomy. More recently, it has been suggested that a microdeletion, affecting a number of contiguous genes, is at the root of WS. The most recent family studies have linked supravalvular aortic stenosis (characteristic of WS) to the elastin gene on the long arm of chromosome 7 (Ewart, Morris, Atkinson, Jin, Sternes,

Spallone, Dean Stock, Leppert, & Keating, 1993; Bellugi & Morris, in press), suggesting indeed that WS is a disorder in which vascular and connective tissue abnormalities are caused by the deletion of one elastin allele and other contiguous genes. All the family cases tested indicate hemizygoty at the elastin locus. Other nonfamilial cases showed in some cases the expected pattern of inheritance, and others a *de novo* deletion of one allele on the elastin gene. Of course, the neurobehavioral features of WS, to which we turn in a moment, are not directly explicable in terms of hemizygoty at the elastin locus. It is possible that elastin influences neurodevelopment through its functional role in the vasculature (Galaburda, Wang, Bellugi, & Rossen, 1994). It is also possible that the severity of mental retardation is related to the size of the microdeletion, again pointing to the involvement of several contiguous genes (Ewart et al., 1993).

High levels of whole blood serotonin have also been found in WS individuals (August & Realmuto, 1989; Mariage, 1994), as well as high levels of calcium (Arnold, Yule, & Martin, 1985). WS affects males and females equally. There are possibly two subgroups, one involving idiopathic infantile hypercalcemia (IIH) and another in which IIH does not seem to be present. Where IIH is diagnosed, reduction in calcium and vitamin D intake reverts calcium levels in the blood to normal, and where IIH is not diagnosed it is still possible that infants recover the correct calcium levels in the blood spontaneously. Despite the subsequent normality of calcium levels, other characteristic physical and mental abnormalities remain, due possibly to a neurotransmitter imbalance in the central nervous system (Bellugi, Bihrl, Neville, Jernigan, & Doherty, 1992; Bellugi & Morris, in press; Udwin, 1990; Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987).

Calcium plays an important role in regulating chemical reactions inside brain cells, capable of binding almost 100% of the internal calcium ions of cells. A number of ion channels on dendrites and cell bodies of neurons are calcium-sensitive, several of which control the fine-grained timing and location of short-term habituation and dishabituation of cell firing. Calcium is also crucially involved in long-term potentiation, weight changes between neurons, and changes in firing thresholds within particular circuits. Calcium currents at the postsynaptic cell feed back into the presynaptic cell, thereby increasing the subsequent probability of neurotransmitter release. Some neurons have slow calcium spikes instead of sodium spikes. This is particularly true early in development. Calcium and sodium channels are both usually excitatory, but the major difference between them is that calcium channels tend to have much slower kinetics (seconds versus milliseconds) (Kandel, Schwartz, & Jessell, 1991). At synapses, calcium is crucial for the fusion between the vesicles and the membrane that releases neurotransmitters.

All of this suggests that elevated levels of calcium in

WS blood during embryogenesis and/or postnatal brain growth, leading to elevated calcium levels inside brain cells, could throw the delicate timing/location/use of calcium ions out of balance, thereby deregulating neurotransmitter release in the developing brain cells. It has been shown that an increased level of calcium can diminish apoptotic cell death (Lipton & Kater, 1989; see, also, Johnson & Karmiloff-Smith, 1992, for a discussion of the relationship between cell death and emergent specialization). How this relates to the particular resulting *cognitive* characteristics of the syndrome is still unknown, but it clearly would affect the timing and patterning of parcellation of different brain circuits, thus leading to aberrant neural organization.

A number of recent studies have compared brain structure in normal adults, and those with WS or autism, using various techniques including PET, MRI, and ERP. WS brains turn out to be only 80–85% of the volume of normal brains (Jernigan & Bellugi, 1990, 1994; Wang, Hesselink, Jernigan, Doherty & Bellugi, 1992). However, compared to normals, there are no obvious left/right symmetry differences and no obvious space-occupying lesions in either the right or the left hemisphere. Total cerebellar volume is entirely normal in WS subjects. Leiner and Leiner (1989) suggested that the neocerebellar versus paleocerebellar regions control different functions in higher cognitive processing and are ontogenetically, anatomically, and developmentally distinct. What is particularly interesting for our present purposes is that the volumetric differences in these regions may be opposite in the brains of people with WS versus those with autism. Neocerebellar, but not paleocerebellar, lobules have been reported to be significantly smaller in people with autism (Courchesne et al., 1988), whereas there is a striking increase in neocerebellar volumes in WS subjects (Jernigan & Bellugi, 1990; Bellugi, Wang, & Jernigan, 1994). If these results are robust and nonartificial, the volumetric differences in WS may turn out to be the reverse of those found in autism, and help explain the different cognitive profiles in the two syndromes.

As for morphological features within the cerebrum, supratentorial volume in WS is reduced. Total cerebral gray matter shows a highly significant reduction compared to normal controls. However, when anterior and posterior cortical regions are analyzed separately, then similarities between WS and normal controls become evident. Although WS subjects' anterior regions are proportionately smaller than controls, the difference does not reach significance. By contrast, the brains of both WS and controls show significantly larger anterior regions than Down syndrome brains (Jernigan & Bellugi, 1990, 1994; Wang, Hesselink, Jernigan, Doherty, & Bellugi, 1992). There is no difference between WS and controls with respect to the limbic structures in the temporal lobe. Indeed, limbic structures of the temporal lobe (including uncus, amygdala, hippocampus, and parahippocampal gyrus) are relatively spared in WS (Galaburda,

Wang, Bellugi, & Rossen, 1994). By contrast, the diencephalic and lenticular proportions are reduced in WS as compared to overall cerebral reductions. WS brains also show reduced caudate nuclei volumes (Jernigan & Bellugi, 1990, 1994).

In sum, WS typically presents no evidence for lateralized/focal lesions. Rather, differing patterns of brain growth during embryogenesis and ontogenesis result in different brain volume proportions. WS cerebrum is small, but the frontal cortex acquires a near normal volume relationship to the posterior cortex (Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Wang et al., 1992). In particular, the ratio of neocerebellar to paleocerebellar structures may be opposite in people with WS and autism. Clearly more work has to be done to determine the relationship between these volumetric differences in the cerebellofrontocortical system and the behavioral profile that emerges, but it is important to stress that WS brains are not lesioned as such. Recent work, however, points to cytoarchitectonic anomalies in the form of exaggerated horizontal organization of neurons within layers, increased cell packing density throughout brain regions, decreased myelination, and abnormally clustered and oriented neurons, particularly in visual cortex (Galaburda, Wang, Bellugi, & Rossen, 1994).

If at the biological/neurological level WS continues to be a puzzle to the neuroscientist, why is it of such interest to the developmental cognitive scientist? As we shall see below, unlike Down syndrome (which seems to suggest an across-the-board deficit in functioning, and could be used to argue for a domain-general view of development), WS and autism both present an uneven pattern of cognitive dissociations that, although different, challenges the domain-general view of cognition.

### **WILLIAMS SYNDROME: LINGUISTIC/COGNITIVE PROFILE**

WS is typified by an uneven linguisticocognitive profile. A number of studies of WS subjects have now demonstrated surprisingly preserved linguistic and face-processing capacities, despite low IQs and serious deficits on tasks requiring the integration of visuospatial information, as well as number, motor, problem-solving, and planning tasks (full details can be found in Arnold, Yule, & Martin, 1985; Bellugi et al., 1990, 1992; Karmiloff-Smith, 1992a,b; Udwin et al., 1987). The full IQ of WS subjects is typically in the 50s-60s, although there is a fairly wide range from 40 through 90. Despite rather good language skills in many WS subjects, Verbal IQ does not always outstrip Performance IQ on the WISC-R (Udwin, Yule, & Martin, 1987; Crisco & Dobbs, 1988; Kataria et al., 1984; Arnold et al., 1985), suggesting that some standardized IQ tests mask the particular linguistic abilities of WS subjects (Bellugi, Bihle, Neville, Doherty, & Jernigan, 1992; Mervis, 1994; Volterra, 1994). However, specific

tests more sensitive to individual components of linguistic ability reveal the special linguistic capacity of WS subjects in comparison to other aspects of their cognition (Bellugi et al., 1992, Wang & Jernigan, 1994, Reilly et al. 1991; Karmiloff-Smith & Grant, 1992; Karmiloff-Smith, Grant, & Berthoud, 1993). However, further work is required to tease apart the syntactic, semantic, and pragmatic aspects of their language (Karmiloff-Smith, Tyler, & Sims, unpublished experiments).

What characterizes WS cognitively is that, like autism, it presents an uneven profile of islets of deficit and proficiency. Compared to their marked difficulties in simple arithmetic, some WS subjects are almost at ceiling on certain tasks measuring understanding of syntactically complex structures (Bellugi, Wang, & Jernigan, 1994; Wang & Bellugi, 1993). Also, in contrast to individuals with autism and Down syndrome, they produce complex narratives that make extensive use of affective prosody (Reilly, Klima, & Bellugi, 1991), a point to which we shall return later. Furthermore, WS subjects score as well as normal adults on the Benton, a face discrimination task (Bellugi et al., 1992) and better than matched controls on the face recognition subtask of the Rivermead Behavioural Memory Test (Udwin & Yule, 1991). More recent work confirms the remarkable sparing of face processing in WS individuals (Rossen, Jones, Wang, & Klima, 1994).

A series of studies have been undertaken using event-related potential (ERP) techniques to assess the timing and organization of neural systems active during sensory, cognitive, and language processing in WS subjects (Neville, Holcomb, & Mills, 1989; Neville, Mills, & Bellugi, 1993). Two of the notable characteristics of the WS behavioral profile have so far been investigated. First, the auditory recovery cycle has been tested for indices of hyperexcitability at any stage along the auditory pathway that might provide clues to the basis of their hyperacusis. Second, auditory sentence processing has been assessed as to whether such processing is mediated in WS by the same pathways that are active in normal age-matched controls.

Auditory brainstem-evoked responses turn out to be normal in WS subjects, indicating that hyperexcitability does not occur at the brainstem level. However, the auditory recovery data suggest a possible cortical mechanism subserving the apparent sensitivity to sounds. This is evident only over temporal cortex and is true solely with respect to auditory input; WS subjects are indistinguishable from normal controls on a visual recovery paradigm. Taken together, these studies suggest that the hyperacusis observed in WS may be mediated by hyperexcitability specifically within the cortical areas that are utilized in processing acoustic information.

ERPs have also been recorded of WS subjects' responses to auditorily presented words in sentences (Neville et al., 1994). One-half of the sentences were highly contextually constrained, ending with a semanti-

cally appropriate word, whereas the other half ended with an anomalous word (e.g., "I take my coffee with cream and paper"). Previous research has shown that normal subjects show a large negative response at 400 msec (N400) to semantically unprimed words and this is considered an index of how the mental lexicon is organized. WS subjects displayed responses that were highly abnormal within the first 200–300 msec following word onset. The abnormality consisted of a large positivity, not seen in normal control subjects at any age. This effect, only apparent over temporal brain regions, may relate to WS hyperacusis. Moreover, the WS responses did not show the asymmetry (left temporal regions more negative than right) that is typical by about 7 years of age in normal children (Mills, Coffey, & Neville, 1993), pointing to an unusual pattern of brain organization underlying the WS language capacities. Furthermore, the effect of the semantic anomaly is larger in WS than in the controls, which may be related to the tendency of WS subjects to generate low-frequency words in certain tasks (Bellugi et al., 1992; Karmiloff-Smith, 1992a).

WS children also show a reversal in the order of acquisition of certain linguistic markers compared to normal controls. For example, French-speaking WS subjects are proficient at many complex syntactic structures such as the passive, while being simultaneously very poor at grammatical gender (Karmiloff-Smith, Grant, & Berthoud, 1993). By contrast, WS subjects perform better than controls in a task involving short-term phonological memory, i.e., repetition of nonce terms obeying French phonotactics (Karmiloff-Smith et al., 1993). Taken together, these data suggest that the identity and/or organization of neural systems involved in the processing of certain aspects of language in WS may be different from that of the normally developing brain. A more lengthy discussion of the literature on WS linguistic and cognitive capacities across a number of domains can be found elsewhere (Karmiloff-Smith, 1992a; Bellugi & Morris, in press; for a corresponding discussion of normal development, see Karmiloff-Smith, 1992b,c).

From a behavioral viewpoint, WS subjects have been shown to have concentration difficulties, excessive anxiety, and poor relationships with peers (Udwin & Yule, 1991). By contrast, they often have good social relationships with adults whom they seek out for linguistic interaction. WS children seem to discover early that language is their area of proficiency, and they make extensive use of affective language in socially oriented ways (Reilly et al., 1991). Parental anecdotes and clinical reports suggest that, unlike individuals with autism, children and adolescents with WS are empathetic and very sensitive to others' emotional displays (perhaps even excessively so). This could, of course, merely be responsiveness to external *behavioral* signs. Of crucial importance for our present purposes is to determine whether WS children are sensitive to the *mental* states of others and whether they realize that others' behavior can be

predicted on the basis of their mental state, even when it differs from the objective state of the world as in the case of false belief.

Because of the good linguistic and face-processing capacities in WS subjects, and in line with the Brothers-Ring hypothesis, we predicted that all aspects of human interaction would capture the attention of WS children and be represented by them, and hence that they would perform well on a variety of theory-of-mind tasks.

## RESULTS

Full details of the population, methods and designs can be found in the final section of the paper. In this section, we present the results of six experiments (with, where necessary to understand the results, a very brief outline of the experiment), covering a battery of theory-of-mind tasks carried out with a total of 18 WS subjects. Statistical tests have been carried out only on the results of Experiment 1, which directly compared normal subjects and those with autism or WS. Experiments 2–6 rely on comparisons with the results of previous studies with children with autism and normal controls, the experiments with WS subjects having been run in separate studies. In cases where statistical analyses are not appropriate, we compare percentage success rates.

Experiment 1 required subjects to use direction of eye gaze depicted on faces in real photographs and in schematic drawings as a means of inferring the intentions and goals of others (Baron-Cohen, Campbell, Karmiloff-Smith, Grant, & Walker, in press; see the Methods section for methodological details). By 4 years of age, between 70 and 95% of normal children successfully use eye direction to infer a character's intentions and goals. Mental age-matched subjects with autism perform significantly worse than normal and mentally handicapped controls on conditions involving the use of eye gaze to infer intention. This turned out to be a selective deficit, because there were no significant differences between individuals with autism and controls on judgments of the emotions (sad/happy) displayed by the faces, nor on conditions in which nonsocial clues such as color, size, and direction of arrows were relevant.

Unlike individuals with autism, WS subjects tested on the same materials showed no such selective deficit. Not only did they score almost at ceiling on emotional and nonsocial clues, but they were indistinguishable from the normal controls and very significantly better than the group with autism on inferring intentions and goals on the basis of eye gaze direction (see Baron-Cohen et al., in press for full details of analyses). In fact, in many of the conditions, the WS subjects were 100% correct.

Experiments 2 and 3 concerned children's understanding of false belief (Perner, Leekam, & Wimmer, 1987; Wimmer & Perner, 1983; see the Methods section for methodological details). Very young normal children incorrectly predict a protagonist's behavior on the basis of

their own knowledge about the state of the world. By around 3½ to 4 years of age, however, children focus on the mental state of the protagonist and correctly predict his or her behavior on the basis of their false belief. Only some 20% of children with autism score successfully on such simple theory-of-mind tasks (Baron-Cohen et al., 1985; Leslie & Frith, 1988; Perner et al., 1989). By contrast, 94% of the WS subjects tested on these two tasks predicted successfully the protagonist's behavior on the basis of his or her mental state and used mental state verbs like *think* to explain the behavior (e.g., "He will look in that box because he thinks that it's still there."). In other words, unlike their deficits in number and spatial cognition, WS subjects show ability in the theory-of-mind domain.

The few subjects with autism (about 20%) who do succeed on the above first-order theory-of-mind tasks have been tested on second-order theory-of-mind tasks (Baron-Cohen, 1989b; Happé, 1991, 1993). These test the child's understanding of one protagonist's thoughts about another protagonist's thoughts. Experiments 4 and 5 tested this and involved two protagonists, A and B, who both actually know the state of the world, but A thinks that B has the wrong information (Perner & Wimmer, 1985; Sullivan, Zaitchik, & Tager-Flusberg, 1993; see the Methods section for methodological details). Unlike first-order tasks, in this case there are no false beliefs about the state of the world. Rather, A's false belief is about B's mental state, because A does not realize that B has updated information about the world. Subjects must therefore make second-order computations on the basis of A's false belief about B's belief (i.e., A thinks that B thinks . . .) and predict that A will act on the basis of a false belief, despite the fact that A does not actually hold a false belief.

Normal 4–6 year olds find this task difficult. By 7 years, however, children start to succeed more consistently, with stable success in this particular task at around 9–10 years of age (Perner & Wimmer, 1985). Easier versions of the second-order tasks are passed by somewhat younger normal subjects. In the case of autism, even high-functioning individuals fail second-order tasks (Baron-Cohen, 1989b; Happé, 1991, 1993). Of the 13 WS subjects tested on Experiment 4, 31% succeeded; all had a Verbal Test Age above 9 years, although their Performance Test Age was around 6 years. By contrast, 88% of the WS subjects tested on Experiment 5 succeeded. This is an easier second-order task than Experiment 4 because it uses the propositional attitude *KNOW* rather than *BELIEVE* (see the Methods section for details). This again reveals the WS capacity in the theory-of-mind domain, which is in sharp contrast to the deficit in even high-functioning subjects with autism with considerably higher IQs than the WS subjects. This suggests that successful performance calls on domain-specific capacities that are not solely dependent on general levels of intelligence.

Experiment 6 measured children's understanding of

metaphor and sarcasm (Happé, 1993; see the Methods section for methodological details). Metaphor requires a mapping between a subject's intended meaning and the real meaning of the expression (e.g. "your head's made of wood"), whereas sarcasm requires a higher-order understanding of the speaker's attitude to an internally represented thought rather than a statement (e.g. "now that's a clever thing to do," really meaning "stupid thing to do") (Sperber & Wilson, 1986; Happé, 1993). Normal subjects between 4½ and 5½ years of age succeed on both the metaphor and sarcasm conditions. Subjects with autism succeed on the metaphor condition if they are among the 20% who pass first-order theory-of-mind tasks, but even high-functioning individuals with autism fail on the sarcasm condition, because they interpret sarcastic statements literally (Happé, 1993). The WS subjects showed the opposite pattern. Fifty percent of them succeeded on both sarcasm and metaphor, but those who had difficulties either failed on both conditions or tended to find the more socially constrained sarcastic statements easier than the more cognitively constrained metaphoric statements. In other words, WS subjects did not show the same pattern of success and failure manifest in individuals with autism.

In sum, unlike their serious deficits in number, spatial cognition, and problem solving, WS subjects show a third islet of relatively preserved ability—together with language and face processing—that of theory of mind.

## GENERAL DISCUSSION

At first, our new results on WS capacities with theory-of-mind tasks, together with the previously attested proficiencies in language and face processing, would seem to be grist for the Brothers-Ring mill. WS subjects succeed in three socially relevant areas, where subjects with autism typically have difficulties. In face processing, although both individuals with WS or autism succeed on judging direction of eye gaze, only WS subjects perform successfully on tasks involving inferring intention from eye gaze. In language, our results showed that WS subjects tend to succeed at tasks involving sarcasm, whereas individuals with autism are better at those involving metaphor. Moreover, in the Introduction we referred to research showing a relationship between the processing of socially relevant information and intact limbic structures (Price, Russchen, & Amaral, 1987; Eslinger & Damasio, 1985), which is supported by the finding previously mentioned that limbic structures in the temporal lobe of the WS brain appear to be normal (Jernigan & Bellugi, 1994). We also noted the interesting fact that neocerebellar, but not paleocerebellar, lobules are significantly smaller in autism (Courchesne et al., 1988), whereas there is a striking increase in neocerebellar volumes in WS subjects (Jernigan & Bellugi, 1994; Bellugi, Wang, & Jernigan, 1994), i.e., the volumetric differences in WS seem to be the reverse of those found in autism.

A plausible conclusion, then, on both psychological and neurological grounds, is that socially relevant representations are processed within a broad cognitive module (Brothers & Ring, 1992), which is preserved in WS and impaired in autism. However, comparisons with other phenotypes challenge this conclusion and suggest a far more complex story than that of an impaired or intact module.

Indeed, although there are syndromes in which aspects of language, face processing, and theory of mind are either all spared (WS) or all impaired (autism), there are other phenotypes that point to dissociations between the three areas. For example, in Down syndrome a serious deficit in face processing and a rather serious deficit in the use of morphology in language can coexist with relatively good performance on theory-of-mind tasks (Baron-Cohen et al., 1985, 1986). By contrast, in hydrocephalus with associated myelomeningocele, exceedingly proficient language output (Cromer, 1992) can coexist with serious deficits in both face processing and many theory-of-mind tasks (Karmiloff-Smith, 1992a). Similarly, while in autism theory of mind and the pragmatics of language are impaired, syntax and face processing are relatively intact (Baron-Cohen, 1994). These different patterns of dissociation seem to challenge the Brothers-Ring hypothesis of a single, broad cognitive module for representing other individuals. However, such a challenge holds only if one thinks in terms of a *genetically* specified module including all the properties of a Fodorian module (Fodor, 1983), outlined in our introduction.

Taking a more *epigenetic* view, we offer the following speculation, which retains aspects of the Brothers-Ring original hypothesis but uses the comparison of different abnormal phenotypes to supplement it. We suggest that in normal development there are distinct, domain-specific, skeletal predispositions for discriminating stimuli relevant to language, face processing, and theory of mind (see discussions in Karmiloff-Smith, 1992b, for all three, and more specifically with respect to language, Kuhl, 1991, and Pettito, 1993; with respect to face processing, Johnson & Morton, 1991; and, with respect to theory of mind, Baron-Cohen, 1994, Baron-Cohen & Ring, 1994, and Leslie, 1987). With the massive early experience of superimposed inputs (i.e., face, voice, and human interaction all take place in a superimposed fashion), these predispositions gradually take over privileged circuits in the brain that become increasingly specialized and progressively interconnected. They are not necessarily spatially adjacent in the brain, but both their macro-development and, subsequently, their on-line processing may be closely related in time. Such temporal co-occurrences can give rise, as a result of development, to the emergence of specialized interconnecting circuitry. As development proceeds in the normal case, a process of modularization (a weaker and more constructive view than the Fodorian view of modules; see Karmiloff-Smith,

1992b for discussion) gives rise to the emergence of separate, modular-like organization for each subdomain relevant to theory of mind. Subsequently, we speculate that the computations relevant to theory-of-mind representations in each domain may with time give rise to an emergent superordinate modular-like organization for the pragmatics of social interaction in general, along the lines of the Brothers-Ring hypothesis.

The linguistic component of the broader organization would include all the communicative/pragmatic aspects of language, such as the understanding of sarcasm. In contrast, the syntactic and lexical components of language are unlikely to be part of such a developing module but separate ones. This would allow for the attested dissociations between different aspects of language in various syndromes and in normal adult brain damage. As far as is known, syntax, semantics, and pragmatics are each relatively spared in WS, although further research is under way to probe this in more depth (Karmiloff-Smith, Tyler, & Sims, unpublished experiments). If this turns out to be the case, then development in individuals with WS may give rise to the emergence of three submodules relevant to social cognition, as well as the modules for language and face processing not relevant to theory of mind, whereas those with autism may lack or have damaged the three submodules involved in social cognition.

The Brothers-Ring hypothesis predicts that in certain cases of adult brain damage, the module responsible for representing other individuals would be impaired in its entirety. By contrast, by retaining the notion of progressive emergence and modularization of each component part of the broader social domain, we hypothesize that any of the submodules of the latter can be selectively impaired in adult brain damage. We further speculate that in abnormal development, the domain-specific predispositions for language, for face processing, or for theory of mind are in some cases missing or impaired, such that the child lacks the head start on learning in one or another of these domains. In such cases, it is unlikely that any broad, overarching modular-like organization emerges, and thus any preserved part will be weaker than in subjects proficient in all three. We have seen above that this indeed holds for individuals with Down syndrome. By contrast, in normal development it is likely that each domain would strengthen the efficiency of the others as they progressively interact and share common emergent circuitry. Obviously further research is required to ascertain whether these are three submodules that share later levels of processing, or whether they come to form a single, broad emergent specialization via temporal co-occurrences as development proceeds. Further in-depth research should also clarify whether the broader module is one for representing socially relevant stimuli (the Brothers-Ring hypothesis) or one more specifically for social interaction.

The fact that these three islets of ability are preserved



in WS suggests that WS children may indeed progressively build up a broad socially relevant module, along the lines of the Brothers-Ring hypothesis (or one specifically for social interaction), that they use to bootstrap other parts of their deficient cognition. Such speculations about gradual modularization weaken Fodor's criteria by suggesting that the modular-like organization of the brain stems from skeletal domain-specific predispositions, and is neither hard-wired nor of fixed neural architecture. But progressive modularization does ultimately give rise to the fast, mandatory, automatic, and stimulus-driven behaviors that rapid theory-of-mind computations seem to involve. Indeed, with the exception of individuals with autism, most humans simply cannot help but compute the mental states of others. People do it almost 100% of their waking hours (indeed, as we write we of course cannot help wondering what our readers will think of our theory!). In sum, our speculation is that if modules such as a broad module for representing socially relevant stimuli exist, they are the *emergent product* of development, not its starting point.

## METHODS

An overall total of 18 WS subjects of chronological age between 9 and 23 years were tested, first on standardized tasks and then with a battery of 6 theory-of-mind tasks. The WISC-R gave an IQ range between 40 (floor) and 65 for these subjects (mean, 52.4; SD, 7.3). Language tests were also performed. Receptive vocabulary was assessed using the BPVS (British Picture Vocabulary Scale, Dunn et al., 1982) or the equivalent American/French Peabody, yielding a test age range between 5;3 and 10;7 (mean, 8;5; SD, 1;9). Comprehension was tested using the TROG (Test for Reception of Grammar, Bishop, 1983) or equivalent French test, yielding a test age range between 5;0 and 9;0 (mean, 6;5; SD, 1;1). Nonverbal scores were obtained from results on the Raven's Coloured Progressive Matrices (Raven, 1986), yielding a test age range between 3;6 and 8;6 (mean, 6;1; SD, 1;4).

Subjects were from middle class to lower middle class socioeconomic status. All were monolingual; 8 were English-speaking (British or American) and 10 were French-speaking. Due to illness and/or unavailability of subjects for second or third testing sessions, not all subjects were tested on all six experiments, but results were obtained from three subjects for all six experiments, from five subjects for five of the six experiments, from nine subjects for three of the six experiments, and from one subject for one of the experiments. In every case, subjects were tested across the full range of conditions for any particular experiment. The number of WS subjects in the different experiments ranged from 11 to 16, with experiment 5 having a subset of 7 subjects (see details below).

Subjects were tested individually, either in their

schools or their homes or, in the case of the American subjects, they were accompanied to the laboratory. All subjects were part of broader studies of overall cognitive and linguistic development in WS. The battery of theory-of-mind experiments was administered to the WS by the same (peripatetic!) scientist (AK-S). Standardized IQ, language, and other tests were run by each of the laboratories involved (Salk Institute for the American WS subjects and Medical Research Council for the British and French WS subjects). For the different tasks, experimental data were already available for young normal controls as well as for groups of autistic subjects of equivalent or higher IQ than the WS subjects.

Twelve WS subjects were tested on Experiment 1. The task comprised a series of conditions that explore subjects' capacity to use direction of eye gaze to infer a character's mental states such as ATTEND TO, DESIRE, GOAL, REFER, and THINK (see Baron-Cohen, Campbell, Karmiloff-Smith, Grant, & Walker, in press, for full details). Subjects were shown pairs of photographs of children's faces, comprising an equal number of male and female models of different ethnic groups. In the first two conditions, the eyes of one of the pair were looking directly at the camera (i.e., at the subject). In one condition, the other face looked away in profile and, in the other condition, the other photograph was full face but had the eyes averted. For each pair, the subject's task was to determine in which of the two photographs the child was looking at the subject. A third condition involved the same problem but the stimuli were two-line schematic cartoon faces instead of photographs. The fourth condition was designed to ensure that subjects were able to make judgments about faces when features *other* than eye gaze are important. This condition used pairs of schematic faces and questioned subjects as to which was the happy/sad face (depicted by different positions of the mouth). A fifth condition, again using schematic cartoon faces, had subjects point out the silly face (one in which the two eyes were in impossible positions with respect to one another), to ensure that they could attend to the details of the eyes on a basis other than direction of gaze. Next were a series of conditions using a central schematic face and four well-known candy wrappings (e.g., Polo, Mars, Milky Way, and Smarties, depending on the nationality of the subjects). These were the crucial theory-of-mind conditions in which we ascertained whether the child could use direction of eye gaze to infer, for instance, what a character desired (e.g. "Which candy does he want?") or had as a goal (e.g. "Which candy is he about to take?"). Finally, subjects were tested on their sensitivity to direction of eye gaze as a more natural clue to intentionality than, say, an arrow pointing to another wrapping. Several other, nonsocial conditions (reference to color, size, etc.) were used as further controls. Full details can be found elsewhere (Baron-Cohen et al., in press).

Fifteen WS subjects were tested on Experiment 2. This

is a typical theory-of-mind task used extensively with both normal children and those with autism (Wimmer & Perner, 1983; Baron-Cohen et al., 1985). There are two experimenters (E1 and E2). The subject is shown two boxes and the following scenario is acted out. E1 hides an object in location A, E2 leaves the room on some excuse, E1 moves the object to location B, and asks the subject three questions: "When E2 comes back, where will he look for the object?" "Where is the object really?" and "Where does E2 think it is?" The subject's task is to differentiate between the mental representation she or he has of the actual location of the object now (in location B) and the mental representation that E2 has of the object (in location A), and to predict that E2's behavior will be based on his or her false belief and not on the object's present location.

Experiment 3 again tests for subjects' capacity to use mental states to infer subsequent actions (Perner, Leekam, & Wimmer, 1987). Sixteen WS subjects were tested. The subject is shown a packet of M&Ms (or a local French/British equivalent) and asked what is inside. All subjects give the reply: "M&Ms." The experimenter then shows the subject that this packet does not actually contain M&Ms, but small pencils. The subject is then asked what another child, a classmate, who has not yet seen the contents, will reply when shown the packet. Once again the subject's task is (1) to differentiate between the mental representation she or he has of the actual contents of the packet (pencils) and the mental representation that the other child will have of its contents as a result of expectancies from world knowledge (candies), and (2) to predict that the other child's response will be based on her or his mental state (what she or he *thinks* it contains) and not on the actual contents known to the subject.

In Experiment 4, 14 WS subjects were tested on a second-order version of the theory-of-mind tasks (Perner & Wimmer, 1985). Subjects watch the following story being acted out with toy dolls and a street scene: "Mary and Bob are playing in the park. An ice-cream seller comes along. Bob decides to buy an ice-cream but hasn't got any money with him, so he tells the ice-cream seller that he will go home and get some money and return to buy an ice-cream. The seller assures him that he'll stay in the park until he gets back. Bob goes off to his house. Mary stays playing in the park. Business is poor, so the ice-cream seller tells Mary that he is not staying in the park after all, but will go to the church yard to sell his goods. He leaves. The subject is asked: "Where is Bob?" "Did Bob hear what the ice-cream man said?" Mary then goes home too. In the meantime, as Bob comes out of his house with his money, he sees the ice-cream seller drive by. The seller calls out to Bob to explain that he decided not to stay in the park after all and is going to the church yard to sell his ice-cream. The child is then asked: "Where is Mary? Does she know that Bob has met the ice-cream seller?" Bob walks off. Then Mary comes

out of her house and goes across to tell Bob what has happened. But he is not in and his mother tells Mary that Bob has gone to buy ice-cream. She doesn't tell her where Bob has gone, simply that he's "gone out to buy ice-cream." The subject is then asked the crucial second-order theory-of-mind question: "Where does Mary go to look for Bob?"

Unlike first-order theory-of-mind tasks, in this case there are no false beliefs about the state of the world. Both protagonists know where the ice-cream seller really is. Rather, the false belief is about another's belief state. Mary thinks that Bob thinks that the ice-cream man is in the park, i.e., that Bob holds a false belief, because Mary doesn't know about Bob's updated information. The subject must therefore make second-order computations on the basis of Mary's false belief about Bob's belief and predict that Mary will go to look for Bob in the park, despite the fact that they each separately know the correct location of the ice-cream man.

Experiment 5 also involved second-order false belief attribution, but was couched in a story using the mental state term *KNOW* (Sullivan, Zaitchik, & Tager-Flusberg, 1993), a much simpler propositional attitude than *BELIEVE*. It was used because normal 4-5-year-olds and many of the WS subjects tested on Experiment 4 had difficulties keeping track of the lengthy story and drawing the correct inferences. The simpler version, which became available only toward the end of the theory-of-mind testing sessions,<sup>2</sup> was used with a subset of 8 WS subjects, as follows: "There's this little boy called David and it's his birthday tomorrow. And guess what he wants? He wants a puppy. But Mummy says she's not going to buy him a puppy. She's going to buy him a toy. David is a little bit sad, because he really wants a puppy. David goes off to play. He goes down to the basement to get his bicycle and guess what he sees? In the corner, there's a tiny little puppy hidden in a basket. David is so pleased." The subject is then prompted with the following question: "Does Mummy know that David has seen the puppy?" The story continues: "While David is in the basement, upstairs Mummy hears the phone. It's grandma." "Hello. It's David's birthday tomorrow, isn't it? What did you buy him?", asks grandma. The subject is then asked: "What does mummy reply to grandma?" Then grandma asks two questions to which the subject must reply: "Does David know you bought him a puppy?" (second-order knowledge); "What does David think you have bought him?" (second-order belief). Finally, the experimenter checks again the child's knowledge by asking: "Does David really know what he is getting for his birthday?"

Experiment 6 involved attributing intentions to linguistic utterances. This task correlates highly with success and failures of subjects with autism on the above theory-of-mind tasks (Happé, 1991, 1993). Eleven WS subjects were presented with a series of stories involving metaphor and sarcasm. A typical story relates an

event in which a child did something silly (e.g., put whole eggs, with shell, into a cake mixture) after which one parent exclaims: "Your head is made of wood" (metaphor) and the other parent: "Now that's a clever thing to do" (sarcasm). The subject is asked to explain what the parent means when making such statements. When reflecting on metaphoric statements, the subject must differentiate between the real meaning and intended meaning. Sarcasm requires a higher order understanding of the speaker's particular attitude to an internally represented thought.

## Acknowledgments

We should like to thank Mark Johnson and an anonymous reviewer for comments on an earlier version. This research was supported by the Medical Research Council to the Cognitive Development Unit, U.K. and in part by NIH Grants HD26022, DC01289, and NS22343 to the Salk Institute for Biological Studies, San Diego.

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## Notes

1. This section on the biological/neurological profile of Williams syndrome is reproduced from Karmiloff-Smith (1992a) and updated on the basis of recent work by Bellugi and collaborators, Ewart and collaborators, and Galaburda and collaborators.
2. We should like to thank Helen Tager-Flusberg for making the puppy story available to us prior to publication.

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