Children with either Autism, Gilles de la Tourette syndrome or both: Mapping cognition to specific syndromes

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Children with either Autism, Gilles de la Tourette Syndrome or Both: Mapping Cognition to Specific Syndromes

Simon Baron-Cohen and Mary M. Robertson

Abstract

Three patients were tested: (1) A case of ‘pure’ autism; (2) A case of ‘pure’ Gilles de la Tourette Syndrome (GTS); and (3) a case comorbid for both autism and GTS. Based on previous studies, we predicted a ‘theory of mind’ deficit in the first case, an ‘intention editing’ deficit in the second, and a co-occurrence of these deficits in the third. Finally, we predicted that an ‘executive function’ deficit would not distinguish these patients, since they might be equally impaired in this. These predictions were supported. The results are discussed in terms of a ‘frontal’ theory to account for both pure and comorbid cases, and in terms of how such dissociations contribute evidence for the independence of these putative frontal systems.

Introduction

Autism is a severe childhood neuropsychiatric disorder, diagnosed on the basis of abnormal social, communicative and imaginative development (DSM-IV; APA, 1994). For reasons that are not yet understood, a proportion of children with autism also receive a diagnosis of Gilles de la Tourette Syndrome (GTS). GTS is usually a separate severe childhood neuropsychiatric disorder, diagnosed on the basis of multiple motor and vocal tics (DSM-IV; APA, 1994). Some studies suggest the rate of GTS in autism to be as high as 20% (Burder et al., 1987). Such comorbidity could occur for a number of reasons. First, it could reflect a referral or ascertainment bias, as most reports are from clinic populations (Berkson, 1946). Secondly, comorbidity could arise for genetic reasons (see Comings and Comings, 1990), and/or as a result of overlapping regions of brain damage. It is this possibility that we pursue here. Clearly, the two syndromes cannot be due to identical forms of brain damage, since many of the symptoms are non-overlapping. For example, autism involves social and communicative impairments of a type not seen in GTS. The two syndromes do however overlap in terms of frontal lobe signs, as expressed by deficits on cognitive tests of ‘executive function’ (Bornstein, 1990, 1991; Bishop, 1992; Ozonoff, 1994). Executive function is defined as flexible and planful shifting of attention (e.g. Shallice, 1988). One possibility, then, is that autism and GTS are independent syndromes, involving abnormalities in the development of different but overlapping frontal systems, which have a significant probability of both being disrupted together. Such a theory has the power to explain why both ‘pure’ and ‘mixed’ cases should exist.

In this paper, this theory is explored in relation to three relevant cases: a child with autism alone, a child with GTS alone, and a child with diagnoses of both autism and GTS. Our interest was in predicting which forms of cognitive deficit each patient might display. From previous studies, we expected that the child with autism alone would show severe deficits in the development of a ‘theory of mind’ (Baron-Cohen et al., 1985; Baron-Cohen, 1995), that is, in the ability to attribute mental states to people, as a way of making sense of and predicting action. Equally, from previous studies, we expected that the child with GTS alone would show deficits in ‘intention editing’ (Baron-Cohen et al., 1994a,b), that is, in the ability to inhibit one of two simultaneously competing intentions. We next predicted that only in the case of the child with both autism and GTS would we see the co-occurrence of theory of mind deficit and an intention-editing deficit. Finally, whilst the above predictions were expected to differentiate these three cases, we expected that on a test of executive function, all three cases should be equally impaired. This latter prediction was made on the grounds that a ‘dysexecutive syndrome’ [as Baddeley and Wilson (1988) call it] does not occur in any one psychiatric syndrome (see Baron-Cohen and Moriarty, 1995).

Regarding a possible brain basis to these deficits, some limited speculation is possible. First, all three cognitive domains are thought to involve different frontal areas. Thus, there is some evidence from single photon emission tomography (SPECT) and positron emission tomography (PET) functional neuroimaging studies that theory of mind may involve either the orbito-frontal cortex (OFC) or medial–frontal cortex (MFC) (Baron-Cohen et al., 1994;
P. Fletcher et al., unpublished material; V. Goel et al., unpublished material). [This may be part of a wider brain circuit involving the superior temporal sulcus and the amygdala (see Brothers, 1990; Baron-Cohen and Ring, 1994).] Secondly, regarding the brain basis of intention editing, the assumption is that this involves the anterior cingulate (and striatal pathways), since these appear to be abnormal in GTS (Moriarty et al., 1993). Finally, regarding the brain basis of executive function, SPECT and PET functional neuroimaging studies suggest that the dorsolateral prefrontal cortex (DLPFC) plays an important role. These different predictions and their relation to each psychiatric diagnosis and its putative brain basis are shown in Fig. 1. In terms of the theory of pure and mixed cases, we relate these to cognition using a Venn diagram in Fig. 2.

In sum, we regarded these three case studies as valuable 'experiments of Nature' in that they afford us the opportunity to test the relationship between psychiatric syndrome and cognitive profile. The design employed here was to give tests of theory of mind, intention editing and executive function to all three patients, in order to test the predictions outlined above.

**Experimental investigations**

**Patients**

We tested three patients, closely matched for sex, chronological age and verbal mental age.

**Patient 1**

JA had a diagnosis of autism but no GTS, using established criteria (DSM-III-R: APA, 1987). He was 13:6 years old, with a verbal mental age (MA), using the British Picture Vocabulary Scales (BPVS) (Dunn et al., 1982) of 9:9 years. He was attending a special school for children with autism in London.

**Patient 2**

ZB had a diagnosis of GTS but no autism. He had been diagnosed using DSM-III-R criteria by M.M.R. at the National Hospital, Queen Square, London. He was attending a mainstream school, though he was receiving remedial help for educational difficulties. He was aged 13:1 years, and had a verbal (BPVS) MA of 9:0 years. He showed facial tics during the test session, and had some vocal tics (without content).

**Patient 3**

DF had received diagnoses of autism and GTS, using DSM-III-R criteria. At the time of testing, he was 13:7 years old, with a verbal MA (BPVS) of 9:0 years. He was attending a college for adolescents with learning difficulties in the Birmingham area (UK). During the testing session, he showed facial and other motor tics, and reported that he was trying hard to control his vocal tics. He also reported getting into trouble at school for involuntary shouting of a coprolalic nature.

<table>
<thead>
<tr>
<th>Theory of Mind</th>
<th>Intention Editing</th>
<th>Executive Function</th>
</tr>
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<tbody>
<tr>
<td>Autism</td>
<td>x</td>
<td>√</td>
</tr>
<tr>
<td>GTS</td>
<td>√</td>
<td>x</td>
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<tr>
<td>Autism + GTS</td>
<td>x</td>
<td>x</td>
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<tr>
<td>Brain basis?</td>
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|x = impaired; √ = intact.

Brain areas: OFC = orbito-frontal cortex; MFC = medial-frontal cortex; DLPFC = dorsolateral prefrontal cortex.

See text.

**Fig. 1. Predictions regarding cognition in the three psychiatric categories**

![Venn diagram depiction of the postulated relations between pure and mixed cases of autism and GTS, and cognitive deficits.](image)


No aetiological factors were evident in patients 1 and 2, though in patient 3 there was a history of tremor, and a family history of epilepsy.

**Method**

Each child was tested individually in a quiet room either at school (patient 1), at the clinic (patient 2) or at home (patient 3). They were each given three tests of theory of mind, three tests of intention editing and a control test of executive function.

The theory of mind tests were (1) The Sally-Anne False Belief Test (Unexpected Transfer) (Baron-Cohen et al., 1985), which uses a puppet story and tests the subject's ability to attribute a false belief to a character, whilst the subject him/herself holds a true belief about the same situation; (2) The Smarties False Belief Test (Unexpected Contents) (Perner et al., 1989); this is similar to the previous one, but the content of the belief is different; and (3) The Penny Hiding Game (Baron-Cohen, 1992), which uses tests whether the subject can produce deception in a hiding game. Scoring of all of these tests exactly followed the original protocols, to which the reader is referred for a full description of the methods.

The intention editing tests were (1) The Yes and No Game (Baron-Cohen et al., 1994), in which the subject has
to answer questions without using the words ‘Yes’ or ‘No’;
(2) The Hand Alternation Task (Baron-Cohen et al., 1994b). This is a task of Luria’s (1966), in which the subject has to produce fluent simultaneous switching of hand positions (open versus closed); and (3) the Children’s Stroop Test (Gerstadt et al., 1994). This is an adaptation of the classic Stroop test (Stroop, 1935), modified for use with pre-literate children, in which the subject sees a picture of the night sky and has to say ‘Day’, and vice versa. Again, scoring and methodological details were based on the original descriptions, to which the reader should refer.

Finally, the control task of executive function was the FAS test (Milner, 1964), in which the subject has to produce as many words beginning with the letter F (then A, then S) as they can, in a minute. This test is a measure of executive function as it involves generativity and flexible shifting. Patients with frontal lobe damage typically show perseveration on this task (Shallice, 1988). As before, scoring and methodological details were based on the original descriptions, to which the reader should refer.

**Scoring**

Two points were awarded if the subject passed on both of two trials of each theory of mind test, and on both of two trials of each intention editing test. One point was awarded if they passed on only one trial. Passing was defined using the published criteria in the original articles. On the executive function test, the number of words produced by a subject on each of the three trials (F-A-S) was averaged.

**Results**

Table 1 shows the results from the battery of tasks. As can be seen, on the theory of mind tasks, patient 2 (with GTS alone) performed significantly better than the other two patients. On the intention editing tests, patient 1 (with autism alone) performed significantly better than the other two patients. Finally, on the executive function test, the three patients did not differ from one another, all of them producing relatively few words in one minute. Note that the deficits were marked, but not all or none in every case. Thus, patient 2 passed on one trial of the Stroop, and patient 3 performed the hand-alternation task perfectly. On deception in the penny-hiding game on one trial.

Significance is not tested statistically, since these are not group comparisons. However, visual inspection of the scores is sufficient to see the marked differences between the three cases.

**Discussion**

In this paper we tested the relationship between psychiatric diagnosis and cognition in three patients: a case of autism alone, a case of GTS alone, and a case comorbid for autism and GTS. As predicted, we found that the patient with autism alone was significantly impaired on theory of mind but not intention editing, the patient with GTS alone showed the opposite pattern, and the patient comorbid for both syndromes was impaired in both of these domains. On tests of executive function, all three patients were equally impaired.

These results are consistent with the notion that (1) the three processes are independent; (2) autism involves a theory of mind impairment whilst GTS involves an intention editing impairment; (3) mixed cases of autism and GTS show both deficits; and (4) an associated (frontal) cognitive process (executive function) is impaired in both psychiatric disorders. These results throw doubt on the idea that theory of mind necessarily depends on executive function (Russell, 1995), since in patient 2 (with GTS), theory of mind was intact whilst executive function was relatively impaired. Naturally, these results should be treated with caution since they have the inevitable limitations of single case studies and because there are as yet no documented cases of intact executive function with impaired theory of mind. Nevertheless, in supporting highly specific predictions from cognitive neuropsychiatry, they may indicate the potential for future research in this area to map syndromes and cognition.

**Acknowledgements**

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**References**


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Number of Neurocases: 20

Table 1

<table>
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<tr>
<th>Primary diagnosis of interest</th>
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<td>DF</td>
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</table>

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Three patients were tested: (1) A case of 'pure' autism; (2) A case of 'pure' Gilles de la Tourette Syndrome (GTS); and (3) a case comorbid for both autism and GTS. Based on previous studies, we predicted a 'theory of mind' deficit in the first case, an 'intention editing' deficit in the second, and a co-occurrence of these deficits in the third. Finally, we predicted that an 'executive function' deficit would not distinguish these patients, since they might be equally impaired in this. These predictions were supported. The results are discussed in terms of a 'frontal' theory to account for both pure and comorbid cases, and in terms of how such dissociations contribute evidence for the independence of these putative frontal systems.

Journal

Neurocase 1995; 1: 101-4

Neurocase Reference Number:

07

Primary diagnosis of interest

1. Autism

2. Gilles de la Tourette Syndrome

3. Autism and Gilles de la Tourette Syndrome

Author's designation of case

1. JA

2. ZB

3. DF

Key theoretical issue

- Theory of mind does not necessarily depend on executive function

Key words: autism; Gilles de la Tourette syndrome; executive function; theory of mind

Scan, EEG and other related measures

Not known

Standardized assessment

Sally-Anne False Belief Test; Smar test; Penny Hiding Game; Yes and No Game; Hand Alternation Task; Children's Stroop Test; FAS test.

Other assessment

Nil

Lesion location

- Not known

Lesion type

- Not known

Language

English