Cognition in Tourette Syndrome: Cognitive Characteristics Associated with Tourette Syndrome, their Neurobiological Bases, & Management Strategies

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Cognition in Tourette Syndrome

Why might Tourette Syndrome imply a distinct cognitive profile?

- The ‘unvoluntary’ nature of tics
- Premonitory urges (PMUs)

Cognition in Tourette Syndrome

Executive Function (EF): A set of neurocognitive processes that help regulate overt and covert behavior (Lezak, 1995). EF is essential for goal-directed action and cognition, and problem solving (Anderson, 2002; Zelazo et al., 2008).

Social cognition: The thought processes that are used in understanding others (Flavell & Miller, 1998). It includes abilities such as emotion recognition in facial and verbal expression, non-literal language comprehension, and social situational problem-solving.

Executive Function

Updating of Working Memory

Inhibitory Control (or Response Inhibition)

Cognitive Flexibility (or Set-Switching)

Working Memory Updating

S

R

Z

U

X

I

W

Stroop Task

When each word appears, state the color the word is typed in:

RED

BLUE

GREEN

YELLOW

BLUE

RED

BLACK

GREEN

YELLOW

ORANGE

‘Letter-Number Task’

When the letter-number pair appear at the top of your screen, then if the letter is a vowel, say ‘Quack’, and if it is a consonant, say ‘Moo’.

When the letter-number pair appear on the bottom of your screen, then if the number is odd, say ‘Quack’, and if the number is even, say ‘Moo’.

B7

T4

09

X8

A3

G5

Social Cognition

- ‘Theory of Mind’ (ToM)

  Jenny put her chocolate away in the cupboard and went outside. Alan moved the chocolate from the cupboard into the fridge. Half an hour later Jenny came back inside.

  Where will Jenny look for her chocolate?

- Empathy

  Visual/Auditory emotion recognition

- Social Problem Solving

Objective

Is ‘pure’ Tourette syndrome, or TS without cooccurring ADHD or OCD, associated with distinct performance characteristics on measures of executive function and social cognition?

Is there a relationship between these cognitive characteristics and symptom severity in individuals with pure TS?

Results

23 studies addressed executive function in children/young adolescents: Only one of these indicates performance deficits for the pure TS group collectively (Crawford et al., 2005).

21 studies that addressed EF in adult populations: Twelve of these report performance deficits associated with pure TS.

These disparate results may relate to the developmental course of TS during the lifespan, and the likelihood that individuals whose TS persists into adulthood may have a more severe form of TS.

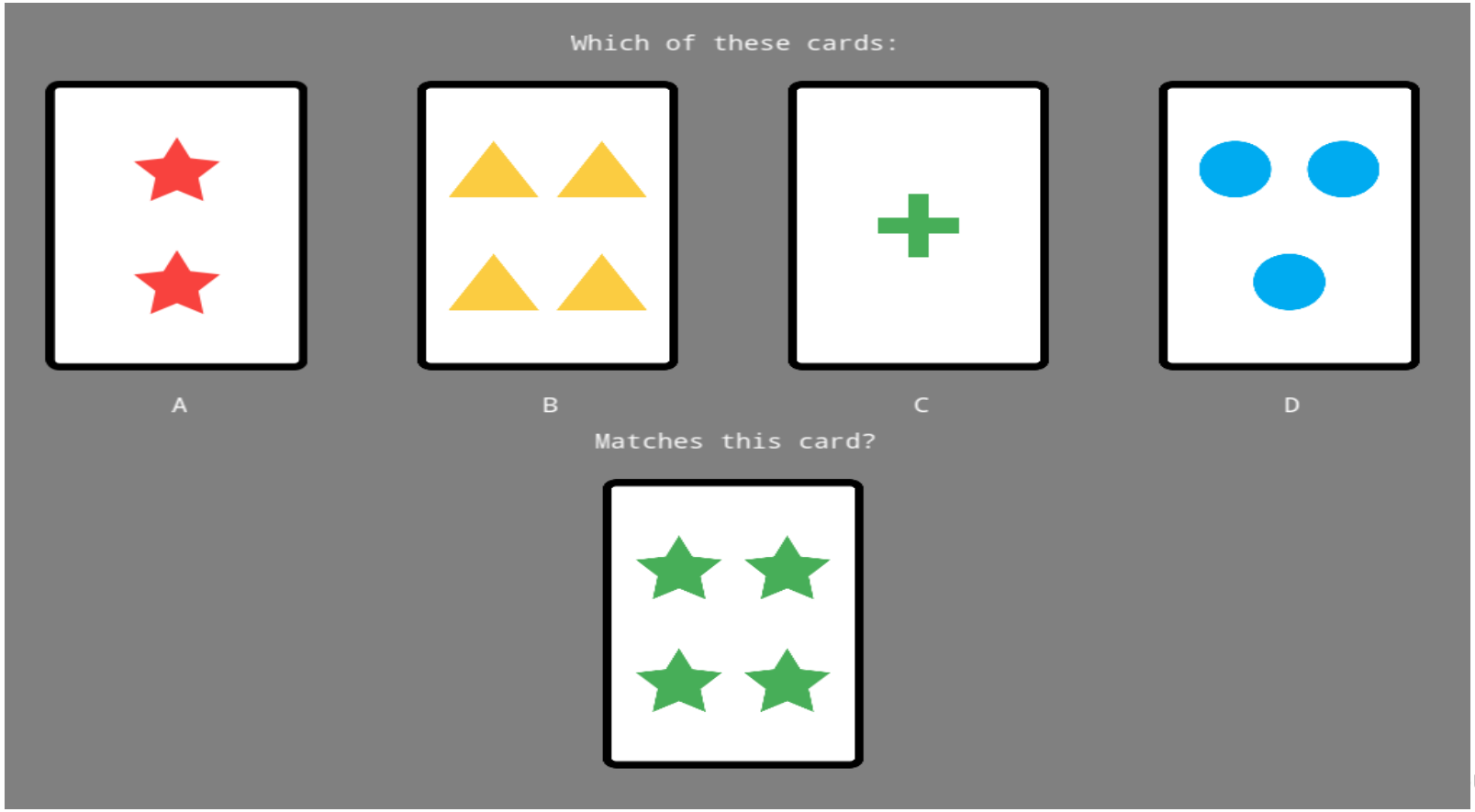
Results

‘Hayling Test’

“The captain wanted to stay with the sinking _____”

Results

‘Wisconsin Card-Sorting Test’



Results

- Social Faux-pas

  Jill had just moved into a new apartment and bought some new curtains for her bedroom. When she had just finished decorating, her best friend, Lisa, came over. Jill asked her, "How do you like my bedroom?" "Those curtains are horrible," Lisa said. "I hope you're going to get some new ones!"

- Indirect Sarcasm

  Alex takes two completely burned pieces of toast from a toaster. Mary says to Alex: “You’re the best cook in the world!” [direct sarcasm] or “I’ll hire you in my restaurant” [indirect sarcasm].

- Conflicting Emotion

Results

Several studies have reported a positive correlation between deficits on EF tasks, and tic severity (Baym et al., 2008; Cavanna et al., 2009; Channon et al., 2006; Delorme et al., 2016a; Eddy et al., 2014; Jeter et el., 2015; Jung et al., 2015), even when a similar relationship was not observed between EF variables and the TS group as a whole.

Neurological Profile of TS

- Functional differences in neural networks are implicated in TS & its symptoms.
- basal ganglia (& striatum), thalamus, anterior cingulate cortex (ACC), dorsolateral and ventromedial prefrontal cortex (DL-PFC, VM-PFC)
- Dysfunction in the production & processing of the neurotransmitters dopamine & GABA

Neurological Profile of TS

- Dopamine plays a central role in reward based learning and habit formation

- GABA is involved in the inhibition of neural signaling

Neurological Profile of TS

Dopamine

- Related to tic production (which possess habit-like qualities) (Beste & Munchau, 2018; Conceicao et al., 2017; Delorme et al., 2016)
- Increased activation in brain areas involved in dopamine production predicts tic severity (Baym et al., 2008; Conceicao et al., 2017)
- A proposed mechanism is that the performance of tics alleviates PMUs, a cycle that results in tics becoming a learned behavior (Brandt et al., 2016; Conceicao et al., 2017)
- Ultimately, *increased levels of dopamine lead to enhanced 'perception-action binding'*

Neurological Profile

GABA

- Reduction in GABA transmission in individuals with TS (e.g. Ganos et al., 2013; Puts et al., 2015) is thought to result in the disinhibition of areas related to motor learning and action selection
- GABA reduction found to produce tic-like behaviors in animals (Jackson et al., 2015) and in children with TS (Puts et al., 2015)
- GABA reduction is associated with higher levels of tic severity (Jackson et al., 2015; Puts et al., 2015)
- Ultimately, *decreased GABA levels result in 'disinhibition', which may be experienced as heightened arousal, or 'neural noise'*

What Neurological Study can Teach us about Cognition in TS

Key neurophysiological characteristics of Tourette Syndrome: strengthened stimulus-response pairings induced by dopaminergic activity, and increased neural excitation and arousal due to a decrease in GABA-related activity, would predict that tasks involving a sufficient level of processing in the context of interference from the elicitation of a strong automatic response would pose a greater degree of difficulty for individuals with TS.

What Neurological Study can Teach us about Cognition in TS

Measures of Executive Function

Hayling Test, Wisconsin Card Sort Test

Measures of Social Cognition

'Faux-pas', indirect sarcasm, perspective taking & emotion recognition with 'emotionally charged' stimuli

What Neurological Study can Teach us about Cognition in TS

The frontocortical-striatal network is also activated in response to anticipated stimuli, which may explain some distinct cognitive traits that are observed in TS in the absence of these conditions:

- Tendency to rate social interactions as ‘more awkward’
- Tendency to ascribe intentions to neutral stimuli
- Link between metacognitions about tics and their severity
- Altered ACC activity for predictable stimuli (musical chords)
- Urge to perform improper actions or verbalizations (e.g. corprolalia)

Management Strategies

Awareness & Acceptance

Mindfulness

Reflection/Verbalization

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