Neuropsychology and Tourette Syndrome

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History

- Until 2000s
  - small Ns
  - lack of control data
  - little consideration of co-occurring disorders
Brain Pathways

- MRI research (Peterson et al, 2003; Raz et al, 2009; Miller et al, 2010; Tobe et al, 2009)
- Dopamine overproduction (neuroleptics)
- DBS
- Premonitory Urge
- Motor circuitry

Cortico-striatal-thalamo-cortical loop

Osman and Smerz, 2005
Studies 1978-2011 suggest no significant GLd
Brand et al. (2002) reported that 23 children with TS-only scored higher IQ than 17 TS+ADHD children
Schuerholz (1996) & Hagin (1982) mean IQ of a TS-only group 1SD > than controls
Huckleba et al. (2008) reported low IQ in N= 47 associated with weak numeracy
Debes (2011) N= 266 (TS only- 125) VIQ = 93, PIQ=87
  - Each year the tics emerged later, PIQ was >1 point increase
  - TS + OCD FSIQ >TS+ ADHD + OCD FSIQ group.
  - No effects of medication or tic severity (YGTSS)
  - TS-only < Controls only differed on BD
  - TS+ADHD < Control on DS

Neuropsychological testing battery including *Purdue Pegboard Test*.

Follow-up 7.5 years later

Weaker performance with the dominant hand on Pegboard predicted worse adulthood tic severity

Weak performance bilaterally on Pegboard predicted worse psychosocial functioning
**Rule governed language (Walenski et al., 2007)**
- Processing of predictable procedurally based regular verb conjugation (e.g. *slip-slipped*) more fluent in TS than controls

**Speech dysfluencies (De Nil LF et al., 2005)**
- Normal (conversational) but not atypical (objectively measured) dysfluencies were more frequent but were present in relatives and self-perceived reading difficulties

**Higher Level Language Skills**
- Legg et al, 2005 TLC - Disorganized output, concreteness and poor language formulation abilities

**Parental Vocab deficits**
- Casey et al (2000)
Executive Function

- **Deficits**
  - Pure TS < Controls on inhibition and strategy, TS+ADHD < Controls on all executive functioning measures (inhibition and strategy use, multitasking, rule following, and set shifting), TS + OCD not different to Controls (Channon, Pratt, & Robertson, 2003, Crawford, Channon, & Robertson, 2005).
  - Use of the BRIEF suggested deficits with WM but not other areas (unless co-occurring ADHD) with little correlation between experimental and self-report measures (Mahone et al., 2002)
  - CANTAB - deficits present especially in older children (Rasmussen et al, 2009; Bornstein et al., 1985)
Is Tic Suppression an Advantage?
Enhanced Cognitive Control

- Osmon and Smerz (2005) Overview
  - Oculomotor switching task greater cognitive control over movements.
  - Sub-cortical locus for the triggering of tics
  - Go/Nogo task
  - Go/Nogo task
  - Omission errors problematic not commission errors
  - Time estimation stronger in children with TS only
  - WM changes in individuals with TS
  - Increased diffusivity: particularly in the corpus callosum and forceps minor
  - Motor-response tasks
What about Neural Correlates?

- Cause or effect? (Jackson et al 2011)
- Continued Inhibition of tics leads to compensatory self-regulation mechanisms
- How can we use this cognitive strength therapeutically?
Causal Model of Tics?

**Genetic Brain Abnormality**

**Inhibition**

**Tic (Suppression) Disorder**

Single Strength Model?
Comorbidity
Prevalence

- ADHD
  - Deficits with Inattention, Hyperactivity and Impulsivity
    - TD - 50-55% cases (Freeman et al, 2007; Rothenberger, 1991)
    - ADHD – 20% have TDs (Gillberg et al, 2004)

- OCD
  - Repeated, unpleasant thoughts or images, repetitive, distressing rituals
    - TD – 30-50% cases (Pauls et al, 1991)
    - OCD – 10-30% cases (Holzer et al, 1994)
## Specific Learning Difficulties

<table>
<thead>
<tr>
<th>SpLD</th>
<th>Estimated Percentage</th>
<th>Cited by</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyslexia</td>
<td>4-8</td>
<td>Snowling, 2009</td>
</tr>
<tr>
<td>Dysgraphia</td>
<td>7</td>
<td>Brook and Boaz, 2005</td>
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<tr>
<td>Dyscalculia</td>
<td>4-6</td>
<td>Butterworth, 2010</td>
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<tr>
<td>ADHD</td>
<td>3-6</td>
<td>Simonoff, 2008</td>
</tr>
<tr>
<td>SLI</td>
<td>7</td>
<td>Bishop, 2011</td>
</tr>
<tr>
<td>Presence of a SpLD</td>
<td>18</td>
<td>Brook and Boaz, 2005</td>
</tr>
</tbody>
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http://www.bis.gov.uk/assets/biscore/corporate/migratedD/ec_group/101-08-FO_on

Need to remember co-existence of many conditions
Academic Learning Difficulties

- **Rates vary**
  - 51% - Burd et al (1991)
  - 23% - Schuerholz et al (1996)
  - 23% - Burd et al. (2005)
- More frequently males, onset before age 8, perinatal problems, > likely to have coprophenomenon.
- Family history was less frequent; other co-morbidities were significantly more frequent, particularly ADHD
- Arithmetic deficiencies Huckeba et al. (2008)
  - ADHD accounted for most of the differences
Prevalence - Clinic at GOSH

- 2006-2011 ADHD/OCD/ASD (HIV, epilepsy..)
- 19/46 more than 1.5 SD between Indices (broader Average range)
- 4 Statement of SEN
- 9 IEP
- Most fulfil criteria for more than 1 SpLD

Cognitive Function
Comorbidity
Specific LDs

SpLD

- Dysexecutive
- SLI
- Dyslexia
- Dyspraxia
- Dysgraphia
- GLD
- Poor Comprehenders
How to Screen carefully for SpLD

- Developmental History
- School reports
- Caution of relying on referral to EP
- Questionnaires are useful
- Standardised tests
- Increased risk for SLd if ADHD present (primary or secondary)
Deficits Associated with ADHD?

Both ADHD and TS
- Childhood onset
- Excessive motor activity
- Identified abnormalities in the cortical-striatal-thalamo-cortical circuit
  - Reduced caudate volumes (Peterson et al, 2003)
  - Thinning of sensorimotor cortices (Sowell et al 2008)
  - Reduced cortical inhibition in motor circuitry (Orth et al, 2009)
Case study - Recommendations

- Time accommodations / Reader / Scribe
- Speech recognition software (Dragon 12)
- Bibliotherapy on SpLD (Snowling, Muter)
- Morphemes for spelling (Nunes & Bryant, 2006)
- Reciprocal reading strategies (Oczkillo, 2003)
- Organisation of written work (www.inspiration.com/kidspiration)
- Working memory intervention (Cogmed)
- Strategies for tics
- Chang et al, 2007
  - Spatial attention deficits in OCD group not TS
- Bloch et al, 2006
  - Higher IQ
- Gresberg and McKay, 2003
  - Visual Spatial deficits
• Time accommodations with study skills of how to use them
• Referral for CBT for OCD
• Graded approach with feedback
• Use of visual timetables, prompts from teacher
• Mentor for problem-solving
• Education for teachers (primary and secondary)
• Availability of visual and verbal information
Conclusion
• How to distinguish multiple SpLDs when they co-exist with TS
• How to figure out what the worst problem is?
Motor tics can interrupt processes such as writing, reading

Absenteeism from school can compromise progress

Tic suppression – brain / behaviour changes - cause or effect? Who will respond to behavioural treatments

Informing Educational supports

Prognosis (Fine motor skills)

Screen for co-morbidity (Behaviour vs Cognition)

Experimental and ecological measures
Queries, questions, comments?