Social and cognitive development in children with Down syndrome: trying to build wider developmental models?

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Keynote Talk to 8th ECIDD
19th June 2010

Support from ESRC, Baily Thomas Trust, NIH, Wellcome Trust, Eranda foundation

IRCD

- Based in School of Psychology at Stratford
- Purpose built Neuro-behavioural & baby labs
- Attention- eye tracking equipment ASL & Tobii
- EEG/ ERP 128-channel
Three parts to talk

- Why we need to understand the relationship between social and cognitive development in infants with DS
- Why we need to develop theories
- How to notate and portray development

What is this?
My starting point (a long time ago...)

• Are there aspects of social perception that operate independently of processes that determine IQ?

• Is there evidence that these are relatively spared in children with MLDs?

• How do the social and cognitive domains relate to each other?

From Allison et al (2000) TICS

Information processing & ID

Simple effects:
*p<0.05
**p<0.01

Social perception


Spared abilities in children with MID?

What of other developmental difficulties

- Do people with general IDs have emotion recognition difficulties with other emotional stimuli?

- Are these over and above developmental level (MA)?

Review of 21 studies

- most employed static stimuli
- only 12 used mental-age matched controls
- of these only 5 had a control task
- only one of these found evidence for a specific emotion recognition problem
- this involved complex cross-modal matching

### Demands of different types of facial emotion recognition tasks

<table>
<thead>
<tr>
<th></th>
<th>Labeling</th>
<th>Identification/Discrimination</th>
<th>Within-mode matching</th>
<th>Cross-modal matching</th>
<th>Rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hold in mind goal</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Identify emotion in more than one stimulus</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Identify emotion in more than one modality</td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td>Employ verbal response</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Make non-categorical judgement</td>
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</table>


“...there is a danger of creating a setting in which one participant's intuitive emotional sensitivity might confer little advantage over another participant's [...] cognitively effective classification abilities” (Hobson, 1991).


Important trends in the data

• Performance worsens with number of stimuli

• Performance worsens when doing labelling tasks or cross-model matching

• Performance very poor when rating ambiguous and neutral stimuli

• Performance worsens with age
“to trace back the ontogenesis of complex social behaviour requires a focus on developmental mechanisms, not static lesions or deficits”

Pollock (2006)

What do these trends suggest?

• Is there a developmental element to this that is missing?

• We need to begin to follow processes from early in development

• We need to consider the relationship between social and cognitive processes as part of an explanation of developmental difficulties

• Need begin to study this in children with IDs as early as possible

• Very few IDs identified in first year but…
Children with DS (Trisomony 21)  
**Behavioural phenotype**

**Key cognitive outcomes**

- General intellectual delays
- Different attentional profiles
- Delays in motor development
- Selective deficits in short term memory
- Relatively slower onset of speech
- Selective problems in expressive language and syntax

But also specific problems with aspects of social communication

See Chapman & Heskith 2000; Fidler, 2005

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**Neuro-pathology**

- Reduced neural proliferation
- Lower density in all cortical layers and reduced inter-neurons
- Variable myelination
- Reduced dendritic arbors and postsynaptic spines
- Spines abnormal
- Reduced synaptic density
- Reduction of brain volume

- See Capone 2001 for gene-brain relationship in DS
Social phenotype

- Stereotyped as sociable and warm
- Good quality of relationships

But...

- There are subtle differences…

DS longitudinal study

<table>
<thead>
<tr>
<th>Time</th>
<th>DS</th>
<th>TD</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 months</td>
<td>10</td>
<td>22</td>
</tr>
<tr>
<td>6 months</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td>7 months</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>10 months</td>
<td>26</td>
<td></td>
</tr>
</tbody>
</table>

ESRC grant R000236722
Problems with using Bayley scales for matching

- performance not independent of motor abilities
- requires a level of social engagement

We excluded items if...

- The motor demands were likely to compromise performance.
- The item was on the social facet.
- The item was on the language facet.
- Items had previously been found to be unreliable in children with Down’s syndrome (based on Wright 1998)

The still-face paradigm

Tronick, Als, Adamson, Wise & Brazelton (1978)

phase 1: 180 secs of face-to-face mother-infant interaction
phase 2: 90 secs (max) where mother holds a ‘still-face’
phase 3: 180 secs of face-to-face mother-infant interaction

Video


Still face responses appear typical...

• It seems that infants with DS have a similar propensity to engage with others and have intact ‘primary intersubjectivity’

But…

• is their social behaviour and environment typical?

There are subtle differences in emotional responding

Differential environment?

• Mother–infant interaction may have a different quality.

See also
Adamson et al 2009; Buckhalt, Rutherford, & Goldberg, 1978; Cielinski, Vaughn, Seifer, & Contreras, 1995; Legerste, Varghese, & van Beek, 2002

Warmth and directiveness

<table>
<thead>
<tr>
<th>Phase</th>
<th>Maternal ratings†</th>
<th>Interaction Mean (SD)</th>
<th>Re-engage Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>DS 4.1 (1.0)</td>
<td>4.0 (1.1)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>TD 3.8 (1.2)</td>
<td>3.6 (1.1)</td>
</tr>
<tr>
<td>Directiveness</td>
<td></td>
<td>DS 4.3 (1.8)</td>
<td>4.5 (.7)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>TD 3.6 (1.2)</td>
<td>3.5 (1.1)</td>
</tr>
</tbody>
</table>

†Group main effect F(1,27) = 5.31, p = .03, partial Eta-squared= .16
* p<.05, ** p<.01, 1-tailed

Different relationship to infant behaviours

<table>
<thead>
<tr>
<th>Maternal ratings†</th>
<th>Interaction</th>
<th>Re-engage</th>
<th>Correlations with infant behaviour in initial interaction (Spearman’s rho)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean (SD)</td>
<td>Mean (SD)</td>
<td>%looking</td>
</tr>
<tr>
<td>Warmth</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DS</td>
<td>4.1 (1.0)</td>
<td>4.0 (1.1)</td>
<td>.46</td>
</tr>
<tr>
<td>TD</td>
<td>3.8 (1.2)</td>
<td>3.6 (1.1)</td>
<td>.46*</td>
</tr>
<tr>
<td>Directiveness</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DS</td>
<td>4.3 (1.8)</td>
<td>4.5 (1.7)</td>
<td>-.55*</td>
</tr>
<tr>
<td>TD</td>
<td>3.6 (1.2)</td>
<td>3.5 (1.1)</td>
<td>.07</td>
</tr>
</tbody>
</table>


- Mothers seem to be working harder to maintain attention
- This is not necessarily negative at this age but may have later effects
- What do we really know about the transaction between cognitive capacities and the social environments of people with DS
Cebula, Moore & Wishart (2010)

- There are relatively few studies of DS that attempt to relate social abilities and core cognitive difficulties in a developmental framework.

- Reviewed literature on social cognition in people with DS
- While there are some excellent studies of social abilities in DS there are large gaps in knowledge
- Tend to focus on specific aspects of social or cognitive but not theory driven
- Is DS poorly served by theoreticians?


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Is there enough research?

<table>
<thead>
<tr>
<th>Google Scholar search term</th>
<th>Articles found for period of search</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2010</td>
</tr>
<tr>
<td>&quot;Autism&quot;</td>
<td>5,900</td>
</tr>
<tr>
<td>&quot;Down(‘s) syndrome&quot;</td>
<td>3,432</td>
</tr>
<tr>
<td>“Trisomy 21”</td>
<td>771</td>
</tr>
</tbody>
</table>

Is there enough theory?

- “Theory of autism” 75 1,470
- “Theory of Down(‘s) syndrome” 0 0
• With ASD there is a clear attempt to link cognitive phenotype to social outcomes; social difficulties central to definition of ASD

• It appears there is less interest in explaining the subtleties of social difficulties in people with DS.

Social phenotype: sparing?

• Stereotyped as sociable and warm
• Possible relative sparing in neonatal imitative abilities (Heimann et al. 1998)
• Empathetic

But…

Social phenotype: early differences

• Differences in temperament – reduced distress (Cichetti et al)

• More attention towards people than objects (Legerstee et al)

• Different style of early interactions (Moore et al 2008; Buckhalt et al 1978; Roach et al 1998 etc.)

• Possible over application of imitative strategies (Wright, 1998)

• Possible differences in emotion recognition (Williams et al, 2005; Wishart et al 2007)

• Fewer social referencing looks (Knieps et al 1994; Kasari et al 1995)

• Reduced frequency of requesting behaviours (Mundy et al.)

Social phenotype: later differences

• Continued differences in interaction styles and fewer emotional and mental state terms in conversation (Kasari et al, 2001; Tingley et al, 1994)

• Very little known about interactions with fathers (see de Falco, 2008, 2009)

• Show reduced motivation to complete tasks and greater inconsistency (Wishart; Cuskelley)

• Still much unknown

“Joint attention in infancy forms a bedrock for shared social realities, a precondition for the acquisition and use of language, and, in the deepest sense, for the formation and maintenance of culture: it depends on sharing the focus, context, and presuppositions about objects that guide attention.” Bruner (1995)

Static directional models are not sufficient

“Understanding development itself is the key to understanding developmental disorders”

(Karmiloff-Smith, 1998)

- To understand development we need dynamic developmental models and theories.
“Despite a large number of studies there is no consensus about whether or not the development of children with DS is delayed or different... Some researchers have argued that focussing on the question of delay versus difference may not be particularly helpful. What is more relevant is to examine how different areas of development are related” Lewis (2003)

Delays can lead to differences
Differences to delays

But how?

• We need a way of notating development so that people working in different fields are able to share in debates about theoretical causal pathways.

• We need to link the important work in genetics to the subtle social outcomes

• We need to make models that incorporate a developmental perspective

- The differences between theoretical positions can be difficult to understand clearly
- Very few theories are made fully explicit in text form
- We need conceptual tools to help us make these comparisons.
- These tools are useful for clinicians and researchers

Morton’s approach

- Genetic factors
  - Brain conditions
  - Cognitive factors
- Environmental factors
  - Behavioural descriptions

Factors are divided into three categories: Biological, Cognitive, and Behavioural.
Improving this framework

- Do not restrict to uni-directional causes
- Make more allowance for changing influences over time
- Adapt the diagrammatic form to allow the description of transactional processes.

**Neurological impairments**

<table>
<thead>
<tr>
<th>Time</th>
<th>Biological</th>
<th>Infant cognition</th>
<th>Infant social behaviour</th>
<th>Maternal behaviour</th>
</tr>
</thead>
<tbody>
<tr>
<td>birth</td>
<td>Neurological impairments</td>
<td>Impaired information processing and representational development</td>
<td>Constrained attention in interactions</td>
<td>Adoption of a style characterised by ‘forceful warmth’</td>
</tr>
<tr>
<td>6 months</td>
<td></td>
<td></td>
<td>Attention becomes ‘locked in’</td>
<td>Mother takes more directive role and less sensitive to infant’s ‘topics’</td>
</tr>
<tr>
<td>12 months</td>
<td></td>
<td></td>
<td>Fewer spontaneous JA bids and requests</td>
<td></td>
</tr>
<tr>
<td>18 months</td>
<td></td>
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</table>

Why psychological theory is needed for DS

Over-simplified explanations may hinder understanding in all levels of explanation. Psychological theories can make the link from neuroscience to social behaviour

- Genetic and Neurosciences need to attend better to subtle phenotypic behavioural descriptions to understand gene expression.

- Cognitive science needs to better consider epi-genetic gene-environment effects to account for individual differences

- Clinicians and social workers need to better understand the neuro-cognitive underpinnings of behaviour

Developing a unified notation for development

• If we are to develop a better interdisciplinary understanding we need to come up with better ways of communicating across different fields.

• A unified notation for modelling causal developmental processes and pathways is required


It is only a theory but…

• A theoretical model can not be completed by one person from any one field!
• While any model and its components will be wrong this approach may allow people to articulate and demonstrate how they think a theory is wrong and why.
• This approach starts to portray development as transactional and dynamic
• This also allows us to portray both group and individual pathways but…
• We need to have a way of showing these processes and pathways in more dynamic ways … This is to follow
Conclusions

• We must consider multiple levels of description to understand developmental disorders and in DS in particular.

• We need to be careful about attributing simple causal models that are based on ‘static’ adult neuropsychology

• Much more data on early behaviours is needed if we are to unpick the developmental processes involved and establish the transactions between biological, psychological and environmental factors

• We need to start developing explicit causal models a-priori and test them!

• You must use a big piece of paper.

People with DS deserve more

• There is considerable within-group variability in people with DS that could be explained

• Taking a developmental perspective across levels of explanation may help to resolve some of the difficulties in each of these areas.

• With more complete theories we can target valuable resources to focus on critical issues

• Clear theoretical models provide more powerful rationales for intervention strategies
With Katie Cebula, Jennifer Wishart, Peter Hobson, John Oates, Julia Goodwin, Mike Anderson, Christine Deruelle and others