Built-in modules vs a developmental process of gradual modularisation: Insights from genetic disorders

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Plan

2. Where do arguments for innate modularity stem from?
   - Adult neuropsychology
   - Evolutionary psychology
   - Early infant competencies
   - Genetic disorders with *uneven* cognitive profiles
     (juxtaposition: preserved/impaired modules)

8. Why preserved module unlikely:
   - what we know about normal infant brain

3. The case of Williams syndrome
   - Domains of behavioural proficiency:
     Face processing and language: preserved modules?

4. The normal case: gradual localisation/specialisation

5. WS: failure of brain localisation/specialisation

6. Importance of tracing development back to origins in infancy:
   Cross-domain relations
Assumptions about development

Infant start state

Adult end state

Number
Syntax
Face Processing

Social cognition
Data that seem to support the nativist assumption

" Early competences in normal children

" Evolutionary psychology

" Genetic disorders seeming to show similar modular deficits as those found in adult neuropsychological patients
For instance, children with Williams syndrome have a barely measurable general intelligence and require constant parental care, yet they have an exquisite mastery of syntax and vocabulary. They are, however, unable to understand even the most immediate implications of their admirably constructed sentences.

(Piattelli-Palmarini, 2001)

In sum, brain volume, brain anatomy, brain chemistry, hemispheric asymmetry, and the temporal patterns of brain activity are all atypical in people with WS. How could the resulting system be described as a normal brain with parts intact and parts impaired, as the popular view holds? Rather, the brains of infants with WS develop differently from the outset, which has subtle, widespread repercussions.

(Karmiloff-Smith, 1998)
Autism is due to a deficit in an innately-specified module that handles theory-of-mind computations only (Leslie, 1992)

& a module that is localized in the orbito-frontal cortex (Baron-Cohen et al., 1999)

Autism affects the interconnectivity among and within various cognitive systems & in autism, functional brain development goes awry such that there is increased intra-regional specialization and less inter-regional interaction (Carpenter et al., 2001)

..examine the crucial role of unbalanced excitatory-inhibitory networks & complex pathogenetic pathways & leading to ASD through altered neuronal morphology, synaptogenesis and cell migration (Persico & Bourgeron, 2006)
Claimed *genetic double dissociation* in developmental disorders

Specific Language Impairment (SLI)

Williams syndrome (WS)

&Pink, 1999, p. 262, *italics added*}
The case of Williams syndrome
WS genotype

WS Critical Region: hemizygotic deletion of ~28 genes on chromosome 7 @ q11.23
Preserved modules: face processing, language

Impaired modules: visuo-spatial cognition, number
Do significantly better scores in one domain indicate a preserved module?

" Comparisons across domains are relative, not absolute.

" What we know about normal brain.

" Good *behavioural* scores might be reached by *different cognitive/brain* processes from the normal case.
Why preserved module unlikely?

What we know about normal brain:

"Cortex: highly interconnected in very young infant" (Conel; Huttenlocher)
"Ratio of white matter to gray matter: different in infancy" (Geidd)
"Corpus callosum: thickness of fibre bundles different in infancy" (Geidd)
"Early on, widespread activity in response to faces or to language: across multiple regions of cortex in both hemispheres" (Casey, Neville, Johnson)
"Subsequent pruning in normal development: gradual specialisation/localisation of function" (Johnson, Rakic)

Genetic mutation in WS: present from conception.
Critical genes are expressed throughout cortex, so effects of WS mutation will be widespread, not specific to a single region of cortex.
Example of face processing

Progressive modularisation
(localisation, specialisation and relative encapsulation of function)

and lack thereof in WS
despite good behavioural scores
Progressive modularisation of face processing in normal infants over developmental time (first 12 months and beyond) (2 decades of research by Johnson, de Haan, de Schonen, Simion and others)

Typically developing infants

& & 6 mo & 12 mo & adult
Different labs (Benton/Rivermead):

WS good at face processing: *in the normal range*

Preserved face processing module in WS?
Cognitive processes underlying good *behavioural* scores: same as normal?

Inversion effect (hallmark of *configural* processing) *doesn’t* emerge developmentally in WS.

Karmiloff-Smith, et al., 2004
What about WS brain?

WS adolescent in Geodesic HD-ERP net

Grice et al., 2001, 2003
Healthy controls:
Progressive restriction of input type

WS: lack of modularisation of function over time

Controls

Healthy controls:
Progressive restriction of input type

WS adults

WS: failure to specialise

WS: failure to localise

Healthy controls:
Progressive restriction of brain localisation

Human Faces

Upright

Inverted

WS

Controls
## Williams syndrome: not only faces

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- Note: change-Y, Contour change-N

Mayada Elsabbagh
Gamma-band bursts: integration/binding of features

Atypical brain function in both syndromes, but cross-syndrome brain difference

Rethink motion of featural at cognitive level& ..
Behavioural scores in normal range: underpinned by *different* cognitive/brain processes.

**Not:** impaired spatial module versus preserved face processing module.

**Common** featural processing bias for processing:
- spatial stimuli
- facial stimuli
- auditory stimuli
Lack of modularisation in developmental disorders common initial processes before, in normal development, these would have become increasingly segregated
What about the other domain of proficiency in WS?
Claims of preserved language module in WS?

Why WS language so delayed?
(often not until 5th/6th year & and as delayed as DS)

Late maturing module?

Or developmental explanation?
WS infants, toddlers and children:

- extremely delayed in onset of babbling
- extremely delayed in segmenting speech stream
- rely more on perceptual cues than linguistic labels
- production precedes pointing
- comprehension doesn’t show normal advance over production
- comprehension in WS infants/toddlers as delayed as in DS
- don’t use or follow eye gaze for referential communication,
  despite fascination with faces (dyadic vs triadic joint attention)
- don’t understand referential function of pointing
- auditory perception follows atypical developmental pathway

No single explanation: *all* contribute, in complex interactions, to late onset and atypical trajectory of WS language
Is *developmental* explanation of WS late language confined to speech/language/communication? confined to *same* domain?
Visuo-spatial precursors to socio-communication in normal/DS infants and toddlers, impaired in WS

DS/WS = different causes for similar language delay
Basic deficits in visual system,
early in WS developmental trajectory
focus on features:
cascading developmental effects over time
across several emerging higher-level linguistic/cognitive systems.
Nature versus Nurture = false dichotomy!
What about role of environment in dynamically shaping developmental outcomes (genetic, brain, behavioural)?

But is environment same for typical/atypical development?

Need for detailed study of how having developmental disorder subtly changes the environment

*changes the environment*

(social, e.g. language; motor, e.g. exploration)

in which atypical infant/child develops.
Domain-specific approaches:
Start state = domain-specific modules; core knowledge/domains
(e.g. Pinker, 1999; Spelke, 2005; Gelman, 2005)

Domain-general approaches:
Start state = single learning mechanism (e.g. McClelland, 2005)

Domain-relevant approaches
(Neuroconstructivism):
Start state = limited no. of domain-relevant biases (slight differences across cortex in brain chemistry, neuronal density, type/orientation of neurons, etc.). These initial biases become domain-specific over developmental time; modules emerge developmentally from competition during ontogenetic process of gradual modularisation (e.g. Karmiloff-Smith, 1992, 1998; Elman, Bates, Johnson, Karmiloff-Smith, Parisi & Plunkett, 1996)
2 developmental processes

1. Progressive modularization
   (specialisation/localisation and relative encapsulation of function)

3. Progressive explicitation
   (RR increasing role of language)

Cognition *not* built-in; gradual change over developmental time -> PLASTICITY FOR LEARNING

Both deficient in WS and many developmental disorders
Concluding thoughts

Choice is *not* between multiple innately-specified modules vs single domain-general learning mechanism.

Small number of *domain-relevant biases* compete (activity across whole brain) until one wins out -> *becomes* domain specific over time.

Progressive modularisation and explicitation of function.

Trace *full developmental trajectories* back to origins in early infancy: developmental changes in brain activity.

Focus also on domains of *proficiency (in the normal range)*: same behavioural scores -> *different* brain and cognitive processes: not preserved modules.

Don't only seek dissociations; *cross-domain associations* outside domain of overt deficit.

Infant cortex starts out *highly interconnected* -> progressive modularisation; atypical brains less pruning so remain more interconnected with widespread cortical activity.

Adult neuropsychological models too static for developmental disorders; need to *think developmentally* + *importance of timing across domains*:

Adult endstate, if modular -> *emergent* from developmental process of specialisation, localisation and relative encapsulation: not a *state*, but a developmental *process*: modularisation.
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http://www.psyc.bbk.ac.uk/research/DNL/personalpages/annette.html