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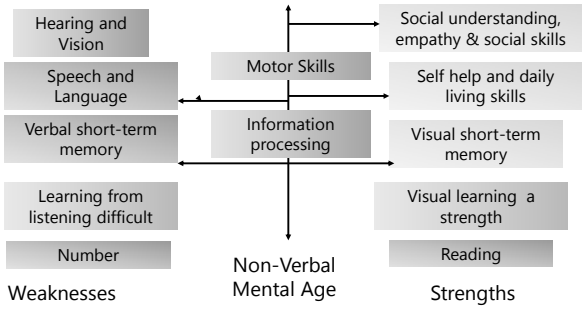
The Down syndrome phenotype: implications for effective early intervention and education

Professor Sue Buckley OBE

Effects of Down syndrome on development

- Most children will have delayed development
- There is a very wide range of individual differences from mild delays to more severe levels of disability
- For most children, severity of disability cannot be predicted at birth or in early years
- Not all aspects of development are equally delayed
- Research in the past 20 years has highlighted a profile of strengths and weaknesses
- We can use this information to be more effective in helping children reach their full potential – development is not fixed at birth

Typical profile associated with Down syndrome (see, Hodapp, Fidler, Buckley in DSRP 9 (3) online & refs



The specific developmental profile (behavioural phenotype) associated with Down syndrome

- Good social interactive skills
- Good empathy and positive personalities
- Sensitive to failure and negative emotional cues
- May use social skills to distract/avoid difficult tasks
- Good behaviour relative to mental ability and communication skills
- Good practical self-help/daily living skills over time
- Delayed early motor development – affects early learning through play and handwriting progress

The specific developmental profile (behavioural phenotype) associated with Down syndrome

Significant risk of vision and hearing impairments

Specific speech and language delays relative to non-verbal mental abilities

Cognitive strengths and weaknesses

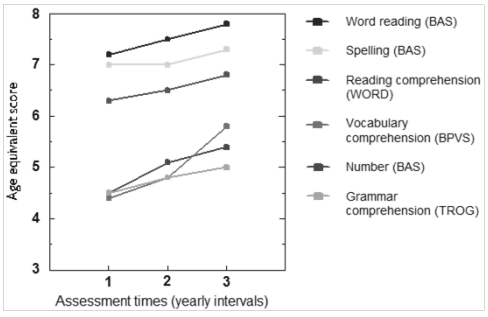
- Specific verbal working memory difficulties
- Strengths in visual memory and processing

Academic learning

Strengths in reading – can be at age level (10%)

Number more difficult – often 2 years or more behind reading

Reading, language and number skills



Children with Down syndrome (aged 6-14 years at end of study) number skills 4 years behind CA and 2 years behind their reading skills (Byrne, 1997)

The importance of the weaker areas – speech, language and working memory

- Language underpins cognitive and social development for all children
- Words for knowledge – vocabulary size
- Language for remembering, thinking, reasoning
- Language for self-control and planning
- Language for dealing with emotions and worries
- Language for communicating with others
- Language for friendships

Any child with language delay will have cognitive (mental) delay

Working memory deficits will affect all learning

Speech and language development

For most children with Down syndrome spoken language is delayed for mental age but they show an uneven profile

- **Communication** skills are usually good
- **Vocabulary** is delayed but grows steadily
  - understanding is ahead of expression
- **Grammar** is more difficult
  - tend to be 'telegraphic' talkers, using key content words
  - understanding is ahead of expression
- Clear **speech** is more difficult and means speech is difficult to understand

Vocabulary/grammar link

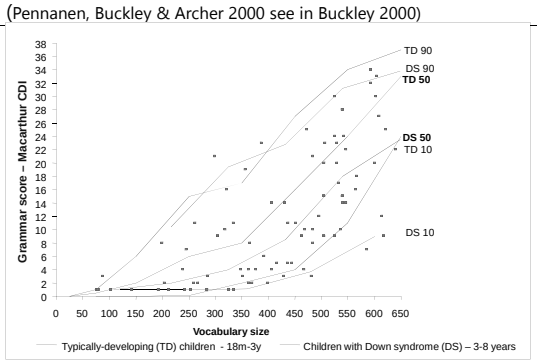
- Vocabulary size pushes along grammar development
- Children with Down syndrome have a vocabulary delay
- 200-250 words are needed before grammar starts
  - Understanding will be ahead of production
  - 200-250 words understood to begin to understand grammar
  - 200-250 spoken words to begin to use grammar

There will be many children with Down syndrome in preschool and primary schools who do not yet have 250 words in spontaneous spoken language

(Vocabulary checklists to 800 words based on MacArthur CDI available http://store.dseenterprises.org/collections/checklists)

Vocabulary/grammar link

(Pennanen, Buckley & Archer 2000 see in Buckley 2000)



Why this learning profile?

- Hearing loss plays a part
- Verbal processing may play a part
- Slow vocabulary learning may delay grammar
- Difficulties with verbal short-term memory play a part
- We know nothing of early speech discrimination in children with Down syndrome
- We know very little about causes of speech-motor issues
  - Not just a motor issue
  - Planning component
  - Verbal short-term memory component

Working memory is important for all children

- 'Working memory is the mental workplace in which information can be temporarily stored and manipulated during complex everyday activities.'
- listening to another speaker
- decoding an unfamiliar word whilst holding the meaning of the previously decoded text in mind
- writing while formulating the next part of the text
- engaging in mental arithmetic
- See excellent book S. Gathercole & T. P. Alloway. Working memory and learning: practical guide for teachers. Sage 2008 and article for teachers at http://www.york.ac.uk/res/wml/PATOSS.pdf

Effects of poor verbal short term memory function

In other children with poor verbal STM

- Speech characterised by short utterance length
- Immature syntax/grammar
- Limited range of vocabulary
- Speech clarity issues
- Storage and processing of sentences
- Poorer reading and poorer maths

See Gathercole et al (2005) Developmental consequences of poor phonological memory in childhood. *Journal of Child Psychology and Psychiatry* 46 (6) 598-611 and also in 47 (1) 4-15 on memory in developmental disorders

Implications for intervention and education

Use social/emotional strengths

- build on emotional responsiveness – encourage social communication, looking, smiling, gesture
- early social communication underpins cognitive and language development
- talk to and play naturally with children
- build on social understanding – encourage 'good' behaviour

Always encourage AGE appropriate behaviour – do not 'baby' or 'spoil' child (or adult), have clear expectations and boundaries

Implications for intervention and education

- Target speech and language difficulties from infancy and through school years
- Remember that children are visual learners
- Use *reading to teach talking* from early (2 to 3 years) and through school years
- *Learning from listening* will be specially difficult but *learning from looking easier* so always use visual supports – signs, pictures, reading, the computer
- Enable understanding to be demonstrated without the need to say it – choosing, pointing, selecting

Implications for intervention and education

- Progress with grammar is linked to total vocabulary size for children with Down syndrome – therefore teaching vocabulary is an important goal from early
- Speech skills start in first year – therefore intervention should start then – games to develop discrimination and encourage production of speech sounds
- Non verbal communication skills predict talking (joint attention and pointing) therefore intervention should start in first year
- Gesture use can close the comprehension/production gap but we need more research on the proper use of signing

Implications for intervention and education

Compensate for 'weaknesses'

- Hearing, vision – regular checks, good health care – speak clearly, use signs, limit background noise. Involve sensory impairment team
- Address working memory difficulties with sound and word discrimination games from infancy, improving spoken language development and playing memory games
- Encourage motor development at all times
  - Active practice
  - Encourage active movement through play
  - Sporting skills are good for fitness as well as social skills

In summary – the phenotype suggests

- Children with Down syndrome are visual learners
- They find learning from listening particularly difficult
- This effects learning to talk and it effects processing spoken language and instruction

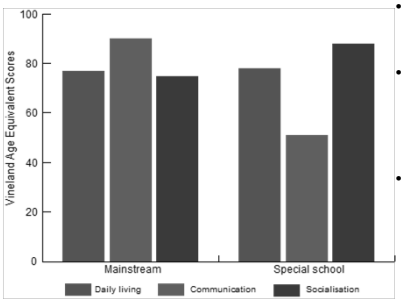
If we plan interventions to

- to focus on teaching spoken language
- support all learning visually – especially with print
- to improve and compensate for working memory

Can we make a difference?

Our data for teenagers taught in this way from preschool years suggests we can

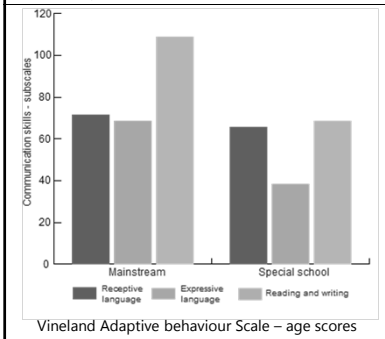
Closing the speech-language/non-verbal ability gap (Buckley, Bird, Sacks & Archer 1996)



Comparison across domains

- This is another version of the earlier coloured profile slide
- One group show the expected profile – social and practical strengths, language weakness
- The 'adapted input' mainstream group show language skills as good as their other skills – it is possible to change the profile

Language and literacy -inclusion study (Buckley, Bird, Sacks & Archer 1996)



Vineland Adaptive behaviour Scale – age scores

- Very significant gains in literacy (mean gain 3.3yrs) and expressive language (mean gain 2.5 yrs) in mainstream education
- Children fully included in mainstream classrooms
- Access the same curriculum with individual targets and in-class support
- Both groups had same range of abilities and social backgrounds at start of school

We can change the profile

- We can make a difference
- Outcome data from a study of teenagers shows significant gains in spoken language as a result of comprehensive interventions from early years
- Significantly better language and clearer speech
- Significantly better reading skills
- Linked to immersion in mainstream school/teaching to the profile of strengths and weaknesses
- Buckley, Bird, Sacks and Archer 1996 – see at http://www.down-syndrome.org/reports/295/

We need better evidence

- I have set out what we think are the practical implications of what we know at present to guide practice
- There is almost no evidence that what we do in early intervention, speech and language therapy or education is effective
- The government is arguing that there should be - see
- https://www.gov.uk/government/publications/test-learn-adapt-developing-public-policy-with-randomised-controlled-trials and https://www.gov.uk/government/speeches/michael-gove-speech-to-teachers-and-headteachers-at-the-national-college-for-teaching-and-leadership
- DSEI has recently conducted 2 RCTs with 2 different approaches – evaluating an adapted reading and language intervention in schools and evaluating a memory training programme

Evaluating the impact of the reading and language intervention

- Randomised controlled trial of 57 children with Down syndrome in 50 mainstream primary schools (aged 5-10 at start of study)
- No exclusion criteria; participating children varied widely in their abilities
- Intervention was delivered by trained teaching assistants working with the children in school on 1:1 basis
- Supported by contact with research team and termly observations
- Tough test – waiting control group also had on-to-one teaching assistants in lessons and inclusion training at the start

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Our approach to reading and language intervention

- Theoretically motivated; based on what works for typically-developing children – partners at Centre for Reading and Language, York
- Adapted to suit the learning profile of children with Down syndrome
- Able to meet a range of individual abilities and needs
- Rigorously evaluated

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Adapting teaching for the learning profile of children with Down syndrome

- **Learn more slowly**
  - Small steps, intensive daily instruction, frequent opportunities for revision and consolidation
- **Short term memory: visual stronger than verbal**
  - Visual supports for learning
- **Comprehension an extra challenge**
  - Emphasise and support reading for meaning from the outset
- **Maintaining attention and managing behaviour**
  - Short, varied activities and teaching approaches that ensure success
- **More support for phonological awareness and phonics**
  - Explicit instruction and frequent practise, alongside sight word instruction
- **Recognising the wide range of ability**
  - Scope to tailor programme for individual abilities

Our main findings

Burgoyne, Duff, Clarke, Buckley, Snowling, Hulme 2012

- On most measures intervention group progressed faster - shown as green bars above the line
- Some small gains, some larger – 4 reached statistical significance = letter knowledge, word reading, phoneme blending, expressive vocabulary
- These reflect directly taught skills – and these gains did not yet transfer to gains in spelling, non word reading or standardised language measures. Maybe not a surprise given the demands of the reading tasks and the extent of language difficulties for children with Down syndrome
- The waiting control group progressed faster when they moved to the intervention – and showed similar gains to the intervention group

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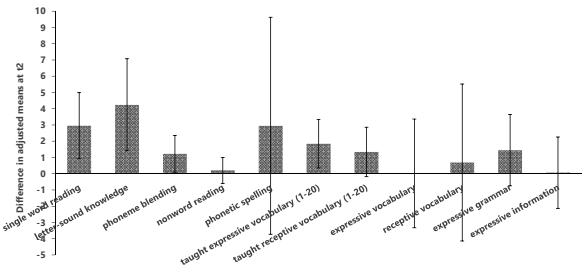
Effect of Intervention: Week 20



Individual differences in progress

- Wide variation in progress made on intervention – some children made rapid progress, other slower progress and some very little progress
- What influenced progress? - age, receptive language and no. of sessions
  - Younger children tended to make more progress – those starting at 5 and 6 years
  - Children with better receptive language tended to make more progress
  - BUT not all 5 and 6 year olds went fast and some older children did, some children with more delayed receptive language progressed with reading
  - Children receiving at least 80% of the intervention sessions made more progress
  - The only advice we can give is to try it – well planned and adapted to the individual child.
  - Other (unmeasured) factors may also contribute

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Feedback from teaching assistants

- Teaching staff delivering intervention reported that:
  - Though the intervention was hard work they enjoyed it
  - Increased confidence and feelings of competence
  - Improved skills and expertise
  - Increased self-esteem and greater job satisfaction
  - Potential for application to other children in the classroom

I am absolutely adamant that this programme (even though it is tailored for and suits children with Down syndrome extremely well) can be successfully used on many more children that struggle with a regular reading scheme

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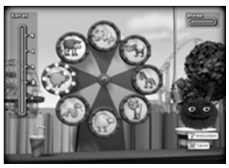
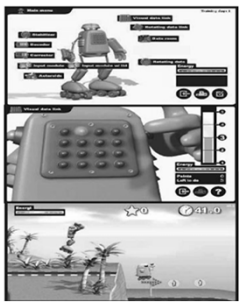
Rolling out the intervention

- Paper published in Journal of Child Psychology and Psychiatry on the RCT in 2012 – see refs
- Handbook and resources published 2012
- Training available in person and online
- Much interest from outreach support teachers, speech and language therapists and educational psychologist in training to deliver local support
- Hertfordshire funding roll-out in this county – a parent and teacher collaboration has achieved this
- Also evaluating and rolling out in Texas, USA

Cogmed JM/RM – See www.cogmed.com





- JM = 75 games.
- RM = 200 games.
- Designed by psychologists and computer games designers.
- Adaptive training on a trial by trial basis constantly adapting to each individuals WM capacity.

Pilot

5 Children with Down syndrome
Trisomy 21. Mainstream Schooling.
Cogmed training completed at **home**.

1. PPVT (Peabody Picture Vocabulary Test)
2. 8 Subtests of the AWMA (Verbal & Visual STM/WM)
3. BRIEF parent version

Main Study

24 Children with Down syndrome
21 Mainstream, 3 SEN.
Cogmed training completed at **school.**
RCT random assignment (G1 N= 12, G2 N=12)

1. KBIT 2 (Kaufman Brief Intelligence Test)
2. 4 Subtests of the AWMA (Verbal and Visual STM/WM)
3. BRIEF P parent version (preschooler)

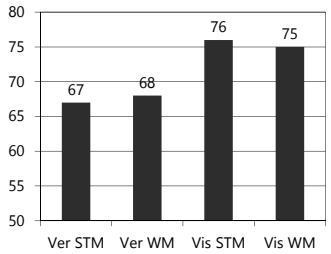
21 children completed training (Group 1 = 10, Group 2 N = 11).

Memory and Down syndrome

Research has shown that Individuals with Down syndrome have a specific deficit in the verbal memory domain.

Their visual memory skills are often less impaired. (e.g. Chapman & Seung, 2005/Hick, Botting & Conti-Ramsden, 2005).

This graph shows baseline AWMA data from our current study (N=25)
M CA 8.6 (range 7-12)
M MA 5.4 (range 4-8)



BRIEF-P – Executive Functioning Measure

Measures EF in 5 key areas.

•**Working Memory** – Hold information in mind for purpose of completing/sticking with an activity.

•**Shift** – Move freely from one situation to another, solve problems flexibly.

•**Inhibition** – Controls impulses and behaviour at correct time/context.

•**Emotional Control** – Modulates emotional responses appropriately to situation.

•**Plan/Organise** – Anticipates future events/consequences.

High scores indicate difficulties in that area – average score for typical child is 50.

RCT findings for memory training

Bennett, Holmes, Buckley in press

- Cogmed training was feasible and improved short term visual memory for children with Down syndrome in our study.
- Cogmed training may be suitable for younger children with appropriate support – also depending on their existing memory skills.
- Children who completed Cogmed training had less problems on WM & SHIFT (BRIEF-P).
- Gains are sustained – children likely need more frequent practise JM intervention programme less intensive than RM (75 activities v 200) – current case study of RM showing continuing gains.
- Planning larger trial in USA hopefully with verbal short term memory component in it

Concluding comments

- The behavioural phenotype is becoming increasingly well defined
- Very important to recognise it is not an inevitable pattern
- Development is a set of dynamic processes over time – the phenotype information can indicate how we can influence outcomes
- We need more funding to evaluate therapy and education interventions - the phenotype information is also informing potential drug trials
- Both need the development of more sensitive measures of change

The evidence for a specific phenotype or profile

- See Deborah J. Fidler (Colorado State University) and colleagues for a recent reviews of the evidence
- The Emerging Down Syndrome Behavioural Phenotype in Early Childhood. *Infants and Young Children* (2005) 18, 2, 86-103
- Education and children with Down syndrome: neuroscience, development and education. *Mental Retardation and Developmental Disabilities Research Reviews* (2007) 13, 262-271.
- The Down syndrome behavioural phenotype: implications or practice and research in occupational therapy. *Occupational Therapy in Health Care* (2011) 25, 7-25

And free access articles - preschool, primary and teenage profile papers

- Down Syndrome Research and Practice 9 (3) special section on the specific profile free at
- http://www.down-syndrome.org/research-practice/

References speech and language

- Buckley, S. J. (2000). *Speech and language development for individuals with Down syndrome: An overview.* Portsmouth, UK: Down Syndrome Education International. (available in print or e book)
- Abbeduto, L, Warren, S.F. & Conners, F.A. (2007) Language development in Down syndrome: from the prelinguistic period to the acquisition of literacy. Mental Retardation and Developmental Disabilities Research Reviews 13: 247-261
- J.E. Roberts, R.S. Chapman, & S.F. Warren (Eds.) (2008) *Communication and language intervention series: Speech and language development and intervention in Down syndrome and fragile X syndrome* Baltimore: Paul H. Brookes Publishing Co.
- Chapman, R.S., Kay-Raining Bird E. (2011) *Language Development in Childhood, Adolescence and Young adulthood in persons with Down syndrome.* In J.A. Burack, R.M. Hodapp, G. Iarocci & E. Zigler. The Oxford Handbook of Intellectual Disability and Development. Pp 167-183. New York: Oxford University Press.

References – literacy and inclusion study

- Buckley, S. J. & Johnson-Glenberg, M.C. (2008) Increasing literacy learning in Down syndrome and Fragile X syndrome. In J. E. Roberts, R.S. Chapman & S.F. Warren (Eds.) *Speech and Language Development and Intervention in Down Syndrome and Fragile X Syndrome.* (pp 233-254). Baltimore: Paul H. Brookes.
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- Buckley, S.J., Bird, G., Sacks, B. & Archer, T (2006). A comparison of mainstream and special school education for teenagers with Down syndrome: Effects on social and academic development. *Down Syndrome Research and Practice* 9 (3), 54-67.
- http://www.down-syndrome.org/reports/295/

References to RLI and Cogmed studies

- Burgoyne, K., Duff, F., Clarke, P., Buckley, S., Snowling, M. & Hulme, C. (2012). Efficacy of a reading and language intervention for children with Down syndrome: a randomized controlled trial. *Journal of Child Psychology and Psychiatry* http://onlinelibrary.wiley.com/doi/10.1111/j.1469-7610.2012.02557.x/pdf
- Burgoyne, K., Duff, F., Snowling, M., Buckley, S. & Hulme, C. (in press). Training phoneme blending skills in children with Down syndrome. *Child Language Teaching and Therapy*.
- See http://www.dseinternational.org/en-gb/resources/teaching/rli/support/ While the Handbook is intended to provide all the information needed to implement the intervention, we are continuing to support the roll-out in schools with training, web seminars and blog
- Bennett, S.J., Buckley, S.J., Holmes, J. Evaluating working memory training for children with Down syndrome. (in press). *American Journal on Intellectual and Developmental Disabilities*

The RLI handbook

- Instructions for how to deliver each component, ideas for teaching activities and adaptations to suit individual abilities
- Practical information on delivering intervention, record keeping and collaboration
- Resources for assessments and teaching, planning and record keeping
- Video illustrating each component and activities for teaching, including examples of children with different starting levels, strengths and weaknesses
- Background information on development and evaluation
- http://store.dseenterprises.org/products/reading-language-intervention-down-syndrome-teachers-handbook

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See and Learn - early intervention materials

- Visual supports for learning
- Language and reading from first words to sentences
- Speech from sounds to words
- Number coming shortly
- http://www.dseinternational.org/en-gb/resources/
- Further information sue.buckley@dseinternational.org

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