Looking in two directions: the implications for linguistic theory and clinical intervention of research on developmental and acquired sign language impairments

Bencie Woll, Deafness Cognition and Language Research Centre, University College London

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Why study sign language disorders?

- Visuomotor channel
- Multiple articulators: hands, face and torso
- Importance of facial actions
- Exploits space to show grammatical relations
- Role of iconicity in language
- Self production is substantially different from perception

Study of atypical signers reveals phenomena that might be otherwise overlooked by both spoken language linguists and signed language linguists

Questions about sign language impairments

- How are they distinct from or similar to impairments in spoken language?
- What is the prevalence of SL impairments?

WHAT CAN THEY TELL US ABOUT LANGUAGE?

Some of the conditions we have studied

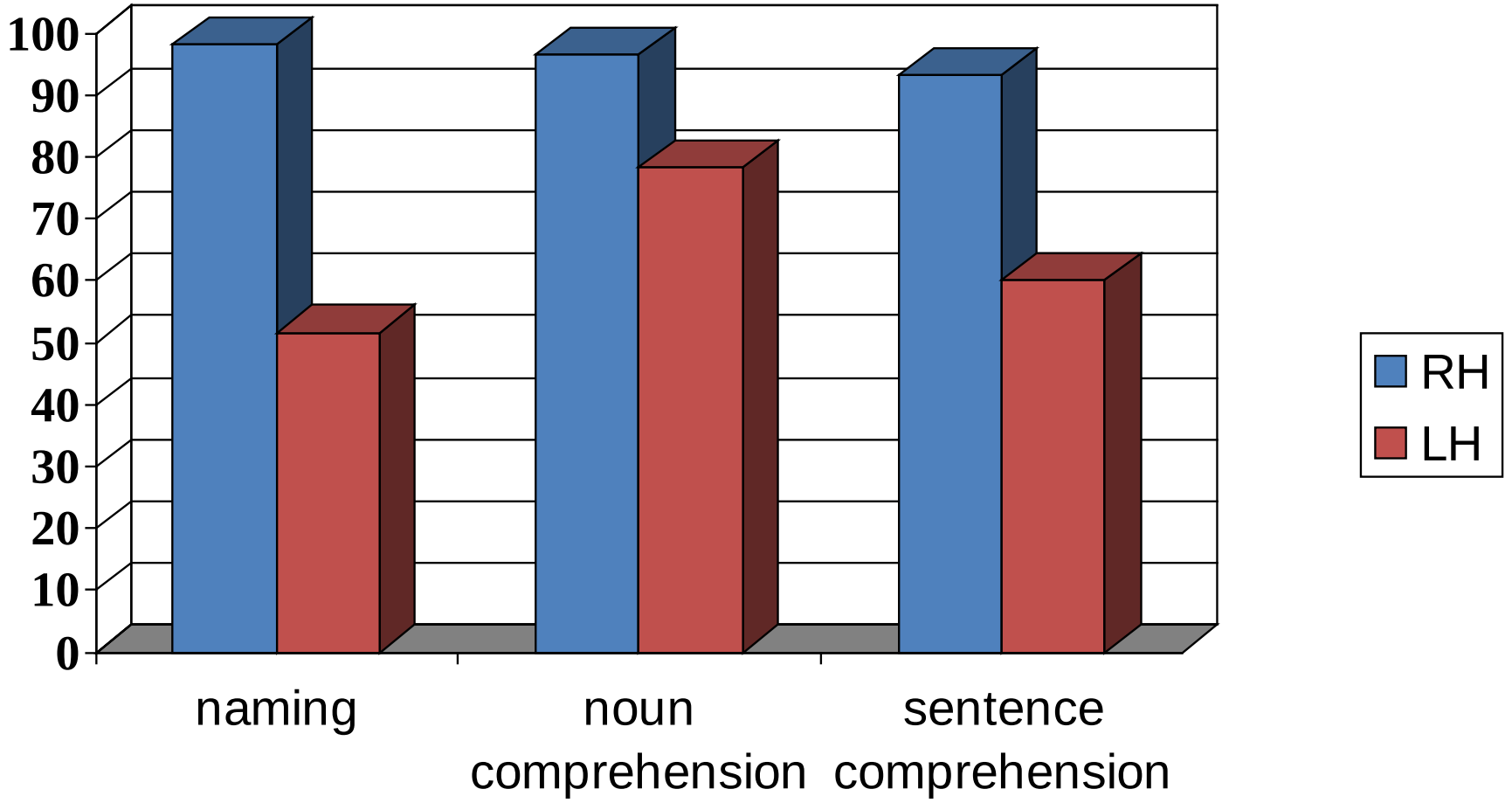
- Developmental impairments
  - HEARING SIGNERS
    - Landau-Kleffner syndrome
    - Down syndrome
  - DEAF SIGNERS
    - Specific language impairment (SLI)
    - Williams syndrome
    - Autistic Spectrum Disorder
- Acquired impairments
  - Stroke
  - Parkinson's disease
  - Progressive supranuclear palsy
  - Schizophrenia
  - Dementia

Signers with stroke

Stroke (CVA)

- Left hemisphere (LH) damage is associated with sign language impairment - Aphasia
- Right hemisphere (RH) damage is not generally associated with sign language impairment

Mean % correct on language tests

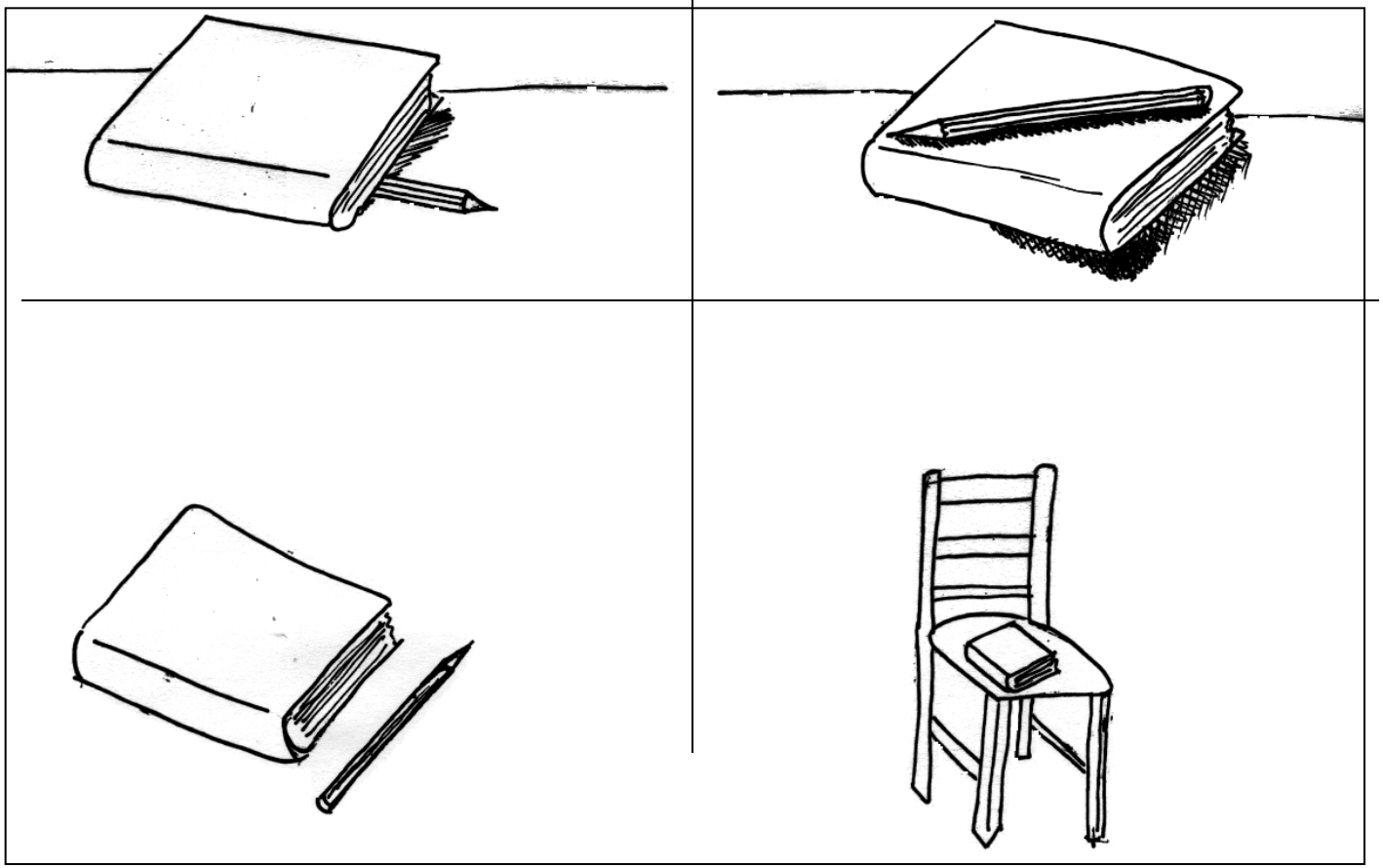


Test of Locative Comprehension

Examiner signs sentence (“the cup is on the box”)

- Match to one of 4 pictures
- 2 administrations:
  - Using prepositional signs (IN, ON, UNDER)
  - Using classifiers

Test item



PENCIL BOOK ON

Construction with preposition



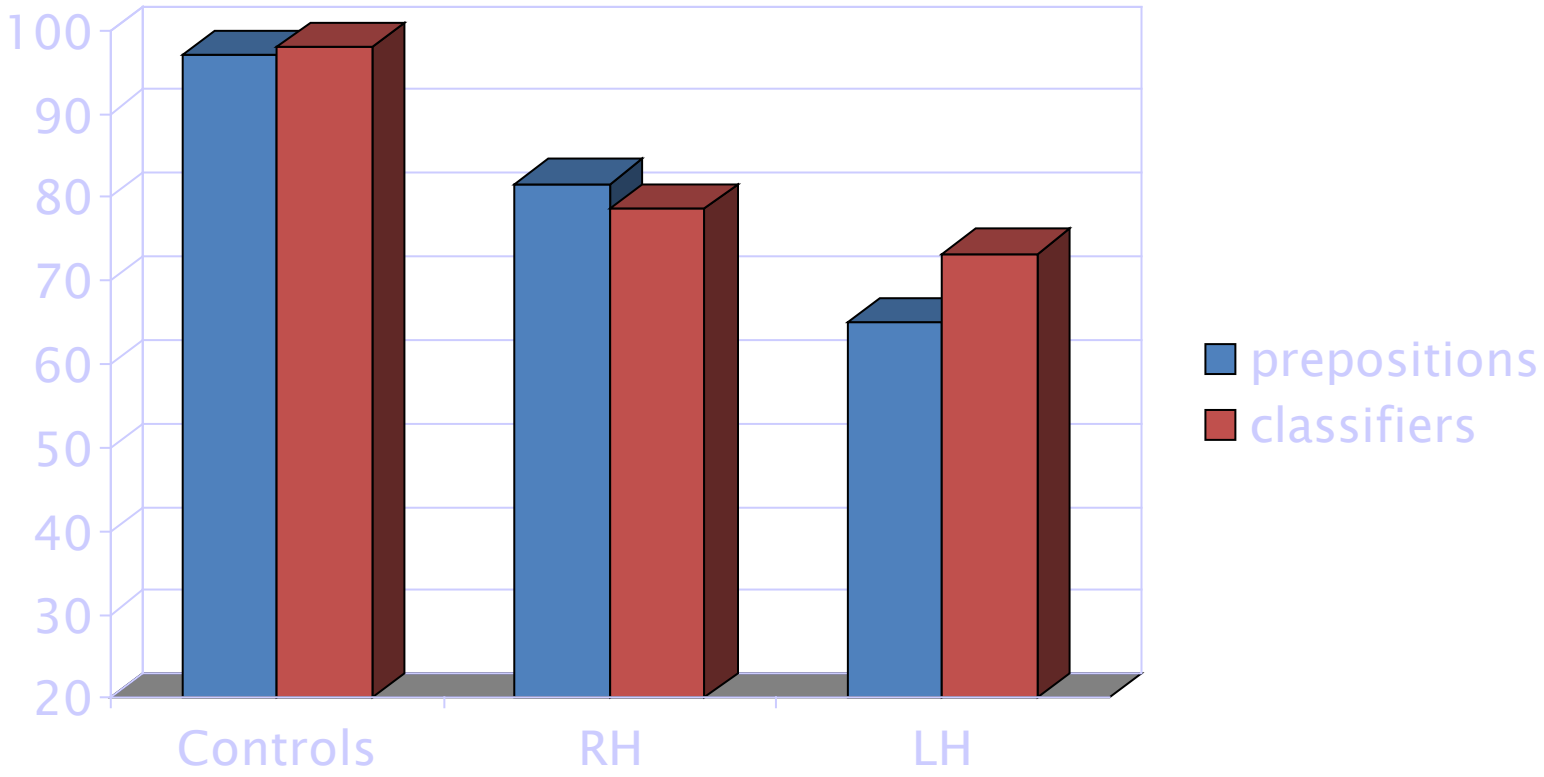
PEN ON PAPER

Construction with classifier



Paper, pen (thin object), thin object on flat object

Mean % Correct on Test of Locative Comprehension



Stroke Conclusions 1

- Participants with RH damage perform within the normal range on language tests
  - None displays aphasia in general conversation
- When testing requires processing of locatives, people with RH damage are impaired.
  - This may be a consequence of more general visuo-spatial impairments, since they are as impaired on prepositions ('linguistic') as on classifiers ('gestural')
- LH patients are worse on prepositions
  - intact visuospatial skills may assist them to process classifiers

    OR
  - Classifier constructions may make fewer demands on short-term memory

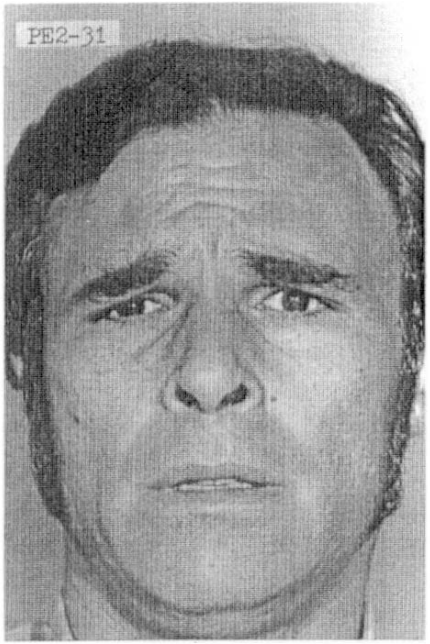
Right hemisphere stroke

Non-manual elements and BSL: role of the RH?

- Visuospatial skills
- Face processing
- Non-manual negation

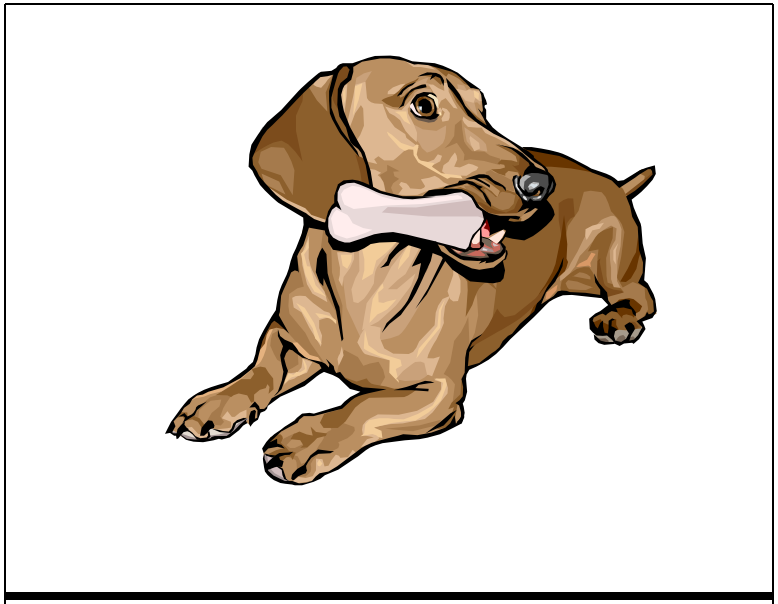
Tests of Face Processing

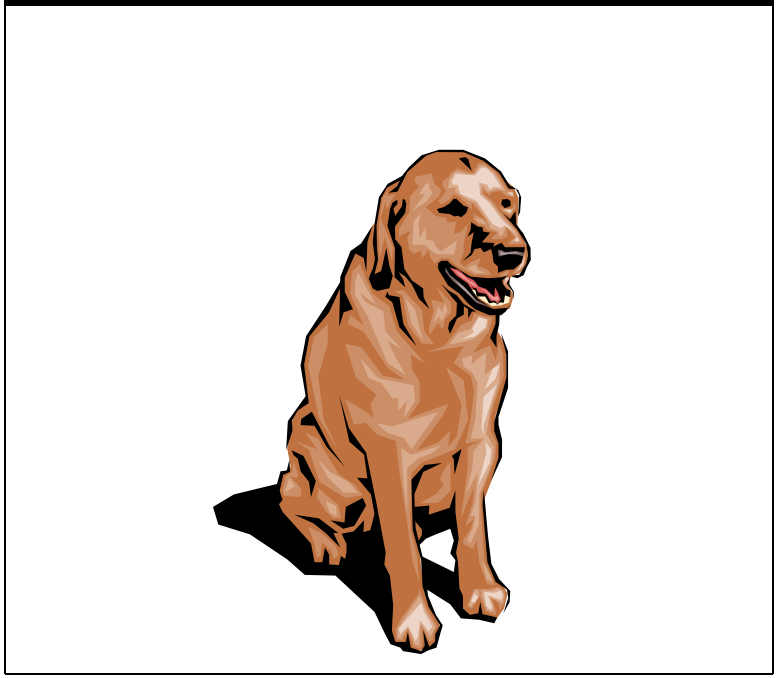
RH participants are impaired on all tests



Stanley: HAPPY

Brian: NONCHALANT



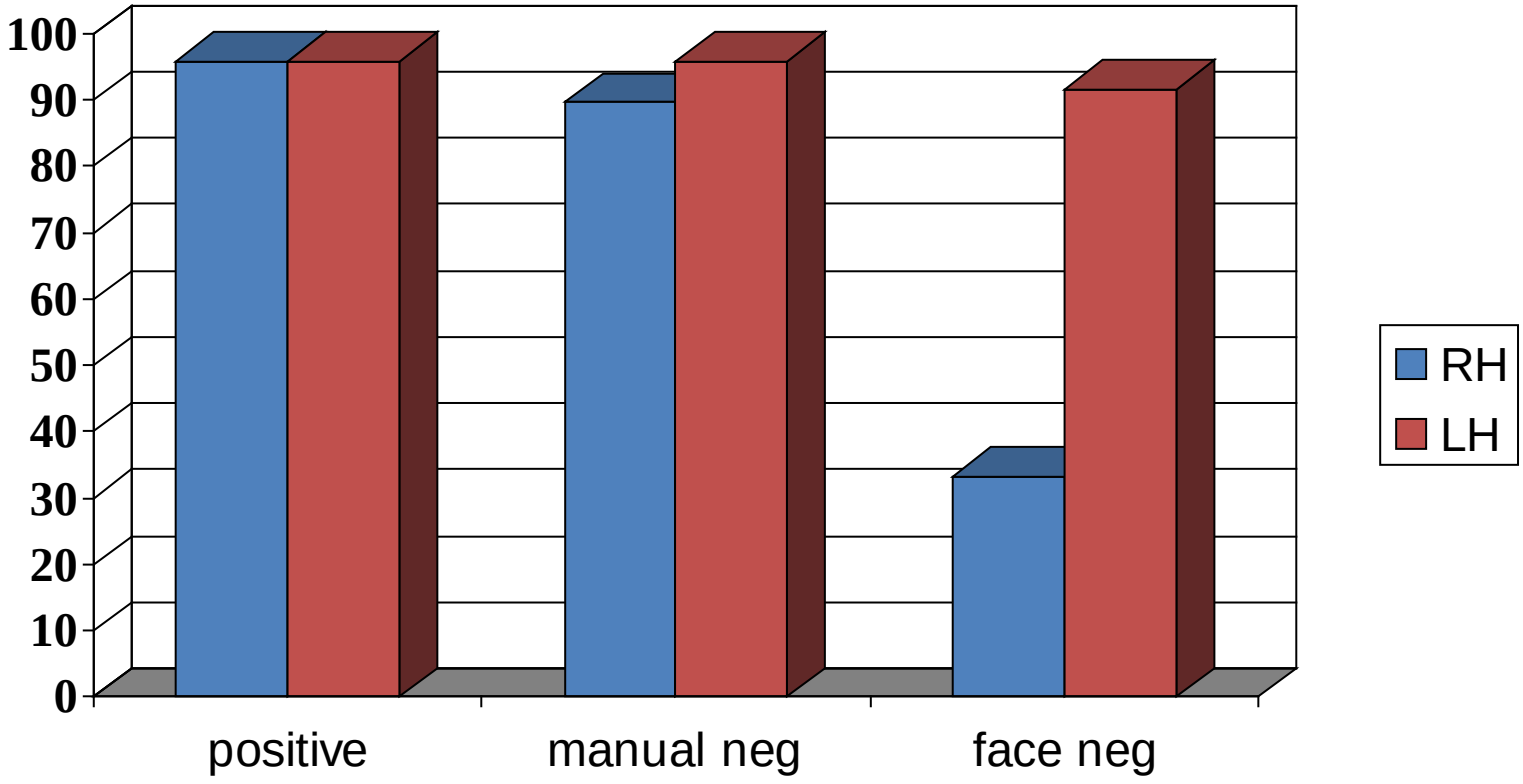


Presentation A:
BONE (HAVE)

Presentation B:
BONE NOTHING
(manual + face)

Presentation C:
BONE (NO)
(face alone)

Results: Mean % Correct



Do RH face processing impairments affect perception of all information on the face?

Lipreading tasks

Task 1: lipreading numbers

- Subject watches the examiner say the numbers 1 – 10 without voice and in random order. S/he has to point to the corresponding digit on a printed response card.

Task 2: lipreading proper names

- Subject watches the examiner say 10 proper names without voice: e.g. Bob, Arthur, Ellen, Amy. S/he has to point to it on a printed response card.

All RH patients made ≤2/40 errors per task

Stroke Conclusions 2

- Sign language can withstand facial processing impairments arising from right hemisphere damage
  - Lexical and syntactic processing are largely unaffected
  - But comprehension of some non-manual actions is affected (i.e. non-manual negation and affective facial expression)

Results from this study and the ASD study provide new insights into linguistic and gestural elements of sign language, and processing of grammar, prosody and affect

Autistic Spectrum Disorder

Autistic Spectrum Disorder (ASD)

Three characteristic difficulties:

- Face processing, using and producing facial expressions
- Limited eye contact and attention to faces
- Impaired Theory of Mind

How do such difficulties affect signers with ASD?

- Affective facial expressions
- Negation
- Adverbials

Affective facial action recognition

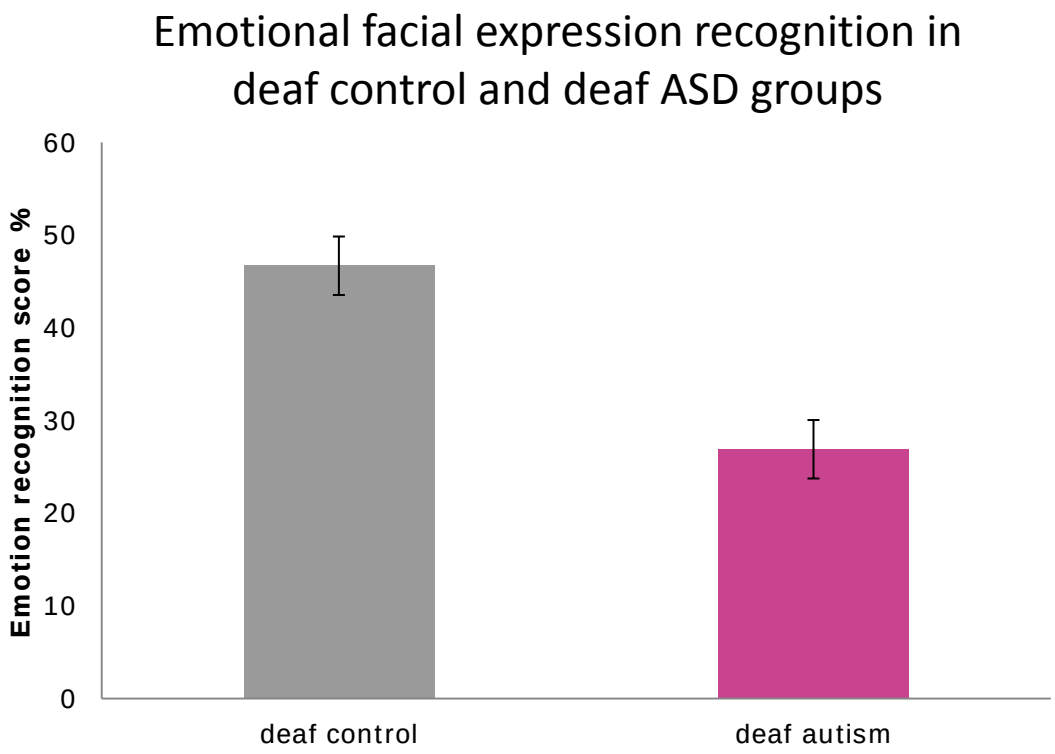
- Sentences with 6 different affective facial actions (happy, sad, neutral, angry, annoyed, disgusted, surprised and mischievous) N=48
- Participants identify emotion from closed set of responses

HAPPY

MISCHIEVOUS

NEUTRAL

Results



- Recognition impairments, particularly for HAPPY, ANGRY and MISCHIEVOUS
- Production impairments for DEMANDING and MISCHIEVOUS

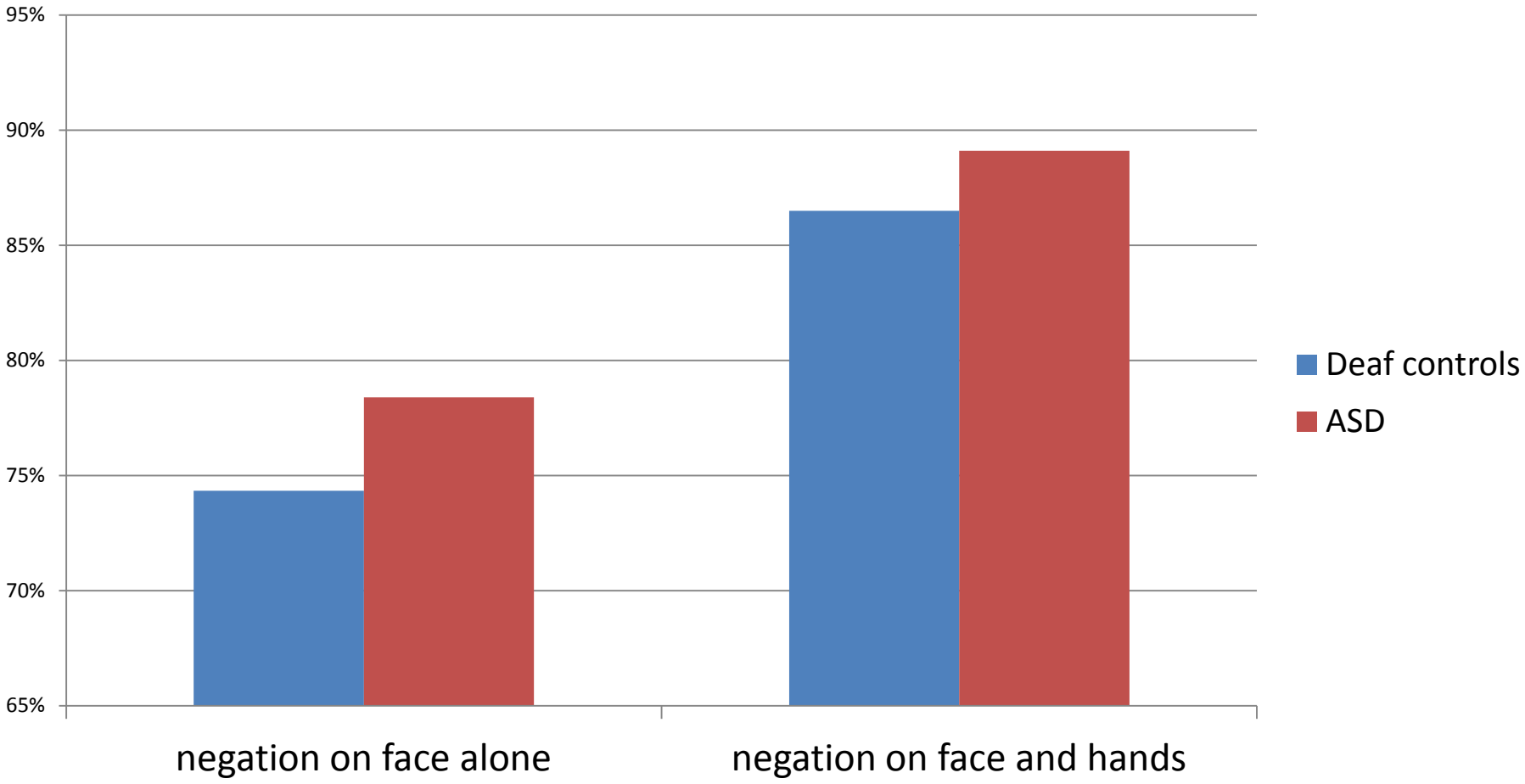
Targets requiring Theory of Mind are most affected

Negation

BREAD with *neg* face + headshake

BREAD with *neg* face + headshake and manual lexical sign NOTHING

Comprehension of negation



No significant group differences

Comprehension of BSL Adverbials

1. Large size

2. Relaxed manner

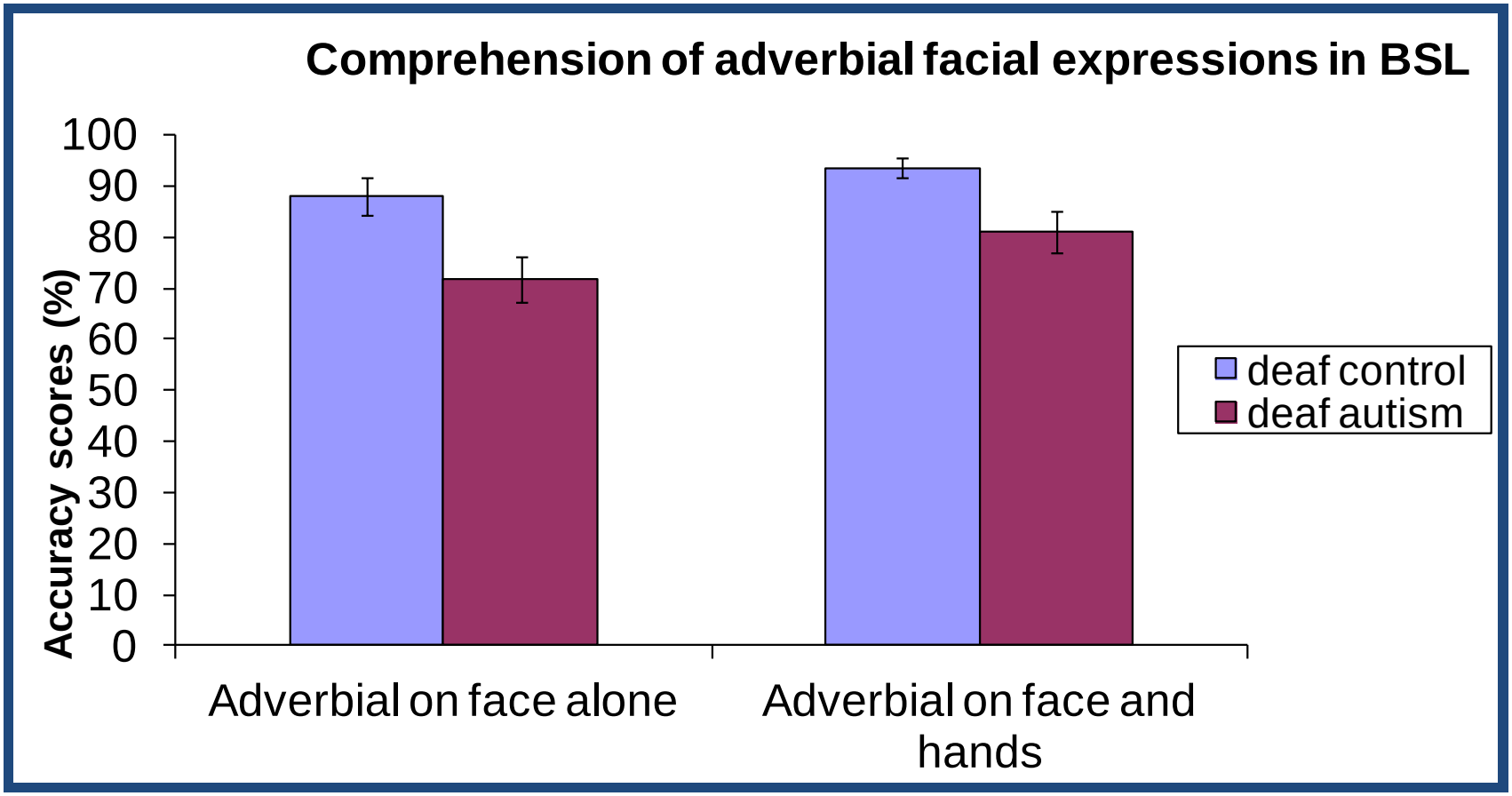
Video stimuli

Signing:

- Face only
- Face and hands

Images

Adverbials



Deaf children with ASD were significantly less accurate than controls in both conditions $F(1, 23)=8.6$, $p<.05$

ASD Conclusions

- Significant impairment in comprehension of both manual and non-manual adverbials. This suggests that such difficulties are not just consequent to facial processing limitations but attributable to a more general deficit
- Explanation? Understanding and producing adverbial facial actions relies on the ability to internalise emotional and mental states of others and to understand their actions (i.e. experiencing something as heavy, painful or slow)
- Face processing impairments are not central to ASD but are likely to be an indirect consequence of social and communication difficulties caused by a central TOM deficit

Reframes our understanding of ASD generally

Results from the stroke and ASD studies provide new insights into linguistic and gestural elements of sign language, and processing of grammar, prosody and affect

What about policy and health care delivery?

Issues

- Unmet need and hidden impairments
- Serious underrepresentation in clinical referrals
- Unique challenges in assessment of signers
- Lack of service provision to meet the needs of Deaf people with developmental and acquired neurological disorders

Estimated figures

- Assuming a BSL community of 50,000, the overall prevalence of adult Deaf BSL users living with a neurological condition is approximately 8,000
- This includes migraine, head injuries, epilepsy, stroke, motor disorders, neurodegenerative diseases etc.
- 6% of deaf children have Specific Language Impairment
- 500-700 signers have Alzheimer's disease

Health Service Access

- 2003 study on referral of Deaf people to Speech and Language Therapy (SLT) services in the UK over the previous 5 years.
  - Of 264 SLT services, only 34 had received referrals of Deaf patients, totalling 39 Deaf individuals. We estimate that at least 100 Deaf people in Britain have strokes every year with around 1/3 of the survivors becoming aphasic. Thus these figures are only 20% of the likely numbers.
- Most referrals were for swallowing disorders – not language
- Most teams did not have signing staff members and access to interpreters was variable.
- The results suggest that many Deaf people are not gaining access to SLT after neurological impairment, and those who are referred are unlikely to receive language therapy

(Marshall et al, 2003)

Why is specialist cognitive assessment necessary?

- Native vs delayed language acquisition
- Experience with visuomotor language
- Auditory deprivation (neural plasticity/adaptation)
- Organic causes of deafness (e.g. meningitis, rubella) may affect other cognitive functions

These contribute to unique cognitive signatures among deaf people which require expertise to accurately assess

- Clinician fluency in BSL is essential

Barriers to diagnosis = unequal access to health care

- Existing tests are unsuitable
- Not normed or validated with deaf signers
- Cultural and educational differences
- Poor literacy levels mean written instructions cannot be used
- Using interpreters is unreliable and error-prone, especially where communication is part of the assessment
- Lack of clinician familiarity with healthy Deaf people - makes detecting abnormalities challenging

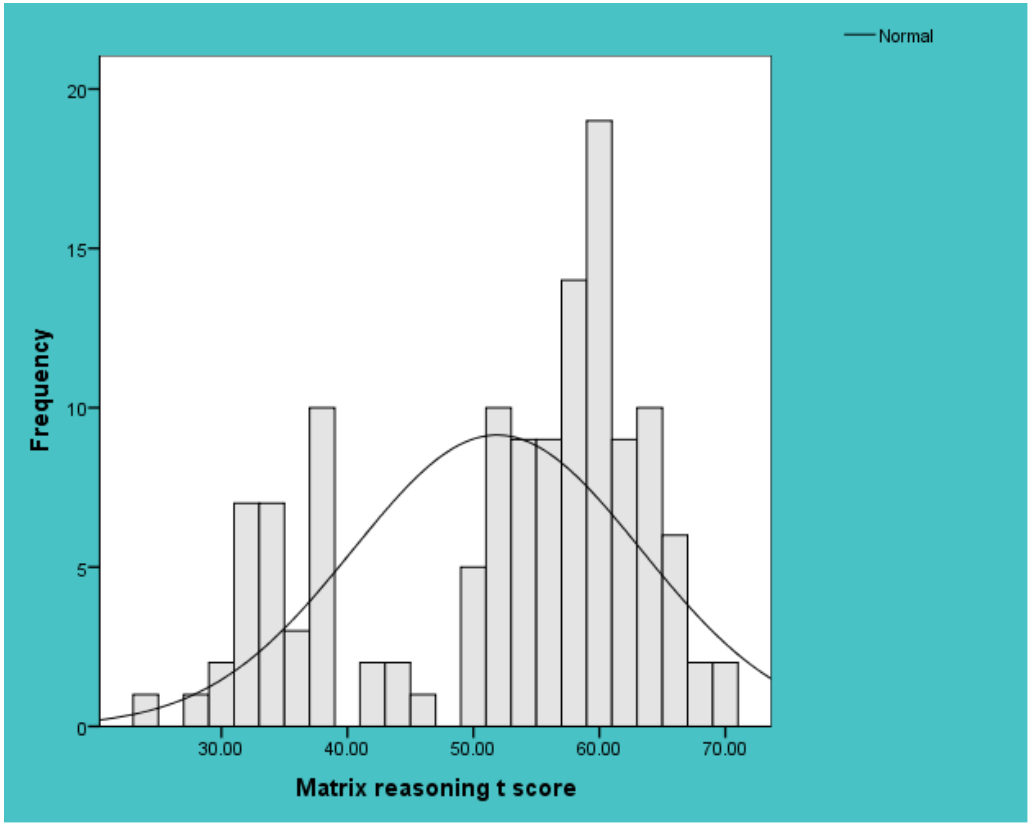
Challenges in assessing signers

- The cognitive assessment field is driven by theoretical ideas and tests for users of spoken languages, acquired by ‘ear’ in early life
- **Simply translating tests is not enough. We are not comparing like with like.**

Hidden impairments

- **Bimodal distribution of non verbal intellectual reasoning**
- Deaf BSL users aged 50-69 years (n=127)
  - 14% fell in *borderline* to *severely impaired* range (n=18)

- Greater prevalence of undiagnosed developmental and acquired cognitive impairments in the Deaf community
- Receiving no support/ treatment
- Risk of escalation and welfare state burden



Case Study 1: A deaf person who received an assessment that was only suitable for a hearing person

My mother has dementia and has since had a stroke. It took a long time to get a diagnosis of dementia because the professionals did not really know what they were looking for and they could not sign.

A person came and did an assessment which was for hearing people. It was not suitable for my mother; the questions just weren't relevant. The sign language interpreter wasn't trained in working with people with dementia, so I had to act as a relay so that my mum could understand.

By the time she was finally diagnosed, her dementia was obvious to everyone. It's sad she was not diagnosed when we first raised our concerns, as we could have had more time with her while she was clearer headed.

Case Study 2: A deaf person who was misdiagnosed because doctors could not communicate with him

When my father had the stroke things were terrible for him in hospital because the doctors couldn't communicate with him. He stayed in hospital for 4 months, much longer than necessary.

The doctors struggled to diagnose changes in his medical condition. They would ask my father a question about his medical needs and he would just nod his head having not really understood what they asked. Consequently they assumed that he was cognitively alert when this was not the case. I had to challenge this when I noticed his signing didn't make sense and he was in a state of confusion.

I have since worked out myself that he has aphasia after his stroke but the hospital did not even pick this up!

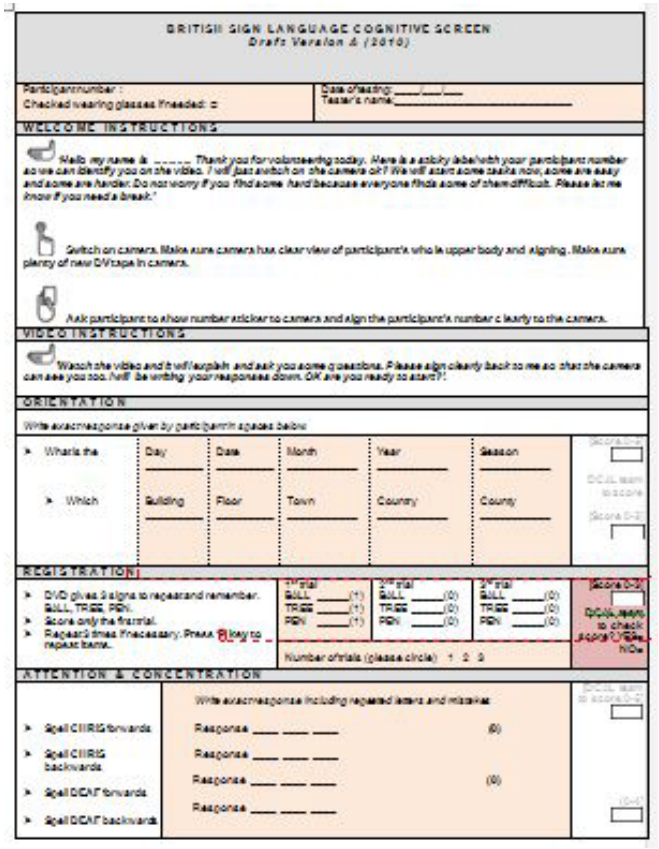
The current situation in relation to dementia

- Lack of interaction is attributed to communication barriers, not dementia, as Deaf people often live their final years in an environment where nobody signs
- Dementia is diagnosed late or not at all
- Cognitive scores contribute to decisions about medication
- Lack of accurate assessment leads to poorer access to services, pharmacological treatment and psychosocial interventions
- Families are not able to plan for future or put care arrangements in place
- Escalation in costs (falls, hospital admissions, carer burden etc.)

Assessing signers

We have developed and normed a new British Sign Language cognitive screening test (BSL-CST) to facilitate earlier diagnosis of dementia

Atkinson et al., (in press)



BRITISH SIGN LANGUAGE COGNITIVE SCREEN
Draft Version A (2010)

Participant number :
Checked wearing glasses if needed: □
Date of testing: ___/___/___
Tester's name: _____

WELCOME INSTRUCTIONS

'Hello, my name is _____. Thank you for volunteering today. Here is a sticky label with your participant number so we can identify you on the video. I will just switch on the camera ok? We will start some tasks now, some are easy and some are harder. Do not worry if you find some hard because everyone finds some of them difficult. Please let me know if you need a break.'

Switch on camera. Make sure camera has clear view of participant's who is upper body and signing. Make sure plenty of new DV tape in camera.

Ask participant to show number sticker to camera and sign the participant's number clearly to the camera.

VIDEO INSTRUCTIONS

'Watch the video and it will explain and ask you some questions. Please sign clearly back to me so that the camera can see you too. I will be writing your responses down. OK are you ready to start?'

ORIENTATION

Write exact response given by participant in spaces below

▸ What is the	Day	Date	Month	Year	Season	Score 0-5
▸ Which	Building	Floor	Town	County	County	DCAL sum score Score 0-2

REGISTRATION

- DVD gives 3 signs to repeat and remember. BALL, TRAIN, PEN.
- Score only the first trial.
- Repeat 5 times if necessary. Press Play to repeat items.

1st trial	2nd trial	3rd trial	Score 0-3
BALL ____(1)	BALL ____(0)	BALL ____(0)	DCAL sum score to check against CRIG score
TRAIN ____(1)	TRAIN ____(0)	TRAIN ____(0)	
PEN ____(1)	PEN ____(0)	PEN ____(0)	

Number of trials (please circle) 1 2 3

ATTENTION & CONCENTRATION

Write exact response including repeated letters and mistakes

- ▸ Spell CRIG forwards
- ▸ Spell CRIG backwards
- ▸ Spell DCAL forwards
- ▸ Spell DCAL backwards

Response ___ ___ ___ (0)
Response ___ ___ ___
Response ___ ___ ___ (0)
Response ___ ___ ___

DCAL sum score Score 0-2

Pilot clinic for deaf patients

We have started to use this test in routine clinical practice by creating a pilot service for Deaf patients within the Cognitive Disorders Clinic at the National Hospital for Neurology and Neurosurgery, Queens Square, London

NHS England is currently considering whether to make this a national specialist service.

Case Study 3: A deaf person who finally received the right diagnosis

After decades of frustrating experiences with the NHS, just two appointments at a specialist deaf cognitive disorders clinic and my mother finally received a diagnosis that explained her perplexing health problems.

No one could explain why she experienced pain, fatigue, reading and memory issues, bizarre visual problems, obsessions and emotional detachment. She had a myriad of investigations with consultants, neurologists, psychiatrists and support services, resulting in treatments which either made no difference or made things worse. A BSL interpreter was needed each time. Half of the appointments were cancelled on arrival because there was no interpreter. The cost to the NHS was unimaginable.

No diagnosis was ever reached. Most professionals had no understanding of deafness or BSL and each time I would have to explain that my mother was a bright, articulate person, not someone with learning difficulties... I would have to challenge the psychology tests used, because the questions were irrelevant for a deaf person or her scores were being judged against those for hearing people, rendering them meaningless.

The clinic was a remarkably different experience for us. The team were fully Deaf aware. Not just at a surface level but they had deep understanding of what is normal and abnormal for a deaf person. The service was fully accessible, building a relationship, providing assessments and diagnostic support in BSL, my mother's first language. At last we were provided with an accurate neurological diagnosis which made sense of all her symptoms. The results of an MRI scan showed brain damage to Mum's right temporal lobe. It was the missing piece of the jigsaw; all her behaviours suddenly made sense.

What is needed?

- Publicly funded specialist neurology services: to achieve earlier diagnosis and intervention
- Appropriate training for interpreters
- Cognitive assessments in sign language
- Networks of support groups for Deaf people with cognitive and language disorders
- Specialist residential facilities
- Improved knowledge and awareness in the Deaf community

Interreg V Northwest 2016-2020 HEALTHSIGN proposal

Three lines of research and provision:

- Health care:
  - The development of a health care platform to facilitate access to health care for deaf people who use sign language.
- Neuroscience and linguistic research
  - The establishment of a platform of linguistic and cognitive research related to deaf patients.
- Education:
  - The creation of a training platform for deaf and hearing people who seek to acquire skills which will allow them to work in health professions related to the care and support of sign language users.

The political agenda

- Meeting the needs of deaf citizens, especially in ensuring full communication access and early diagnosis, is a challenge we must meet
- Issues are not limited to sign language users: better understanding can also inform provision for other deaf and hearing impaired people