CASE REPORT

An inability to learn to read caused by shaken baby syndrome

Leonardo Bevilacqua,¹ Adam Kuczynski,² Merle James-Galton,¹ Alex P Leff¹

¹Institute of Neurology & National Hospital for Neurology and Neurosurgery, University College London, London, UK

²Department of Clinical Neuropsychology, Great Ormond Street Hospital, London UK

Correspondence to
Leonardo Bevilacqua,
lululrossi@hotmail.it

Accepted 21 March 2014

SUMMARY

We report a 12-year-old boy who suffered from shaken baby syndrome at the age of 4 months and has been unable to learn to read even high-frequency, three-letter words, despite slow but accurate letter naming. He had a right homonymous hemianopia and evidence of impaired higher visual function, but not at a severe enough level to account for his inability to read. Speech production and reception of language were impaired for his age but at least of an order of magnitude better than his reading performance. MRI scanning revealed focal damage to the dorsal and ventral reading pathways. This case challenges the Kennard principle, a widely accepted assumption which claims that the earlier a brain injury occurs, the better the recovery. It also adds to the growing literature suggesting that early damage to multiple parts of the language learning network can result in relatively selective impairments later in life.

BACKGROUND

Abusive head trauma in infancy, including the 'shaken baby syndrome', is a significant cause of mortality and morbidity.¹ Brain recovery after brain injury in infancy is associated with injury severity²; however, two established principles of brain function appear at odds: the inherent plasticity associated with a developing brain,³ which predicts a good outcome after early injury, and the concept of 'critical periods' where the immature brain has not yet established stable, experience-dependent neural systems,⁴ which predicts a poor outcome. The former is in line with the Kennard principle, which claims that early brain injuries result in better recovery. Here we present the case of a child who sustained abusive head trauma in infancy. Cranial MRI showed bilateral brain damage to the posterior cortex, with greater damage to the left-sided cortex. Neuropsychological assessment at the age of 12 years indicated that while there was evidence of general cognitive impairment, this was out of keeping with his profound inability to learn how to read or write.

CASE PRESENTATION

The patient, referred to as P1 hereafter, suffered from 'shaken baby syndrome' at the age of 4 months. A CT of the brain at this time showed bilateral subdural haematomas and a left parietal contusion. He was treated conservatively. Six months after his injury, he was noted to have 'minimal visual responses' when reviewed by a consultant ophthalmologist. P1 received light therapy for his vision in the first 5 years of life and speech and language therapy between 2 and 3 years. At the

age of four, he was found to have 6/9.5 (near perfect) acuity, but with a clear right-sided homonymous hemianopia. He was registered as partially sighted at this point. At five, he was noted to have problems with higher visual function ('cortical visual impairment') and was provided with a statement of special educational needs. At seven, he was assessed with the Wechsler Intelligence Scale for Children—third UK Edition. All scores were below the expected level for his age, but significantly higher for verbally mediated tests than visuospatial ones. He attended a school for children with learning difficulties where it was reported that he had compensated well for his hemianopia (eg, playing football), had reasonably intact language and motor skills, but had severe problems with reading and writing.

P1 was seen in a specialist hemianopia clinic at Queen Square, London at the age of 12 years and 11 months. All data presented here are from this assessment. He wore his glasses throughout the formal procedures and often looked closely at any materials. He occasionally misrecognised and misnamed pictured objects (eg, when shown a padlock, he said it was a 'bag' and, 'Oh, it's not a bag. It's a keylock'). He was able to identify single letters and letter pairs corresponding to phonemes, he could read the word 'up' but could not read any three letter words. He could write his own name but this was laboured and poorly formed. Numeracy was similarly rudimentary.

The patient was assessed by two specialists on the same day: AK, a clinical paediatric psychologist, and MJG, a neuropsychologist with an expertise in assessing higher visual function. The behavioural measures of cognitive abilities and academic achievements are listed in [table 1](#). He also had a standard clinical MRI scan to better characterise his brain injury.

The results of the measures of cognitive abilities are summarised in [tables 2 and 3](#). [Table 2](#) includes indices composed of several separate subtests, providing reliable summary estimates of functioning in several key domains. [Table 3](#) provides a detailed description of specific abilities based on individual tests or subtests. [Table 4](#) summarises the results of tests of academic attainments, including reading and writing.

General cognitive function

Composite scores on tests of intelligence, language and memory were significantly below the expected level for his age and consistent with the presence of general learning difficulties in the moderate range.

To cite: Bevilacqua L, Kuczynski A, James-Galton M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203070

Table 1 Cognitive assessment protocol

Test	Abbreviation	Normative data (years)
Beery-Buktenica Developmental Test of Visual-Motor Integration—Fifth Edition (Beery and Beery, 2004)	VMI-5	2–18
Children's Memory Scale (Cohen, 1997)	CMS	5–16
Clinical Evaluation of Language Fundamentals—Fourth UK Edition (Semel, Wiig, and Secord, 2006)	CELF-4 ^{UK}	5–16
Comprehensive Test of Phonological Processing (Rashotte, Torgesen, and Wagner, 1999)	CTOPP	5–24
Cortical Vision Screening Test (Warrington, Plant and James, 2001)	CORVIST	Adult
Test of Everyday Attention for Children (Manley, Robertson, Anderson and Nimmo-Smith, 1998)	TEA-Ch	6–15
Unusual Views Test	—	Adult
Visual Object and Space Perception Battery (Warrington and James, 1991)	VOSP	Adult
Wechsler Individual Achievement Test—Second UK Edition (Wechsler, 2005)	WIAT-II ^{UK}	4–16
Wechsler Intelligence Scale for Children—Fourth UK Edition (Wechsler, 2004)	WISC-IV ^{UK}	6–16

In this context, there was also some meaningful variation in the profile. Although still consistent with learning difficulties, verbal intelligence was significantly higher than non-verbal intelligence ($p < 0.05$, cumulative percentage 11.8 in children with Full Scale Intelligence Quotient ≤ 79). Estimates of short-term phonological memory and long-term recognition memory were within the broad normal range.

Specific cognitive abilities

While the composite scores tended to indicate widespread impairment of cognitive development, inspection of individual test scores differentiated more clearly between specific areas of relatively spared function and pronounced weakness. Within the domain of verbal cognition, several measures indicate that aspects of auditory verbal learning were broadly normal. For example, P1's ability to retain phonological information in short-term memory was estimated to be at the 21st centile. Formulation and recall of sentences were at the 16th centile or above. In line with these findings, long-term recall of meaningful short stories was at the 25th centile and recognition memory was similarly intact. His ability to define spoken words was marginally below the expected level for his age but still at the 9th centile.

P1 tended to obtain somewhat lower scores on tests of conceptual understanding and rate of new learning (5th centile),

and far lower scores on tests involving more complex processing or reasoning, such as when required to rearrange information in short-term or working memory, follow multistep instructions or demonstrate everyday social understanding (≤ 1 st centile).

As noted above, non-verbal visuospatial abilities tended to be significantly lower than verbal abilities. Scores at or below the 1st centile were evident across a wide range of tests drawing on visual-spatial and visual-motor functions, ranging from relatively simple tasks such as visual search, object recognition, shape-matching, tracing and rote coding to more complex constructional and abstract reasoning. Even so, it was notable that P1 passed the majority of tests of basic object and spatial perception from batteries developed for adult populations.

Academic attainments

P1's scores on tests of single word reading and spelling, and of written calculation, were very low (< 0.1 centile for his age). Even in the context of moderate learning difficulties, P1's academic attainments were disproportionately low compared to most estimates of his intellect, including his verbal ability.

INVESTIGATIONS

MRI

P1's MRI scan revealed two areas of posterior cerebral damage, affecting the medial occipital lobes bilaterally. The

Table 2 Composite measures of cognitive function

Cognitive domain	Test	Composite	Standard score	Centile
Intelligence	WISC-IV ^{UK}	Verbal Comprehension Index	65	1
		Perceptual Reasoning Index	51	0.1
		Working Memory Index	62	1
		Processing Speed Index	53	0.1
		Full Scale IQ	49	< 0.1
Language	CELF-4 ^{UK}	Receptive Language Index	64	1
		Expressive Language Index	75	5
Phonological processing	CTOPP	Phonological Awareness	70	2
		Phonological Memory	88	21
		Rapid Naming	61	< 1
		Alt. Rapid Naming	49	1
Long-term memory	CMS	Visual Immediate Index	60	0.4
		Visual Delayed Index	75	5
		Verbal Immediate	78	7
		Verbal Delayed Index	78	7
		General Memory	62	1
		Delayed Recognition Index	88	21
		Learning Index	63	1

Alt., alternate; CELF-4^{UK}, Clinical Evaluation of Language Fundamentals—Fourth UK Edition; CMS, Children's Memory Scale; CTOPP, Comprehensive Test of Phonological Processing; WISC-IV^{UK}, Wechsler Intelligence Scale for Children—Fourth UK Edition.

Table 3 Tests and subtests of cognitive function

Cognitive domain	Scale	Subtest/compound	Scaled score	Centile
Verbal learning and understanding	WISC-IV ^{UK} Verbal Comprehension	Similarities	5	5
		Vocabulary	6	9
		Comprehension	1	0.1
Visual-spatial learning and reasoning	WISC-IV ^{UK} Perceptual Reasoning	Block design	3	1
		Picture concepts	2	0.4
		Matrix reasoning	1	0.1
Short-term/working memory	WISC-IV ^{UK} Working Memory	Digit span	6	9
		Letter-number sequencing	1	0.1
		Arithmetic	3	1
Psychomotor processing	WISC-IV ^{UK} Processing Speed	Coding	2	0.4
		Symbol search	4	2
		Cancellation	4	2
Attention	TEA-Ch	Sky Search time per target	3	1
		Sky Search attention score	3	1
		Score	5	5
		Creature counting correct	2	0.4
		Creature counting timing	—	—
		Sky Search DT	10	50
Language	CELF-4 ^{UK} Receptive Language	Word-Classes-R	5	5
		Concepts and following directions	2	0.4
	CELF-4 ^{UK} Expressive Language	Word-Classes-E	3	1.0
		Recalling sentences	7	16
		Formulating sentences	8	25
Phonological processing	CTOPP Phonological Awareness	Elision	4	2
		Blending words	6	9
	CTOPP Phonological Memory	Memory for digits	7	16
		Non-word repetition	9	37
	CTOPP Rapid Naming	Rapid digit naming	4	2
		Rapid letter naming	3	1
	Alt. Rapid Naming	Rapid colour naming	2	0.4
		Rapid object naming	1	0.1
Long-term memory	CMS Visual Immediate	Dot locations total	3	1
		Faces immediate	4	2
	CMS Visual Delayed	Dot locations long delay	8	25
		Faces delayed	4	2
	CMS Verbal Immediate	Stories immediate	8	25
		Word pairs total	5	5
	CMS Verbal Delayed	Stories delayed	8	25
		Word pairs long del	5	5
	CMS Delayed Recognition	Stories delayed recognition	9	37
		Word pairs. Delayed recognition	7	16
	CMS Learning	Dot locations learning	3	1
		Word pairs learning	5	5
Visual-motor integration	VMI-5		Standard score	Centile
		Visual-motor integration	73	4
		Visual perception	55	0.3
		Motor coordination	58	0.6
			Raw score/maximum	Pass (>5 centile)
Object perception	CORVIST	Face perception	7/8	Pass
		Colour perception	4/4	Pass
	VOSP	Shape detection	20/20	Pass
		Incomplete letters	18/20	Pass
		Object decision	14/20	Fail
	Unusual Views Test	Unusual views	2/20	Fail
		Usual views	20/20	Pass
Space perception	VOSP	Dot counting	10/10	Pass
		Position discrimination	16/20	Fail
		Cube analysis	8/10	Pass

Alt., alternate; CELF-4^{UK}, Clinical Evaluation of Language Fundamentals—Fourth UK Edition; CMS, Children's Memory Scale; CORVIST, Cortical Vision Screening Test; CTOPP, Comprehensive Test of Phonological Processing; TEA-Ch, Test of Everyday Attention for Children; VMI-5, Beery-Buktenica Developmental Test of Visual-Motor Integration—Fifth Edition; VOSP, Visual Object and Space Perception Battery; WISC-IV^{UK}, Wechsler Intelligence Scale for Children—Fourth UK Edition.

left lesion was the larger of the two and contiguous damage was noted to: (1) the grey and white matter around the primary visual cortex, which extended inferiorly into the lingual gyrus and fusiform gyrus; (2) the lateral extension of the lesion just involved the inferior temporal gyrus and (3) superiorly, the damage included the precuneus, the medial

part of the superior parietal lobule and part of the posterior cingulate cortex (figure 1). On the right, the occipital lobe was affected but to a lesser extent. The primary visual cortex was spared but the regions inferior to this were not, including portions of the lingual and fusiform gyri. Importantly, there was no evidence of any damage to more anterior brain

Table 4 Tests of academic attainments

Scale	Subtest/composite	Scaled score	Centile
WIAT-II ^{UK} Reading	Word reading	40	<0.1
WIAT-II ^{UK} Mathematics	Numerical operations	43	<0.1
WIAT-II ^{UK} Written Language	Spelling	48	<0.1

WIAT-II^{UK}, Wechsler Individual Achievement Test—Second UK Edition.

structures on either side. The lateral parietal and temporal lobes appeared normal as did the whole of the frontal cortex.

DISCUSSION

P1 suffered brain damage at the age of 4 months, causing a widespread, but not global, cognitive impairment. Of particular interest was the fact that despite useful expressive and receptive spoken language and other verbal cognitive abilities, he remained unable to acquire the ability to read or write. The behavioural data collected at the age of 12 years and 11 months suggest that his striking illiteracy cannot be explained by either general visual or language impairments, although neither of these domains was unaffected. After briefly discussing the results of behavioural testing, we will focus on a neuropsychological account of his behaviour.

Figure 1 Patient's clinical MRI of the brain. Coronal fluid-attenuated inversion recovery (FLAIR, upper half), axial T2-weighted (lower half) and single sagittal image (bottom right). Damaged regions show as high signal (white) on the FLAIR and T2-weighted images. The sagittal image shows the atrophy of the posterior portion of the corpus callosum with the isthmus (arrow) highlighted.

P1 has a striking impairment in his reading skills. He performed very poorly on the standardised test of reading ability for children (<0.1% centile, table 4), such that he cannot read any high-frequency three-letter words, despite being able to name letters correctly, even fragmented ones (table 3). This suggests that his impairment is not mediated solely by impaired visual perception. While weak, his expressive vocabulary (9th centile, table 3) suggests that he has been able to develop central language skills, presumably via the auditory route. His other cognitive scores, however (table 3), indicate a more general learning difficulty, with impairments in multiple areas of cognitive function.

Although his brain injury was acquired, he does not have an acquired reading deficit (alexia) in the classical sense, as this term is reserved for people who suffer a brain injury after having learnt how to read. Rather, he has an inability to learn how to read or write. He clearly has visual perceptual difficulties including a right-sided hemianopia and evidence of higher level visual perceptual problems, so his visual impairment is not 'pure' in the sense that only visual language is affected, but by the same token, these associated visual impairments do not seem severe enough to explain his inability to learn how to read.

In terms of neuropsychological correlations, the easiest to explain is his right visual field defect, which is due to involvement of his left primary visual cortex (calcarine sulcus) with sparing of that on the right (coronal images 19–21 in figure 1). Retinotopy is coded at an early point in the mammalian cortex⁵ and while there is considerable debate over the upper limit of plastic recovery that can occur once it has been damaged,⁶ the evidence is that the first few weeks of life are critical for establishing retinotopic maps in the primary visual cortex.⁷ While right-sided hemianopias are associated with acquired reading disorders, both hemianopic and 'pure' alexia,⁸ the visual field is not causative in this case as hemianopia has only a mild effect on single word reading speeds.⁹

The involvement of the fusiform gyrus and its connections is more likely to be relevant. The damage on the left fits with abundant evidence that the left fusiform mediates efficient word recognition in normal participants and patients with pure alexia.^{10–11} P1's reading behaviour is in some ways comparable to the more severe cases of pure alexia,⁸ except that while he can identify individual letters (albeit slowly), he cannot then map these percepts onto long-term representations of word forms, presumably because these have never been established. The damage to the right ventral occipital region may well be relevant, especially in relation to the posterior callosal fibres which will have been subjected to a 'double hit' from both sides.

P1 also has damage to a more dorsal region (the left medial superior parietal lobe) and this is likely to play a part in his cognitive picture. A tractography study comparing illiterate adults who subsequently learnt to read with those who never attempted to learn demonstrated structural changes involving callosal white matter tracts connecting the dorsal posterior cortex.¹² Also, a recent study in young adults who had been born preterm showed that atrophy of the most posterior portion of the corpus callosum, the splenium and isthmus, was associated with language impairment in adolescence.¹³ The isthmus contains interhemispheric connections between the parietal lobes¹⁴ and is clearly affected in P1 (figure 1). Northam *et al* stress the importance of interhemispheric connections for language development. A functional imaging study on 76 normal skilled readers found that the neural networks they employed to

read did not cluster around a single 'average' network; rather, participants fell into four distinct patterns, some dorsal, some ventral.¹⁵

Learning points

- The development of normal reading is reliant on establishing multiple networks or routes, some which emphasise visual processing (more ventral) and some which emphasise print-to-sound processing (more dorsal). Compensation following damage to one set of routes may be mediated by those that remain.
- However, the following points should be kept in mind:
 - This case suggests that if multiple posterior brain regions are damaged early in life, both dorsal and ventral, it may not be possible for patients to develop any of the range of possible reading networks.
 - This case shows that the Kennard principle, and more recent theories inspired by it, can be violated: the concept that the earlier the injury the better the recovery is not observed in the majority of cases.^{16–17}

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Kemp AM. Abusive head trauma: recognition and the essential investigation. *Arch Dis Child Educ Pract Ed* 2011;96:202–8.
- 2 Barlow KM, Thomson E, Johnson D, *et al*. Late neurologic and cognitive sequelae of inflicted traumatic brain injury in infancy. *Pediatrics* 2005;116:E174–85.
- 3 Johnston MV, Ishida A, Ishida WN, *et al*. Plasticity and injury in the developing brain. *Brain Dev* 2009;31:1–10.
- 4 Rice D, Barone S Jr. Critical periods of vulnerability for the developing nervous system: evidence from humans and animal models. *Environ Health Perspect* 2000;108(Suppl 3):511–33.
- 5 McLaughlin T, Torborg CL, Feller MB, *et al*. Retinotopic map refinement requires spontaneous retinal waves during a brief critical period of development. *Neuron* 2003;40:1147–60.
- 6 Wandell BA, Smirnakis SM. Plasticity and stability of visual field maps in adult primary visual cortex. *Nat Rev Neurosci* 2009;10:873–84.
- 7 Cang JH, Renteria RC, Kaneko M, *et al*. Development of precise maps in visual cortex requires patterned spontaneous activity in the retina. *Neuron* 2005;48:797–809.
- 8 Leff AP, Crewes H, Plant GT, *et al*. The functional anatomy of single-word reading in patients with hemianopic and pure alexia. *Brain* 2001;124(Pt 3):510–21.
- 9 Sheldon CA, Abegg M, Sekunova A, *et al*. The word-length effect in acquired alexia, and real and virtual hemianopia. *Neuropsychologia* 2012;50:841–51.
- 10 Binder JR, Mohr JP. Topography of callosal reading pathways. *Brain* 1992;115:1807–26.
- 11 McCandliss BD, Cohen L, Dehaene S. The visual word form area: expertise for reading in the fusiform gyrus. *Trends Cogn Sci* 2003;7:293–9.
- 12 Carreiras M, Seghier ML, Baquero S, *et al*. An anatomical signature for literacy. *Nature* 2009;461:983–6.
- 13 Northam GB, Liegeois F, Tournier JD, *et al*. Interhemispheric temporal lobe connectivity predicts language impairment in adolescents born preterm. *Brain* 2012;135:3781–98.
- 14 Hofer S, Frahm J. Topography of the human corpus callosum revisited—comprehensive fiber tractography using diffusion tensor magnetic resonance imaging. *Neuroimage* 2006;32:989–94.
- 15 Kherif F, Josse G, Seghier ML, *et al*. The main sources of intersubject variability in neuronal activation for reading aloud. *J Cogn Neurosci* 2009;21:654–68.
- 16 Ewing-Cobbs L, Prasad MR, Kramer L, *et al*. Late intellectual and academic outcomes following traumatic brain injury sustained during early childhood. *J Neurosurg* 2006;105(Suppl 4):287–96.
- 17 Demir OE, Levine SC, Goldin-Meadow S. Narrative skill in children with early unilateral brain injury: a possible limit to functional plasticity. *Dev Sci* 2010;13:636–47.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow