

# Age of onset and outcome in Landau-Kleffner syndrome (1985)

DMCN   
50<sup>th</sup>  
Anniversary

*Another in our series of commentaries on notable papers from the DMCN archives. The full paper is available at [www.mackeith.co.uk](http://www.mackeith.co.uk)*

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Bishop DVM. Age of onset and outcome in 'acquired aphasia with convulsive disorder' (Landau-Kleffner Syndrome). *Dev Med Child Neurol* 1985; 27: 705–12.

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In 1957 Landau and Kleffner described an unusual childhood disorder which they termed 'syndrome of acquired aphasia with convulsive disorder', in which language regresses after a period of normal development and seizures develop. Children with this disorder typically have severe receptive language difficulties and often are at first thought to be deaf. However, pure-tone audiometry shows normal peripheral hearing, and usually there are no abnormal neurological signs. Some children do not have seizures but do have an abnormal EEG, typically with spike activity in one or both temporal lobes. Since the original description by Landau and Kleffner several further cases have been reported ...

## Commentary

DMCN shares its 50th birthday with the first description by Landau and Kleffner<sup>1</sup> of the devastating epileptic dysphasia that bears their name. Just over 20 years ago, in 1985, the Journal published Dorothy Bishop's paper on this perplexing condition which examined outcome in relationship to the age of onset and she reported on 50 cases who were a combination of those sought from the literature and her own series.

Her paper drew attention to two key aspects of the disorder and these were its heterogeneity and the initially counter-intuitive finding that a younger age of onset appeared to be related to a poor prognosis for language recovery. These continue to be important considerations in clinical practice. Firstly, Bishop acknowledged the limitations of her study that arose from its largely retrospective nature which reduced the information available about the language regression. However, she hypothesized that the disorder was one of disrupted auditory processing and yet at the same time she highlighted the inconsistencies in the clinical picture that did not entirely fit with this explanation. She also made the important observations that some children not only suffered from an aphasia, but had prominent behavioural and autistic-like presentations. Since these early descriptions, neuropsychological and linguistic assessment tools have advanced so that we can now examine, for example, differential effects on types of sound discrimination, word finding, and associated disruption of literacy, and confirm that the primary difficulty in Landau-Kleffner syndrome is a dysphasia.

Neurophysiological techniques such as magneto-encephalography can also now help explain why children have limited potential to relocate the devastated language area as there is bilateral involvement of the cortex. It appears that the likely 'pacemaker' for the electrical disruption of the language arises from the intrasylvian cortex but spreads to the contralateral sylvian cortex.

Bishop's interest in the detail of the language disruption is not just of theoretical importance. If in clinical practice we are to understand how best to support or augment communication for children during their illness and to detect change, then we need to employ appropriate assessment tools. These need to be able to accommodate the fact that the child may be contending with very low levels of language and communication. Assessment tools such as the Pragmatics Profile of Communication Skills in Children and the MacArthur Communicative Development Inventories can be helpful in this regard as they cover respectively 0 to 4 years and 8 to 30 months. Emerging signs of recovery may however only be captured by employing outcome measures such as early joint attention on video analysis of standardized scenarios that can be helpful in other clinical settings such as those used in autism. Such measures are also essential if we are to evaluate intervention and understand the influence of baseline clinical features. Bishop acknowledged that whilst she was not reporting on electro-encephalography findings she did note that seizures were not invariable and it is now well accepted that they cannot be used as a proxy for the response of the dysphasia to treatment.

Bishop's paper finishes with the conclusion that longitudinal research studies are needed to confirm whether younger age of onset is indeed associated with a poorer prognosis in the Landau-Kleffner syndrome. She suggested that this could be an artefact in that children recovering at a younger age were not reported in the literature or followed up in the longer term in clinical practice.

In November last year, the European Paediatric Neurology Society held its meeting '50 Years of the Landau-Kleffner Syndrome' to pool expertise in this difficult condition. Let's hope that there is a willingness to combine these talents so that we can establish the evidence into outcomes and effective intervention into this serious aphasia of childhood and shed light on the observations that Bishop reported on back in 1985.

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## Reference

1. Landau WM, Kleffner F. Syndrome of acquired aphasia with convulsive disorder in children. *Neurology* 1957; 7: 523–30.