

Dyslexia

The word dyslexia comes from the Greek, 'disordered words'; it was coined by an ophthalmologist, Berlin, in 1887 to describe patients who had lost the ability to read due to a stroke dividing visual processing from language areas in the left hemisphere. We would now call this 'acquired dyslexia'. Acquired dyslexia can take many forms depending on the precise areas lesioned. The features described separately below are combined to greater or lesser extents in most patients; 'pure' examples are extremely rare because strokes do not respect functional borders in the brain.

Both acquired and developmental dyslexias are most easily understood by reference to the two main routes by which we read words. The phonological or 'sublexical' route converts seen letters into their sounds. This process is essential for children reading new words and even experienced readers reading words they've never seen before, such as made up nonsense words like 'tegwop'. We can read this word by translating its letters into sounds, even though it has no meaning.

The second route to reading is the visual, semantic or 'lexical' route whereby the whole visual form of words is recognised, hence their meaning is understood very quickly, because slow translation of the separate letters into their sounds is not required. This enables familiar and irregular words to be read quickly.

'Phonological' acquired dyslexia is when the patient's sublexical, phonological route is most affected. He will have particular problems with letter to sound translation; and will not be able to read nonsense words like tegwop at all; because he has never seen them before, they are not represented in his visual lexicon. Hence he cannot use the other visual route for reading which may still be intact. This kind of acquired dyslexia occurs when the stroke involves the left supramarginal gyrus and also it is often combined with Broca's motor dysphasia when the left inferior frontal gyrus is damaged.

Visual, 'letter by letter' or 'surface' varieties of acquired dyslexia are seen when the alternative visual lexical reading route is damaged. Here the patients have particular difficulties with reading irregular words, such as 'yacht' or 'bough', where the letter sounds give little help to its pronunciation and the visual form of the word has to be learnt as a whole in order to read it correctly. Phonological 'regularisation errors' are therefore common, whereby the patient has to sound out each letter in the word, bough, and perhaps read it as 'bog'. The most common site for lesions causing visual acquired dyslexia is in the 'visual word form area' that is situated in the anterior part of the left fusiform gyrus on the under surface of the occipito-temporal junction.

The third main kind of acquired dyslexia, 'deep' dyslexia, is when the patients' misreading is semantically related, eg 'ship' is read as 'boat'; the misreading has only a similar meaning, with no visual/orthographic or phonological resemblance to the target word.¹ This occurs when there is partial disconnection between the semantic store of word meanings and their visual/orthographic and phonological representations, again often following lesions in the left angular gyrus. Normally when reading, both these are activated so that the word retrieved can be checked for accuracy against its phonological and orthographic forms.

Developmental Dyslexia

Deep acquired dyslexia is the most interesting kind from a theoretical linguistic point of view. But semantic errors of this kind are seen less often in 'developmental' dyslexia because this occurs in children learning to read, before they have acquired many visual or detailed phonemic representations of words. However phonological and visual

versions of developmental dyslexia are frequently found, which is why considering acquired, is helpful for understanding developmental, dyslexia.

Developmental dyslexia (hereafter called simply 'dyslexia') is much more common than acquired. It affects perhaps 10% of all children, particularly boys, and is a potent source of individual and family misery. These children with normal or high intelligence unexpectedly find it very difficult to learn to read despite normal schooling and other opportunity. Most neurologists now view this condition as a neurodevelopmental syndrome, because its effects are not confined to reading, but instead reveal more fundamental underlying sensory, motor and attentional features. However, there is still fierce debate. Many psychologists still take the view that dyslexia is a specifically linguistic phonological condition without more basic underlying neurological causes.²

Aetiology

The neurological case rests on many kinds of evidence: genetic, neuropathological, functional imaging, physiological, psychophysiological and behavioural. First dyslexia has a strong genetic basis. Comparing dizygotic and monozygotic twins has shown that 60% of the variability in dyslexics' reading can be attributed to the particular alleles they have inherited.³ Recent high resolution linkage studies have identified at least 6 chromosomal sites associated with reading difficulties. None seem to make any distinction between visual or phonological problems. Recently 4 genes, ROBO1, KIAA 0319, DYX1C1 and DCDC2 have attracted particular interest because they have all been implicated in the way in which neuronal migration is controlled early in brain development and with how new connections are formed later in development.⁴ For example, using RNA interference techniques, KIAA 0319 has been shown to provide essential surface active signals that control neuronal migration from the germinal ventricular plate up the radial glia to form the 6 layers of the mature cerebral cortex.⁵ However the slight underexpression of this gene that has been found in dyslexics is probably balanced by slightly increased expression of other genes that may explain the talents in areas other than reading that many dyslexics demonstrate.

The few dyslexic brains that have been examined in detail neuropathologically have confirmed that they contain many sites of mild mismigration of neurones, that must have occurred in utero early in brain development. They show surface 'ectopias' which are small (c. 1 mm) outgrowths beyond the cortical surface where large migrating neurones seem to have failed to observe stop signals at the surface.⁶ These ectopias are particularly common in the homotypical association areas that form the language network in the left hemisphere. They are associated with disorganised connectivity of these neurones not only in the cortex immediately below them but also in the homotopic areas connected to them via the corpus callosum in the opposite right hemisphere.

In addition mild abnormalities have been detected in subcortical structures. For example the large neurones that should be confined to the magnocellular layers of the main visual relay in the thalamus, the lateral geniculate nucleus, were found to be 30% smaller and more disorganised than in control brains.⁷ Again they seem to have failed to observe stop signals, so that they had infiltrated the parvocellular layers of the LGN. Likewise in the left medial geniculate (auditory relay) nucleus of the thalamus the neurones were found to be smaller and more disorganised. To summarise the neuropathological findings, large, 'magnocellular' neurones in both subcortical and cortical structures tend to be mildly abnormal and disor-



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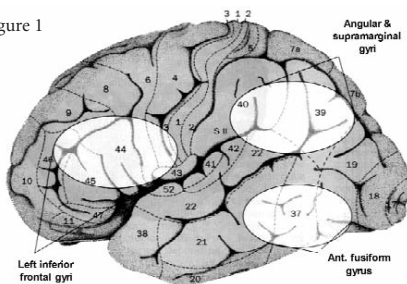
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ganised in dyslexic brains. But it must be admitted that only a small number of dyslexic brains have been examined in this detail and these could have been exceptional.

However recent functional imaging studies in live subjects have corroborated this neuropathological story to a large extent. Figure 1 shows that the same areas that showed ectopias in the language network in the left hemisphere, namely the left anterior fusiform, angular and supramarginal and inferior frontal gyri, are found to be less activated when dyslexics read compared with good readers, whereas, perhaps in compensation, language area homologues in the right hemisphere together with the left middle frontal gyrus seem to be relatively overactive in dyslexics.⁸

Figure 1



Neurophysiological studies have shown that visual, auditory and language related event related potentials are slower and smaller in many dyslexics, providing further evidence that dyslexia may be due to mild abnormalities in the overall development of the brain.⁹ But this conclusion is still hotly disputed because not all dyslexics show them. So they have been dismissed as 'epiphenomena' and not causal. But such thinking exhibits the logical fallacy known as the 'undistributed middle' - like saying that because not all diabetics suffer eye problems, eye problems are never caused by diabetes.

A fifth strand of evidence is behavioural. Developmental dyslexics have difficulty with reading of course. Although they make both visual and phonological errors most have the most trouble with splitting down the sounds of words into their constituent phonemes, so, like phonological acquired dyslexics, they make more errors trying to read nonsense words. They also tend to have poor short term phonological memory, so that they are also poor at repeating nonsense words read out to them. Others are more like visual acquired dyslexics; they have particular problems with reading irregular words, making characteristic visual errors such as continuing to confuse bs and ds and misreading god for dog or was for saw.

However in addition all developmental dyslexics are characterised by a variety of non-reading problems.¹⁰ They often report other members of the family who were affected. During development they may have been late crawlers or walkers, failed to learn to ride a bicycle easily and been generally clumsy. They tend to have difficulty distinguishing their left from their right sides. They usually have big problems with recalling a string of digits read out to them, particularly if they are asked to repeat them backwards. They tend to have difficulty with focusing attention and concentration, so that they are slow at reading out a simple list of digits, or naming a sequence of pic-

tures, or reciting the days of the week or months of the year in the right order. None of these features are particularly dependent upon reading experience; hence they suggest that dyslexics tend to have a general problem with rapid linear sequencing, again suggesting a more fundamental neurological causation and casting doubt on a purely linguistic explanation.

This doubt is reinforced by the large number of studies that have shown low level sensory anomalies in dyslexics. Many have reduced visual sensitivity to flickering coarse black and white stripes, ie they have lowered contrast sensitivity to low spatial and high temporal frequency 'gratings'. In line with their histological magnocellular abnormalities this is the hallmark of a selective impairment of the visual magnocellular system. This hypothesis that dyslexics have slightly impaired visual magnocellular function has been supported by numerous studies that have demonstrated that many dyslexics have reduced visual motion sensitivity, since this is mainly mediated by the visual magnocellular system.¹¹

Likewise dyslexics tend to have reduced auditory sensitivity to changes in the frequency and amplitude of simple acoustic stimuli; identifying these is thought to be carried out mainly by large neurones in the auditory system. Since these temporal changes mediate our ability to distinguish between the different sounds of letters, this is further evidence that dyslexics may have mild impairments in basic sensory processing that underlie their phonological reading problems.¹²

Again however, since not all dyslexics can be shown to have these impairments, it is vigorously argued that their basic problem is not sensory but at a higher phonological level. Nevertheless it is likely that many psychophysical tests lack the sensitivity required to detect the mild deficits characterising most dyslexics, but that together these slight auditory and visual deficits compound to make it difficult for them to learn to read.

Diagnosis

It is not surprising, given the amount of disagreement about its aetiology, that there is little agreement about how to identify developmental dyslexia reliably. Its hall mark is difficulty with reading, despite normal intelligence. But thereafter agreement ends. Those who believe that a linguistic phonological deficit is paramount argue that it is only necessary to demonstrate this to diagnose dyslexia. Yet children of low average intelligence experience the same kinds of difficulties with acquiring the phonological skills required for reading. So an influential body of opinion has concluded that there is no real difference between dyslexia and poor reading due to low intelligence, often known as 'garden variety reading difficulty'. The logical conclusion to all this is that there is no such thing as specific reading difficulty or developmental dyslexia.¹³

However this argument ignores the genetic, neurological and physiological evidence that dyslexia is a neurological syndrome. We can argue from this point of view that dyslexia should be diagnosed whenever there is a discrepancy between reading performance and general intelligence in the presence of many of the non-reading symptoms discussed above, such as family histo-

ry, clumsiness, poor recall of digits, slow naming, poor sequencing, visual errors, and it is this discrepancy/ syndromic approach that neurologists should follow.

Treatment

As might be expected there is great controversy about the best means of treating dyslexics. Everybody agrees however that the most important issue is to recognise the problem as early as possible, by the age of 8 before the child loses all self confidence, and descends into a downward spiral of misery and depression, even suicide, or frustration, anger and delinquency. Those who survive their usually horrible educational experiences, later develop the compensating talents that many dyslexics turn out to possess, regularly reporting that what made all the difference was someone, often a grandparent, who recognised their talents and gave them unstinting support.

Although the mainstay of treatment according to the phonological view is to train children in phonics, this is not always successful by itself. The most successful treatment regimes adopt a multi-sensory, visual, auditory and motor, approach,¹⁴ tacitly accepting that dyslexia is more than just a phonological problem. There is also evidence, some supported by randomised controlled trials, that basic visual or auditory perceptual training can often be highly cost effective for some dyslexics.^{15,16} There is much that we can do to help dyslexics, but they must be identified as early as possible, because all these treatments are more effective the younger the child, and will abort the negative emotional consequences of failure.

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