Developmental dyslexia: the cerebellar deficit hypothesis

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Surprisingly, the problems faced by many dyslexic children are by no means confined to reading and spelling. There appears to be a general impairment in the ability to perform skills automatically, an ability thought to be dependent upon the cerebellum. Specific behavioural and neuroimaging tests reviewed here indicate that dyslexia is indeed associated with cerebellar impairment in about 80% of cases. We propose that disorders of cerebellar development can in fact cause the impairments in reading and writing characteristic of dyslexia, a view consistent with the recently appreciated role of the cerebellum in language-related skills. This proposal has implications for early remedial treatment.

Developmental dyslexia is traditionally defined as ‘a disorder in children who, despite conventional classroom experience, fail to attain the language skills of reading, writing and spelling commensurate with their intellectual abilities’. Dyslexia researchers have focused on two alternative hypotheses: the phonological deficit account, which holds that the reading difficulties derive initially from problems in breaking spoken words down into their constituent sounds (syllables or phonemes), and the magnocellular deficit account, which holds that the reading problems derive from impaired sensory processing, caused by abnormal auditory and/or visual magnocellular pathways.

Unfortunately, in spite of extensive research, these approaches have failed to account for the full range of difficulties established for dyslexic children. It is therefore timely to present the case for our alternative hypothesis that the full range of deficits experienced by dyslexics are those that experience cerebellar damage later in life similarly affected? Is the cerebellum the sole contributor to dyslexia?

Two pairs of experts in this field, Thomas Zeffiro and Guinevere Eden, and Richard B. Ivry and Timothy C. Justus, discuss these and other questions. It becomes clear during the debate that the acquisition of reading-related skills requires the co-ordination of many areas of the brain involved in visual, motor and cognitive activities, and that an increased understanding of dyslexia could provide insights far beyond the disorder itself. The conclusion to this debate is provided by Roderick Nicolson and his colleagues.
It is possible to present an ontogenetic causal model for the development of the reading-related problems and other problems of dyslexic children, with the major causal factor being impaired implicit learning as a result of cerebellar abnormality\(^{15}\).

**Empirical evidence**

**Behavioural symptoms of dyslexia**

All major theories make a reasonable attempt at explaining the major behavioural symptoms - reading, writing and spelling. Consequently, crucial tests often derive from domains outside of literacy. In the studies mentioned below, the dyslexic subjects are defined in terms of significant reading delay (at least 18 months); IQ of at least 90; without attention deficit hyperactive disorder (ADHD) or serious emotional problems. Control subjects were matched with the dyslexic subjects for age and IQ, and had no reading delay. In early work we assessed the ‘profile’ of difficulties of dyslexic children by testing a range of skills, within and outside the literacy domain. Interestingly, we established that the dyslexic children tested showed difficulties ‘across the board’, in information processing speed, memory, motor skill and balance, in addition to phonological and literacy skill\(^{16,17}\). This pattern was obtained not only for group data but also for the individuals within the group. In particular, taking three disparate tests – balance (while also undertaking a secondary task), phonemic segmentation (e.g. “say ‘stake’ without the ‘t’”) and picture naming speed - 90% of the dyslexic children had ‘marked impairment’ (at least one SD below normal performance) on at least two of the tests. We concluded that the data supported our ‘dyslexic automatization hypothesis’ – that dyslexic children have difficulties automatizing skill, whether or not the skill is in the literacy domain\(^{18}\).

**Behavioural tests of cerebellar function**

Problems in automatization point to the cerebellum, which has traditionally been considered a motor area\(^{19-20}\). We have established extensive multi-disciplinary evidence directly consistent with the cerebellar deficit hypothesis. An influential study\(^{21}\) established that patients with acute cerebellar damage show a characteristic dissociation between time estimation and loudness estimation, with a significant deficit only for the former. We established that the same dissociation occurred for our dyslexic panel\(^{15}\). Next, we reimplemented the classic clinical tests of ‘cerebellar signs’ – both dystonia and dyscoordination, described in Ref. 22, and applied them to our panels. The dyslexic children showed highly significant impairments on all the cerebellar tests, and significant impairment compared even with reading-age controls on 13 of the 14 tasks, with effect sizes equivalent to those found on the earlier literacy-related tests\(^{24}\). The study was subsequently replicated with a larger sample of dyslexic children taken from the whole cohort of 8–16-year-olds at a special school for dyslexic children\(^{12}\). A similar pattern of difficulties again occurred, with highly significant deficits on balance and muscle tone comparable in magnitude to their reading and spelling deficits, and greater than their deficits on segmentation and nonsense word repetition. Particularly noteworthy was that 51 of the 59 dyslexic children were markedly impaired on muscle tone.

**Direct tests of cerebellar function**

The above tests of cerebellar function were necessarily indirect. In considering the design of a direct test, we wished to implement a functional imaging study. However, rather than select one in the reading-related domain (for which differences in performance affect interpretation of imaging data) we preferred to study a task outside the literacy domain in which there was clear evidence of strong cerebellar activation in normal subjects. Fortunately, a PET study\(^{25}\) provided a perfect opportunity. Jenkins and colleagues had their subjects learn a sequence of eight button presses by trial and error using a four-key response board with one key per finger. They established clear increases in cerebellar activation (compared with rest), both when the subjects were executing a previously overlearned (automatic) sequence of presses and also when they were learning a new sequence of presses. We undertook a precise replication, using the oldest members (now adult) of our dyslexic and control panels. Compared with the control subjects, our dyslexic subjects showed significantly less cerebellar activation in the ipsilateral (right) hemisphere. Interestingly, similar results were obtained for both tasks – executing the previously overlearned sequence, and learning the new sequence (Fig. 1).

Overall the dyslexic group showed barely any increase in activation in the right cerebellar hemisphere and...
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probable problems outside the phonological domain, and indicate that the difficulties in learning to characteristics of speech, and subsequently to the well-established difficulties in phonological articulatory fluency leads in turn to an impoverished representation of the phonological processes involved. Of particular interest is the progression highlighted as a central feature by the known difficulties in reading, writing and spelling. The text provides a fuller explanation of the impairment Cerebellar cerebellar. A hypothetical causal chain. The abscissa represents both the passage of time (experience) 5 years that lead to subsequent problems in learning to read. Other routes outline the probable problems outside the phonological domain, and indicate that the difficulties in learning to read, spell and write might derive from a number of inter-dependent factors.

vermis (~10% of the controls). This PET study therefore confirmed that the behavioural cerebellar signs of these subjects did indeed reflect abnormal cerebellar function, and therefore lends weight to the above behavioural studies.

Converging direct evidence of cerebellar dysfunction is also provided by a recent study24 of metabolic abnormalities in dyslexic men. Rae and colleagues obtained localized proton magnetic resonance spectra bilaterally from the temporo-parietal cortex and cerebellum of 14 dyslexic men and 15 control men of similar age. Bilateral MR spectroscopy indicated significant differences in the ratio of choline-containing compounds to N-acetylaspartate (NA) in the left temporo-parietal lobe and the right cerebellum, together with lateralization differences in the cerebellum of the dyslexic men but not the controls. The authors concluded that 'These differences provide direct evidence of the involvement of the cerebellum in dyslexic dysfunction'.

**Toward a causal explanation**

The above analyses indicate a correlation between dyslexia and abnormal cerebellar function in ~80% of the dyslexic children tested. A key question that arises is whether cerebellar impairment can provide a causal explanation of the development of the specific cognitive difficulties of dyslexic children.

Figure 2 (adapted from Ref. 15) outlines one hypothetical ontogenetic causal chain, linking cerebellar problems, phonological difficulties and eventual reading problems. Note that the three critical difficulties of writing, reading and spelling are all accounted for in different ways. It might be useful to distinguish between direct and indirect cerebellar causation. Cerebellar deficit provides a natural, direct, explanation of the execrable quality of handwriting frequently shown by dyslexic children. Handwriting, of course, is a motor skill that requires precise timing and coordination of diverse muscle groups. Literacy difficulties arise from several routes. The central route is highlighted. If an infant has a cerebellar impairment, initial direct manifestations will be a mild motor difficulty - the infant might be slower to sit up and to walk - and crucially, the direct effect on articulation would suggest that the infant might be slower to start babbling, and, later, talking. Even after speech and walking emerge, one might expect that the skills would be less fluent, less 'dextrous', in infants with cerebellar impairment. If articulation is less fluent than normal, then one indirect effect is that it takes up more conscious resources, leaving fewer resources to process the ensuing sensory feedback. An additional indirect effect is that reduced articulation speed leads to reduced effective 'working memory' as reflected in the 'phonological loop'24. This, in turn leads to difficulties in language acquisition25. Furthermore, reduced quality of articulatory representation might lend directly to impaired sensitivity to onset, rime, and the phonemic structure of language26 - in short, one would expect early deficits in phonological awareness. Cerebellar impairment would therefore be predicted to cause, by direct and indirect means, the 'phonological core deficit'27 that has proved such a fruitful explanatory framework for many aspects of dyslexia. For spelling, the third critical skill, problems arise from several indirect routes - over-effort in reading, poor phonological awareness and difficulties in automatizing skills.

It is valuable to consider how the above framework relates to alternative theoretical formulations for dyslexia. Note that there is a qualitative difference between the three hypotheses discussed above: the magnocellular deficit and cerebellar deficit hypotheses are both phrased in terms of an underlying neural substrate - the 'biological' level, whereas the phonological deficit hypothesis is framed in terms of a non-biological theoretical mechanism - the 'cognitive' level28.

At the biological level, cerebellar deficit is an alternative, or perhaps parallel, mechanism to magnocellular abnormality. It is possible that dyslexic children might show either or both of these abnormalities. This remains an open research issue. From the behavioural and functional data that we have established, it would appear probable that the majority of dyslexic children suffer from abnormal cerebellar function.

At the cognitive level of explanation we have outlined how cerebellar deficit accounts naturally for phonological deficit and for automatisation deficit. It also provides a natural explanation of the more recent
‘double deficit’ hypothesis. This is based on the established difficulties that dyslexic children have on ‘rapid automatised naming tasks’, in which the child has to name as rapidly as possible a page full of common pictures or standard colours, and suggests that dyslexia is characterized by a deficit not only in phonological skills but also in naming speed (reflecting a lower speed of processing). Naming speed difficulties are precisely those predicted by the cerebellar deficit hypothesis, given its established role in speech, inner speech and speeded processing. Consequently, all three cognitive level hypotheses appear to be directly consistent with, and indeed, subsumed by, the cerebellar deficit hypothesis.

Summary and conclusions
In summary, we have argued the following points.

1. A high percentage of diagnosed dyslexic children show behavioural evidence of abnormal cerebellar function – in skill automatisation, in time estimation, balance and the classic cerebellar signs of dystonia.

2. In the dyslexic adults tested, the behavioural evidence of cerebellar abnormality was accompanied by direct evidence of cerebellar abnormal function, both for executing an ‘automatic’ sequence of button presses and for learning a new sequence of button presses.

3. The difficulties in skill automatisation correspond directly to the traditional role of the cerebellum. The hypothesised role of the cerebellum in articulation-related cognitive skills is directly consistent with recent evidence of its role in speech-related cognitive tasks.

4. Finally, we provided a plausible, albeit speculative, causal analysis that explains the difficulties in reading, writing and spelling within a consistent and coherent developmental framework. Furthermore, two of the major alternative cognitive-level explanations of dyslexia, namely the phonological deficit hypothesis and the double-deficit hypothesis, might be integrated naturally within this framework.

We would like to conclude by emphasizing that the cerebellar deficit hypothesis should be seen as speculative at this stage, because the dyslexia-related data provided are mostly from small scale studies in our own laboratory. One important research requirement therefore is to establish the extent to which other groups of dyslexic children show ‘cerebellar signs’. The approach raises many further theoretical questions: are there subtypes of dyslexia corresponding to different loci of abnormality in the cerebellum; to what extent do cerebellar and magnocellular deficits co-occur; and how do these specific issues relate to underlying genetic endowment? We consider these are all potentially fruitful research issues, and we consider their investigation will continue to illuminate the complex interplay between the brain, the environment and behaviour, in both normal and abnormal development.

References
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