Cognitive deficits in developmental disorders

UTA FRITH

MRC Cognitive Development Unit, London, UK and University College London, UK


The existence of specific developmental disorders such as dyslexia and autism raises interesting issues about the structure of the normally developing mind. In these disorders distinct cognitive deficits can explain a range of behavioural impairments and have the potential to be linked to specific brain abnormalities. One possibility is that there are specific mechanisms dedicated to particular types of information processing. These mechanisms may function independently of more general information processing systems and may have a distinct anatomical basis in the brain.

Key words: Developmental disorders, autism, dyslexia, cognitive deficits.

Throughout my working life I have been interested in the distinct cognitive difficulties that characterize developmental disorders, such as dyslexia and autism. What is the nature of these difficulties? What is their locus in the brain? What do cognitive deficits tell us about the normal mind? The nature of the underlying cognitive component is always conjecture, but by imagining and systematically testing what a deficit in the critical component might mean, it is possible to arrive at powerful explanations for the origin and course of the disorder. The basic bold idea is that the disorder is caused by a discrete cognitive deficit, independent of other cognitive abilities. This is an extraordinary claim. If true, it would be evidence that the mind has a modular architecture right from the start. This is roughly how I see it: The mind possesses general and specific cognitive mechanisms for transforming information; there are all-purpose processing devices for general use, and there are some few tailor-made devices that are geared to transform only particular kinds of information. These specialist devices are vulnerable to developmental hazards. If they fail, a disorder of development will result. The head start that is provided by the dedicated devices is irretrievably lost, and this will have consequences for development as a whole.

What dyslexia and autism have in common is a genetic origin, an anatomical basis in the brain, and extremely variable behavioural manifestations. At present, neither dyslexia nor autism can be diagnosed by biological markers. Instead the diagnosis is based on behavioural criteria. These are changeable over time and subject to a whole array of general factors including environmental and cultural influences. In a sense both can be considered disorders of communication, but of very different impact and severity. Dyslexia is a mild handicap due to a deficit that has only slight impact on everyday life adaptation, while autism is a severe handicap due to a deficit in that aspect of communication that strikes at the very heart of human social adaptation.

The causal modelling of developmental disorders at three levels: biological, cognitive and behavioural has been a key element of my approach to the study of developmental disorders (Morton & Frith, 1995). A common feature of these studies is that they highlight the distinction between behavioural description and neurocognitive explanation of disorders.

DYSLEXIA STUDIES

The hypothesis of a deficit in the phonological processing of language as a cause of dyslexia has been of particular interest to me (Snowling, 1995; Frith, 1997). The suggestion is that the critical cognitive component that is dysfunctional in dyslexia is a component that is involved in the processing of speech. The nature of the critical component is at present unclear; whatever it is, it underpins the remarkable human facility in saying and hearing the right name of a word at the right time.

Figure 2 illustrates the causal modelling notation applied to the phonological deficit hypothesis. The relationship between biological and cognitive levels is indicated by a

Figure 1. Basic causal model of dyslexia as a phonological deficit.
causal arrow. Such causal arrows also occur between cognitive and behavioural levels. The relationship here is complicated by the component ‘phoneme-grapheme system knowledge’ (labelled g-p). This component is influenced by the particular orthography that is being acquired. Some of the behavioural manifestations of a phonological deficit are poor acquisition of reading, poor performance on phoneme awareness tasks, slow naming speed, and impaired verbal short term memory. The theory is that performance on these tasks is facilitated by a dedicated phonological processing device.

The claim that a phonological deficit can cause dyslexia was highly speculative at first, but has withstood testing of the following predictions: a) Problems should predate the acquisition of literacy, b) they should be independent of general intelligence, c) they should persist over time, d) they should be universal, regardless of language, e) their neurological basis should be found in the language processing areas of the brain.

Measures of phonological ability. Performance on any test is influenced by many factors. Thus, a cognitive deficit may be masked, to be revealed only by novel and taxing tasks. We therefore developed a battery for children aged 6 to 16 (Frederickson et al., 1997) which includes rhyme detection, digit naming speed, and phonemic fluency (Frith et al., 1995). This now standardized battery distinguished poor and good readers after IQ has been partialled out (Gallagher & Frederickson, 1995). Hence it might aid in the diagnosis of children whose reading disability is due to specific phonological impairments (Frith, 1995).

How early does a phonological deficit emerge? Because dyslexia has a genetic origin it is possible to study young children from affected families. This approach has been pioneered by Scarborough (1990) who found direct evidence for early language difficulties in children later diagnosed as dyslexic. Findings from our own prospective study which started at age 3 (Gallagher et al., submitted) indicate that children at risk for dyslexia had poorer than expected vocabulary knowledge due, we presume, to difficulties in phonological processing. Such difficulties were shown for example in a relative impairment in repeating novel words with unusual stress patterns.

Does the phonological deficit persist? The time it takes to spoonerise word pairs (rocket-tulip becomes tocket-rulip by exchanging initial consonants) has proved to be a particularly sensitive measure of phonological problems. We showed that able adolescents, previously diagnosed as dyslexic, who had achieved normal reading levels, and who were equal to their peers on academic grades, were significantly slower at spoonerisms (Gallagher et al., 1996). The same was found with well compensated dyslexic university students (Pauls et al., 1996).

Is a phonological deficit also found in non-English speaking dyslexics? The officially recognised prevalence of dyslexia is different in different countries. My own German background made me keen to compare reading problems in English and German. The two languages have many words in common, but their orthography is strikingly different, with German being more regular. We asked English and German children to read words that have a common root (e.g. hand, summer, baker) and found that young German children performed this task faster and more accurately than young English children until about age 12 (Frith et al., 1998). For dyslexic children, the difference was even more dramatic. The regular German orthography facilitates accurate reading even in dyslexics. The irregular English orthography appears to be less dyslexia-friendly. However, while the German dyslexics were faster and more accurate at reading, the underlying phonological impairment was the same (Landerl et al., 1997).

What is the neuroanatomical basis? In a brain imaging study on well compensated dyslexics we measured brain activity during simple phonological tasks which involved inner speech (Pauls et al., 1996). Dyslexics showed less activation in the speech processing areas of the left hemisphere compared to controls. In particular, they did not activate the areas in concert. On the basis of this study we hypothesized that the phonological deficit in dyslexia may be a result of weak connectivity between anterior and posterior language areas. At the cognitive level this may indicate a lack of fusion between orthographic and phonological codes. A prediction from this hypothesis was that dyslexics should show less interference from one code to the other. This was indeed borne out (Landerl et al., 1996). In a brain imaging project funded under the Biomed 2 programme we are now comparing normal and dyslexic readers in three languages with different orthographies, Italian, French and English. This comparison should enable us to explore how cultural and biological factors interact at the level of brain function.

Too simplistic? There have been other suggestions of cognitive deficits with a different neurocognitive basis that also make clear and testable predictions. Perhaps the best known of these is the hypothesis of an impaired magnocellular system resulting in impaired processing of fast temporal sequences (Lovegrove & Williams, 1993). This theory and the phonological deficit theory are not mutually exclusive. It may well be that we will find several distinct cognitive deficits that coexist in one and the same individual. This still does not make the deficits general, but will make the whole picture more complex. This is just what we should expect with a developmental disorder where there are large individual differences.

AUTISM STUDIES

The concept of a spectrum of autistic disorders (Wing, 1996) is now widely recognised. A triad of impairments in social interaction, communication and imagination is the common denominator in this spectrum, varying from mild...
to severe with other handicaps often superimposed. This triad can be explained by a single cognitive deficit, a lack of ‘theory of mind’ (Baron-Cohen et al., 1985). We hypothesised that there is a dedicated cognitive mechanism that allows human beings to represent thoughts, feelings and beliefs about the world, but which is not present in individuals with autism (Leslie, 1987). The term ‘mentalising’ has been used as a shorthand for this ability (Frith et al., 1994).

At the cognitive level ‘mentalising’ implies that people constantly attribute mental states to each other and predict their behaviour on the basis of inferred beliefs rather than on the basis of physical facts. As illustrated in Figure 2, the biological level, we assume that there is a neural substrate for a critical mechanism involved in mentalising which is faulty in autism. A genetic basis is considered very likely. At the behavioural level, there are a number of consequences of the lack of mentalising. One is the inability to acquire an intuitive everyday ‘theory of mind’; another, the inability to understand intended meaning; a third, the inability to understand pretend play. A number of questions arise from the claim that the triad of autistic impairments has a cognitive basis in a dedicated processing mechanism that underlies the ability to conceptualize mental states as different from real states of affairs (Frith, 1989). How early does the deficit in ‘mentalising’ emerge? Is it persistent and universal—even in case of good compensation? What are its effects on other aspects of cognitive development? What is its physiological basis in the brain?

Lack of eye gaze monitoring, lack of pointing and showing, and lack of pretend play are early signs of the triad of impairments. These could be discerned in a prospective study in children aged 18 months, and confirmed by diagnosis at age 3 (Baron-Cohen et al., 1996). The persistence of the deficit has been demonstrated in able individuals who can pass standard ‘theory of mind’ tasks. They still make tell-tale slips on advanced tasks involving persuasion, white lies, and double bluff (Happe´, 1994). Nevertheless, there are some data suggesting that such individuals have gained a measure of real social insight. Thus, they are judged by their teachers to show behaviours which benefit from a ‘theory of mind’, such as keeping secrets, and lying (Frith et al., 1994). Typically, these successful individuals who are often given the label Asperger syndrome (Frith, 1991, 1996, Attwood, 1998), have high verbal ability.

Repercussions on other abilities: Theory of mind and language. It is hard to exaggerate the importance of mentalising in language acquisition, and specifically in learning words through ostension (see Bloom, 1996). The young child needs to track the speaker’s gaze in order to associate the word with the intended referent. Thus idiosyncratic and delayed acquisition of language can be seen as a further consequence of the inability to mentalise (Frith & Happe´, 1994). On the other hand, we believe that the ability to acquire grammatical and phonological competence in a particular language is dependent on its own dedicated processing device, and that this is intact in autism. Since children with autism require a far higher verbal mental age to pass standard false belief tasks than other children (Happe´, 1994), it is possible that they pass such tasks thanks to verbal skill, perhaps using a verbally mediated strategy.

Exploring the brain basis of ‘theory of mind’. In all our studies on specific cognitive deficits we attempt to apply a ‘fine cuts’ technique (Happe´ & Frith, 1997). In order to demonstrate a specific as opposed to a general deficit it is necessary to contrast performance on mentalising and non-mentalising tasks that have identical requirements for problem solving and memory. This method was used for a Swedish-English collaborative PET study. Volunteers were scanned while reading stories which either did or did not require thinking about the protagonists’ mental states. Normal volunteers activated a circumscribed region of left medial frontal cortex only while reading ‘theory of mind’ stories (Fletcher et al., 1995). Volunteers with Asperger syndrome did not activate this region, but a neighbouring area instead. (Happe´ et al., 1996). The groups did not differ when reading non-mentalising tasks. These first results indicate that it will eventually be possible to identify the neural basis of ‘mentalising’ and to understand the physiological causes of its malfunction.

Understanding the nonsocial features of autism. A major limitation of the theory of mind deficit account of autism has been its inability to address the non-social features of autism; e.g. the restricted repertoire of interests and activities and the islets of ability. We have suggested that these features of autism may arise form a particular cognitive style: weak central coherence (Frith, 1989; Frith & Happe´, 1994). Unlike individuals with mental handicap, people with autism appear to pay more attention to parts than to wholes, and retain the surface form of information while failing to extract gist. Recently, Happe´ (1996) showed that individuals with autism succumb to visual illusions less than control subjects. She argued that this is due to a tendency to perceive even simple line figures in a fragmented way. This finding raises new questions about how people with autism perceive the world.
A number of studies support the idea that weak central coherence and impairments in theory of mind may be independent, though interacting, aspects of autism. At present we suggest that people with autism are at the extreme end of the normal continuum for central coherence, but have also suffered a specific impairment to the theory of mind mechanism. We expect coherence to vary in the normal gene pool and suggest that weak central coherence, which carries both benefits and disadvantages, may be an essential aspect of the extended phenotype of autism. We are currently exploring the hypothesis that relatives of people with autism may show special skills in areas where weak coherence is beneficial.

Too simplistic? The variability of the clinical picture of autism is staggering—huge differences are observed not only between individuals, but there are also marked changes over time in one and the same individual. The severity of the handicap may often point to additional problems, so that the idea of a single specific deficit seems far too simple. The interaction of a deficit in ‘theory of mind’ with general factors, such as ‘weak central coherence’ and impairments in theory of mind may be an essential aspect of the extended phenotype of autism. It is clear that the study of developmental disorders still has a long way to go before we can fully understand the nature of the underlying cognitive deficits and their links to behaviour as well as to their neural substrates. The unravelling of the origin and course of developmental disorders is an exciting task. It is possible to make a start even though much is still unknown.

Conclusion. It is clear that the study of developmental disorders still has a long way to go before we can fully understand the nature of the underlying cognitive deficits and their links to behaviour as well as to their neural substrates. The unravelling of the origin and course of developmental disorders is an exciting task. It is possible to make a start even though much is still unknown.

REFERENCES


Accepted 20 January 1998

© 1998 The Scandinavian Psychological Associations.