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A clinico-genetic study of psychiatric disorder in Huntington's chorea

by David C. Watt and Anneke Seller

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A clinico-genetic study of psychiatric disorder in Huntington's chorea

Huntington's chorea (HC) is a dominantly inherited neurological disorder of movement accompanied by dementia. It is progressive, leading to severe disability in five to ten years and death within a further ten. In the course of the illness, and sometimes before the onset of organic symptoms, periods of functional psychiatric disorder (fpd) of varying types occur; these add to the burden of the illness and of caring for affected subjects. An opportunity of examining these conditions was provided in 1983 by a discovery that enabled a genetic test to be developed. This test brought to genetic departments young adult couples who were contemplating marriage or procreation, where one adult from each couple was from an HC family, seeking information about the test. These young adults were more numerous and provided a more open and informative attitude about their family than had formerly been the case. This gave an enhanced opportunity, facilitating the examination of a larger number of affected subjects and their unaffected relatives.

A detailed description of the genetic basis of HC and of the test is given here. The incidence of diagnostic categories of fpd is compared for affected HC subjects with subjects of 50% initial risk of HC, with subjects of zero risk in a similar environment, with subjects affected with long-standing, disabling, incurable disease and, for those who by pre-symptomatic genetic testing are shown to be carrying the HC gene, with those in whom it is shown to be absent. The interval between the onset of organic symptoms of HC and the onset of fpd was measured and the bearing of all these results on the relation of HC to fpd is discussed, together with the future prospects of the genetics of HC.

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