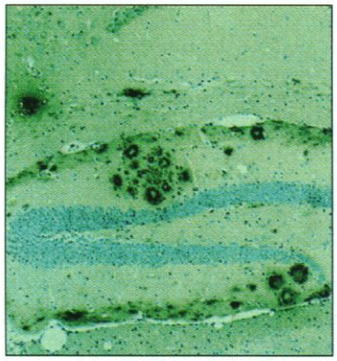
Neurodegeneration

Neurodegenerative disorders are some of the most feared illnesses in society. For example, Alzheimer's disease, which affects 5 to 10% of all people over 65 years of age, causes the progressive loss of memory and other mental faculties, leaving the individual confused and incompetent to care for him- or herself. Needless to say, this is extremely distressing for the victims and their caregivers. Another relatively common neurodegenerative disorder (affecting 1 in 10,000 individuals) is Huntington's disease. Patients with Huntington's make involuntary movements and become severely emotionally disturbed as well as cognitively impaired. The prion diseases, including the new variant form of Creutzfeldt-Jakob disease, lead to mental and physical decline and eventual death.



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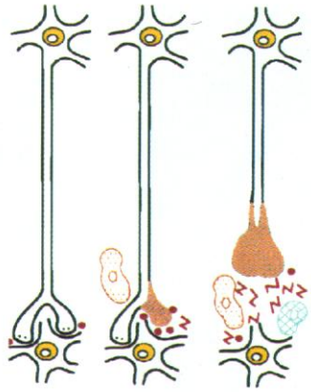
No successful treatments are yet available for any of these disorders. Research on neurodegeneration aims to promote understanding of the pathology of these diseases in order to develop successful treatments and to prevent the onset of symptoms in patients. Many neurodegenerative disorders are linked to particular genetic traits, although sporadic cases are also observed. For example, the chromosome and gene linked to Huntington's disease, the prion genes linked to spongiform encephalopathies, and the triplet repeat mutations responsible for myotonic dystrophy, Freidrich's ataxia, and other ataxias as well as for Huntington's disease have all been identified. This special issue of *Science* focuses on molecular aspects of neurodegeneration research, an area that is proving particularly productive in our understanding of the basic pathology of these disorders. Price *et al.* review how transgenic mice have come to the forefront of neurodegenerative research to provide insight into the molecular pathology of human disorders in an easily manipulated experimental model system. Hardy and Gwinn-Hardy describe the classification of genetically predetermined neurodegenerative disorders, leading to groupings of disorders that have different pathological outcomes for patients, depending on the brain region affected, but may have very similar cellular pathologies and turn out to be amenable to similar treatment or prevention strategies. Shoulson looks at the role of clinical trials of potential therapies for neurodegenerative disorders. He points out the problems in arranging for appropriate, and large enough, patient and control groups for disorders that present themselves in very diverse ways, depending on the complex interplay of the patient's complete genetic makeup with the particular disease gene.



A News story (p. 1030) also looks at potential treatments for patients with neurodegenerative disease. Marcia Barinaga describes some behavioral strategies aimed at helping patients with Alzheimer's disease by finding ways to maximize functionality and minimize some of the more distressing symptoms.

The field is now set to exploit model systems in which to test potential therapies and prevention strategies that should eventually lead to beneficial treatments for some of these particularly cruel and distressing diseases.

–STELLA M. HURTLEY

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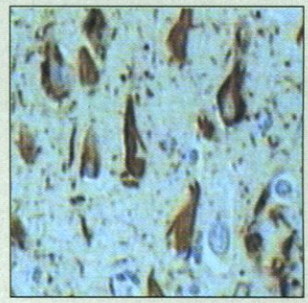

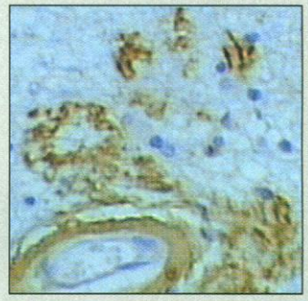

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See also related News story on p. 1030.

Science