PRESS RELEASE
WINNERS ANNOUNCED
1997/1417H KING FAISAL INTERNATIONAL PRIZE
IN MEDICINE

Topic: DEGENERATIVE DISEASES OF THE NERVOUS SYSTEM

The King Faisal International Prize in Medicine has been awarded for research on degenerative diseases of the nervous system. Between 3 and 5% of the world's population suffer from one or other type of incurable, degenerative disease of the nervous system. Many individuals are afflicted as young adults which results in a loss of their ability to work, thus imposing a considerable social and financial burden. An increased understanding of the genetic, molecular and biochemical bases of several of the most prevalent of these diseases is anticipated to lead to improvements in their diagnosis and management.

Professor Colin Louis Masters, an Australian national born in Perth in 1947, graduated in medicine in the University of Western Australia in 1960 and is now Professor and Head of the Department of Pathology in the University of Melbourne as well as Chief of Neuropathology at the Mental Health Research Institute of Victoria.

Professor Konrad Beyreuther, a German national born in Leutersdorf, Germany in 1941, obtained his PhD at the Max Planck Institute for Biochemistry in Munich in 1968. Since 1987 he has been Professor of Molecular Biology and Head of the Laboratory for Molecular Neuropathology in the University of Heidelberg. Working in close collaboration these two investigators have made major advances in knowledge of the molecular biology and chemistry of the abnormal amyloid tissue that characterizes the development of the widespread condition known as Alzheimer's disease. This disease causes dementia and death in over 20% of adults over 80 and 50% over 90 years of age, as well as affecting many younger people. These investigators have identified a protein known as 13A4 that is a major component of the amyloid plaques and have shown that a gene on chromosome 21 encodes for this protein which is part of a lamel protein called amyloid precursor protein (APP). They later studied the regulation of the synthesis and function of APP and its ability to bind to
metallic ions. They hypothesized that the abnormal accumulation of pA4 protein underlies the neuronal changes that lead not only to Alzheimer's disease but also to other degenerative diseases such as Creutzfeldt-Jakob disease. Their research has opened the way to the rational development of novel drugs that can interfere with these pathological processes and which it is hoped will offer some chance of limiting or ameliorating these devastating diseases in the near future. Professors Masters and Beyreuther have published 124 joint papers relating to this field as well as numerous other individual papers.

A rarer but equally distressing condition is Huntington's disease which, developing in early adulthood, results in totally disabling motor disorders functional and psychiatric changes. Professor Francis James Gusella, a Canadian national born in Ottawa in 1952, obtained his PhD in biology at the Massachusetts Institute of Technology in 1980 and is currently the Bullard Professor of Neurogenetics in Harvard Medical School, Boston. In 1983, employing the then new techniques of "reverse genetics", Professor Gusella and his associates succeeded in mapping the gene locus for Huntington's disease (HD) on the human chromosome 4. Ten years later he identified the gene locus which encodes for the protein that he called Huntingtin. This seminal discovery enabled him to lead a major collaborative investigation that has culminated in the unraveling of the detailed structure of the HD gene and the expanded CAG repeats that underlie the neurodegenerative changes that characterize this condition. This work has made possible the prenatal and presymptomatic testing of individuals which, in turn, provides the opportunity for genetic counseling in families with a history of BD. Subsequently Professor Gusella and his team were able to identify the chromosomal localization of genes involved in two other neurodegenerative conditions, Alzheimer's disease (on chromosomes 21 and 14) and familial amyotrophic lateral sclerosis (on chromosome 21). He and his team are now developing animal models in which to investigate further HD and other hereditary neurodegenerative conditions. In parallel with this pioneering work Professor Gusella has made an outstanding contribution to the training of large numbers of research workers and his laboratory has acquired a unique international reputation as a focus for neurobiological research. Professor Gusella and his colleagues have published 292 papers as well as 78 reviews, symposium contributions and chapters in books.