## Nuclear battle (Contd. from P. 4)

fective in stopping the Communist takeover of Europe and that policies would continue to change and adapt in the future.

"Deterrence buys time for the politicians and diplomats to work it out around the table, rather than over the top of the world," said Lane.

Lane rejected the idea that the arms race was out of control, saying that the limitations in SALT (Strategic Arms Limitation Talks) agreements were being adhered to.

Epstein, however, said that the SALT agreements actually provided no limitations at all. He said that prior to the SALT talks, the U.S. had an overkill capacity of some 36 times, while Russia's was 11 times.

After SALT, these overkill factors rose to 50 and 20 times respectively, he said.

The present policy of deterrence developed from the military wondering "what the hell are we going to do with all that overkill?"

"The drive to infinity-killing power is what will kill us all," said Epstein.

While both debaters agreed that nuclear proliferation is a danger, Lane said one must assume there was a degree of rational behaviour behind proliferation. "The UN could bring enormous pressure to bear on small powers who try to use the weapons," he said.

(From an article by Les Leyne in the March 30, '77 issue of The Ring.)

# Breakthrough in identifying the cause of Batten Disease

Under the leadership of a neurochemist at the Montreal Neurological Hospital and Institute, a McGill University teaching hospital, a team of neuropathologists, neurologists and geneticists have made a key finding which will result in the identification of the cause of Batten Disease. This is an inherited disease which causes severe cerebral degeneration, convulsions, blindness and ultimately the death of afflicted children before they reach the age of ten.

Named after an English neurologist who clinically described it in detail at the turn of the century, Batten Disease is a hereditary condition with a clearlydefined clinical course associated with severe brain degeneration.

There are currently over 50 reported cases of Batten Disease in the province of Quebec, and it has also been found in Newfoundland, New Brunswick and Nova Scotia, as well as in many Eastern American states. In Europe it is particularly common in Scandinavia, Britain, Denmark and Holland. Unlike Tay Sachs Disease, another inherited metabolic and degenerative disease of the nervous system, Batten Disease is considerably more common, widely distributed and unrelated to any particular ethnic group. Until recently, apart from a clinical and pathological description and many theories, the cause of it was unknown.

Dr. Leonhard Wolfe is a neurochemist and head of a McGill University team of researchers including chemist Dr. N.M.K. Ng, Ying Kin, neurologist Dr. Frederick Andermann, geneticists Dr. Eva Andermann and Dr. Charles Scriver, and neuropathologist Dr. Stirling Carpenter, who have spent over five years searching for the cause of Batten Disease. They have succeeded in identifying the molecule which appears to be its basic cause. They were aided by Dr. A.S. Perlin's Department of Industrial and Cell Chemistry of the Pulp and Paper Research Institute.

### A normal child at birth

In most cases it is not before they reach three years of age that children display the first symptoms of Batten Disease. Usually the onset of the disease is marked by the occurrence of an epileptic seizure. The disease progressively worsens as the seizures become more and more frequent and difficult to control. There ensues progressive intellectual deterioration, visual impairment and finally the child becomes demented and suffers so severely from mental retardation and degeneration that death occurs, some time before the age of ten.

### Cause

Batten Disease is caused by the massive accumulation of a material in the child's neurons (nerve cells) the nature of which was unknown until recently. The success of the work of Dr. Wolfe and his team of scientists lies in the identification of the composition of this material.

Ceroid, the non-specific name frequently applied to the material present in the neurons of Batten Disease victims, is peculiar in that it is autofluorescent, i.e. it gives off a brilliant greenish-yellow fluorescence when exposed to ultraviolet light. In fact, sections of the brain of a child who has died from the disease need not be stained prior to examination under an ultraviolet microscope. Almost all the neurons will be visible owing to their fluorescence. This is extremely unusual and abnormal. Scientists in general agree that this fluorescent material (ceroid) is the primary storage material causing the degeneration of the nerve cells on a massive scale as its accumulation increases. The disturbed neurological functions are directly related to this process. This storage phenomenon occurs not only in the brain but in many other tissues as well. However, the basic cause of death is neurological.

Diseases which involve the storage of chemical substances in special particles in the cells (lysosomes) are often caused by the absence of activity of a key enzyme which is responsible for the metabolism of the particular material. It is generally felt, therefore, that the first step in discovering the cause of these diseases is to identify the chemical nature of the accumulated material. This has provided the clue in the past ten years to the nature of the enzymatic defect in a number of other diseases of this type, including Tay Sachs Disease. In the case of Batten Disease, no one knew the specific nature of the autofluorescent material and consequently no one had any clue to the specific enzyme defect.

#### Physical aspects of affected neurons

Over a year ago, Dr. Wolfe and his colleagues studied the brain of a child who had died from Batten Disease and immediately decided to make a fresh attempt to isolate and chemically characterize the storage material in the neurons of the brain. After about six months they managed to isolate from the brain by ultracentrifugation methods (the use of a radially outward force at a very high speed to separate substances according to weight) a very pure subcellular fraction which contained particles of identical structure