INVERTEBRATES

Discovery in Devon

THESE fearsome little creatures have been discovered between the sand grains of the Exe Estuary in South Devon during an ecological survey of free living nematodes. These two, the male (a) and female (b) of *Enoplolaimus denticulatus*, belong to one of fourteen new species of nematodes described from the estuary by R. M. Warwick (*Bull. Brit. Mus. (Nat. Hist.) Zool.*, 19, No. 4; 1970). On the basis of Warwick's measurements of six specimens of *E. denticulatus*, the heads in the picture are likely to be about 0.05 mm in diameter. Males were on average 3.7 mm long and females 4.3 mm long; this is apparently a relatively large member of the genus.

METABOLIC DISEASES

More Clues from Enzymes

from our Medical Biochemistry Correspondent

NEW methods of diagnosis may be possible for two serious conditions involving inborn errors of complex lipid metabolism. They could stem from the results of enzyme assays of cases of Fabry's and Tay-Sachs diseases.

Fabry's disease seems to result from a lack of galactosyl hydrolase, which acts on the glycosphingolipid galactosyl galactosyl glucosyl ceramide, known as GL-3. Patients with this disease accumulate GL-3

in their plasma, urine and most tissues. The enzyme is found in a particulate fraction in normal spleen, small intestine, kidney, brain and liver; it is absent from the mucosa of males with Fabry's disease and present in smaller than normal concentrations in tissues from heterozygous females (this disease is sex linked). Mapes *et al.* (*FEBS Letters*, 7, 180; 1970) have found that normal human plasma contains at least one enzyme with this activity, whereas the plasma of patients contains no detectable GL-3 galactosyl hydrolase activity.

In the plasma of four normal subjects they found maximum GL-3 galactosyl hydrolase activity at pH 5.4 and 7.2, which probably means that two different enzyme molecules catalyse the same reaction. The optimum activity at pH 5.4 is similar to that found in the tissues; the four normal subjects all had about sixteen units of activity in their tissues at this pH. In the patients there was no activity in the plasma at either pH, which means that their plasma could not contain more than 1.5 enzyme units. The activity of GL-3 galactosyl hydrolase in the plasma not only provides an easy method of diagnosis, but also suggests that the condition could be treated. If the normal plasma enzyme activity could be restored artificially the accumulation of glycosphingolipid in the tissues might be prevented.

Tay-Sachs disease is a similar condition which is usually rapidly fatal, and Schneck *et al.* (*Lancet*, i, 582; 1970) have shown that it can easily be diagnosed before birth. They investigated a subject who had given birth to a child who died from Tay-Sachs disease, due to the absence of hexosaminidase A activity—the enzyme hexosaminidase is normally present as fractions A and B. They removed amniotic fluid at fifteen and twenty weeks in a later pregnancy and assayed it and uncultured foetal cells for hexosaminidase A activity, which turned out to be present in trace amounts only. By contrast, uncultured cells from a normal subject contained 28 per cent hexosaminidase in the A form. When the pregnancy was terminated the foetal brain contained 4.5 times as much of the ganglioside GM2 (which is cleaved by hexosaminidase A and B) as a normal foetal brain at fifteen weeks.

The results of the enzyme assays show that it is easy to detect Tay-Sachs disease according to the level of activity of hexosaminidase A at a relatively early stage in pregnancy without the need to culture foetal cells. Schneck *et al.* point out that if further work substantiates their findings effective genetic counselling will be available for the first time for mothers of infants with Tay-Sachs disease.

BRAIN CELLS

Neurons that Count

from our Neurophysiology Correspondent

A TEAM at the University of California claims that the cat's brain contains cells which count. R. F. Thompson, K. S. Mayers and C. J. Patterson (*Science*, 168, 271; 1970) have found that certain cells in the "associative cortex" apparently respond to a particular stimulus in a sequence of stimuli, for example, the fifth or seventh. Thus these cells seem to be coding for number.

The "receptive field", a concept introduced by Adrian in 1929, has become the investigational unit