

to those seen by electronmicroscopy in affected neurons. The particles examined by electronmicroscopy were composed of tiny curved lamellated structures given the name curvilinear bodies by the pathologists.

These isolated curvilinear bodies were intensely fluorescent and their fluorescence was insoluble in all common organic solvents. Moreover, they had exactly the same structural features as the bodies seen in neurons of the intact brain.

Solving the autofluorescence puzzle

After a number of efforts lasting several months, Dr. Wolfe and his group in the Neurochemistry Department found that the autofluorescent material was a derivative of Vitamin-A, which becomes physically associated with phospholipids and cholesterol. The chemical name for the Vitamin-A derivative is retinoic acid, known to be a highly fluorescent compound. Retinoic acid, one of three Vitamin-A compounds, is involved in the growth, maintenance and differentiation of cells. The other two, Vitamin-A alcohol and aldehyde, are necessary for the formation of visual pigment. Victims of Batten Disease display no Vitamin-A deficiencies. In the affected neurons, retinoic acid is present in a complex with a small peptide. The indications of the unique presence of retinoic acid led Dr. Wolfe and his colleagues to seek more direct chemical proof for the nature of the material.

Through a series of chemical reactions and physico-chemical methods, the group was able to indicate clearly that a retinoic acid derivative was the autofluorescent material in the affected cells of the patients. They were able to match the spectra of retinoic acid and methyl retinoate (derivative of retinoic acid) derived from the patients with authentic spectra of these compounds. "The realization that retinoic acid complexes accumulate in the brain," Dr. Wolfe suggests, "can reveal with further research that the basic enzyme defect lies somewhere in the pathway of the metabolism of retinoic acid."

Although they are still trying to identify the defective enzyme, members of the research team are now able to detect an excess of retinoic acid derivative in the urine of Batten

Disease children which has similar properties to the material stored in the brain. Dr. Wolfe is currently working on a method for the diagnosis of Batten Disease through urine and blood analysis in order to determine the presence of retinoic acid derivatives in above normal amounts. An important object of this research is to detect not only cases but also carriers of Batten Disease. For example, where a sibling already suffers from the disease, a skin biopsy of the younger children would show whether or not they are likely to suffer from the condition.

Moreover, Dr. Wolfe believes that there is a distinct possibility that Batten Disease could be controlled through the restriction of Vitamin-A intake before the onset of symptoms. The team is now working on the development of a dietary therapy.

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Heart's content remembers

The provincial historical site at Heart's Content, Newfoundland, is a vivid reminder of how Newfoundland was involved in the development of communications. On the site is an interpretation centre recently opened by the provincial government, which is filled with memorabilia pertaining to the history of telegraphy and transatlantic cables.

The little Avalon Peninsula community, the oldest town in Trinity Bay, is 137 km (85 miles) from St. John's, the capital.

The first lodge of the Society of United Fishermen was opened at Heart's Content in 1873 and, today, it is still a quiet fishermen's town with the interpretation centre its chief tourist attraction.

More than 100 years ago, the then-tiny village of Heart's Content was a hive of activity as the first successful transatlantic telegraph cable reached shore.

But it had not been easy. It took millions of dollars and 15 years before final victory was achieved.

In 1858, a cable was laid across the

Atlantic and its completion raised cheers on both sides of the ocean. But the rejoicing didn't last. After three months, the cable insulation failed.

Seven years later, in 1865, another attempt was made. All went well for several days but, 1,932 km (1,200 miles) out to sea, the cable broke.

Undaunted, the organizers tried again the following year, this time with a newly-built liner *Great Eastern*, a ship that made history, which could store and release cable easily and safely.

So, with British funds, a British crew and a staff of scientists that included one American, Cyrus Field, it sailed from Valentia, Ireland, July 13, 1866.

For Cyrus West Field, the transatlantic cable was an obsession. He was one of the founders of the New York, Newfoundland and London Telegraph Co., formed for one purpose: the laying of a cable across the Atlantic Ocean.

The job took only two weeks. The *Great Eastern* laid anchor opposite Heart's Content on July 27 after a 3,436-km (2,134-mile) historic journey.

After a well-deserved rest, the *Great Eastern* crew returned to mid-ocean where the cable broken the previous year was recovered, spliced and stretched to Heart's Content.

What had seemed a failure in 1865 had become an asset. The company now had an insurance cable for its communications between the two continents.

Canada-France cable

In 1869, the French Atlantic Cable Co. laid a cable from Brest, France, to the Island of St. Pierre, off the coast of Newfoundland, which became the first Canada-France cable.

It was only in 1956 that the first transatlantic telephone cable was laid, from Clarenville, Newfoundland, to Oban, Scotland.

Newfoundland, because of its relative proximity to Europe, also boasts the site of the first transatlantic wireless signal, received by Marconi at St. John's in 1901 and the field, also at St. John's, from which Alcock and Brown left on the first successful airplane flight across the Atlantic.

A museum at the airport in Gander, Newfoundland, relates the history of early attempts to fly across the ocean.