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José Ramón Ricoy, neuropathologist

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ABSTRACT

José Ramón Ricoy, who studied under Ludo van Bogaert at the Bunge Institute in Antwerp, is a very influential professor who has trained specialists in general anatomical pathology and neuropathology throughout his professional career.

He has published more than 130 scientific papers in both national and international journals. Stellar contributions include his 1974 doctoral thesis on orthochromatic (or sudanophilic) leukodystrophies. They also include his studies of other leukodystrophies, Pelizaeus-Merzbacher disease, mitochondrial behaviour in myopathies, and CSF and serum studies in patients with subacute sclerosing panencephalitis. Additional examples are his histochemical and ultrastructural studies of striated muscle fibres in Prader-Willi syndrome, the case series of neuronal ceroid lipofuscinosis or Batten disease, and his clinical and neuropathological examination of progressive myoclonus epilepsy of Unverricht-Lundborg. I should also mention his contribution to the study of toxic oil syndrome, a neurological disease of great social concern that struck Spain in 1981.

This article also presents the anatomical pathology collection which the doctor recently donated to the SEN's historical archive. This collection consists of 56 histological slides illustrating various nervous system diseases. It is particularly interesting from a didactic point of view since it showcases rare diseases and an excellent slide preparation technique. Key examples are the slides presenting Huntington disease, tuberous sclerosis, and Binswanger disease because their image quality is especially sharp.

KEYWORDS
José Ramón Ricoy, neuropathology, Huntington disease, tuberous sclerosis, Bourneville disease, Binswanger disease

Introduction

José Ramón Ricoy Campo was born in 1942 in As Ermidas, Ourense province. He attended medical school in Santiago de Compostela and graduated in 1966. Neuropathology was his calling from an early age. A year after earning his medical degree he began working at the Bunge Institute in Antwerp, where he was trained in that specialty by Ludo van Bogaert in the three years from 1967 to 1970.

He rapidly acquired a solid background, which is exemplified by his study, with J.H. Bruens as co-author, of a case of familial amaurotic idiocy. The conclusions of this article address the challenges in characterising this myoclonic variant of non-ganglioside familial amaurotic idiocy according to its clinical and pathological features. Based on cellular neuropathological findings, the authors highlight the selective localisation and nature of build-up in neurons.[1]

Ricoy returned to Spain and was hired as an associate professor of anatomical pathology by Universidad Autónoma, Madrid, in 1973, and in 1976, by Universidad Complutense. He held these positions until 1974 and 1984, respectively.

In 1985, he was made a senior lecturer in pathology at Universidad Complutense, Madrid, later becoming the chair of anatomical pathology at that university's Faculty of Medicine.

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Lines of research

Dr Ricoy Campo has presented more than 30 oral communications on a wide variety of diseases at the annual meetings held by the Spanish Society of Neurology (SEN). His contributions include descriptions of leukodystrophies, Pelizaeus-Merzbacher disease, mitochondrial behaviour in myopathies, and CSF and serum studies in patients with subacute sclerosing panencephalitis. We also find his histochemical and ultrastructural studies of striated muscle fibres in Prader-Willi syndrome, the study of seven cases of neuronal ceroid lipofuscinoses or Batten disease, and his clinical and neuropathological study of two patients with the Unverricht-Lundborg form of progressive myoclonus epilepsy.

Special mention should be made of his first oral presentation, given with A. Vázquez and G. Moya at the 1969 SEN Annual Meeting, on the neurological aspects of hypoglycaemic comas. Here, a neuropathology study of two autopsy cases revealed mild neural loss with homogenising necrosis and diffuse demyelination, with no accompanying glial reaction.[2]

In the same meeting, he also presented his own study of the description of the myoclonic variant of familial amaurotic idiocy. He concluded that this variant was simply a less severe form of the histopathological process that takes place in diseases of this type.[3]

At the 1970 annual meeting, Ricoy was the lead author and presenter of a comparative study of the ultrastructure of striated muscle fibres in two motor neuron diseases: amyotrophic lateral sclerosis (ALS) and Wohlfart-Kugelberg-Welander (WKW). Here, he presented the results from a study of striated muscle in ten cases of ALS and in one patient with WKW. The conclusions from this study are that in ALS, the observed myofibrillar alterations are secondary to Z-band changes that indicate denervation. In WKW disease, the entire myofibrillar complex becomes disorganised, whereas Z-band changes are secondary and manifest differently.[4]

In 1971, he co-authored a case study of McArdle disease (glycogenesis V). The authors believed that this case study was probably the first one published in the world, and they concluded that in addition to the characteristic alterations in glycogen storage, there was also an active myopathic process.[5]

Dr Ricoy has published more than 130 scientific papers in Spanish and international journals. His remarkable doctoral thesis, defended in 1974, addresses orthochromatic (sudanophilic) leukodystrophies.[6] This thesis reviews the concept of diffuse sclerosis and distinguishes three subtypes: myelinoclastic diffuse sclerosis (Schilder's disease), van Bogaert encephalitis, and the leukodystrophies. Within this last category, only the orthochromatic or sudanophilic varieties are considered true leukodystrophies. Ricoy studied these entities using the classification system set out by Peiffer, and proposed 2 types: pure forms and special forms (Pelizaeus-Merzbacher disease and variants).

The pure forms include cases in which the underlying disease consists of a change in the endoplasmic reticular system of oligodendroglia. This gives rise to alterations in the makeup or maintenance of myelin. In special forms, however, microscope imaging reveals patchy demyelination, limited axonal lesions, and a scarce or absent astroglial and sudanophilic reaction. Remarkable ultrastructure findings in these forms are absence of normal myelin, degeneration of myelin sheaths, spiral wrapping in periaxonal membrane, limited axonal degeneration, oligodendrocyte deficit, and swollen astrocytes poor in glial filaments and containing lipid bodies.

I should also point out his contributions to the study of toxic oil syndrome, a neurological disease of great societal concern that struck in Spain in 1981. It affected approximately 20 000 people and caused 330 deaths. The study published in *Brain* in 1983 distinguishes between lesions in muscle tissue, peripheral nerves, and the central nervous system.[7] It also describes two different and time-dependent types of muscle system lesions. Patients initially presented myalgia and inflammatory infiltration in the perimysium, the muscle spindle capsules, and the intramuscular nerves. Muscle tissue displayed electron-dense material similar to Z-bands. Observations at later stages of the disease also included pronounced neurogenic amyotrophy with endomysial fibrosis. The onset of denervation atrophy is secondary to impairment of peripheral nerves, which is one of the distinguishing features of this disease.

The most obvious lesion in this syndrome was impairment of the peripheral nerves; researchers observed perineuritis and perineural fibrosis as typical features in toxic oil syndrome.

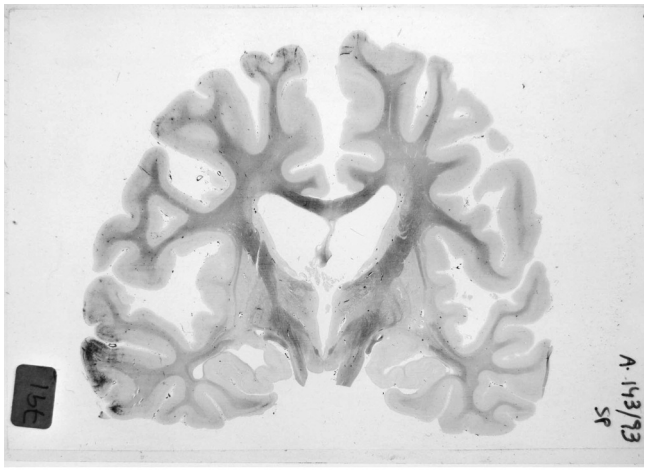
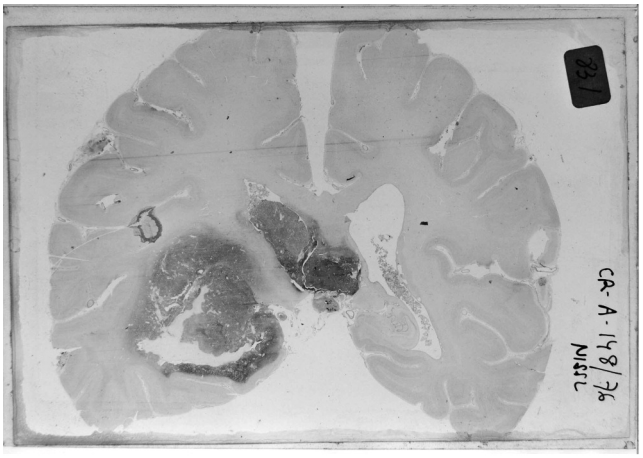
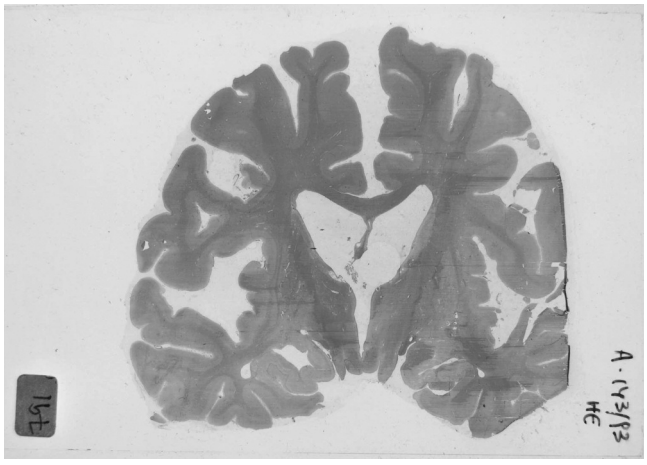


Figure 1. Huntington Disease





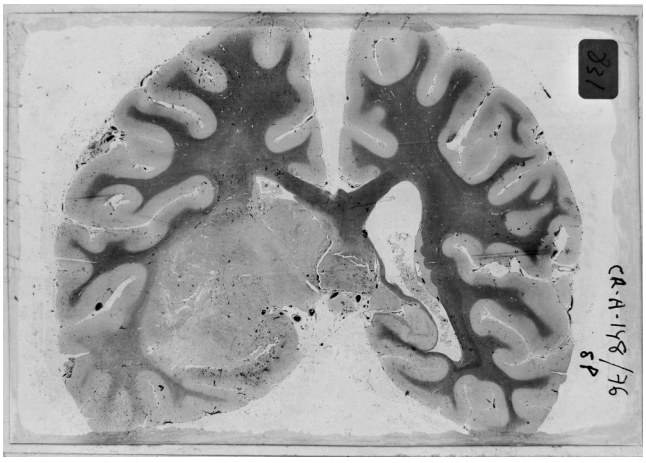


Figure 2. Tuberous sclerosis or Bourneville disease

The central nervous system consistently displayed lesions in cells of the anterior horns, and in the nuclei of cranial nerves and reticular neurons. Astrocytes of the brainstem showed hypertrophy and abnormal nuclei. The study also found areas of chromatolysis in locations with microglial proliferation.

The adulterated oil had a high linoleic acid content. This led to an excess of arachidonic acid, which may have played a role in lesion pathogenesis.

In addition to his clinical, diagnostic, and research contributions, José Ramón Ricoy was outstanding as a professor. He trained specialists in general anatomical pathology and neuropathology during his career as a neuropathologist at several hospitals (Gran Hospital del Estado [1970-1973], Clínica Puerta de Hierro [1973-1976], and Hospital Universitario 12 de Octubre from 1985 until his retirement).

Between 1983 and 1996, he developed his considerable abilities as an administrator and manager while directing Hospital Universitario 12 de Octubre. He served as the Deputy Director General of Spain's National Institute of Health, and also directed the Spanish health research fund (*Fondo de Investigaciones Sanitarias*), the research and education management strategy (*Ordenación de la Investigación y Formación*), and Institute of Health Carlos III.

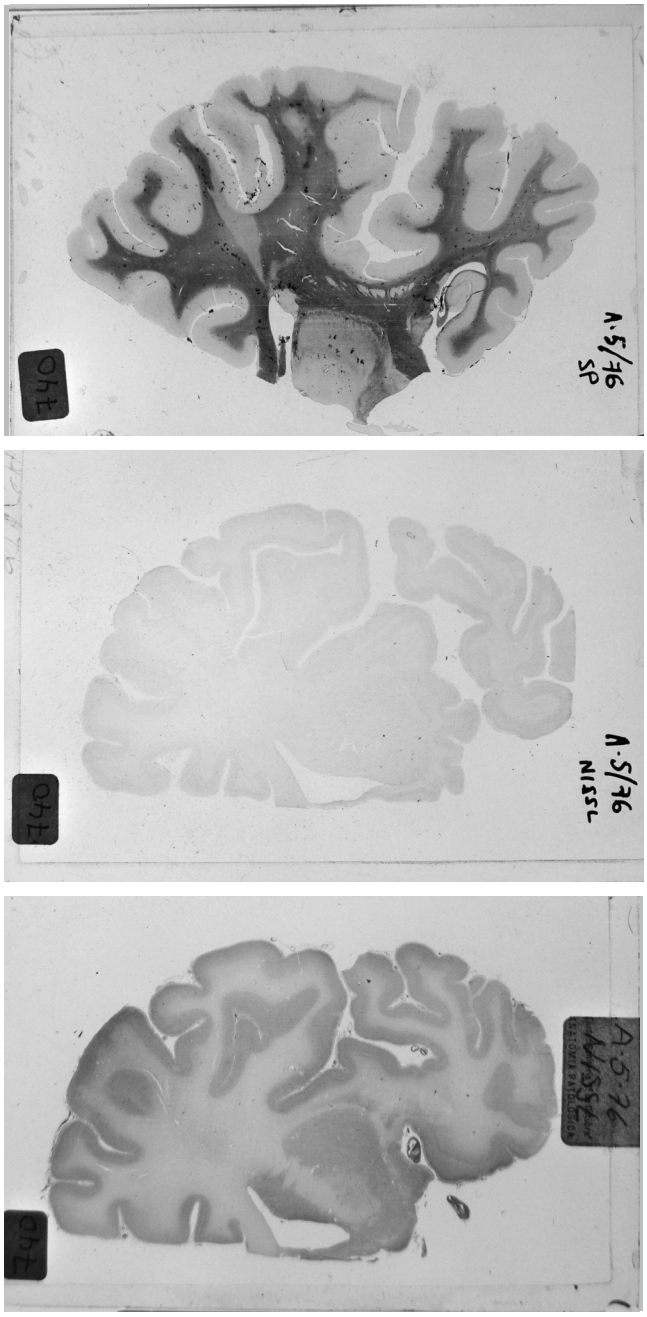


Figure 3. Binswanger disease

Anatomical pathology collection

José Ramón Ricoy has donated a collection of 56 histological slides illustrating different pathological findings in the nervous system to the SEN historical archive.

This collection is particularly interesting from a didactic point of view, since it showcases rare diseases and the technique used to prepare the slides is excellent.

Key examples are the slides presenting Huntington disease, tuberous sclerosis, and Binswanger disease because the quality of the images that illustrate these rare diseases is especially sharp.

— Huntington disease (Figure 1): the coronal section shows diffuse cortical atrophy and pronounced atrophy of the caudate nucleus of the globus pallidus and putamen. Atrophy of the basal ganglia results in an increase in ventricle size.

— Tuberous sclerosis or Bourneville disease (Figure 2): this coronal section of the brain exhibits glial nodules arranged in rows. Some invade the ventricular spaces and they also form multiple hardened areas of the cerebral cortex. Nodules are formed by very large subpial astrocytes and enlarged neurons. Tumours evolve from these nodules, and some display characteristics of spongioblastomas. The disease profile also includes presence of sebacious adenomas, rhabdomyomas in the myocardium, and multiple renal cysts in the fetus.

— Binswanger disease (Figure 3): coronal section showing foci of myelin destruction in the white matter of the temporal lobe and widening of the horns; both processes are secondary to white matter atrophy. The cortex and subcortical fibres remain largely intact.

Acknowledgements

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