Friedrich Hugo Kufs (1871–1955)

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Hugo Kufs [1, 10] lent his name to the eponymous Kufs disease, the adult form of different lysosomal storage diseases that have now been grouped under the common term neuronal ceroid lipofuscinosis (NCL). Kufs was born on December 16, 1871, into the family of a minor landowner in Großzössen. He enrolled at the nearby Leipzig University to study medicine, and then worked as assistant surgeon at the Chemnitz Municipal Hospital and Brandenburg County Asylum in Eberswalde before changing over to the

Saxon asylums at Pirna-Sonnenstein, Hochweitzschen, and Hubertusburg in 1900. During World War I, he was not conscripted into the army, as he suffered from rheumatism. On the basis of his autodidactic studies and his work on brain histopathology, he was appointed prosector and head of a newly established laboratory for brain pathology at the Leipzig-Dösen Municipal Asylum in 1919. Due to the lack of competent staff at the histopathological laboratory at the neuropsychiatric department of Leipzig University, headed by Paul Schröder, Kufs also supervised work carried out there and was subsequently awarded the title of professor in 1925. Even at this early stage of his career, experts testified to Kufs's excellent reputation. Walther Spielmeier, head of the Neuropathological Department of the German Research Institution for Psychiatry in Munich, called him an "extremely talented histopathologist", whose works were "extraordinarily scrupulous", "simply excellent" or, with regard to his study of focal degenerative processes in nerve fibers, "of importance for pathology in general". Spielmeier even admitted that he had tried in vain to recruit Kufs for his institute. Likewise, Hamburg neurologist and neuropathologist Alfons Maria Jakob counted Kufs "among the best experts in the histopathology of the central nervous system in Germany", who had "a good feeling for important questions". He "admired" the "clarity with which he formulated the problem, the critical evaluation" and his "profound knowledge of the literature" [10].

During the 1910s and 1920s, Kufs gained prominence with his studies on tuberous brain sclerosis and on the differential diagnosis of neurosyphilis. Through his investigation of a family with cerebral, retinal, and cutaneous cavernous malformations, he pioneered the description of hereditary angiomas of the brain [5] and he also provided new insight into the hereditary character of the

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Von-Hippel-Lindau syndrome [2]. Amaurotic idiocy, which today is part of the NCL group, also intrigued Kufs for decades. NCL is probably the most common variety of progressive neurodegenerative disease in children, but it is rather rare in adulthood. His three descriptions of inherited forms with later onset made Kufs the name-patron of the adult variant of the syndrome (CNL 4) [4, 6, 7]. In these studies, he identified most clinical features of the late forms of NCL acknowledged today. Kufs also described the protracted course of the disease, spanning many years until the patient's death, after decades of normal mental and neurological behavior. Apart from the comprehensive clinical descriptions, his histological findings on the disease were stunning. Kufs recognized that the disease represented a disturbance in lipid metabolism and identified morphological changes in different parts of the body and organs. In particular, Kufs discovered the extensive storage of ceroid-lipofuscin in the central nervous system. He also investigated the relationship between amaurotic idiocy and Niemann–Pick disease. The nosological classification of Kufs disease and related disorders has been a major point of discussion for a long time. Over the past 15 years, numerous gene mutations have been identified and recent biochemical and molecular genetic findings have contributed to the classification and pathogenesis of this devastating brain disorder [3].

In 1936, Kufs, who was never a member of the National Socialist Party or any of its subdivisions, retired, yet continued his scientific work. In 1940, his wife passed away, and in December 1943, his flat was destroyed in an air raid. Together with his daughter, he found accommodation with relatives, yet he was completely destitute. With the support of Richard Arwed Pfeifer, who had also been brought back from retirement and appointed provisional head of the Neuropsychiatric Department of Leipzig University, Kufs was reappointed in 1946 to rebuild the clinic's brain anatomical research institute after its total destruction during World War II. Pfeifer even succeeded in achieving promotion for Kufs, who then became Senior Scientific Assistant and prosector in 1947 [10]. Each year he analyzed some 150 cases, mainly from the Neuropsychiatric Department and its Neurosurgical Ward, but also from other departments of the university hospital. Furthermore, Kufs was approached for advice and testimonials by clinics in both East and West Germany [10]. His later scientific

works include characteristics of multiple and diffuse sclerosis. He also identified ova of *Taenia solium* in the brain as the cause of cysticercosis. After 1951/52, there was a clash between Kufs, then 80 years of age, and Pfeifer, 6 years his junior; they heavily insulted each other, testimonials of which have survived in the archives. In their dispute, Kufs accused Pfeifer of scientific incompetence in histopathology as well as of 'bossy manners' and of stealing his preparations. Kufs even went as far as referring to himself as 'the one and only histopathologist in East Germany'. It is no secret that Pfeifer often proved to have a conceited side. Sources nonetheless suggest that Kufs was quite haughty himself, at least in his later years. In retrospect, both neuroscientists may have been showing signs of old-age obstinacy [8, 10]. Upon Pfeifer's request, Kufs was pensioned off in August 1954. One year later, on September 7, 1955, he died in Leipzig at the age of 84 [1, 9].

Conflict of interest The author declares that he has no conflicts of interest.

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