## PIONEERS IN NEUROLOGY



## **William John Adie (1886–1935)**

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William John Adie, MD Ed, FRCP (Fig. 1) was remembered after his passing as the "Geelong boy who made good in London" by the daily newspaper of his hometown in Australia. He made his name as a prolific neurologist and neuro-ophthalmologist in the early 1900s. He was born in Geelong, a small port city on the southern coast of Victoria, Australia, on October 31, 1886. His passion for medicine became clear when he had a transitory encounter with a physician from his hometown. He eventually moved to the United Kingdom and enrolled at University of Edinburgh to pursue his medical education due to the excessive tuition costs in his home-country. He received a gold medal for his outstanding academic performance and also overachieved in his extracurricular activities as an avid rower.

In 1911, Adie was the recipient of the prestigious McCosh Scholarship with which he gained clinical experience in Berlin, Munich, Vienna, and Paris. He developed his passion in neurology when he worked on the serology of cerebrospinal syphilis and other cerebrospinal fluid-related pathology in the Bacteriology department of the "Rudolf Virchow Krankenhaus" in Berlin with Dr. Liefmann.

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Adie served as a Regimental Medical Officer in France during World War I. He briefly returned home in 1916 to marry Lorraine Bonar, a woman he had met in Edinburgh. They soon had a son and daughter. He eventually returned to the battle front to serve as a neurological specialist and became a consultant for the management of traumatic head injuries. Soon after the war had concluded, Adie became a medical registrar at Charing Cross Hospital in London, where he met Gordon Holmes, a fellow neurologist who soon became his friend. Adie eventually practiced as a neurologist at the Royal Northern Hospital, National Hospital for Nervous Diseases, Mount Vernon Hospital, Northwood Hospital, and Moorfields Eye Hospital. After a decade of clinical practice, in 1929, Adie joined the British Medical Association as the secretary of the Section of Neurology and Psychological Medicine. Subsequently, in 1932, at a meeting at the house of Gordon Holmes, Adie co-founded the Association of British Neurologists.

Adie's most recognized accomplishment came in 1931 when he published his findings on a clinical syndrome involving a dilated, poorly reactive pupil (i.e. a tonic pupil) with reduced or absent deep tendon reflexes [1]. By initially reporting six cases, he was one of the first to acknowledge this association. He further bolstered this finding in 1932 when he reported additional cases of tonic pupil associated with absent deep tendon reflexes in his clinical essay for *Brain* [2]. This eventually became known as Adie syndrome associated with the hallmark feature of the tonic pupil, now also known as Adie pupil. In addition, he confirmed the benign nature of Adie syndrome and pupil and rejected the misunderstood equivalence between the Argyll Robertson pupil of syphilis and the tonic pupil [1, 2].

An important point of discussion is the naming of the pupil and the syndrome. A year after Adie's death, Bramwell suggested naming the syndrome as "Holmes-





Fig. 1 William John Adie, MD Ed, FRCP. Courtesy of the Queen Square Archives and Museum

Adie syndrome". This is because both Adie and Gordon Holmes simultaneously, yet independently, provided clinical descriptions of this phenomenon in the same year (1931). This is why the syndrome and the associated pupil are also referred to as the "Holmes–Adie" syndrome and pupil, respectively. It must also be noted that the syndrome had been recognized previously by several individuals. However, Adie is given most of the credit because of his assertion of the benign nature of the disorder and distinction from neurosyphilis [1, 2]. In 1933, Collier was the first to name this phenomenon as "Adie Syndrome", which is the way it is primarily referred to in the United States and Europe. In the United Kingdom, however, Bramwell's suggestion of Holmes–Adie syndrome is preferred.

Adie was also more than productive in his native field of neurology. With James Collier, he co-authored the neurology section in Price's Textbook of Practical Medicine. When he joined Moorfields Eye Hospital, he authored case reports on neuro-ophthalmological topics such as pseudo-Argyll Robertson pupil, pituitary tumours, and the ophthalmic aspects of disseminated sclerosis. During the 1920s and 1930s, he published a number of case reports and clinical essays on various degenerative and autoimmune neurological disorders: congenital amyotonia; progressive familial cerebral degeneration; progressive muscular atrophy; disseminated sclerosis; myotonic dystrophy; familial periodic paralysis; hepatolenticular degeneration; myasthenia gravis; and a plethora of other disorders [3–6]. These publications added a new dimension and valuable epidemiological and clinical evidence in the medical literature for these rare neurological diseases.

In 1921, Adie conducted and published a detailed literary review and explanation of the pathogenesis of tabes

dorsalis. He critically reviewed the work of many experts of tabes dorsalis in an attempt to broaden the understanding of the subject [7].

In 1924, Adie reported on a form of epilepsy in children and interestingly termed it "pyknolepsy" (from Greek, "densely packed"). He described it as "a disease with an explosive onset ... of frequent short, very slight, monotonous minor epileptiform seizures..." [8]. His description of pyknolepsy provided the fundamentals which form the modern-day syndromic complex of Childhood Absence Epilepsy.

In 1923, Adie wrote a detailed clinical essay on myotonic dystrophy. He provided an account of the history, the naming, and the characteristic cataracts of the disease. He speculated about the pathophysiology behind the muscular symptoms of the disease [3].

In 1926, Adie diagnosed and published the youngest case of narcolepsy at the time in a patient of only 12 years of age. It was previously thought to be a disease prevalent only in the adult population. In the same year, Adie also published a large case series of narcolepsy in association with cataplexy in a clinical essay in *Brain* [9]. He described narcolepsy as "attacks of irresistible sleep without apparent cause, and curious attacks on emotion in which the muscles relax suddenly so that the victim sinks to the ground fully conscious but unable to move" [9]. This definition is still in line with the modern understanding of narcolepsy and has been cited on many occasions in the medical literature.

In 1932, by way of a case series, Adie described the causation and presentation of disseminated sclerosis. He studied two sets of patients, one with acute retrobulbar neuritis and the other with disseminated sclerosis. Adie compared the results from these patients to the findings from Russell Brain and other experts on the disease. Together with his critical review of previous descriptions, he synthesized a thorough narrative on disease aetiology and symptomatology [10].

William John Adie's career was short in duration but splendid in quality. His work elucidated the relationship between the tonic pupil and absent deep tendon reflexes, and confirmed its benign nature by distinguishing the tonic pupil from the pupil of syphilis. His many case reports and descriptions provided needed insight into narcolepsy and many other neurological disorders. He was described as a polite, humble, and pleasant individual by his students and colleagues. Adie was invited as a guest speaker to many events where his clinical teachings and lectures were always treasured—he was particularly known for simplifying the most sophisticated topics. On March 17, 1935, Adie passed away at the age of 48, in London, from a myocardial infarction after suffering from angina pectoris for years. Many tributes were written in his honour. William John Adie certainly made his mark and is still



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remembered to this day for his many contributions to medicine.

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**Informed consent** All authors gave their informed consent prior to their inclusion in the study.

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