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on Controversies in Neurology (CONy)
March 22-25, 2018 | Warsaw, Poland

Symposium on Wilson's disease

Saturday March 24, 2018

Kinnier Wilson and Wilson's disease

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Disclosure of interest : Nothing to disclose

French Network on Wilson's disease

Broussolle et al., Rev Neurol (Paris) 2013; 169: 927-35

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History of Neurology

Samuel Alexander Kinnier Wilson. Wilson's disease, Queen Square and neurology

Samuel Alexander Kinnier Wilson. La maladie de Wilson, Queen Square et la neurologie

E. Broussolle[a,*,b], J.-M. Trocello[c], F. Woimant[c], A. Lachaux[d], N. Quinn[e]

Samuel Alexander Kinnier Wilson
(1878-1937)
London, UK

Presentation in two parts

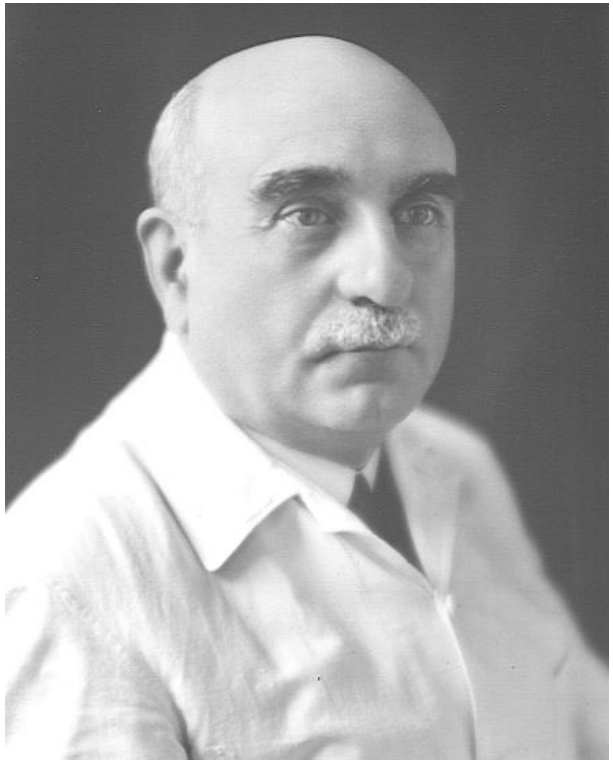
1- Kinnier Wilson the neurologist
2- Wilson's disease

Samuel Alexander Kinnier Wilson, the neurologist

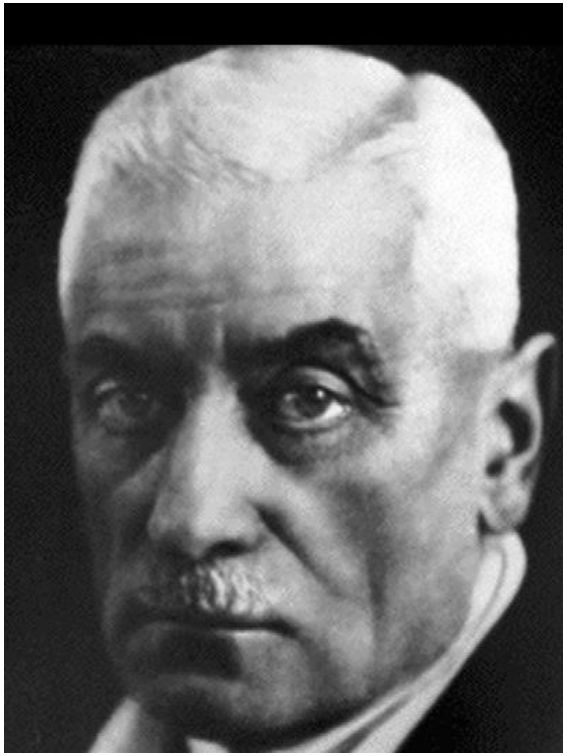
- Born in the USA in 1878, went then to Scotland
- Qualified in Medicine in Edinburgh
- Was inspired to study neurology by Sir Byron Bramwell
- Spent one year in Paris in 1903 where he worked under Pierre Marie (1853-1940) at Bicêtre Hospital, and met other neurologists (Babinski, Crouzon, Lhermitte, Souques, Foix, Meige) and published in French medical journals

Samuel Alexander Kinnier Wilson, the neurologist

Pierre Marie
(1853-1940)



Louis Edouard
Octave Crouzon
(1874-1938)
(craniosynostosis)



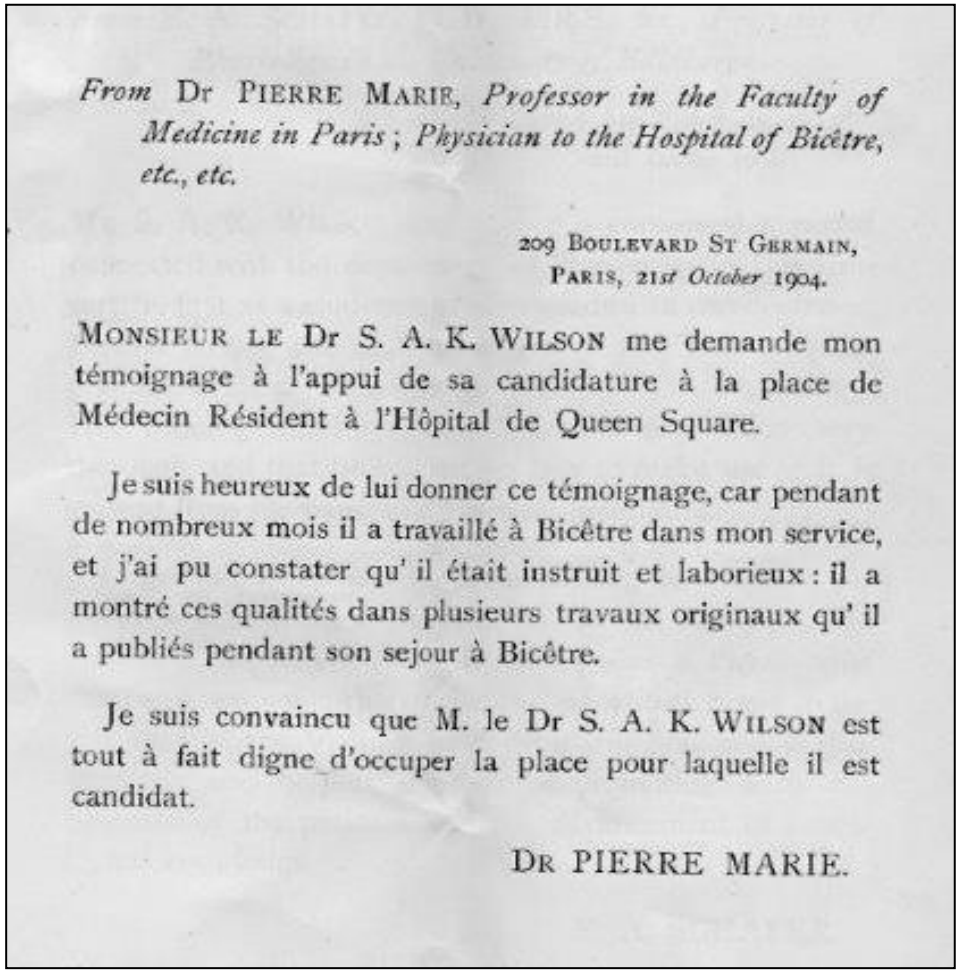
Georges Guillain
(1876-1961)

Samuel Alexander Kinnier Wilson, the neurologist

- Wilson had therafter uninterrupted links with French neurology, particularly with Georges Guillain (1876-1961) who had a remarquable academic career in Paris at the same time as Wilson did in London
- In 1904, Wilson went to Germany and visited in Leipzig the reknown anatomist Paul Flechsig (1847-1929); he thus could learn from German and Central Europe neurological schools

Samuel Alexander Kinnier Wilson, the neurologist

- In 1904, Wilson returned to London with a warm letter of recommendation from Pierre Marie ===>
- He was appointed at the National Hospital , Queen Square, also later at King's College Hospital
- became the first pure neurologist in the UK, and one of the most brilliant and renowned specialists in neurology in the world



From Dr PIERRE MARIE, *Professor in the Faculty of Medicine in Paris; Physician to the Hospital of Bicêtre, etc., etc.*

209 BOULEVARD ST GERMAIN,
PARIS, *21st October* 1904.

MONSIEUR LE Dr S. A. K. WILSON me demande mon témoignage à l'appui de sa candidature à la place de Médecin Résident à l'Hôpital de Queen Square.

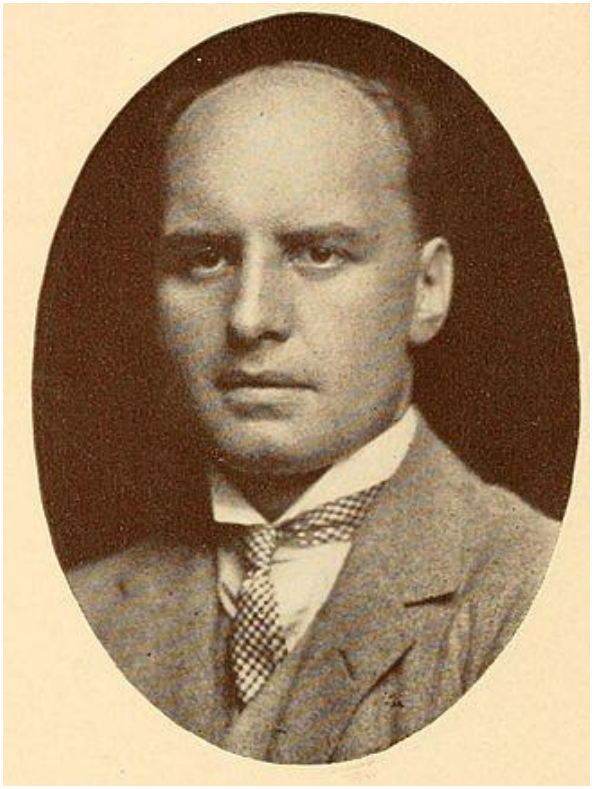
Je suis heureux de lui donner ce témoignage, car pendant de nombreux mois il a travaillé à Bicêtre dans mon service, et j'ai pu constater qu' il était instruit et laborieux : il a montré ces qualités dans plusieurs travaux originaux qu' il a publiés pendant son sejour à Bicêtre.

Je suis convaincu que M. le Dr S. A. K. WILSON est tout à fait digne d'occuper la place pour laquelle il est candidat.

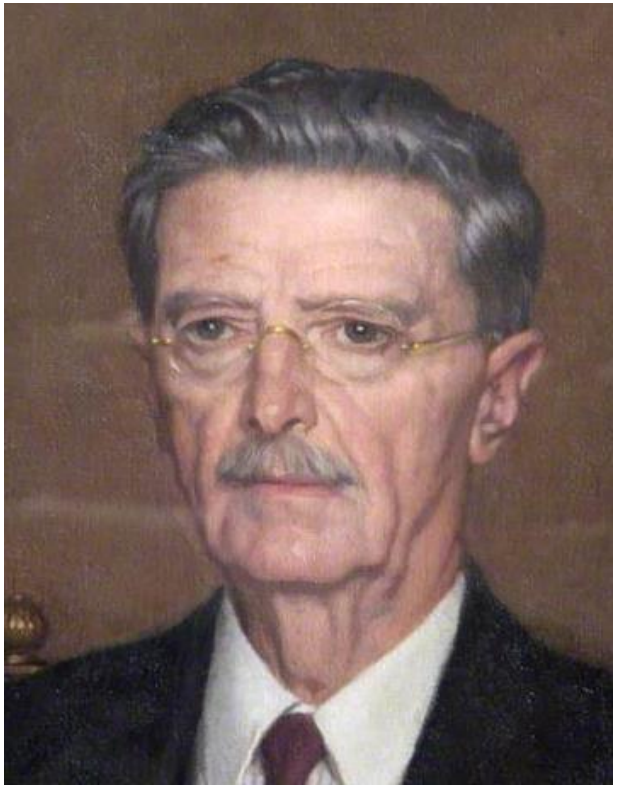
DR PIERRE MARIE.

Samuel Alexander Kinnier Wilson, the neurologist

John Huglings Jackson (1835–1911)



Robert Foster Kennedy (1884-1952)



Gordon Morgan Holmes (1876–1965)

Samuel Alexander Kinnier Wilson, the neurologist

- Wilson had an encyclopedic knowledge, was fluent in French and in German, and thus covered all the neurological literature in English and in the two other languages
- He published extensively, did his medical thesis on progressive lenticular degeneration, founded in 1920 the *Journal of Neurology and Psychopathology* (today JNNP)
- Wilson did Important contributions to the study of disorders of mobiliy and motor tone, epilepsies, aphasia, apraxia, tics and other movement disorders, pathological laughing and crying, …
- He prepared a 2-volume textbook, *Neurology*, published 3 years after his death in 1937 by Dr. A.N.Bruce in 1940

Samuel Alexander Kinnier Wilson, the neurologist

NEUROLOGY

BY

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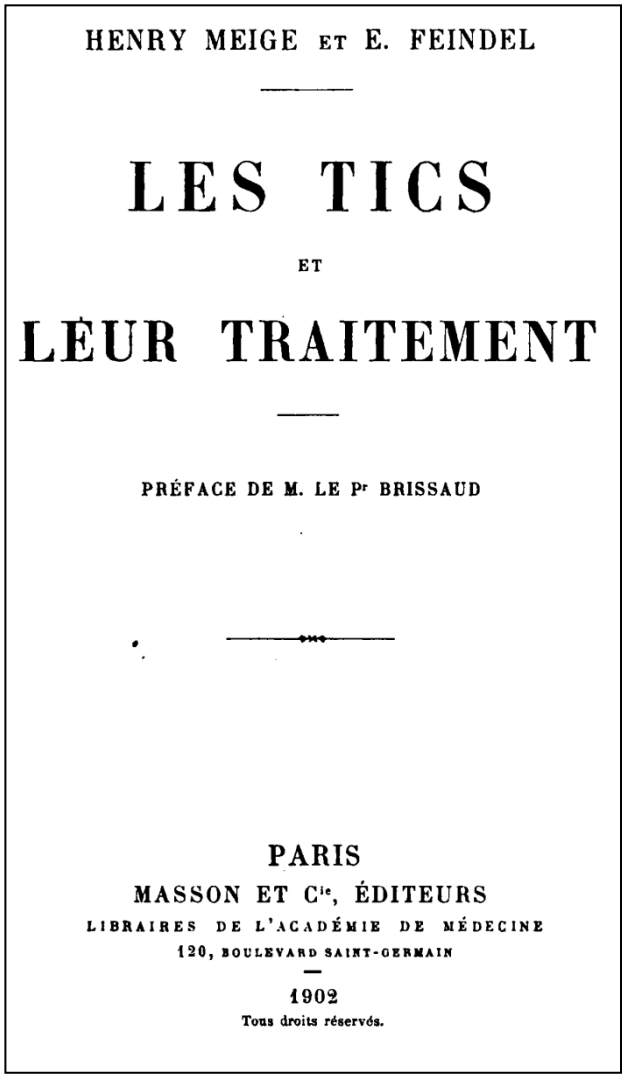
Samuel Alexander Kinnier Wilson, the neurologist

Early special interest in movement disorders



HENRY MEIGE ET E. FEINDEL

LES TICS

ET

LEUR TRAITEMENT

PRÉFACE DE M. LE Pr BRISSAUD

PARIS

MASSON ET Cie, ÉDITEURS

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120, BOULEVARD SAINT-GERMAIN

1902

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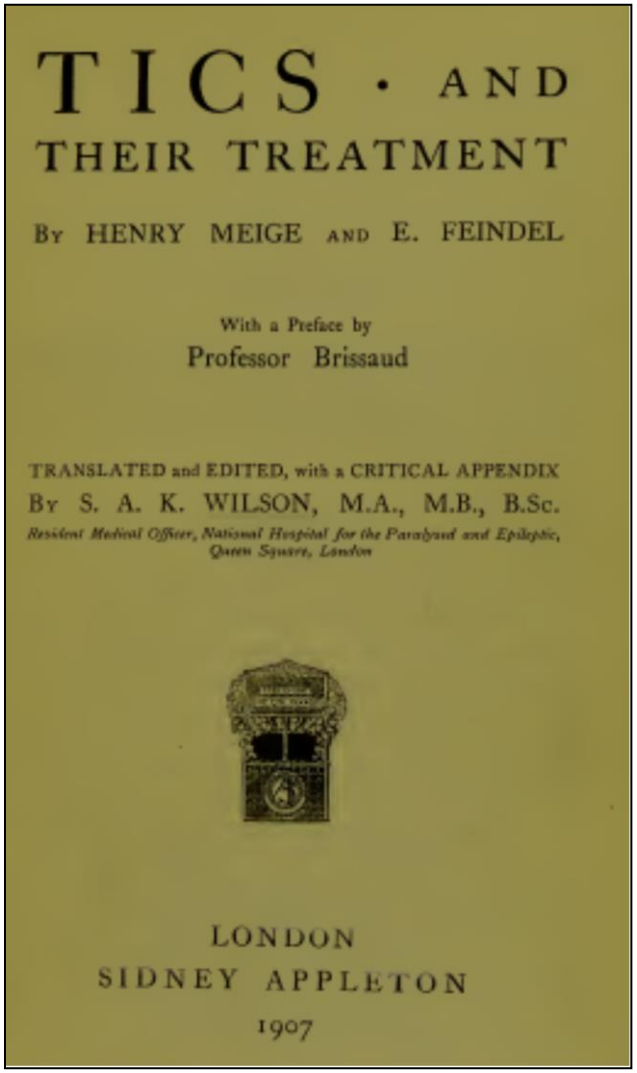

TICS AND THEIR TREATMENT

By HENRY MEIGE and E. FEINDEL

With a Preface by

Professor Brissaud

TRANSLATED and EDITED, with a CRITICAL APPENDIX

By S. A. K. WILSON, M.A., M.B., B.Sc.

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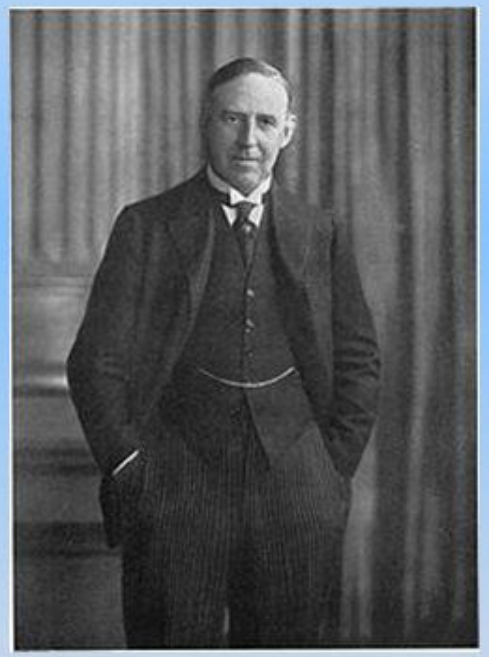
LONDON

SIDNEY APPLETON

1907

Wilson's disease

2012 – centenary of the first publication on Wilson's disease



BRAIN [MARCH, 1912.]

PART IV., VOL. 34.

Original Articles and Clinical Cases.

PROGRESSIVE LENTICULAR DEGENERATION: A FAMILIAL NERVOUS DISEASE ASSOCIATED WITH CIRRHOSIS OF THE LIVER.[1]

BY S. A. KINNIER WILSON, M.D., B.Sc.EDIN., M.R.C.P.LOND.
Registrar to the National Hospital, Queen Square, London.

(*From the Laboratory of the National Hospital, Queen Square.*)

215 pages, the longest paper ever published in Brain

Wilson's disease

Main features of the 12 patients in the 1912 Brain article (including 4 cases W1-W4 followed by Wilson)

Age at onset:10-25 yrs; survival: 5.5 mths-7 yrs; familial cases

Case	Name/Sex	Age at onset (years)	Survival (mo or yrs)	Brain pathology	Liver pathology	Consultant & years of onset & death
H1	Sydney Moor[d]/M	10	5.5 mo	"normal" (macro only)	cirrhosis	W. Gowers 1886–1886
H2	Charlotte Moor[d]/F	15	14 mo	"normal"	cirrhosis	W. Gowers 1887–8
W5	Samuel Moor[d]/M	10	4.5 yrs	no p/m	no p/m	W. Gowers 1874–9
H3	Walter William S/M	10	7–8 mo	lentiform	cirrhosis	J.A. Ormerod 1889–90
H4	Alfred K[e]/M	20	3 ½ yrs	lentiform	cirrhosis	E.A. Homén 1886–90
H5	Wilhelm K[e]/M	12	7 yrs	lentiform	cirrhosis	E.A. Homén 1882–9
H6	Anna K[e]/F	20	6 yrs	lentiform	cirrhosis	E.A. Homén 1882–8
W1[a]	Sylvia Tylor/F	25	4 yrs & 2 mo	lentiform	cirrhosis	D. Ferrier 1904–8
W2	DP[f]/F	17	2 yrs & 8 mo	lentiform	cirrhosis	D. Ferrier 1904–7
W3[b]	EP[f]/M	19	3 yrs	lentiform	cirrhosis	D. Ferrier 1907–10
W4[c]	Charlotte Tolputt/F	18	still living; 22 mo	still living	still living	H. Tooth 1910–
W6	Christopher Johnson/M	11	2yrs	no p/m	no p/m ascites & oedema	J.A. Ormerod 1887–9

mo: months; yrs: years; no/pm: no post-mortem examination; lentiform: lentiform nuclei lesions; H4: Homén's case 2; H5: his case 3; H6: his case 1.

[a] Still living in 1911 with 18 months of follow-up; transient jaundice aged 21.

[b] Psychic troubles onset when aged 17.

[c] Jaundice onset when aged 13.

[d] Sibs.

[e] Sibs.

[f] Sibs.

Broussolle et al., Rev Neurol (Paris) 2013; 169: 927-935

Wilson's disease

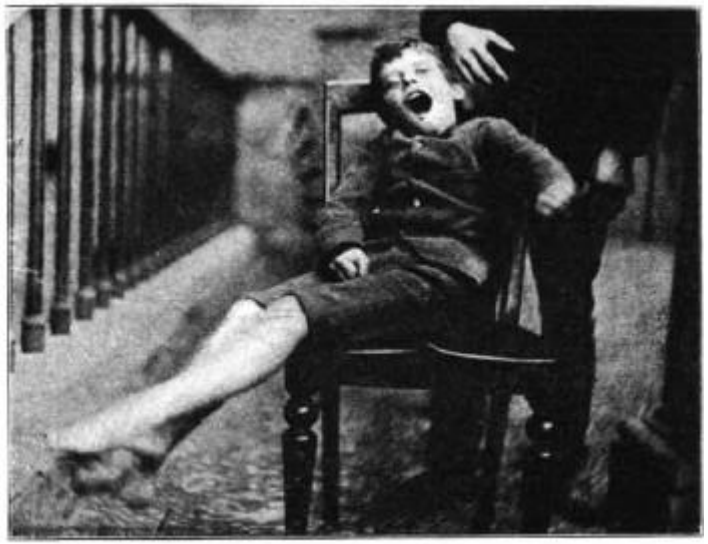
Clinical presentation

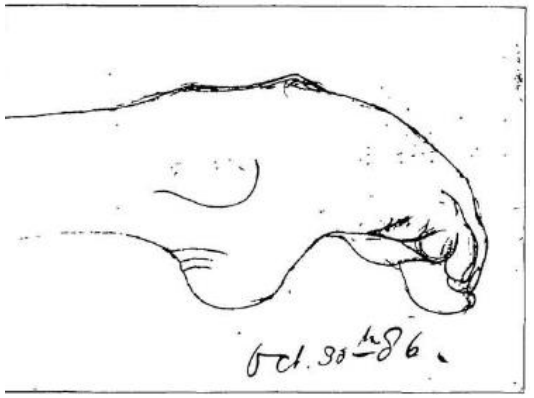
- Abnormal movements: Tremor
- Muscular rigidity
- Contractures/spasms
- Facial involvement/Dysarthria/dysphagia
- Muscular weakness, but no true paralysis nor pyramidal signs
- Emotionalism
- Mental disability, psychosis
- Jaundice
- Progressive emaciation, fatal issue

Wilson's disease

Clinical presentation (Gowers patient, 1887)

=> *contractures, spasms*, ... dystonia appears today as the main feature but this term was not available at that time



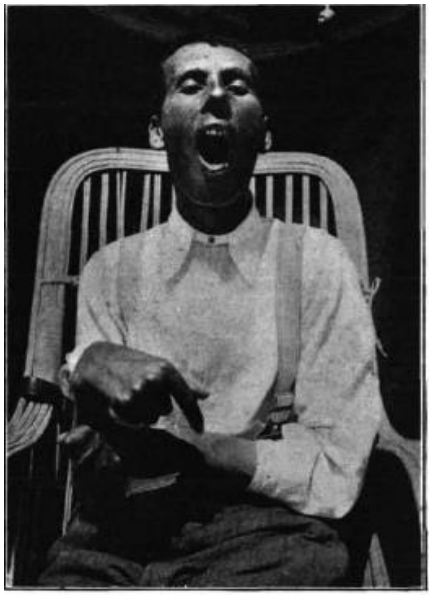


Wilson's disease

Clinical presentation (Wilson's patient W1)

Wilson's disease

Wilson's patients W2, W3 & W4

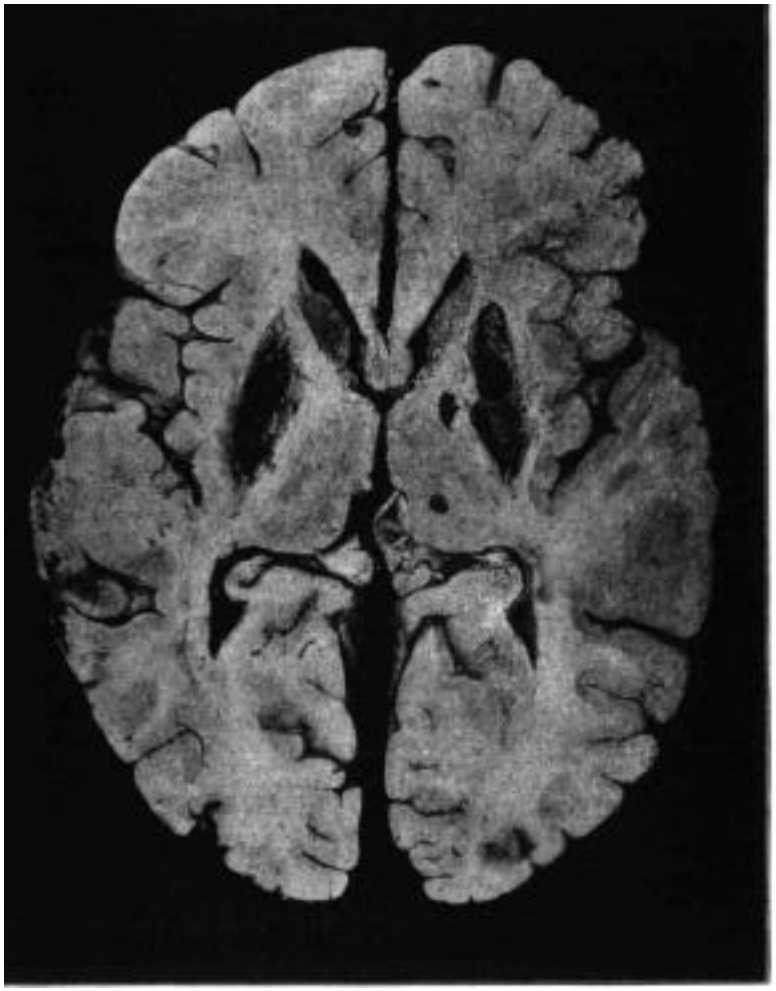


severe orofacial dystonia with large mouth opening

Wilson's disease

Brain and liver pathology

(9 autopsied cases)



- Lentiform lesions in seven cases (two had only macroscopic examination)
- Liver cirrhosis in all 9
- Illustration with patient W1

Wilson's disease

Previous reports related to the illness

- 1888 & 1898: Westphal C then Strümpell A : Pseudosclerosis, but no clear lenticular lesion; 1/5 had liver disease
- 1911: Völsch M: Pseudosclerosis with cirrhosis and no macroscopic lesion of the lenticular nuclei (acknowledged by Wilson)
- 1902 & 1903, Kayser B then Fleischer B : pericorneal ring

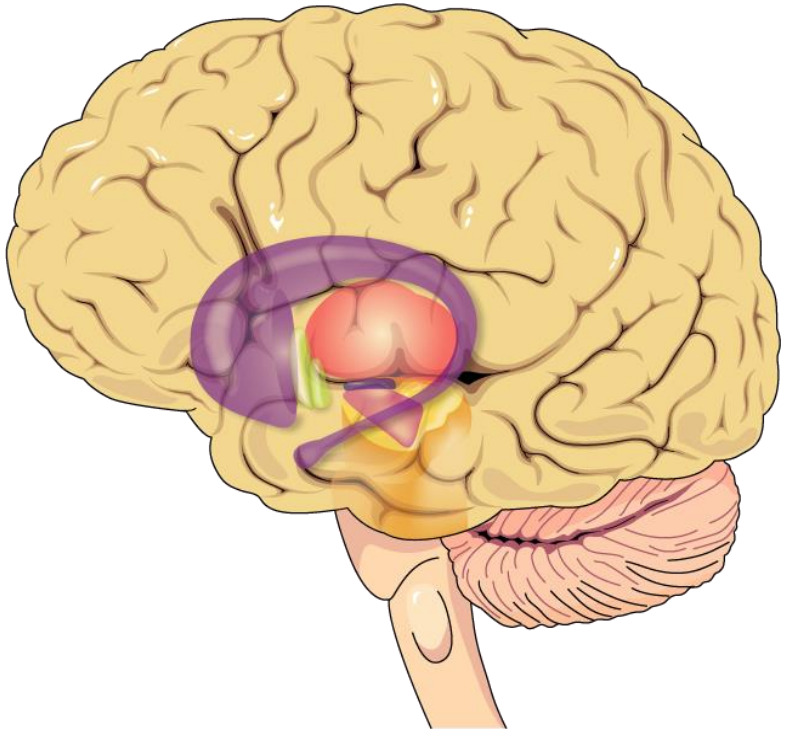
Wilson's disease

Subsequent reports and delineation of the toxic role of copper

- 1913: Rumpel A : Excess copper in the liver (forgotten finding)
- 1921: Hall HC (thesis, DK): New denomination: « dégénérescence hépatolenticulaire » (*hepatolenticular degeneration*); autosomal recessive inheritence
- 1934: Gerlach W & Rohrschneider W : Excess copper in corneal ring
- 1948: Excess copper in brain and liver (Cumings JN) and in the urine (Mandelbrote BM)

Wilson's disease

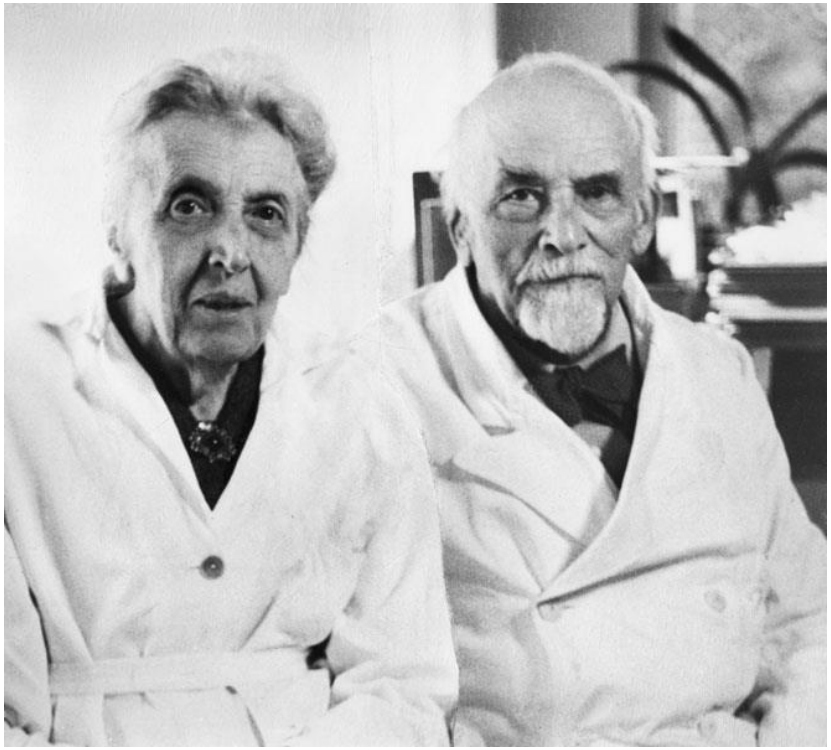
Wilson contributed to determine the role of the basal ganglia in motion and introduced the concept of extrapyramidal system



No definite idea of the role of the basal ganglia at the turn of the 19th and 20th centuries

Wilson's disease

Cécile Vogt, 1911: role of the corpus striatum (basal ganglia) in motility



Cécile Vogt-Mugnier (1875-1962)
& Oskar Vogt (1870-1959), Berlin

- Cécile Vogt: "état criblé" and double athetosis

In:

- Vogt C. Demonstration anatomischer Präparate (Syndrom des Corpus Striatum). *Neurol Zentralbl.* 1911;30:397-399.

Wilson's disease

Wilson, in his 1912 Brain article, emphasizes the role of the basal ganglia in motor function

Concept of extrapyramidal syndrome

- Wilson's patients have severe contracture and muscle weakness but no real paralysis, no Babinski sign, and no lesions of the cortico-spinal tract
- He suggests the role of the basal ganglia in motility and proposes the terms extrapyramidal system to depict the basal ganglia role and extrapyramidal signs to describe the motor signs associated with damage to this system
- Wilson will later speak about the «old » and the « new » motor system and further explore the syndrome of the corpus striatum in his famous 1925 Croonian lectures

Kinnier Wilson and Wilson's disease

Conclusion :

- Kinnier Wilson was one of the most renowned neurologists of the first half of the 20th centuty
- He did a great contribution to many aspects of neurology
- Wilson identified a disease in 1912 which shares his name and contributed to the delineation of the role of the basal ganglia in motor function

Thank you for your attention