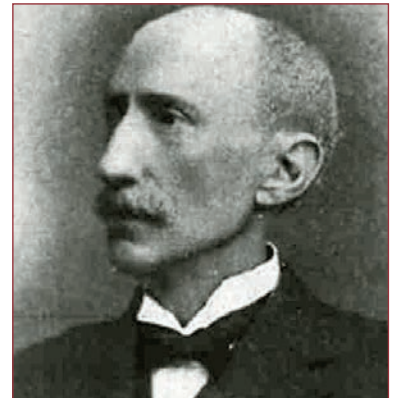


LLUÍS BARRAQUER I ROVIRALTA BARRAQUER-SIMONS SYNDROME

The eponym

Barraquer-Simons syndrome. A condition initially described by Lluís Barraquer i Roviralta (1907) as atrophy of adipose tissue and later by Simons (1911) as progressive lipodystrophy. It is characterised by extraordinary weight loss in the upper half of the body, especially in the face (mummy face), that begins in childhood or around puberty. The lower half of the body maintains the adipose stores –they even increase. The syndrome is more common in women than in men; it may be accompanied by lipoatrophic diabetes, mental retardation, otosclerosis, bone cysts, and nephritis¹. It is also called Barraquer-Simons disease, Barraquer syndrome, Barraquer disease, and Barraquer progressive cephalothoracic lipodystrophy.



Lluís Barraquer i Roviralta
(1855-1928)

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Lluís Barraquer i Roviralta is considered the founder of Catalan neurology. Furthermore, prestigious physicians, such as Pedro i Pons² and Barcia Goyanes³, consider him the founder of Spanish neurology, although his personality was an impediment to his teaching fully reaching beyond Catalonia's borders. Barraquer was fully dedicated to the medical profession; in his grandson's words, he "lived with a Benedictine devotion to his work"⁴. This dedication resulted in the development of a new medical specialty in Catalonia, following the example of other European countries such as France, Germany, and Austria.

Lluís Barraquer i Roviralta was born in Barcelona in 1855 and graduated from the *Universitat de Barcelona* in 1878. During his time at the Faculty of Medicine he was notably influenced by two teachers⁵. The first was his brother Josep Antoni, an ophthalmologist with an intimate knowledge of the nervous system. The other, Bartomeu Robert, had a passionate interest in clinical neurology and would go on to become very well known for his political activity as the mayor of Barcelona. Barraquer's early interest in diseases of the central nervous system was rewarded when in 1882, just four years after graduating, he was appointed director of the Dispensary of Electrotherapy and Diseases of the Nervous System, later known as the Department of Neurology and Electrotherapy and eventually as the Department of Neurology. At this time, no such department yet existed anywhere in Spain⁶.

It was, perhaps, portentous that 1882 was a significant year for European neurology: Charcot took on the first Chair of Neurology in Paris, and Obersteiner opened his Institute of Neurology in Vienna⁷. When the *Hospital de la Santa Creu* moved to its new headquarters in Sant Antoni Maria Claret Street to be renamed *Hospital de la Santa Creu i Sant Pau*, Barraquer transferred there too. He would not leave his position in this hospital until his death.



Three generations of neurologists: Lluís Barraquer i Roviralta (right), his son Lluís Barraquer i Ferré (left), and his grandson Lluís Barraquer i Bordas (centre). Summer of 1923, Sant Climent de Llobregat

All biographic references to the figure of Barraquer i Roviralta highlight his single-minded dedication to his profession, only interrupted by periods of relaxation at his home in Sant Climent de Llobregat, near Barcelona. Such an intense professional life enabled him to develop a new medical specialty, with no teachers or foreign references, since Barraquer i Roviralta, contrary to custom, did not train under any of the great figures of the period. He had only his own effort, his intelligence, his patients, and his books. He was, in fact, a self-taught neurologist. This important undertaking deserves to be even more highly valued considering that Barraquer i Roviralta had a weak constitution, and his character was infused with a certain sadness and irritability. He had his

reasons, since he was widowed for a second time when he was 35 years old, and a few years later he lost the son from his first marriage, Enric Barraquer i Solà. Furthermore, his financial situation was not very good, since he had given up the practice of psychiatry in favour of neurology, an exceptional situation for the time⁴.

Barraquer i Roviralta's work was not limited to the *Hospital de la Santa Creu*; he also founded the Department of Neurology at the *Hospital de Nostra Senyora del Sagrat Cor*. Moreover, he took the first steps toward establishing neurosurgery in Barcelona. From 1910 onward, under his instruction, Salvador Cardenal and Enric Ribas operated on patients affected by focal epilepsies, often of a traumatic origin⁶. He also indicated the excision of expansive brain lesions such as cerebral cysticercosis, an operation undertaken by Cardenal, and a cerebellar tumour, operated on by Antoni Raventós⁸.

In 1922, he became a member of the *Reial Acadèmia de Medicina de Barcelona*, giving a speech on the semiologic value of idiomuscular contraction. Corominas, who was president years later, recalled an anecdote that occurred at the Academy with Barraquer i Roviralta at its heart⁹:

“At that time it was common for the Academy to pronounce on court cases concerning accidents at work; when the matter was not very clear, the worker was summoned to plenary session and was asked to enter the hall, where all the academics could examine him and give their opinion after the worker had left. At one of these sessions, in which the case of a worker affected by sciatica was being contended, some of the academics claimed he could be malingering, but Barraquer, who had seen him and knew that he was not, without saying a word, took a bunch of keys from his pocket, flung them at the standing worker's feet and asked him to pick them up. The good man remained perplexed for a moment, but finally decided to crouch down. However, he did so through a series of contortions so as to keep

his sciatica relaxed to avoid the pain it would cause him to bend his spine as a healthy person would. Once the worker had left, alone with the academics, Barraquer explained the reason for the movements the worker had executed to pick up the keys. He went into so much anatomical detail about the situation and the path of the sciatic nerve that all present were convinced, and nobody dared to suggest that the worker was faking it.”

Barraquer i Roviralta passed away on 12 October 1928 at the age of 73, at home in his beloved Sant Climent de Llobregat. Only thus was this indefatigable worker halted. Two years after his death, his study on brain compression, the last he had undertaken, was published. All his efforts were not in vain: his school would continue the work he undertook under the direction of his son, Lluís Barraquer i Ferré.

Barraquer-Simons syndrome

Among his many contributions to clinical neurology, perhaps the best known was his description of a case of progressive lipodystrophy. Rodríguez Arias gave a thorough account of the story of this description more than 70 years ago¹⁰. In 1906, Barraquer described a case of a little-known entity, lipoatrophy, which was published the following year in *Neurologisches Zentralblatt*. In fact, only a single similar case, but exclusively affecting the upper part of the body, had been presented before, by Weir Mitchell in 1885. Barraquer i Roviralta provided the first full description; the German physician Simons characterised it as a separate disease entity, systematised it, and coined the name “progressive lipodystrophy” in 1911.

Barraquer-Simons syndrome is not the only eponym associated with Lluís Barraquer i Roviralta. The eponym Barraquer reflex is also widely used to refer the grasp reflex of the foot¹¹, the athetosis associated with

Patient with progressive lipodystrophy described by Barraquer i Roviralta included in the original publication⁹



childhood encephalopathies, Barraquer anaesthetic mask, and Barraquer generalised hemilateral atrophy. With time, as would be expected, his work was recognised in Spain and abroad. Martí-Vilalta⁵ recounted an anecdote that reflects the professional esteem in which other prestigious doctors held him:

“Two brothers, affected by the atrophy described by the renowned French neurologist, whose name is associated with Pierre-Marie, visited Charcot, at the Hôpital de la Salpêtrière in Paris. ‘Well then’, said the great French master, upon realizing they came from Barcelona and their doctor was Dr Barraquer, [...] ‘You needn’t have come to me. In your country you have a man who knows your disease as well as I do’.”

The syndrome is also known as acquired partial lipodystrophy, cephalothoracic lipodystrophy, and progressive lipodystrophy. The first seems now the preferred name for describing it.

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