Joaquín Costa's (1846-1911) scapuloperoneal atrophy: self-reported observations through his personal diary and correspondence

S. Giménez-Roldán[1], J. C. Ara Torralba[2]

[1]Former head and professor of the Department of Neurology. Hospital General Universitario Gregorio Marañón, Madrid, Spain.
[2]Department of Linguistics and Hispanic Literature. Universidad de Zaragoza, Zaragoza, Spain.

ABSTRACT

Joaquín Costa (1846-1911) was regarded as an intellectual and moral reference in the promotion of democratic regeneration. A polymath, he produced a vast body of written work despite suffering a neuromuscular disease from the age of 20 years, which resulted in a severe disability that progressed over four decades. Between 1864 and 1878, he wrote personal notes and autobiographical texts about daily events, ideas, and projects, in addition to describing the progression of his condition. Costa's rare symptoms, including head drop, baffled the many doctors he consulted. His close friend Luis Simarro, a renowned neurologist and neurohistologist who was training in Paris at the time, got him an appointment with Jean-Martin Charcot in 1882. Charcot told Costa that he had a primary muscle disorder, and requested permission to take pictures of his back. Through Costa's letters and autobiographical notes, which are available digitally at the Historical Archive of the Province of Huesca (in Aragon, Spain), the disease presents as a hereditary scapuloperoneal atrophy, with eight affected family members spanning four generations. Costa's detailed self-reported observations over the last third of the 19th century and the first decade of the 20th century preceded Davidenkov's groundbreaking descriptions between 1927 and 1934.

KEYWORDS

Joaquín Costa, scapuloperoneal atrophy, Luis Simarro, Jean-Martin Charcot, Romain Vigouroux, Daniel Urrabieta Vierge

School and pantry
Epitaph on Joaquín Costa's tomb

Introduction

Joaquín Costa Martínez (14 September 1846 - 8 February 1911) was a jurist, historian, sociologist, and politician. He was born in Monzón, a town in the region of Alto Aragón, in the province of Huesca, to a humble peasant family.[1] Deeply patriotic, Costa was an enthusiastic advocate of democratic regeneration, an ideology and doctrine that was widespread across different sectors of Spanish society, aware that the country was falling behind.[1-4] His famous motto "School and pantry," symbolised with a book and an ear of wheat, denounced the chronic hunger and the lack of competent school teachers. Through the territorial organisation of Aragon,[1] he sought to trigger political action and to promote legal and structural reforms in education, culture, and research. He also promoted water resources management, a much-needed initiative in a country that periodically experienced destructive floods and prolonged droughts. The contemporary image of Joaquín Costa in Spain is that of a self-made man, both a myth and a legend (Figure 1).[5]

Several incomplete biographies of Costa have been published, including those by the writer and journalist Antón del Olmet in 1917[A] and Manuel Ciges Aparicio in 1930. However, it was the Hispanist George James

[A]http://ifc.dpz.es/publicaciones/ver/id/3027

Corresponding author: Dr Santiago Giménez-Roldán
E-mail: sgimenezroldan@gmail.com

Received: 23 March 2025 / Accepted: 30 April 2025

Figure 1. A) Joaquín Costa at 23 years of age. Photograph taken at "Rivas Foto," a studio located at number 5, Puerta del Sol, in Madrid (picture from Gil Novales, 2024). B) Portrait of Joaquín Costa by Victoriano Balasanz (1854-1929), showing a "leonine head of grey hair and a doctoral beard."[13] Source: Biblioteca Nacional de España.

Gordon Cheyne (London, 15 November 1916 - Newcastle upon Tyne, 29 December 1990) who published the most reliable and comprehensive biography of Joaquín Costa, in 1972.[6-9] However, no author to date has focused on the nature of the disease that caused him such great suffering. Cheyne proposed that Costa may have presented progressive muscle dystrophy (Figure 2A).[9] On the other hand, Venancio Díaz Castán, a writer and rural physician who was very familiar with Costa's circumstances, since he was born in Graus and shared a kinship with Costa's family, suggested that Costa had "limb-girdle muscle dystrophy," assuming that his disease was inherited through maternal transmission (Figure 2B).[10] In view of the limited knowledge about Costa's disease, he humbly made the following observation: "I have always found it very surprising that specialists never showed an interest in Joaquín Costa's process" (personal correspondence with the author, November 2023).

The purpose of this study is to review the disabling neurological condition that affected Costa over nearly five decades (1864-1911): its clinical manifestations, natural history, and possible affected relatives. This study is based on information sources that until recently were only partially available.

Materials and methods

In 1864, at the age of 18 years, Costa began keeping his personal diaries, or *Memorias*, using cheap ruled notebooks[11] where he described private details, including his observations about the development of his disease. To locate, organise, and decipher Costa's "small, terrible handwriting," including corrections and countless notes organised in notebooks and folders, Juan Carlos Alba Torralba, the chair of linguistics and Hispanic Literature at the Universidad de Zaragoza, required the patience of a saint. The publication of Costa's *Memorias*, a book

spanning 583 pages, represents a historical milestone, Alba's 1980s dream come true.[12] The Historical Archive of the Province of Huesca, created in 1931, preserves documents from as early as the 12th century, as well as Costa's original writings[B]. *Anales de la Fundación Joaquín Costa* (1984-2021), a journal of the Instituto de Estudios Aragoneses, publishes research articles on Costa and his time[C]. Four renowned writers who knew him personally (Alberto Insúa, José Martínez Ruíz-Azorín, Miguel de Unamuno, and Rafael Altamira) hinted at his evident physical limitations.[13] The numerous letters exchanged over the course of five years (1904-1909) between Costa and Laureano Rosso, an unscrupulous physician from Málaga, constitute another interesting source of information.[14]

Results

Chronology of Joaquín Costa's symptoms, functional limitations, and complications (Table 1)

Costa travelled to Paris on 6 March 1867, at the age of 21 years, to work as a builder at the Spanish pavilion at the Paris Universal Exposition of 1889; this eye-opening experience made him reflect on the situation of his country. He was forced to interrupt his stay in Paris to attend an examination at his hometown's army recruitment centre, where he was deemed unfit for military service.[2] By age 54, Costa was no longer able to care for himself. At the age of 57 years, he had ceased his professional activity, and was under the care of his sister Martina at their family house in Graus. He had a small office on the third floor, where he would sit on a rocking chair, leaning back to rest his head against the wall.[10] From his window, he enjoyed a view of the wide river Ésera and the beautiful medieval bridge.

At age 62, Costa summarised the course of his disease and the resulting limitations in daily living activities as follows:

> It is a hereditary condition whose manifestations become evident at the age of 25 or 28 years. It progresses imperceptibly over several years. Then, the progression seems to halt. Over the course of approximately a decade, it has progressed and become more marked […]. The serratus anterior muscles are weakened; the right shoulder blade is loose and the shoulder drops […]. I can only use my left hand to tip my hat. To bring food to my mouth, I need to rest my left arm on the table, which is considered bad table manners, and still I suffer greatly. When I comb my hair, I look distracted. When I attempt to write or draw, the situation is beyond words. When I walk, I drag the tips of my shoes, so I risk tripping over any little pebble and falling. Years later, the condition of my left arm had progressed nearly to the degree of involvement of my right arm. My brain seems to retain all or nearly all its function, since I already had this condition when I practised simultaneously as a notary public and as a lawyer […]. In recent years, atrophy has begun to affect new muscle groups; I don't know what they are called—maybe the cervical muscles—, but it's the muscles that hold the head upright, preventing it from tilting forward. They have presented weakness since my university years. Some time later, my mother's head began to tilt forward; the same occurred to one of her brothers, a priest who is still alive but can no longer lift his head […]. Writing is extremely draining and exhausting, even though I take breaks every few minutes and drink coffee […]. Fatigue starts from the moment I get up; pain mainly affects the area around the knees. My condition is tolerable only when I sit on a slatted rocking chair. Climbing up the stairs, even when pushed and pulled by two people, is torture […]. Not to mention how unbearable it is when the weather is hot; I just can't do it, I need cold […]. My poor muscles, sick and powerless, turn against me […] the labour of lifting and bearing 90 kilograms.[14]

Around the age of 60, Costa developed recurrent respiratory infections and constipation secondary to prolonged immobility. On 18 December 1910, he presented a cerebral haemorrhage that caused left hemiplegia; he fell into a coma and died at the age of 65 years, on 8 February 1911. Costa's family home, a four-storey house, is the subject of dispute between his heirs and the provincial government of Huesca; a project has been developed to convert the building into a house-museum, displaying Costa's library, office, and famous rocking chair, to which he was confined for over two decades (Fig. 3A). Graus pays homage to Joaquín Costa with a monument designed by the architect Fernando García Mercadal and sculpted by José Bueno. On the initiative of the journalist Mariano de Cavia, it was inaugurated in 1929 (Figure 3B).

[B]https://dara.aragon.es/opac/app/simple/apjc; accessed 22 September 2023
[C]https://dialnet.unirioja.es/servlet/revista?codigo=70.

Tabla 1. Chronological description of Joaquín Costa's disease based on his correspondence and self-reported symptoms.[15-22]

1867 (21 years old). Initial symptoms. While working as a builder at the Spanish pavilion at the Paris Universal Exposition of 1889, he presented difficulty raising his right arm. In August of that same year, he was exempted from military service due to his physical limitations.[15]
1868 (22 years old). Weakness extended to the left arm.
1869 (23 years old). Weakness extended to the right leg: "When I walk, I drag the tips of my shoes, so I risk tripping over any little pebble and falling. I have tripped several times, and I need to use orthopaedic boots; without them, I cannot take a single step."[15]
1870-1872 (24-26 years old). He studied law at the University of Madrid: he began to feel "weakness and laxity in the muscles of the head."[9]
1870-1898 (24-52 years old). He consulted numerous physicians in search of a diagnosis and possible treatment. At the age of 30, he mentioned difficulty walking during a hunt in Arro, in Huesca, and soon after, while walking up to the Royal Monastery of San Victorián, in the Aragonese village of El Pueyo de Araguás, Huesca.[11]
1882 (36 years old). Charcot examined Costa on Saturday 27 July 1882. He referred the case to Romain Vigouroux, who would treat Costa with electrotherapy; this forced him to travel to Paris a second time, probably during the summer of 1883.[10]
1885 (39 years old). Difficulty holding his head upright worsened. In a letter to Giner de los Ríos, Costa states: "My problems [now] involve the legs, arms, and head."[21]
1891 (45 years old). Generalised weakness. Replying to a letter from Giner de los Ríos suggesting that he should publish a book about customary law, he wrote: "I was unable to do so, as I was chained to my general paralysis."[21]
1895 (49 years old). In reference to Costa climbing down the steep stairs of the Ateneo de Madrid: "We saw that man climbing down the stairs. He descended slowly and heavily, with the overwhelming difficulty of a sick person. This view filled us with grief and compassion."[11]
1900 (54 years old). Dependent for self-care. His brother Tomás, who worked as an assistant at Costa's notary office, stated: "Joaquín's disease has progressed, and he now has difficulty standing up, getting dressed, and bathing."[14]
1901 (55 years old). Severe difficulty walking. Dr Chabás wrote: "He was unable to walk for years and years." Costa accepted the incurable nature of his disease: "I am a slave to my nervous disorder […], my amyotrophy is incurable; it's been four years since I voluntarily and forcibly stopped looking for a cure."[22]
1903 (57 years old). Confined to a rocking chair, he abandoned all professional activity and retired to Graus, remaining under the care of his sister Martina. He gave up writing, except for his personal correspondence. However, encouraged by Giner de los Ríos, he travelled to Switzerland to consult with the rehabilitation physician Heinrich S. Frenkel.[14]
1906 (60 years old). Prolonged, recurrent respiratory tract infections associated with chronic bronchitis. He ventured to travel to Madrid to gather information at the library of the Ateneo. Two men carried him on their backs, with his head tilted forward, a view that inspired compassion and pity among those who saw him.
1907 (61 years old). He planned a trip to Tarragona and Lérida, but warned that he would "need to be supported by the arms of two strong men."
1908 (62 years old). He started exchanging letters with the physician Laureano Rosso, from Málaga (13 January - 4 July); Rosso pressed Costa to hire him as his personal doctor.[14]
1909 (63 years old). Confined to bed, Costa suffered a "recurrent intestinal catarrh and persistent constipation."
1910 (64 years old). "I have made up my mind, I have resisted, but today I decidedly give up."
1911 (65 years old). After several years with diabetes and arterial hypertension, Costa developed left hemiplegia on 18 December 1910, but was not examined until eight days later. He was brought to the second floor, which had a bathroom, and a young man was hired to carry him. Costa developed pressure ulcers on the buttocks. Several physicians attended him, including Miguel Gayarre, a neurologist of Cajal's school in Madrid, and Royo Villanova, chair of medicine in Zaragoza. On Tuesday 7 February, he presented confusion, and entered a coma shortly thereafter. He died at 4 o'clock in the morning.[10]

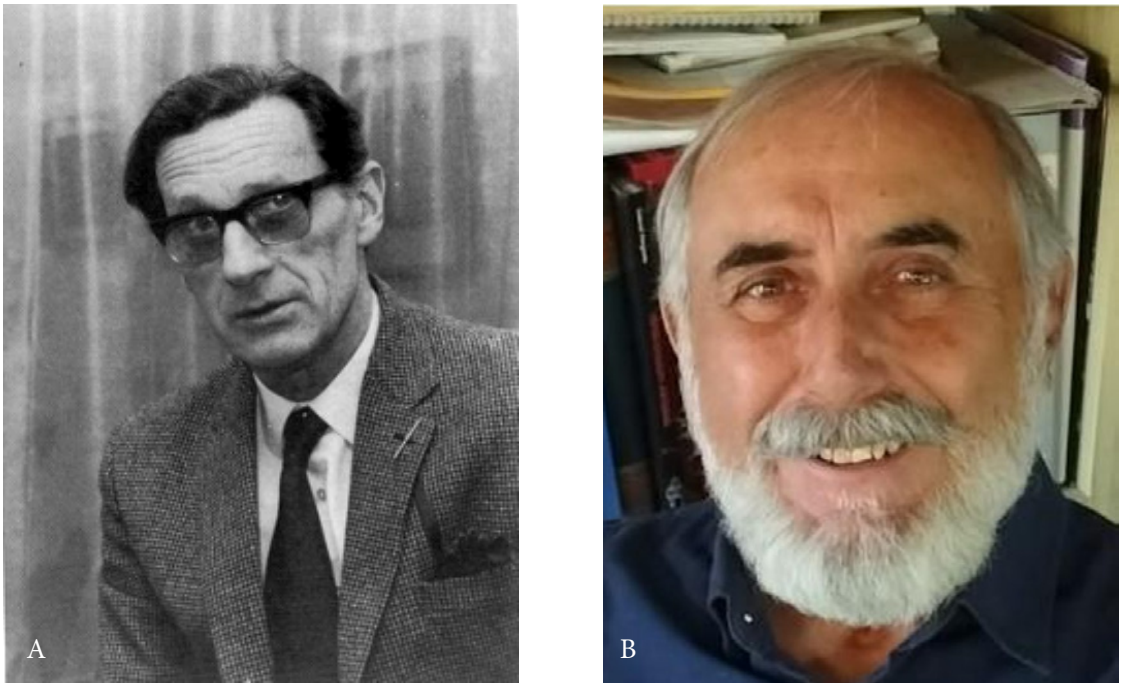


Figure 2. A) George J.G. Cheyne. Photograph by Manuel Compañy (1908). Public domain image. B) Dr Díaz Castán (personal donation to the author, September 2023).

A hereditary disease

Joaquín Costa was aware of the familial nature of his disease, which he referred to as "the curse of a race." He identified his own symptoms in his mother, María Martínez Gil, Joaquín Costa Larrégola's second wife, as well as her brother, the priest Lucas Martínez. Little is known about Costa's grandparents, although it has been noted that María Gil, his maternal grandmother, was always ill. Joaquín Costa Larrégola and María Martínez Gil had four children, of whom only the eldest, Joaquín Costa, developed the disease, with his siblings Martina, Vicenta, and Tomás apparently being spared.[9] Tomás died without offspring, but none of his sisters' six children developed symptoms.[9] Costa had a relationship with Isabel Palacín Carrasco (Elisa), the widow of the engineer Teodoro Bergnes, with whom she had two children. In 1883, when both Joaquín Costa and Isabel Palacín were living in Madrid, they had a daughter, whom they named Antígona and whom Costa acknowledged belatedly. Antígona was later baptised as María del Pilar Costa Palacín (1883-1970). According to Greek mythology, Antigone disobeyed human law out of respect for a higher divine law.

María del Pilar married José María Ortega Ballesteros, chief public works engineer in Barcelona, with whom she had 13 children. She was widowed at the age of 40 and died at the age of 87, apparently asymptomatic.[13] However, she passed the disease to five of her 13 children (two men and three women). In 1960, her son Alfonso Ortega Costa, Joaquín Costa's sixth grandchild, accompanied Cheyne and his wife, the psychiatrist Assumpciò Vidal Parellada, on their first visit to Monzón and Graus, two relevant towns in Costa's life.[23] In a letter to Dr Venancio Díaz Castán, dated 20 November 1989, Alfonso Ortega Costa, who was president of the Joaquín Costa Foundation, narrates the second part of this story: "The manifestations of my disease were identical to those of my grandfather," based on Costa's descriptions in his letters to Laureano Rosso. Regarding the future of his family, he wrote: "The affected relatives had no direct heirs,

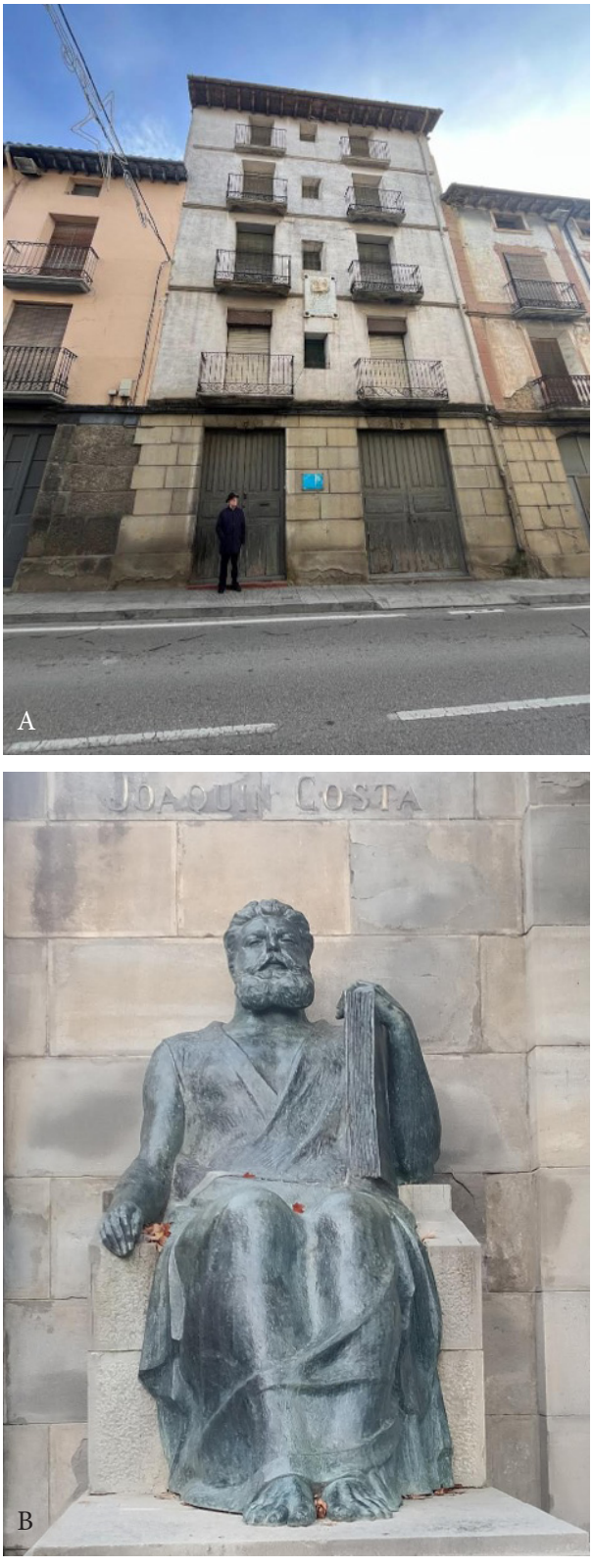


Figure 3. A) Costa's family house in Graus, Huesca, where he spent his last 21 years (photograph by the author, November 2024). B) Monument to Joaquín Costa in the town of Graus (photograph by the author).

either because they died without offspring or because they were not married." He died in 1994, after spending six years confined to bed due to a femur fracture from a fall at home, according to the obituary published that year in *Anales de la Fundación Joaquín Costa*. In summary, Joaquín Costa's family included eight members with documented clinical manifestations, affecting four generations over the 19th and 20th centuries. The available data suggest that the alleged mutant gene inherited by Joaquín Costa from his mother disappeared in the fourth generation. Two large families with scapuloperoneal syndrome reported in Barcelona and Madrid do not seem to be related to the Ortega-Costa family.[24,25]

Costa in Paris: in search of Charcot's clinical judgement

Over his lifetime, Costa was examined by at least a dozen of Spain's most prestigious physicians.[10] He even travelled to Heiden, in Switzerland, in 1890 to attend the consultation of the rehabilitation physician Heinrich S. Frenkel, who supplied him with a pair of special boots that became essential for walking.[9] Costa and Dr Luis Simarro Lacabra (1851-1921), a famous neurologist and neurohistologist, had a close friendship that began at the Institución Libre de Enseñanza (1876-1939), an influential educational project based on Krause's ideas and intellectual freedom as the essential pillar of progress.[26] Neuroscientific knowledge was spreading; particularly noteworthy were Simarro's lectures on nervous system physiology in the Ateneo de Madrid[27,28]

In late 1880, Simarro travelled to Paris to train with the French alienists, with whom he had ideological affinity. He probably attended Charcot's lessons at La Salpêtrière; therefore, we may assume that they established a friendship.[29] He frequently visited the laboratory of Louis-Antoine Ranvier (1835-1922) at the Collège de France, where he learnt the Golgi silver staining technique, which highlighted nervous cells in black (*reazione nera*).[30] Years later, he would teach Cajal this technique, which represented a milestone for the Spanish neurohistological school.

Worried about the nature of Costa's disease, Simarro suggested that he travel to Paris to consult with Jean-Martin Charcot, the most famous neurologist at the time.[31] Charcot's high professional fees were a disadvantage, given Costa's precarious financial situation. Charcot's elegant consultation was located in his family home at Hôtel de Varengeville, at number 217, Boulevard

Saint-Germain.[31,32] Millionaires and influential people from all over the world, including the emperor of Brazil, had to accept the long waiting list. Simarro managed to arrange a quick, free-of-charge consultation with Charcot through the Administration Générale de l'Assistance Publique, a French non-profit organisation established in 1849. Upon arrival, Costa completed only a section of the admission form, with nearly illegible handwriting. Except for his age (36 years at the time), he deliberately provided incorrect information, giving a fake surname, a different profession (builder instead of university professor and lawyer), and home town (Madrid, instead of Monzón, in the province of Huesca) (Figure 4). We may assume that he wanted to avoid the attention of journalists, who were eager to publish information about him.

Charcot examined Costa on Saturday 27 July 1882, as noted on the form, probably at the polyclinic in La Salpêtrière, on Boulevard de l'Hôpital, where he was able to attend male patients at a hospital for women (Figure 4). Only seven months had passed since Charcot was appointed, on 2 January, to the chair of the new department of nervous system diseases, thanks to the prime minister Léon Gambetta (1838-1882).[31]

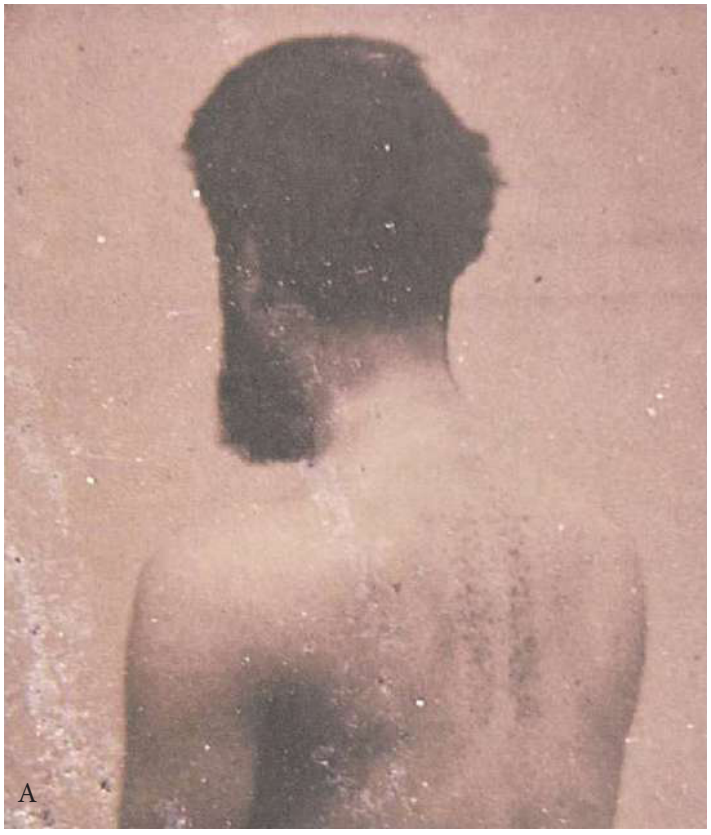
Charcot told Costa that his disease did not cause lesions to the spinal cord, as he himself had previously described (referring to amyotrophic lateral sclerosis), but rather to the muscles. The French physician prescribed him silver nitrate (internal use), *claviceps purpurea*, weekly thermocautery sessions on both sides of the spinal column, massages, and showers. He also recommended daily electrotherapy sessions, which would be administered by Dr Vigouroux at his own laboratory at La Salpêtrière. Charcot requested permission to take an undressed photograph of Costa, which is currently held at the Historical Archive of the Province of Huesca (circa 1890; ref. Costa/000056/011-06).

In 1878, Charcot created a clinical photography laboratory at La Salpêtrière, under the direction of Albert Londe (1858-1917),[33] who presumably took Costa's picture. By 1882, Charcot was delighted with the good results of electrotherapy, particularly for the treatment of patients with hysteria and hystero-epilepsy: "Application of static electricity for a few weeks freed them of their attacks." After the death of Guillaume Duchenne de Boulogne (1806-1875), Charcot opened an electrotherapy laboratory in a wooden building located near the kitchens of La Salpêtrière, which would be managed by Romain Vigouroux (1831-1895). Both Charcot and Duchenne used electrotherapy as a treatment for different neurological diseases. Duchenne, who lacked a service of his own, selected eligible patients across different hospitals in Paris, whereas Vigouroux applied electrotherapy at La Salpêtrière.[34]

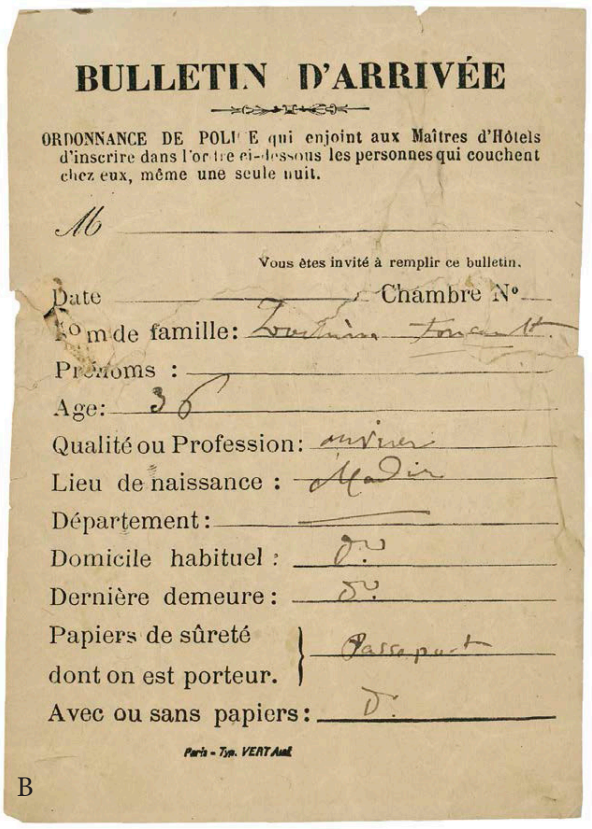
The technique was applied not only to Joaquín Costa, who had evident muscular atrophy, but also to Daniel Urrabieta Vierge (Madrid, 1851 - Boulogne sur Seine, 1904), a Spanish illustrator of books and periodicals.[35] Following an episode that caused right hemiplegia, Vierge was treated by Vigouroux at La Salpêtrière. Vierge captured the sinister scene of an electrotherapy session in one of his illustrations: wearing his usual black hat, Vigouroux uses a giant electrotherapy device to apply electrical current to a half-naked woman who appears to be in pain, while an impassible nurse contemplates the scene (Figure 5).[36] In André Brouillet's famous painting *A clinical lesson at the Salpêtrière* (1887), Vigouroux can be seen close to Charles Féré, to his left, and Gilles de la Tourette, to his right.[37-39] He frequently used the device invented by Du Bois-Reymond for therapeutic faradisation.[39] Costa had to return to Paris the following summer, this time to attend Vigouroux's private consultation. Vigouroux informed him succinctly: "The technique's lack of success is confirmed, and I have no further treatment to prescribe. After six months of follow-up, not a single muscle tissue has reacted or made the least movement, maybe due to fatty degeneration." Romain Vigouroux referred Costa to Dr Serafín Buisen, who was in charge of the department of electrotherapy and the outpatient neurology consultation at the Instituto de Terapéutica Operatoria in Madrid, founded by the surgeon Federico Rubio Galí (1827-1902).[40]

Unusual clinical signs of Costa's disease

Dr José Chabás Bordehore (1877-1963), the last physician who examined Costa when he was 55 years old, wrote: "In contrast with his mighty, coarse voice [this earned him the nickname of 'the lion of Graus']," and his hefty, giant-sized build, "his hands and feet were surprisingly small."[22,41] The weekly satirical magazine *Gedeón* (1895-1912) published a caricature of Costa in 1900 in which he is depicted with very small feet, though they are not deformed; this points to the hypotrophy associated with conditions causing global atrophy of the intrinsic muscles of the feet (Figure 6A).[42]



A



BULLETIN D'ARRIVÉE

ORDONNANCE DE POLICE qui enjoint aux Maîtres d'Hôtels d'inscrire dans l'ordre ci-dessous les personnes qui couchent chez eux, même une seule nuit.

M

Vous êtes invité à remplir ce bulletin.

Date Chambre N°

Nom de famille: Costa

Prénoms :

Age: 36

Qualité ou Profession: ouvrier

Lieu de naissance : Madrid

Département :

Domicile habituel : D°

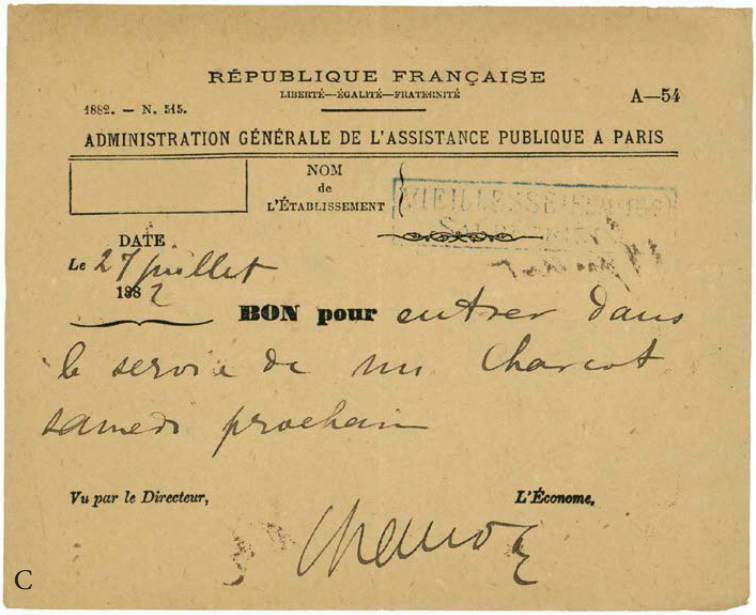
Dernière demeure : D°

Papiers de sûreté dont on est porteur. } Passeport

Avec ou sans papiers : D°

Paris - Typ. VERT Aîné

B



RÉPUBLIQUE FRANÇAISE

LIBERTÉ—ÉGALITÉ—FRATERNITÉ

1882. — N. 545. A—54

ADMINISTRATION GÉNÉRALE DE L'ASSISTANCE PUBLIQUE A PARIS

NOM de L'ÉTABLISSEMENT

DATE

Le 27 juillet 188 2

BON pour entrer dans le service de M. Charcot samedi prochain

Vu par le Directeur,

L'Économe,

C

Figure 4. A) Photograph of Costa's back after an examination performed by Jean-Martin Charcot in 1882. B) Costa's admission form upon his arrival in Paris. C) Appointment receipt for a consultation with Charcot on Saturday 27 July (available at the Historical Archive of the Province of Huesca, in Spain).

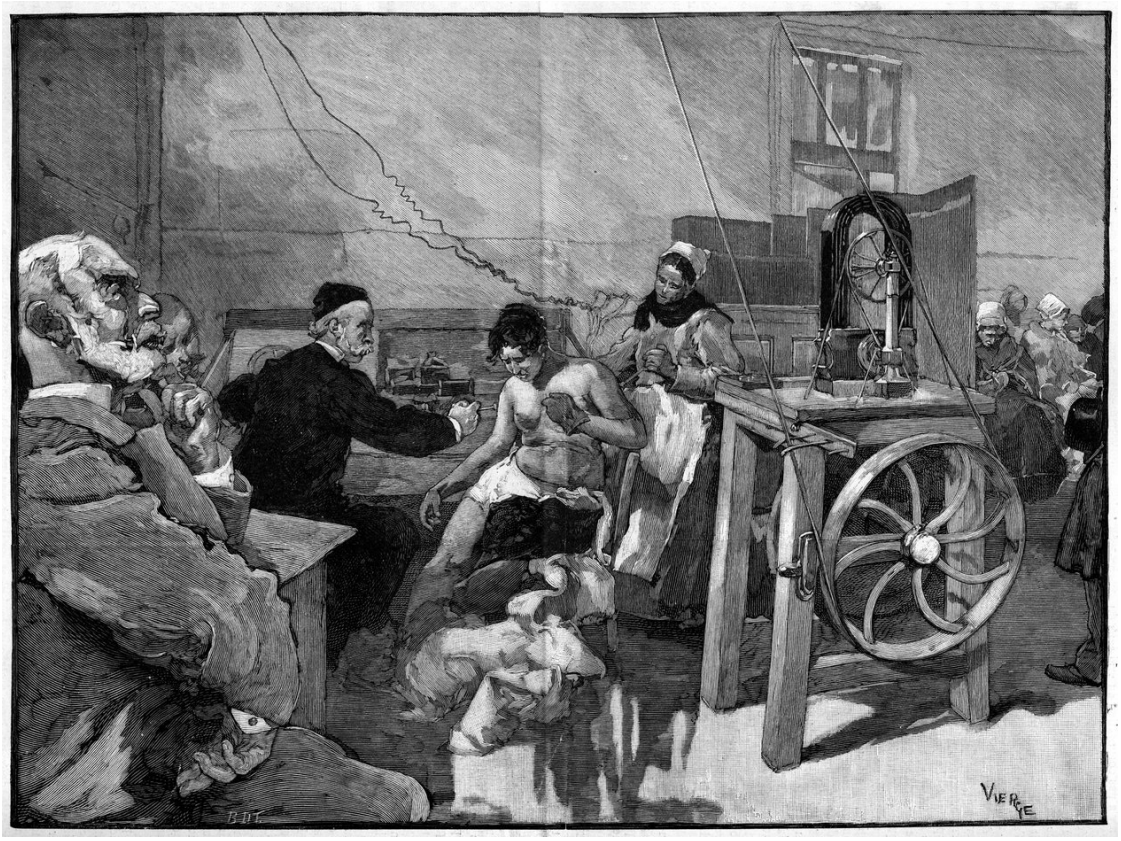


Figure 5. Illustration by the Spanish illustrator Daniel Urrabieta Vierge, known as Vierge in France. Vierge moved to Paris after his hospitalisation at La Salpêtrière due to a stroke. The illustration shows Dr Vigouroux during an electrotherapy session. Source: *Manuel théorique, instrumental et pratique d'électrologie médicale*[36].

At 24-26 years of age, muscular atrophy of all four limbs had worsened, which may explain Costa's difficulty holding his head upright; his head would tilt forward, with his chin resting on his chest, and slightly sideways. At the age of 60 years, Costa insisted on climbing the steep stairs of the Ateneo de Madrid to the library, but had to do so "in the arms of two strapping men, with his head down and tilted"; this inspired sympathy among those who saw him.[43] A pencil drawing (1923) shows Costa with his head tilted forward and to one side (Figure 6B).

Another striking feature was the marked asymmetry in the development of limb weakness and atrophy, which initially affected the right arm, extending to the left arm one year later, and to the legs after another year. Howard H. Tooth had already included marked asymmetry in his description of the peroneal form of progressive muscular atrophy.[44,45]

Discussion

Natural history in a family with scapuloperoneal atrophy

In his *Memorias*, Joaquín Costa described the course of his progressive neurological disease, which limited him for 44 years and ultimately left him severely disabled. The disease manifested at 21 years of age with "a loose right shoulder blade and dropped shoulder," probably due to scapular winging as a result of atrophy of the serratus anterior muscle at the level of the shoulder girdle, a topic that fascinated Charcot in 1882. The condition subsequently extended to the distal muscles of the right leg, causing foot drop, presumably due to the weakness and atrophy of the muscles in the anterior compartment of the legs. This marked the beginning of Costa's difficulty walking, with frequent falls after tripping over the tips of his shoes, which forced him to use orthopaedic boots.

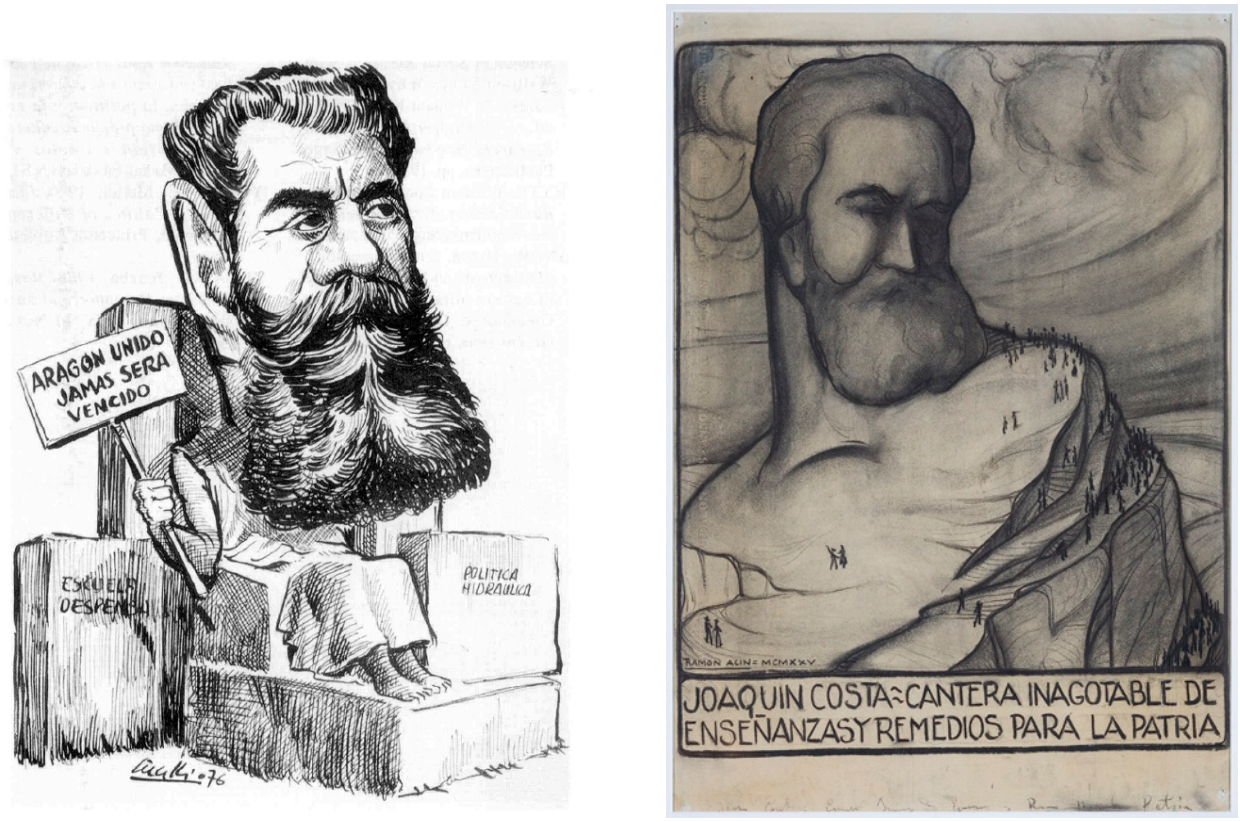


Figure 6. A) Costa's tiny feet, showing clear disproportion to his voluminous figure. Lithograph on Silenus paper. Source: the satirical weekly magazine *Gedeón*, no. 253, of 26 September 1900 (Víctor Juan, www.fundacionacin.org). B) Sketch of the monument to Joaquín Costa, by Ramón Acín Aquilué (1888-1936); pencil drawing made in 1923. The drawing shows the bust of Costa carved into a mountain, which a group of people are walking up. The inscription reads: "Joaquín Costa, endless source of lessons and remedies for the fatherland." Photograph taken by Fernando Alvira Lizano (Museo de Huesca).

After all four limbs were affected, at the age of 24-26 years, Costa began to present difficulty holding his head upright, with his chin frequently resting on his chest (dropped head syndrome). This is caused by weakness of the extensor muscles of the neck, and may present in a wide range of neuromuscular disorders.[46] In Costa's family, his mother, uncle, and some grandchildren also developed the disease. On the other hand, the lack of a formal clinical examination prevents us from confirming the integrity of the facial muscles, a feature that is not evident in the picture of Costa at 23 years of age nor in later pictures, when he had a thick beard.

As Costa himself proposed and his grandson Alfonso Ortega Costa (1990) confirmed, this familial aggregation of the disease strongly suggests an autosomal dominant disorder whose clinical characteristics were highly consistent across all affected family members (Figure 7). We should mention the notable exception of María del Pilar Costa Palacín (Antígona), Costa's only daughter, who apparently remained asymptomatic until her death at the age of 87 years. We cannot rule out the possibility that a detailed clinical examination would have detected signs of a muscle disorder; in fact, this may be the case in patients with a scapuloperoneal phenotype and chromosome 4q deletion.[47] Apparently, the responsible gene disappeared in the Ortega-Costa branch of the family, with no new cases reported to date.

Heterogeneous pathology of scapuloperoneal atrophy

Scapuloperoneal atrophy may present in myopathies, neuropathies, and spinal muscular atrophy.[48,49] After the examination performed in 1882, Jean-Martin Charcot

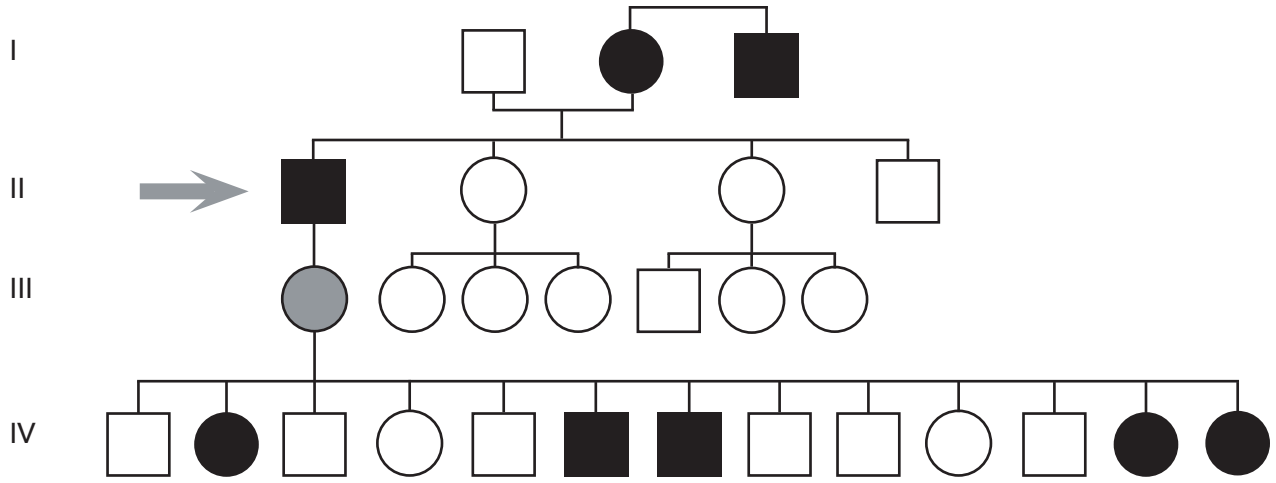


Figure 7. Joaquín Costa's family tree. Pilar Costa Palacín, his only daughter (grey circle), necessarily transmitted the disease, even though she herself did not present clear manifestations. Joaquín Costa is indicated with an arrow. Circles represent women, squares represent men. Black symbols represent affected individuals, while white symbols represent unaffected individuals. Source: created by the author.

concluded that Costa presented a primary muscle disease. Some years later, in 1906, Costa was examined by Ricardo Royo-Villanova (1868-1943), a distinguished professor of medicine who had a special interest in the nervous system. Royo-Villanova succinctly concluded that "Costa's disease is a condition of the feet, not of the head; of movement, not of ideation; of the muscles, not of the nerves […]. The weakness observed in his arms and legs may explain the extraordinary vigour of his lobes […]. Costa's disease is called primary progressive myopathy and is currently known as Landouzy-Dejerine disease."[50]

The limited value of self-observation

The medical literature includes numerous examples of self-observation from historically relevant figures. The Norwegian professor of neuroanatomy Alf Brodal (1910-1988), who had suffered a stroke, published the anatomical correlations of his symptoms a year later in *Brain*.[51,52] This is also the case of Alexander von Humboldt (1769-1859), who, like Joaquín Costa, was a political reformer and a scholar, and authored over 8000 letters. At the age of 61 years, he described with remarkable precision the clinical manifestations of his parkinsonism and the progressive disability it caused. He had a long romance with Charlotte Diede, who, against Von Humboldt's wishes, posthumously published his memoirs.[53] Costa's detailed self-reported manifestations, recorded over decades, strongly suggest that his muscle atrophy had a peculiar scapuloperoneal distribution. On the other hand, given the lack of confirmatory data from supplementary laboratory tests, the validity of his notes may be questioned, particularly the myopathic or neuropathic nature of his condition, whether due to degeneration of the peripheral nerves or the anterior horn of the spinal cord.

History of scapuloperoneal atrophy: a matter of priority

The history of scapuloperoneal atrophy dates back to the 19th century, with the Soviet neurologist and geneticist Sergej Nikolajevich Davidenkov (1880-1961). Born in Riga, in present-day Latvia, he developed most of his vast oeuvre (331 publications) in Leningrad, with a special focus on hereditary neuromuscular disorders, for which he coined the term "neurogenetics."[54,55] In a series of publications,[56] he proposed that "scapuloperoneal amyotrophy" was a variant of Charcot-Marie disease,

characterised by involvement of the shoulder girdle and symptoms in the distal lower limbs. In a study published posthumously, Davidenkov changed his initial interpretation, now considering scapuloperoneal amyotrophy to be a hereditary myopathy with a well-defined identity, although he observed that some characteristics were similar to those of the facioscapulohumeral muscular dystrophy found in Landouzy-Dejerine disease.[57] The 1975 review of the original K. kindred, in which 18 members were examined with modern diagnostic techniques, confirmed the presence of muscular dystrophy.[58,59]

Acknowledgements

A very special thanks to Dr Venancio Díaz Castán (RIP), who, in very difficult personal circumstances, encouraged us to continue the neurological study of Joaquín Costa. We also thank Juan José Generelo Lanaspa, director of the Historical Archive of the Province of Huesca (ahphuesca@aragon.es), for providing relevant data for our study.

Conflicts of interest

The authors have no conflicts of interest to declare.

References

1. Urbieta Arteta A. Historia de Aragón: la formación territorial. Vol. 1. Zaragoza (ES): Anubar; 1981.
2. Fernández Clemente E. Educación y revolución en Joaquín Costa y breve antología pedagógica: cuadernos para el diálogo. Madrid: Edicusa; 1969.
3. Forcadell Álvarez C. Costa en su centenario. Consideraciones acerca de Joaquín Costa y otros textos autobiográficos de su juventud. In: Salomón P, Alares G, Rújula P, coords. Historia, pasado y memoria en el mundo contemporáneo. Teruel (ES): Instituto de Estudios Turolenses; 2014. p. 117-23.
4. Dueñas Lorente J. Compromiso y modernidad en la obra de Joaquín Costa Martínez (1846-1911). AFJC. 2018;30:6-20.
5. Mérida Donoso JA. Joaquín Costa: mito, memoria e historia (la construcción de una imagen). Anales de la Fundación Joaquín Costa. 2014;28:41-126.
6. Fernández Clemente E. Una relectura bibliográfica de Joaquín Costa. Temas de Antropología Aragonesa. 1996;6:95-134.
7. Fernández Clemente E. La obra costista de George J.G. Cheyne. In: Gómez Benito C, Ara Torralba JC, Calvo Carrillo JL, Fernández Clemente E, Fontana J, eds. En torno a Joaquín Costa. Zaragoza (ES): Institución Fernando el Católico; 2012. p. 55-74.
8. Cheyne GJG. Ensayos sobre Joaquín Costa y su época. Gil Novales A, ed. Huesca (ES): Fundación Joaquín Costa; 1992.
9. Cheyne GJG. A bibliographical study of the writings of Joaquín Costa. London: Tamesis; 1972.
10. Díaz Castán V. Joaquín Costa y los médicos. Anales de la Fundación Joaquín Costa. 2020;3:11-44.
11. Antón del Olmet L. Los grandes españoles: Costa. Madrid: Imp. de Juan Pueyo; 1917.
12. Costa J. Memorias. Ara Torralba JC, ed. Zaragoza (ES): Prensas Universitarias de Zaragoza; Institución Fernando el Católico; 2011.
13. Peiró Martín I. Razones de los otros: perfiles, memoria e historia de Joaquín Costa. Huesca (ES): Fundación Joaquín Costa; 2013.
14. Díaz Castán V. El final de Joaquín Costa, el enfermo de la mecedora. Anales de la Fundación Joaquín Costa. 2020;32:45-6.
15. Fernández Clemente E. Estudios sobre Joaquín Costa. Zaragoza (ES): Prensas de la Universidad de Zaragoza; 1989.
16. Ara Torralba JC. El Costa de Luis Antón del Olmet o la interesada biografía al uso periodístico de un ex datista. In: Ara Torralba JC, ed. Los grandes españoles: Costa. Zaragoza (ES): Institución Fernando el Católico; 2010. p. 1-9.
17. Ara Torralba JC. Costa íntimo: diarios y escritos de juventud. Anales de la Fundación Joaquín Costa. 2013;27:353-62.
18. Ara Torralba JC. Costa según Costa: notas y escritos autobiográficos (1864-1878). In: En torno a Joaquín Costa: conferencias de Barcelona, 2010. Zaragoza (ES): Institución Fernando el Católico; 2012. p. 28-36.
19. Gómez Benito C. Una introducción al pensamiento reformista de Joaquín Costa. In: Gómez Benito C, Ara Torralba JC, Calvo Carilla JL, Fernández Clemente E, Fontana J, eds. En torno a Joaquín Costa: conferencias de Barcelona, 2010. Zaragoza (ES): Institución Fernando el Católico; 2012. p. 13-26.
20. Gómez Tabanera JM. Joaquín Costa y los idearios de la llamada Generación del 98. In: Sevilla Arroyo F, Alvar Ezquerra C, eds. Actas del XIII Congreso de la Asociación Internacional de Hispanistas (Madrid, 6-11 Jul 1998). Vol. 2. Madrid: Castalia; 2000. p. 219-26.
21. Cheyne GJG. Joaquín Costa, el gran desconocido. Barcelona: Ariel; 1972.
22. Chabás J. Don Joaquín Costa, precursor y actuante de la "generación del 98". Confidencias orales y epistolares. Med Clin (Barc). 1954;22:420-5.
23. Ortega Costa A. Homenaje al profesor G. J. G. Cheyne. Anales de la Fundación Joaquín Costa. 1990;7:89-96.
24. Pou Serradell A, Oliveras Ley C, Periz A, Rives A. Síndrome escapuloperoneal con cardiopatía. Rev Neurol (Barc). 1974;2:41-7.
25. Calandre L, García Albea E, Ricoy JR, Rodríguez Vallejo A, Trueba JL, Peña P. Síndrome escápulo-peroneal. Descripción de un caso con afección miopática y neuropática. Arch Neurobiol (Madrid). 1977;40(4):291-306.
26. Giménez-Roldán S. Dr. Simarro and his time. Neurosci Hist. 2021;9:135-56.

27. Simarro L. Fisiología general del sistema nervioso. Boletín de la Institución Libre de Enseñanza. 1878;2:167-8, 176-7.
28. Simarro L. Fisiología general del sistema nervioso. Boletín de la Institución Libre de Enseñanza. 1879;3:22-3, 31-2, 37-8, 46-7, 53-4, 61-3, 79, 126-7.
29. Kaplan T. Luis Simarro, Spanish histologist. In: Actas del III Congreso Nacional de Historia de la Medicina (Valencia, 10-12 Apr 1969). Vol. II. Valencia (ES): Sociedad Española de Historia de la Medicina; 1969. p. 523-33.
30. Simarro L. El curso de anatomía general de Mr. Ranvier. Boletín de la Institución Libre de Enseñanza. 1881;94:5-7.
31. Guillain G. Charcot (1825-1893). Sa vie, son œuvre. Paris: Masson; 1959. p. 34-8.
32. Guizoni Teive HA, Branco Germiniani MB, Ferreira Camargo CH, Walusinski O, Lees A. "Flâneur Neurologique in Paris": a guide to pinpointing the houses of famous neurologists. J Clin Neurosci. 2018;52:32-6.
33. Aubert G. From photography to cinematography: recording movement and gait in a neurological context. J Hist Neurosci. 2002;11(3):255-64.
34. Scherrer J. Histoire de la physiologie à La Pitié-Salpêtrière de Duchenne de Boulogne. Histoire d'une discipline. La physiologie, fragile? Paris. 16 Oct 2012.
35. Holcomb HS. A wood engraving by Daniel Urrabieta y Vierge (1851-1904). J Hist Med Allied Sci. 1967;22:181-2.
36. Trouvé G. Manuel théorique, instrumental et pratique d'électrologie médicale. Paris: Octave Doin; 1893.
37. Signoret JM. Une leçon clinique à la Salpêtrière (1887) par André Brouillet. Rev Neurol (Paris). 1983;139:687-705.
38. Germiniani FMB, Moro A, Munhoz RP, Teive HAG. Where is Gilles? Or the little mistake in a copy of Brouillet's painting: A clinical lesson at the Salpêtrière. Arq Neuropsiquiatr. 2013;71:327-329.
39. Brigo F, Balasse A, Nardone R, Walusinski O. Jean-Martin Charcot's medical instruments: electrotherapeutic devices in La Leçon Clinique à la Salpêtrière. J Hist Neurosci. 2021;30:94-101.
40. Vázquez de Quevedo F. Instituto de Terapéutica Operatoria (1880–1939): Instituto Rubio y Galí, Instituto Moncloa. Contribución a las especialidades médicas y enfermeras de España. Anales de la Real Academia Nacional de Medicina. 2005;(3):411-32.
41. Barahona JL. José Chabás Bordehore (1877-1963). Tuberculosis y medicina social en la Valencia del primer tercio del siglo XX. Valencia (ES): Generalitat Valenciana; Consell Valencià de Cultura; 2007.
42. Ingram G, Barwick KES, Hartley L, McEntagart M, Crosby AH, Llewelyn G, Morris HR. Distal hereditary motor neuropathy with vocal cord paresis: from difficulty in choral singing to a molecular genetic diagnosis. Pract Neurol. 2016;16:247-51.
43. Goetz CG, Bonduelle M, Gelfand T. Charcot, constructing neurology. New York: Oxford University Press; 1995.
44. Tooth HH. The peroneal type of progressive muscular atrophy [Doctoral thesis]. London: H. K. Lewis; 1886.
45. Pelayo-Negro AL, Carr AL, Laura M, Skorupinska M, Reilly MM. An observational study of asymmetry in CMT1A. J Neurol Neurosurg Psychiatry. 2015;86:589-90.
46. Gourie-Devi M, Nalini A, Sandhya S. Early or late appearance of "dropped head syndrome" in amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry. 2003;74:883-6.
47. Felice KJ, North WA, Moore SA, Mathews KD. FSH dystrophy 3q35 deletion in patients presenting with facial-sparing scapular myopathy. Neurology. 2000;54:1927-31.
48. Kaeser HE. Scapuloperoneal muscular atrophy. Brain. 1965;88:407-18.
49. Orrell RW. Facioscapulohumeral dystrophy and scapuloperoneal syndromes. In: Amato AA, Griggs RG, eds. Muscular dystrophies. Handbook of Clinical Neurology. Vol. 101. Amsterdam: Elsevier BV; 2011.
50. Brodal A. Self-observations and neuro-anatomical considerations after a stroke. Brain. 1973;96:675-94.
51. André CA. Brodal's stroke in 1972: a brilliant self-report by a neuroanatomy professor. Rev Neurol (Paris). 2023;179:251-5.
52. Valko PO, Baumann CR. Sergej Nikolajevich Davidenkov. J Neurol. 2011;258:338-9.
53. Horowski R, Horowski L, Vogel S, Poewe W, Kielhorn F-W. An essay on Wilhelm von Humboldt and the shaking palsy: first comprehensive description of Parkinson's disease by a patient. Neurology. 1995;45:565-8.
54. Marco Igual M. Sergei Davidenkov, the father of Soviet neurogenetics. Neurosci Hist. 2018;6:21-7.
55. Davidenkov SN. Über die neurotische Muskelatrophie Charcot-Marie. Klinisch-genetische Studien. Z Ges Neurol Psychiatr. 1927;107:259-320.
56. Davidenkov SN. Scapulo-peroneal type of progressive muscular atrophy. J Clin Med (Moscow). 1928;6:337-43.
57. Davidenkov SN. Scapulo-peroneal amyotrophy. Arch Neurol Psychiatry. 1939;41:694-701.
58. Kazakov V. What is Davidenkov's scapuloperoneal amyotrophy: is it a myopathic entity or a neurogenic syndrome? What was Davidenkov's opinion concerning this knotty problem? Neuromuscul Disord. 2003;13:91-2.
59. Kazakov V, Bogorodinsky DK, Skorometz A. Myogenic scapuloperoneal syndrome - muscular dystrophy in the K kindred. Eur Neurol. 1975;13:350-9.