

# GASPAR CASAL I JULIÁN

## CASAL NECKLACE

### The eponyms

**Casal necklace or Casal collar.** An area of erythema and pigmentation that appears around the neck in patients suffering from pellagra<sup>1,2</sup>. A pellagrous eruption in the form of an arc around the neck<sup>3</sup>. Brownish or pale red annular erythema, generally with well-delimited borders, that forms around the neck, a typical sign of pellagra<sup>4</sup>. Dermatitis that partially or fully encircles the lower part of the neck, appearing in pellagra<sup>5</sup>.

**Casal disease. Pellagra**<sup>1,3</sup>, a disease due to vitamin B3 (niacin) deficiency. Clinically, it is characterised by gastrointestinal symptoms (diarrhoea, stomatitis), skin disorders (rosy erythema on the skin resembling a necklace glove, or sock), nervous disorders (hyperreflexia, spastic ataxia symptoms, and paresthesia), and psychological disorders. It is also known as Gaspar Casal disease or *Mal de la rosa*<sup>1</sup>.



Gaspar Casal i Julián  
(1680-1759)

## Gaspar Casal i Julián (1680-1759)

The information available about the life of Gaspar Casal i Julián is full of gaps and contradictions. Catalan by birth but Asturian by adoption, his birthplace was a source of controversy for many years. He was ascribed to many nationalities, including Aragonese, Castilian, Asturian, Italian, and Catalan, until his birth in Catalonia was definitively confirmed thanks to the efforts of the dermatologist Jaume Peyrí. Convinced by certain documents that he had been born in Girona, Peyrí asked Josep Vila i Sabater and Tomás Noguer i Musquera, the canon and archivist of Girona Cathedral, to help locate his birthplace in the parish registries pertaining to the diocese of Girona<sup>6</sup>. Finally, in January 1936, Gaspar Casal's birth certificate was found in the records of the parish of Santa Susanna de Mercadal, establishing that he had been born there on 31 December 1680. With the publication of a monograph<sup>6</sup> that contained a photograph of this document, the controversy was laid to rest. The finding was truly a stroke of luck, since five months later all the documents in this church were destroyed by fire at the outbreak of the Spanish Civil War. Without Peyrí's persistence, it is highly likely that Casal's birth would still remain a mystery. No doubt adding to this confusion is the fact that, four years before his death, Casal signed his will as a "native of Utrilla, diocese of Sigüenza" in Soria<sup>7</sup>.

Casal was the son of a Lombard subaltern born in Pavia who, in some documents, such as Gaspar's birth certificate, appears as Federico Xacon i Casal and in others as Federico Casal Dajon. His mother was Magdalena Julián, a native of Utrilla (Soria). It seems that Gaspar soon left Catalonia to settle in his mother's country. It is not known what he did there until he reappeared working as a physician in several villages in the Alcarria region, such as Somolinos, Medinaceli, and Romanillos, and above all, in the city of Atienza (Guadalajara) from 1706 to 1712<sup>6,8</sup>. This period of his life is also controversial, since there is no evidence that Casal had earned a degree in medicine<sup>9</sup>. For many years, nobody questioned his

qualifications until Marañón did so: "Gaspar Casal was lucky enough not to be from the university. Had he been so, his innate capacity for observation would have been drowned in the stupidly theoretical atmosphere that pervaded classes"<sup>10</sup>. At that time, it was not uncommon to practice medicine without a degree, but it is hard to believe he was not awarded a degree at some point in time, considering the important posts that he went on to occupy. In any case, it seems no documentary evidence exists anywhere to confirm his qualification. While some authors have suggested he would have studied at the *Universidad de Alcalá*, the university's records do not include Casal among its graduates<sup>8</sup>.

Casal graduated with a Bachelor in Arts from San Antonio de Porta Coeli College at Sigüenza University in 1713<sup>9</sup>. In this city lived the apothecary Juan Manuel Rodríguez de Luna, who had been trained in Rome, and who taught Casal physics and chemistry. The same year, Casal went to work in Madrid, but it seems that he did not adapt to the city's climate; in 1717 he left for Oviedo<sup>6</sup>. Some authors have surmised that he might have been fleeing from the Inquisition, which had issued an ignominious sentence against his wife, María Ruiz, that also affected him and their children<sup>7</sup>. This is another obscure aspect of his life, since no documentary evidence has been found to substantiate the Inquisition's accusation. Yet there are no doubts that the Asturian ecclesiastical authorities ordered that Casal's children were to be banished from the Principality of Asturias on reaching the age of twelve years. Certain authors have questioned why Casal abandoned his clients in Madrid and travelled to Asturias, a difficult place to make a living in the early 18th century, suggesting that his wife may have been exiled to Asturias by the Inquisition<sup>11</sup>.

In Asturias, Casal would undertake his most interesting work, for which he is remembered both nationally and internationally. Initially he was protected by the Duke of Parque, but he did not earn enough to maintain his large family. So in March 1720, he applied for and was awarded the

post of municipal physician in Oviedo, a post he would leave on 5 March 1729 when he was chosen as physician of the *cabildo* (council) of the Cathedral<sup>10</sup>. The responsibilities of this position included care of the hospitals of Santiago, San Juan, and Santa María de los Remedios<sup>8</sup>. He maintained good relations with Father Benito Feijoo, one of the key intellectuals of the Enlightenment in Spain. Casal himself gradually became a respected physician thanks to his critical view of life and significant contributions to the fields of natural history, epidemiology, nutrition, and medical pathology.

Though he had declared that he wished to remain in Asturias, Casal returned to Madrid in 1751 to attend the Spanish queen, Barbara of Portugal. It is hard to understand why a 70-year-old man would choose to move to the capital and begin a new life, though it appears Father Feijoo had a notable influence in his decision. On 24 August he was appointed temporary (later permanent) physician of Fernando VI Royal Chamber; on 8 January 1752, he was appointed chief physician of the Kingdom of Castile<sup>6,7</sup>. He was later elected a permanent member of the *Real Academia de Medicina de Madrid* reaching the highest pinnacle of recognition for a physician of this period.

Casal died in Madrid on 10 August 1759, the same day as King Fernando VI. He was buried in secret, albeit with the vicar's permission, in the church of San Sebastián in Madrid<sup>6,8</sup>. No documents explaining the need for such secrecy have been found.

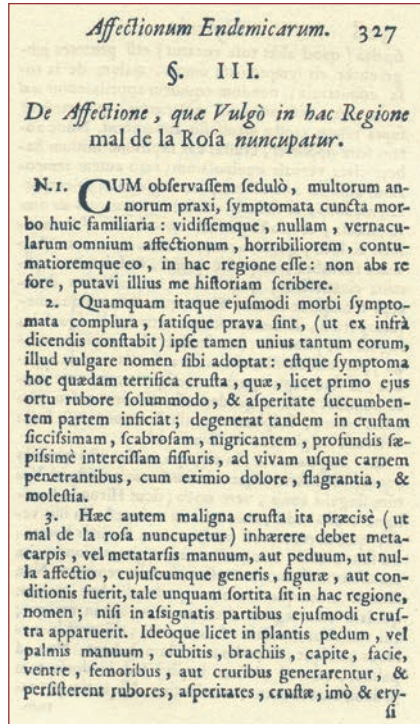
Gaspar Casal is the author of a single surviving work, which seems to have been written about 1735 and published posthumously in Madrid in 1762, entitled *Historia Natural y Médica del Principado de Asturias* (Natural and Medical History of the Principality of Asturias). This work is in fact a compilation of several books, whose only apparent connection is that they were penned by Casal. The most important are *Historia physico-medica del Principado de Asturias* (Physical and medical history of the

Principality of Asturias); *Historia de la constitución de los tiempos, y de las enfermedades epidémicas, y particulares, que en este Principado de Asturias observamos desde el año 1719 hasta el de 1721* (History of the constitution of the times, and of epidemic –and individual– diseases, that in this Principality of Asturias we have observed from 1719 until 1721); *Constitución de los tiempos* (Constitution of the times); and *Historia affectionum, quarumdam regionis hujus familiarum* (History of some diseases endemic to this region). The original work was not republished until 1900 and has since been reprinted several times. A facsimile edition was published by the government of Asturias in 1988.

## Casal necklace and Casal disease

The third chapter of *Historia affectionum, quarumdam regionis hujus familiarum* contains a description of what Asturian peasants called the “*mal de la rosa*”. This point is noteworthy because some writers credit Casal with creating this expression. In his work, Casal described an endemic disease observed in this region among poor farmers whose diet was based almost exclusively on maize, with no ingestion of fresh meat. This clinical presentation was called “*mal de la rosa*”, as the chapter heading describing it states: “*De affectione, que Vulgò in hac Regione mal de la Rosa nuncupatur*” (In this province, this affliction is known among the common people as “*mal de la rosa*”), since affected individuals displayed a typical reddening of the skin and an exanthema with vesicles on the back of hands and feet, as well as a similar outbreak on the neck. Casal had mentioned that the skin displayed “reddening, roughness, a scab, and erysipelas”. Casal described the disease in the following way:

“Having, over many years of practice, carefully observed all the peculiar symptoms of this disease, and having noted that, of all the common afflictions in this country, none is more horrific nor stubborn, I judged that it would not be inopportune to write its history.”

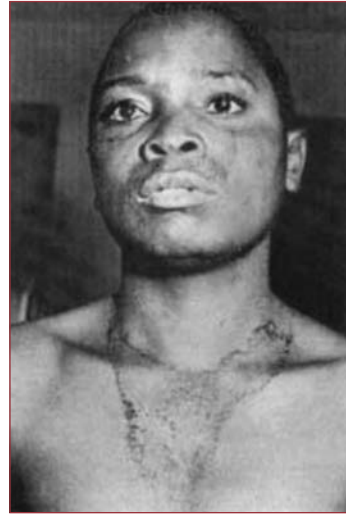


First page of the chapter describing the “mal de la rosa” in the original edition of Casal's *Historia physico-medica del Principado de Asturias* (Physical and medical history of the Principality of Asturias) published in Madrid in 1762

In the following lines, Casal accurately described the dermatological characteristics of the disease. The last paragraph of this description is devoted to the sign for which it is especially well known:

“Another visible sign appears in this class of patient, though not in all, and that is a scabby roughness of a dark ashy colour in the lower frontal neck region, like a necklace, extending from one side of the nape to the other, over the collar bones and the upper edge of the sternum, having the width of one or two fingers, like a strip, and which, often leaving the upper part of the nape intact, just reaches, at its edges, the borders of both trapezius muscles without extending further. In its central section, an appendix of equal width descends

Casal necklace on a patient with pellagra<sup>15</sup>



from the sternum to the mid-point of the thorax. No such sign can be found in any person, sick or healthy, except in patients suffering from the "*mal de la rosa*", from which I infer that it occurs solely in them, though not in all".

Casal also suspected that there might be a relation between diet and the disease, though he had doubts, as the lines below show:

"1) The cause of this disease should be sought in climatic or atmospheric conditions, or in the patients' constitution or diet. Yet, since I have analysed the climatic conditions extensively enough, in the *Physical and Medical History* of this region, which I myself wrote and keep in my house, and therefore not to repeat what has been said, I currently propose treating solely diet. 2) Maize or Indian corn is the main food of almost all those who suffer from this affliction; in fact, the flour of this cereal is used to create their bread, and from it are made the gruels which some mix with milk or with milk butter, for their ordinary fare [...]. Only rarely do they eat fresh meat;

furthermore, only occasionally do they eat cured meat, since almost all those having this disease are poor farmers, so it is not possible to eat bacon or the cured meat of another animal daily, not even once in three days [...]. 3) Such a diet might unexpectedly seem to some enough to produce not just this disease but also other, worse ones. Yet he who, having thought about this calmly, had carefully observed all the concurrent circumstances, would truly not fail to find great difficulties that have nothing to do with those who maintain such an opinion: 1) because throughout almost the entire province all the peasant farmers eat such foods and, notwithstanding, not all are afflicted by this disease... 2) because this "*mal de la rosa*" is not found equally throughout the region, but especially in one section or territory of it... 3) because a theory as abstruse as diet is, as is said, *a priori* and obscure, so that I do not think any clear reason can be deduced from it."

The original description of the *mal de la rosa* became famous thanks to its dissemination by François Thiéry, the former physician of Louis XIV of France, who remitted the definition of the disease to the dean of the Faculty of Medicine in Paris, A. F. Chomel, in 1755<sup>11,12</sup>. This enabled it to be included in Saunders' Medical Nosology under the name *Lepra asturianensis*<sup>7</sup>, but did not stop the *mal de la rosa* being better known in the future as pellagra, an adaptation of the term popularised by Frapolli when in 1771 he used "pellagra" to refer to *mal de la rosa* (from *pelle agra*, or "rough skin")<sup>12</sup>. It would also be known –and this denomination remains valid today– as Casal disease, or Gaspar Casal disease<sup>1</sup>.

In ensuing centuries, the *mal de la rosa* was recognised in the poorest classes of Lombardy, Venice, and the south of France. The disease remained endemic among poor farmers with diets almost exclusively of maize, until it was also described in the US in the early 20th century, where it took on epidemic proportions in the southern states<sup>13</sup>. Once more, poverty and consumption of maize were recognised as the main



risk factors, causing physicians at the time to pinpoint this cereal as being responsible for the disease. In 1914, it began to be suggested that a nutritional deficiency could be behind it, but it would take 18 years before Goldberger and Tanner would suggest that the disease was associated with the lack of an amino acid. It was Goldberger himself who confirmed that it was in fact a dietary deficiency, finally identifying the preventive factor of pellagra as nicotinamide<sup>13</sup>. It is now known that primary pellagra is a consequence of a niacin (nicotinic acid) deficiency, resulting from inadequate intake of niacin and/or its precursor, the essential amino acid tryptophan. Secondary pellagra appears when there is sufficient intake but pathological interferences hindering adequate availability, such as prolonged diarrhoea, anorexia nervosa, chronic alcoholism, chronic colitis, severe ulcerous colitis, Crohn disease, or

Engraving of Casal's work, displaying the characteristics of the "mal de la rosa" (Casal disease), clearly showing Casal necklace



hepatic cirrhosis, among others<sup>13</sup>. It can also appear in association with isoniazid treatment when pyridoxine is not administered concomitantly, as the latter is necessary for the synthesis of nicotinamide.

The clinical manifestations of pellagra are the classic triad of dermatitis, diarrhoea, and dementia, although they do not always appear in this order. Atypical forms exist that are oligosymptomatic. The dermatological manifestations are very constant; when absent, it may be difficult to reach the diagnosis. These consist of a symmetrical, bilateral eruption in the areas exposed to sunlight, which can be painful to the touch in the acute phase. It begins as an initially acute or intermittent erythema, progressing to an exudative eruption on the back of the hands, face, neck, and thorax with a stinging, burning sensation. The affected areas are initially red and clearly defined, but progressively change to cinnamon and brown tones.

They also appear in areas subject to pressure, friction, or heat. Initially, they can be confused with sunburn, but are differentiated by later darkening<sup>13</sup>. The distribution of affected areas depends on exposure to the sun. Initially flaky and erythematous areas are observed on the face, back of the hands, and forearms, accompanied by itching. In a more advanced phase, the dermatosis worsens and the skin becomes rough. Lichenification and pigmentation processes appear, and end up cracking. It is common for the neck, nape, and sternal region all to be affected, creating the typical Casal necklace.

The back of the hands is the most frequently affected area (77%-97%), though in fact, many other areas can be affected: feet, legs, shoulders, elbows, forearms, and knees. On the face, an erythema appears that may follow the innervation of the trigeminal nerve and a butterfly-wing eruption similar to erythematous lupus may appear. Equally frequent is the appearance of a well-defined eruption on the front of the thorax, extending in a wide band or necklace completely around the neck to the

regions of dermatomes  $C_3$  and  $C_4$ . The upper limit reaches to the hairline in the back and the larynx in the front, while the lower limit starts in the hollows of the spinous processes, reaching the sternal manubrium. The following may appear in its evolution: vesicles or phlyctenae, ulcers, intense dryness, hyperpigmentation, and flakiness in areas not exposed to light. It is believed that a low concentration of urocanic acid in the skin could explain the photosensitivity to sunlight, given the capacity of this compound to absorb ultraviolet light<sup>14</sup>.

Casal's work is striking for its clarity and scepticism, features that make him one of the most notable scientists of the Spanish Enlightenment. In this sense, García-Valdecasas<sup>12</sup> words are apt:

"His clarity of understanding surprises me, as does his objectivity and lack of prejudice, observing the facts, without allowing himself to be influenced by a number of philosophical principles and systematic deductions on which the ingenuities of his age became shipwrecked. His independence of criteria compared to the prejudices of the period are not found in any author from those times, not even a hundred years later. While his discovery of the mal de la rosa is

*El médico* (The doctor, 1779)  
by Francisco de Goya y  
Lucientes. It is argued that  
represents Gaspar Casal with  
two young students



notable, even more worthy, in my view, was his clarity of thought and his scientific criteria, independent of the prejudices of his time, which place him among the great men of his period”.

Casal was a man of his time: as well as a physician, he was one of the most important Spanish scientists in the first half of the 18th century. He precisely described for the first time a disease that was the result of a dietary deficiency. His is one of the Catalan medical eponyms that is most used internationally. Even today, numerous publications and images refer to Casal necklace, as illustrated in with its inclusion in a FAO publication<sup>15</sup>.

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