

Historical Note

First description of polycystic kidney disease in a Portuguese journal

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The first description of polycystic kidney disease (PKD) in a Portuguese medical journal occurred in 1918 when Dr Custódio Cabeça published a manuscript in *Medicina Contemporânea*, a medical journal founded in 1883 by Dr Miguel Bombarda (1851–1910) and Dr Sousa Martins (1843–1897), two important teachers at the Medical School in Lisbon [1]. A reprint of the manuscript dedicated to his colleague, Professor Daniel de Mattos (1845–1921), who was the teacher of Surgery in Medical Faculty of the University of Coimbra, was found by manual searching in the files of the library of this institution (Figure 1). In this manuscript Dr Cabeça reported his experience in treating eight patients with PKD (two of whom he nephrectomised) and reviewed the current knowledge on the pathophysiology of PKD. In his paper, Dr Cabeça started reviewing the pathophysiology of the disease and gave preference to the theory of ‘fetal papillitis’, or obstructive theory, that was proposed by Virchow (1821–1902), as opposed by the theories of neoplastic degeneration and the theory of congenital origin of the disease.

According to this theory, polycystic degeneration is in part of congenital origin and in part acquired. In its essence the lesion is inflammatory and cyst arise by retention. Chemical agents with irritant properties are transmitted to the son by the mother and, therefore, initiating its effects in the fetal kidney. Moreover there is an interstitial nephritis and the new conjunctive tissue compresses the urinary canals inducing urine retention and, later, dilatation of the canals forming cavities. Other authors, following Virchow, believe that urinary retention can be caused by occlusion of the tubuli by colloid, hyaline or fibrinoid casts formed as a consequence of the nephritis.

His choice was based on the findings in pathological analysis of the specimens he nephrectomised where signs of chronic inflammation and vascular congestion resulted in obstruction of the straight tubules in the renal papilla. He rejected the neoplastic theory, based on the slow evolution of the disease, as well on the signs of benignity in the proliferative epithelium of the cysts. Interestingly, he

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referred to a recent publication in Surgery Gynaecology and Obstetrics that appeared in 1916, that we assumed as that of Braasch [2] in Mayo Clinic, and divided the clinical aspects of polycystic kidneys into those with medical treatment, that frequently died on uraemia, and those with surgical treatment where haematuria, abdominal mass and pain were the main symptoms.

The evolution of polycystic kidney can be slow and could last 10 to 20 years. In my opinion it is practical to differentiate polycystic kidney in two separate forms:

- medical form, presenting with symptoms of chronic nephritis and progressing to uraemia.
- surgical form, presenting as tumour, pain or haematuria.

Additionally I will add a third form, unilateral disease, also a surgical form that can be treated by nephrectomy.

It is in the surgical presentation that complications such as cyst rupture or infection can be found.

Prognosis of polycystic kidney is always ominous as the disease has a tendency to affect both kidneys. Even in the surgical presentation that can be nephrectomised we cannot exclude that the other kidney will be affected as well.

Treatment

- (a) medical form, should be treated conservatively.
- (b) surgical form, should be nephrectomised.

In this later form are included unilateral cases associated with sufficient renal function of the other kidney. In symptomatic cases (pain, haematuria compression of other organs) when the other kidney is insufficient we should use nephrostomy.

In his paper, entitled ‘Surgical treatment of Polycystic kidney’, Dr Cabeça reported two cases that were treated by unilateral nephrectomy because of the volume of the kidneys that were detected by abdominal palpation. One case, of a young woman of 22 years old, had a follow-up after 10 years and remained well, except for albuminuria (1 g/l). In the other case, in order to assure that the opposite kidney had adequate renal function, separate ureteral catheterization with urine biochemistries was performed. Eosinophilia

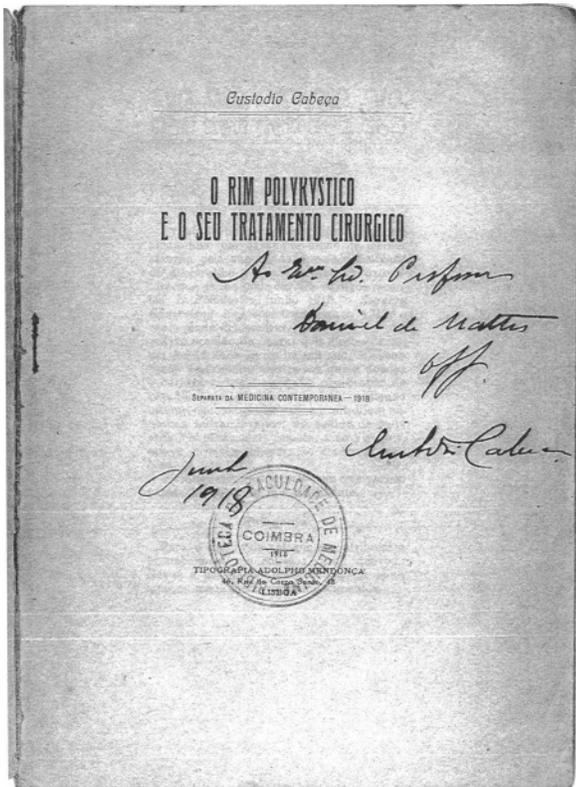
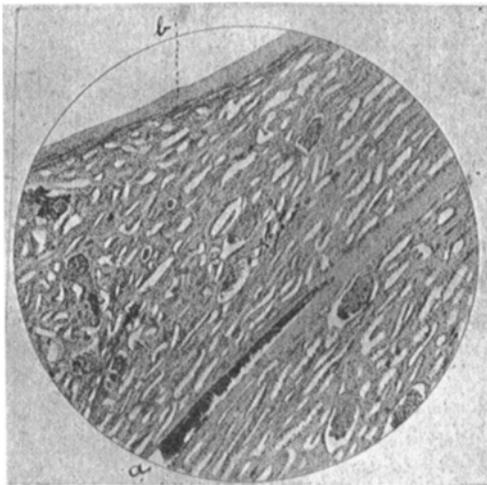


Fig. 1. Reproduction of the cover page of a reprint of the original article published by Custódio Cabeça, in *Medicina Contemporânea*, in 1918 (Courtesy: Biblioteca da Faculdade de Medicina da Universidade de Coimbra).



a. grande foco hemorragico n'um tubuli recto.
b. parede d'uma cavidade kystica constituída por espessa membrana conjunctiva, onde se veem raros e pequenos nucleos de celulas epiteliaes achatadas lembrando o endotelio vascular. (Ocul. 2. — Obj. 3. — 160).

Fig. 2. Reproduction of the image showing pathological analysis of a renal cyst wall (original legend: *a*—haemorrhage in a straight tubule; *b*—wall of a cystic cavity showing a conjunctive membrane with rare and sparse nuclei of flattened epithelial cells resembling vascular endothelium) (Courtesy: Biblioteca da Faculdade de Medicina da Universidade de Coimbra).



(Face posterior)

Fig. 3. Reproduction of a photograph included in the manuscript showing a polycystic kidney (*posterior view*) (Courtesy: Biblioteca da Faculdade de Medicina da Universidade de Coimbra).



PROF. CUSTÓDIO CABEÇA
(1866 — 1936)

Fig. 4. Reproduction of a photograph of Custódio Cabeça that appeared in a manuscript of his homage published after his death [4].



Fig. 5. Professor Egas Moniz in a Conference in the Faculty of Medicine of Lisbon presenting his results of the first cerebral arteriography. In the first row of the audience, Professor Custódio Cabeça is the second from the left [13] (Courtesy: Camara Municipal de Estarreja).

was a criterion to conclude or exclude the diagnosis of hydatidic cysts.

Pathological report was performed by Professor Azevedo Neves (1877–1955), teacher of Forensic Medicine at the Medical School (Escola Médico-Cirúrgica de Lisboa), who noted the large size and weight of the kidney (17 cm and 472 g, respectively) and that described the organ as a bunch of grapes filled of cysts ranging from the size of a grain of corn up to that of a hazelnut. On microscopic examination of the cysts, he noted that the epithelia lining in the cysts is flattened and stretched and resembling the endothelium. However this epithelial lining was missing in large extension of the cystic wall that is lined by a thick layer of conjunctive tissue (Figure 2). The manuscript also included photographs of a polycystic kidney (Figure 3).

Custódio Cabeça

Professor Custódio Cabeça (1866–1936) studied at the Medical School in Lisbon and started his surgical activity in 1891 performing more than 7000 interventions (Figure 4). In 1898, he made his application as a teacher at the Medical School in Lisbon teaching surgery until 1936 where he was fellow of Egas Moniz (1874–1955) who was awarded the Nobel Prize of Medicine in 1949 (Figure 5). He published several books on surgical subjects such as tumours of the breast, and tumours of the ovary, as well as several manuscripts in Portuguese medical journals. His experience in the surgical treatment of bone sarcomas was well recognised, e.g. by Professor Askanazi and Professor Leriche who visited him in his clinic, in 1930 and 1932, respectively [4].

PKD in Portugal

Although cystic kidneys were probably noted since antiquity [5], and descriptions in the seventeenth century's works by renowned anatomists are available [6], PKD as an independent clinical entity was first suggested in the work of Pierre Rayer (1793–1867) [5]. However, the term 'polycystic' was used for the first time by Felix Lejars (1863–1932) in 1888 [7] in his PhD thesis, and it is interesting to find that in 30 years the term was well accepted worldwide.

Unfortunately the publication of Dr Cabeça was not followed by systematic review of other cases of PKD and another important publication in this subject, in a Portuguese journal, appeared only in 1940, when Cândido da Silva (1914–2008) reported the first renal angiography in a PKD patient [3]. For long time, thereafter, only case reports were published in the Portuguese medical literature and only after the advent of the genetic analysis in the 1990s that studies in cohort of patients with PKD appeared in indexed medical journals [8–12].

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