

THE EMPEROR DOM PEDRO II

His convulsive seizures when a boy

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ABSTRACT - Introduction: Dom Pedro II, the Prince Heir and Emperor under regency, in a delicate period of the construction of the Brazilian nation, had convulsive seizures. **Objective:** To investigate the convulsive seizures and related syndromes of Dom Pedro II and his family, besides the physicians in charge of the health care. **Method:** Narrative review based on primary and secondary sources. **Conclusion:** The scattered and self-limited convulsive seizures associated with physical and mental integrity favored a benign prognosis. Dom Pedro and his family presented rich history of epileptic seizures and febrile convulsion. This variety resembles the diagnosis of generalized epilepsy with febrile seizures plus that seems to be a combination of several syndromes with shared genetic susceptibility.

KEY WORDS: Dom Pedro II, epilepsy, genetics, history of Brazil, monarchy.

O Imperador Dom Pedro II: as suas crises convulsivas quando menino

RESUMO - Introdução: Dom Pedro, o príncipe herdeiro e imperador sob regência, em período delicado da formação da nação brasileira, apresentou crises convulsivas que geraram preocupação para o país. **Objetivo:** Investigar a história da epilepsia de Dom Pedro II e da sua família e procurar identificar quais tipos de crises epiléticas estavam presentes, além dos médicos envolvidos com os cuidados de saúde. **Método:** Revisão narrativa baseada em fontes primárias e secundárias. **Conclusão:** As crises convulsivas esparsas e auto-limitadas associadas a hígidez física e mental de Dom Pedro II sugerem um prognóstico benigno. A história de epilepsia idiopática e convulsões febris no imperador e em outros membros da sua família aponta para o diagnóstico mais provável de Epilepsia Generalizada com Convulsões Febris Plus que é determinada por uma combinação de alguns tipos de manifestações epiléticas com suscetibilidade genética compartilhada.

PALAVRAS-CHAVE: Pedro II, epilepsia, genética, história do Brasil, monarquia.

Brazil, from 1824 until the declaration of the republic, was a monarchy¹. The formation of the second Brazilian Emperor - Dom Pedro de Alcântara Bragança e Habsburgo, 1825-1889 - was *sui generis*: an orphan raised by the State and for It. Dom Pedro II spent much of the his infancy isolated, and he was educated by servants and tutors. Two sisters would be the only members of the family to coexist with him lengthier. He was an insecure child, and he suffered convulsions, situations that brought great concern to the Brazilian Government^{2,3}. In this period, the Brazilian Medicine and the country were consolidating.

BIRTH AND UPBRINGING OF A NATION'S ORPHAN AND PUPIL

Pedro II was member of the Brazilian royal family as son of Dom Pedro I de Bragança e Bourbon, person with epilepsy⁴, Liberator of Brazil from Portugal, and

of Portugal from the absolutists, and of Dona Leopoldina de Habsburgo e Bourbon. D. Pedro II was an intellectual. He was born at Quinta da Boa Vista, in Rio de Janeiro, on December 2, 1825. He was the heir of the Brazilian's crown heir, of the dynasty of the House of Bragança, and son of one of the oldest and noblest European royal families⁵. However, this child was born in circumstances less than desirable: in an incipient and unstable empire; mother orphan; with father abroad and, in short time, died. Dom João VI, his grand father, left behind him his son, Dom Pedro (later called Dom Pedro I of Brazil and IV of Portugal), in 1821, who left also behind him his heir son, the Heir, the future Pedro II, in 1831⁶. The Emperor Boy destiny was kept in the hands of the regents who would make decisions concerning with the future of Brazil, the monarchy and the boy himself^{7,8}. The period of the regencies, waiting for the Dom Pedro II corona-

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tion, was from 1831 until 1840. The importance of the role of the tutor at the time of many rebellions, and in a nation still not consolidated is well understood. On July 23, 1840, the Brazilian Imperial Parliament abolished the regency and declared that Dom Pedro II was mature to govern, despite his 14 years of age⁵. Not surprisingly, Brazilian politics expressed skepticism about the capacities of an inexperienced adolescent emperor. One year after, he was consecrated and crowned. He initiated a reign that lasted 49 years, and that finished with the proclamation of the Republic.

DOCTORS OF THE IMPERIAL CHAMBER. THE BUILDING OF THE HOSPICE OF DOM PEDRO II

The doctors always had been next to the Brazilian Monarchy, and were politically influential. Medical education deserved great attention. During the Trina Regency, and on behalf of the Emperor, on October 3, 1832, the Reformation of the Medical Education was homologated, when the course was fixed in six years and the two existing Schools of Medicine had been transformed into the School of Medicine of Bahia and Rio de Janeiro⁹. The Emperor coming of the age was celebrated, among other events, with the construction of the Hospice Pedro II under the influence of Jose Clemente Pereira, that joined the duties of Minister of State with the ones of Provider of the Santa Casa de Misericórdia. The first hospital for the treatment and isolation of the insane people in Brazil (Fig 1) was, then, built in the region of the Praia Vermelha, the old Praia da Saudade. Its construction was initiated on November 2, 1842 and its inauguration took place on November 30, 1852¹⁰. In this hospital, great names of the Brazilian medicine pontificated, and in the following century, they constructed in its backyards the Institute of Neurology as well as the Institute of Psychiatry of the present-day Federal University of Rio de Janeiro (UFRJ). In the main hall of the Forum of the Science and Culture of the UFRJ, ex Hospice of Dom Pedro II, stands the marble statue of Pedro II, at the age of 15, with the garment of the coronation (Fig 2), and in front of the Emperor, stands the



Fig 1. Hospice Dom Pedro II. Foto by Augusto Stahl - CoordCOM / UFRJ – reproduction authorized.

statue of Jose Clemente Pereira¹⁰. Dom Pedro II kept narrow relationships with the Faculty of Medicine of Rio de Janeiro as well as with the Imperial Academy of Medicine, currently Faculty of Medicine of the UFRJ and National Academy of Medicine, respectively. Some doctors of the Imperial Chamber were members of these institutions, and the monarch frequently attended their sessions. Since his birth, in his daily life, due to his misfortunes of health, especially in the convulsive seizures, Dom Pedro II was cared for by the doctors of the Imperial Chamber. These doctors were also assigned to determine the temperature of the royal bath, and to observe the meals of the Emperor, evaluating the food and preventing its excessive ingestion⁷. In the initial phase of the life of Dom Pedro II, the most distinguished doctors were: Domingos Ribeiro dos Guimarães Peixoto who attended Dona Leopoldina at Dom Pedro II birth⁵, besides Francisco Manoel de Paula, Baron of Inhomirim. Both attended the small prince in his seizures of August 3-4, 1827¹¹.

EPILEPTIC SEIZURES IN THE HOUSE OF BRAGANÇA

There are reports of some Dom Pedro II convulsive seizures, from 1827 until 1840. Some sources comment his three seizures on August 3-4, 1827, at the age of



Fig 2. Statue of Dom Pedro II in honor to his coronation, located in the old Hospice of Dom Pedro II, currently Forum of the Federal University of Rio de Janeiro. Foto by Bira Soares - CoordCOM / UFRJ - reproduction authorized.

two years, that had been considered "bothering proper of the age and aggravated at the time of the dentition"². Three eminent physicians had leaned over the small bed of the patient, according to the report of Rangel². The seizures had occurred at hours 18, 23 and 6, being followed by fever¹. At the age of 8 years, on the night of October 4 to 5, 1833, Dom Pedro was victim of a "indigestion" followed by manifestation of the "tremendous hereditary harm" that was described as similar to the tetanus convulsions. The fever early appeared and abated no sooner than two days after the "accident". Although the interval between the two registered episodes was 6 years, apparently the seizures were more frequent, as suggested by the Representative of the Austrian government, Baron Daiser, on April, 1834, *apud* Barman³: "Pedro II's physical growth led him to eat more, and the heavy meals gave him indigestions which almost always had as the result a small nervous movement which disappears after a very few days". The Austrian considered that the cause of this discomfort would be the type of diet: "noxious to the health, above all the practice of constantly eating sweet things"³. To the 14 years, on March 23, 1840, Dom Pedro II suffered new seizures preceded by mild pain in the left eye, loss of consciousness and convulsive movements, mainly in the muscles of the face and eyes, being the inferior extremities in spasm. The consciousness was recovered within few minutes. He woke up with pain and feeling a heavy head. Friction with water of colony after the return of the consciousness was carried out; sinapisms were applied to the foot². The doctors of the Royal House recommended that the studies of the Emperor be reduced, that any intellectual effort after eating be prevented, and that exercises such as horse riding and gymnastics be recommended². Another seizure occurred on the first day of the following month, although less intense³. On March 27, 1840, so as to reduce speculations about the health of Dom Pedro II in the court, a daily, *Jornal do Commercio*¹² published a note, signed by the doctor of the week – J.C.S. de Meirelles - that the bulletins about the health of the Emperor would be suspended, once there was not fear of relapse of the disease that attacked him. However, on April 14, 1840, the same Baron Daiser who commented 6 years before about "the indigestions" of the boy would have declared that: "very strong signs of his attachment to the imperial family and of the importance it places in the life of the young monarch, on whom the hope and all the future of Brazil rests", *apud* Barman³. Dom Pedro II had a rich family history of epilepsy or convulsive seizures: his father was epileptic and presented generalized seizures predominantly tonic-clonic, possibly begin-

ning at the age of 13 years; aunts on the paternal side also presented them; Fernando I of Habsburgo-Lorena, uncle on the maternal side in the same way⁴. His mother, Dona Leopoldina, and his grandfather on the paternal side, Dom João VI, died in the course of symptomatic epileptic seizures underlined by a cerebral acute insult. Two brothers of Dom Pedro II died of convulsions: in 1822, Dom João Carlos, "who died of an inflammation of the liver, in epileptic seizures that lasted twenty eight hours"; and Dona Paula Mariana died of fatal encephalitic secondary seizures². Dona Januária, another sister of Dom Pedro II, "had, for the third time, an attack of epilepsy on June 8, 1830² and Dom Pedro I linked this last seizures to the fever, *apud* Rangel². Dom Pedro II had four children: Afonso Pedro (1845-1847), Isabel (1846-1921), Leopoldina (1847-1871), and Pedro Afonso (1848-1850). The first son died of convulsions ("at midday he was playing in the corridors....") as well as the second male child². Princess Isabel, on May 30, 1847, had convulsions that lasted three days, as Rangel tells². There is also reference that a grandson of Dom Pedro II, Prince August de Saxe-Coburgo had seizures, and according to Conde d'Eu it was associated with fever...."et en outre est sujet à des accès de fièvre et d'épilepsie", *apud* Rangel². Summarizing, a significant number of ascendants and of descendants of Dom Pedro II presented convulsive seizures, primary or secondary. His paternal grandfather, mother, two brothers and two children had died in the course of convulsive seizures, at the time that the medicine did not count on efficacious therapeutical measures for the majority of the illnesses, nor efficacious anti-epileptic drugs. It is not improbable that the two children of Dom Pedro II died of a status epilepticus febril (SEF), considering that: the febrile convulsions (FC) are the most common form of convulsive symptomatology in infancy; the age of both is one of FC occurrence; great percentage of children who have FC presents SEF¹³; other relatives suffered from FC; the report of Dom Afonso death mentions that hours before he was in excellent conditions of health as can be observed in the context of the FC. Dona Januária, sister, and Prince August, grandson of Dom Pedro II, suffered from FC as registered by Dom Pedro I and Conde D'Eu². Possibly, the convulsions suffered by Princess Isabel were of the same nature, taking into account, one more time, the age at which they happened (1 year). The situation lived by Princess Isabel on May, 1847, characterized by the repetition of the convulsions for three days defines one of the aspects that characterize the atypical febrile convulsions, complicated or complex¹⁴. It seems that the father, paternal aunts and maternal uncle of Dom Pedro II suffered from

generalized tonic-clonic type of epilepsy - probable from idiopathic epilepsy⁴. The history of Dom Pedro II signals the presence of convulsive manifestations since 2 years of age - until 8 years associated with fever - and, thereafter, afebrile seizures, focal or generalized tonic-clonic. The recurrence of the seizures was sporadic and the last manifestation seems to have occurred in the adolescence. The clinical findings of Dom Pedro II and his family fulfill the diagnostic criteria of the so called Generalized epilepsy with febrile seizures plus (GEFS+). This entity is a chanelopatía, defined by the presence of FC that extend after the age of 6 years, associated to different types of afebrile epileptic seizures, mainly tonic-clonic, of strong genetic influence, with some biological markers already known¹⁵. Nordli¹⁶ emphasizes that the family members have experienced febrile seizures and usually mild or relatively infrequent convulsions. However, some have exhibited more intense forms of epilepsy that could be classified as myoclonic-astatic epilepsy and severe myoclonic epilepsy in infancy¹⁶. Although, The ILAE indeed considers that GEFS+ a syndrome in evolution¹⁶. Despite the evidences in favor of the diagnosis of the GEFS+, we must mention the possible alternative diagnosis regarding the benign and precocious epileptic picture of Dom Pedro II. One of them, would be the benign childhood epilepsy with centrotemporal spikes (BCECTS) regarding the description of Dom Pedro "shake in the face". BCECTS is the most frequent epilepsy in school-age, the onset is between 3 and 13 years, remission before age 15-16 years, and there is a high genetic predisposition, is characterized by brief, simple, partial, hemifacial motor seizures, with or without somatosensory symptoms, sometimes evolving into generalized tonic-clonic seizures, usually related to sleep¹⁷. "Early-onset benign childhood seizure susceptibility syndrome with occipital or extra-occipital spikes", as suggested by Panayiotopoulos can also be mentioned, regarding the so called "indigestions" related to the convulsions of the Emperor boy. It is the second most frequent epilepsy syndrome of childhood. Age of onset is between 2 and 8 years, with a peak at age 5 and remission before age 12. Clinical semiology is characterized by brief or prolonged, usually nocturnal, infrequent partial seizures, consisting of a combination of autonomic and behavioral disturbances, vomiting, deviation of eyes, and often impairment of consciousness that can progress to hemi and/or generalized tonic-clonic convulsions. Among the generalized idiopathic syndromes, the benign myoclonic epilepsy of infancy deserves mention regarding the differential diagnosis of the Emperor seizures. It has a family history of epilepsy or febrile seizures in about 40% of cases,

according to Nordli¹⁶. Jerks rarely cause falls, and children present with head drops, upward eye deviation, or brief jerks of the arms, daily, or sometimes as clusters¹⁶.

In conclusion, the health of The Emperor Boy - educated to be the Head of the Brazilian State and agglutinator joiner of a fragmented nation - raised doubts about his competence to take on these functions. The convulsions, observed since the age of 2 years, were one of the main reasons for this skepticism. Dom Pedro II as well as several of his ascendants and descendants presented epileptic manifestations: idiopathic seizures, acute symptomatic seizures, and also febrile convulsions. The analysis of the different types of seizures suffered by the family group suggest the diagnosis of GEFS+ that is a combination of many epileptic syndromes with shared genetic susceptibility¹⁶.

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