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# "La Surdité Familiale" by Giuseppe Gradenigo 1921

## ABSTRACT

Giuseppe Gradenigo was born in Venice in 1859, and he was a member of the noble Venetian family of the Gradenigo. After graduation in medicine in Padua, he went to Vienna where he deepened his studies in otology and embryology. He obtained, in 1888, the "libera docenza" in otology in Padua, and in 1889, he began to direct the section of otolaryngology of the polyclinic of Turin. After a brilliant career in 1917, he took over the chair of otolaryngology at the University of Naples. Gradenigo was a pioneer in otolaryngology and audiology and a reference figure in the clinical and scientific fields. In this paper, the most important aspects about his works and his biography will be discussed.

Keywords: Audiology, Giuseppe Gradenigo, otology

Gradenigo published in 1921 Contribution a l'étude de l'hérédité morbide chez l'homme: La Surdité Familiale. Sa prophylaxie, son traitement. This monography is the third of the Monographies Oto-Rhino-Laryngologiques Interantionales, Amédée Legrand éditeur in Paris, written in French with extensive summaries in German, English, Spanish, and Italian<sup>1</sup> (Figure 1).

Gradenigo was a great otologist with a profound interest in audiology (see the extensive report in Stephens, Orzan and Galletti di San Candido: *Giuseppe Gradenigo and His Contributions to Audiology, 1997*),<sup>2</sup> and he is well known for his contributions to otology (particularly important the ones on embryonal development of the ear, 1924)<sup>3</sup> and for his description of Gradenigo's syndrome (1904)<sup>4</sup> (Figure 2).

In this monography, Gradenigo wrote: "Biological doctrines on hereditary are of a very recent date. Clinicians, and otologists in particular, have not paid to them the attention that they deserve. There exist, however, two important forms of deafness which may be ascribed to heredity and that may be called family deafness." Gradenigo reports his personal experience of collecting "more than one hundred of genealogical trees upon patients affected with family deafness," and this proves an interest in this field for many years before (Figure 3).

Even if the interest regarding heredity is long lasting, just before Hippocrates (c. 460 to c. 375  $_{BC}$ ) and Aristotle (384-322  $_{BC}$ ), scientific evidence for patterns of genetic inheritance was due to Gregor Mendel (1822-1884). Mendel published his theory in 1865, but this work was ignored by the scientific community for many years.<sup>5</sup>

Gradenigo cites in particular William Bateson (1861-1926)<sup>6</sup> (who was the first person to use the term genetics to describe the study of heredity) and James Arthur Thomson (1861-1933).<sup>7</sup>

After discussing the normal and pathological heredity in man, he reports on the distinction between "family diseases and weakness of constitution" and on the "influence of consanguinity of parents." In chapter II, after discussing the possible influence of "oto-sclerosis and chronic catarrhal middle otitis," Gradenigo presents the possible symptomatology of familial deafness. In his study, "oto-sclerosis has been noted 96 times out of 132 genealocal trees i.e. 72 per cent; primitive neuro-labynthine injury, 10 per cent; weakness of constitution of the ear, 19 times-14 per cent, and family sordomutitas, 4 times-3 per cent." In Table II, he reports the difficulties of studying genetic disorders in humans (impossibility of making experiments; human life is relatively long; races are numerous, so there is much heterozygosity; families are small; children born in adultery are numerous...).



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Figure 1. Giuseppe Gradenigo, title page of "La Surdité familiale." *Monographies Oto-rhino-laryngologiques internationales No 3*. Legrand, Paris. 1921.





In a later paper (1923), Gradenigo<sup>2,8</sup> reported on different audiometric configurations in dominant genetic disorders, observing that the mid-frequency hearing loss is almost associated with dominant genetic disorders and very rarely with other etiologies.

Only 30 years later, Fisch<sup>o</sup> demonstrated a correspondence between audiometric patterns and the etiology of perceptive deafness. He wrote that "a flat audiogram suggests rubella, a saucer-shaped audiogram kernicterus, a gently sloping audiogram with the high tones affected more than the low is often seen in dominant deafness, and a sharply sloping audiogram with residual island of hearing in the low tones suggests autosomal recessive deafness."

In 1970, Fraser<sup>10</sup> reported on a large sample of 3534 individuals "who have been profoundly deaf from childhood," describing many syndromic cases and paying particular attention to the causes based "on family history." In 1971, Nance<sup>11</sup> published "Genetic Counselling for the Hearing Impaired"; in addition, the same year Bruce W. Konigsmark titled his contribution to "The second Conference on The Clinical Delineation of Birth Defects," Syndromal Approaches to the Nosology of Hereditary Deafness. He wrote: "There are about 70 types of hereditary deafness in man. The differential diagnosis of these familial deafness syndromes is aided by using the following five characteristics of the syndrome: 1) the mode of genetic transmission, 2) the characteristics of the deafness, 3) the age of onset, 4) the sonic frequencies involved and 5) the associated abnormalities." Bruce Konigsmark's fundamental book (completed by Robert Gorlin after Bruce's premature death in 1973) *Genetic and Metabolic Deafness* was published in 1976.<sup>12</sup> This book, and subsequent editions of it,<sup>13,14</sup> as well as McKusick's *Mendelian Inheritance in Man* and its online version<sup>15-17</sup> have been important reference sources for clinicians for decades.

Victor Almond McKusick (1921-2008) widely known as the "father of medical genetics" and editor of *Mendelian Inheritance in Man* was born just the same year of Gradenigo's monography. McKusick published more than 800 scientific papers and books, and he gave a great contribution also to the study of the genetics of deafness, especially in collaboration with Bruce W. Konigsmark.

#### Giuseppe Gradenigo and His Family Biographical Aspects

Giuseppe Gradenigo was one of the most important figures of otolaryngology between the end of the 19th century and the



Figure 3. a,b. Genealogical trees from "La Surdité familiale."



Figure 4. Pietro Gradenigo. Source: Katalog der Deutschen Nationalbibliothek.

beginning of the 20th century. His scientific work also had an international impact. Born in Venice in 1859, he was a member of the noble Venetian family of the Gradenigo. This had illustrious exponents in the history of the Serenissima Republic of Venice. The Gradenigo family can boast other prominent figures in its history: many were bishops, archbishops, patriarchs, politicians, soldiers, and gave Venice 3 doges: Pietro Gradenigo (1251-1311), Bartolomeo Gradenigo (1260-1342), and Giovanni Gradenigo (c. 1279-1356).

Giuseppe's grandfather, Vettor, was a politician and diplomat. He was a member of the Council of Ten, an ancient and prestigious *governing instrument* of the Republic of Venice and also ambassador to France and later Secretary of the Venetian Legation in Vienna. He witnessed the capitulation of the Republic of Venice in 1797 and the succession of French occupation and Austrian domination of Venice.

It was a tradition of Venetian families not to participate in the academic activity of Padova University, but the rapid political upheavals of those years led first Pietro Gradenigo and then his son Giuseppe to take a different path.

Pietro Gradenigo (Venice 1831 to Padua 1904) (Figure 4) had participated in the uprisings of 1948-1949 against Austria at a



Figure 5. Giuseppe Gradenigo by courtesy of prof. Umberto Barillari.

very young age and later graduated in medicine and surgery in Padua in 1855. Here, he also perfected himself in ophthalmology and obstetrics. He began his clinical activity in Venice, also becoming head physician in 1863. Meanwhile, he also became an assistant in the ophthalmology university department in Padua, and in 1873, he took over the reins of the same taking the chair of ophthalmology. He gave great impetus to the development of ophthalmology. He designed numerous instruments for the ophthalmology practice, took care of the modernization of the clinic in Padua, and enhanced the teaching of the subject.

Giuseppe (Venice 1859 to Treviso 1926) (Figure 5), as his father had done, devoted himself to science and the academy. In 1883, he graduated in medicine and surgery in Padua, and soon after, he decided to go to Vienna, the city where he deepened his studies in otology and embryology. Here, in fact, he was a pupil of Adam Politzer (1835-1920), one of the fathers of otology, and of the embryologist Samuel Leopold Schenk (1840-1902). This experience greatly contributed to forging Gradenigo's scientific interests. He obtained, in 1888, the "libera docenza" in otology in Padua, and in 1889, he began to direct the section of otolaryngology of the polyclinic of Turin. In 1890, he began teaching otology and rhino-laryngology, and in 1896, he was appointed extraordinary professor of otolaryngology in Turin; in 1891, he was one of the founders of the "Società Italiana di Laringologia, Otologia e Rinologia" (Figure 6). In parallel, he carried on the assistance practice in the otolaryngology field. As the resources available to him for clinical practice were insufficient, on January 29, 1900,



(Per gentile concessione della nipote Sig.ra Anna Maria Trifiletti).

Figure 6. Congress of the Italian Society of ENT, 1900, also published in "I cento anni della Otorinolaringoiatria Italiana" by Dino Felisati, Giunti ed. 1991; by courtesy of prof. Umberto Barillari.

he opened the Gradenigo private hospital which also offered free outpatient care for the indigent and gave seat to specialization school in otolaryngology for doctors. In 1902, he refused the chair in Rome and had to wait until 1910 for his appointment as full professor in Turin. In 1911, he had the opportunity to take over from Vincenzo Cozzolino (1853-1911) in Naples, an opportunity that he temporarily decided to decline. During the First World War, he assumed the rank of medical lieutenant colonel and devoted himself to the care of soldiers. The important scientific collaboration with the physiologist Amedeo Herlitzka (1872-1949) regarding the medicine of aviators dates back to that period.

In 1917, he took over the chair of otolaryngology at the University of Naples, succeeding Ferdinando Massei (1847-1917). During his brilliant scientific career, he was interested, among other things, in embryology of the auditory organ, bacteriology, audiology, and also in the history of medicine. On March 15, 1926, he passed away in the family house in Lancenigo, near Treviso: one of the most illustrious otolaryngologists in medicine of the early 20th century disappeared.<sup>18-20</sup>

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