Special Report

The Two Anomalies of Wilhelm Ebstein

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The year 2016 was the 180th anniversary of Wilhelm Ebstein's birth and the 150th anniversary of his description of the congenital tricuspid valve malformation that came to be known as Ebstein anomaly. We present a brief history of the life and work of Dr. Wilhelm Ebstein. Despite his distinguished career, he is seemingly forgotten in his own birthplace. We include a review of the relevant historical medical literature. ***(Tex Heart Inst J 2017;44(3):198-201)***

The Lower Silesian city of Jawor (pronounced Yahvor) was mentioned historically as early as the 11th century. Present-day Jawor (population, approx. 25,000) is about 40 miles west of Wroclaw, Poland. Wilhelm Ebstein (Fig. 1) was born in Jawor to a German-Jewish family on 27 November 1836. His father, Louis Ebstein, was a tradesman, and his mother was Amalie Ebstein. At age 28 years, he discovered and described the congenital tricuspid valve (TV) malformation known today as Ebstein anomaly, and thereafter he had a long, distinguished career. Strangely, we found no mention of this prominent medical researcher on the official Jawor website, nor has any street or landmark been named for him, as was done for other notable Silesian doctors and scientists. We present a brief history of Dr. Ebstein's life and work, review the historical medical literature on the Ebstein anomaly, and speculate on the oddity—arguably an anomaly, in view of the recognition of others—of why he is seemingly forgotten in his own birthplace.

Early Years

At age 19, Wilhelm began his medical studies at the University of Breslau, in the capital city of Prussia. After a few months, he transferred to the more prestigious medical school at the University of Berlin. There, he met the esteemed pathologist Rudolf Virchow and learned from him the importance of writing detailed, scholarly medical descriptions. Ebstein graduated in 1859 and, in 1861, began working as an assistant physician at the Allerheiligen (All Saints') Hospital in Breslau (Fig. 2).[1,2]

First Description of the Ebstein Anomaly

On 28 June 1864, Dr. Ebstein's hospital admitted Joseph Prescher, a 19-year-old laborer whose childhood dyspnea and palpitations had worsened with age. The patient's attending physician, Hermann Kornfeld, described the patient's cachexia; severe cyanosis; jugular veins that throbbed in synchrony with the heart's rhythm; a systolic cardiac murmur; and an enlarged cardiac silhouette, detected during cardiac percussion. The clinical picture indicated a congenital heart defect. Eight days later, the patient died; the next day, the 28-year-old Dr. Ebstein performed the autopsy. In 1866, he published his case report, "A very rare case of tricuspid regurgitation caused by a congenital defect" (Fig. 3).[3,4]

Ebstein's paper included meticulous illustrations by his colleague, Dr. Wyss, and described anatomic and pathologic factors with elegant clarity and precision. Ebstein presented connections between the clinical findings and cardiac pathologic conditions, suggesting embryonic defects as a likely cause of the patient's anomaly. He observed that the TV was extremely malformed: its anterior leaflet was elongated and had numerous fenestrations, and its hypoplastic, thickened posterior and septal leaflets adhered to the right ventricle. The atrialized part of the right ventricular wall was thin, the right atrium was enlarged, and a patent foramen ovale was present. Chief among the anomaly's 3 major components, Ebstein wrote, was the severe TV malformation. He concluded that a second component, an absent Thebesian valve, played no role

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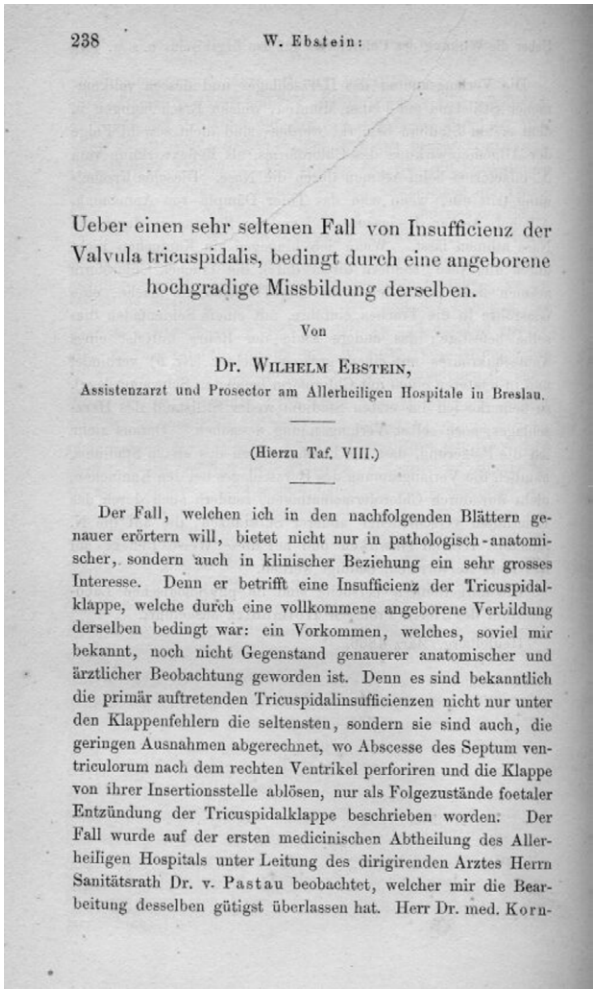
Fig. 1 *Photograph shows Wilhelm Ebstein, ca. 1885.*

(© Städtisches Museum; Göttingen, Germany)

Fig. 2 *Photograph shows the hospital in Wroclaw, Poland (then Allerheiligen Hospital in Breslau, Prussia), where Wilhelm Ebstein performed the autopsy of the patient who had a tricuspid valve anomaly.*

(Photograph from Magdalena Mazurak's collection.)

in the defect's pathophysiology. The third component, the patent foramen ovale, enabled right-to-left shunting with consequent cyanosis—an aspect overlooked by Ebstein and arguably the only weak point in his analysis. In his paper, Ebstein included a thorough review of the available literature on TV defects and concluded that his case was unique.[3,4]



238 W. Ebstein:

Ueber einen sehr seltenen Fall von Insufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben.

Von

Dr. Wilhelm Ebstein,

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(Hierzu Taf. VIII.)

Der Fall, welchen ich in den nachfolgenden Blättern genauer erörtern will, bietet nicht nur in pathologisch-anatomischer, sondern auch in klinischer Beziehung ein sehr grosses Interesse. Denn er betrifft eine Insufficienz der Tricuspidalklappe, welche durch eine vollkommene angeborene Verbildung derselben bedingt war: ein Vorkommen, welches, soviel mir bekannt, noch nicht Gegenstand genauerer anatomischer und ärztlicher Beobachtung geworden ist. Denn es sind bekanntlich die primär auftretenden Tricuspidalinsufficienzen nicht nur unter den Klappenfehlern die seltensten, sondern sie sind auch, die geringen Ausnahmen abgerechnet, wo Abscesse des Septum ventriculorum nach dem rechten Ventrikel perforiren und die Klappe von ihrer Insertionsstelle ablösen, nur als Folgezustände foetaler Entzündung der Tricuspidalklappe beschrieben worden. Der Fall wurde auf der ersten medicinischen Abtheilung des Allerheiligen Hospitals unter Leitung des dirigirenden Arztes Herrn Sanitätsrath Dr. v. Pastau beobachtet, welcher mir die Bearbeitung desselben gütigst überlassen hat. Herr Dr. med. Korn-

Fig. 3 *The title page from Ebstein's original paper.*

(Courtesy of BIU Santé.) Available from: http://www.biusante.parisdescartes.fr/histmed/medica/page?epo0270&p=1.

Subsequent History

In 1869, Ebstein defended his habilitation thesis, "Die Recidive des Typhus," and earned the right to give university lectures; however, his 9-year career at Allerheiligen Hospital was interrupted by the Franco-Prussian War. After returning from the front, he served as a medical officer in Breslau's shelters for the poor. In 1874, he accepted a professorship of medicine at the University of Göttingen. He died of a stroke at age 75 in Göttingen on 22 October 1912.[2] He was survived by his wife, Elfriede Ebstein. Her brother, Arthur Nicolaier (also Silesian), had identified the causative bacillus of tetanus and injected it into animals.[5,6]

Ebstein's daughter Amalie, named after her paternal grandmother, was born in 1876 in Göttingen. She married the mathematician Otto Blumenthal, with whom

she had 2 children. The couple were victims of the Nazi Holocaust.[7] Wilhelm's son Erich, born in 1880, was a doctor, a collector, a bibliophile, and the author of more than 500 publications on the histories of medicine and literature. He and his wife had 2 children. Like his father, Erich died of a stroke (in 1931, at age 51).

After World War II, the Prussian/German city Breslau became the Polish city Wroclaw, and Allerheiligen Hospital was renamed as Jozef Babinski Hospital. This hospital—where the 28-year-old Dr. Wilhelm Ebstein had performed his landmark autopsy of a patient with a TV anomaly—closed its doors in 2007.

The Seeming Rarity of Ebstein Anomaly

In 1867, Hermann Lebert, then a professor of medicine in Breslau, was probably the first to catalogue Ebstein's seminal publication.[8] Until the 1950s, a mere handful of cases of patients with Ebstein anomaly had been reported. Theodor Marxsen used one case as the subject of his doctoral dissertation, "A rare case of anomaly of the tricuspid valve," in 1886.[9] The term "Ebstein's disease" did not appear in the medical literature until 1927.[10] A decade later, Yater and Shapiro[11] confirmed the diagnosis of "Ebstein's disease" at autopsy in a patient whose congenital cardiac defects they had evaluated with the use of radiography and electrocardiography; they concluded that the malformation "may be suspected with other possibilities but cannot be positively diagnosed during life." At last, in 1951, Soloff and colleagues[12] reported the case of a living patient in whom cardiac catheterization and angiocardiography revealed Ebstein anomaly. In a detailed clinical study of 23 patients (published in 1959), Schiebler and associates[13] mentioned 140 cases of Ebstein anomaly in the world medical literature. Christiaan Barnard surgically replaced an affected TV with a bioprosthesis in 1962.[14]

More than 150 years after Ebstein anomaly was first reported, remarkable progress has been made in its repair. Since the introduction of various corrective surgical techniques, operative mortality rates have decreased.[15-21] Nevertheless, the diagnosis and treatment of Ebstein anomaly can still be challenging because of operator inexperience and the relatively few patients who present with the malformation.[21]

The "Anomaly" of Wilhelm Ebstein's Legacy

During his lifetime, Wilhelm Ebstein was a respected medical practitioner and a prolific man of science. Of note, only 12 of his 237 publications dealt with heart disease.[1] His fame was chiefly as a specialist in metabolic diseases, especially renal diseases, diabetes mellitus, and obesity—yet even these topics constituted only a fraction of his broad research, clinical, literary, and artistic interests. Ironically, the publication that eventually immortalized his name[3] came early in his career and was substantially recognized only after his death.

Lower Silesia's other 2 eponymous cardiac physicians, Jan Purkynie (Johannes Evangelista Purkinje) and Adam Christian Thebesius (Thebesian valve), are honored throughout the region. There is a Purkynie Street in Wroclaw, and a marble bust of Purkynie graces the Physiology Department of Wroclaw Medical University. Thebesius Street is found in Jelenia Góra, a Silesian town 28 miles from Jawor and 62 miles from Wroclaw, where Thebesius had established his medical practice. However, no traces of Wilhelm Ebstein can be found in this geographical area.[22] We speculate that Dr. Ebstein's broad range of research interests beyond cardiology and the rarity of the Ebstein anomaly are factors in why he has not specifically been honored.

Despite the absence of local recognition, to this day Wilhelm Ebstein deserves credit for his detailed description of the congenital malformation that bears his name.

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