Profiles in Cardiology

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Etienne-Louis Fallot: Is It His Tetralogy?

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The congenital cardiac disturbance characterized by pulmonary stenosis, interventricular septal defect, hypertrophy of the right ventricle, and dextroposition of the aorta carries the eponym "tetralogy of Fallot." The classical paper by Etienne-Louis Fallot (Fig. 1), describing in detail the components of this common cause of "maladie bleue," appeared in 1888 in an obscure French journal.^{1,2} The issue in which it appeared devoted 98 pages to a series of six articles on the pathologic anatomy of congenital heart disease with cyanosis. Fallot's article was titled *Contribution à l'anatomie pathologique de la maladie bleue (cyanose cardiaque)*.³ Four years later, a paper was presented to the Royal Academy of Medicine in Ireland by Ambrose Birmingham, professor of anatomy in the Cecilia Street School of Medicine. It came to be known as the pentalogy of Fallot since, in addition to the components described by Fallot, Birmingham added a fifth, namely, an interatrial communication.⁴ In retrospect, this seems superfluous, since Fallot did also say that "At times, there is an additional entirely accessory defect; namely, patency of the foramen ovale."

The malformations described by Fallot as a cause of blue babies were, however, originally described by Niels Stensen, the discoverer of the parotid duct, as far back as 1671.⁵ Indeed, in the two centuries that followed, many notables were to add

their descriptions to the malady; these included Eduard Sandifort (1777), William Hunter (1784), Farré (1814), Gintrac (1824), Peacock (1866), Debely (1878), and Roger (1879).

Steno (the latinized surname of this Danish anatomist and theologian) came upon this congenital abnormality in a fetus, describing in accurate detail the multiple cardiac defects as seen at autopsy. A little more than a century later, the Dutch physician, teacher, and linguist, Eduard Sandifort, described in his book *Observationes Anatomico-Pathologicae* what he thought was both a rare and unusual malady of the heart. The patient had cyanosis as an infant and complained of extreme fatigue, headaches relieved with venesection, fainting spells, palpitations, and pedal edema. When the patient died at the age of 12½ years, Sandifort described at postmortem the very same four salient defects that Stensen had originally reported in the fetus in 1671.⁶ Hunter's description of the malady seen at autopsy was rather vivid and so typical of the British in their ability to transmit imagery via the written word. Who can fault his description where "the passage from the right ventricle into the pulmonary artery, which should have admitted a finger, was not so wide as a goose quill; and there was a hole in the partition of the two ventricles, large enough to pass the thumb from one to the other. . . The greatest part of the blood in the right ventricle was driven with that of the left ventricle into the aorta, or great artery, and so lost all the advantage which it ought to have had from breathing."⁷

Fallot's comprehensive account of the malformation, his demonstration that the disease could be readily diagnosed prior to death, and that, among the congenital abnormalities of the heart, these multiple cardiac defects were common rather than rare, were probably the three most important elements responsible for the designation of his name as the marker for the entire syndrome. The eponym persists to this day, despite the fact that Fallot quite candidly acknowledged the contributions of his predecessors. In particular, Fallot referred to the thesis of Debely that had been published just a decade before.⁸

Fallot's paper itself is rather terse and to the point; however, he did manage to include a review of the literature in the decade that followed the publication of Debely's account. Of a

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FIG. 1 Etienne-Louis Fallot, 1850–1911. Courtesy of New York Academy of Medicine.

total of 55 cases reported during this decade as examples of the "maladie bleue," Fallot pointed out that only 39 of these had the four components characteristic of his own cases. His delineation of the pathogenesis of the multiple defects was similar to that of William Hunter. Both were of the opinion that the multiple defects were a consequence of pulmonary stenosis. Fallot and Hunter were in error, of course; neither one recognized the embryologic nature of the defects.

Fallot had expressly forbidden the publication of any eulogy after his death, and this may be the reason for the scarcity of biographical data. Serendipitously, I was able to find a short biography in a text written by Mattei.⁹ All that can be said is that he was born on September 20, or 29, 1850, in Cette, France. He was probably a very good student, as implied by the prize he received for high scholastic ability while attending the Lycée in Marseilles. His medical degree was obtained from the École de Médecine in the same town. He stayed in Marseilles throughout his life, serving first as substitute professor of medicine at the University of Marseilles, later taking charge of the course in pathologic anatomy, and finally serving as professor of hygiene and legal medicine until his death.

Fallot was renowned as an excellent clinician who was painfully accurate in the physical examination of the patient and who was apparently well respected for his diagnostic skills. In addition to his description of the tetralogy, Fallot also reported a case of congenital pectoral dysplasia, a case of hysterical hemiplegia, the incidence of encephalitis among Corsicans, and his observations during a local cholera epidemic. Strangely enough, he did not publish any medical articles during the last 10 years of his life. The only publication during this

time was his description of a Neolitt cave in Reillane. Mattei reported that he died in May 1911 "after a period of purifying loneliness."⁹ Other sources place the time of his death on April 30, 1911.

Probably the most interesting individual to have had the tetralogy of Fallot was the patient described by White and Sprague in 1929.⁸ That individual was Henry F. Gilbert, who, despite being born with this malady, managed to overcome his handicap and carry on a very active life as a talented composer of music. Gilbert managed to live to the age of 60 just shy a few months. The oldest patient in Fallot's original series was 36 years of age at the time of his death. Unable to exert himself even in the slightest without marked discomfort, plagued by cyanosis, clubbing, dyspnea, weakness, and fainting episodes, Gilbert even managed to survive a successful European tour, only to die from apoplexy a year later. He left a legacy of native American musical compositions, which included "Comedy Overtures on Negro Themes," "Negro Rhapsody," and "Indian Sketches."⁸

Merely discovering or being the first to describe a disease should not mandate that the disease be identified eponymously with the discoverer's name. In actuality, there are many instances and cogent reasons when such has not been the case. Duke aptly points this out, when he quotes Hans Selye, who wrote in his book *From Dreams to Discovery* that "it is not to see something first, but to establish solid connections between the previously known and the hitherto unknown that constitutes the essence of scientific discovery."¹⁰

Fallot's role in this regard can be easily justified and thus allows us to continue honoring his memory by retaining "tetralogy of Fallot" as the eponymous designation for *morbus caeruleus*.

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