Frederic Chopin and Michael Jackson: What could they have in common?

Frédéric Chopin et Michael Jackson : qu’ont-ils en commun ?

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If you open a biography of Frederic Chopin (1810–1849), whose 200th birthday anniversary is celebrated this year, you will almost invariably read that he was affected, during most of his life, by tuberculosis and that he died of it [1–3]. Is it that simple?

A long-lasting illness

Let us look first at the clinical history. Chopin is a delicate child, who fatigues easily and avoids physical activity. He suffers from the age of at least 16 years (perhaps earlier) of repeated episodes of productive cough, asthenia, fever and haemoptysis. The first of these recurrent episodes occurs in 1826 (at age 16): according to George Sand and Franz Liszt [4], at that time, he has an illness lasting 6 months, with respiratory complaints, and severe headaches. He suffers similar trouble while in Vienna in 1830, and then again in 1835 when he returns to Paris after a trip to Carlsbad (where he visited his parents) and Dresden. The episode lasts about 2 months and all symptoms disappear completely in December. Thereafter, a similar attack takes place each winter. In February 1837, a diagnosis of ‘‘flu’’ is made, with fever, followed by a first haematemesis. A second haematemesis will take place in 1849, a few months before his death. In October 1838, during his stay in the Island of Majorca with George Sand, he has a severe attack of cough, fever and haemoptysis and decides to return to France to benefit from the sunny weather of Provence. In Marseilles, Chopin consults Dr. Cauvière, a renowned physician of the time. Dr. Cauvière excludes the diagnosis of tuberculosis and speaks of ‘‘irritated’’ lungs. Shortly thereafter, George Sand writes, ‘‘Thanks God, he had only just felt the dry air of Provence that he resuscitated before one’s very eyes and he is perfectly well, growing relatively stout, almost without cough’’. Such a rapid ‘‘cure’’ is highly unlikely for tuberculosis. Several similar episodes occur regularly, with progressive exertional dyspnoea and chest pain. In the summer of 1842, he consults George Sand’s family physician, Dr. Papet, who, again, excludes tuberculosis and speaks of ‘‘bronchial mucosities’’. His dyspnoea worsens. Laennec himself is consulted and makes no firm diagnosis. Recurrent episodes of cough and fever occur, with, often, prompt recovery, until June 1849: at the occasion of a severe attack, his friends call on Professor Jean Cruveilhier (who took care, among others, of Talleyrand, Chateaubriand and Alfred de Vigny). He makes a diagnosis of terminal tuberculosis and declares that there is no hope of cure. Chopin dies at two
o’clock in the morning of October 17, 1849. He had asked that an autopsy be performed to make sure that he would not be buried alive, a great obsession, and also to have his heart brought back in Warsaw. The autopsy is carried out by Cruveilhier (who is not only a famous clinician, but also professor of pathology). The certified cause of death is tuberculosis of lungs and larynx, but Cruveilhier (whose report has been lost) admits, according to Chopin’s sister Ludwicka and several other witnesses, that the autopsy actually did not reveal the cause of death and that Chopin had a disease that was unknown to him [5].

A striking family history

Let us look now at the family history. Frederic (originally Fryderyk) has three sisters: Ludwicka, born in 1807, Isabela, born in 1811, and Emilia, born in 1812. His father, Nicolas, born in Lorraine but who emigrated to Poland and took the first name of Mikolaj, suffered throughout his life of recurrent respiratory infection. So did his elder sister, Ludwicka, who died of it at the age of 47 years. Emilia, the youngest of the family, suffered repeatedly from cough, wheezing and dyspnoea. She died at the age of 14 years, from massive upper gastro-intestinal bleeding. The family tree is depicted in Fig. 1.

A very likely hereditary disease

If we consider the family history and the long duration of Chopin illness (at least 25 years), a diagnosis of tuberculosis appears unlikely. Long-lasting tuberculosis is usually associated with finger clubbing, and we know from a cast of Chopin’s hand taken after his death that he lacked this sign [4]. This diagnosis would explain the repeated haemoptysis, but not the two episodes of haematemesis. We must also take into account the opinion of Cruveilhier, a recognized authority in tuberculosis pathology, who declared after the autopsy that Chopin did not have that disease, but a condition he had not previously encountered.

The family history strongly suggests a hereditary condition. Mucoviscidosis (cystic fibrosis) has been considered by several authors [6] and is still proposed, among others, by Professor Wojciech Cichy, a polish specialist of the disease [7]. It is a common autosomal recessive disease, associated with recurrent pneumonia. There is pancreatic insufficiency causing chronic diarrhoea with steatorrhea. Cirrhosis of the liver is possible, but rare. Death from infection and right-sided cardiac failure usually occurred in the first decade of life in the preantibiotic age. Mucoviscidosis could explain most of Chopin’s symptoms and signs. It appears however unlikely in his case, since survival until the age of 39 years would have been extraordinary at that time.

The hereditary disease that best explains Chopin’s medical history, in my opinion, is α1-antitrypsin (α1-AT) deficiency. This hypothesis was drawn to my attention by a remarkable analysis by Adrian Reuben a few years ago [8]. α1-AT deficiency is a genetic metabolic disease caused by the lack of the protease inhibitor α1-AT [9]. The α1-AT gene has been mapped to the long arm of chromosome 14 [10]. It causes chronic lung disease with emphysema, bronchiectasis, chronic bronchitis and recurrent bronchopulmonary infections. Chopin most probably had emphysema, as depicted in a caricature drawn by his friend, the singer Pauline Viardot, in 1844, showing a barrel-chested skinny man with extraordinary thin limbs (Fig. 2). Haemoptysis, occasionally massive, are possible [11,12]. The bronchopulmonary manifestations may be associated with liver cirrhosis and portal hypertension, failure to thrive and pancreatic insufficiency. The common disease is associated with the ZZ phenotype, the incidence of which is around 1:1000 in Western Europe, and the number of heterozygous carriers is about 4% of the population. Both homozygotes and heterozygotes are at increased risk of developing respiratory problems.

This diagnosis would explain:

- Emilia’s death at the age of 14 from massive upper gastrointestinal bleeding secondary to portal hypertension caused by cirrhosis;
have been blamed to spread the

Some authors have also discussed the possible infertility complaints. For example, he had frequently pain in the ankles, feet and even hands. This could be due to pulmonary hypertrophic osteoarthropathy, which manifests by painful swelling of distal joints and soft tissue swelling. Some authors have also discussed the possible infertility of Chopin: despite several sexual encounters and a long-standing relationship with George Sand, Chopin did not have any child. Finally, poor exercise tolerance and failure to gain weight are also possible consequences of his disease. It is also very likely that Nicolas, Frederic’s father, and Ludwicca, his elder sister, also had $\alpha_1$-AT deficiency.

Some long-term complications of a chronic lung disease such as $\alpha_1$-AT deficiency could also explain other Chopin’s complaints. For example, he had frequently pain in the ankles, feet and even hands. This could be due to pulmonary hypertrophic osteoarthropathy, which manifests by painful swelling of distal joints and soft tissue swelling. Some authors have also discussed the possible infertility of Chopin: despite several sexual encounters and a long-standing relationship with George Sand, Chopin did not have any child. Finally, poor exercise tolerance and failure to gain weight are also possible consequences of his disease. It is fair to acknowledge, however, that none of these possible complications is specific to $\alpha_1$-AT deficiency, or even to chronic respiratory disease.

A four-thousand-year-old mutation

The mutation causing $\alpha_1$-AT deficiency appeared anywhere from 107 to 228 generations ago [13,14], and the Vikings have been blamed to spread the $\alpha_1$-AT Z gene throughout the world [15]. Study of the Z gene frequency in European countries show a gradient of distribution from northwest to southeast, with a likely site of origin of the mutant Z gene in southern Scandinavia. The distribution of type S is quite different; the gene frequency is highest in the Iberian peninsula and the mutation is likely to have arisen in that region [13]. The Vikings, therefore, do not deserve alone credit or blame for disseminating the mutations around the world. To return to Chopin, given the gene frequency in France (0.0114) and in Poland (0.0041) and the total population of these two countries, it can be calculated that the population at risk for $\alpha_1$-AT deficiency in Poland and France together is 2-fold greater than in Scandinavia. The occurrence of the disease in a francopolish individual is, therefore, genetically possible.

June 25, 2009: death of a King

One hundred and fifty years after Chopin’s death, another music star dies, with, allegedly, the same hereditary disease. For the hundreds of thousands of fans of Michael Jackson, a dramatic event takes place on June 25, 2009. On that day, the ’’King of Pop’’ dies, according to the official report of autopsy, of ’’acute propofol intoxication’’, an anaesthetic agent used by his personal physician to facilitate sleep. This practitioner who injected propofol will have to justify himself before the justice of his country.

But, apart from chronic anxiety and sleep problems, vitiligo and other complaints, the pop star might also have suffered from $\alpha_1$-AT deficiency. In December 2008, one can read in the press that Michael Jackson has $\alpha_1$-AT deficiency and that his life expectancy does not exceed a few months [16]. In a biography published soon after his death, the journalist and investigator Ian Halperin (who is at the origin of the rumour diffused by the press in December) alleges again that Jackson has had this disease for several years and was actually on a waiting list for lung transplantation [17]. The author claims that a physician who took care of the respiratory problems of Michael Jackson, and who accepted to answer his questions, confirmed the diagnosis. This allegation raised considerable emotion throughout the world. In fact, there is little evidence to support this hypothesis. Michael had eight brothers and sisters and none is known to have a chronic respiratory disease. The singer displayed considerable energy on stage during his professional career, starting at the age of 11 years, an observation hardly compatible with a long-standing chronic disease. And finally, the autopsy report does not mention any major abnormality of the bronchopulmonary system or of the liver [18], the two main targets of $\alpha_1$-AT deficiency.

In conclusion, there is convincing evidence, although not conclusive at this time, that one of the greatest composers of the 19th century, Frederic Chopin, suffered from $\alpha_1$-AT deficiency. Many musicologists even argue that this long-lasting and devastating disease, which made Chopin conscious that he would probably not live a long life, probably accounts for the dramatic character of many of his works. In contrast, the allegation that Michael Jackson, one of the most popular artists of the 20th century, also had $\alpha_1$-AT deficiency should at this time be taken with great caution. Recently, a DNA analysis was performed on the mummy of Tutankhamun, the Egyptian pharaoh [19]. Since he died circa...
1324 BC, some 34 centuries ago, why not consider performing such an analysis on Chopin’s heart and on Jackson’s tissues taken at autopsy, so that future historians of music have a definite answer to these medical mysteries.

Conflicts of interest

The author has not declared any conflicts of interest.

References
