

Chapter

New Insights into Beethoven's Deafness

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Abstract

There have been many theories proposed to explain the deafness of Ludwig van Beethoven, because his history is complex. Evidence of otosclerosis is lacking, because close gross examination of Beethoven's middle ear at autopsy did not find any otosclerotic foci. His slowly progressive hearing loss over a period of years differs from the reported cases of autoimmune hearing loss, which is rapidly progressive over a period of months. The absence of mercury in Beethoven's hair and bone samples leads us to conclude that his deafness was not due to syphilis, because in that era, syphilis was treated with mercury. Microscopic examination of bone samples and examination of the middle ear have found no evidence of Paget's Disease. High levels of lead found deep in the bone suggest repeated exposure over a long period of time. The finding of shrunken cochlear nerves at his autopsy is consistent with axonal degeneration due to heavy metals such as lead. Chronic low-level exposure, like Beethoven's, causes sensory and autonomic findings rather than the classic wrist drop due to motor neuropathy. Beethoven's physicians thought that he had alcohol dependence. He particularly liked wine to which lead had been added to improve the flavor. A live patient reported in 2021 from Italy with a slowly progressive hearing loss and other symptoms like Beethoven had, was found to have lead poisoning. Therefore, the most likely cause of Beethoven's deafness was his consumption of wine tainted with lead.

Keywords: lead, alcohol, otosclerosis autoimmune, syphilis, Paget's disease

1. Introduction

The reader may wonder why this chapter is part of a book about the pharynx. Although Beethoven had no problem with his pharynx, he did have problems with his ears and his gastrointestinal tract at both ends of the pharynx. The reason this is relevant is that new insights have come to light from information not available prior to this century. This has allowed a better picture of a problem that has been poorly understood for many years.

2. The eustachian tube

The pharynx, the incredible rendezvous site of gas, liquid, and solids, is connected in its superior aspect, or nasopharynx, to the ear *via* the eustachian tube. In its lower

extension, it connects to the gastrointestinal tract posteriorly *via* the esophagus and anteriorly to the lungs *via* the larynx. The eustachian tube is a very interesting structure that is about 36 millimeters long. It is divided into two segments. The bony, nearest to the ear, comprises $\frac{1}{3}$, and the cartilaginous, nearest to the nasopharynx, the other $\frac{2}{3}$ of its length. The cartilaginous portion is part of the pharynx [1]. A popular misunderstanding is that the eustachian tube drains fluid from the ear, because it projects downward from the ear to the nasopharynx. However, its function is to ventilate the ear. The eustachian tube is normally closed. It opens with swallowing in response to the pull of the tensor and elevator veli palatini muscles. They are innervated by the pharyngeal plexus of the vagus nerve. When swallowing occurs, air from the nasopharynx goes up into the ear to maintain a balance of pressure between the closed cavity of the ear and the outside air. If the eustachian tube does not open, a negative pressure develops in the middle ear cavity that draws fluid from the mucus membrane into the middle ear cavity, resulting in a hearing loss.

Many things can impede the eustachian tube from opening. These include viral upper respiratory infections, such as the common cold, nasal allergies, and sinus infections. These cause swelling of the eustachian tube opening, preventing it from opening properly. These can be treated by decongestants, antihistamines, nasal steroids, and antibiotics. Adenoid hypertrophy or enlargement can block air getting into the ear and can be treated by surgery or adenoidectomy. Tumors, such as nasopharyngeal carcinoma that is common in people of oriental origin, can be treated by surgery, radiation, and chemotherapy.

In cases of chronic eustachian tube dysfunction that has not responded to conservative measures, fluid can be drained from the middle ear by means of a hole or myringotomy made in the tympanic membrane or ear drum. If the eustachian tube remains occluded, however, fluid will re-accumulate, because the tympanic membrane will heal in a few days. This will necessitate placement of a small tube through the tympanic membrane or tympanostomy, to allow air to enter the middle ear space so the fluid can reabsorb. In recent years, a new technique called eustachian tube dilation has been performed from either the middle ear [2] or the nasopharynx [3], to restore normal eustachian tube function.

3. A diagnostic dilemma

Beethoven had no problem with his eustachian tube, but he did have hearing loss, chronic gastrointestinal problems, intermittent pain in his extremities, depression, and alcohol dependence. There are many opinions regarding the etiology of Ludwig van Beethoven's hearing loss. This is due to lack of confirmatory evidence by present techniques such as hearing tests, radiological and blood studies. His overall medical history was also complex with many symptoms that lead to conflicting diagnosis. Two papers demonstrate this sharp difference of opinion that can be formed even about the same diagnosis.

One paper examined three fragments of what was felt to be "nearly with certainty" Beethoven's skull bone that did not show signs of Paget's disease and emphasized that "it must therefore be concluded that Beethoven's deafness was not caused by Paget's Osteitis Deformans" [4]. The other paper stated, thus Paget's disease, complicated by hyperparathyroidism, gout, and attempts to find relief of symptoms through the use of alcohol, quinine, and possibly salicylates, can explain virtually all of Beethoven's medical problems [5]. The title of the latter included: "A Pathologist Sounds a final Note."

Obviously, there is no final note sounded, given the opposite opinions formed by these authors. A major problem is the approach that physicians have used. In their search for a diagnosis, they try to fit Beethoven's multiple symptoms with a cause of hearing loss that they know little about and do not have personal experience with. This contrasts with the approach of someone familiar with causes of hearing loss making a diagnosis and then seeing if Beethoven's other problems can be explained by this diagnosis. Relevant literature regarding his hearing loss was critically examined and felt not to explain his loss. It was then discovered that by reexamining a previously considered diagnosis with current information, a diagnosis could be made that explained not only his hearing loss, but his other symptoms as well. This satisfied Ockham's razor, which is a principle of parsimony that postulates that among competing theories, the hypothesis with the fewest assumptions is the one best selected [6]. Although this principal does not apply in all cases where there are multiple symptoms, it does work here.

3.1 Approach to the dilemma

An extensive review of the musical and medical history of Beethoven's life was done as part of a master's degree in music history and literature at the University of Utah School of Music and is published in *The Laryngoscope* [7]. Literature subsequent to the publishing of that paper has also been critically examined to complete this chapter. All diagnoses of Beethoven's deafness previously considered are not reviewed in this chapter due to length constraints, but attention is given to those most compelling.

3.2 Important historical evidences

Although confirmatory evidence is not available, the history of his hearing loss is, available and reveals much regarding it. At 27 years of age, Beethoven first noticed a hearing problem with ringing in his ear and became aware that he missed words and phrases. He confessed this to a childhood friend and physician in a letter. He said, "for the last several years my hearing has grown weaker and weaker-I cannot hear the high notes of instruments or voices-I can hear sounds, but I cannot make out the words" [8]. His loss slowly and progressively got worse so that he stopped playing the piano in 1815. In 1822, he had to stop conducting. What does the history of his hearing loss tell us? First, it was slowly progressive over a period of years. Second, it involved the high frequencies, at least initially, and third, he had reduced discrimination. Reduced discrimination is often seen in individuals that have hearing loss in the high frequencies. This causes them to miss the consonant sounds of words, because consonant sounds involve high frequencies. They hear words, but do not understand them, because they hear the vowels that are largely composed of lower tones, but not the high-tone consonants that separate the vowels.

3.3 Otosclerosis

One of the popular theories regarding Beethoven's deafness is that of otosclerosis [9]. This is a disease that causes a slowly progressive loss and often begins in the teens or twenties like that experienced by Beethoven. However, people with otosclerosis usually do not have difficulty understanding words when the sounds are sufficiently loud. In other words, otosclerosis is usually associated with good discrimination. It is also inherited, and although it can skip several generations, it is striking that there is no history of hearing loss in Beethoven's musical family. The most significant feature

against otosclerosis is that a close examination of his temporal bone, including the middle ear, at the time of his death, found no evidence of otosclerotic foci. These are white plaques that would have been easily seen by the naked eye. These would certainly have been present given the many years of his loss before death.

3.4 Autoimmune hearing loss

Autoimmune hearing loss has been described in association with autoimmune bowel disease, and since Beethoven had gastrointestinal problems, his hearing loss might be autoimmune [10, 11]. However, autoimmune hearing loss usually progresses rapidly over weeks or months often with accompanying episodes of vertigo. A review of the literature did not find any cases of autoimmune loss with such a slow progression like Beethoven's. In addition, autoimmune bowel disease is invariably accompanied by bloody diarrhea. Beethoven's loss was slowly progressive and lacked vertigo, and no bloody diarrhea was ever mentioned. At his autopsy, there were no adhesions, strictures, or perforations in the bowel that are seen with autoimmune bowel disease. This diagnosis therefore does not fit.

3.5 Syphilis

Another diagnosis suggested by a prominent otologist is acquired syphilis, because of the shrunken cochlear nerves found at autopsy [12]. However, none of Beethoven's 17 physicians considered the diagnosis of syphilis, and evaluation of Beethoven's hair and bone samples in 2000 and 2005 showed no evidence of mercury [13, 14], which was used to treat syphilis in his era. Congenital syphilis is also not a valid consideration, because even though Beethoven's father was an alcoholic, there is no evidence that he was sexually promiscuous. Beethoven also lacked any of the accompanying features of congenital syphilis such as skin rash, nasal discharge, ulcers of the nose and palate, bony deformities, corneal scarring, or dental abnormalities.

3.6 Lead poisoning

Lead poisoning has been previously considered to be the cause of Beethoven's hearing loss, but has been dismissed because he lacked wrist drop, or weakness of the extensor muscles that elevate the wrist. This is invariably present with classic lead poisoning. This classic lead toxicity, however, usually occurs with exposure to high lead levels over a few years' time. Those with low-level exposure over a longer time have mood disturbances, abnormal liver and kidney function, gastrointestinal disturbances, and pain and tingling in the hands and feet. Elevated levels of lead were found in Beethoven's hair (see **Figure 1**) and deep in the bone, consistent with exposure over a long period of time [13, 14]. This constellation of symptoms was described in a group of 151 patients in Latvia [15] and was similar to those experienced by Beethoven. It is important to note that a black lead line on gingival tissues, seen with classic lead poisoning, nor disturbance in the sense of taste, was reported neither in these patients nor by Beethoven.

3.7 The sense of taste

One of the amazing functions of the pharynx is the sense of taste. Taste buds for sweet, sour, bitter, and salt are found in the oral cavity and the or-pharynx, and even

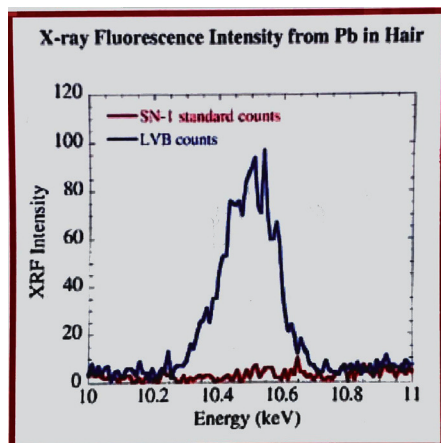


Figure 1.
X-ray fluorescence intensity levels in Beethoven's hair compared with normal levels.

in the hypopharynx. The sensation of taste or gustatory sensation is transmitted by way of both the lingual nerve *via* the seventh cranial nerve, and the pharyngeal, or ninth cranial nerve. The sense of smell is integrally involved with the sense of taste in that part of tasting foods is smelling them. Therefore, a loss of the sense of smell will make food taste bland. This has been very prevalent during the pandemic and can cause temporary as well as persistent loss. Taste disturbance is most commonly a side effect of medicine that often produces a metallic taste in the mouth. It can also be caused by viral illnesses, head injury, oral hygiene problems, tonsillectomy, stroke, and heavy metals such as lead.

3.8 Gastrointestinal problems

A recent article discusses the many medical symptoms Beethoven had during his life [16]. Although lead can account for several symptoms, such as abdominal pain or colic, it is also possible that he had irritable bowel syndrome to explain his gastrointestinal problems, and the pain in his hands and feet could have represented a rheumatological syndrome. Cirrhosis and chronic pancreatitis seen at death were attributable to his alcohol consumption, but lead could also have contributed to the cirrhosis. His kidney damage could have been caused by lead, but also by pyelonephritis.

3.9 Paget's disease

The provocative article written in 2017 mentioned in the introduction [5] deserves close scrutiny as it is well written by Oiseth, a pathologist. He disagrees with the original finding of prior pathologists [4]. There appear to be a number of flaws with his approach, which is largely based on the findings of a thickened skull on gross examination at the autopsy, which is consistent with Paget's disease. However, as he himself wrote, there are other causes of a thickened skull. One of these is hyperparathyroidism which the author suggests he had due to chronic renal disease. A gross examination is the initial examination to draw up a differential diagnosis, and then, the microscopic examination is used to confirm the diagnosis. The prior pathologists did a microscopic exam and found no evidence of Paget's disease. Oiseth

dismisses this with the fact that Paget's disease of the skull is not uniform and may undergo remodeling, so normal bone may be seen in the final phase of the disease. This is something those who did the original exam were well aware of, but perhaps they did get a sample that lacked microscopic evidence of Paget's disease. This seems extremely unlikely, as they examined three large pieces of bone from the parietal and occipital areas of the skull. However, Oiseth fails to point out that a gross exam was also done on the middle ear, which was found to be normal. It would seem likely that microscopic fractures, fixation of the stapes footplate, or fissures of the otic capsule would have been seen as sequelae of the active process, since the hearing loss that is associated with Paget's usually involves the middle ear. It is also likely that an irregular surface of the temporal bone would have been noted from prior active disease.

Oiseth admits that lead poisoning cannot be dismissed entirely, but then makes the same error as other authors that have dismissed lead poisoning, by noting the lack of other neurologic manifestations. Although the article in *The Laryngoscope* was cited by him [4], he apparently read only the title and did not even read the abstract that states that chronic low-level lead exposure like Beethoven had does not result in motor deficits that are usually seen with acute lead poisoning. This also explains why the acoustic nerve, a sensory nerve, was shrunken with lead damage, but the facial nerve, a motor nerve, was left untouched. Beethoven's loss started in his twenties, whereas only 3% of patients with Paget's disease are under 40.

He also mentions that Beethoven had recruitment, which is an abnormal sensitivity to loud sounds, due to impingement of Paget's disease on the cochlear nerve as evidence of a sensorineural loss and not of otosclerosis. By this, he demonstrates his lack of understanding of the term sensorineural, which is an inclusive term comprising a loss from either the nerve, the cochlea, or both. Patients with hearing loss from the cochlear nerve do not have recruitment, as this is a symptom of disease in the cochlea itself. Again this underlies the difficulty of someone not familiar with hearing loss to conjecture as to its causes. Recruitment is fully discussed in the article in *The Laryngoscope* [7]. Therefore, it is very unlikely that Paget's disease caused Beethoven's hearing loss.

3.10 Where did the lead come from?

Although lead can come from a variety of sources such as dishes, lead-lined wine flasks, lead crystal, or mineral water at spas, Beethoven's expenditures for wine were excessive [17]. He drank wine at every meal. Although Beethoven's personal secretary never mentioned that he had a problem with alcohol, his physicians were well aware of it. His secretary did say that "Beethoven preferred wine of the heights around buda to every other" [18]. Lead was added to this inexpensive wine to improve the flavor. In addition to his father, Beethoven's family had a history of alcohol abuse. It is interesting that George Frederic Handel also had lead poisoning [19].

3.11 Recent additional evidence

A fascinating patient in Italy was reported in 2021 that dramatically emphasized the relationship that lead has to Beethoven's deafness [20]. A 64-year-old woman presented with a slowly progressive hearing loss. She also complained of abdominal pain and tingling in her upper extremities like Beethoven. An audiogram showed a moderately severe hearing loss. Elevated levels of lead were found in her blood and urine. An examination of possible lead sources revealed that she used a cooking pan with a worn ceramic surface that released lead into her food. She underwent chelation

with ethylenediaminetetraacetic acid disodium salt which was injected into her to bind to the lead and allow it to be excreted from the body. After 1 year, her symptoms had all resolved, and her hearing was stabilized.

3.12 How did hearing loss affect Beethoven as a person?

Hearing loss can be very debilitating. This was especially true with Beethoven whose livelihood was dependent on his hearing. His loss caused him to be depressed, and at one time, the depression became so severe that he considered taking his own life [8]. Poignant is the picture of him when his ninth symphony was first performed. He was the honorary conductor and faced the orchestra. When the performance was finished, the contralto soloist had to turn him around to face the audience in order for him to see their applause, as he could not hear it [21]. It also significantly influenced his relationships with others. He was largely a solitary figure. He would very much have liked to have been married, but women generally found him eccentric. One cannot comprehend the depth of sorrow that this disability engendered in this gifted man.

3.13 How did his hearing loss affect his ability to compose?

On long walks that he liked to take, he would hear music in his head and write it down on a sketch pad. He did not just hear the melody, and later harmonize the parts, he heard it in its entirety [22]. With his loss of hearing, he was not able to check his music on the piano, but his superb ability to hear intervals in his head left his ability to compose unaffected. His music has been held in high esteem for over two centuries and continues to inspire millions. He recognized the great gift that he had to give to mankind and was consummately successful in doing so. Although Beethoven's hearing loss did not affect the quality of his music, his *Eroica* and 5th Symphony, the opera *Fidelio*, and the *Egmont* and *Coriolan* overtures reflect his heroic resolve and determination to conquer his suicidal thoughts and go on creating [23]. This heroic attitude is also heard in the *Waldstein* and *Appassionata* Sonatas, three string quartets opus 59, and the oratorio *Christ on the Mount of Olives*.

4. Conclusion

The best current evidence is that the most likely cause of Beethoven's hearing loss was his consumption of wine tainted with lead. Fortunately, it did not affect his ability to compose.


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