

Deafness and liver disease in a 57-year-old man: a medical history of Beethoven

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Ludwig van Beethoven had a number of medical conditions, including deafness and chronic liver disease, for which there are contemporary descriptions. An autopsy was performed on the day after his death. Physicians and historians have tried to reinterpret original sources to determine the causes of his deafness and systemic illnesses. We have reviewed the differential diagnoses that have been proposed by otologists and physicians. Clinical and post-mortem findings point to renal papillary necrosis and liver cirrhosis of unknown aetiology. In the absence of further histological examination, there is no definitive answer to the cause of Beethoven's deafness and gastro-intestinal symptoms.

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Introduction

Ludwig van Beethoven (1770-1827) symbolises the archetypal Romantic hero.¹ Despite having physical disability, he created music that is central to the modern repertoire.² His symphonies figured prominently during the German unification and the return of Hong Kong to the People's Republic of China. Since his death in 1827, Beethoven's medical biography has been the subject of speculation by historians and doctors. Attempts to explain the reason for his deafness and systemic complaints are hampered by the limitations of medical science in his time. The otoscope was not invented until 1854 by Toynbee, whereas the microscope, thermometer, and stethoscope were not then in widespread use.³ Corti's neuroanatomical work on the organ named after him and Ferrier's discovery of the auditory cortex were still to come.⁴ The pattern of diseases in Europe was also different from that of today. For example, smallpox, typhoid, tuberculosis, and syphilis were more common. Nevertheless, modern reviewers know much about the medical history of Beethoven from his diaries, extensive correspondence, conversation books, and contemporary accounts.⁵ He was treated by some of the most distinguished

physicians of his day, including Anton Braunhoffer; Jakob Staudenheim, personal physician to the Emperor Franz Joseph I; Giovanni Malfatti, founder of the Viennese Society of Physicians; and Johann Frank.⁶ An autopsy was performed by Johann Wagner, assisted by a young Karl von Rokitansky, who is now considered to be the father of modern morbid anatomy.⁷ In this article, we review the current English literature on the theories behind Beethoven's deafness and medical illnesses, based on a *Medline* search from 1966 to 2000.

Beethoven's deafness

Beethoven first revealed his symptoms in a letter dated 29 June 1801 to Dr Franz Wegeler, who was Professor of Medicine at the University of Bonn, Germany⁸:

For the last three years my hearing has grown steadily weaker, and the first cause of this is said to be my bowels, which as you know were already troublesome in the old days...I really lead a wretched life, for nearly two years I have been avoiding almost all company, just because I find it impossible to say to people: I am deaf...Sometimes I can scarcely hear a person who speaks softly...but if he shouts, I can't bear it.

This description suggests a recruiting sensorineural loss. In a letter dated 2 days afterwards, on 1 July 1801, he admitted to his friend Karl Amenda⁹:

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Let me tell you that my most prized possession, my hearing, has greatly deteriorated.... You will realize what a sad life I must now lead, seeing that I am cut off from everything that is dear and precious to me.

These sentiments were echoed in the Heilingenstadt testament of October 1802, which was discovered posthumously by his admirer Anton Schindler.^{10,11} The deterioration in his hearing coincided with his second period of musical development; the disability ended his career first as a virtuoso and then as a conductor. Spohr reported on one performance by the former child prodigy¹²:

It was not a treat; for one thing the piano was badly out of tune... his deafness had robbed him of nearly all his once-celebrated virtuosity. At *forte* passages the poor deaf fellow banged the key so vigorously that the strings twanged while in the *piano* passages he played so softly that whole groups of notes went unheard...

At a poignant scene at the end of the first performance of the Ninth Symphony on 8 May 1824, the contralto, Caroline Ungher, had to induce him to turn round and face the appreciative audience as he could not hear the applause.¹³

Beethoven's deafness began when he was 28 years of age. The left ear was involved first, and initial symptoms included tinnitus and high-tone hearing loss, which was associated with poor discrimination and recruitment. The symptoms were intermittent but after 1 year, they became persistent.¹⁴ The ear trumpets that he used are preserved today at the museum in Bonn. Czerny recorded that from 1812 to 1816, Beethoven's friends needed to shout to be understood.¹⁴ A piano with a more powerful tone was specially designed for him by Konrad Graf, the imperial court piano maker.¹⁵ He was forced to abandon organ recitals by the time he was 30 years old and was dissuaded from conducting at 42. In 1817, he started using notebooks to communicate with the outside world; these conversation books provide a tantalising glimpse into his personal life, from the mundane to the inspired.^{16,17} According to one account, he used a wooden rod, which he held at one end between his teeth; he placed the other end at the soundbox of the piano, suggesting an element of conductive hearing loss.^{14,16} The post-mortem examination in 1827 revealed the following^{18,19}:

The Eustachian tube [and]...the facial nerves were very much thickened. The acoustic nerves

on the other hand were wrinkled and were without a medulla. The auditory arteries running near them were dilated beyond the size of the lumen of a raven's quill and were cartilaginous. The left acoustic nerve was much the thinner...the right had a much thicker white root, the brain substance in the region of the fourth ventricle was much denser in consistency and more vascular than those nerves which arose from it.

The petrous bones and ossicles were removed. They were last seen at the Pathology Museum of Vienna but have since been lost. Otologists have tried to piece together the above findings in the light of modern medicine and have proposed the following diagnoses.

Chronic otitis media

In chronic otitis media, there is typically a history of ear discharge, acute exacerbations, and earache. These features were not reported by Beethoven's doctors. In addition, autopsy did not reveal perforation of the tympanum or the presence of cholesteatoma.¹⁵

Autoimmune sensorineural loss

Beethoven's long history of abdominal symptoms is considered by some to be due to inflammatory bowel disease. In rare cases, this is associated with hearing loss, which is thought to be secondary to vasculitis.¹⁵ This theory, like many others, is difficult to prove or disprove.

Ménière's disease

A diagnosis of Ménière's disease (endolymphatic hydrops) is considered unlikely, because dizziness and vestibular features are prominent and would have been noted by Beethoven.^{14,16}

Postinfective syndrome

There was no record of meningeal symptoms, and other pathological changes were absent at the brain autopsy.¹⁶ Toxic neuritis or labyrinthitis (otitis interna) may occur secondary to typhoid, and 5% of cases are complicated by serous effusion. The onset would be expected to be more acute, with tympanic perforation and signs and symptoms of acute otitis media.^{14,15}

Syphilis

Syphilis was once a popular diagnosis.¹⁷ If the congenital form is considered, this must be of improbably late onset and no other associated features such as a high arched palate or nasal stenosis were observed. The acquired form would be expected to cause more vertiginous symptoms. In addition, there was no historical evidence suggestive of rash, tabes dorsalis,

or dementia, and the auditory artery did not show chronic inflammatory changes.^{17,18}

Paget's disease

The combination of headaches, tinnitus, and deafness, which can be a presenting complaint in early disease, may be explained by Paget's disease (osteitis deformans); the terminal event being partly due to high-output heart failure. Proponents of this theory point to the thickness of the skeletal remains and drawings of Beethoven by artists such as JD Böhm and JPT Lyser, and descriptions by friends which highlight his unusual characteristics: large asymmetrical head, protruding forehead, and short legs in proportion to the trunk.²⁰⁻²² But the age of onset before 40 years is unusual. The skull specimen was incomplete and the measurements taken at the first exhumation are disputed. Osteodystrophy involving other bones was not mentioned. Furthermore, the sketches by the above artists were surely caricatures. A more accurate representation of the composer, made more than 15 years after the onset of the deafness, such as the bust by the sculptor F Klein or the drawing by August von Klöber, did not show the features of long-standing Paget's disease.²² In addition, X-rays that were taken of bone fragments allegedly from the composer did not show bone abnormalities. The current German literature as quoted by Davies' review does not support this diagnosis.¹⁵

Otosclerosis

Bilateral progressive hearing impairment starting in early adult life in the absence of any reason for deafness suggests a probable diagnosis of otosclerosis (otospongiosis), in which there is ankylosis of the stapedovestibular joint. In advanced cases, cochlear degeneration with secondary atrophy of the acoustic nerve may occur and result in tinnitus and sensorineural deafness. But a positive family history is found in 50% of cases, and females are twice as likely to be affected.¹⁴ Mercury was prescribed by his doctors—whom Beethoven called “medical asses”. This heavy metal may have exacerbated his hearing, as it is ototoxic.

Beethoven's systemic disease

Alcoholism featured prominently in the composer's family history. From adolescence, he would have episodes of asthma, and headaches in the winter months probably due to pansinusitis.^{17,18} Beethoven had also had a childhood illness that resulted in facial scarring, which is usually attributed to smallpox. Alternatively, it has been regarded as lupus pernio or the malar rash of systemic lupus erythematosus. The problems that

dogged him from his late 20s were diarrhoea alternating with constipation and abdominal colic.²² From his 40s onwards, Beethoven had episodes of polyarthralgia, which contemporary physicians labelled as “rheumatism”. With increasing ill health and relationship difficulties, the composer neglected his personal appearance, became preoccupied with his finances, and became more abrasive and distrustful. At restaurants, he would argue with bemused waiters over the price of a roll and in 1821, he was arrested by the police, thinking he was an itinerant beggar.^{23,24}

Final illness

On 5 December 1826, Beethoven's health deteriorated after he was exposed to cold weather during a journey to Vienna. His swollen abdomen and feet had already been noted that autumn. His last physician, Dr Andreas Wawruch, Head of Medical Services at the Allgemeines Krankenhhaus in Vienna gave an account of his terminal decline¹⁸:

...his feet were tremendously swollen. From this time on dropsy developed, the segregation of urine became less, the liver showed plain indication of hard nodules, and there was an increase of jaundice.

Paracentesis was performed by Dr Johann Siebert and more than 10 L of ascitic fluid were tapped, but the procedure was complicated by erysipelas. The patient survived to be tapped on three more occasions; these were uncomplicated. For 4 months, he was confined to bed while friends, knowing the gravity of the situation, came to pay their last respects.¹⁶

On 23 March 1827, Beethoven finalised his will, appointing his attorney, Dr Bach, as trustee and leaving his estate to his nephew, Karl. He received the last sacraments 1 day later, when he was still coherent. His last words referred to the arrival of a case of Riesling, which he realised he was too ill to drink. The next day, he fell into a coma and died in the late afternoon on 26 March during a thunderstorm.²⁵ The autopsy was performed by Dr Johann Wagner on 27 March. The original report, written in Latin, was rediscovered only in 1970 at the Vienna Museum of Anatomical Pathology²⁶:

The corpse was very emaciated, especially in the limbs, and sown over with black petechiae; the abdomen, which was unusually dropsied, was distended and stretched...In the cavity of the abdomen four quarts of a greyish-brown turbid fluid were effused. The liver appeared

shrunk up to half its proper volume, of a leathery consistence and greenish-blue colour, and was beset with knots...The spleen was found to be more than double its proper size, dark coloured and firm. The pancreas was equally hard and firm. Both kidneys were invested by a cellular membrane an inch thick...every one of their calices was occupied by a calcareous concretion of a wart-like shape and as large as a split pea.

Systemic lupus erythematosus

In 1823, there was an 8-month period when Beethoven was afflicted with eye pain, which has been regarded as uveitis.^{16,22} The constellation of signs and symptoms which included episodes of 'rheumatism', eye pain, and facial scarring were attributed to connective-tissue disorders such as systemic lupus erythematosus. Larkin first argued the case for this diagnosis on the bicentennial of Beethoven's birth.¹⁸ However, systemic lupus erythematosus is nine times less common in males.²² Isolated deafness in the early stages cannot be explained, and liver cirrhosis is not part of the natural course of progression; death from renal involvement is more common. Alopecia, lymphadenopathy, pericarditis, and a bleeding tendency were not reported by contemporary sources.^{16,22}

Tuberculosis

As both his mother and brother Karl died of tuberculosis, Beethoven may have contracted tuberculosis in his early life.²² But to explain his gastro-intestinal and neurological symptoms would imply a diagnosis of miliary tuberculosis lingering for 30 years. Pachymeningitis involving the eighth cranial nerve is again unlikely, due to the high mortality and morbidity of this condition.¹⁶

Inflammatory bowel disease

A diagnosis of inflammatory bowel disease would account for the abdominal pains, chronic diarrhoea, arthritis, eye disorder, and chronic liver disease.^{15,16,22} Deafness has also been associated with ulcerative colitis, but there is no history of rectal bleeding—an alarming development which Beethoven's voluminous correspondence would have noted. The autopsy did not reveal evidence of the strictures, perforations, or adhesions that are found in Crohn's disease. The possibility that a diarrhoeal illness in 1792 was dysentery, which progressed to seronegative enteropathic arthritis is intriguing: sacroileitis and recurrent uveitis are associated with this condition.^{15,16,22}

Inflammatory bowel disease

Chronic diarrhoea and abdominal pain are features of inflammatory bowel disease, but this does not account

for the severe pain and dehydration that the composer reported at times.²² The polyarthralgia and liver disease would also remain unexplained.

Alcoholism

Alcoholism is a traditional cause of hepatic disease, and from Thayer's records of Beethoven's wine receipts, he certainly drank enough alcohol for a sufficiently long period to suffer from alcohol-induced liver damage.²⁷ Distractors point out that this condition is associated with a micronodular cirrhosis. Chronic pancreatitis would explain the diarrhoea and abdominal pain, but steatorrhoea would have been evident.

Sarcoidosis

Both Palfermann²² and Drake²⁸ have suggested sarcoidosis as the cause of Beethoven's systemic illness. Lupus pernio would cause the facial scarring, sarcoid infiltration or portal hypertension would result in splenomegaly, cranial neuropathy would explain the hearing loss and the colic would be due to coexisting renal colic. Five percent of patients with sarcoidosis have neurological abnormalities: bilateral seventh and eighth nerve palsies are suggestive of this condition.²⁸ Apart from cranial neuropathy, hypothalamic dysfunction, and hydrocephalus, aseptic meningitis and peripheral neuropathy are manifestations of neuro-sarcoidosis. With severe liver involvement, other main sarcoidotic organ damage would have been expected but these were not found. Gastro-intestinal involvement is unusual and typically subclinical.

Renal papillary necrosis

The description of the kidneys at autopsy has been interpreted as evidence of perirenal fibrosis with calcified necrotic papillae,^{26,29} which would make this the first anatomical description of renal papillary necrosis.³⁰ This can be due to severe liver disease or analgesic abuse: salicylate precursors were available at the time and Beethoven is known to have taken regular amounts of analgesics for his headaches and rheumatism. Calcium deposition in nephrocalcinosis due to hypercalcaemia (eg sarcoidosis) or tuberculosis would have been more extensive.

Beethoven was buried on 29 March 1827. An estimated 10 to 30 thousand people attended the funeral. Among the mourners was Franz Schubert, who was to die himself the following year. Both were buried at Währing Cemetery in Vienna. The actor Anschutz delivered the funeral ovation,³¹ which included the following lines:

...the last master of tuneful song, the organ of soulful concord, the heir and amplifier of Handel

and Bach's, of Haydn and Mozart's immortal fame is now no more, and we stand weeping over the riven strings of the harp that is hushed.

Discussion

Musicologists and physicians have long speculated on the influence of illness on creativity.³²⁻³⁴ Smetana, Rossini, Fauré, and Vaughan-Williams also had significant hearing impairment, but at a later stage in their careers. By concentrating his energy and by freeing his creativity from the distractions of external stimuli, Beethoven—through his disability—produced the masterpieces of his mature work.³⁵ His compositions for the piano decreased in number with the end of his career as a virtuoso pianist, while his other musical output increased. Naturally, there were influences apart from his disability: the years in which few pieces were produced, from 1817 to 1820, was a time overshadowed by legal proceedings regarding the guardianship of his nephew.⁹ And there was no doubt a stylistic evolution, which cannot be attributed merely to his state of health.

The mechanism of musical perception and the cortical organisation to form musical ideas are not understood. Yanz and Liston have shown that there was no decrease in Beethoven's use of high tones from an analysis of his symphonic work, which suggests that he perceived these overtones in his mind, rather than relying on auditory feedback.³⁶

Can the composer's illnesses be explained by a single unifying multisystem disease or by a combination of the mundane such as alcoholism and irritable bowel syndrome? The systemic effects of Whipple's disease have been implicated, as well as chronic active hepatitis and selective immunoglobulin A deficiency; the latter can be associated with recurrent sinusitis, otitis media, ulcerative colitis, and liver cirrhosis.^{16,37} Specialists have attributed Beethoven's symptoms to uncommon or fashionable diagnoses that have not survived close scrutiny. Clinical and post-mortem findings point to renal papillary necrosis and liver cirrhosis of unknown aetiology. In the absence of further histological examination, there is no definitive answer to the cause of his deafness and gastro-intestinal symptoms.

Addendum

On 17 October 2000, William Walsh at the Pfeiffer Research Center in Illinois, United States, announced the discovery of high levels of lead from a sample

of hair from Beethoven.³⁹ This particular sample, consisting of 160 hairs, was obtained at the deathbed by Dr Ferdinand von Hiller and was donated by Dr Alfredo Guevara for testing. The source of lead was not commented on but could have been derived from the soil, drinking water, medicines, or earthenware. Contamination from the burial environment is unlikely. Assuming that it is authentic (DNA comparison with other material said to be from the composer will be performed) and that it is free from contamination during storage, lead poisoning is a plausible diagnosis, because it explains the colic, arthralgia, headache, and behavioural problems. This is an attractive theory because tissue lead is quantifiable. Former President of the United States Andrew Jackson was also diagnosed with plumbism after retrospective analysis of his hair.³⁸ As with other diagnostic possibilities, however, gaps remain: the deafness and cirrhosis is not accounted for. The hair ranged from 7 to 15 cm in length and represents only the last 6 to 12 months of the composer's life. Due to lead's chronic toxicity, one would also expect complaints of metallic taste, wrist-drop, and numbness (due to neuropathy); these were not reported by contemporaries. The last word has not been said on this topic.

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