## **History of Medicine:**

# **Beethoven's Deafness**

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#### SUMMARY

Ludwig van Beethoven died in 1827, aged 57 years. During the last 30 years of his life he suffered a progressive bilateral hearing loss, which left him profoundly deaf during his later years. The possible causes of this deafness, the subject of much conjecture, are discussed.

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The tragedy of Beethoven's deafness has become a legend. That one of the greatest musicians the world has ever known should suffer this handicap is barely credible. For more than half his life and for most of his productive years as a composer he suffered a deafness that was progressive to the eventual point of total loss of hearing. This disability greatly influenced his way of life, his personal happiness and doubtless his creative art, but it was during those years after he had lost the ability to converse that most of his finest and most monumental works were completed.

Born in Bonn in 1770, the second child of Johann and Maria Magdelene van Beethoven, his life was a succession

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of personal, and at times extreme, sorrows, difficulties and tragedies. During the early period of his adulthood in Vienna, Beethoven showed signs of the ill health that was to plague him for the rest of his life. The most serious problems began at the turn of the century and included a chronic diarrhoea, recurrent severe headaches and a progressive deafness.

He first noticed his hearing loss, initially of the left ear and soon afterwards of the right, in his late twenties and by the age of 30 years he was shunning social gatherings in order that its detection be avoided. At this time, in panic and despair, he consulted a number of physicians and with equal ineffectiveness was prescribed numerous remedies including oil of almonds, cold baths, Danube baths, vesication of his arms with bark, herbs on the belly and galvanism. Of all these treatments only the Danube baths helped, and that only his tinnitus and intestinal upset.

He was advised to visit the country to avoid exposure to noise, and so in 1802 he retired to Heiligenstadt. No improvement occurred. This and the marriage of an earlier *amour* plunged him into renewed depths of depression, prompting his writing of the famous Heiligenstadt testament,<sup>3</sup> a letter to his brothers Karl and Johann that took the form of his will. This document, best read in full, gives dramatic insight into the emotions of the deaf.

In Vienna Beethoven settled back into his work in which he must have found some solace, and despite various minor setbacks, the following decade was his most 1586

productive. He appears to have accepted his handicap and to have again enjoyed a normal social existence. 'Let your deafness no longer be a secret, even in art', he pencilled onto one manuscript. In his writings thereafter he rarely referred to this affliction.



Fig. 1. Beethoven's ear trumpets.

His deafness progressed relentlessly and by 1814 he was using ear trumpets which had been made for him by Mälzel, a fellow musician. A few years later even these hearing aids had become ineffectual. He had already been using his famed *Konversationshefte* (notebooks), of which 138 found their way into the Berlin Museum, for to converse with him in the normal manner had become impossible. He also used a special wooden drumstick, one end held between his teeth and the other against the piano in order to improve his hearing while composing, but for how long he used this is not known.

His fame was now world-wide, but his public appearances had become an embarrassment. His piano performances suffered a forced ending in 1814, and in 1822 he conducted for the last time.

In later life Beethoven became a man of strange habits. Extraordinarily industrious in his creative work, he became withdrawn, depressed and introspective, took progressively less care of his person and consequently his health suffered. He ate sporadically and indifferently, drank liberally and consumed vast quantities of medicine of dubious merit prescribed by his numerous medical advisers. His tenacity, his emotionally labile and naturally aggressive personality, and his inconsistent and difficult personal relationships with others all remained evident.

A 2-day return journey to Vienna in 1826, made in an open coach in bad December weather, eventually led to his death. He initially went down with pneumonia and later liver failure, from which he never recovered. He eventually died on 26 March 1827.

A postmortem examination was performed 2 days later by Wagner and Rokitansky and their findings (in translation) are recorded here in full.<sup>2</sup> The temporal bones were removed and put into the National Museum in Vienna, but half a century later, when further examination was decided upon, they were missing and as yet have not been found.

### THE AUTOPSY FINDINGS

'The cadaver, and especially the limbs, was very wasted and covered with black petechiae; the abdomen very much swollen and tense with water.

'The auricular cartilage was large and irregularly formed; the scaphoid fossa, but particularly its conchal portion, was very roomy and deeper by half the normal; the various projections and depressions were considerably exaggerated. The external auditory canal appeared to be lined with glistening cutaneous scales; this was especially marked near the concealed tympanic membrane.

'The Eustachian tube was very thickened and its mucosa rugose and rather narrowed at the bony part. At its pharyngeal opening, and near the tonsils, scarred depressions were to be seen. The mastoid process was large and not marked off by any groove; its cells were conspicuous and lined by highly vascular mucosa. The whole of the petrous portion of the temporal bone, showing large vessels, was similarly well supplied with blood, especially in the region of the cochlea, the spiral lamina of which seemed slightly reddish.

'The facial nerves were of considerable thickness. The auditory nerves on the other hand seemed shrunken and without pith; the accompanying arteries were of the calibre of a crow-quill, and of cartilaginous consistency. The left auditory nerve, much the thinner, arose by three very thin greyish roots; the right by one root, stronger and pale white; the superficial substance surrounding the fourth ventricle was much denser in consistency and more vascular than these nerves which arose from it. The brain was softer and more watery than normal, and the convolutions appeared to be twice as deep and more numerous than usual. The vault of the skull was markedly and uniformly dense and about half an inch thick. The thoracic cavity, and its contents, were normal.

'The abdominal cavity contained 8 litres of turbid, greyish fluid. The liver appeared shrunken to half its size, leathery in consistency, and greyish-blue in colour; its tuberculated superficial surface and its cut surface were full of knotted masses the size of a bean; all the vessels were very contracted, thickened and devoid of blood. The bile was a darkish-brown fluid, containing a gritty sediment.

'The spleen was twice its normal size, blackish in colour and tough; similarly the pancreas was large and firm, and its duct was the width of grey quill. The stomach and the intestines were very distended with gas. Both kidneys were contained in a cellular membrane one inch in thickness, dripping a turbid brownish fluid, its structure was loose, and pale red in colour; each calyx contained a chalky deposit, wart-like in shape, and similar to a pea cut across.'

His body was exhumed in 1863 and again in 1888, but little was added to Wagner's original findings. Apart from the illnesses already mentioned, it is known that Beethoven also suffered at one time or another from small-

Otosclerosis was not diagnosed at postmortem examination; a macroscopic defect of stapes mobility was not reported by Wagner, and it is unlikely that it was overlooked. The importance of ankylosis of the stapes had first been recognised and reported by Valsalva in 1735 and since that time this middle ear dysfunction had been further and extensively investigated by others. The effective use by Beethoven of his drumstick does not necessarily confirm a middle ear cause of his deafness. The audiological weaknesses in the argument for this type of deafness have been excused by the inclusion in the diagnosis of cochlear otosclerosis4 and its consequential perceptive hearing loss. Since the widespread use of surgery for this disease numerous reports of progressive, to the point of severe, bilateral hearing loss of onset in early adulthood, and due to manifest otosclerosis, have been made.<sup>4</sup>

Until the missing temporal bones are microscopically examined this diagnosis, indeed any suggested diagnosis, must remain unconfirmed. It must, however, be added that he experienced an auditory phenomenon, an initial high-tone hearing loss, which is out of keeping with uncomplicated conductive deafness including otosclerosis.

Syphilis has been, especially in earlier writings, strongly suggested as the cause,6 but again histological confirmation was not available and neither was there adequate support on serological or clinical grounds for this diagnosis. It was at that time a very common disease and was popularly discredited as the cause of a multitude of misunderstood disorders. Beethoven's deafness, with its insidious onset, its progressive course and the absence of accompanying vestibular symptoms (vertigo), is unlikely to have been the result of syphilitic labyrinthitis, congenital or acquired. The audiological picture of this disease may be bizarre and vestibular symptoms are not necessarily present in all cases. Cases of the congenital form of the disease in which the deafness is bilateral, rapidly progressive and presenting in early adult life, have been reported.

The postmortem finding of changes in the surround of the brain stem suggests a chronic localised meningeal reaction, thought to be syphilitic meningovasculitis. Syphilis was the cause that his contemporary medical attendants diagnosed and for which they treated him. His cirrhosis was at that time attributed to the same disease.

It has recently been suggested<sup>7,8</sup> that Beethoven suffered from osteitis deformans or Paget's disease, a condition that in its clinical manifestations would explain his physical appearance, especially of his skull with its hyperostosis frontalis that altered and progressed with age, his headaches, his deafness and the tinnitus. The fact that in later life he wore his hat far back on his head, leaving his forehead exposed, is as likely the result of enlargement of his head as a quirk of fashion.

The postmortem findings of a uniformly dense and thickened skull and hypervascular petrous temporal bones support this diagnosis. Further, the conductive deafness which may occur in the early phase of the disease is not necessarily the result of stapes ankylosis, but is more frequently due to attic impaction of the incudo-malleolar complex. This would, unless specifically looked for, pass unnoticed at postmortem examination. The disease, by involvement of the petrous temporal bones, produces a bilateral sensorineural deafness, especially of the high tones, and its advanced state produces multiple fractures and fissures in the labyrinthine capsule, degeneration of the cochlear epithelial elements and partial atrophy of the ganglion cells and auditory nerves. It could also account for the nephrocalcinosis found at postmortem examination and the facial spasm that Beethoven suffered in his later years. An early age of onset, although exceptional, can occur in this common disease that was originally described by Sir James Paget 50 years after Beethoven's death.

Apart from otosclerosis and osteitis deformans any other middle ear disorder is most unlikely. There is no history of otorrhoea or childhood ear disease and the normal postmortem findings, except for the vascular mucosal lining of the mastoid cell system, exclude such a diagnosis. The presence of a large and cellular mastoid is also indicative of the normal development and pneumatisation of a pathology-free middle ear cleft.

Cochlear deafness progressive to the point of total hearing loss, first appearing in early adulthood and without any other clinical stigma, is unusual. An hereditary origin for such is unsupported by Beethoven's family history. In his lifetime the onset of his hearing loss had variously been related to a fall, typhoid and a chill, all of which are most improbable causes. However, the necropsy findings of atrophic auditory nerves cannot be ignored.9

No firm conclusion regarding the cause of this most renowned of hearing losses arises from this publication. Those put forward during his lifetime had no scientific foundation and can be rejected; those put forward after his death remain conjecture. The truth awaits discovery in a missing Viennese specimen jar.

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