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TYPE:	Article CC:CCL
JOURNAL TITLE:	Journal of medical biography
USER JOURNAL TITLE:	Journal of medical biography
ARTICLE TITLE:	Beethoven's autopsy revisited: A pathologist sounds a final note
ARTICLE AUTHOR:	Oiseth, Stanley J
VOLUME:	25
ISSUE:	3
MONTH:	
YEAR:	2017
PAGES:	139-147 / 139-147
ISSN:	0967-7720
OCLC #:	

Processed by RapidX: 5/19/2022 3:40:02 AM

Original Article

Journal of Medical Biography
2017, Vol. 25(3) 139–147

DOI: 10.1177/0967772015575883
journals.sagepub.com/home/jmb
SAGE

Beethoven's autopsy revisited: A pathologist sounds a final note

Stanley J Oiseth

Abstract

This review of the original autopsy report of Beethoven's remains indicates Paget's disease within the skull, which was dense and twice normal thickness, with dilated vessels at the petrous bone. The facial nerves were enlarged and the eighth nerves atrophied despite their sharing a common meatus at the internal auditory canal. Nephrolithiasis and pyelonephritis with cortical and perinephric abscesses were also reported. The hypercalcaemia was probably caused by hyperparathyroidism, which may be associated with Paget's disease, and both may have played a role in his psychiatric symptoms as well as in his abdominal pain and gastrointestinal complaints. Since Paget's disease may also be associated with gout, some of the joint pains could be attributable to this as well. Hypovitaminosis A from chronic pancreatitis is suggested as a cause of painful eyes and either quinine abuse or severe hypercalcaemia as a cause of arrhythmias. Beethoven died of terminal cirrhosis with chronic pancreatitis, most likely related to chronic excessive intake of alcohol. 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, some of which appear to have influenced his musical compositions.

Keywords

Music, cirrhosis, Paget's disease pancreatitis, osteitis-deformans, hyperparathyroidism, nephrolithiasis, deafness, Beethoven

The last half of Ludvig van Beethoven's life was marked by progressive deafness and headaches, recurrent colicky abdominal pain, alternating bouts of diarrhoea and constipation, rheumatic disorders including joint pains and superficial 'abscesses', bizarre misanthropic behaviour, and a painful eye condition in the third year before his death on 26 March 1827 at the age of 56 from complications of cirrhosis.[1–3] The autopsy was undertaken one day after death by Dr Johann Wagner of the Vienna Pathological Museum with his assistant, Dr Karl von Rokitansky (1804–1878). The original one-page autopsy protocol in Latin has been reproduced previously and selected parts of Alexander Thayer's English translation,[1] placed in italics, are interpreted here.

> *The cavity of the chest, together with the organs within it, was in the normal condition.*

This tells us that there was no gross pathology of the lungs or heart, and there was no hilar or mediastinal lymphadenopathy. Therefore sarcoidosis, systemic lupus erythematosus and tuberculosis, as suggested by various other authors, are unlikely.[3–5]

> *The corpse was very emaciated, especially in the limbs, and sown over with black Petechien; the abdomen, which was unusually dropsied, was distended and stretched.*

The body had massive ascites with the cachexia of end-stage cirrhosis, which was not relieved by the continuous drainage of ascitic fluid through an open wound that developed at the site of one of multiple paracenteses undertaken during the previous three months, as described by his last attending physician, Dr Andreas Warwuch.[6(pp. 40–43)] Petechiae are typically

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due to thrombocytopenia and are seen in disseminated intravascular coagulation, which was most likely due to bacterial sepsis or septic shock but may also be seen in end-stage cirrhosis.

> *In the cavity of the abdomen four quarts of a grayish-brown turbid fluid were effused.*

Ascites from cirrhosis alone is clear yellow, so the added turbidity was most likely caused by a superimposed pyogenic bacterial infection related to the open abdominal wound. Other causes of turbid ascites include an open pancreatic duct or cyst draining into the abdomen ('pancreatic ascites') or tuberculous peritonitis. However, evidence of a pancreatic cyst or draining duct was lacking in Dr Wagner's description of the gland nor was there mention of the well-known effects of tuberculosis in the lymph nodes or in any other tissue.

> *The liver appeared shrunk up to half its proper volume, of a leathery consistence and greenish-blue colour, and was beset with knots, the size of a bean, on its tuberculated surface, as well as in its substance; all its vessels were very much narrowed and bloodless. The spleen was found to be more than double its proper size, rose-coloured, and firm.*

This is the description of macronodular cirrhosis, since common beans measure more than 0.3 cm, the measurement distinguishing micronodular from macronodular cirrhosis. Although alcoholic cirrhosis is typically micronodular, the regenerating nodules of a micronodular liver may enlarge to macronodular size and therefore macronodularity alone is not very helpful in arriving at a specific diagnosis. From his housekeeper's records, we know that Beethoven had been consuming one to three litres of wine per day,[7] and there is evidence that he used alcohol for pain control, so it is possible that his cirrhosis was due to alcohol ingestion although a viral aetiology as a primary or contributing factor in his cirrhosis cannot be excluded. The spleen was enlarged due to cirrhotic portal hypertension, and the hepatic blood vessels were narrowed because of the compressive growth of the regenerating cirrhotic nodules; they appeared void of blood due to the premortem hypotensive and haemorrhagic states, combined with his chronic anaemia from the coagulopathy of cirrhosis as noted by his two-year history of repeated episodes of epistaxis and haemoptysis.[8]

Dr Warwuch had described his patient as jaundiced, which explains at least in part the green colour of the liver, but the blue colour was most likely attributable to infection with *Pseudomonas aeruginosa* which produces the blue-green pigments pyocyanin and pyoverdin.[9] This Gram-negative bacterium is ubiquitous in nature and could easily have gained entrance through the contaminated, draining abdominal wound. The bacterial pigments also probably contributed to the dark appearance of the blood in the spleen and skin petechiae. It is conceivable that the 'Petechien' may have been the initial haemorrhagic vesicles of ecthyma gangrenosum, seen in some patients with *P. aeruginosa* bacteraemia. Post mortem production of sulfhaemoglobin and methaemoglobin production may also have contributed to the dark appearance of the blood and spleen, which is nonspecific and not uncommonly found at autopsy.[10,11] Haemochromatosis as a cause of cirrhosis and the dark colour of the spleen has been proposed[12] but is unlikely since the distinct rust-brown colour that disorder imparts to the heart, liver, and pancreas was not mentioned, and excessive skin pigmentation was not described.

> *The pancreas was equally hard and firm, its excretory duct being as wide as a goosequill. The stomach, together with the bowels, was greatly distended with air.*

This is the typical picture of end-stage chronic pancreatitis with scarring and dilated ducts and often associated with even moderate consumption of alcohol. A normal duct is 4–5 mm in diameter, and a goosequill is wider than that. Evidence against Crohn's disease is provided by the greatly distended stomach and bowels since the thickened rigid bowels of that condition would not be distensible. There was also no mention of ulcers, pseudo-polyps or blood in the intestine, nor was there any critical history of haematochezia or chronic malnutrition, which would be expected after several decades of ulcerative colitis or Whipple's Disease although these have been suggested.[1,13–15] Before his pre-terminal stage, Beethoven was always described as appearing quite healthy, and he had actually been taking long walks in the countryside just several months before he died.[6(p. 89)]

Post mortem bacterial overgrowth was responsible for the abundant gastrointestinal gas and can be quite marked in a patient who died in septic shock and whose body is not refrigerated before autopsy.

> *Both kidneys were invested by a cellular membrane of an inch thick, and infiltrated with a brown turbid fluid; their tissue was pale-red and opened out. Every one of their calyces was occupied by a calcareous concretion of a wart-like shape and as large as a split pea.*

Well-developed bilateral cortical and perinephric abscesses are nicely described here, as well as nephrolithiasis. Most of the cortical abscesses were ruptured

and described here as cortical kidney tissue 'opened out'. Since there would have been perinephric inflammatory and fibrotic reaction to each cortical abscess after it ruptured, and since the abscesses would have ruptured at different times, the larger, secondary perinephric abscesses were likely made from the fusion of multiple secondary abscesses with fibrotic septa to form individual 'cells' containing liquefied purulent necrotic material in each. This created the appearance of a 'cellular membrane' infiltrated with brown turbid fluid. Before the antibiotic era perinephric abscesses were a quite common sequel of acute pyelonephritis, commonly initiated by bacteraemia, unlike nowadays when the ascending route of infection predominates.

Erroneous interpretations of these classic pathologic findings have included perirenal fibrosis or even retroperitoneal fibrosis. This autopsy description has also been misinterpreted as a case of papillary necrosis with calcified papillae[16,17] but the calyces contain the calcifications – not the papillae. The anatomy of the kidney was well known at the time.

> *The calvarium exhibited throughout great density and a thickness amounting to about half an inch. The facial nerves were of unusual thickness, the auditory nerves, on the contrary, were shrivelled and destitute of neurina: the accompanying arteries were dilated to more than the size of a crow quill and cartilaginous. The Eustachian tube was much thickened . . . and somewhat contracted about the osseous portion of the tube.*

The skull was thickened to twice its normal size and dense throughout. A reportedly excellent life mask taken in 1812 when Beethoven was 41 years old shows, as described by the art historian Alessandra Comini (with the word '*is*' italicized by that author) that 'the forehead is large: pronounced, powerful, wide, rounded and prominent without being really high. In profile view it bulges slightly forward above the furrowed brow'.[18] A photograph of the skeletonized cranium taken at the time of the first disinterment of Beethoven's body in 1863 highlights even better the unusual physiognomy (see Figure 1). The notably irregular and large zygomatic bones made obvious in the skeletal remains are not usually commented upon but are a known occasional manifestation of PDB and contributed to the mildly leonine appearance of the composer. Although there has been some debate about the translation from Latin of the adjectives used to describe the skull, the measurement of "half an inch" is not disputed. One inch refers to the Viennese Zoll, an official measurement used at that time which is equivalent to 2.63 cm. Therefore, the skull measured 1.3 cm in thickness, which is approximately twice normal. To add emphasis to the point that it was indeed thickened, it helps to know that the pathologists at the time usually only measured something if it was abnormal. There are multiple causes of calvarial bone thickening with increased density, including Paget's disease of bone (PDB), haemolytic anaemia, hyperparathyroidism, acromegaly, osteopetrosis and rare bone dystrophies.

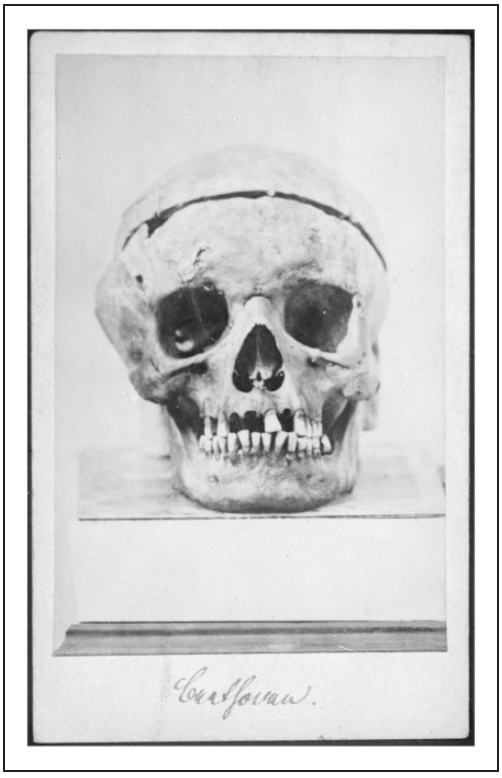

Figure 1. This photograph of the skull was taken at the time of Beethoven's first disinterment in 1863. Note the prominent frontal bossing and the irregular and markedly enlarged zygomatic bones, producing a "leonine facies" of which Paget's disease is a known cause. The bulging parietal area of the skull seen on the right side of the skull in the photograph layout may only be artifactual, due to an attempt by Dr. Gerhard von Breuning, a physician who had personally known Beethoven, to reconstruct the nine skull fragments and "fill in" missing portions of the skull with clay at the time of the disinterment.[68] Reprinted with permission from the Archives and Graphics Department of the Austrian National Library.[69]

Paget's disease

PDB stands out as the most likely cause in this case since it best matches the clinical findings, including the irregularity of the skull shape. The incidence of PDB is approximately 3% in Austria and Germany and, although most patients are more than 40 years old, 3% of cases occur under the age of 30[19];

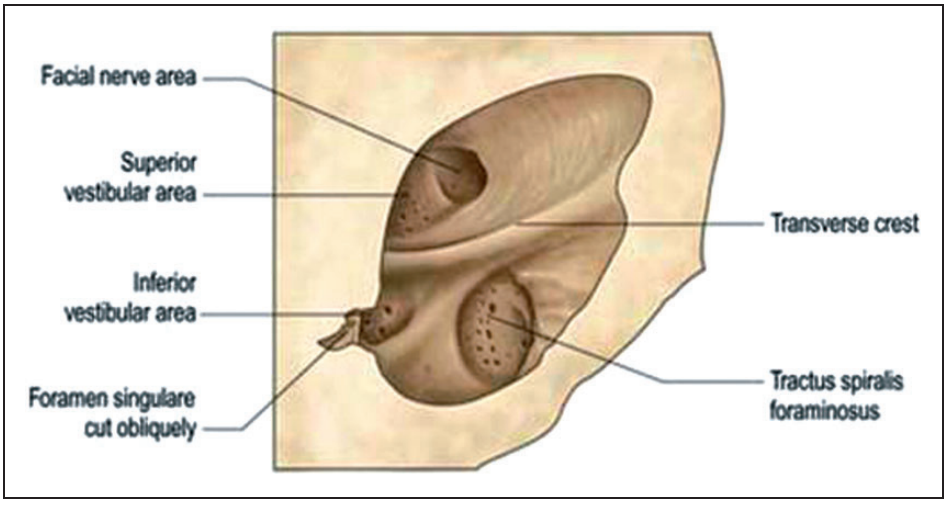

Figure 2. The fundus of the left internal acoustic meatus, exposed by a section through the petrous part of the left temporal bone nearly parallel to the line of its superior border. The facial nerve is already a well-formed bundle as it passes out through its foramen in the meatus, while the many individual peripheral fibers of the cochlear nerve must pass through small foramina of the tractus spiralis foraminosus before entering the cranium. Reprinted from Gray's Anatomy, 40th ed., with the kind permission of the Elsevier Publishing Company.[70]

Paget's disease has even been reported in children.[20] Approximately 50% of patients with Paget's disease of the temporal bones have hearing loss which typically is bilateral and of mixed type, usually with a conductive component at low frequencies and down-sloping sensorineural hearing loss at higher frequencies.[21–23]

Detailed histopathologic studies of the temporal bone in PDB have revealed various abnormalities due to bone remodelling, involving focal lysis and sclerosis with abnormal thickening which is the basic pathologic feature of PDB.[23,25] While loss of bone mineral density in the cochlear capsule was favoured in one imaging study of 68 temporal bones from patients with PDB as the major mechanism underlying the hearing loss in PDB, the author of that study conceded that there were probably multiple mechanisms of hearing loss in PDB, including damage to the auditory nerve.[24] Severe hearing loss may have multiple additive pathogeneses. The most frequent finding in one study of 15 temporal bones from 8 patients with PDB was obliteration of the bony canal of Cocugno that carries the inferior cochlear vein.[25] Other abnormalities that have been described include microfractures, fractures and fixation of the stapes footplate, microfissures and microfractures of the otic capsule, bleeding in the scalae, strial atrophy, and cystic lesions of the spiral ligament.[25,26] If compression caused the observed eighth nerve atrophy in Beethoven's case, one could ask how it could be possible to have coexistent enlarged facial nerves since the seventh and eighth nerves pass through the same canal? To understand this requires review of the anatomy of the nerve pathways in the internal auditory canal (see Figure 2). The eighth nerve is a sensory nerve, formed by the fusion of small vestibular and cochlear nerve twigs after they pass through small foramina before and within the internal auditory canal so they are quite susceptible to pressure ischaemia caused by any overgrowth of bone in this area, which would first compromise the small foramina. Microneuromas of eighth nerve branches have been described in the temporal bones of patients with PDB and hearing loss, consistent with blockage of cribrose areas by Pagetic bone causing injury to small nerve fibres followed by aberrant regeneration.[23] The facial nerve, on the other hand, is mostly a motor nerve and is already a well-formed single bundle before it passes through its single large foramen at the meatus and so it can more easily resist compression by growing Pagetic bone, although the nerve's lymphatic drainage could be easily obstructed, leading to lymphoedema and the enlargement of the right and left nerves noted by the pathologist. Although compressive effects on the eighth nerve can occur due to the effect of pressure while passing through multiple tiny obstructed foramina, the internal auditory canal itself may be of normal size, narrowed by bony growth, or even enlarged by pulsating blood vessels because of increased localized blood flow in Pagetic bone.[23,26,27] Dr Wagner did indeed describe dilated thickened intracranial arteries having a 'cartilaginous' quality, providing evidence of atherosclerotic plaques that commonly occur when blood flow is increased within enlarged tortuous vessels. Beethoven wrote in 1815 that he lost all of his hearing after a fall,[28]

a history consistent with acute nerve damage secondary to pathologic microfractures in Pagetic bone, occurring in nerves already severely damaged by chronic disease. Pathologic fractures are a well-known complication of PDB.

The bony part of the Eustachian tube was narrowed, consistent with extensive Pagetic involvement of the skull. Clinical evidence for bony impingement upon the cochlear nerve with neurosensory loss is given by Beethoven's initial intolerance for loud sounds despite inability to understand spoken words, as Beethoven explained in a letter of 1801.[29(p. 76)] This is evidence of nerve recruitment and is not seen in pure otosclerosis and so the other major competing diagnosis put forth to explain his deafness has been cochlear sclerosis or 'otosclerosis combined with nerve deafness'.[13,30,31] In addition, the high-frequency loss described by Beethoven is not typical of the condition. None of these diagnoses which have suggested Paget's disease as a cause of Beethoven's deafness explain the enlarged facial nerves.[32,33] Dr Wagner dissected the temporal bones but did not describe fixation of the stapes. The temporal bones were removed for study and never seen again, but it is clear from the gross description that Beethoven most likely had Paget's disease. Fragments of skull bones alleged to have belonged to Beethoven have been analysed and reported to show no evidence of Paget's disease.[34] This is not surprising, if indeed the attribution were genuine, since Paget's disease of the skull is not uniform and the histologic mosaic pattern of active Pagetic bone may undergo remodelling just like any other bone once the pathological stimuli cease and so the lamellar histologic pattern of normal bone may be seen in the final, fourth phase of the disease.[23] Hypercalcaemia and hypercalciuria occur in Paget's disease when the patient is immobilized by bedrest, a likely therapy when Beethoven was troubled by headache or joint and abdominal pains. He also may have had continuous hypercalcaemia since hyperparathyroidism is associated significantly with Paget's disease.[35] There was no mention of enlarged parathyroid glands but these glands were not described in animals until 1850 and Virchow first identified them in man in 1863[36] so, if they were enlarged, they were probably considered part of the thyroid gland. Sir James Paget did not describe the disease entity bearing his name until 1877,[37] but Dr Wagner showed the keen observational skills of a good pathologist by describing and quantifying the abnormal size of the cranium, even though he could not explain it.

Beside deafness, the symptoms that have been associated with Paget's disease are varied and numerous, including pain in the back, hips and extremities, enlargement of the skull, headache, tinnitus, and 'paranoia'.[19,38,39] This last is mentioned because the composer showed signs of bizarre and eccentric thought and behaviour in his own letters and actions, with bouts of melancholy and irritability.[6(pp. 99–100),40] He changed lodgings two or three times a year and had difficulty retaining housekeepers and cooks.[29(p. 161)] In his Heiligenstadt Testament, written when he was only 32 years old, he actually pleaded with his brothers to help his doctor find and make public a medical explanation for his 'disease' after his death, since he thought the world would judge him differently if the underlying physical reasons for his behaviour could be explained.[29(pp. 78–81)] Although bipolar disorder has been posited as a possible aetiology for his behaviour,[40] emotional instability is occasionally seen with hypercalcaemia and behavioural improvement is noted in patients who have undergone parathyroidectomy for hyperparathyroidism.[41] Some authors have commented that Beethoven had many signs and symptoms suggestive of the 'moans, groans, bones, and stones' of hyperparathyroidism.[42] Hypercalcaemia can also cause lethargy,[43] and this was a distinct complaint of the composer, since he could work until the early hours of the morning when he was young and needed only four or five hours' sleep before resuming his work; he would later write how tired he always felt.[44(p. 82)] Mental confusion may be referable to either hypercalcaemia or Paget's disease, which is associated with mental impairment including confusion, presumably because of hypoxia due to a shunting mechanism.[45]

Gout and pseudogout (calcium pyrophosphate disease)

Several other rheumatic disorders are associated with and can contribute to articular and musculoskeletal pain in patients with Paget's disease, including gout and pseudogout; in one series one quarter of male patients with Paget's disease had gout,[46] while another study showed that one quarter of patients with gout had Paget's disease.[47] Evidence of tophaceous gout in Beethoven includes occasional finger and foot abscesses, as well as the 'large, deformed ears' described in the external autopsy examination. Calcific nephrolithiasis accounts for 15% of the kidney stones in gout and may have contributed to Beethoven's nephrolithiasis.[48] Some authors have speculated that Beethoven may have taken salicylates for pain,[4] which would have aggravated his kidney condition because of their known interference with the excretion of uric acid. When PDB affects bones close to joints it can mimic arthritis and, over time, can cause arthritis when enlarged and misshapen bones put extra stress on adjacent joints. If pseudogout played any role in his joint pains, it would have been exacerbated by dehydration caused by attacks of pancreatitis.

The complexities of his medical condition caused difficulties in diagnosis, treatment and prognostication, which help explain the contempt that Beethoven had towards most of his attending physicians.[2] The prolonged episode of photophobia with decreased vision suffered by Beethoven several years before his death may very well have been the 'hot eye' of gout although this is not as common as once believed.[49] Since Beethoven had been prescribed quinine for analgesia,[50] and since we know he liked to self-administer large doses of his medications, quinine toxicity, with consequent mydriasis and retinopathic changes might also explain these symptoms.[51,52] Chronic pancreatitis may lead to decreased absorption of lipid-soluble vitamins including vitamin A, and hypovitaminosis A can cause eye pain with decreased vision.[53,54]

The intermittent episodes of abdominal pain and diarrhoea alternating with constipation may also have been manifestations of coeliac disease, which has a prevalence of approximately 1% in the general population. This would be an ironic connection since Beethoven was of Dutch origin and the discovery of the link to wheat bread was first made and confirmed by the Dutch paediatrician Willem-Karel Dicke in the 1940s while treating children on the Coeliac Ward in his hospital.[55] These gastrointestinal symptoms could also have been explained by irritable bowel syndrome – the commonest gastrointestinal disease in current clinical practice[56] – and some later episodes were likely caused by recurrent attacks of pancreatitis. He suffered from intermittent episodes of abdominal pain since his teenage years[57] but inflammatory bowel disease is unlikely for reasons noted earlier. Plumbism has also been invoked as an aetiologic agent for some of his symptoms,[21,58] and one study of Beethoven's hair indeed showed elevated levels during his last 110 days,[59] possibly related to lead-laced poultices applied after repeated paracenteses by his physicians during his final days. However, reliable studies of cranial bone fragments by Dr Andrew C Todd, a lead-poisoning expert at Mount Sinai School of Medicine in New York, did not show elevation above the low-level norm for that period.[60] Even if lead poisoning cannot be dismissed entirely as a contributory cause for some of his symptoms, including abdominal pain and behavioural disorders, it is unlikely that poisoning significant enough to cause deafness that started before he was 30 years old would not have other serious neurologic manifestations. An additional significant negative sign was the lack of a description of a typical black 'lead line' in the gingival tissues at autopsy; this sign indicates heavy metal intoxication and is not seen in edentulous individuals – and Beethoven had teeth. Renal colic and peptic ulcer disease must also enter into the differential diagnosis of abdominal pain. Since hypercalcaemia by itself is known to cause polyuria, dehydration, anorexia, nausea, vomiting, abdominal pain, and constipation, it may also have contributed to these symptoms. Either hypercalcaemia or quinine ingestion could have caused his frequent cardiac arrhythmias, and some authors believe that the rhythm of the Piano Sonata in E-flat major, Opus 81a, and other compositions may have been set to Beethoven's irregular heartbeat.[61–63] Although primary hyperparathyroidism rarely causes cardiac arrhythmias,[64] it has been associated with the tachy-brady syndrome in cases of severe hypercalcaemia.[65]

Syphilis, one of Sir William Osler's 'great mimics', has been asserted as a cause of at least some of Beethoven's problems, with support given by finding mercury – often used as a treatment for syphilis – in post mortem hair samples.[66] However, there is no definitive autopsy or clinical evidence of syphilis, and mercury was a constituent of multiple drugs used for other diseases at that time.

If one assumes that Beethoven abused alcohol as well as quinine and possibly salicylates in his attempts to alleviate the headaches and joint pains of Paget's disease, and if hyperparathyroidism and gout are not just associated but secondary (in some yet unknown manner) with Paget's disease, then it is possible to attribute all or most of his signs, symptoms, and social problems to Paget's disease.

The general problem with medical histories of historical figures is that they must be speculative to a certain degree, and this analysis is no different. Mark Twain noted 'There is something fascinating about science. One gets such wholesome returns of conjectures out of such trifling investment of fact'. [67] This type of analysis places even more importance on the accuracy of the facts used for speculation, and one should always be able to recognize speculation from fact. In the case of Beethoven, the original autopsy provides the best source of information, and this paper analyses each autopsy observation and attempts faithfully to correlate each one back with documented clinical signs and symptoms. The Viennese pathologists at the time were good observers but only described positive findings without mentioning significant negatives – a common practice then. Beethoven was a legend in his own time, so naturally his autopsy was performed by a recognized master anatomist, Dr Johannes Wagner, of the University of Vienna. It would be the first of approximately 60,000 autopsies for his assistant, Karl von Rokitansky, who would eventually inherit Dr Wagner's professorship and become known as one of the best descriptive anatomic pathologists in history.

Beethoven's music has been divided into three periods corresponding roughly to his changing states of

health, so it is probable his work would have been quite different if he had not been plagued by the deafness and pain caused by Paget's disease. Although he may have had an associated primary mental ailment, Beethoven would probably not be unlike many people who suffer behavioural changes because of their physical ailments, although outsiders attribute their behaviour to personality problems or antisocial tics. Any illness with sufficient severity will always have both psychological and physical dimensions. Beethoven suffered much during the last half of his life but it is possible, 188 years after his death and 201 years after the birth of Sir James Paget, to fulfil Beethoven's desire to give the world 'a description of my illness, and the story of my malady'.[6(p. 87)]

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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Journal of Medical Biography
2017, Vol. 25(3) 147–152

DOI: 10.1177/0967772015608052
journals.sagepub.com/home/jmb
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Charles Bernard Puestow (1902–1973): American surgeon and commander of the 27th Evacuation Hospital during the Second World War

Anand N Bosmia[1] and John D Christein[2]

Abstract

Dr. Charles Bernard Puestow (1902–1973) was an American surgeon who is well known for developing the longitudinal pancreaticojejunostomy, which is known as the "Puestow procedure" in his honor. Puestow served in the American military during the Second World War and commanded the 27th Evacuation Hospital, which provided medical and surgical services to wounded individuals in Europe and North Africa. In 1946, he founded the surgical residency training program at the Hines Veterans Hospital, which was the first such program in the United States based at a veterans hospital.

Keywords

Charles Puestow, surgery, pancreas, Second World War

Introduction

Dr. Charles Bernard Puestow (1902–1973) and his colleague Dr. William J. Gillesby (1905–1989)[1] introduced the longitudinal pancreaticojejunostomy in 1958[2] for the management of ductal obstruction and dilation in the setting of chronic pancreatitis.[3] This procedure is known today as the "Puestow procedure"[4] or "Puestow-Gillesby pancreaticojejunostomy".[2] It consists of a longitudinal incision of the pancreatic duct and implantation of the tail of the pancreas into a

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