Classical notes: Beethoven’s medical history. Variations on a rheumatological theme

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Introduction
Beethoven’s medical history is available from contemporary sources1-3, since when it has been comprehensively reviewed and reinterpreted4,5. Larkin has recently amended his invaluable account of 19706 and in the light of increasing knowledge of rheumatology and immunology has written in support of the growing impression that many of Beethoven’s disorders can be brought together under the umbrella of a ‘rheumatic disease’.

A further appraisal is now offered from the viewpoint of the practising rheumatologist. Beethoven’s clinical history is reconsidered leading to a differential diagnosis with particular emphasis on the likely rheumatic diseases. In addition, a single explanation for all the multi-system aspects, including the deafness, is suggested. Such a diagnosis has not previously been mentioned in any medical literature on Beethoven.

Prelude
Ludwig van Beethoven was baptised in Bonn on 17 December 1770, suggesting that he was born the day before. He died in Vienna on 26 March 1827. Beethoven’s father Johann was, by all accounts, an alcoholic tyrant. Beethoven’s mother, Maria Magdalena was a meek, gentle woman adored by Beethoven; her death from tuberculosis on 17 July 1787 affected him profoundly.

There were six other children of the marriage of whom only two boys survived. The older of these, Casper Carl, was baptised on 8 April 1774; he died from tuberculosis on 15 November 1815 leaving his own son, Karl, to be brought up by his widow whom Beethoven considered immoral, inadequate and unsuited to the task. His quixotic struggle to secure guardianship over Karl is well known.

The younger of his brothers, Nikolaus Johann, was baptised on 2 October 1776 and outlived Beethoven.

The family deaths from tuberculosis were by no means exceptional for the times. The only other family history of note was that of alcoholism. Beethoven’s father and paternal grandmother were both tipplers.

Bonn remained the family home until November 1792 when the Beethovens moved to Vienna, primarily for young Ludwig to receive lessons from Haydn. Father, Johann, died a month later on 18 December.

Traditionally, Beethoven’s compositions are grouped into three periods representing his achievements to 1802, those from 1802 to 1812 and from 1812 to 1827.

From his medical history it will be appreciated that 1802 was a pivotal year. This was followed by the decade of Beethoven’s second period, medically one of relative calm when his musical output was prodigious.

The final period was one of increasing ill-health. Rheumatic symptoms became prominent and his hearing deteriorated to the point of paradox when Beethoven’s outside world was silent yet the sounds in his mind’s ear must at times have been overwhelming.

As Beethoven’s decline accelerated during the last year or so his music became more innovative and profound.

Arguably his greatest masterpieces are the five late string quartets including the Grosse Fuge which Stravinsky, more than a century later, acknowledged as being eternally contemporary. A conspectus of his medical history is presented first. The rheumatic diseases are subsequently considered in more detail followed by a discussion of the differential diagnosis with arguments forwarded for the more likely contenders. A case is then made for a newly suggested diagnosis to explain all Beethoven’s medical problems including the deafness.

Exposition
It is likely that Beethoven suffered smallpox in childhood. This was a common affliction from which he bore the facial pock marks. The first documented illness occurred in 1787 when, coinciding with the death of his mother, he suffered a chest complaint associated with fever and depression. In a letter to Wegeler of 18017 he states that he had been suffering from an abdominal complaint which dated from before the family had moved to Vienna a year earlier. This consisted of diarrhoea, debility and ‘dreadful attacks of colic’, a complaint which plagued him recurrently throughout life. Deafness is first mentioned also in 1801 in letters to Wegeler and Amend8.

Detailed analysis of the causes of his deafness will not be attempted here. However, it is appropriate to mention the profound depression in which his early hearing loss resulted. In 1802 while spending the summer at Heiligenstadt Beethoven wrote his famous ‘Testament’, which took the form of a letter to his brothers, never dispatched, and only discovered posthumously. The significance of the Heiligenstadt Testament, how superficially it related to Beethoven’s despair over his increasing deafness and its deeper psychological implications have been thoroughly debated.

1804 saw a severe fever; an abscess which almost caused the loss of a finger was reported in 1807 and another abscess, this time in the jaw, suffered a year later.

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In May 1810, he wrote again to Wegeler admitting his mounting depression from the increasing deafness; then in 1811 Beethoven visited the Bohemian spa at Teplitz to take the waters.

Between 1813 and 1825 he suffered episodes of bronchitis, rheumatism, anxiety and bouts of prostration from his intestinal trouble and colic. By 1817 Beethoven was virtually stone deaf. In 1821 jaundice developed. Mid-1822, Pancreas: Large arteries: Auditory A few or the afternoon of Rites. Beethoven returned (Table March, peaceful by February including 1. peas: the following day severe, February was Physically devastated Braunhofer. Beethoven performed, we have become Anton Braunhofer. Beethoven was at the time, when he reports haemoptysis and spontaneous epistaxis.

A letter to Braunhofer in 1826 mentions back pain 'not severe, I admit, but it shows the trouble is still there'. A few days later he begs the same doctor not to visit, 'for I have been plagued for some time with rheumatism or gout'.

After supervising the convalescence of his nephew Karl, who, in July 1826 had attempted suicide, Beethoven returned on 1 December to Vienna from his brother Nikolaus's estate in Gneixendorf. On reaching home Beethoven was extremely ill and on 5 December dropsy was diagnosed. Eight days later the history is that of gram negative septicemia with abdominal pains, rigors, oliguria and significant increases in oedema, ascites and jaundice. Hepatomegaly was elicited by Dr Wawruch with the description of the liver as like 'hard knots'. On 20 December 1826, 11 litres were allegedly drained at abdominal paracentesis. Similar large volumes were removed on 9 January, 2 February and 27 February 1827.

Surrounded by squalor and chaos, detached from the world by his deafness Beethoven was apparently relatively calm, writing and receiving visitors, including Schubert. Physically devastated with limbs leaden, his mind remained, for a while, relatively mercurial since we know that he was at work on what might have become his 10th symphony. On 24 March, peaceful and lucid, he received the Last Rights. Beethoven lapsed into coma and died late in the afternoon of 26 March 1827.

The following day a postmortem examination (Table 1) was performed, supervised by Professor Wagner. He was accompanied by Dr Rokitansky who was to become the 'father of modern morbid anatomy' and this was the first of the 59 786 autopsies with which he is credited during his professional life.

### Development

A list of the main features of Beethoven's medical history is shown in Table 2. The likely terminal event was sepsis with liver failure and portal hypertension from a macronodular cirrhosis. The finding of a shrunken liver after death contrasts with the hepatomegaly observed by Dr Wawruch 3 months earlier. The significance of the renal calculi and thickening at the base of the skull are considered later. There were no other bony findings in the skull to explain the nerve deafness. It is uncertain how thoroughly the ossicles were examined.

Apart from a septic foot in 1813, the first of many references to rheumatic complaints occurs in a letter of 18 December 1816 to Charles Neate, pianist, composer and a founder member of the Philharmonic Society in London. The relevant rheumatic symptoms are shown in Table 3. The question to be addressed is whether the problems listed in Table 3 can be reconciled with those of Table 2 to produce an acceptable diagnosis of one of the multisystem rheumatic diseases.

Larkin suggests the whole picture is compatible with a 'connective tissue disease' or 'immunopathy' and singles out systemic lupus erythematosus (SLE) as being most likely. Bearing in mind that SLE has probably replaced syphilis as the disease most prevalent in its manifestations, superficially there are many attractive reasons to support this hypothesis. The 'thoracic gout' for example alludes to associated chest symptoms:

### Table 2. Summary of Beethoven's medical history

| Probable smallpox in childhood |
| Contact with tuberculosis - from which mother and at least one brother died |
| Deafness |
| Recurrent bouts of depression |
| Recurrent infections |
| Chest symptoms and 'asthma' |
| Repeated episodes of 'kolik', ie diarrhoea and abdominal pain, sometimes debilitating |
| Attacks of rheumatism |
| Jaundice - first appearing at 50 |
| Eye pains - at 53 |
| Dropsey - peripheral oedema and ascites |
| Hepatomegaly |
| Death 1827 |

### Table 1. Autopsy findings

| Body 5 feet 6 inches. Wasted. Covered in petechiae |
| No significant abnormalities in the bony parts of his ears |
| Auditory nerves: shrivelled and narrowless |
| Auditory arteries: Dilated to 'more than a crow's quill' and cartilaginous |
| Facial nerves: considerably thickened |
| Skull vault: uniformly dense, ⅛ inch thick |
| Liver: Shrunken half volume. Leathery, greyish-blue, tuberculated surface. Cut surface - bean sized nodules |
| Spleen: Twice normal size, black and tough |
| Pancreas: Large and indurated with a dilated duct |
| Kidneys: Pale with chalky calyceal concretions size of split peas: the whole enclosed in cellular membrane, one inch thick and dripping turbid fluid |
| Bowel: Distended with gas |

### Table 3. Beethoven's main rheumatic symptoms

| 1816 | December: 'Rheumatic feverish cold' |
| 1817 | Autumn: 'Fearful attack of rheumatism' |
| 1820 | Bad year for health culminating in rheumatic fever. 6 weeks in bed |
| 1822 | 'Gout on my chest' ('thoracic gout') |
| 1823 | April-September: Sore eyes(s) |
|  | Eyes bandaged at night |
|  | Violent diarrhoea: 'Abdomen and eyes wretched' |
|  | 5 September: Baden taking waters and drinking mineral waters |
| 1826 | February: 'Plagued long with rheumatism or gout' |
|  | February: 'Pain in my back not severe but still there' |
and joint disease, a feature of SLE in about 50% of cases. Similarly, the commonest presenting complaint of SLE is joint pain, of which Beethoven had more than his fair share. However, the case for SLE looks weaker when other aspects of his rheumatic history are scrutinised.

To start with, SLE is rare in males. When developing after childhood and before the age of about 40 it is up to 9 : 1 in favour of women. Larkin proposes that the facial scars present from adolescence were from lupus and suggests further that the fever Beethoven suffered aged 16 signalled the onset of SLE. It is true that in children and adolescents the commonest manifestation is skin disease; set against this, however, is the lower incidence of joint and patients, to fulminating disease. (Beethoven had further examples in favour of SLE by Larkin) and the greater risk of renal disease with its poor prognosis. The evidence is, on balance, against juvenile onset SLE in Beethoven. Smallpox was a common disease and much more likely to have caused Beethoven’s facial scarring. Before leaving his early afflictions it is worth recording that there is no reference to deafness occurring in SLE in Dubois standard work.8

Many of the common characteristics of SLE such as lymphadenopathy, alopecia, pericarditis or renal disease were inconspicuous in Beethoven. The clotting abnormalities which resulted in Beethoven’s spontaneous haemorrhages towards the end were surely from the cirrhosis plus portal hypertension leading to hypersplenism, rather than the thrombocytopenia associated with SLE alluded to in Larkin’s chapter.

A consideration of the most serious of all Beethoven’s illnesses and that which killed him, his liver disease, provides the greatest objection to the diagnosis of SLE. Working from the premise that cirrhosis was present with hepatitis as the likely cause, the point at issue is whether these might be related to SLE. The condition ‘lupoid hepatitis’ encourages this possibility, but is misleading.

Lupoid hepatitis refers to a chronic active hepatitis (CAH) without a viral aetiology in which antibodies to smooth muscle are detectable. A few patients have antinuclear antibodies and a smaller number possess high titres of antibodies to double stranded DNA. Nonetheless, it is very uncommon indeed for these patients, to fulfill the American Rheumatism Association’s criteria for SLE. CAH and SLE co-exist so rarely as to warrant publication of individual case reports. It is generally accepted that ‘lupoid hepatitis’ is unrelated to SLE and that significant liver involvement in SLE is rarely of clinical importance. Jaundice and ascites occur in about 4% and 11% of cases respectively.

If Beethoven’s fatal liver disease cannot be blamed on SLE, other possibilities must now be entertained. CAH itself results in a macronodular cirrhosis which if untreated can lead to end stage liver disease. It is also associated with arthralgia, ulcerative colitis and pulmonary disease. However, CAH is three times more common in women and many of Beethoven’s other complaints appeared long before his jaundice. These factors, while not excluding CAH, weigh against such a diagnosis, which also fails to explain other of Beethoven’s problems and a later discussion on his gut disease contributes to such doubt. A macronodular cirrhosis similarly results from cryptogenic/idiopathic disease, that unsatisfactory label attached to a diminishing group of patients in whom no specific cause can be identified, but this too fails to explain several other of Beethoven’s complaints.

Finally, alcoholic cirrhosis has been propounded yet this seems unlikely. It results in a micronodular cirrhosis and whereas Beethoven enjoyed a drink, there is no evidence in his personal history that he abused or consumed alcohol to a degree that predisposed him to cirrhosis. On the contrary, there are indications that Beethoven leaned towards asceticism.

Having dismissed SLE (and similar objections are levelled at the other connective tissue diseases) as a likely explanation for Beethoven’s joint disease and looking beyond the arthralgia of CAH three other rheumatic diseases merit closer attention. The first, an arthropathy associated with inflammatory bowel disease; the second a reactive, possibly post-dysenteric arthritis progressing to chronic spondylarthropathy. The third, Paget’s disease of bone, has also been forwarded as a likely explanation for his deafness10,11 and the evidence for this is analysed.

Beethoven’s long history of abdominal complaints hint at an enteropathic arthropathy, if that is, it can be accepted he had inflammatory bowel disease. In addition, he had painful eyes and hepatitis, both recognized as extra-intestinal complications. Certainly, as Beethoven’s letters indicate his joint and gut symptoms paralleled one another, as is often the case in enteropathic arthropathy. There is however scant evidence to support a diagnosis of inflammatory bowel disease. The history of his intestinal complaints was long with attacks of pain often severe, in which case at postmortem more florid findings should have been present. With Crohn’s disease, complications would certainly have been found and at autopsy there were no references to adhesions, perforations or strictures.

Had ulcerative colitis been present then rectal bleeding, an almost invariable accompaniment should have been a dominant symptom. There is considerable room for doubt about its presence in Beethoven. In his final illness he was questioned by Dr Wawruch about the presence of ‘haemorrhoidalleiden’. The tone of his conversation books (tantalisingly recording only the questions and comments of those with whom he discoursed) suggest that Beethoven answered in the affirmative. Haemorrhoidalleiden implies rectal bleeding but literally means bleeding from any orifice. Spontaneous haemorrhage from nose and bronchi were present in Beethoven and it would not have been surprising, in view of the obvious clotting deficiencies, if he had noticed blood per rectum in his terminal illness. However, rectal bleeding is such a striking and worrying symptom to most patients that its absence earlier in Beethoven’s life can probably be accepted.

A more likely explanation for his colic is irritable bowel syndrome (IBS), which does not explain Beethoven’s joint problems. There is, nevertheless, an alternative explanation for the relationship between his gut and rheumatic symptoms. Once again, this takes into account the eye pains but now includes his backache. Dysentery was likely, conditions of personal hygiene or public health were often poor. Enteritis was a common problem and there were particular times when this would have been a greater risk. It should be remembered that Vienna was under siege by Napoleon’s army while Beethoven was resident
and water supplies in cities under siege are notoriously vulnerable. In addition, we know that he drank the murky waters of several spas, surely suitably contaminated with enteropathic organisms.

His relapsing joint symptoms do indicate a seronegative 'reactive arthritis'. Further, the description of his eye symptoms raises the possibility of uveitis and again this coincided with a bad bout of his diarrhoea in 1823, while backache, mentioned in 1826 had obviously been present for some time. These points all favour a post-dysenteric reactive arthritis possibly progressing to a chronic spondylarthropathy with sacroilitis or full-blown ankylosing spondylitis. What an intriguing prospect, could it be that Beethoven was HLA B27 positive and the first and most famous recorded case of HLA B27 associated rheumatic disease?

The third rheumatological disorder, Paget's disease of bone, has been proposed as a likely explanation for Beethoven's deafness. The evidence presented in defence stems from three sources. First, a sketch by J P T Lyser of 1833 (Figure 1) purports to show the stocky figure of Beethoven with an ill fitting hat atop an outsized Pagetic head. This is unconvincing, the figure depicted is surely a caricature on to which too much has been projected. Another sketch by Lyser and that of Böhm of 1820 (Figure 2) rather more convincingly show that it was Beethoven's mop of unruly hair which was responsible for his hat appearing undersized.

Secondly, a photograph by Rottmayer of Beethoven's reconstructed skull, taken after the first exhumation of 1862 was supposed to show a swelling over the right temporo-parietal region and Paget's disease suggested as the likely cause. However, other contemporary sketches, both then and following the second exhumation of 1888, fail to show this feature. It seems likely, therefore, that the deformity was an artefact.

The case for Paget's disease so far rests on the possible morphology of Beethoven's skull. The most accurate facsimile is considered to be that of Klein's bust of Beethoven (Figure 3). Taken from a life mask in 1812 when deafness was well established and severe, this does not demonstrate any convincing
signs of advanced Paget’s disease. Since Beethoven was first aware of his deafness in his late twenties, 15 years or so before Klein’s work, it appears highly improbable that Paget’s was so well established at that early stage as to cause eighth nerve compression.

Cadenza
In every clinical problem with multi-system aspects, however seemingly obscure, variable or unrelated, an attempt must be made to apply Occam’s Razor. It is possible that no such unifying diagnosis exists, but before abandoning all hope, it is necessary to consider two chronic granulomatous diseases.

First, tuberculosis (TB) - its presence in Beethoven has previously been suggested by several authors11-12. Certainly Beethoven suffered chest complaints and fever. Nothing from when he was 16, however, appears to indicate that TB was a cause of his deafness. Nevertheless, although TB affects many systems it is unlikely that intra-abdominal disease would have produced symptoms for 35 years, since diarrhoea and abdominal pain from intestinal involvement or associated peritonitis would have resulted in rapid decline. Tuberculous granulomatous hepatitis, in conjunction with eye, bone, abdominal and renal disease with calcification, indicates tuberculous involvement. Tuberculosis cannot, therefore, be accepted as the single explanation for Beethoven’s problems.

A second chronic disease, however, does have great possibilities. Such is sarcoidosis, which can present with all the following features: skin involvement13; eighth nerve involvement14; hepatic involvement15; portal hypertension16; splenic involvement17; arthropathy, arthralgia18; uveitis19; chest symptoms20; hypercalcaemia21; ureteric colic22. Of the granulomatous conditions sarcoidosis is much more likely to run a chronic, intermittent, progressive course. Germane to Beethoven’s case is the continuing controversy over the relationship between the development of sarcoidosis in those exposed to tuberculosis. At least two close members of Beethoven’s household died from tuberculosis and an hypothesis exists that individuals in contact with mycobacterium tuberculosis respond in an immunologically inappropriate manner resulting in the non-caseating granuloma of sarcoid23. Many aspects of Beethoven’s medical history draw one to sarcoidosis.

Consider again the most significant component to Beethoven’s final illness, his liver disease. The macronodular cirrhosis described (although not in these modern terms) would do well for granulomatous sarcoidosis, with the splenic enlargement relating either to associated portal hypertension or to direct sarcoid infiltration, both well described.

Remaining in the abdomen, now reflect on Beethoven’s colic, the abdominal pain which often laid him low. Sarcoidosis of the gut is very rarely symptomatic although on occasions the histology is confused with that of Crohn’s disease. A case was made earlier against inflammatory bowel disease and irritable bowel syndrome suggested. However, the abdominal pain was frequently of a severity to prostrate him and yet another possibility exists. The autopsy report records renal calculi. Sarcoidosis results in renal stone formation from hypercalcaemia and hypercalciuria. Ergo, might Beethoven’s most evil abdominal pains have resulted from renal and ureteric colic against a background of irritable bowel and dysentery? Chest symptoms although common in sarcoidosis are variable in severity. Joint disease usually represents a non-deforming acute or chronic arthropathy or is related to changes in adjacent bone. Very occasionally a chronic granulomatous synovitis presents. Uveitis is well recognized; often it is associated with facial nerve involvement, not infrequently bilateral and usually lower motor neurone in type. The finding of facial nerve thickening in Beethoven is puzzling and contrasts with the atrophied eighth nerve since in sarcoidosis both can be affected together. There are no reports in his letters or notebooks to suggest Beethoven suffered facial palsy and it is not clear which parts of the seventh nerves were examined at autopsy. Presumably the thickening relates to their emergence from the brainstem in which case granulomatous infiltration is a likely explanation.

A further contention is that Beethoven’s facial scars were from lupus pernio. Lupus pernio is well accepted as occurring with chronic widespread sarcoidosis and may well be the presenting feature. The probability that Beethoven suffered smallpox in childhood need not preclude skin infiltration from sarcoidosis developing subsequently. It is established that existing scars can later be affected by sarcoidosis in a variety of ways, from lupus pernio to nodular granulomatous infiltration. Interestingly when sarcoidosis does infiltrate pre-existing scars the incidence is higher in men, that opposite to when unscarred skin is involved.

Finally, what about the deafness? Much has been written about Beethoven’s deafness by a distinguished list of contributors10,11,14,24-25 and superficial speculation on the subject here must therefore be forgiven.

Eighth nerve involvement is well recognized in sarcoidosis, sometimes presenting so early that the diagnosis is overlooked because many other features of the disease have not yet evolved. When deafness does result it has been found in association with widespread disease including uveitis and hepatomegaly22.

Could it be that sarcoid affected Beethoven’s vestibuloneural function early on, with more widespread disease smouldering for many years before becoming an overwhelming blight?

Recapitulation
Based on the foregoing arguments the following differential diagnosis is offered.  
1 Sarcoidosis, as the possible single diagnosis encompassing all aspects  
2 Chronic active hepatitis with resultant cirrhosis and multi-system aspects  
3 Cryptogenic/diopathic cirrhosis (and 2 leading to portal hypertension)  
4 Seronegative arthropathy  
(a) Post dysenteric ‘reactive’ arthritis  
(b) Sacroiliitis/ankylosing spondylitis from (a)  
(c) Associated with inflammatory bowel disease  
5 Irritable bowel syndrome  
6 Incidental Paget’s disease  
7 Deafness of undetermined type. See specialized literature
Coda
An academic pastime is to contemplate how Beethoven's prognosis might have been improved had today's knowledge of disease, investigation techniques and range of treatments been available at the turn of the 19th century.

On reviewing the differential diagnosis it is striking to appreciate just how valuable our routine, everyday laboratory and radiological services would have been to Beethoven's doctors and recalling their limited therapeutic tools, what they would have given for non-steroidal anti-inflammatory drugs, steroids and immunosuppressants. With the 'high-tech' end of modern treatment in mind, had all else failed to control Beethoven's liver disease he surely would have been considered a likely recipient for a liver transplant.

Yet would all this, if feasible, have been in the best interests of posterity or Beethoven's genius? The history of art in all its forms is strewn with those in whom anguish, disadvantage, despondency and suffering catalysed or enhanced their creative abilities. Many of Beethoven's works, particularly during his last period, can be considered in this light. He, himself, acknowledged such in one of the most famous examples - the slow movement of Op 132 No 15 string quartet - Beethoven's transcendent 'Holy Song of Thanksgiving, in the Lydian mode, of one recovered from an illness'.

Possibly his disappointments and struggles against the curse of ill health resulted in the darker side of his nature: yet maybe those same adversities provoked and released emotions which were translated into the apocalyptic, sublime music left to us. Callous though it might appear, perhaps Beethoven's increasing wretchedness and misery were ultimately to the benefit of mankind.

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