



# Primary Sclerosing Cholangitis may have caused the death of Beethoven

In a recently published article by two American otologists it is claimed that the different physical symptoms of Beethoven's could be explained by an immunopathy associated with inflammatory bowel disease (IBD) [1]. His deafness may have been caused by a sensorineural hearing loss, and the liver failure Beethoven died of in 1827 may have been caused by Primary Sclerosing Cholangitis (PSC). This article did not however provide any strong arguments that Beethoven actually suffered from PSC. In 2002 Keynes and colleagues wrote that Beethoven may have suffered from PSC, but dismissed this as most unlikely [2].

A good deal of the medical focus on Beethoven has been aimed at his deafness, since it's an extraordinary mystery that one of the musical history's most eminent composers created several of his immortal works without being able to hear a single tone. Beethoven did however have multiple other physical symptoms. The diagnostical grounds which can be found today are above all the letters and "conversation notes" that Beethoven wrote, of which some are preserved,

describing his symptoms. Beethoven was also examined by several of Austria's and Germany's most prominent doctors. What's missing, essentially, are objective findings of his status. Mostly there are diagnostical accounts such as "typhoid fever", "rheumatic fever" and "thorax gout" – all being diagnoses having a totally different meaning in Beethoven's days than today.

We also have access to the results from the three autopsies carried out on Beethoven. The diagnostic "arsenal" was of course quite sparse, which has given room for countless different hypotheses about Beethoven's possible medical diagnoses. Many authors have tried to gather the different symptoms under one medical entity and others yet have chosen to describe each symptom by itself. The medical entities stated has been different diagnoses such as sarcoidosis, SLE, syphilis, alcoholism, tuberculosis, Paget's disease, and lead poisoning [3-8]. Neither of these conditions however explains why Beethoven had so many diarrhoeas with abdominal pain, fever and macronodular cirrhosis.

**LUDWIG VAN BEETHOVEN**, asleep by the piano, exhausted after strenuous composing. He's dreaming of allegoric shapes.  
Photo by IBL Bildbyrå

**B**eethoven was born in Bonn in 1770. There's no data supporting a presence of gastrointestinal disease in his family. It seems like several of Beethoven's physical and psychological symptoms appeared after some traumatic events in his youth. His younger sister died from tuberculosis in 1787. The same year Beethoven, seventeen years of age, applied for and was granted to be the head of family and responsible for his younger siblings since the father was an alcoholic. In letters to his friend and moreover his doctor, Dr. Frantz Wegeler, Beethoven described, at the age of twenty, episodic problems with diarrhoeas and colic reminding spasms and pains centrally in the abdomen, which had been present for several years [9]. When the hearing problems later occurred he related them to his bad stomach. In the year of 1822 Beethoven said to his colleague Louis Schlösser that his hearing problems were originated from his stomach and also that he felt mistreated by his different doctors [10].

In the beginning of the 19th century Beethoven had many inexplicable fever episodes which were interpreted as rheumatism; later on abscesses also occurred - in one finger and also in his jaw. He had trouble with his joints which sometimes was interpreted as gaut, sometimes as rheumatism. The trouble with the joints often appeared when the intestines were worse. Beethoven also suffered from bronchitis and something what was called "thorax gout", which could be interpreted as chestpain. After being offered various treatments for his abdominal problems he was recommended hot baths in Danabue (Danube), which worked miraculously well and became something he often used as treatment [11]. In the spring of 1821 he got troubles with his joints which once again were interpreted as rheumatism. Shortly after this he became jaundiced, which lasted during the summer and decreased gradually. During 1823 to 1824 he got pain in his eyes and became sensitive to light, which was treated with bandage as well as resting in a dark room. These problems have been interpreted as uveitis.

In 1825, at 55 years of age, Beethoven got intense abdominal pain and yet again became jaundiced, which decreased gradually after a period of several weeks. During this period of illness changes in the skin, which resembled pyoderma gangrenosum, was described. One year later, following upon an infection Beethoven caught during a journey, his abdomen swelled up due to ascites. Peripheral oedema and jaundice occurred and he finally died from liver failure. To make the abdominal and perspiration problems somewhat easier to live with he got his abdomen drained at four different occasions which led to an infection. Until shortly before his death Beethoven was still mentally active and produced new compositions. He then started to write something that later on has been interpreted as the beginning of the 10th symphony. One day after his death an autopsy was made by Professor Wagner and Dr. Rokitansky. In the autopsy report they described a man covered



### LUDWIG VAN BEETHOVEN,

1770 – 1827, created several of the musical history's masterpieces without hearing a single tone. Besides deafness he suffered from various other complaints, such as smallpox, bronchitis, depression-melancholy, colic, diarrhoea, recurring infections, problems with joints, aching eyes sensitive of light, tinnitus, spontaneous bleeding, jaundice, back pain and "thorax gout".

Photo by IBL Bildbyrå



by petechiae, 9 liters of infected ascites, and a blue green grey leather looking liver being half the normal size and shattered by noduli. His spleen was twice the normal size, the intestines were ballooned with gas, but no strictures or adhesions in the intestines were described. The pancreas was solid and abnormal, and the excretory passage was described as wide as a pen from a goose, which has been interpreted as chronic pancreatitis. The gallbladder contained dark brown fluid and sediments and/or stones.

**W**hat then, has been the earlier explanation to Beethoven's periodic proceeding liver disease which later took his life? There are guesses that Beethoven, depending on his taste for Oysters, got viral hepatitis which should have caused "post-hepatitis cirrhosis". This is however unlikely since the hepatitis spread via Oysters is hepatitis A, which does not cause any chronic hepatitis. Others have guessed that he in 1821, when the first icteric episode occurred, got an acute viral hepatitis and that this later caused cirrhosis with liver failure in 1825. This also seems less likely since the time elapsing from acute hepatitis to serious liver failure seldom is as short as four years. It mostly takes 10 to 30 years for liver cirrhosis to be developed from an acute hepatitis B.

As with many other people with liver diseases he was also accused with self inflicted liver failure due to overconsumption of alcohol, and the liver failure that caused his death could have been alcoholic related [12, 13]. There has been, for a long period of time, a debate whether Beethoven overconsumed alcoholic beverages or not. His childhood friend and doctor Wegeler claimed that Beethoven drank modest amounts. Anton Schindler, another good friend of Beethoven's from the later years, told that alcohol affected Beethoven's abdomen in an unfavourable way, but that he despite this liked a glas of good Hungarian vine and some glas of beer in the evening – quantities that wouldn't

cause liver failure [14]. Schindler also pointed out that Beethoven's favorite beverage was a glass of fresh water. Dr. Wawruch, one of the doctors who treated Beethoven during the last years, claimed however that Beethoven during his last years developed an interest for consumption of alcohol, which still was regarded as modest [15]. This statement indicates that the interest for alcohol was something that came during the last years and consequently could not have been of such an extent that it would have led to liver cirrhosis.

E Larkin wrote in 1970 that Beethoven's diseases could depend on an immune-mediated disease, from which he thought SLE was most plausible [16]. P J Davies and later A K Kubba suggested that the trouble with the intestines could be dependent on inflammatory bowel disease [12, 17]. Inflammatory bowel disease (IBD) consists of above all two different diseases: Crohns disease and Ulcerative Colitis. Since Beethoven had so much pain it has been suggested that it was Crohns disease he suffered from. The fact that there were no strictures or adhesions in the abdomen when the autopsy was performed would more likely support that Beethoven suffered from Ulcerative Colitis than Crohns disease. Ulcerative Colitis rarely gives abdominal pain, opposite Crohns disease, and thus the bowel disease can't explain the intense abdominal pain that tormented Beethoven during so many years.

In presence of IBD there is occurrence of several different extra intestinal complications which commonly are divided into activity and non activity related complaints. The most common activity related complaints are changes in the skin (such as pyoderma gangrenosum), problems with the joints and eye problems (iritis, uveitis). Of the non activity related complaints, liver diseases are the most common. Approximately 10 to 15 percent of all patients with IBD develop liver disease, the most common of these being PSC. There is also described an association between IBD and an early debut of sensorineural hearing loss.

**W**hat then is PSC and what symptoms does the disease give? The first time PSC was described was in Germany 1867 by Dr. Hoffman – but it was presented in the English literature not earlier than 1927. The disease was regarded as a medical rarity up to the 1970's. Since then the knowledge about the disease has grown considerably, and PSC is today one of the most common cholestatic liver diseases in the western world. There is no evidence that the disease has increased in incidence – rather that knowledge about the disease has grown and better diagnostic methods have made it easier to diagnose. The disease preferably affects men who have Ulcerative Colitis [18]. The age at onset of disease could vary from 2 to 80 years of age; the average is about 40 years of age. Most often the IBD appears first and the liver disease later on, but the other way around could also be the case. With PSC the bile ducts are affected by a fibro inflammatory process that causes stenosis. In the majority of the patients the intrahepatic as well as the extrahepatic bile ducts are affected, but one third only have



**ANDREAS WAWRUCH,**  
1773 - 1842, was  
Beethoven's last  
doctor.



**BEETHOVEN SICK IN HIS BED.**  
Pencil drawing by  
Josef Teltcher,  
1827.

engagement of the intrahepatic bile ducts. The cause of this disease is unknown, but very much supports that it is an immune-mediated disease [19]. The proceeding is varying. Some patients remain asymptomatic for decades while others develop symptoms such as abdominal pain, fever, itching and icterus. The abdominal pain can radiate up to the right shoulder and also to the back – which could explain the back pain Beethoven had and also what was described as “thorax gout”. The symptoms of PSC often come in short periods and could disappear by themselves without any known predisposing factors [20]. With most patients the disease progrediate to liver cirrhosis and increasing signs of liver failure in the shape of ascites, enlargement of the spleen, esofagusvarices, encephalopathy and icterus [21]. Unfortunately there is no possibility to cure the disease, and PSC is today the most common cause of liver transplant in the Nordic countries [22]. With PSC there is an increased occurrence of diseases such as Diabetes, thyroid disease, SLE and coeliac disease [23]. Furthermore 10 to 20 percent of the patients have affected pancreas [24]. Consequently it seems that Beethoven on basis of Ulcerative Colitis could have had a lot of activity related symptoms such as problems with the joints, skin disease and uveitis. Though one can not look away from the possibility that some of the problems with diarrhoea also could have been caused by bacterial infections, which in turn could have caused phases of Ulcerative Colitis. In addition he could have had a PSC which caused abdominal pain and fever episodes that came and went leading to two icterus episodes before the final liver failure began.

The enlarged spleen could have been due to portal hypertension, causing hypersplenism with thrombocytopenia leading to the troubles with bleeding Beethoven lived with during later years, as well as the petechiae which were present all over his body when he died. The episode with icterus in 1825 was preceded by more intense abdominal pain, which point towards a more pronounced stenosis, alternatively stone, in the large bile duct. When the autopsy was performed, stones or sediments were found in the gall bladder, and a stone could very possibly have transferred down in coledocus. The findings at the autopsy – macronodular cirrhosis with blue green grey colouring – also supports that the liver disease was of cholestatic type and not caused by alcohol. The widened pancreatic duct could speak for an obstruction of the outflow, but there is no description of the big bile duct that supports a stricturing process, alternatively a stone, there. Some of the diarrhoea and pain could surely be related to the chronic pancreatitis, but the periodic proceeding supports more that PSC and Ulcerative Colitis were the foremost reasons for these complaints.

**D**oes it anyhow matter what disease Beethoven suffered from? One thing that was evident with Beethoven was the feeling he had of being misunderstood by his doctors. He consulted the most prominent doctors of this era, but the relation to them is described as tense. He describes them as “bumbling doctors and

medical asses" [25]. Beethoven did also suffer from recurring periods of depression. There are letters preserved where he describes a depression over his physical symptoms as "...would like to have left this world a long time ago and even have helped my self doing so. This life is beautiful but for me it is for ever poisoned" [26]. Beethoven wrote as early as 1802 in a letter to his brothers that he had a strong wish for a professor Schmidt to describe his diseases so that the world should know what he suffered from [27]. This was surely one of the reasons why the autopsy was to be executed already on the day after his death. What is a great mystery is the fact that Beethoven, despite his physical as well as psychological complaints, still kept his creative ability and high working capacity until the day he died.

Maybe this was the best treatment for his PSC. Maybe some of the pain from a chronic periodic proceeding disease gives echo in his wonderful music. Though in Beethoven's music there is also very much light, and hopefully this music could bring comfort to some of the PSC patients that still today have to endure that many of their questions concerning cause, proceeding and therapy can not be answered – even if they unlike Beethoven have a name on their disease.

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#### Reference:

1. Karmody CS, Bachor ES. The deafness of Ludwig van Beethoven: an immunopathy. *Otol Neurotol.* 2005;26(4):809-14.
2. Keynes M. The personality, deafness, and bad health of Ludwig van Beethoven. *J Med Biogr.* 2002;10(1):46-57.
3. Larkin E. Beethoven's illness-a likely diagnosis. *Proc R Soc Med.* 1971;64(5):493-6.
4. Palferman TG. Beethoven: a medical biography. *J Med Biogr.* 1993;1(1):35-45.
5. Jacobsohn L. Beethovens Gehörleiden. *Deutsch Med Wschr* 1910;343:1282
6. Kubba AK, Young M. Ludwig van Beethoven: a medical biography. *Lancet.* 1996;347(8995):167-70.
7. Naiken VS. Paget's disease and Beethoven's deafness. *Clin Orthop Relat Res.* 1972;89:103-5.
8. Martin R. Beethoven's hair. New York: Broodway Books 2000
9. Wegeler FG, Ries F. Biographische Notizen über Ludwig van Beethoven. Auflage, Koblenz: Bädecker; 1838:23
10. Schlösser L. Erinnerungen an Ludwig van Beethoven (Teil 1). *Allgemeine Deutsche Musik-Zeitung* 1880;7:401-405
11. Thayer AW. *Life of Beethoven.* Princeton, NJ: Princeton University Press 1973.
12. Kubba AK, Young M. Ludwig van Beethoven: a medical biography. *Lancet.* 1996;347(8995):167-70.
13. Bjornrtorp P. [What did Beethoven die of?] *Lakartidningen.* 1998;95(20):2364-7.
14. Schindler A. I : Sonnek OG, ed. *Beethoven: Impressions by his Contemporaries.* New York: Dover Publications; 1967:168.
15. Wawruch AJ. Ärztlicher Rückblick auf die Todeskrankheit. I: Nohl, Ludwig, ed. *Beethoven nach den Schilderungen seiner Zeitgenossen.* Stuttgart: J.G. Cotta;1877:247-254
16. Larkin E. Beethoven's illness-a likely diagnosis. *Proc R Soc Med.* 1971;64(5):493-6.
17. Davies PJ. Was Beethoven's cirrhosis due to hemochromatosis? *Ren Fail.* 1995;17(1):77-86.
18. Broome U, Bergquist A. Primary sclerosing cholangitis, inflammatory bowel disease, and colon cancer. *Semin Liver Dis.* 2006;26(1):31-41.
19. Xu B, Broome U, Ericzon BG, Sumitran-Holgersson S. High frequency of autoantibodies in patients with primary sclerosing cholangitis that bind biliary epithelial cells and induce expression of CD44 and production of interleukin Gut. 2002;51(1):120-7.
20. Olsson R, Broome U, Danielsson A, Hagerstrand I, Jarnerot G, Loof L, Prytz H, Ryden BO. Spontaneous course of symptoms in primary sclerosing cholangitis: relationships with biochemical and histological features. *Hepatogastroenterology.* 1999;46(25):136-41.
21. Broome U, Olsson R, Loof L, Bodemar G, Hultcrantz R, Danielsson A, Prytz H, Sandberg-Gertzen H, Wallerstedt S, Lindberg G. Natural history and prognostic factors in 305 Swedish patients with primary sclerosing cholangitis. *Gut.* 1996 Apr;38(4):610-5.
22. Brandsaeter B, Friman S, Broome U, Isoniemi H, Olausson M, Backman L, Hansen B, Schrupf E, Oksanen A, Ericzon BG, Hockerstedt K, Makisalo H, Kirkegaard P, Bjoro K. Outcome following liver transplantation for primary sclerosing cholangitis in the Nordic countries. *Scand J Gastroenterol.* 2003;38(11):1176-83.
23. Saarinen S, Olerup O, Broome U. Increased frequency of autoimmune diseases in patients with primary sclerosing cholangitis. *Am J Gastroenterol.* 2000;95(11):3195-9.
24. Epstein O, Chapman RW, Lake-Bakaar G, Foo AY, Rosalki SB, Sherlock S. The pancreas in primary biliary cirrhosis and primary sclerosing cholangitis. *Gastroenterology.* 1982;83(6):1177-82.
25. London SJ. Beethoven: Case report of a titan's crisis. *Arch Intern Med.* 1964;113:442-8.
26. Huxtable RJ. The deafness of Beethoven: a paradigm of hearing problems. *Proc West Pharmacol Soc.* 2000;43:1-8.
27. Kastner E, Kapp J. *Ludwig van Beethovens sämtliche Briefe* (ed. 2). Leipzig, Becker Verlag, 1923.