

**Sensorineural Hearing Loss in Inflammatory Bowel Disease**Aman Madaik<sup>1</sup>, Tulika S. Madaik<sup>2</sup>, Anju Madan Gupt<sup>3</sup><sup>1</sup>M.O., Department of ENT Head and Neck Surgery, IGMC, Shimla;<sup>2</sup>Associate Professor, Department of Physiology, IGMC, Shimla;<sup>3</sup>Associate Professor, Department of Physiology, IGMC, Shimla.

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Conflict of interest: Nil

**Abstract**

**Background:** Inflammatory bowel disease (IBD) is an immune mediated chronic intestinal disease. It is of two major types, Ulcerative Colitis (UC) and Crohn's Disease (CD). Though IBD is primarily an intestinal disease, it involves various organ systems such as eyes, skin, musculoskeletal and hepatobiliary systems. Sensorineural hearing loss is one of such extraintestinal manifestations which is attributed to the autoimmune inner ear disease. The objective of this study is to find the prevalence of sensorineural hearing loss in patients of inflammatory bowel disease.

**Materials and Methods:** 28 patients of inflammatory bowel disease in the age group of 18-50 years visiting outdoor patient department or admitted in the Department of Gastroenterology, IGMC Shimla over a period of one year (August 2014 to July 2015) fulfilling the inclusion and exclusion criteria were included for the study. The subjects were then subjected to a battery of audiological tests: Tuning fork tests, Pure Tone Audiometry and Brain Stem Evoked Response Audiometry (BERA). The data was tabulated in Microsoft Excel spread sheet and Epi Info version 3;4,3 statistical software was used for statistical analysis.

**Results:** A total of 28 patients were included in the study, 15 of the which were females and 17 males. Mean age of the study population was  $37.79 \pm 8.14$  years. 23 (82.14%) patients were diagnosed to have Ulcerative Colitis while 5 (17.85%) patients were diagnosed to have Crohn's disease on endoscopy and further histopathological examination. A total of 8 (28.57%) patients were found to have sensorineural hearing loss. Out of these 8 patients of SNHL, 6 (75%) had UC and 2 (25%) had CD. 6 (75%) patients had Cochlear type of hearing loss while 2 (25%) had Retrocochlear type of hearing loss. The mean Duration of IBD of the patient population with SNHL was  $6.37 \pm 4.90$  years while that of the patients without SNHL was  $1.34 \pm .82$  years.

**Conclusion:** Inflammatory bowel disease is a treatable cause of otherwise permanent sensorineural deafness. Early audiometric evaluation may be advised in all inflammatory bowel disease patients.

**Key words:** Sensorineural Hearing Loss, Inflammatory Bowel Disease, Extraintestinal Manifestation, Ulcerative Colitis, Crohn's Disease.

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**Introduction**

Hearing impairment is the most frequent sensory deficit in human populations, affecting more than 250 million people worldwide. [1] Consequences of hearing impairment include inability to interpret speech sounds, often producing a reduced ability to communicate, delay in language acquisition, economic and educational disadvantage, social isolation and stigmatization. It may also be worsened by some associated medical conditions such as hypothyroidism, diabetes and other systemic diseases.

Hearing loss can further be divided into three types as conductive, sensorineural and mixed. While Sensorineural hearing loss (SNHL) is the type of hearing loss which results from lesions in the vestibulocochlear nerve (Cranial nerve VIII), the

inner ear, or central processing centres of the brain. It may be present at birth (congenital) or start later in life (acquired) caused by age related degenerative changes in the organ of Corti (presbycusis), infections such as labyrinthitis, noise exposure and trauma. SNHL has many different presentations, ranging in severity from mild to profound, including low and high pitch patterns; and can affect people of any age.

Sensorineural hearing loss is a well recognized condition of diverse aetiology. The possibility that part of the previously considered idiopathic SNHL were autoimmune in origin was first noted by Lehnhardt in 1958. [2] McCabe later described this condition known as autoimmune inner ear disease (AIED), autoimmune cochleopathy or immune-

mediated cochleovestibular disease in his study on patients with progressive SNHL. [3] SNHL is currently a well-recognized Extra intestinal manifestations (EIM) of Inflammatory bowel disease (IBD). [4]

IBD comprises conditions characterized by chronic or relapsing immune activation and inflammation within the gastrointestinal tract. It comprise of three types of chronic intestinal disorders: Crohn's disease (CD) Ulcerative colitis (UC) and indeterminate colitis(IC). The incidence of IBD has dramatically increased during the 20th century, and in the developed nations is as high as 20 and 24 cases per 100,000 person-years for CD and UC, respectively. As developing countries, such as India and China, became industrialized, the incidence of IBD has risen in parallel. This shift in risk suggests that changes in environment, in addition to genetic predisposition, contribute to the development of IBD. [5,6]

IBD are now considered as systemic disease since they are associated with clinical manifestations involving organs outside the alimentary tract. The Extra intestinal manifestations (EIMs) are found in more than 50% of all the IBD patients. [7,8] The neurological involvement in IBD as a subgroup of the EIMs may precede the appearance of digestive symptoms or develop after diagnosis of IBD. In addition, neurological symptoms may exacerbate during flare-ups of IBD or evolve independently from intestinal manifestations without responding to treatment provided for the underlying bowel disease. [9,10,11]

The pathogenesis of SNHL associated with IBD is not fully understood, and several immune mechanisms may lead to inner ear dysfunction. [12,13,14] The hearing level often fluctuates, with periods of deterioration alternating with partial or even complete remission. Inner ear involvement associated with IBD is frequently underreported. Some authors have discussed the EIM of IBD that involve virtually all organ systems but make no mention of hearing loss. [15,16] Hence, in order to investigate this further we undertook the present study to find out the prevalence of SNHL in patients with IBD and to study its relationship with various disease parameters.

### Material and Methods

The present study was a hospital based observational study conducted in the Department of Otorhinolaryngology - Head and Neck Surgery and the Department of Gastroenterology, IGMC Shimla from August 2014 to July 2015. All consecutive patients of inflammatory bowel disease in the age group of 18-50 years visiting outdoor patient department or admitted in the Department of Gastroenterology, IGMC Shimla over a period of

one year (August 2014 to July 2015) without any other systemic illnesses or history of hypertension, Diabetes mellitus, Chronic Kidney disease etc. and consenting to participate were included for the study. Patients with any congenital ear anomalies /ear pathology or having history of head trauma / noise exposure / exposure to ototoxic drugs/ exposure to radiotherapy, or with family history of hearing loss were excluded from the study.

IBD diagnosis was based on the typical clinical course of the disease and endoscopic examination with histologic confirmation of UC or CD. Twenty eight diagnosed patients of IBD were then included in the study. A pre structured Proforma was used to record the information ascertained. Patients found to be eligible for the study based on specified inclusion and exclusion criteria were then subjected to a battery of audiological tests after thorough systemic and ENT examination.

Audiological assessment included the Tuning fork tests, Pure Tone Audiometry and Brain Stem Evoked Response Audiometry (BERA). Pure tone audiometry was done in all the subjects to know the subjective pure tone audiometry with interacoustics AC40 Clinical Audiometer [interacoustics A/S. Assens, Denmark,] in a fully sound attenuated room situated in the Department of Otorhinolaryngology IGMC, Shimla. BERA was performed using EPIC-PLUS LABAT BERA machine in a fully sound attenuated room situated in the Department of Otorhinolaryngology IGMC, Shimla with patient lying down, eyes closed and preferably asleep. Following parameters of BERA were studied :- (i) Latency of the wave – absolute, interwave & Interaural, (ii) Amplitude of the wave – both absolute & relative and (iii) Wave morphology

**Statistical Analysis:** Data was entered in Microsoft Excel spread sheet and Epi Info version 3;4,3 statistical software was used for statistical analysis. The clinical characteristics of the study population were reported as percentages and Mean $\pm$ SD for categorical and continuous variables respectively. All the tests with regard to significance were two tailed. Statistically significant value was defined as  $p < 0.05$ .

### Results:

Twenty eight diagnosed patients of IBD were included in the study. The sociodemographic and Clinical profile of study population is described in detail in Table 1. In brief the mean age of the patients in the study group having IBD was  $37.79 \pm 8.14$  years that included 15 male and 13 female participants.

Amongst the study group, 23 (82.14%) patients were diagnosed to have Ulcerative Colitis while 5 (17.85%) patients were diagnosed to have Crohn's disease on endoscopy and further histopathological

examination. Out of 28 patients of IBD, 15(53.57%) patients presented with various gastrointestinal symptoms like altered bowel habits and only 3 (10.71%) subjects presented with various auditory symptoms like tinnitus, vertigo and ear discharge. While 8 (28.57%) patients presented with various other Extra intestinal manifestations like erythema, aphthous ulcers, episcleritis, pyoderma and ankylosing spondylitis. The mean duration of illness in the study group suffering from IBD was 2.75 ± 3.51 years.

Table 2 shows the Prevalence of SNHL in the study population. A total of 8 (28.57%) patients were found to have sensorineural hearing loss. Out of these 8 patients of SNHL, 6 (75%) had UC and 2 (25%) had CD.

Severity / degree of hearing loss by PTA has been described in Table 3. Among the 8 patients of SNHL on PTA, 6 (75%) patients had Grade I hearing loss while 2 (25%) patients had Grade II hearing loss. None of the patients had Grade III hearing loss. While table 4 shows the Type of hearing loss on BERA. 6 (75%) patients had Cochlear type of hearing loss while 2 (25%) had Retrocochlear type of hearing loss. Out of 6 patients of Cochlear type of hearing loss, 5 (83.33%) patients had Bilateral

hearing loss while only 1 (16.66%) had Unilateral hearing loss. While in the retrocochlear type both the patients (100%) had Bilateral hearing loss.

Table 5 Compares the baseline characteristics between patients with and without SNHL. Amongst the patients having SNHL, 6(75%) had UC and 2(25%) had CD while in patients without SNHL 17(85%) had UC and 3 (15%) had CD. The mean Duration of IBD of the patient population with SNHL was 6.37 ± 4.90 years while that of the patients without SNHL was 1.34 ± .82 years. A statistically significant difference was observed in the mean Duration of IBD among the two groups (p=0.000). Among the SNHL patients 3 (37.58%) had Other Extra intestinal manifestations while among the patients without SNHL 5 (25%) had Other Extra intestinal manifestations. No statistically significant difference was observed (p=0.566). Among the SNHL group, 1 (12.5%) patient and among the non SNHL group 2 (10%) patients had one or the other Auditory Symptoms like tinnitus, vertigo and ear discharge at the time of presentation to the hospital. No statistically significant difference was observed (p=0.388).

**Table 1: Sociodemographic and Clinical profile of study population (N=28)**

VARIABLES	
Age, Years (Mean±SD)	37.79 ± 8.14
Gender, Males (Percentage)	15 (53.61%)
Ulcerative colitis	23 (82.14%)
Crohn’s Disease	5 (17.85%)
Family history of IBD (percentage)	2 (7.14%)
GI symptoms	15 (53.57%)
Auditory Symptoms (percentage)	3 (10.71%)
Other EIM (percentage)	8 (28.57%)
Smokers (percentage)	8 (28.57%)
Alcohol (percentage)	5 (17.93%)
Duration of IBD(years) Mean±SD	2.75 ± 3.51

**Table 2: Prevalence of SNHL in the study population (N=28)**

VARIABLE	IBD (N=28)	UC (N=23)	CD (N=5)
SNHL N (%)	8 (28.57%)	6 (75%)	2 (25%)

**Table 3: Severity / degree of hearing loss by PTA**

GRADE	Group	No. of Patients (8) N (%)	Hearing level
Grade I	Mild	6 (75%)	26-40dB
Grade II	Moderate	2 (25%)	41-55dB (moderate)
	Moderately severe	0	56-70dB (moderately severe)
Grade III	Severe	0	71-90dB (severe)
	Profound	0	Above 90 dB(profound)

**Table 4: Type of hearing loss on BERA**

TYPE OF SNHL	Number of patients With SNHL (N=8)	Laterality B/L U/L	
Cochlear N (%)	6 (75%)	5 (83.33%)	1 (16.66%)
Retrocochlear N (%)	2 (25%)	2 (100%)	0

**Table 5: Comparison of baseline characteristics between patients with and without SNHL**

VARIABLES	WITH SNHL(N=8)	WITHOUT SNHL (N=20)	p value
Ulcerative colitis N (%)	6 (75%)	17 (85%)	0.786
Age Years, Mean $\pm$ SD	42 $\pm$ 8.90	36.10 $\pm$ 7.40	0.083
<b>Gender (Percentage)</b>			
Males N (%)	4 (50%)	11 (55%)	0.862
Smokers N (%)	2 (25%)	6 (30%)	0.862
Duration of IBD (Years)	6.37 $\pm$ 4.90	1.34 $\pm$ .82	0.000
EIM N (%)	3 (37.58%)	5 (25%)	0.566
Family H/O IBD N (%)	1 (5%)	1 (5%)	0.201
Auditory Symptoms N (%)	1 (12.5%)	2 (10%)	0.388

## Discussion

Crohn's disease and Ulcerative Colitis are indolent disabling diseases and form the major components of the IBD complex whose fundamental pathology is a severe noninfectious inflammation of the mucosa of the gastrointestinal tract. The immunologic abnormalities, however, that are associated with IBD and the increased risk of other autoimmune diseases among patients with IBD lend some support to the concept of an immunopathy-based process. [17] The extra intestinal manifestations of IBD, however, are not of less importance. In some cases they are the first clinical manifestation of the disease and may precede the onset of gastrointestinal symptoms by many years. [18,19] During the last few years many authors have reported many serious complications of IBD manifesting in the Ear-Nose-Throat (ENT) and influencing disease morbidity. Sensorineural hearing loss being a seldom recognized extra intestinal manifestation of both UC and CD often remains under reported. We thus carried out the present study to find out the prevalence of SNHL in patients with IBD and to study its relationship with various disease parameters.

In our study, out of twenty eight diagnosed cases of IBD, 23 patients were found to have UC and 5 patients were confirmed as cases of CD on colonoscopy and biopsy. The higher incidence rate of UC as compared to CD in our study population was in accordance to the statistics of previous studies done on Asian and Indian cohorts. [20]

15(53.57%) patients in our study presented with various Gastrointestinal symptoms, while 8 (28.57%) patients presented with other extra intestinal manifestations. The mechanisms involved in the pathogenesis of extra intestinal manifestations of IBD are not clear, but increased bowel permeability during active disease may cause luminal antigens to be presented to the systemic

immune system. [21,22] The inner ear, nose and throat, like other extra intestinal involvement sites in IBD, can become targets of an autoimmune attacks. There is considerable evidence to suggest that hearing and vestibular function can be influenced by autoimmune processes. The most important diagnostic finding in their studies was the improvement in hearing seen with a trial of immunosuppressants like corticosteroids and cyclophosphamide which further substantiates the autoimmune characteristics of progressive or sudden sensorineural hearing loss seen in patients of IBD.

In 80% of our patients with Bilateral hearing loss the auditory deficit first occurred in one ear which gradually/suddenly progressed to the other ear. The subsequent involvement of the contralateral ear, suggests the possibility that degeneration of inner ear tissues in one ear lead to the production of anti-cochlear antibodies that eventually damaged the second ear leading to hearing loss in the opposite ear. This assumption challenged the theory of the inner ear being an immunologically privileged site due to the presence of a blood-labyrinthine barrier. [23]

A significant difference was noted in the mean years of duration of IBD in the SNHL group when compared with the other group (6.37  $\pm$  4.90 vs 1.34  $\pm$  .82 years,  $p < .05$ ). The occurrence of SNHL correlates with several parameters of IBD, total duration of the disease being one of them, which usually parallels the activity of intestinal inflammation. But these findings need to be elucidated by further clinical and experimental studies performed with larger patient populations of IBD. An investigation of the natural course and autoimmune pathogenesis, including possible relations with autoantibodies for IBD such as pANCA (perinuclear antineutrophil cytoplasmic antibodies) and ASCA (anti-Saccaromycescerevisiae antibodies) may also be required to postulate this further.

### Limitations

The results of the present study are applicable to a small cohort of IBD patients where the duration of the study is likely to be short. Since the sample size was small the trends of reduced hearing observed in patients of IBD might be truly reduced but due to limited power of the study the null hypothesis may have been falsely accepted. Study of a larger cohort with controls would be more valuable.

The observations reported in the study are on hospital based patient population thus with inherent selection biases. Another limitation of our study was the lack of precise information about the time of onset and progression of the SNHL because most of our case patients were unaware of their hearing status before their first visit.

### Conclusion

Our study lends further credence to the theory that SNHL can be an extra intestinal manifestation of IBD. Knowledge of the entity and its variable presentation is essential to facilitate prompt recognition and treatment, so that permanent disability may be prevented. Larger long term studies are needed to better understand the natural history of SNHL in IBD as well as common etiological mechanisms linking these two disorders. As it is unknown whether asymptomatic hearing loss could progress to severe overt hearing loss or not, follow up of patients with positive findings is required periodically.

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