Treatable Sensorineural Hearing Loss
Essay

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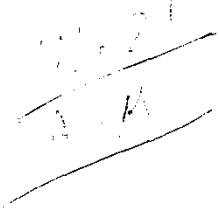
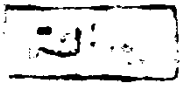
By

Abdalla Mahmoud Abdalla

M.B., B.Ch.





Supervised By

Prof. Dr. Abd El Rehim Sarwat

Prof. of E.N.T., Faculty of Medicine Ain Shams University

and

Dr. Mahmoud El Sammaa

Lecturer of E.N.T., Faculty of Medicine Ain Shams University

Faculty of Medicine

Ain Shams University

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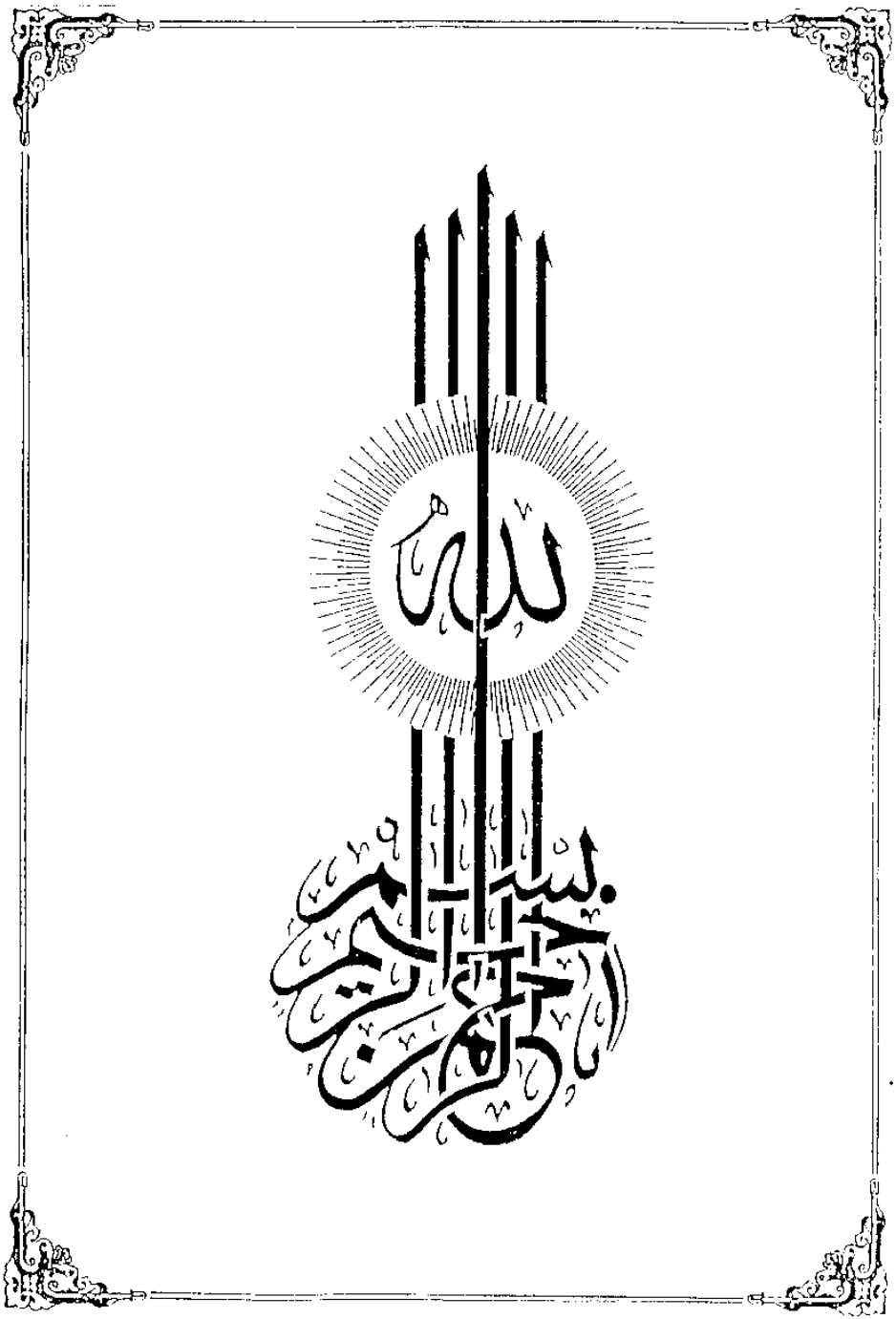

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INTRODUCTION

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The old concept was that sensorineural hearing loss is untreatable and the patient may be fixed with a hearing aid and encouraged to acquire lip-reading habbits in a rehabilitation programme in the severe cases .

Recently , this concept has been changed as a small but increasing number of conditions producing sensorineural hearing loss can be helped with specific medical or surgical treatment to restore hearing partially or completely or at least stabilize it and stop its progression .

The aim of this essay is to discuss the various causes of treatable sensorineural hearing loss , their clinical presentation , diagnosis and try to treat these causes to improve the hearing loss or to stabilize it .

These conditions include : sudden sensorineural hearing loss , perilymph fistula , Meniere's disease , allergic causes metabolic causes , syphilis , cochlear otosclerosis , acoustic neuroma , stenosis of the internal auditory canal , cervical nerve lesions , Mondini's deafness and autoimmune inner ear disease .

SUDDEN SENSORINEURAL HEARING LOSS

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It is an otologic emergency . It is characterized by rapidity of onset which may be within a moment or over a few days , Goodhill (1979) .

Causes , Shambaugh (1980) :

I. Popular proven causes :

- . Meniere's disease
- . Virus
- . Membrane break
- . Trauma
- . Syphilis
- . Autoimmune disease
- . Vascular occlusion
- . Metastatic cancer
- . Multiple sclerosis
- . eighth nerve tumour .

II. Rare causes :

- . Embolic phenomenon
- . Sickle cell crisis
- . Psychogenic
- . Ceagulapathies
- . Diabetes mellitus
- . Vitamin defficiency
- . Ototoxicity
- . Blood sludging
- . Allergy
- . Renal dialysis
- . Hypercholesterolaemia (vasospasm) .

III. Idiopathic .

Clinical features :

Sudden sensorineural hearing loss may be mild, moderate or severe in degree . It may occur alone , or it may be preceded , accompanied , or followed by vertigo . Tinnitus is frequently present . Ataxia also has been reported, Healey et al. (1976) .

The patient usually requests examination within hours or days of onset . However , a week or more may have elapsed . The patient may emphasize the tinnitus as a major complaint with secondary reference to hearing loss , regardless of its severity . Vertigo may accompany the onset of hearing loss and tinnitus , but more often , it follows by hours or days and may be the only symptom . It may vary from a mild "giddy " feeling to severe true vertigo with spontaneous nystagmus , nausea and vomiting or ataxia . In addition to hearing loss , tinnitus , and vertigo , frequent complaints include : a " dead feeling " , a " numbness " or a " hollowness " in the affected ear . Slight pain and / or a feeling of itching deep in the ear occur in some patients, Goodhill (1979) .

The history of hearing loss may be preceded by awareness of viral infection i.e. " flu " or " cold " or sore throat , sinusitis or viral exposure .

The patient may give a history of cardiac

or hypertensive disease and may give a history of diabetes , arteriosclerosis , hypercholesterolaemia , hyperlipidaemia or other systemic disease involving microvasculature . The onset is usually not preceeded by any stress-related event . There may be also a history of barotrauma or stress due to unusual lifting , coughing sneezing , difficult micturition or defecation or sexual intercourse immediately prior to the onset of hearing loss , Goodhill (1980) .

Medical history , should include previous general medical problems , cardiocirculatory disease , anticoagulant drug intake , ototoxic drug use , recent viral or bacterial upper respiratory infections , and recent unexplained fever , chills or malaise , Goodhill (1979) .

Otologic history should be obtained in details concerning previous ear problems or surgery , previous ear examinations , vertigo or tinnitus experiences . Inquiry is made into the possibility of trauma or noise exposure or family history of deafness , Nadol and Wilson (1980) .

Investigations :

Otologic examination should include otoscopy , standard audiologic tests , special cochlear-retrocochlear auditory tests battery , tympanometry , stapedius reflex and vestibular E.N.G. studies , cranial nerve and basic neurologic examination as well as Schuller and Stenever view radiographs . The clinical findings of all patients regardless of etiology are usually similar , namely normal otoscopic findings and audiological evidence of moderate or total sensorineural hearing loss involving high-frequency losses with retention of low-frequency hearing . Bekesy tracings are not usually diagnostic in terms of types , Mattox and Simmons(1977) .

Vestibular studies show varied responses , and although caloric responses may be abnormal , the findings are only of importance in ruling out previously unrecognised difinitive mastoid or petrous pyramid lesions , Goodhill (1979) .

Medical examination should include special search for bradycardia , arrythmia and other cardiac and circulatory problems , bruits , and urine and blood studies including sedimentation rate , and FTA- ABS tests . Special tests for hypercoagulation , lipidemia hyperviscosity , and macroglobulinemia and other syndromes may be indicated , Meyerhoff and Paparella (1980) .

Differential diagnosis :

The differential diagnosis from fluctuant hearing loss of Meniere's disease is difficult , but in fluctuant hearing loss , the onset is usually less sudden , there is the characteristic fullness not present in sudden hearing loss, and the response in the tranetympanic electrocochleograph to glycerin dehydration and intravenous hypaque is quite different . The differential diagnosis of idiopathic sudden hearing loss from that due to round or oval-window membrane rupture is made with the history , examination of the patient , and surgical exploration if necessary, Shea and Emmett , (1980) .

Treatment :

The patient must be hospitalized and rest in bed . In cases due to proven aetiology , such as trauma , toxins , haematological disorders , neoplasms and syphilis , treatment should be directed to the cause . In cases of syphilis , a good response is usually obtained with antibiotics and steroid therapy which may be continued for life to avoid relapses , Hendershot (1973) .

The hearing loss associated with systemic toxins , both iatrogenic and enviromental is usually irreversable except in the case of aspirin , quinine , ethacrynic acid and furosemide Meyerhoff and paparella (1980) .

Shambaugh (1980) stated that , sudden sensorineural hearing loss due to ototoxicity can be prevented by the following measures :

1. Avoidance of ototoxic drugs especially ototoxic antibiotics .
2. Proper dosage of the drugs .
3. Serial determination of concentration of the drug and serial determination of serum creatinine .
4. Stoppage of the drug if worning manifestations appear (e.g. tinnitus , vertigo or and audiometric changes) .

Treatment of idiopathic group :

1. Anticoagulants : Meyer (1958) stated that the use of heparin and other anticoagulants in the treatment of idiopathic sudden sensorineural hearing loss is borrowed from their use in cerebral vascular occlusion , in which anticoagulants seem to promote better collateral circulation whereasе the effect in ischaemic area itself is to produce an increase in perivascular haemorrhage . In the cochlea , however , there is a little or no collateral circulation , so that the only logical expectation for these drugs would be an increased risk of haemorrhage within the inner ear with no improvement of blood flow .

Meyerhoff & Paparella(1980)reported that,the adminsteration of heparin 5000 to 10.000 units subcutaneously every 12 hours in combination with ACTH gel , 40 units by intramuscular injection daily , papaverine hydrochloride (150 mg.) by mouth every 12 hours and low molecular weight dextran (10 ٪) , 500 ml. given intravenously over a 4-hour period twice daily gives good results .

2. Corticosteroids : Jaffe and Maassab (1967) reported that , if the aetiology of idiopathic sudden sensorineural hearing loss is indeed viral , corticosteroids are potentially harmful , since the immune respose to the viral infection is diminished by corticosteroids , as has been demonstrated in herpes infections of the cornea and they added that , the possible mechanism of injury secondary to viral infection may be ischaemia due to swelling of the endothelial layers of the capillary wall . However, reduction in swelling occurs only many hours after corticosteroid adminsteration.

Haynes (1975) on the other hand , atated that , corticosteroids have a vasoconstrictive effect that develops over a similar period and that would further reduce perfusion to the affected tissue.

3. Vasodilators : The use of vasodilator therapy assumes that vasospasm has caused transitory cochlear ischaemia and a subsequent sudden hearing loss and that the use of vasodilators will reverse the vasospasm and induce recovery of cochlear function . These therapeutic hypotheses are not supported by experiemental evidence , Nadol and Wilson (1980) .

- Histamine : although histamine has a powerful dilatory effect on the cerebral vasculature , it has the undesirable side effect of causing marked tissue oedema . The changes in cochlear blood flow following histamine adminsteration varied in some animals,there was an increase in blood flow whereas in others , there was a decrease , Suga and Snow (1969) .

-Nicotinic acid : although nicotinic acid does cause a peripheral vasodilatation , there is no associated effect on cerebral or cochlear blood flow and in fact , the net result of nicotinic acid adminsteration may be to lower the systemic blood pressure and reduce perfusion of affected cerebral vasculature , Shaia and Sheehy (1976) .

-Nylidrin hydrochloride and papaverine hydrochloride: have been shown to produce dilatation of of the normal cerebral vasculature , including the cochlear vasculature, however , the use of these drugs in acute and chronic iscaemic conditions of the peripheral and cerebral vasculature has been disappointing , Eisenberg (1960) .

-Inhalation of 5 % carbon dioxide and 95 % oxygen : will dilate normal cerebral and cochlear vasculature, Suga and Snow (1969) .

4. Diatrizoate Meglumine : (Hypaque) ,

Morimitsu (1977) reported on the successful treatment of sudden hearing loss with diatrizoate meglumine , an iodinated dye used in radiologic studies , which has a trade name of Hypaque or Urografin . The initial case involved a patient who had a sensorineural hearing loss of 40 days' duration and a significant improvement in hearing following a 1 cc test dose of intravenous Hypaque prior to vertebral angiography study . Subsequently , he compared 39 patients treated within 2 weeks following onset of sensorineural hearing loss with 10 cc of Hypaque daily until maximum recovery with 419 patients followed by the Sudden Deafness Rresearch Committee of Japan , who were treated with vasodilators within the same period . Of the the patients treated with Hypaque , 54 % (21) had complete recovery of hearing compared with 19 % (80) of those treated with vasodilators.

The systemic pharmacological actions of Urografin or Hypaque such as on blood pressure , heart rate , and respiration rate are extremely rare . When injected intravenously they pass through the capillaries of the glomerulus of the kidney only and because of their molecular weight and structure they are not reabsorbed by the renal tubules . They can not permeate the blood barrier and also the blood cochlear barrier . The structure and biological function of the stria vascularis are similar to that of the renal tubules.In idiopathic sudden sensorineural hearing loss , the blood cochlear barrier breaks down in that portion of the stria vascularis exposed to the mixture of endolymph and perilymph . The site of action of Hypaque is limited to the vessels of the stria vascularis damaged by the mixture of endolymph and perilymph in wich the cochlear barrier breaks down . The molecular weight and structure of Hypaque are such as to allow it to pass into the damaged stria vascularis and somehow repair and reactivate the sodium-potassium pump to produce endocochlear potential again , Shea and Emmett (1980)