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Hearing Loss in Infectious and Contagious Diseases

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Abstract

Hearing loss can occur for various reasons, whether it is of a genetic, congenital or acquired character. Infectious diseases stand out among those causing this type of deficiency and account for approximately 25% of all cases of profound hearing loss. Of these, one-fifth are due to congenital causes. As to classifying hearing loss, this can be done according to where this is in the hearing system, to whether the loss is unilateral or bilateral, and to its intensity or degree. Regarding where the hearing system is affected, hearing loss can be about transmission (or conduction), perception (sensorineural), or mixed. Hearing losses arising from any affection of the outer and middle ear are called transmission or conductive losses. Sensorineural losses occur due to lesions on the hair cells of the cochlear organ of Corti (inner ear) and/or of the cochlear nerve. When there is concomitant conductive and sensorineural affection, the loss is classified as mixed. Hearing loss can interfere in the lives of affected individuals, since besides affecting communication, it can influence the quality of life, when the loss leads to feelings of sadness and anxiety, or even to social isolation. In children, it can moreover represent consequences for development. Thus, appropriate treatment and/or monitoring of infectious diseases is important, the purpose of which is to see to it that hearing loss is prevented or diagnosed early.

Keywords: Hearing loss, infectious diseases, etiology

1. Introduction

The hearing process begins when sound waves enter the outer ear and travel along the ear canal to the eardrum, causing it to vibrate. These vibrations are transmitted to the ossicles of the middle ear, which cause the sound vibration to be amplified before transmission to the inner ear. The inner ear has a part called the cochlea, which is filled with liquid and contains hair cells.[1]

The frequencies and intensities of the sound determine which hair cells will move. This causes electrical impulses to be generated and sent through the auditory pathways to the brain so that it may process the information. These electrical impulses are the codes that the brain can process and, on understanding them, assigns them various specific meanings.[1]

Hearing losses can be classified according to the location of the portion of the hearing system affected, to whether the loss is unilateral or bilateral, and to its intensity or degree. The location of the portion affected of hearing loss has to do with transmission (or conduction), perception (sensorineural), or a mixture of these (mixed). Sensorineural losses arising from some affection of the outer and middle ear are called transmission or conductive losses. Sensorineural losses result from lesions on the hair cells of the cochlear organ of Corti (inner ear) and/or of the cochlear nerve. When there are concomitant conductive and sensorineural affections, the hearing loss is classified as mixed.[2]

Hearing loss can occur due to a genetic, congenital, or acquired cause.[3] Among the acquired causes, many could sometimes be avoided, e.g., infections that occur during pregnancy, meningitis, and even due to using ototoxic medication.[4]

2. Epidemiology

Deafness is a global problem that affects individuals, families, societies, and governments. According to the World Health Organization (WHO), deafness affects between 1 and 4 people per 1,000 individuals, and there has been a considerable increase in poor countries. In 2005, for example, about 278 million people had degrees of hearing loss between moderate and profound, and 80% of them live in poor and developing countries.[5] Prevalence greater than 1 per 1000, however, indicates a serious public health problem that needs urgent attention.[6]

Infectious diseases are the leading cause of hearing loss and produce about 25% of profound losses. Of these, the causes of one-fifth are congenital.[7] The main infections include diseases such as rubella, cytomegalovirus, and measles.[7]

In the newborn, congenital infections are an important cause of hearing loss, which may have implications for the development of the child.[8]

3. Pathogenesis

The mechanisms that lead to the onset of viral hearing loss may include infections of the upper airways, may progress to subsequent involvement of the middle ear, and may occur with conductive hearing loss.[3]

Moreover, viral invasion of the inner ear can occur.[3] The viruses that can damage the inner ear may do so at different stages of the life cycle: during intrauterine life, childhood, adolescence, or adulthood. The pathological changes that predominate in the basal cochlea include

degeneration of the organ of Corti, atrophy of the stria vascularis, displacement and distortion of the tectorial membrane, and degeneration of the saccule. The utricle and semicircular canals tend to be preserved.[9]

4. Main infectious and contagious diseases related to loss of hearing

4.1. Infections by virus

4.1.1. Epidemic parotiditis

Epidemic parotiditis or mumps is an acute systemic and contagious viral infection. A *Paramyxovirus*, with an RNA genome, is involved.[10]

The most typical clinical manifestations are sialadenitis, epididymo-orchitis, pancreatitis, meningitis, and hearing loss. Sensorineural hearing loss occurs in up to 5/10,000 cases and may appear some days or weeks after the parotiditis.[11]

Deafness is usually sudden, profound, associated with or without nausea, vomiting, dizziness, and tinnitus.[9] Hearing loss, which is unilateral in 80% of cases, is more common for high frequencies of sound and may present reduced caloric responses to the vestibular test.[11]

There may be atrophy of the organ of Corti and of the stria vascularis, with minimal effect on the vestibular system. Also observed are endolymphatic hydrops and obliteration of the endolymphatic duct.[11]

4.1.2. Infection by cytomegalovirus

Cytomegalovirus (CMV), which belongs to the herpesvirus family, is an enveloped virus that has the largest genome among the viruses that infect animal species. In immunocompetent individuals, it is generally responsible for asymptomatic infections.[12]

The highest incidence of the primary infection occurs in two peak periods: the first is in childhood, with early acquisition as a result of perinatal infection, and the second is in adolescence, through sexual transmission or by kissing.[12] It infects up to 70% of children who spend the day in kindergartens, and about 1–2% of infants are infected with CMV.[13]

In the congenital form, clinical manifestations range from the unapparent to the severe and widespread. Cytomegalic inclusion disease develops in about 5% of the infected fetuses. The most common manifestations at presentation are petechiae, hepatosplenomegaly, and jaundice. The occurrence of microcephaly with or without intracranial calcifications delayed intrauterine growth, and prematurity in 30–50% of cases is observed.[12] Deafness occurs in 20–65% of infants with this disease, which is typically bilateral.[13]

In patients with hearing loss, a consistent pattern follows, and this can develop over a period of years. Among asymptomatic patients, the rate of hearing loss of such children ranges from 7% to 13% and should therefore be considered in patients with nonsyndromic and nongenetic hearing loss.[13]

4.1.3. Rubella

This is a disease with an acute rash caused by an RNA virus of the genus *Rubivirus* and the Togaviridae family, which is highly contagious and mainly affects children.[13]

The clinical state is characterized by a maculopapular and diffuse pinpoint rash, starting on the face, scalp, and neck, later spreading to the trunk and limbs.[13]

The infection acquired after birth usually causes a mild or even subclinical disease. The main symptoms of this form are retroauricular, cervical and suboccipital lymphadenopathy, rash, and fever. Complications are uncommon.[10]

Maternal infection during early pregnancy can lead to infection of the fetus, resulting in congenital rubella.[10] Congenital rubella syndrome (Gregg's syndrome) affects most organ systems, causing cataracts, microphthalmia, heart defects, skin rash, retardation of growth, and hearing loss. In general, hearing loss affects about 50% of individuals with the disease and is normally severe to profound. Auditory manifestations may occur months to years after the initial infection.[11]

4.1.4. Measles

Measles is an acutely infectious viral disease that is potentially serious, transmittable, and extremely contagious. Its etiologic agent is an RNA virus of the genus *Morbillivirus*, family Paramyxoviridae.[13]

Among the clinical manifestations, it is characterized by high fever, above 38.5°C, a widespread maculopapular rash, cough, coryza, conjunctivitis, and Koplik spots (small white spots on the oral mucosa, prior to the rash).[13]

It may cause severe degeneration of the organ of Corti, the stria vascularis, cochlear neurons, and vestibular damage. Inflammation, fibrous deposit, and ossification in the basal turn of the cochlea may also be present. Hearing loss tends to be asymmetrical, bilateral, and severe. Vestibular abnormalities are not rare.[11]

4.1.5. Viral meningitis

Viral meningitis is characterized by a clinical state of neurological changes, which usually develops benignly. Approximately 85% of cases are due to the group of Enteroviruses, among which the poliovirus, echovirus, and coxsackievirus stand out. Other less common groups are arboviruses, herpes simplex virus, and varicella, mumps, and measles viruses.[14]

It occurs most frequently in children over two years old and can lead to sensorineural hearing loss.[3]

4.1.6. Herpes simplex

Herpes simplex has been considered one of the most common viral contamination agents in humans and is subdivided into two groups: type 1 and type 2.[15]

Infections caused by herpes simplex type 1 usually affect areas such as the lips, mouth, intraoral region, nose, eyes, while infections caused by herpes simplex type 2 are mainly found in the genital and surrounding areas. Trigger factors include fever, exposure to cold temperatures or ultraviolet rays, skin or mucous abrasions, emotional stress, and nerve injury. In the case of occurrence in newborns, the onset of infection can be at different periods: prenatal (congenital infection), perinatal (infection through the birth canal), or postnatal (infection through contact with infected individuals).[15]

The virus of the herpes group is regarded as causing sensorineural loss. In pregnancy, it can also cause spontaneous miscarriages, still births, and congenital defects.[15]

4.1.7. Infectious mononucleosis and other viral agents

Infectious mononucleosis (IM) is caused by the Epstein–Barr virus (EBV), characterized by fever, pharyngitis, lymphadenopathy, and atypical lymphocytosis. EBV is a member of the Herpesviridae family.

One of the main viral agents associated with sensorineural hearing loss in adulthood is the IM virus. Other agents that can also often affect this age-group and are related to hearing loss are adenovirus, enterovirus, influenza, and parainfluenza.[3]

4.2. Infections by bacteria

4.2.1. Bacterial meningitis

Meningitis is frequently associated with a high mortality rate. A large portion may still present sequelae of the disease, among which is hearing loss. This disease is held to be among the main ones responsible for postnatal acquired hearing impairment.[16]

Among the mechanisms elucidated, as being responsible for hearing damage, is the direct invasion of the bacteria into the cochlea and labyrinth, lesion of cranial nerve VIII, by toxins, and blockage of small vessels and ototoxic action of the antibiotics used. Regarding the degree of loss, a high percentage of profound hearing loss (66.95%) has been evidenced. However, hearing loss of all degrees (mild to anacusis) was observed.[16]

In a study of 124 children recruited from 21 hospitals in England and South Wales, aged between 4 weeks and 16 years old, with a recent diagnosis of bacterial meningitis, 92 (74%) had meningococcal and 18 (15%) had pneumococcal meningitis. All cases showed obvious hearing loss in the first assessment. Three children had permanent sensorineural hearing loss. Thirteen children (10.5%) had reversible loss, nine of which were resolved within 48 hours of diagnosis.[17]

The impact on the development of the child after meningitis can be devastating. In the postmeningitis period, a possibility of rehabilitation for patients with severe and profound sensorineural loss is a cochlear implant.[18]

In cases of postmeningitis hearing loss, it is particularly important to do the implant as early as possible due to the intracochlear ossification that may occur, thus preventing the placement of electrodes in the lumen of the cochlea.[19]

4.2.2. Syphilis

During the decade of 2003–2012, the diagnosis of primary syphilis increased 61% in men in England, while in contrast, this diagnosis in women decreased by 16%.[20] In the 2004 Sentinela Parturiente (Mother in Labour Sentinel) Study of the Ministry of Health in Brazil, the prevalence of syphilis in pregnant women was 1.6%, about four times higher than HIV infection in the same group, the estimate being that a total of 48,425 pregnant women were infected in that year. Between 2005 and June 2012, 57,700 cases of syphilis in pregnant women were registered in SINAN (the Brazilian statutory body for notifiable diseases), most of which occurred in the Southeast and Northeast regions.[21]

Syphilis is an infectious disease caused by a bacterium, *Treponema pallidum*, which is predominantly transmitted sexually. If left untreated, the disease can progress to stages that adversely affect the skin and internal organs such as the heart, liver, and nervous system central.[18] Hearing loss can occur because of syphilis, but currently this is rare, and this being most often in the tertiary phase.[3]

Otosyphilis may be present in the form of a sudden and fluctuating sensorineural loss, episodic vertigo, with progressive unilateral or bilateral loss.[11]

Acquired syphilis may also affect the inner ear, simulating Ménière's disease. Hearing loss can progress rapidly progressive, initially with good discrimination; tinnitus and vestibular symptoms disappear to the extent that the destruction of the labyrinth is completed.[9]

Congenital syphilis is due to the hematogenous spread of *Treponema pallidum* of pregnant women who have not been treated or inadequately treated for their unborn child, via the placenta. Transmission can occur at any stage of pregnancy and in any stage of the disease.[22]

Congenital syphilis can cause severe deafness and separately affect both ears. Manifestation occurs when a child is around two years old or between 8 and 20 years old.[9]

4.3. Protozoan infections

4.3.1. Toxoplasmosis

Toxoplasmosis is caused by infection with the obligate intracellular parasite *Toxoplasma gondii*. Both in its acute and in its chronic form, it is related to the appearance of a clinically evident disease, including lymphadenopathy, encephalitis, myocarditis, and pneumonitis.[10]

In immunocompetent individuals, acute toxoplasmosis is habitually asymptomatic and goes unnoticed in 80–90% of adults and children with acquired infection. In the congenital form, the infection of the placenta determines the hematogenous infection of the fetus. The proportion of fetuses that are infected increases as pregnancy progresses, but the severity of the infection declines.[10]

Toxoplasma gondii has been associated with lesion of the auditory pathways with a demonstration of calcium deposits (similar to the calcifications found in the brains of children with congenital toxoplasmosis) in the spiral ligament and the cochlea. A hearing deficit has been reported in about 20% of cases of congenital toxoplasmosis.[23]

5. Final remarks

Hearing loss can interfere with the life of affected individuals because in addition to affecting communication, this can influence the quality of life, on expressing feelings such as sadness and anxiety, or can even lead to social isolation. In infancy, hearing loss can still represent consequences for development.

Thus, proper treatment and/or monitoring of infectious diseases for the purpose of establishing the prevention or early diagnosis of hearing loss is important. With regard to congenital infections, public measures that encourage primary prevention and early identification of these affections in newborns are needed. Therefore, hearing health will depend on epidemiological studies of each location and on a perfect integration between health and education authorities working in an integrated way with all other sectors of society.

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