Human Immunodeficiency Virus and Acquired Immune Deficiency Syndrome AIDS-related Hearing Disorders

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Abstract

After a brief discussion of the nature of the human immunodeficiency virus (HIV)/acquired immune deficiency syndrome (AIDS) disease process and its consequences, the article considers implications for the ear and hearing. One of the newest etiologic considerations for audiologists is pediatric autoimmune deficiency syndrome (PAIDS). Babies born to HIV-AIDS-positive mothers, and children who have acquired the disease, represent a new challenge to clinics. Symptoms, audiologic care, and management are considered.

Key Words: Acquired immune deficiency syndrome (AIDS), hearing disorders, human immunodeficiency virus (HIV)

Since the first descriptions of the acquired immune deficiency syndrome (AIDS) in adults (Centers for Disease Control [CDC], 1981a, b; Gottlieb, 1981), followed by the initial reports of its appearance in children in 1982 (Simonds, 1992), AIDS has been shown to be a disease with multifaceted clinical manifestations. A growing pattern of involvement of the otorhinolaryngologic structures has been widely documented. In the late 1980s, it was reported that 40 percent of patients with AIDS would present with head and neck symptoms (Flower and Sooy, 1987). More recently, Lalwani and Sooy (1992) stated that such involvement approaches 100 percent of the AIDS cases during the course of the disease. It is clear that medical professionals in the field of otorhinolaryngology, as well as medical and nonmedical clinicians in audiology, speech-language pathology, and other related areas of special education, will sooner or later be in contact with AIDS patients, children and adults.

The human immunodeficiency virus (HIV) virus, first identified and isolated by Luc Montagnier from the Pasteur Institute in May 1983, is a RNA retrovirus whose genome has been well characterized (Flower and Sooy, 1987). HIV shows a dense cylindrical core under the electron microscope and encases two molecules of RNA. The external envelope is formed out of two layers of lipids and glycoproteins. The viral particle includes regulatory genes for the replication and pathogenic actions of the virus. HIV has a selective tropism for T4 lymphocyte cells, the helper/inducer subset of T-lymphocytes that express the CD4 phenotypic marker (Fauci, 1988) and are a critical inducer of most immunogenic functions. The resulting depletion causes a significant impairment in the individual immune system. So far, the complex genomic characteristics of the virus, its high mutability rate, and our lack of understanding of its pathogenic mechanisms and of what cofactors fire the replication phase have frustrated most efforts to produce a useful vaccine or to generate more effective antiviric agents.

From the neuro-otologic point of view, the clinician deals with a changing pattern of infections and with a neurologic involvement that compromises the auditory nerve and the hearing abilities of the individual with AIDS, both peripherally and centrally. It is reported that, currently, most infections are caused by common microorganisms, with good response to standard therapeutic measures, and that infections...
by unusual opportunistic pathogens are less common (Lalwani and Sooy, 1992).

**COMMON OTORHINOLARYNGOLOGIC MANIFESTATIONS OF AIDS**

Mainly due to abnormal immune response, but also as a direct effect of the virus on the central nervous system (CNS) and the auditory pathways, we must expect a high prevalence of ear abnormalities, as well as nose and throat findings in the HIV/AIDS patient. Since the initial reports by Sooy (1987), Oleske (1987), and Williams (1987) in the mid 1980s, we have learned to expect as much as 40 percent of head and neck manifestations in AIDS patients, mucocutaneous candidiasis in 31 percent of cases, and 8 percent with rapidly enlarging neck masses (Williams, 1987). The most frequent findings to be originally reported were serous otitis media, acute and chronic otitis media, cervical adenopathy, and parotid enlargement (Williams, 1987). Kaposi's sarcoma can be a common finding in HIV/AIDS patients. It has been reported with a prevalence from 27 percent to 95 percent (Patow, 1992), and, when in the larynx, it can be responsible for voice disturbances. Cervical lymphadenopathy continues to be one of the most common manifestations. Oropharyngeal, esophageal, and laryngeal candidiasis is another frequent finding (Williams, 1987). Hiccups as a presenting symptom has also been reported in AIDS patients with lower esophageal candidiasis (Herrera, 1992). Cytomegalovirus infections are also described in the literature involving the larynx and, again, causing voice disorders that have to be differentially diagnosed (Marelli et al, 1992). Principi et al (1991), in their otitis media study in Milan, described the agents isolated from tympanocentesis in their HIV/AIDS patients as Streptococcus pneumoniae, Haemophilus influenzae, Streptococcus pyogenes, Proteus mirabilis, and Escherichia coli. They also state clearly that such etiologic pattern is similar to the one usually found in acute otitis media (AOM) in normal children in Italy. One interesting aspect of the Milan study is that it does not seem to demonstrate that HIV modifies the general occurrence of acute otitis media in the “P-1” group of children (from the CDC classification for pediatric AIDS), that is, asymptomatic HIV (+), compared with normal children. HIV-AIDS-infected children — the “P-2” group — experienced significantly more episodes of AOM than paired normal controls and P-1 group children. They concluded that HIV infection does not seem to favor the occurrence of AOM per se, but it predisposes to higher recurrence. They also show a greater failure rate to treatment. The pathogenic reasons for higher recurrence in P-2 children remain to be investigated. In Pittsburgh, Haddad et al (1992) have reported a retrospective study in sinuses, ears, and head and neck infections in children with various types of primary immunodeficiencies but with no AIDS. The causative bacteria was no different from the previous references. They stated that ear infections that required hospitalization in this group were caused by community-acquired bacteria. Moreover, only in one case — a scalp abscess culture — an Aspergillus fumigatus was isolated. Currently, it is more infrequent to see infections by rare pathogens (Lalwani and Sooy, 1992).

Probably, the most common pathogen related to HIV/AIDS patients is Pneumocystis carinii, isolated from both pulmonary and extrapulmonary...
sites. The presence of *P. carinii* has been reported in middle ear infections by Park et al in 1992 in mastoiditis (Gherman et al, 1988) and cutaneous pneumocystosis (Coulman et al, 1987), among others. As expected, unusual and opportunistic pathogens have also been described in the literature: otitis media by *Nocardia asteroides* (Forret-Kaminsky et al, 1991), otomastoiditis by *Aspergillus fumigatus* (Strauss et al, 1991), malignant otitis by *Pseudomonas aeruginosa* (Rene et al, 1990), sinusal infection by *Candida albicans* (Poole et al, 1984), and herpes zoster oticum (Mishel and Applebaum, 1990). Lalwani et al (1991) and Sooy (1992) have reported eustachian tube obstruction by nasopharyngeal mass, leading to serous otitis media with effusion.

Any of the already mentioned clinical conditions can present with a mild to moderate conductive hearing loss, which will be characterized by the classic air-bone gap and the expected immittance findings: low-mobility tympanograms, negative pressure in the middle ear, and absence of acoustic reflexes. The overall prognosis of response to conventional antimicrobial therapy is good in general (Kohan et al, 1990), and the hearing problem is expected to be reversed.

**INNER EAR AND CENTRAL NERVOUS SYSTEM: SENSORINEURAL AND CENTRAL HEARING LOSS**

The HIV virus is characterized by lymphotropic and neurotropic activity capable of producing severe neurologic or systemic disease in both adults and children. The virus has been isolated in the brain, spinal cord, cerebrospinal fluid, peripheral nerves, and muscles in the HIV/AIDS patient. Nevertheless, the neurons do not seem to harbor the virus, although it is found in the microglia cells, giant multinucleated cells, and central and peripheral macrophages, as well as in the lymphocytes (Mehta and Kula, 1992).

Over 90 percent of necropsies in AIDS patients show CNS abnormalities (Koralnick et al, 1990), and more is being learned about AIDS polyneuropathy and vascular myelopathy, opportunistic infections in the CNS and the structural and functional correlation of the HIV with brain damage and AIDS encephalopathy, and cranial nerve neuropathy. One of the most fearsome consequences of HIV/AIDS is brain destruction (Koralnick et al, 1990) and dementia as part of the fatal outcome of the disease (Ollo et al, 1991). Estimates of the frequency and degree of motor, cognitive, and behavioral abnormalities in adults with AIDS (group IV, CDC classification for adults) vary widely in the literature (32–78%), while in asymptomatic HIV (+) individuals (group II, CDC), the cognitive abnormalities tend to fall from 5 to 20 percent (Goethe et al, 1989).

The pediatric population infected with HIV seems to be more susceptible to CNS involvement. In 96 percent of children dead from AIDS, the CNS showed some gross or microscopic abnormalities (Kozlowski, 1992). The exact mechanism of CNS destruction is unclear, but Kozlowski suggests that the virus could produce a direct effect on the brain, affect the process of CNS maturation, actually cause opportunistic infection of the CNS and generate lesions that may be primary to HIV infection but whose relation to the virus is unknown (CNS neoplasia) or lesions that are secondary to other organ involvement (hypoxic encephalopathy). In children infected with HIV, the brain involvement continues to develop slowly, even in the absence of other clinical signs. According to Ho (1992), there is no "dormant" or "latent" period for the virus. HIV is in the infected CNS cells with a low rate of duplication. What is yet to be defined is what causes the "phenotypic change" and the firing of the replication high rate state that leads to cell damage and immunosuppression. It is now clear that AIDS is a primarily viral phenomenon and only secondarily an immunologic event (Ho, 1992). This subclinical action in the child's developing brain affects higher mental functions. Developmental language and perceptual delays, loss of developmental milestones, and motor and cognitive deficits are to be expected in the HIV/AIDS pediatric population (Kozlowski, 1992). According to Kastner and Friedman (1988), pediatric AIDS has been recognized as a cause of mental retardation and developmental disabilities, as well as motor and expressive language problems not seen in children without AIDS. In the US, a New Jersey Task Force for the prevention of mental retardation expects pediatric AIDS soon to become a major cause of mental retardation and learning disability in their state (Kastner and Friedman, 1988).

In adults, recent research, exemplified by Ollo et al in 1991, used the known sensitivity of the event related (brain) potentials to examine the integrity of information processing, in order to detect cognitive changes in asymptomatic HIV patients and in patients classified within the CDC groups III/IV. Reduced P300 amplitudes
and increased $P_{300}$ latencies occurred in AIDS cases in response to both auditory and visual stimuli, while in the HIV patients, the finding only took place in the visual modality. As they stated, “The $P_{300}$ results demonstrate alteration in stimulus evaluation and processing speed in the earliest stages of HIV disease, even before the cognitive deficits can be detected by more traditional measures” (Ollo et al, 1991).

In relation to the inner ear and the hearing pathways, the sensorineural involvement of hearing in HIV-positive individuals is reported to be anywhere from 20.9 percent to 49 percent (Lalwani and Sooy, 1992). Kohan’s series (1990) of 18 AIDS patients with otologic complaints such as hearing loss, otalgia, otorrhea, vertigo, and tinnitus in the initial examination demonstrated a 10/18 correlation of CNS and/or otologic involvement, through contrast-enhanced computed tomographic scans of the temporal bone and brain. Sooy (1987) describes a prevalence of 45 percent sensorineural hearing abnormalities in a prospective study with AIDS patients. Smith and Cannalis reported in 1989 a series of five cases where an HIV infection altered the course of a latent syphilis and favored the development of otosyphilis, with unilateral or bilateral hearing loss, tinnitus, ear pressure sensation, and labyrinthine symptoms. Vertigo and other neurootologic findings are not frequently described in the literature, except as part of the terminal phase of AIDS. A well-documented case was reported by Hart et al (1989). The patient’s pattern of dizziness, dysequilibrium, and emotional disturbances, along with the alteration of his optokinetic nystagmus and saccadic pursuit with a total abolition of the caloric responses, showed in the autopsy an anatomopathologic correlation. It was possible to demonstrate cortical, subcortical, cerebellar and brainstem lesions consistent with the known histopathologic findings of HIV.

Regarding the electrophysiologic changes in the auditory brainstem response (ABR) of HIV/AIDS patients, Pagano et al (1992) reported a review in which all cases had normal audiograms, but ABR testing showed central conduction times (latency in the I–V and III–V intervals) significantly longer than normal controls (non-HIV). Similar findings are reported by Welkoborsky and Lowitzsch (1992). The absolute and interwave latencies were prolonged.

There are a few reports of sudden sensorineural hearing losses in HIV/AIDS patients. The first report was made by Real et al (1987). They describe a homosexual male patient (group IV, CDC) who developed a cervical and inguinal adenopathy, a Pneumocystis carinii pneumonia, and a cryptococcal meningitis. After a symptom-free period of 5 months, he developed a sudden sensorineural unilateral drop in hearing, evolving to a massive cerebral hemorrhage, and he eventually died. Timon and Walsh (1989) presented two sudden deafness cases in HIV-positive patients with no overt AIDS (group II, CDC). Both cases were unilateral, and one showed an additional left-sided facial palsy with complete remission of the audiologic findings without a clear explanation. Kwartler et al (1991) describes the anatomopathologic temporal bone changes in a full-blown AIDS case, although the sudden deafness was associated with a cryptococcal meningitis. The meningeal infectious complication in AIDS is not infrequent, and the incidence of residual hearing loss is reported as high as 27 percent (Maslan et al, 1985). In 1992, Madriz reported an AIDS case with bilateral sensorineural moderate hearing loss, of sudden onset, rapid progression, and no otherwise explained origin. Due to the effect of HIV in the CNS, another aspect of HIV/AIDS hearing disorders is being documented in those cases with a more central origin of the disturbance. We would expect the virus to affect the cortical and subcortical areas of the temporal lobe as well as the diencephalic relays of the auditory pathways. We also should expect such patients to have a disproportionate inability to discriminate language in relation to their tonal audiometric curves, which worsen as the signal-to-noise ratio becomes more negative. Strutz (1991) stated that AIDS should be considered a new cause of acquired central auditory disfunction. Madriz reported in 1992 a case whose speech-tonal discrepancies suggested a more central component, given the very mild sensorineural high-frequency sloping audiometric profile. As the negative signal-to-noise ratio was incremented in the Synthetic Sentences Identification Test (Jerger et al, 1968), the performance versus intensity function deteriorated significantly. The case could be explained as a central auditory signal processing disorder, the etiology of which was attributed to HIV/AIDS.

**AIDS AND THE PROFESSIONAL IN COMMUNICATION DISORDERS**

As the AIDS epidemic expands, and as we are able to document an increasing otorhinolaryngologic involvement in the HIV/AIDS patient — both in adults and children — there
is a growing awareness that, as professionals in the field of communication disorders, we will soon become deeply involved with HIV/AIDS and its consequences in hearing, voice, speech-language, and learning disabilities.

From the audio-otologic point of view, we can expect to find in the AIDS patient sensorineural abnormalities of hearing, labyrinthine manifestations, and facial palsy as a result of CNS and cranial nerve involvement, as well as central auditory processing disturbances, as the cortical ability to manage the acoustic linguistic signals is deteriorated due to the HIV lesions in the CNS. From the speech pathology point of view, we must consider organic lesions of the Kaposi’s sarcoma type that can be responsible for disturbances in the laryngeal function, with secondary dysphonia. In the US, Pressman reported in 1992 an overall prevalence of 12.5 percent of voice disorders in a group of 96 HIV/AIDS children from 4 months to 17 years of age. Hull et al (1976) had reported a prevalence of 3 percent of voice disorders in the school-aged population. Voice problems have also been related to vocal chord paralysis (Pressman, 1992) and Candida infection in AIDS patients. Intubation and artificial respiratory means can also condition the development of additional voice disorders. Dysphagia has also been reported (Pressman, 1992). The few reports available do not support a significant influence on speech disorders in the young AIDS patient. In the Pressman series, loss or regression in expressive language or dysarthria was found in 7.3 percent of cases, in correlation with progressive encephalopathy. Articulation deficits were seen in 27 percent of cases, mild to moderate in degree, and considered developmental errors, because almost 80 percent of this subgroup overall was developmentally delayed.

AIDS patients in terminal phases, in common with the average, terminally ill patient with severe respiratory failure who is dependent on assisted respiratory units, are confronted with devastating communication barriers. Intensive care unit personnel should become familiar with and develop lipreading skills (Flower and Sooy, 1987). Under these and other less dramatic circumstances, the speech-language pathologist will have to consider assistive devices such as the electric artificial larynx or augmentative communication devices with electronic amplification of voice signals. Weak AIDS patients, although not intubated, may be unable to overcome the environmental noise of a support system, and would benefit from small amplifiers to facilitate basic linguistic exchange. Audiologists will have to prescribe hearing aids and assistive listening devices and will have to learn about specific counselling, orientation, and support of these patients.

Ear, nose, and throat (ENT) surgeons and operating room personnel must implement firm preventive measures to minimize inadvertent transmission of HIV. ENT and audiologic clinicians will have to learn about universal body fluid management and precautions to avoid contamination (blood, purulent secretions, etc.) (Davidson and Stabile, 1991).

CONCLUSIONS

A large proportion of the patients infected with HIV do present with medical problems in the head and neck region. For that reason, medical specialists in otorhinolaryngology and audiologists will play an important role in its diagnosis and treatment (Tami and Wawrose, 1992). The most important audio-otologic manifestations of HIV/AIDS include the ear’s Kaposi’s sarcoma, otitis by Pneumocystis carinii, eustachian tube obstruction by nasopharyngeal mass, serous otitis media, abnormal auditory response in the brain stem, sensorineural hearing loss, and facial palsy (Lalwani and Sooy, 1992).

Given the described audio-otologic and communicative implications of AIDS, audiologists, speech-language pathologists, and other special education professionals will have to participate in initial evaluations that could lead not only to the diagnosis of HIV/AIDS, but also to the rehabilitative efforts to compensate HIV-AIDS-related hearing, voice, speech-language, and learning problems, both in children and adults. We also must expect to have our special education facilities increasingly populated with HIV/AIDS children. Preventive measures will have to be stressed, not as much for ourselves, but because AIDS patients can be extremely vulnerable to infections carried by their caregivers. Learning about AIDS, and learning to live with the AIDS patient, will soon become a new challenge and a new responsibility in our daily clinical routine.

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