OPTIC ATROPHY IN CHILDREN ASSOCIATED WITH PROGRESSIVE HEARING LOSS.CLINICAL CASE AND REVIEW OF LITERATURE

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Introduction: Optic atrophy is one of the most common cause of vision loss in children, the differential diagnosis, clinical investigation and genetic testing are of extreme importance to rule out secondary optic atrophy with potentially reversible processes.
Aim of study. The report aims to describe the genetic and clinical characteristic of hereditary optic neuropathies through the presentation of a case of dominant optic atrophy with loss of visual acuity associated later with progressive bilateral neurosensory hearing loss.
Methods: We summarize current literature, describe genotype and clinical aspect correlations .A systematic literature search was conducted in electronic database PubMed /Medline , and Cochrane Library The analysis of existing literature has been conducted.
Conclusion: Autosomal Dominant Optic Atrophy and Deafness represent a syndromic form of Dominant Autosomal Optic Atrophy, To date OPA 1 is the major gene responsible for Dominant Optic Atrophy accounting for 80% of all the patients. So understanding the molecular pathogenesis of OPA1gene linked to DOA may elucidate many other mitochondrial diseases connected later in life with neurodegenerative progression of glaucoma, Parkinson, dementia and others.