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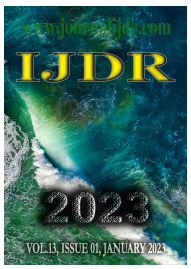

RESEARCH ARTICLE OPEN ACCESS

USHER SYNDROME

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ABSTRACT

Usher syndrome is the one among the rare genetic disorder it is characterized by sensory impairment of the auditory and visual system with various level of vestibular dysfunction. This syndrome born when child is born and it is usually found out during child hood itself. Usher syndrome has three types and it has different levels of impairment in hearing and vision problem. This syndrome cannot able to cure because there is no specific treatment but research is going on and using hearing aids and cochlear implants helps to improve the quality of life of the persons and be as independent.

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INTRODUCTION

Usher syndrome is first diagnosed in the year 1858 , by Albrecht von Graefe , he was named that Charles usher and a Scottish eye doctor find out this condition is caused by hereditary means recessive gene. Among 100,000 children 4 to 17 are born with usher syndrome and this 3% to 6% children were born with deaf in this both parents have responsible to cause it.Usher syndrome has three types Type 1, Type 2, Type 3, it is causing various level of hearing and visual loss and some children may show balance problem. Usher syndrome Type 1 and Type 2 are more common in most of the countries. And it is more common in Eastern and central European and French and French Acadian.

Usher Syndrome is a genetic condition characterized by partial or total hearing and vision loss and also some may experience by balance problems.

Etiology it is caused by mutations in several different types of genes. Type 1 MYO7A gene mutations, Type 2 USH2A gene, and Type 3 USH3A gene. Most of the conditions find out two copies of gene one from the mother and other one are received from the father, and high risk in consanguineous marriages.

Genemutation affects the nerve cells in cochlea so they don't transmit sound. Due to a retinitis pigmentosa affects the rods and cons.

Types are in the usher syndrome are TYPE 1in this babies are born with severe hearing loss or deafness , some children may develop balance problems and until they are not reach 18 months they will not start walking , children's when they reach 10 years of age vision starts getting loss with night vision as age increases condition became worsen. TYPE2 here children's born with hearing loss it may be moderate to severe. Vision problem start at the age of teenage and start with worsen and children don't have balance problems. TYPE 3 it is very rare observe only 2% cases, at birth both hearing and vision loss but hearing loss usually begins in late childhood. But vision loss starts in early to mid-adult hood, usually start with night blindness and balance problems.

Clinical Manifestation Symptoms varied from types of usher syndrome Hearingloss; observe at birth it may be moderate to severe. Vision loss; due to a retinitis pigmentosa, difficulty in seeing in low light (night blindness) peripheral vision also loss. Balance problem; due to damage to ear and eye can lead to problem with balance and coordination.

Complications are delay in sit and walk without support, always need extra help from the others means depended to others.

Diagnosis Genetic test, Hearing tests, Vision tests (peripheralvision).

Management At present no specific treatment for cure this, only supportive treatment using hearing aids, cochlear implant, using glasses, vitamin A supplementation for improving the ability to communicate and independently.

Prevention Usher syndrome can cause due to gene mutations. So through genetic testing and counseling can prevent.

Conclution There is no cure for Usher syndrome, but persons can be improve through hearing and vision therapy with early diagnosis and treatment, getting the correct assistant to the child can lead a life full active, easy to communicate and stay independent.

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