INTRODUCTION
For humans, hearing is an important mechanism for communication. Its importance is rarely appreciated until it is impaired or lost. Hearing impairment affects the speech and language acquisition.

Anatomically, human ear can be divided into three distinct compartments-the outer, middle and inner ear. The outer ear or pinna collects the sound waves and channels them down the ear canal to vibrate the ear drum. The vibrations are transmitted as mechanical waves by the three ossicles of middle ear and transfer into oval window of inner ear where they are converted to nerve impulses. These impulses are transmitted along the auditory nerve to the brain and interpreted as sounds. Inability to perceive the sounds is more appropriately called as hearing impairment/hearing loss (Petit et al., 2001).

Hearing impairment can be classified based on several criteria: Type of ear defect-conductive (outer or middle ear defects), sensorineural (anomaly in the pathway from inner ear to cortical auditory centers of the brain) or mixed hearing impairment; degree of hearing loss-mild (25-40dB), moderate (45-60dB), severe (65-85dB) or profound (>85dB); age at onset-prelingual (early enough to impair
language acquisition) or postlingual. Finally, the hearing impairment could be classified as non-syndromic or syndromic (Kalatzis and Petit, 1998).

Prelingual forms of hearing loss are the most severe forms. One in 125 children are affected by severe or profound hearing impairment during early childhood in India (ICMR, 1991). Impairment during prelingual period is always of the nerve type (Petit et al., 2001). About fifty percent of prelingual cases are thought to be due to environmental factors and the remainder to genetic causes. Examples of the former include acoustic trauma, ototoxic drugs (i.e., Amino glycosides, Cisplatin, Gentamicin etc.) and bacterial or viral infections such as rubella or cytomegalovirus (CMV; Bussoli et al., 1998).

Approximately 70 percent of congenital genetic cases are classified as non-syndromic. The remaining 30 percent are accounted by more than 400 syndromic cases (Cryns and Van Camp, 2004). Usher’s, Pendred and Waardenburg are the most common forms of syndromic cases of hearing impairment. The auditory pathology varies widely among syndromic cases and includes both conductive and sensorineural deficits; unilateral or bilateral, symmetrical or asymmetrical and progressive or stable (Resendes et al., 2001).
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One can observe almost all modes of inheritance in cases with congenital non-syndromic hearing impairment. Approximately 77 percent are transmitted as autosomal recessive, 22 percent as autosomal dominant and one percent as X-linked. Less than one percent is due to mitochondrial inheritance. Their proportion may vary by 10 to 20 percent in different populations (Jacobs et al., 2005). As a general rule, individuals with autosomal recessive non-syndromic hearing impairment have severe-profound prelingual deafness, while dominant mutations lead to a more variable phenotype (Nance, 2003).

Over the past one decade, remarkable progress has been made in identifying the hearing impairment loci and cloning the genes for deafness. More than 120 independent genes for deafness have been identified (Nance et al., 2003). To date (www.uia.ac.be/dnalab/hhh/accessed, Aug 15, 2005), around hundred twenty-three hearing impairment loci have been mapped (autosomal dominant-54, autosomal recessive-61 and X-linked-8). More than 100 genes involved in causing syndromic hearing impairment. Some of them are PAX3, MITF, EYA1, SOX10, EDN3, USH2A, USH1C, Myosin7A, Harmonin, NF2, and PDS. In some instances, different mutations at the same locus have been found to cause both syndromic and non-syndromic forms of deafness (Steel et al., 2001).
Identification of deafness genes enables us to understand the molecular process of hearing and this offers prospects of DNA testing for the hearing impairment. Although a large number of genes are involved in deafness, in many populations DFNB1 locus (GJB2; 13q11-q12) encoding Connexin26 (Cx26) protein is the most frequent cause of recessive deafness (Cryns and Van Camp, 2004).

Connexins are a family of proteins that form intercellular gap junction channels. They allow ions and small molecules to move between adjacent cells. In humans, at least twenty-two connexin genes have been described (www.genecards, 2004). Many of them harbour germ-line mutations and are associated with a variety of human diseases (Common et al., 2004).

Connexin 26 protein mainly expresses in the supporting cells of the cochlea and maintains the local circulation of the potassium ion between the fluids of inner ear (Kikuchi et al., 2000). Mutations of Cx26 gene mostly cause non-syndromic hearing impairment (NSHI). There are some ethnic-mutations that account for the majority of Cx26 related hearing loss. For example, 35delG in Caucasians, R143W in Africans, 167delT in Ashkenazi Jews, 235delC in Japanese and W24X in Indians (Alvarez et al., 2005).
Therefore, epidemiological studies on the mutational spectrum of Cx26 gene and their frequency in a given population are always useful in molecular diagnosis and genetic counseling.

The aim of the present study is to study the contribution of Cx26 gene mutations to the childhood hearing impairment among children who would be attending special schools for the hearing impaired with particular reference to Tamilnadu, South India. This study also aims at estimating the carrier frequency for the most common pathogenic mutation among normal hearing persons.