INTRODUCTION

Hearing is the mean by which the newborn comes into contact with the world of sound and with language structures. It is through oral language that humans are able to make contact with their fellowmen, and develop the ability to share their experiences, thoughts and ideas in the search for new knowledge ***(Lima et al., 2006).***

Normal speech and language development depend upon a child's ability to hear spoken language. Early infancy is the most appropriate time for a child to acquire the foundation of language and communication. The most important period for language and speech development is generally regarded as the first 3 years of life. Therefore, early detection and early identification of hearing loss is very important. This should be followed by a timely and effective therapeutic intervention and rehabilitation programs to minimize the negative effects of hearing loss on the development of cognitive, psychosocial and verbal communication skills and social interactions ***(Moeller , 2000).***

The impact of hearing loss on early language development has been well documented. Although published studies on efficacy of early intervention are more limited, the majority demonstrates that children with hearing loss who

received early intervention had higher score of improvement on acquired language than those not connected to service early ***(Hille et al., 2007)***.

Early identification followed by proper intervention as early as 6 months of age results in essentially normal language acquisition later on and minimize the negative effects of hearing loss. In contrast, a delay in detection of up to 2 to 4 years may result in abnormal language acquisition ***(Flynn et al., 2004).***

The hearing loss risk indicators, as well as the use of objective methods for performing hearing screening and follow-up were established and reviewed by the Joint Committee on Infant Hearing**,** Risk factors for hearing loss including family history, in utero infections, craniofacial anomalies, birth weight <1500 g, hyperbilirubinemia at serum levels requiring exchange transfusion, ototoxic medications, bacterial meningitis, postnatal asphyxia, mechanical ventilation lasting 5 days or longer, stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductional hearing loss However, the significance of various aetiological factors in the pathogenesis of sensorineural hearing loss is not fully understood and controversy remains regarding their respective roles ***(JCIH, 2000).***

Mild to moderate oxygen deprivation at birth temporarily depresses auditory function. More severe respiratory compromise associated with hypoxic-ischemic encephalopathy (HIE), prolonged ventilation, and persistent pulmonary hypertension increases the risk of a permanent hearing loss ***(Garcias et al., 2004).***

The diagnosis of hypoxic-ischemic encephalopathy was defined by the presence of clear-cut neurological signs of neonatal encephalopathy as depressed level of consciousness, hypotonia and hypoactivity (neurological examination compatible with profiles 2, 3 or 4) associated with documented birth asphyxia (depressed Apgar scores, need of immediate resuscitation in the delivery room and, when information available, cord blood acidosis) and / or neuroradiological evidence of hypoxic-ischemic lesions ***(Garcias et al., 2004).***

Neonatal seizures are usually an acute manifestation of disturbance of the developing brain and are very common in the first weeks of life. Its incidence varies from 1-5 per 1000 live births ***(Van Rooij et al., 2010).***

Neonatal seizures have been associated with adverse neurologic outcomes. Some suggest that seizures themselves cause brain injury and contribute to adverse outcome and others maintain that the underlying cause of seizures, such as asphyxia,

hemorrhage, or infection, is the primary contributor to poor outcome ***(Mizrahi and Clancy, 2000).***

Early detection of hearing loss makes it possible to refer positive cases for medical therapy and rehabilitation programs ***(Ciorba et al., 2007).***

The Joint Committee of Infant Hearing recommends identifying children with hearing loss by universal hearing screening at the moment children are discharged from hospital or within the first month of life. If screening tests are positive, children should be referred to the appropriate medical expert and a speech therapist. A test battery is then undertaken to confirm the diagnosis of hearing loss; this diagnosis should be made by the third month of life, and therapy should be started by the sixth month of life ***(JCIH, 2007).***

AIM OF THE WORK

1- To determine the prevalence of hearing loss during the study period.

2- To determine distribution of associated risk factors in newborns with hearing loss.

3- To assess the effect of hypoxia and/or convulsions on auditory function of neonates.

Research Questions / Hypothesis

1- What is the prevalence of hearing impairment in Special Care Nursery?

2- What are the common risk factors in newborns with hearing loss?

3- Can hypoxia and/or convulsions affect auditory function of neonates?

HEARING

Early Hearing Detection and Intervention (EHDI) programs for infants have been implemented with universal newborn hearing screening and treatment for congenital hearing loss in many developed countries.

As per Yoshinaga-Itano (2003), there is a critical period for early language development within the first six months of life. This sensitive period derives from behavioral and neurological factors that can affect timelines for aspects of development, such as socio-emotional, auditory, speech, and language ***(Yoshinaga-Itano, 2004).***

Permanent childhood hearing impairment in children affects the development of auditory speech perception, speech production, and English language acquisition ***(Yoshinaga-Itano, 2003).***

Permanent childhood hearing impairment has prevalence rates of 1 to 2 per 1000 live births as defined by 40 decibels (moderate) or worse hearing loss in the better ear ***(Fortnum, 2003).***

However, when mild hearing loss cases are included, with a definition of loss greater than 25 dB unilaterally, the prevalence is about 2-3/1000 ***(Hyde, 2005).***

The severity of hearing impairment varies and there are challenges in identifying some permanent childhood hearing impairments, such as progressive or late onset hearing loss ***(Hutt & Rhodes, 2008).***

Universal newborn hearing screening programs result in earlier diagnosis of hearing loss and early detection enables early intervention services for affected families ***(Yoshinaga-Itano, 2003).***

I. Anatomy of the auditory system:

Auditory experience begins with a hearing ear. The role of human ear is to convert sound waves into nerve impulses. The peripheral auditory pathway is divided audiologically into a conductive and a sensorineural pathway ***(Stinmeiz Sand Leel, 2006).***

The conductive part consists of the pinna, external auditory canal (EAC) and the middle ear. Sensorineural pathway includes the cochlea and part of the eighth cranial nerve (Fig. 1) ***(Isaacson and Vora, 2003).***

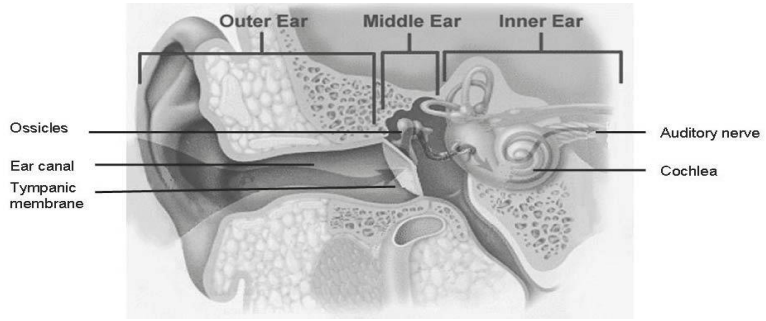

Fig. 1: Overall structure of the human ear

1. The external ear:

Two structures make up the external ear: a flexible, oval shaped structure "pinna" attached to the head, and the auditory canal that leads to the middle ear ***(Lalwani, 2007).***

The pinna is the folds of cartilage surrounding the ear canal. Sound waves are reflected and attenuated when they hit the pinna, and these changes provide additional information that will help the brain determine the direction from which the sounds came (Fig. 2) ***(Lalwani, 2007).***

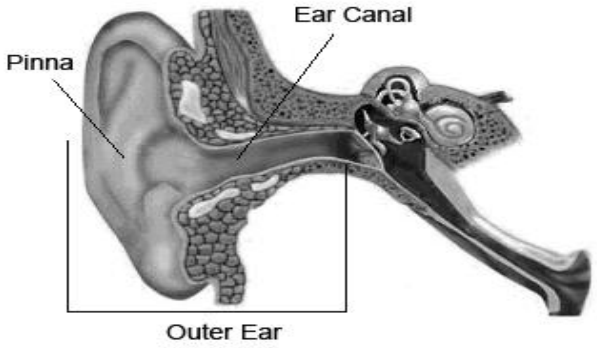

Fig. 2. The outer ear and its main elements.

The sound waves enter the auditory canal: it consists of a lateral cartilaginous portion and a medial bony portion. The tragus forms the anterior cartilaginous canal, directly in front of it lies the parotid gland. The facial nerve exits the stylomastoid foramen 1 cm deep to the tip of the tragus ***(Stinmetz and Lee, 2006).***

Within the anterior and inferior portions of the cartilaginous ear canal, there are small fenestrations through the cartilage called the fissures of Santorini. Infection of the ear canal (otitis externa) can spread to the parotid gland through these fissures and may lead to skull base osteomyelitis**.**The tympanic portion of the temporal bone forms most of the bony ear canal. Anterior to the bony canal is the temporomandibular joint ***(Stinnetz and Lee, 2006).***

The skin of the ear canal is thicker in the cartilaginous canal and contains glands that secrete cerumen (ear wax). The skin of the bony ear canal is very thin and fixed to the periosteum. No cerumen is secreted in the bony ear canal ***(Haddad, 2007).***

The main function of EAC is to collect and transmit sounds to the tympanic membrane (TM) located at its medial end and marks the beginning of the middle ear. The tympanic membrane forms a barrier between the EAC and the middle ear.

It serves as a resonator through which sound waves are transmitted to the manubrium of the malleus, which is attached to its medial side ***(Stinmetz and Lee, 2006).***

Vibrations of the tympanic membrane are dampened almost immediately once the sound wave stops. The loudness of the sound correlates with the amplitude of the sound waves ***(Haddad, 2007).***

2. The middle ear cavity and eustachian tube:

The middle ear has a volume of approximately 2 cc (fig. 3). The eustachian tube is 17 to 18 mm length at birth and 35 mm in adult. It is horizontal at term but achieves a 45-degree inferior incline to the pharyngeal orifice in adult ***(Stinmetz and Lee, 2006).***

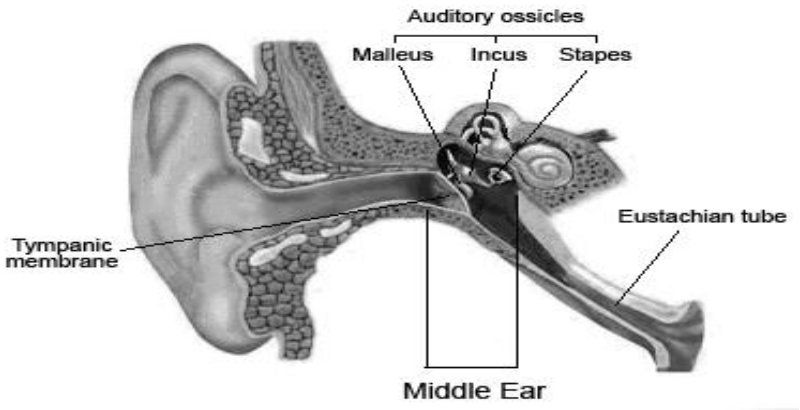

Fig. 3. The middle ear and its main elements.

The eustachian tube is lined by respiratory epithelium and surround for a short distance near the middle ear by bone, but for most of its length it is surround by cartilage ***(Stinmetz and Lee, 2006).***

The opening of the eustachian tube protects the middle ear from nasopharyngeal secretions, provides drainage into the nasopharynx of secretions produced within the middle ear, and permits equilibration of air pressure with atmospheric pressure in middle ear ***(Doyle et al., 2004).***

The tube is normally closed at rest and opens with swallowing by action of the tensor veli palatine which runs from the skull base and inserts laterally into the soft palate ***(Nffenegger, 2008).***

Thus a cleft palate patient will have poor function of eustachian tube with the aberrant insertion of the tensor veli palatine muscle ***(Nfffenegger, 2008).***

Ossicles: Sound waves traveling through the ear canal will hit the tympanic membrane. This wave information travels across the air-filled middle ear cavity via a series of delicate bones which are the main content of the middle ear cavity; the malleus (hammer), incus (anvil) and (stapes) (fig. 4) ***(Nffenegger, 2008).***

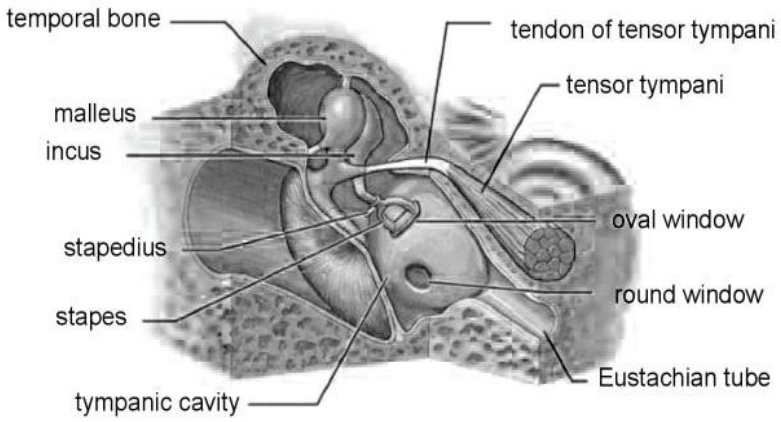

Fig. 4. The middle ear space and ossicle chain

These ossicles act as a lever and a teletype, coverting the lower-pressure eardrum sound vibrations into higher- pressure sound vibrations at another, smaller membrane called the oval window ***(Lalwani, 2007).***

The malleus articulates with the tympanic membrane via the manubrium, where the stapes articulates with the oval window via its footplate. Higher pressure is necessary because the inner ear beyond the oval window contains liquid rather than air ***(Lalwani, 2007).***

The sound is not amplified uniformly across the ossicular chain, The stapedius muscle reflex of the middle ear muscles helps protect the inner ear from damage. The middle ear still contains the sound information in wave form; it is converted to nerve impulses in the cochlea ***(Oghalai, 2004).***

3. <u>The inner ear:</u>

The inner ear consists of the cochlea and several non-auditory structures. The cochlea: It has three fluid-filled sections, and supports a fluid wave driven by pressure across the basilar membrane separating two of the sections (Fig.5). Strikingly, one section, called the cochlear duct or scala media. contains an extracellular fluid similar in composition to endolymph, which is usually found inside of cells ***(Oghalai 2004).***

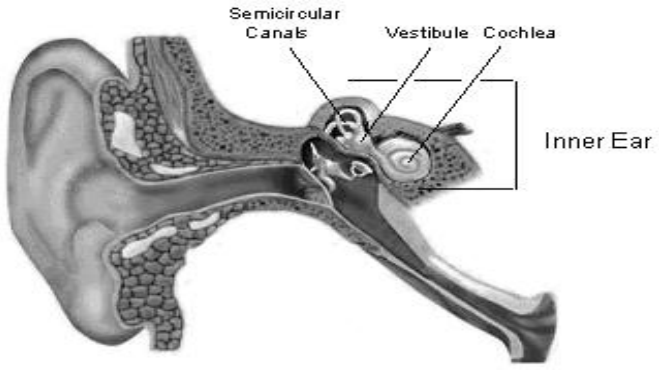

Fig. 5: Elements of inner ear.

The other two sections are known as the scala tympani and the scala vestibuli; these are located within the bony labyrinth which is filled with fluid called perilymph ***(Lalwani, 2007).***

This motion of inner ear fluid excites the nerve cells in the organ of corti. producing electrochemical impulses that are

gathered together and transmitted to the brain along the cochlear division of eighth cranial nerve ***(Fettiplace and Hackney, 2006).***

The chemical difference between the two fluids (endolymph and perilymph) is important for the function of the function of the inner ear.

Within the cochlea lies the most important component of hearing, the organ of corti. It consists of a series of epithelial function of the external ear, middle ear, inner ear (cochlea) and ascending brainstem pathways (fig. 6) ***(Haddad, 2007).***

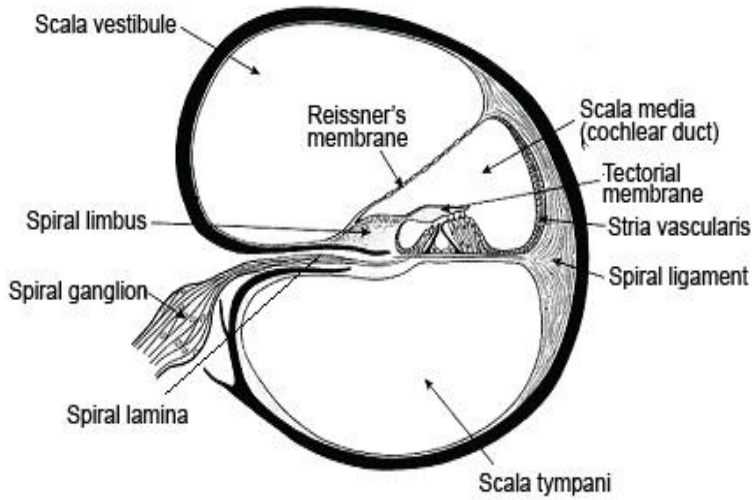

Fig. 6: Cross section of the cochlea and the structure of the organ of Corti.

The hair cells are disposed in a single row of inner hair cells and three or four rows of outer hair cells. When vibrations created by sound waves reach the inner ear causing the

membranes over its openings to vibrate, this cause the fluid in the inner ear to be set in motion (fig. 7) ***(Fettiplace and Hackney, 2006).***

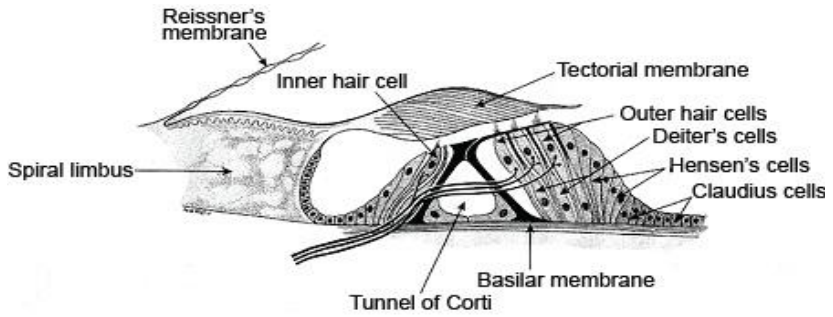

Fig. 7: Organ of corti.

This motion of inner ear fluid excites the nerve cells in the organ of corti, producing electrochemical impulses that are gathered together and transmitted to the brain along the cochlear division of eighth cranial nerve ***(Fettiplace and Hackney, 2006).***

II. Physiology of hearing:

The process of normal human hearing requires proper function of the external ear, middle ear, inner ear (cochlea) and ascending brainstem pathways ***(Haddad, 2007).***

The process of hearing is initiated as sound pressure waves travel through the external auditory canal and vibration of the tympanic membrane. The ossicular chain in the middle ear space then transmits the acoustic energy to the cochlea ***(Kung, 2005).***