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Newborn hearing screening: examining the presence of risk factors for hearing loss

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NEWBORN HEARING SCREENING: EXAMINING THE PRESENCE OF RISK FACTORS FOR HEARING LOSS

by

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INTRODUCTION.

Hearing is one of our most precious senses, facilitating communication through the use of language. Although somewhat treatable through amplification, hearing loss in children inhibits the development of speech and language. It has been established that hearing loss occurs in approximately 3 infants per 1000 (Finitzo et al, 1998; Downs, 1995). In comparison to other conditions, hearing loss is one of the most common congenital anomalies. In fact, hearing loss has been estimated to occur more than twice as often as other disorders often screened at birth including phenylketonuria (PKU), hypothyroid disease, and sickle cell disease (Finitzio and Crumley, 1999). Many states mandate neonatal screens for these conditions. It is true that conditions such as hypothyroidism may have more serious medical ramifications than hearing loss, but the developmental, social, and academic ramifications of hearing loss must not be overlooked when they can be prevented through early identification and intervention. Screening tests such as the automated auditory brainstem response test (AABR) and otoacoustic emissions (OAE) have significantly reduced the cost of screening. Universal screening for hearing loss is a small cost when compared to the amount of funding required for special education when treatments are available which help speech and language development. There is a large body of evidence showing the need and efficacy of early intervention for hearing loss. The most well-known data indicates that infants with hearing loss who receive early intervention (before six months) achieve language development on a level matching their level of cognition (Yoshinaga-Itano, 1995; Yoshinaga-Itano, Dedey, Coulter & Mehl, 1998). Currently, 38 states have passed legislation mandating universal newborn hearing screening (UNHS), with several additional states pending legislation. The Joint Committee on Infant Hearing (JCIH), in the 2000 position statement, defines the goal of UNHS programs as targeting "permanent bilateral or unilateral,

sensory or conductive hearing loss, averaging 30 to 40 dB or more in the frequency region important for speech recognition (approximately 500 through 4000 Hz)."

BACKGROUND INFORMATION: RISK FACTORS.

Before the advent of UNHS, a high-risk registry (HRR) was used to identify infants who were at an elevated risk of having hearing loss. The JCIH compiled a list of risk factors for which hearing screening should be performed during the first two months of life. Factors that placed an infant on the HRR were originally limited to: hereditary childhood hearing loss, rubella, craniofacial anomalies, low birth weight, and elevated bilirubin levels. Over the years, JCIH has updated the registry to include bacterial meningitis, asphyxia, ototoxic medications, mechanical ventilation, and syndromic stigmata. Although the HRR is still recommended when UNHS is not possible, it is not widely used because 50% of hearing impaired infants do not have any risk factors (Mauk et al, 1991). The HRR did not identify children for audiologic monitoring if they did not present with a risk factor at birth. It is apparent why the HRR is not an adequate tool to screen for hearing loss when used alone, as half of the hearing-impaired babies would be missed. Despite its lack of sensitivity, the HRR model helped to build a strong foundation for UNHS models. Perhaps the most valuable lesson learned from the HRR is the use of risk factors to identify babies at risk for progressive hearing loss. A strong recommendation for audiological follow-up for at-risk neonates has become part of the standard of care. Due to a body of research which focused on progressive hearing loss, the JCIH (2000) currently recommends audiologic monitoring (every six months until age three) for infants in the following cases: parental concern regarding speech and hearing, family history of permanent childhood hearing loss, stigmata or other findings associated with hearing loss, postnatal infections associated with sensorineural

hearing loss, in utero infections, neonatal conditions such as hyperbilirubinemia and persistent pulmonary hypertension, syndromes associated with progressive hearing loss, head trauma, or recurrent otitis media for at least three months. Furthermore, infants cared for in a neonatal intensive care unit (NICU) may be at risk for auditory brainstem dysfunction. Kraus and colleagues (1984) described several cases in which peripheral hearing was near normal or moderate while ABR data was abnormal. A majority of these patients had a medical history of asphyxia, hyperbilirubinemia, or perinatal intracranial hemorrhage. Admission to the NICU for greater than 48 hours is now a risk factor included on many state forms, including Missouri.

It is helpful to understand the mechanisms which cause hearing loss in infants due to the conditions which label them at risk, as well as how often hearing loss occurs in these infant populations. There is a large body of research focusing on risk factors for hearing loss, but until UNHS, it was often unclear exactly how often these risk factors resulted in hearing loss. Incidence and prevalence data is more common in recent years as UNHS programs are assessed. Epstein and Reilly (1989) estimated that 10 to 12% of all infants have a risk factor for hearing loss, with up to 5% being identified with sensorineural hearing loss. Data from a large, multicenter study funded by a grant from the National Institute on Deafness and Other Communicative Disorders identified the prevalence of risk factors in both the well baby and NICU nurseries. Particular risk factors found in the NICU include ototoxic medications (44%), low birth weight (18%), ventilation (16%), and low Apgar scores (14%), while the most common risk factors in the well baby nursery were family history of hearing loss (7%), craniofacial anomalies (3%), and low Apgar scores (3%) (Vohr et al, 2000). This group also found 90% of NICU babies had other risk factors in addition to admission to the NICU for more than two days. The same group found that 30% of babies had risk factors in the well baby nursery (Prieve et al,

2000). Another study conducted by Mason and Herrmann (1998) considered all NICU infants to have a risk factor for hearing loss, while less than two percent of babies in the well-baby population were identified with a risk factor. A review of incidence data estimates hearing loss occurs in about 3 per 1000 live births in the universal population, ranging from 0 to 1.2 per 1000 for well baby populations, and ranging from 5 to as high as 60 per 1000 births in the NICU and high risk population (Mason and Herrmann 1998).

OBJECTIVES OF THE STUDY.

The purpose of this study was to determine which risk factors were the most common in the well baby nursery (WBN) and NICU, which risk factors were most likely to result in congenital hearing loss, and how many of the babies identified with hearing loss also had a risk factor.

METHODS.

For this project, data from St. John's Mercy Medical Center (SJMMC) were examined with regard to risk factors and incidence of hearing loss in a well baby, well baby nursery. A six month sample of babies admitted to the hospital's NICU was also included for comparison. The screening protocol (see Appendix A) requires documentation of risk factors prior to testing through chart review. Infants at high risk for hearing loss are administered an automated auditory brainstem response test (AABR). If a high-risk infant refers, the baby is referred for a diagnostic ABR following discharge from the hospital and within the first few months of life. If a high-risk infant passes, the screening process is complete. However, audiologic monitoring at six to eight months of age is recommended for infants with risk factors due to the higher

possibility of progressive hearing loss. Risk factors indicating need for follow up are the risk factors listed by JCIH (2000) as described in the list below. When risk factors are present a letter is sent to the parents when the child is six months of age, reminding them that follow-up is recommended. In all cases, before being discharged, parents receive an informational pamphlet from the state about UNHS, a copy of their baby's screening results including which tests were used, a list of risk factors for progressive childhood hearing loss, and a copy of normal speech and language milestones. This procedure is in accord with the families' right to be informed as outlined by JCIH (2000).

RISK FACTORS REQUIRING AUDIOLOGICAL FOLLOW-UP:
Joint Committee on Infant Hearing, Position Statement (2000)

- Parental concern regarding speech, language, and hearing.
- Family history of permanent childhood hearing loss.
- In utero infections such as toxoplasmosis, rubella, cytomegalovirus, syphilis, herpes.
- Neonatal conditions: hyperbilirubinemia, persistent pulmonary hypertension requiring ventilation, extracorporeal membrane oxygenation.
- Syndromes associated with progressive hearing loss such as: neurofibromatosis, osteopetrosis, Usher syndrome.
- Neurodegenerative disorders such as Hunter syndrome, Friedreich's ataxia, Charcot-Marie-Tooth syndrome.
- Postnatal infections associated with hearing loss including bacterial meningitis.
- Stigmata or findings associated with a syndrome known to include hearing loss.
- Head trauma.
- Recurrent or persistent otitis media with effusion for at least 3 months.

Screening Procedure.

For babies admitted to the well baby nursery, screening takes place in a quiet room in a restricted area of the nursery, allowing uniform testing conditions for all babies. The Clarity screening system (SonaMed Corp. Waltham, Massachusetts) is designed to administer both DPOAE and automated ABR tests. Screening is performed by a trained hearing technician. Unless an infant has been identified with risk factor for hearing loss, testing begins with a DPOAE screening. The intensity levels used (L_1 and L_2) for the stimuli are 65 and 55 dB SPL.

respectively. DPOAEs are measured across the following frequencies: 2000, 2500, 3200, 4000, and 5000 Hz, with the ratio of F_2 being 1.2 times that of F_1 to evoke a robust emission. Passing criteria requires the amplitude of each response to be at least 7 dB above the noise floor. The highest acceptable noise floor is 0 dB. An acceptable response must be obtained on at least four of five frequencies to achieve an overall pass result in each ear. The ABR is administered within the same software system. Disposable pre-gelled electrodes are placed at the forehead (Fpz) and the nape of neck, with a ground electrode placed on the shoulder. Impedances are checked and rejected if greater than 15K ohms or if interelectrode impedances are unbalanced. A 35 dBnHL click is presented at a rate of 38.1 per second using alternating polarity. Depending on the response waveform, up to 12000 sweeps may be collected. The Clarity system uses a template model to match waveforms to normative limits, as set by the test parameters. If an infant does not pass the first day they are tested, a rescreen may be performed the following morning unless the baby is discharged before a rescreen is possible. Results are documented in the infant's medical chart so the nursing staff and pediatrician are aware the test has been completed.

For babies screened in the NICU, or special care nursery, testing is completed when the infant is stable or prior to discharge (See Appendix B). Testing is usually completed by an audiologist at bedside. A handheld, portable AUDIOScreener (Grason Stadler. Visays Healthcare) was used to administer AABR screenings. The screening is done using pre-gelled, snap electrodes at the forehead and each mastoid. Impedances are checked and rejected if greater than 12 kOhms, or if interelectrode impedances are greater than 5 kOhms. A 35 dBnHL click stimulus is used. For infants who pass the initial screening, follow up is recommended as medically or audiologically indicated (See Appendix B. Protocol). If an infant refers after being retested, a complete diagnostic ABR is recommended to determine the level of hearing. OAE

testing is also recommended at this time to rule out auditory neuropathy, since this condition occurs more commonly in the NICU population.

Demographics.

SJMMC is located in a fairly large suburban area in St. Louis County with a smaller satellite hospital (St. John's-Washington) located approximately 45 miles from the main medical center. Well-established as a major birthing hospital in the area, nearly 7000 infants were born at SJMMC in the year 2004. The hospital in Washington, Missouri experiences approximately 1000 deliveries annually. Over 23,000 babies have been screened at the medical center since Missouri state law for UNHS was enacted January 1, 2002. The main hospital also has a Level III NICU, the only of its type in St. Louis County. Although infants may be retested at another facility if more convenient for the parents, most referrals return to this hospital for follow-up, allowing the program to track data over time. Results discussed in this paper were obtained from records of infants screened between January 2002 and December 2004. Well baby nursery data from SJMMC and the Washington medical center were combined.

RESULTS.

Risk Factors in the Well Baby Nursery.

Of the 14,141 babies screened in the well baby nursery from January 2003 through December 2004, 870 babies (6.2%) were identified with a risk factor. Family history of hearing loss was the most common risk factor, found in 2.2% of the total population screened. Maternal infection and hyperbilirubinemia occurred in 1.77% and 1.70% of all the babies, respectively. Craniofacial anomalies and syndromic stigmata were found less frequently in the well baby

nursery, present in only 0.36% and 0.07% of the babies, respectively. The relative occurrence of each risk among the 870 babies identified with a risk factor is shown below in Figure 1. As illustrated, family history accounts for approximately 40% of the babies who present with risk factors. History of maternal infection and hyperbilirubinemia also account for many of the risk factors in this population. It is important to note that in this case maternal infection, such as herpes, the baby is usually not infected but it is not routinely tested. The baby is treated as at risk if any of the TORCH infections are noted in the chart. This is to ensure an ABR is performed given the risk of neurologic damage to the auditory pathway. Audiologic follow up is recommended for maternal herpes if the infant is known to be positive for the infection. Babies with hyperbilirubinemia are administered an ABR when levels decrease or as close to the day of discharge as possible. Peak bilirubin levels are recorded, as well as the level at the time of the test. If bilirubin levels increase after the screening has taken place, it is recommended that another ABR be considered.

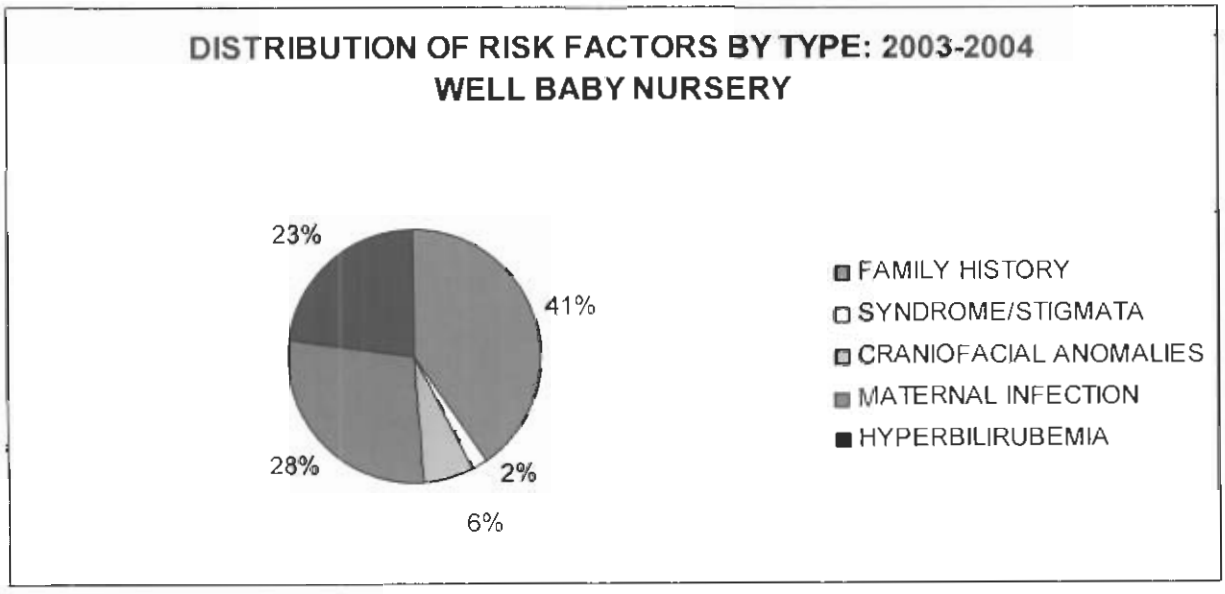


Figure 1.

Of the 870 babies identified as having a risk factor, 2.2% (n=19) referred as a result of the initial AABR screening. Figure 2 shows the total number of babies with each risk factor, results of the initial screening, and the number of babies identified with hearing loss. As expected, babies with craniofacial anomalies and stigmata associated with hearing loss were more likely to refer, accounting for nearly 60% of all high risk referrals. These two categories occurred in over half of the high-risk babies identified with hearing loss. It is not surprising that babies at risk due to maternal infection were most likely to pass, given that the babies in this population were not themselves identified with the disease. In addition, hearing loss due to maternal infection may be progressive; hearing at birth often appears to be normal.

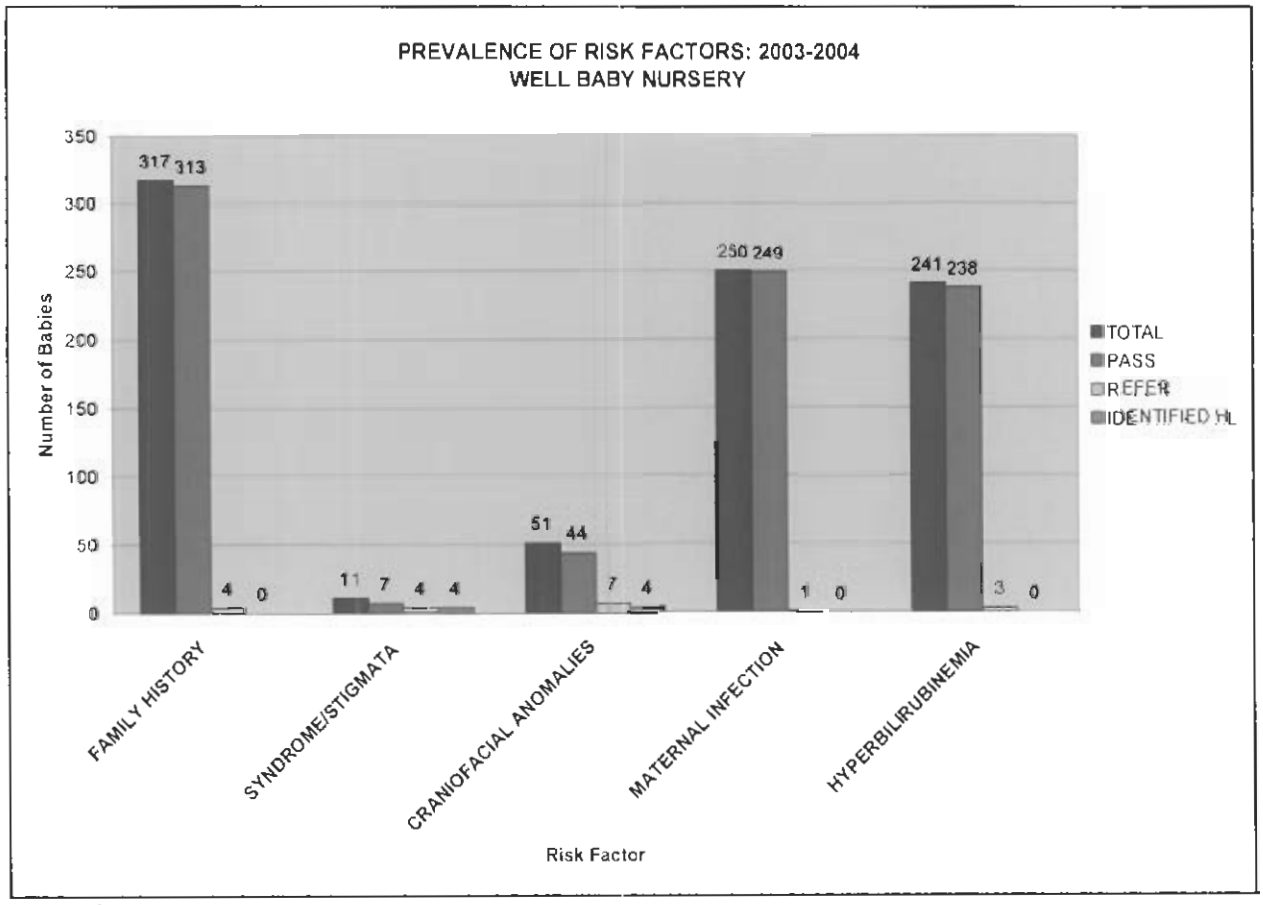


Figure 2.

Of the babies identified with craniofacial anomalies, ear tags and pits were the most common, accounting for over half of the cases. Figure 3 below shows the distribution of anomalies among the population studied in the well baby nursery. Approximately one-fourth of the babies with craniofacial anomalies were identified with cleft lip or palate. Atresia, malformed pinnae, and other anomalies such as malformed jaw all occurred with the same frequency, each accounting for 6% of the babies in this category.

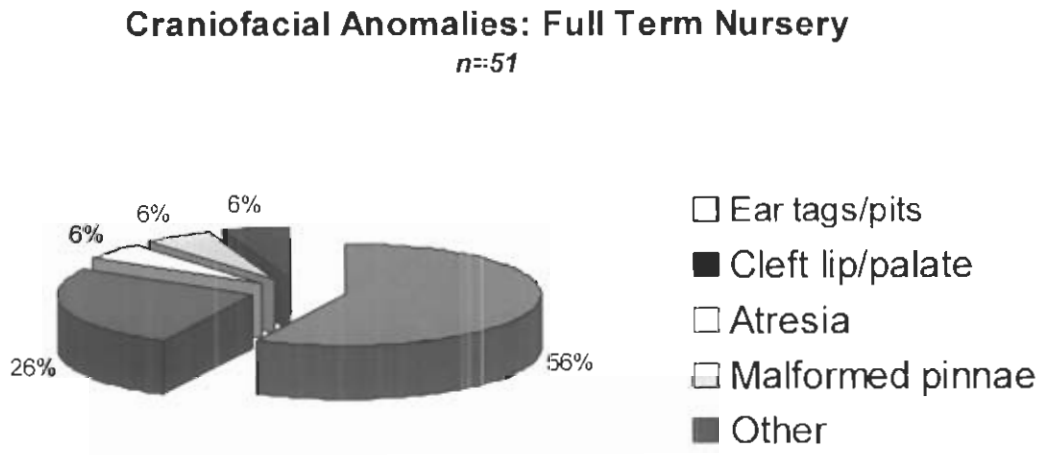


Figure 3.

Well Baby Nursery Screening Program Data.

The number of referrals was also examined to compare babies with risk factors to all babies tested. In 2002, 6483 babies were tested at the main hospital and satellite location. Of these babies, 0.8% (n=51) referred for further testing. By far, this figure surpasses the JCIH recommendation of less than 4% referral rate in the well baby nursery. Of the 51 babies who referred, 24% (n=12) were identified with a risk factor. These babies comprised only 0.2% of the entire well baby nursery population. Return rate for diagnostic testing was 98%, although

documented efforts for follow up were made for 100% of the cases. Diagnostic ABR and OAE testing identified 8 babies (16% of referrals) with hearing loss. Of the total well baby nursery population, these hearing-impaired babies comprised 0.1%. Hearing loss occurred in 1.2 per 1000 babies. Risk factors were identified in 50% (n=4) of the hearing impaired infants. One baby with a mild unilateral conductive hearing loss also had unilateral cleft palate. Another baby had a skin tag and maternal history of herpes. Two of the hearing impaired infants had family history of childhood hearing loss. One of these babies, identified with a bilateral sensorineural hearing loss, had a hearing impaired sibling with Mondini malformation of the cochlea. The other infant, identified as having a unilateral mild-to-moderate hearing loss, had a maternal uncle with hearing loss. All hearing impaired infants were referred for appropriate medical and audiological intervention as necessary.

In 2003, 47 of 6728 babies were referred for further evaluation, yielding a refer rate of less than one percent (0.7%). Of these 47 babies, 5 (11%) had a risk factor. Referrals with a risk factor comprised only 0.07% of the total population. Documented attempts for follow up were made for 100% of the cases, and 96% of the families returned for testing. Diagnostic ABR and OAE data showed a hearing loss in 6 (13%) of these babies. The percentage of babies with hearing loss was only 0.08% of all babies screened. For this year, the incidence of hearing loss per 1000 was 0.9. Only one of the 6 hearing impaired babies (17%) was also identified with a risk factor. This infant had a unilateral malformation of the pinna and presented with a severe unilateral conductive loss.

In 2004, 92 of 7413 babies referred in the WBN, a refer rate of 1.24%. A risk factor was identified in 13 (14%) of the 92 referrals, which seemed consistent with data from the previous year. Again, a documented effort was made to contact 100% of the families for follow up

testing, and 99% returned within three months. Of the babies retested, 12 were identified with hearing loss. The incidence rate for this year was 1.6 per 1000. Over half of these babies (n=7) did have a risk factor for hearing loss, the most common being craniofacial anomalies and stigmata associated with hearing loss. One baby was identified with Trisomy 21 and presented with a bilateral moderate loss. Another baby exhibited syndromic stigmata (low set ears and possible Trisomy 21) and also had a bilateral mild to moderate hearing loss. Goldenhar syndrome was identified in one baby with stigmata such as ear tags and micronathia. This baby was found to have a bilateral severe-to-profound hearing loss. Bilateral mild-to-moderate conductive hearing loss was found in three babies who all had cleft palate. Another infant with atresia was identified with a unilateral hearing loss. All of the hearing impaired infants were referred for the appropriate services and intervention.

When comparing screening results of babies with risk factors to babies without risks, results from 2003 and 2004 were analyzed because of availability of data. as the number of babies who passed with risk factors was not available for 2002. The graphs below divide babies into two categories: those with and those without risk factors. Figure 4 indicates that for both categories, a vast majority of the infants pass the initial screening. Figure 5 shows the number of babies who referred and were then identified with hearing loss for both categories. As illustrated, hearing loss is more common in the at risk population.

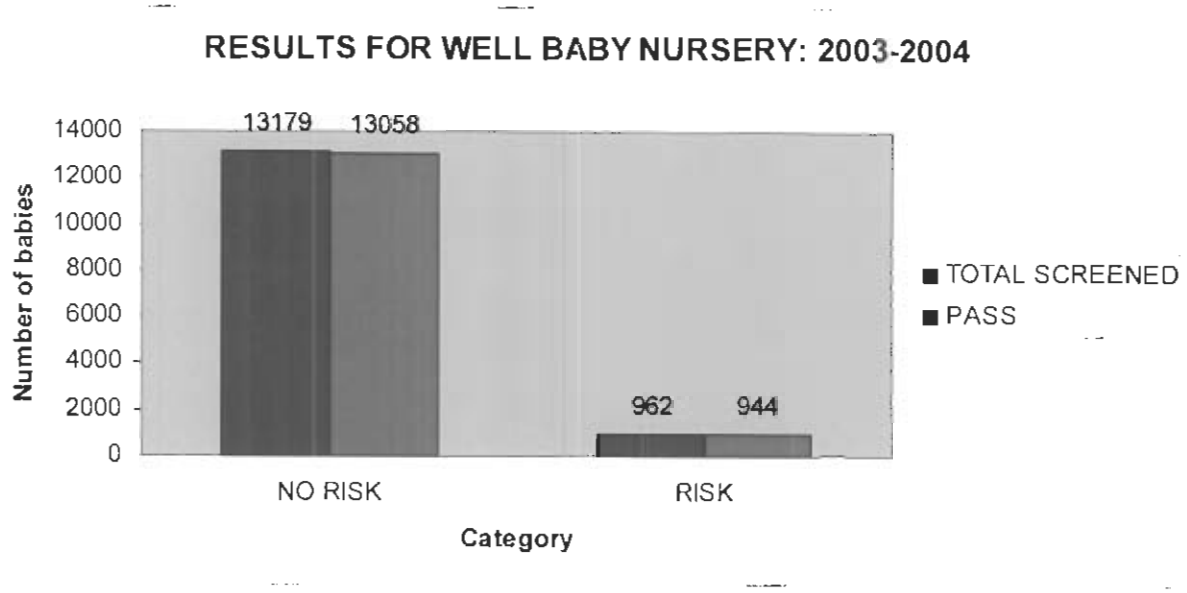


Figure 4.

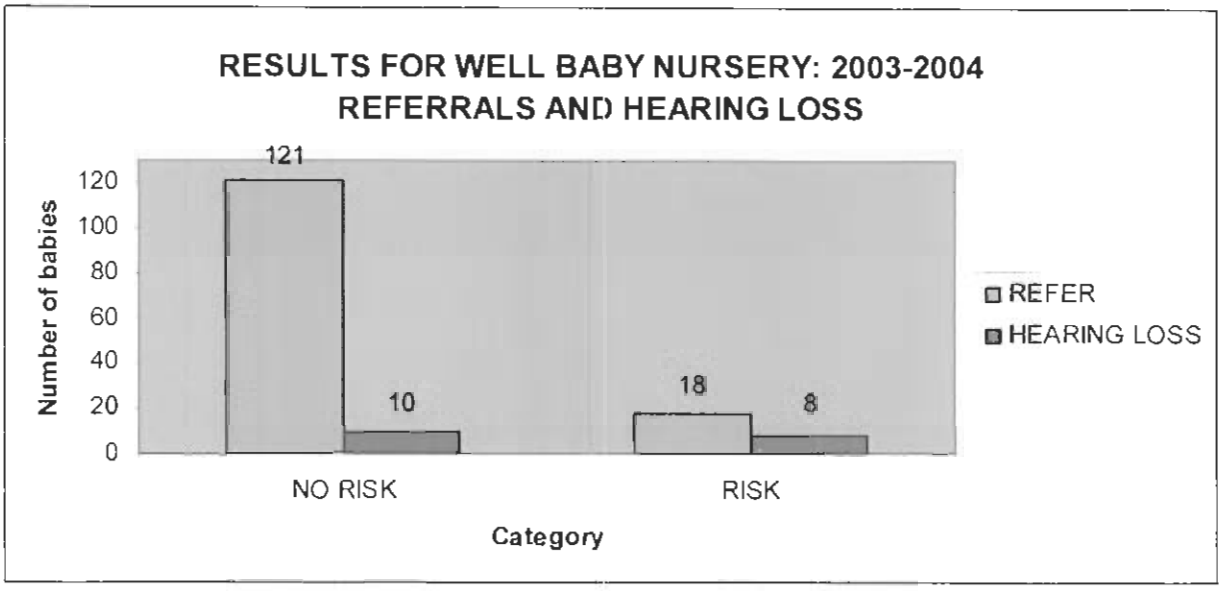


Figure 5.

NICU Sample.

To provide comparison data, infants screened in the NICU from July through December 2004 were also examined. Of the 483 babies screened within this time period, 395 had no risk factors besides admission to the NICU. There were 88 babies, however, that presented with additional risk factors. Figure 6 shows the presence of risk factors in the NICU among the

babies in this sample. The most common risk factors in this population are low birth weight, low Apgar scores, and ototoxic medications. These three conditions are each present in approximately a quarter of the NICU population surveyed. It is not surprising that these risk factors are seldom encountered in the WBN. Presence of a syndrome associated with hearing loss was the fourth most common risk factor in the NICU. Of the 127 babies identified with a risk factor, 8% (n=10) had been diagnosed with a syndrome. These babies comprised approximately 1.4% of all babies tested in the NICU. None of these babies were identified with hearing loss, as all passed the screening at birth. As seen in the WBN, Trisomy 21 was the most frequently diagnosed syndrome in the NICU. This is not surprising given the fact that it is the most common birth defect, occurring in one out of every 660 births (National Institutes of Health).

Another issue in the NICU is multiple risk factors. There are several risk factors which tend to occur together due to their medical manifestations. For example, low Apgar scores and PPHN tend to occur together, that is, that babies with severe respiratory distress will also receive low Apgar scores. In this population, 18% of infants had at least one risk factor, but only 57% of these babies had only one risk factor. There were 38 (43%) babies that had multiple risk factors, and most commonly the combination was low birth weight and low Apgar scores, or ototoxic medications and low birth weight.

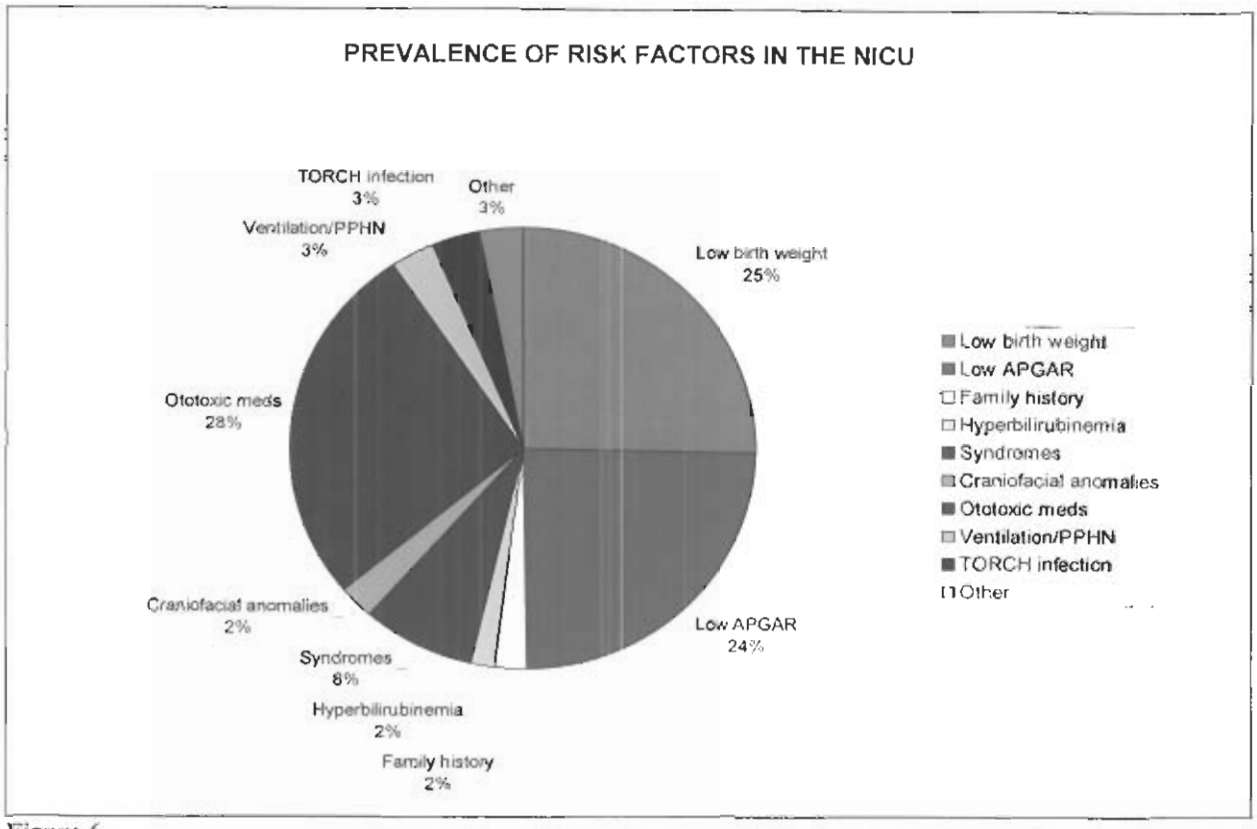


Figure 6.

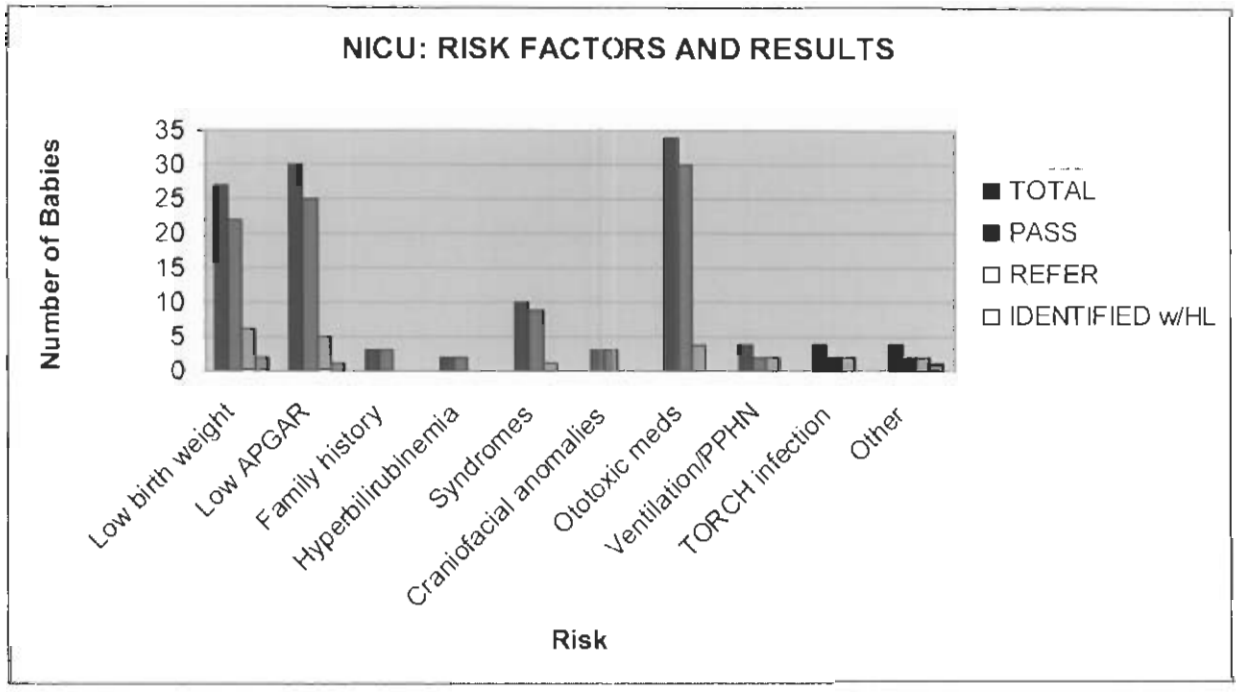


Figure 7.

Of the 483 infants screened in the NICU, 5.6% (n=27) referred for additional testing. This is well within the standards set by the JCIH which state that referral rate must be less than 10% for NICU babies. Of the babies who referred, 18 (67%) had normal hearing in both ears. There were 7 babies (26%) diagnosed with hearing loss in one or both ears, 4 of which had a risk factor for hearing loss. The results for two babies are unknown at this time. Of the 11 babies with risk factors who referred initial screening, 4 (36%) presented with hearing loss at the diagnostic evaluation. Figure 7 above shows the number of babies who referred with each risk factor and results of the initial screening. When divided into categories of risk factors and without risk factors, referrals and hearing loss were both more common in the group with risk factors. This is illustrated in Figure 8. Compared to the WBN, hearing loss was considerably more common among NICU babies with risk factors, as expected. In all, 1.4% of the NICU babies had hearing loss, 50% of which had risk factors. The incidence of hearing loss per 1000 in the NICU was 14.5, a much higher rate than found in the WBN.

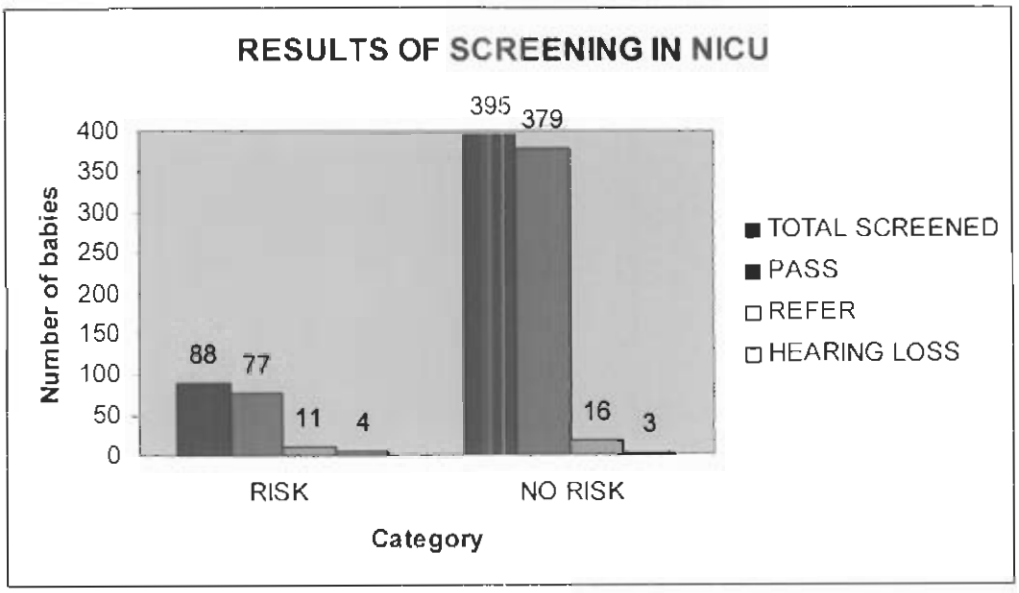


Figure 8.

DISCUSSION.

As noted at the beginning of this paper, it has been previously estimated that 10-12% of all infants have a risk factor for hearing loss (Epstein and Reilly, 1989). In this study, risk factors were present in approximately 6% of infants in the WBN and 18% of NICU infants. There are few published studies for comparison. since many studies group well baby infants with risk factors in the same category as NICU infants. Mason and Herrmann (1998) did calculate the percentage of babies with risk factors in the WBN population alone and found it to be less than 2%. It is important to note the presence of risk factors will vary greatly depending on what criteria is used. If admission to the NICU for more than 48 hours is counted, such as in the Mason and Herrmann study, then the majority of NICU babies would be considered at risk. In this study, admission to the NICU alone was not considered a risk factor, therefore only 18% were considered at risk for hearing loss. As stated by Vohr and colleagues (2000), "Risk factor data are difficult to collect and evaluate, except in a large clinical trial because of both the low incidence rates of individual risk factors and the low incidence of neonatal hearing loss."

The most common risk factors in this study were similar to the most commonly found risk factors in the Mason and Herrmann study: skin tags, cleft palate, family history, and ototoxic antibiotics. Family history was present in 2.2% of the total babies screened in the WBN. In a study by Vohr and colleagues (2000), family history was found in 6.6% of the WBN. They did, however, report that babies with risk factors were actively sought for the study. Another group reported positive family history for 10% of the infants with hearing loss (Sutton and Rowe, 1997). Kountakis et al (1997) found 3% of the infants screened had positive family history. As illustrated in Figure 2, although family history is the most common risk in the WBN, less than 2% of these babies referred the initial screening, and none were identified with hearing loss for

2003 and 2004. There were, however, two babies identified with hearing loss in 2002 who had a positive family history. The low incidence of hearing loss with positive history may be due, at least in part, to the progressive nature of some genetic hearing losses and delayed onset. It has been estimated that one-third of deaf infants do have a recessive gene for hearing loss, but parents deny family history because they are unaware of it (Frasier, 1971). Except for younger parents, a vast majority of previous generations did not receive a hearing screening at birth. Hearing loss thought assumed to be congenital may actually be a result of a disease in infancy or delayed onset hearing loss. For this reason, it is imperative that infants with positive family history are monitored for hearing loss. There have already been a few children who have been diagnosed with hearing loss upon returning to this medical center because of recommended follow up. The overall incidence of progressive hearing loss among the group in this investigation has not yet been determined. In the future, as these children are monitored, perhaps more data can be collected to observe any trends.

Another relatively common risk factor, which seems consistent with the literature, is craniofacial anomalies. In this study, craniofacial anomalies accounted for 6% of the babies with risk factors in the WBN, but only 2% of the high risk infants in the NICU population. Syndromes occurred less frequently in the WBN, present in only 0.09% of the total population, compared to 2.07% of NICU babies. This is similar to results published by Vohr et al (2000), who reported, "The presence of stigmata or syndromes associated with hearing loss were more common among NICU infants (1.8%) than well baby nursery infants (0.5%)." It is important to note that syndromes may not be identified until after the baby is discharged. NICU infants usually spend a longer time in the hospital under observation whereas well infants are discharged much sooner. These infants may have syndromes but have not yet exhibited the symptoms.

Regarding hearing loss, a study conducted by Van Riper and Kileny (1999) indicated that craniofacial anomalies and syndromes "appeared to have high predictive value." In the WBN, over half the babies that referred with craniofacial anomalies were identified with hearing loss, but all NICU babies with craniofacial anomalies passed the initial screening. This difference is probably due to the small sample of NICU babies that was analyzed. Perhaps if data were examined over a longer period of time, there would be a higher number of babies with craniofacial anomalies in the NICU. Within the category of craniofacial anomalies, ear tags and pits were the most common in this investigation. This is congruent with data reported in a study funded by the National Institutes of Health (NIH) (Cone-Wesson et al; Vohr et al. 2000).

The most common risk factors specific to the NICU in this study are also in agreement with findings from previous studies. Ototoxic medications were administered to 7% of the NICU infants. Exposure to ototoxic medications was also the most common risk factor among high risk infants in the Van Riper and Kileny study (1999). Results were fairly similar to this investigation, as most of the infants passed the screening. Only 0.7% of these babies were identified with hearing loss in the 1999 study, compared to the present study which did not identify any hearing impaired babies who had been exposed to ototoxic medications.

Looking at the results of this study, not as many babies were administered phototherapy for hyperbilirubinemia as one might speculate. Rhee and colleagues (1999) noted that "the incidence of hearing loss due to hyperbilirubinemia is decreasing with the current management of neonatal hyperbilirubinemia with timely phototherapy and exchange transfusion. However, once developed, bilateral, symmetrical sensorineural hearing loss in hyperbilirubinemic babies leaves them with great difficulty in communication and mandates some type of rehabilitation." Hyperbilirubinemia occurred in 1.6% of the NICU babies and 0% of the well babies in the NIH

study (Vohr et al, 2000). In the current investigation hyperbilirubinemia occurred in approximately 2% of NICU babies and 1.7% of well babies. All of these infants passed either the initial screening or were within normal limits as determined by the diagnostic evaluation. There were no infants identified with hearing loss in this group. Kountakis et al (1997), however, did find that hyperbilirubinemia was significantly associated with hearing impairment. The study reported over half of the infants with hyperbilirubinemia had hearing loss, and "most infants with bilirubin levels [greater than or equal to] 10 micromoles per liter failed the ABR screening." Their study population involved an equal number of babies who failed the initial screening and who were known to have normal hearing. The vast majority of babies in the SJMMC group passed the screening. In addition, this study is several years old and, as noted previously, management of hyperbilirubinemia has been more aggressive in recent years. These differences could account for why hearing loss due to hyperbilirubinemia was not observed in the SJMMC study. Other risk factors commonly found in the NICU, low birth weight and low Apgar scores, were present in 5.6% and 6.2% of the population, respectively. These same conditions occurred in 18% and 14% of the population in the NIH study (Vohr et al, 2000).

Overall incidence of hearing loss in this study seemed to be in accord with previously reported data. Over three years in the WBN alone, incidence of hearing loss is 1.26 per 1000. White and colleagues (1993) also reported 1.2 hearing impaired infants per 1000 in a well baby population using only OAE for screening. Mason and Herrmann (1998) used AABR to detect hearing loss in a WBN, and found hearing loss occurred in .9 per 1000 infants. It is unusual for incidence studies to focus only on the well baby population (Mason and Herrmann, 1998) and therefore comparative data is limited. There is more data in the literature, although perhaps showing some inconsistencies, regarding hearing screenings for the NICU population. For the

small NICU sample in this investigation, incidence was estimated to be 15 per 1000. The figure for hearing loss in the NICU varies greatly among studies with incidence reports as low as 5 per 1000 (Mason and Herrmann, 1998) to as high as 60 per 1000 (Bradford et al, 1985). The variance may be attributed to the different populations examined, as some studies took a sample of only very preterm infants in the NICU, while others sampled all babies admitted to the NICU. Outcomes of a major investigation (Prieve et al, 2000) indicated hearing loss was present in 1.96 infants per 1000 for the universal population, or 8 per 1000 in the NICU and 0.9 per 1000 in the WBN. Among the literature, the most commonly cited incidence of hearing loss in universally screened populations is approximately 3 per 1000. In this investigation, when a sample is taken from the same six months for both the WBN and the NICU, hearing loss occurred about 3.6 times per 1000 in the universal population.

It is of great interest to know how many hearing impaired infants have a risk factor for hearing loss. The graph below (Figure 9) shows the results of several studies. Of the 25 hearing impaired infants, 14 (56%) were identified with a risk factor while 11 (44%) did not. This number is very close to the 50% estimate which is a frequently reported figure. In the NIH study, 121 hearing-impaired infants (72%) were identified with a risk factor, while only 47 (28%) had no risk factors (Cone-Wesson et al, 2000). The risk factors that seemed to be most predictive of hearing loss were syndromes, craniofacial anomalies, and family history. Cone-Wesson and colleagues suggested the possibility of autosomal recessive genes for deafness to account for some of the hearing-impaired babies with no risk factors. In the study conducted by Mauk et al (1991), parents of 70 hearing impaired children were surveyed with regard to the child's birth history. Exactly half of these children exhibited high-risk characteristics at birth. A

similar trend was reported by the investigators of the Colorado screening project. Of the 126 hearing impaired infants, only 63 (50%) exhibited risk factors (Mehl and Thomson, 1998).

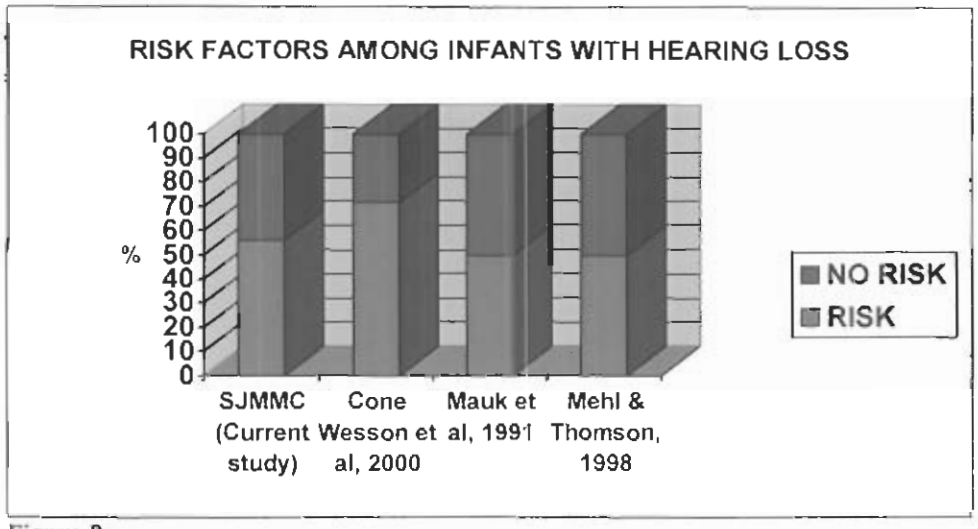


Figure 9.

Another point for discussion is screening protocol. When interpreting data from a screening program, it is also important to discuss the differences in protocol for the different nurseries. In the WBN, the mothers are inpatients and are available to answer questions about family history of hearing loss. In the NICU, parents are not always in the hospital when the AABR is completed, and family history of hearing loss is often unknown by the screener. Another factor is length of stay. Babies are usually discharged from the WBN within two or three days. Syndromes are often not identified until the first few months of life when babies do not have any pressing medical issues at birth. In the NICU, where the population is not as healthy, syndromes may be diagnosed sooner. For example, it is easy to understand why there are so many more babies with Trisomy 21 found in the NICU, since congenital heart defects are often associated with this syndrome. Babies experiencing respiratory problems have low Apgar scores. These infants are often also premature, therefore having low birth weight. The need for

ventilation, intravenous antibiotics and fluids, and monitoring of vital signs may necessitate a long stay in the NICU. When examining risk factors one must question if hearing loss is a direct or indirect result of a particular condition. It is important to keep these differences in mind when comparing data to previous studies and across nurseries.

FURTHER STUDY.

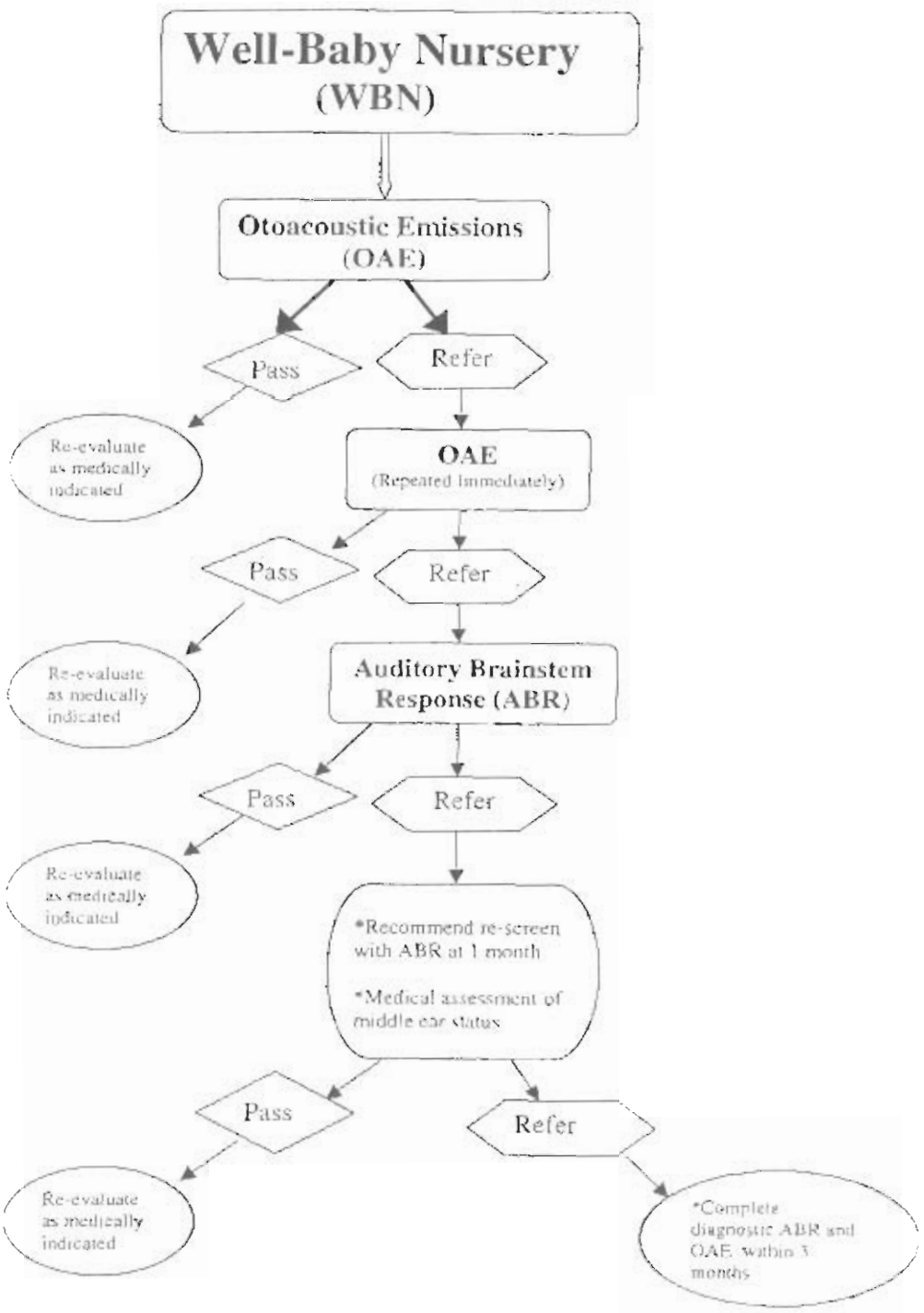
There are many avenues to pursue regarding risk factors and hearing loss. One of the goals of UNHS is to identify babies who are most likely to have a progressive hearing loss and "flag" them for audiologic monitoring. At this medical center, parents and pediatricians of babies at risk for progressive or late onset hearing loss are notified that the baby should have follow up testing at six to eight months of age. Reminder letters help increase the number of families who return. It is difficult to follow up on these infants, however, because parents may receive audiologic testing at another facility. Since UNHS has only been in effect since 2002, there is not enough data to show how often progressive hearing loss occurs in this population. Additional data is needed to determine if state-mandated UNHS is successful in identifying babies with progressive or late onset hearing loss.

Another area of interest is comparing statistics between area hospital UNHS programs. It may be that some hospitals have not yet determined the most effective way to screen. Comparing protocols, test parameters, and results of screening may be beneficial for programs to develop the most reliable and cost-effective set up. In addition, socioeconomic status and access to prenatal care may have an effect on program data.

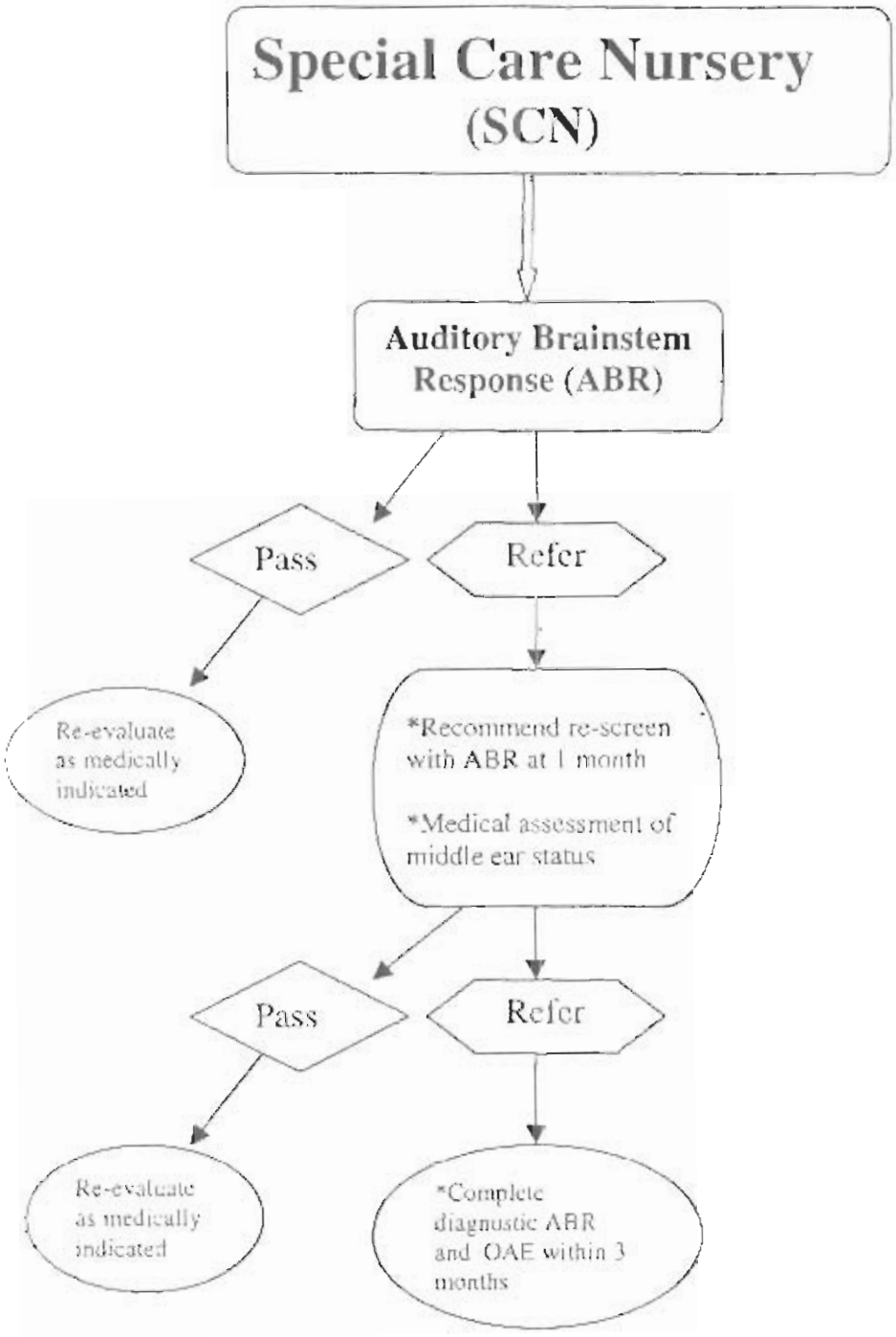
During a time where much needed emphasis is placed upon evidence-based practices, UNHS programs must be able to prove their efficacy. As states continue funding for UNHS,

they will demand data supporting such programs. The most valid way to obtain thorough outcome measures is over time. At present, it has only been three years since UNHS was put in effect for the state of Missouri. Several more years of results will help to provide statistically significant measures.

APPENDIX A. SCREENING PROTOCOL: WELL BABY NURSERY.



APPENDIX B. SCREENING PROTOCOL: NICU (SPECIAL CARE NURSERY).



APPENDIX C. WELL BABY NURSERY DATA.

		2002	2003	2004	TOTAL
A	**Total screened**	6483	6728	7413	**20624**
B	**N refers**	51	47	92	**190**
C	**Refer rate**	.8%	.7%	1.2%	**.9%**
D	**% returned**	98%	96%	97%	**98%**
E	**Documented efforts for follow up**	100%	100%	100%	**100%**
F	**N referred w/RF**	12	5	13	**30**
G	**% referred w/RF of all babies**	.20%	.07%	.18%	**.15%**
H	**% referred w/RF**	24%	11%	14%	**16%**
I	**% referred w/o RF**	76%	89%	86%	**84%**
J	**N identified w/HL**	8	6	12	**26**
K	**% HL of all babies**	.10%	.08%	.16%	**.13%**
L	**% HL of all refers**	16%	13%	35%	**14%**
M	**N HL and RF**	4	1	7	**12**
N	**% HL and RF/all HL**	50%	17%	58%	**46%**
O	**N HL w/o RF**	4	5	5	**14**
P	**% HL w/o RF/all HL**	50%	83%	42%	**54%**

N= number; HL=hearing loss; RF=risk factors

DESCRIPTIONS:

A. Total tested includes all babies screened in the well baby nursery at the main hospital and satellite hospital from January 2002 through December 2004 The three year total is over 20,000.

B. Refers include all babies who do not pass the inpatient hearing screen in one or both ears.

C. Refer rate is calculated by dividing the number of referrals by the total number screened.

D. Percentage returned shows how many families returned for diagnostic testing during the first three months of life. This statistic may be used as a quality indicator of a UNHS program. and according to JCIH (2000), an "ideal" return rate is at least 70%.

E. According to JCIH recommendations, documented efforts for follow up must be made for at least 95% of the referrals. According to program data documented attempts for follow up were made in 100% of the cases.

F. The number of referrals who were identified as having a risk factor for hearing loss.

G. The number of babies who referred initial screening and were identified with a risk factor divided by the total number of babies screened.

H. The number of babies who referred and had a risk factor divided by the total number of referrals.

I. The number of babies who referred and did not have a risk factor divided by the total number of referrals.

J. The number of babies identified with hearing loss.

K. The number of hearing-impaired babies divided by the total number screened.

L. The number of hearing-impaired babies divided by the total number who referred screening.

M. The number of babies who were identified with hearing loss AND a risk factor.

N. The number of babies with hearing loss AND risk factors divided by the total number of hearing-impaired babies.

O. The number of babies who were identified with hearing loss AND NO risk factor.

P. The number of babies with hearing loss AND NO risk factor divided by the total number of hearing-impaired babies.

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