PROJECTS COMPLETED BY STUDENTS OF THE NATIONAL LEADERSHIP TRAINING PROGRAM IN THE AREA OF THE DEAF
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ABSTRACT


The purpose of this study was to determine how many residential schools for the deaf are currently screening their students for the Usher Syndrome and how many of these schools offer support services to the identified Usher Syndrome student and his family. Forty schools responded. Residential schools were chosen because they have large concentrations of high-risk individuals, the congenitally deaf. The results indicate that 53% of the schools that responded are screening students for the Usher Syndrome. Eighty-four percent of the identified students are older than twelve years old. Eighty-one percent of the students have a profound hearing loss. One-half of the schools with screening programs offer some support services: personal counseling, genetic counseling, curricular modifications, and vocational counseling.
# TABLE OF CONTENTS

ABSTRACT ...................................................... i
LIST OF TABLES ................................................... iii
LIST OF APPENDICES .......................................... iv
ACKNOWLEDGEMENTS ............................................ v
INTRODUCTION .................................................. 1
METHODOLOGY .................................................. 11
RESULTS ......................................................... 15
DISCUSSION ....................................................... 21
RECOMMENDATIONS FOR FURTHER STUDY ......................... 23
APPENDICES ....................................................... 25
REFERENCES ....................................................... 54
LIST OF TABLES

TABLE 1: Percent of Respondents and Screening Programs for the Usher Syndrome............15
TABLE 2: Percent of Schools Having a Screening Program for the Usher Syndrome by Size......16
TABLE 3: Percent of Students with the Usher Syndrome by Age.................................17
TABLE 4: Decibel Loss of Students with the Usher Syndrome....................................17
TABLE 5: Frequency of Screening for the Usher Syndrome........................................18
TABLE 6: Evaluations Used in Screening for the Usher Syndrome...............................19
LIST OF APPENDICES

APPENDIX A: A Recommended Bibliography..................25

APPENDIX B: Cover Letter Sent to Residential
Schools for the Deaf.................................29

APPENDIX C: Questionnaire Sent to Residential
Schools for the Deaf.................................31

APPENDIX D: List of Residential Schools for
the Deaf...............................................37

APPENDIX E: Results of Screening for Visual
Acuity..................................................46

APPENDIX F: Regions of the United States as
Established by the National
Association of the Deaf..............................48

APPENDIX G: Comments Made by Respondants...............50
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Thank you McCay Vernon for your inspiration, your guidance and your strict timeline.
INTRODUCTION

The Usher Syndrome* is a hereditary genetic disorder which results in a congenital or a progressive hearing loss and progressive retinitis pigmentosa. The latter refers to a gradual degeneration of the retina. In the following sequence, the symptoms include: a hearing loss (usually profound); a gradual loss of night vision (poor dark adaptation); a gradual loss of side vision (reduced visual field); and a gradual loss of central vision usually leading to legal blindness (Bergsma, 1973).

Although the Usher Syndrome is rare in the general population (3 per 100,000), 2 to 5% of the genetic deaf population have the disease (Vernon, 1973). This inherited condition also causes more than half of deaf-blindness in adults. Ophthalmologists are familiar with the symptoms of retinitis pigmentosa, but they do not always recognize

* "Notice that I'm using the terminology 'the Usher Syndrome' rather than 'Usher's Syndrome' because medical parlance has changed; so instead of using the possessive, we usually say 'the such and such syndrome'" (Bergsma, 1976).
its relationship to hearing loss. In addition, schools for the deaf have not always provided thorough visual screening programs which would detect suspected cases of the Usher Syndrome. Lack of information among relevant professionals about the Usher Syndrome is the primary reason why so little has been done to identify and help people with this double handicap, even though it was discovered over one hundred years ago.

For example, facts about the genetics of autosomal recessive inheritance of Usher's though readily understandable, have often not been communicated to the patient because this information is not widely known except by geneticists interested in the problem. In fact, many of the specialists who are most likely to see deaf or blind children are not even aware of the existence of Usher's Syndrome. The auditory and visual problems of the disease are often thought to be separate and unrelated. Appropriate knowledge is rare even among those who work as consultants to schools for the deaf or in speech and hearing clinics. This is particularly unfortunate in the case of audiologists, otologists, pediatricians, and general practitioners because it is they who first see these patients, usually during preschool years at a time when referral to an ophthalmologist should be made. (Vernon, 1969, p. 136)

Early identification of the Usher Syndrome is important for the students safety. His safety is the most obvious reason. Young children may need additional supervision at night. It may be inadvisable for older students to drive if their visual field and dark adaptation abilities have deteriorated. Joseph Widermeyer describes one of his experiences while driving a car in
Italy, before he was told he had the Usher Syndrome.

Bing, bang, clump! I had driven across a concrete safety isle in Milan, Italy. Endeavoring to make a left turn, I had not seen the raised concrete platform along side of my car. Fortunately, I stopped in time to miss an oncoming bus. My passengers were scared, so I said, "Gosh, these Italians don't care where they pour their concrete." That wisecrack relieved the tension. That shakeup occurred in 1950 and I soon forgot about it until some weeks later when I had an eye infection. I went to a famous Italian ophthalmologist and surgeon for examination and treatment. He cauterized my right eye and then he said to me, "You need eyeglasses but you also have RP." ...That was the first time I knew I had RP. (Widenmeyer, 1973, p. 39)

Without proper identification and counseling, the person with the Usher Syndrome is unaware of his unique visual difficulties.

According to Arthur A. Roehrig, who has the syndrome, there are three levels of awareness of visual deterioration in the Usher Syndrome victim. The first level is unconscious awareness, when the individual realizes he cannot see well at night. The student may not be aware that other people can see well in dimly lit places, so he may not be concerned by his inability to do so. Mr. Roehrig describes an experience he had during the unconscious awareness level.

*Throughout this study, masculine pronouns, in reference to students, have been used for easier reading; however, the students afflicted with the Usher Syndrome include females as well as males.
When I was ten years old and a pupil at St. John's School for the Deaf in Milwaukee, Wisconsin, some friends and I were playing a game of hide and seek. It was the first time I played the game at night, and it was the first event that made me realize I had difficulty seeing at night. When we started playing the game, I saw all of the boys running to many places where they thought they could not be seen. I thought I could do this and started running. All of a sudden, I ran into trees and tripped over things, such as rocks and bushes. However, what I did that night did not bother me because I thought all the boys were running into things also. But when the game was over, we went into a well-lighted building. My supervisor, a nun, and all the boys suddenly stared at me and the supervisor said to me, "What happened to your face, arms and clothes?" I became puzzled and did not know what she meant. So when I went to the bathroom, I looked in a mirror and then at the other boys' faces, arms, and clothes. Although I began realizing my inability to see at night, I thought all the boys also couldn't see well but had avoided the obstacles because they had memorized all they saw during the day. (Roehrig, 1973, p. 33)

The second level is semi-conscious awareness. During this level, the individual notices that his vision is continually deteriorating.

I remained unconsciously aware of my visual impairment for a long time, even after I found out that I have tunnel vision, until one afternoon in a classroom at the Maryland School for the Deaf in Frederick, Maryland. At that time, I was employed as a second-year teacher in the academic year 1970-1971. I had been reading a newspaper and when I rubbed my left eye while continuing to read, I suddenly noticed that I could not see with my right eye as before. My vision was wavy. It made me realize that my eyes would continue deteriorating. Because of this new realization and because I had been unconsciously aware of the problem for a long time, I at this point, became semi-consciously aware of my visual impairment. (Roehrig, 1973, p. 35)
The final level is when the individual has been told that he has the Usher Syndrome. This level is called conscious awareness.

I finally entered as an in-patient at the National Institute of Health in Bethesda, Maryland. I stayed there for about a month for a complete head to toe examination. During my stay at the institute, my social worker, Mrs. Schumann, and my ophthalmologist, Dr. Donald Bergsma, worked very closely with me. One day, Mrs. Schumann and I started talking about my future. At that time, she, in a friendly and nice way, tried to talk me into thinking a little more about the things that blind people are encouraged to learn, such as braille and cane-mobility, and I then became consciously and hopelessly aware of what would happen to my eyes in the next few years—I mean blindness. The night following my talk with Mrs. Schumann, I was naturally depressed and cried all night because I knew I would be blind some day even before Dr. Bergsma explained to me about my progressive blindness. (Roehrig, 1973, p. 36)

Mr. Roehrig then points out several important facts that the counselor needs to explain to the student who has just discovered he has the Usher Syndrome. Being born with the syndrome is not the fault of the parents. Every person is unique, but he is equally as important as any other individual. "It is impossible to know why some of us are born deaf or blind just as it is impossible to know why some of are born black and others, white." (Roehrig, 1973, p. 37) Being honest with the student helps him to be honest with himself. He was not responsible for his genetic anomaly, but he is responsible for knowing what to do with his handicap. The school must help him understand
and evaluate his options.

Although a person with the Usher Syndrome may be able to accept his condition intellectually, it will take a long time before he can accept it emotionally. To help him accept his condition, the school should allow the student to participate fully in all activities of his choice and should encourage him to participate if he seems reluctant. To shelter him is to deny him the opportunity to enjoy life normally like other children and to develop fully. Because of the syndrome's progressive nature, the student need not be given special training, cane or mobility training, braille or daily living skills right away. He will need time to adjust to his handicap. However, he should be encouraged to learn to communicate in as many ways as possible. During a recent workshop in San Diego, California, Linda Annala, who still has fifteen degrees of peripheral vision left, told the author that she has practiced closing her eyes and reading her deaf friends' signs and fingerspelling in her hands, in preparation of becoming totally blind.

The counselors, teachers, medical staff and other related persons can help the student become informed and involved in decisions regarding his handicap. When he is young, it may suffice to tell him that he has visual problems which will become worse as he gets older. As the child matures, personal counseling, curricular modifica-
tions, realistic vocational training and eventually genetic counseling may be provided as necessary. These support services will enhance the student's chance of accepting and adjusting to his handicap.

An adjunctive benefit of this early diagnosis would be better educational planning for the detected cases. Under present circumstances, the eventual blindness is generally unknown until late in a student's academic career, at which time it is often too late to plan appropriately. Ironically these youths are usually taught vocational and educational skills emphasizing vision as this is suitable in programs for the deaf which they attend. (Vernon, 1969, p. 134)

Deaf-blindness creates serious communication and mobility problems. Along with these problems comes social isolation, loneliness and frustration. The individual used to being a member of the deaf community, soon finds himself left out of many activities due to the difficulty others experience in communicating with him. Counseling is a must for these individuals.

Early identification is also important for the families of children with the syndrome. Once a student has been identified as having the Usher Syndrome, it is advisable to refer his family for audiological and visual examinations. The parents need to know the implications this genetic disorder has on their child's future and the genetic possibilities of other children in the family having and/or being carriers of the disorder. Because the Usher Syndrome is an autosomal recessive trait, the
affected individual must inherit an abnormal gene from each parent. Usually, the parents are both carriers although they may be unaware of their genetic link to the syndrome. If the parents are planning to have additional children, genetic counseling should be available if they desire it. Genetic counseling is the most misunderstood discipline of our time. It is not meant to manipulate, counsel or advise. Genetic counseling is intended to inform people of what they can expect under certain conditions. To perform genetic counseling, one must have a reliable diagnosis, a second opinion, a proper family history, and a knowledgable genetic counselor.

A reliable diagnosis is necessary because the Refsum Syndrome and Rubella also cause deafness and blindness and therefore may be confused with the Usher Syndrome. Furthermore, there are at least four types of the Usher Syndrome. Type I, the most well-known, involves congenital deafness and progressive retinitis pigmentosa. Type II involves progressive hearing loss and progressive retinitis pigmentosa. This variety appears to have less severe visual deterioration than Type I. Type III involves congenital deafness, usually profound, progressive retinitis pigmentosa, and cerebellar ataxia. Type IV is similar to Type III, however the individual is also mentally retarded. There is no consistent pattern when looking at families. Some geneticists believe that Type
III and IV are variations of Type I. They are often named the Hallgren Syndrome.

Obtaining a proper family history is not always easy, but it is important to the genetic counselor. For example, a person who has Type I and a person who has Type II have a 100% chance of having normal children because the genes are not located on the same spot on the chromosome. Because of the genetic nature of the Usher Syndrome, the counselor needs to know: if other family members have the Usher Syndrome; if there is a history of consanguineous marriage in the family and; the ethnic origin of the family because there are areas with pockets of the syndrome due to consanguineous marriages, e.g., Louisiana, Finland, Sweden and Denmark.

Because genetic counseling is a sensitive issue and because the discovery that you or your child has the Usher Syndrome is overwhelming, correct timing is crucial. The individuals involved need time to accept and adjust to the medical and personal implications of the Usher Syndrome. When the individuals involved desire more information, or an apparent need for support services becomes necessary, it is the correct time to begin counseling, both personal and genetic. Once the family history has been established, the genetic counselor should explain the basic genetic facts and clarify the risks, pointing out any options to reduce these risks.
The goal of such counseling is to provide accurate, appropriate information, to be certain the family understands the information, and to refrain from making decisions for the family, no matter how strong his or her own personal opinions concerning the appropriateness of those decisions. There are no wrong decisions in this view of genetic counseling, as long as they are truly informed decisions. (Harrod, 1978, p. 377)

There have been several workshops on the Usher Syndrome within the last decade which offer substantial information to the reader interested in knowing more about this genetic disorder. A recommended list of references may be found in Appendix A.
METHODOLOGY

A cover letter (Appendix B) and a questionnaire (Appendix C) were sent to the Superintendents of 59 residential schools for the deaf in the United States (Appendix D). The names of the schools and superintendents were taken from the 1981 director of the American Annals of the Deaf, Volume 126, Number 2. Because of the number of schools involved, it was decided that a questionnaire would be the most efficient and accurate method of obtaining data. The purpose of the questionnaire was to determine how many residential schools for the deaf are currently screening their students for the Usher Syndrome and how many of these schools are offering support services to the individual and/or his family.

The questionnaire was divided into four parts. Section I inquired about general background information, such as the name of the school, the position of the respondent, and the size of the school. In tabulating the data, it was evident that there was no notable difference in services provided by schools with different sized populations, so this section was discarded.

Section II inquired about screening services for visual acuity. Although visual acuity was not the intent of the investigation, it was decided that questioning this
aspect of the screening program first would eliminate confusion and false data in Sections III and IV, which asked questions specific to the Usher Syndrome. Data obtained from Section II will not be discussed in this paper, but the results may be found in Appendix E.

Section III covered the incidence of students who have been diagnosed as having the Usher Syndrome by age, by hearing loss and by sex.

Section IV determined the existence of a screening program for the Usher Syndrome, the population screened, the frequency with which students are screened, the position of the person who conducts the screening, the types of evaluations used, and support services offered to the identified student as well as his family.

The major fault of the questionnaire is its length, five pages. Although 67.8% of the schools questioned did respond, several of those that did not, indicated that they did not have time to answer such a lengthy questionnaire. Another difficulty encountered was in attaching particular evaluations to the students identified as having the Usher Syndrome. Although a school may have access to a particular evaluation, it was not determined whether all evaluations were used on all students.

There were several omissions in the questionnaire: etiology of individual student's hearing losses; age of student when diagnosed; number of other relatives having
the Usher Syndrome; ethnic origin of the family; and any known family members of consanguineous parentage. These omissions may be found in the last section of this text called Recommendations for Further Study.

The "yes-no-not sure" format on most questions lent itself well to being easily answered as well as tabulated. In discussing the results, only the "yes" answers are included in the text, because the purpose of the study was to determine what is currently being done. The other data may be found in the Tables accompanying the text.

In tabulating the results found in Table 1, the total number of schools contacted was divided by the total number of responses for each region. The regions used were established by the National Association of the Deaf (Appendix F). To determine what percentage of respondents have screening programs for the Usher Syndrome, the total number of respondents in each region was divided by the total number of schools which indicated having a screening program.

In Table 2, each student was grouped by age and sex. The total number of students for each age group was divided by the total number of students to determine the percentages.

In Table 3, students were grouped by sex and decibel loss. The total number of students in each decibel category was divided by the total number of students to
determine the percentage.

To establish the frequency with which schools screen their students (Table 4), each respondent was grouped according to the yearly interval indicated in their response. The number of responses for each interval was then divided by the total number of schools that responded.

In Table 5, schools were asked to indicate possible types of evaluations used in their screening programs for the Usher Syndrome. Each test represents a question on the questionnaire. Table 5 was tabulated according to the number of responses each question or evaluation received. For each category, "yes," "no," "not sure," and for those schools which did not respond at all, the total number of responses per answer category was divided by the total number of responses for that evaluation or question.

The fifth page of the questionnaire concentrated on the existence of support services offered to students who have the Usher Syndrome and their families. More research is needed to discover more in-depth information regarding these services (Recommendations for Further Study). Because several schools did not respond to all questions, the total number of "yes" responses was divided by the total number of respondents for each question. A list of comments made by the schools may be found in Appendix G. All respondents received a summary of the findings.
RESULTS

A questionnaire was sent to 59 residential schools for the deaf in the United States. Only the section of the questionnaire inquiring about screening procedures for the Usher Syndrome will be reported here. Forty schools (68%) responded. Fifty-three percent of these schools have screening programs for the Usher Syndrome (Table 1).

Table 1
Percent of Respondants and Screening Programs for the Usher Syndrome

<table>
<thead>
<tr>
<th>Regions</th>
<th>Total Number Contacted</th>
<th>Respondants</th>
<th>Screening Programs</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>N</td>
</tr>
<tr>
<td>West</td>
<td>11</td>
<td>7</td>
<td>64</td>
</tr>
<tr>
<td>Mid West</td>
<td>16</td>
<td>10</td>
<td>63</td>
</tr>
<tr>
<td>Northeast</td>
<td>19</td>
<td>12</td>
<td>63</td>
</tr>
<tr>
<td>South</td>
<td>13</td>
<td>11</td>
<td>85</td>
</tr>
<tr>
<td>Total</td>
<td>59</td>
<td>40</td>
<td>68</td>
</tr>
</tbody>
</table>

aThe percentages below are based on the number of schools that reported.

bOne school indicated that their program will start in the Fall of the 1981-1982 school year.
It was originally thought that the size of the school might determine the existence of a screening program for the Usher Syndrome. However, in scrutinizing the data (Table 2), it is evident that there is little difference in the population groupings.

Table 2
Percent of Schools Having a Screening Program for the Usher Syndrome by Size

<table>
<thead>
<tr>
<th>Number of Students</th>
<th>Number of Schools</th>
<th>Number of Schools with Screening Programs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fewer than 100</td>
<td>1</td>
<td>1 (100)</td>
</tr>
<tr>
<td>100-199</td>
<td>13</td>
<td>7 (54)</td>
</tr>
<tr>
<td>200-299</td>
<td>8</td>
<td>4 (50)</td>
</tr>
<tr>
<td>300-399</td>
<td>8</td>
<td>4 (50)</td>
</tr>
<tr>
<td>400-499</td>
<td>5</td>
<td>2 (40)</td>
</tr>
<tr>
<td>500+</td>
<td>3</td>
<td>3 (100)</td>
</tr>
</tbody>
</table>

Table 3 shows the percent of students who have actually been diagnosed as having the Usher Syndrome. Although some respondents indicated that a number of students were suspected of having the syndrome, this information is not included in the data presented because the etiology of the deaf-blindness could be attributed to the Refsum Syndrome or Rubella. Fifty-one percent of all students diagnosed as having the Usher Syndrome are 16 years old or older, 33% are between the ages of 12 and 15, while only 16% are younger than eleven. There was no
notable difference between the prevalence of male and female subjects. Fifty-five percent are male and 45% are female.

Table 3
Percent of Students with the Usher Syndrome by Age Groups

<table>
<thead>
<tr>
<th>Ages</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>N</td>
<td>N %</td>
</tr>
<tr>
<td>Birth to 5 years old</td>
<td>3</td>
<td>0</td>
<td>3 2</td>
</tr>
<tr>
<td>6 to 11 years old</td>
<td>10</td>
<td>11</td>
<td>21 14</td>
</tr>
<tr>
<td>12 to 15 years old</td>
<td>31</td>
<td>19</td>
<td>50 33</td>
</tr>
<tr>
<td>16 years old to graduation</td>
<td>32</td>
<td>37</td>
<td>76 51</td>
</tr>
<tr>
<td>Total</td>
<td>83</td>
<td>67</td>
<td>150 100</td>
</tr>
</tbody>
</table>

Eighty-one percent of the students have a profound hearing loss which is common with the Usher Syndrome (Table 4).

Table 4
Decibel Loss of Students with the Usher Syndrome

<table>
<thead>
<tr>
<th>db Loss in Better Ear</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>N</td>
<td>N %</td>
</tr>
<tr>
<td>30-50</td>
<td>2</td>
<td>0</td>
<td>2 1</td>
</tr>
<tr>
<td>51-70</td>
<td>8</td>
<td>2</td>
<td>10 7</td>
</tr>
<tr>
<td>71-90</td>
<td>9</td>
<td>8</td>
<td>17 11</td>
</tr>
<tr>
<td>91-110 +</td>
<td>63</td>
<td>58</td>
<td>121 81</td>
</tr>
<tr>
<td>Total</td>
<td>82</td>
<td>68</td>
<td>150 100</td>
</tr>
</tbody>
</table>
There is substantial variation in the frequency with which schools screen their students for the Usher Syndrome (Table 5). Thirty-three percent screen annually, 19% screen every three years and 19% screen upon request or referral. In one school, only Seniors are screened.

**Table 5**

Frequency of Screening for the Usher Syndrome

<table>
<thead>
<tr>
<th>Yearly Intervals</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Annually</td>
<td>7</td>
<td>33</td>
</tr>
<tr>
<td>Every 2 years</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td>Every 3 years</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Every 4 years</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Only Seniors</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Upon Request or Referral Only</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>21</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 6 shows the types of evaluation used. Although 71% of the schools have access to electroretinography (ERG) or electrooculography (EOG), it was not determined which students were given these evaluations.
### Table 6

**Evaluations Used in Screening for the Usher Syndrome**

<table>
<thead>
<tr>
<th>Evaluation</th>
<th>Yes</th>
<th>No</th>
<th>Not Sure</th>
<th>No Response</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>N</td>
<td>%</td>
</tr>
<tr>
<td>Retinoscopy</td>
<td>13</td>
<td>62</td>
<td>5</td>
<td>24</td>
</tr>
<tr>
<td>Ophthalmoscopy</td>
<td>12</td>
<td>57</td>
<td>6</td>
<td>29</td>
</tr>
<tr>
<td>Ophthalmoscopic Exam</td>
<td>14</td>
<td>67</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Visual Field Tests</td>
<td>16</td>
<td>76</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Dark Adaptation Tests</td>
<td>9</td>
<td>43</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td>Electroretinography</td>
<td>11</td>
<td>52</td>
<td>5</td>
<td>24</td>
</tr>
<tr>
<td>Electrooculography</td>
<td>4</td>
<td>19</td>
<td>8</td>
<td>38</td>
</tr>
</tbody>
</table>

**NOTE:** Twenty two schools responded to this question.

Relatives of students who have been identified as having the Usher Syndrome are referred for visual testing by 52% of the schools. Forty-eight percent of the schools provide or refer the student to genetic counseling after he has been diagnosed as having the Usher Syndrome. Thirty-eight percent of the schools provide or refer the student's family for genetic counseling after the student has been identified. Fifty-nine percent of the schools make curricular modifications for the Usher Syndrome student. Fifty percent of the schools provide special vocational counseling to the student.

A list of comments made by individual respondents may be found in Appendix G. The most common other evaluations used in screening include: Titmus, Color.
The most common curricular modifications mentioned include: mobility training, cane training, large print, copies of school work made by copiers rather than ditto machines.

There were no specific vocational modifications mentioned.
DISCUSSION

It is encouraging to see an increasing number of residential schools for the deaf demonstrating concern for identifying students with the Usher Syndrome. However, the data (Table 3) shows evidence that not enough is being done to provide early identification of students with the Usher Syndrome. Students older than age twelve comprise 84% of the diagnosed cases whereas students younger than age eleven comprise only 16%. Because the Usher Syndrome involves a gradual loss of vision, early detection is difficult without a comprehensive screening program, backed up with a knowledgable ophthalmologist. Identification of young children can be improved. It is important to remember that the purpose of screening is to identify individuals who may be in need of a thorough eye examination by a qualified ophthalmologist. It is no longer true that the Usher Syndrome cannot be detected in very young children. Early identification is crucial to provide the maximum support services to the child and his family.

To increase the effectiveness of the screening program, it is recommended that schools provide screening on an annual basis or at least every two years. Gallaudet College has recently published a Screening Program for the Usher Syndrome which is intended for use by schools for the deaf. It provides information for developing a cost-
effective, easily understood screening program which can be performed by the classroom teacher who has had in-service training. It is very important for teachers to be involved in the screening process because they work with the students daily and can provide valuable insights to the student's behavior. It is also important for the teacher to be aware of which of his students has the syndrome in order to provide curricular modifications when necessary, and perhaps informal support, if the student feels comfortable with the teacher.

Hopefully, with improved identification procedures, support services will also increase. The right to be informed cannot be overemphasized. This right includes the family, the student, and the professionals who work with the student. Important decisions and modifications dealing with the individual's personal life, family planning, education and career potential have to be made. Screening for the Usher Syndrome is only the first step toward helping the individual and his family live with and adjust positively to this double handicap.
RECOMMENDATIONS FOR FURTHER STUDY

Further study of the Usher Syndrome is greatly needed. In the area of identification, the following questions need to be answered: the number of diagnosed individuals with the Usher Syndrome currently enrolled in the school; the etiology of the hearing loss of these diagnosed students; the number of relatives who have this genetic disorder; the age of the student when he was identified; the position of the person who made the diagnosis; the family's decision about informing the student of his handicap; and the age of the student when he was informed.

Once the identification program has been established, support services are needed for the student and his family. The researcher interested in this area should investigate: specific curricular and vocational modifications being made; the progression of these modifications in relation to the student's visual deterioration; the availability of genetic counseling services to the student and his family; and specific issues involved in personal counseling.

Answers to these questions could be of great value to programs for the deaf and to rehabilitation agencies which are currently serving clients with the Usher Syndrome. Research on the Usher Syndrome could then be shared with
the National Retinitis Pigmentosa Foundation and/or the Helen Keller National Center which could serve as the umbrella agency for the Usher Syndrome.
APPENDIX A

A RECOMMENDED BIBLIOGRAPHY*

*These references are in addition to those found in the Reference Section
A RECOMMENDED BIBLIOGRAPHY


Fellendorf, G. W. One Man's Search of the Literature on Usher's Syndrome. Workshop on Usher's Syndrome,


Prause, R. J. Placement of Individuals with Usher's Syndrome. Workshop on Usher's Syndrome, Gallaudet
Ranier, J. D. Psychiatric Aspects of Usher Syndrome.  


Spainer, J. Evaluating the Individual's Potential.  
APPENDIX B

COVER LETTER SENT TO RESIDENTIAL SCHOOLS FOR THE DEAF
Dear Colleague:

I am involved in a graduate project on the evaluation programs of hearing-impaired students for visual acuity and the Usher's Syndrome. This survey/questionnaire is being sent to all the residential schools for the deaf in the United States. In order for the study to be valid and meaningful, it is very important that I receive as many responses from as many residential schools as possible. Confidentiality will be respected. Data will be presented by percentages of total responses. I would greatly appreciate your cooperation in answering the enclosed questionnaire. If you would like to receive a copy of my findings, please check the last statement on the questionnaire.

Your cooperation is urgently needed and appreciated. Thank you very much.

Sincerely yours,

Creagh Walker Day

California State University, Northridge
20th NLTP Class
APPENDIX C

QUESTIONNAIRE SENT TO RESIDENTIAL SCHOOLS FOR THE DEAF
STATEMENT OF PURPOSE:

The intent of this survey is to discover what residential schools for the deaf are doing to evaluate their hearing-impaired students for visual acuity and for the Usher's Syndrome. "Usher's Syndrome is a genetic condition resulting in the double handicap of congenital deafness and a progressive blindness known as retinitis pigmentosa."

I. GENERAL BACKGROUND INFORMATION:

A. Name of School -

B. Position of Respondant -

C. Please indicate the number of hearing-impaired students in your school for the categories below:

1. Total number of hearing-impaired students
2. Total number of hearing-impaired students between the ages of birth and 5 years old
3. Total number of hearing-impaired students between the ages of 6 and 11 years old
4. Total number of hearing-impaired students between the ages of 12 and 15 years old
5. Total number of hearing-impaired students between the age of 16 and graduation

II. SCREENING FOR VISUAL ACUITY:

A. Please check the following questions with yes, no, or not sure.

1. Are all students enrolled in your school evaluated for visual acuity?
2. If yes, please indicate the frequency with which students are evaluated for visual acuity. (Check yes, no or not sure for each time period.)
   a. Upon admission to school
   b. Upon request or referral
   c. Annually
   d. Every _______ years (Please write the yearly interval in the blank.)
B. What is the position of the person who conducts the testing for visual acuity?


C. Below is a list of evaluations. Please answer yes, no, or not sure by the name of each evaluation depending on whether or not the evaluation is used in testing for visual acuity.

<table>
<thead>
<tr>
<th>Evaluation</th>
<th>YES</th>
<th>NO</th>
<th>NOT SURE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Questionnaires</td>
<td>1.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Snellen Chart</td>
<td>2.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Cover Test</td>
<td>3.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Ophthalmoscopy</td>
<td>5.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Other, please specify</td>
<td>8.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

II. INCIDENCE OF THE USHER'S SYNDROME:

A. How many students in your school have been diagnosed as having the Usher's Syndrome in the following age groups:

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Number of Students</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Birth to 5 years old</td>
<td>MALE</td>
</tr>
<tr>
<td>2. 6 to 11 years old</td>
<td>1.</td>
</tr>
<tr>
<td>3. 12 to 15 years old</td>
<td>2.</td>
</tr>
<tr>
<td>4. 16 years old to graduation</td>
<td>3.</td>
</tr>
<tr>
<td>5. 16 years old to graduation</td>
<td>4.</td>
</tr>
</tbody>
</table>
B. How many students with the Usher's Syndrome have a decibel loss in their better ear in the following ranges:

<table>
<thead>
<tr>
<th>NUMBER OF STUDENTS WITH THE USHER'S SYNDROME</th>
</tr>
</thead>
<tbody>
<tr>
<td>MALE</td>
</tr>
<tr>
<td>------</td>
</tr>
<tr>
<td>1. 30-50 dB loss in better ear</td>
</tr>
<tr>
<td>2. 51-70 dB loss in better ear</td>
</tr>
<tr>
<td>3. 71-90 dB loss in better ear</td>
</tr>
<tr>
<td>4. 91-110+ dB loss in better ear</td>
</tr>
</tbody>
</table>

IV. SPECIFIC EVALUATION FOR THE USHER'S SYNDROME:

A. Please answer all the following questions with yes, no or not sure.

1. Does your school test students specifically for the Usher's Syndrome?

   If your answer is no, please return the questionnaire in the stamped-addressed envelope provided. Thank you for your time and assistance.

   If your answer is yes or not sure, please answer the following questions.

2. Whom do you evaluate for the Usher's Syndrome?

   a. All hearing-impaired students
   b. All congenitally hearing-impaired students
   c. All suspected Usher's Syndrome students
   d. All high-risk hearing-impaired students (any student who has a history of the Usher's Syndrome in his family)
   e. All hearing-impaired students of consanguineous parentage (descended from the same parent or ancestor)
   f. Other, please explain

   ______________________________________________________
3. What is the frequency with which your school evaluates students for the Usher's Syndrome? Please check yes, no or not sure.

<table>
<thead>
<tr>
<th></th>
<th>YES</th>
<th>NO</th>
<th>NOT SURE</th>
</tr>
</thead>
<tbody>
<tr>
<td>a.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>b.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>c.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>d.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>e.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

a. Upon admission to school
b. Upon request or referral
c. Annually
d. Every _______ years (please write the yearly interval in the blank)
e. Other, please explain ______________________

B. What is the position of the person who conducts the testing for the Usher's Syndrome?

C. Below is a list of evaluations. Please check yes, no or not sure by the name of each evaluation depending on whether or not the evaluation is used in testing for the Usher's Syndrome.

<table>
<thead>
<tr>
<th></th>
<th>YES</th>
<th>NO</th>
<th>NOT SURE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>3.</td>
<td></td>
<td></td>
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<tr>
<td>4.</td>
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<td>5.</td>
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<tr>
<td>6.</td>
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<td></td>
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<td>7.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

1. Retinoscopy
2. Ophthamoscopy
3. Ophthamoscopic Examination
4. Visual Field Tests
5. Dark Adaptation Tests
6. Electroretinography (ERG)
7. Electrooculography (EOG)
8. Other, please explain ______________________
D. Please answer the following questions with yes, no or not sure:

<table>
<thead>
<tr>
<th>YES</th>
<th>NO</th>
<th>NOT</th>
<th>SUPE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

1. Does your school refer relatives of hearing-impaired students who have been identified as having the Usher's Syndrome for visual testing?

2. Does your school provide genetic counseling to the student after he/she has been diagnosed as having the Usher's Syndrome?

3. Does your school provide genetic counseling to the student's family after the student has been diagnosed as having Usher's Syndrome?

4. Does your school make curricular modifications for the student who has been diagnosed as having the Usher's Syndrome? If yes, please explain below:

________________________________________________________________________
________________________________________________________________________
________________________________________________________________________

5. Does your school provide special vocational counseling to the student who has been diagnosed as having the Usher's Syndrome? If yes, please explain below:

________________________________________________________________________
________________________________________________________________________
________________________________________________________________________

E. If funding were not a problem, what changes would you make in your testing program for the Usher's Syndrome?

________________________________________________________________________
________________________________________________________________________
________________________________________________________________________

If you have any additional comments that you would like to make, please feel free to do so on the back of this page. Thank you very much for your time, assistance, and cooperation. If you would like to receive a copy of my findings, please check below.

_______ I would like to receive a copy of the findings of this study.
APPENDIX D

LIST OF RESIDENTIAL SCHOOLS FOR THE DEAF
LIST OF RESIDENTIAL SCHOOLS FOR THE DEAF

Alabama Institute for Deaf and Blind
205 East South Street
Talladega, Alabama 35160
(205) 362-1500
President: Jack Hawkins, Jr., Ph.D.

Arizona State School for Deaf and Blind
1200 West Speedway
Post Office Box 5545
Tucson, Arizona 85703
(602) 882-5357
Superintendent: Ralph L. Hoag, Ed.D.

Arkansas School for the Deaf
2400 West Markham
Post Office Box 3811
Little Rock, Arkansas 72205
(501) 371-1555
Superintendent: Tom J. Hicks, Ed.S.

California School for the Deaf
39350 Gallaudet Drive
Fremont, California 94501
(415) 845-4629
Superintendent: Henry Klopping, Ed.D.

California School for the Deaf
3044 Horace Street
Riverside, California 92506
(714) 683-8140
Superintendent: Robert K. Lennan, Ed.D.

Colorado School for Deaf and Blind
Kiowa and Institute Streets
Colorado Springs, Colorado 80903
(303) 636-5186
Superintendent: Robert T. Dawson, M.S.

American School for the Deaf
139 North Main Street
West Hartford, Connecticut 06107
(203) 236-4149
Executive Director: Ben E. Hoffmeyer, LHD
Margaret S. Sterck School for the Hearing Impaired
Chestnut Hill Road
Newark, Delaware 19713
(302) 454-2302
Director: F. Eugene Thomure, Ed.D.

Model Secondary School for the Deaf
Florida Avenue at 7th Street, N.E.
Washington, D.C. 20002
(202) 651-5800
Vice-President, Pre-College Programs: R. Davila, Ph.D.

The Florida School for the Deaf and Blind
San Marco Avenue
Post Office Box 1209
St. Augustine, Florida 32084
(904) 824-1654
President: William J. McClure, M.A.

Fort Lauderdale Oral School
3100 S.W. 8th Avenue
Fort Lauderdale, Florida 33315
(305) 525-7251
Director: Florence A. Ras, M.A.

Georgia School for the Deaf
Cave Spring, Georgia 30124
(404) 777-3310
Superintendent: J. H. Whitworth, M.Ed.

Hawaii School for the Deaf and the Blind
3440 Leahi Avenue
Honolulu, Hawaii 96815
(808) 734-0297
Principal: Santiago Agcaoili, M.S.

Idaho State School for the Deaf and Blind
202 14th Avenue, East
Gooding, Idaho 83330
(208) 934-4457
Superintendent: Keith W. Tolzin, M.S.

Illinois School for the Deaf
125 South Webster
Jacksonville, Illinois 62650
(217) 245-5141
Superintendent: William Page Johnson, Ph.D.
Iowa School for the Deaf
1600 South Highway 275
Council Bluffs, Iowa 51501
(712) 366-0571
Superintendent: Joseph Giangreco, Ed.D.

Kansas State School for the Deaf
450 East Park Street
Post Office Box 471
Olathe, Kansas 66061
(913) 782-2530
Superintendent: Gerald L. Johnson, M.S.

The Kentucky School for the Deaf
South 2nd Street
Post Office Box 27
Danville, Kentucky 40422
(606) 236-5132
Superintendent: Winfield McChord, Jr., M.S.

Louisiana School for the Deaf
Post Office Box 3074
Baton Rouge, Louisiana 70821
(504) 342-6350
Superintendent: Harvey Jay Corson, Ed.D.

Governor Baxter School for the Deaf
Post Office Box 799, Mackworth Island
Portland, Maine 04104
(207) 781-3165
Superintendent: Joseph P. Youngs, Jr., Litt.D.

Maryland School for the Deaf (Columbia Campus)
Post Office Box 894
Columbia, Maryland 21044
(301) 465-9611
Superintendent: David M. Denton, Ph.D.

Maryland School for the Deaf (Frederick Campus)
101 Clarke Place
Frederick, Maryland 21701
(301) 662-4159
Superintendent: David M. Denton, Ph.D.

Beverly School for the Deaf
6 Echo Avenue
Beverly, Massachusetts 01915
(617) 927-7070
Executive Director: Joseph W. Panko, Ph.D.
The Clarke School for the Deaf
Round Hill Road
Northampton, Massachusetts 01060
(413) 584-3450
President: George T. Pratt, L.H.D.

The Boston School
800 North Main Street
Randolph, Massachusetts 02368
(617) 963-8150
Superintendent: Hollis W. Wyks, M.A.

Michigan School for the Deaf
West Court and Miller Roads
Flint, Michigan 48502
(313) 238-4621
Superintendent: Robert R. Gates, Ph.D.

Minnesota School for the Deaf
Post Office Box 308
Faribault, Minnesota 55021
(507) 334-6411 X369
Research School Administrator: Melvin Brasel, M.A.

Mississippi School for the Deaf
1253 Eastover Drive
Jackson, Mississippi 39211
(601) 366-0315
Superintendent: Alma L. Alexander, Ed.D.

Missouri School for the Deaf
5th and Vine Streets
Fulton, Missouri 65251
(314) 642-3301
Superintendent: Peter H. Ripley, M.A.

Montana State School for the Deaf and Blind
3911 Central Avenue
Great Falls, Montana 59401
(406) 453-1401
Superintendent: Floyd J. McDowell, M.A.

Nebraska School for the Deaf
3223 North 45th Street
Omaha, Nebraska 68104
(402) 554-2155
Superintendent: George W. Collins, M.A.
Marie H. Katzenbach School for the Deaf  
320 Sullivan Way  
West Trenton, New Jersey 08628  
(609) 883-2600  
Superintendent: Philip E. Cronlund, M.A.  

New Mexico School for the Deaf  
1060 Cerrillos Road  
Santa Fe, New Mexico 87501  
(505) 983-3321  
Superintendent: James A. Little, M.S.  

St. Mary's School for the Deaf  
2253 Main Street  
Buffalo, New York 14214  
(716) 834-7200  
Superintendent: Sister Nora Letourneau, Ph.D.  

Lexington School for the Deaf  
30th Avenue and 75th Street  
Jackson Heights, New York 11370  
(212) 899-8800  
Executive Director: Leo E. Connor, Ed.D.  

Rochester School for the Deaf  
1545 St. Paul Street  
Rochester, New York 14621  
(716) 544-1240  
Superintendent: Leonard G. Zwick, M.A., M.Ed.  

New York State School for the Deaf  
401 Turin Street  
Rome, New York 13440  
(315) 337-8400  
Superintendent: Robert Seibold, M.A., M.S.Ed.  

New York School for the Deaf  
555 Knollwood Road  
White Plains, New York 10603  
(914) 949-7310  
Superintendent: Kendall D. Litchfield, M.A.  

Central North Carolina School for the Deaf  
Summit Avenue, Extension North  
Post Office Box 6070  
Greensboro, North Carolina 27405  
(919) 621-6490  
Superintendent: John W. Hudson, Jr., M.S.
North Carolina School for the Deaf
Rutherford Road, Highway #64
Morganton, North Carolina 28655
(704) 433-2955
Director of North Carolina Schools for the Deaf: Rance Henderson, L.H.D.

Eastern North Carolina School for the Deaf
Highway 301 North
Wilson, North Carolina 27893
(919) 237-2450
Superintendent: R. M. McAdams, B.S.

North Dakota School for the Deaf
Devils Lake, North Dakota 58301
(701) 662-5031
Superintendent: Allen J. Hayek, M.S., M.A.

Ohio School for the Deaf
500 Morse Road
Columbus, Ohio 43214
(614) 888-1550
Superintendent: Edward C. Grover, M.A.

Oklahoma School for the Deaf
East 10th and Tahlequah Streets
Sulphur, Oklahoma 73086
(405) 622-3186
Superintendent: David F. Kamphaus, M.S.

Oregon State School for the Deaf
999 Locust Street, N.E.
Salem, Oregon 97303
(503) 378-3825
Director: Bill J. Peck, M.Ed.

Pennsylvania School for the Deaf
7500 Germantown Avenue
Philadelphia, Pennsylvania 19119
(215) 247-9700
Headmaster: Joseph P. Finnegan, Jr., M.A.

Western Pennsylvania School for the Deaf
300 East Swissvale Avenue
Pittsburgh, Pennsylvania 15218
(412) 371-7000
Superintendent: William N. Craig, Ph.D.
Scranton State School for the Deaf
1800 North Washington Avenue
Scranton, Pennsylvania 18509
(717) 961-4546
Superintendent: Victor H. Galloway, Ed.D.

South Carolina School for the Deaf and Blind
Highway 56
Spartanburg, South Carolina 29302
(803) 585-7711
President: N. F. Walker, M.A.

South Dakota School for the Deaf
1800 East 10th Street
Sioux Falls, South Dakota 57103
(605) 339-6700
Superintendent: Gordon L. Kaufman, M.A.

Tennessee School for the Deaf
2725 Island Home Boulevard
Knoxville, Tennessee 37920
(615) 577-7581
Superintendent: William E. Davis, M.S.

Texas School for the Deaf
1102 South Congress
Austin, Texas 78704
(512) 442-7821
Superintendent: Virgil E. Flathouse, Ph.D.

Utah Schools for the Deaf and Blind
846 20th Street
Ogden, Utah 84401
(801) 399-9631
Superintendent: Harlan M. Fulmer, M.S.

The Austine School for the Deaf
120 Maple Street
Brattleboro, Vermont 05301
(802) 254-4571
Headmaster: Richard K. Lane, CAOS

Virginia School for the Deaf, Hampton
700 Shell Road
Hampton, Virginia 23661
(804) 245-0052
Superintendent: Philip A. Bellefleur, Ph.D.
Virginia School for the Deaf and Blind  
East Beverly Street  
Staunton, Virginia 24401  
(703) 885-9533  
Superintendent: Sheldon O. Melton, M.A.

Washington State School for the Deaf  
611 Grand Boulevard  
Post Office Box 5187  
Vancouver, Washington 98668  
(206) 696-6525  
Superintendent: Archie Stack, M.Ed.

West Virginia Schools for the Deaf and the Blind  
Romney, West Virginia 26757  
(304) 822-3521  
Superintendent: Jack W. Brady, M.Ed.

The Wisconsin School for the Deaf  
309 West Walworth Avenue  
Delavan, Wisconsin 53115  
(414) 728-2677  
Superintendent: John S. Shipman, M.A.

St. John's School for the Deaf  
3680 South Kinnickinnic Avenue  
Milwaukee, Wisconsin 53207  
(414) 744-0136  
Director: Reverend Donald F. Zerker, M.A.
APPENDIX E

RESULTS OF SCREENING FOR VISUAL ACUITY
## FREQUENCY OF SCREENING FOR VISUAL ACUITY

<table>
<thead>
<tr>
<th></th>
<th>No Answer</th>
<th>Yes</th>
<th>No</th>
<th>Not Sure</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N   %</td>
<td>N   %</td>
<td>N %</td>
<td>N   %</td>
</tr>
<tr>
<td>Upon Admissions</td>
<td>11  29</td>
<td>20  53</td>
<td>6  16</td>
<td>1  2</td>
</tr>
<tr>
<td>Upon Request/Referral</td>
<td>11  29</td>
<td>27  71</td>
<td>0   0</td>
<td>0   0</td>
</tr>
<tr>
<td>Annually</td>
<td>0    0</td>
<td>25 66</td>
<td>13  44</td>
<td>0  0</td>
</tr>
<tr>
<td>Every two years</td>
<td>0    0</td>
<td>8  21</td>
<td>30  79</td>
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<tr>
<td>Every three years</td>
<td>0    0</td>
<td>5  13</td>
<td>33  87</td>
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## EVALUATIONS USED FOR VISUAL SCREENING

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<th></th>
<th>Number of Responses</th>
<th>No Answer</th>
<th>Yes</th>
<th>No</th>
<th>Not Sure</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N   %</td>
<td>N   %</td>
<td>N %</td>
<td>N %</td>
<td>N %</td>
</tr>
<tr>
<td>Questionnaires</td>
<td>32 84%</td>
<td>6 16</td>
<td>14 37</td>
<td>16 42</td>
<td>2 5</td>
</tr>
<tr>
<td>Snellen</td>
<td>38 100%</td>
<td>0   0</td>
<td>33 87</td>
<td>4 11</td>
<td>1 3</td>
</tr>
<tr>
<td>Cover</td>
<td>31 82%</td>
<td>7 18</td>
<td>17 45</td>
<td>11 29</td>
<td>3 8</td>
</tr>
<tr>
<td>Retinoscopy</td>
<td>30 79%</td>
<td>8 21</td>
<td>10 26</td>
<td>19 50</td>
<td>1 3</td>
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<tr>
<td>Ophthalmoscopy</td>
<td>32 84%</td>
<td>6 16</td>
<td>13 34</td>
<td>18 47</td>
<td>1 3</td>
</tr>
<tr>
<td>Visual Field</td>
<td>31 82%</td>
<td>7 18</td>
<td>17 45</td>
<td>12 32</td>
<td>2 5</td>
</tr>
<tr>
<td>Dark Adaptation</td>
<td>28 74%</td>
<td>10 26</td>
<td>6 16</td>
<td>20 53</td>
<td>2 5</td>
</tr>
<tr>
<td>Other</td>
<td>14 37%</td>
<td>24 63</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
APPENDIX F

REGIONS OF THE UNITED STATES AS ESTABLISHED BY THE NATIONAL ASSOCIATION OF THE DEAF
APPENDIX G

COMMENTS MADE BY RESPONDANTS
1. Students are supposed to have an eye examination by their family optometrist or ophthalmologist on a yearly basis.

2. If the child fails the Snellen Chart and Cover Test during screening for visual acuity, then other tests are conducted by the doctor.

3. Our school does not test students specifically for the Usher Syndrome unless history indicates it or a documented change of visual acuity is seen.

4. All who do not pass the Snellen Chart are referred for in-depth Ophthalmological Evaluation.

5. In January of 1978 the entire school population was screened for the Usher Syndrome. There are no current plans for further screening.

6. The diagnostic teacher is responsible for the administration of Retinitis Pigmentosa Screening. (She administers R. P. Screening, but uses none of the formal tests listed in Question IV.C.)
1. The student is not presently receiving curricular modifications, but is being observed and evaluated every six months.

2. Curricular modifications include: individual and small group counseling; preferential seating; large print when needed on worksheets, etc.; outside speakers to speak to the whole student body on Usher's Syndrome.

3. Curricular modifications include: dark ditto papers; appropriate colored chalk for the chalkboard; darker lead pencils; changed class arrangements, etc.

4. The student is given support services in the Deaf-Blind Unit if necessary.

5. Notetakers are provided to students who have the Usher Syndrome.

6. Curricular modifications include: classroom seating modifications (3-5 feet); use white chalk only; large script writing; reduce class visual distraction; concentration on manual communication rate of transmission; large print materials; individualized testing.
1. When the student moves into High School, vocational counseling will be needed.
2. Plans are being made for vocational counseling.
3. We try to steer the students away from visually oriented vocations.
4. Our school does not provide the student with vocational counseling because of parental resistance.
5. Vocational Counseling is provided through the State Commission for the Blind and Vocational Rehabilitation.
6. Both the Guidance Department personnel and staff affiliated with the Educational Support/Field Services Division provide specialized psychological and vocational counseling services and follow up for diagnosed Usher's Syndrome students.
7. Our school does not provide genetic counseling, but does provide general counseling to the student and his family about their problems and feelings about their visual handicap.
REFERENCES
REFERENCES


Roehrig, A. A. Steps for a Deaf Person's Adjustment to Blindness. Paper presented at the Tenth Southeast Regional Institute on Deafness, Nashville, Tennessee, November 18, 1980.


