AUDIOMETRIC AND VESTIBULAR EXAMINATIONS IN RETINITIS PIGMENTOSA*

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Deafness is one of the most frequent degenerative processes associated with retinitis pigmentosa (Duke-Elder, 1940; Fowler, 1947), the frequency being variously reported as between 2 and 43 per cent. (Azzolini and Cortesi, 1951). These variations in the incidence in different series may be due to the diversity of methods employed to measure hearing and different criteria in evaluating the degree of impairment. Since the introduction of audiometry as a routine method of measuring hearing loss, the number of reports of affected hearing audiometrically examined in many diseases has greatly increased. In retinitis pigmentosa however, the reported cases which have been examined audiometrically were almost always in small series (Sirles and Slaughter, 1943; Givner and Bruger, 1947; Azzolini and Cortesi, 1951; dell'Acqua, 1952; Bietti, 1952; Alagna and Nizetic, 1953; Lavalle, 1953).

During the past 3 years it has been possible to perform audiometric tests on a number of patients and vestibular tests in over half of them. The results of this study are now presented.

Materials and Methods

The material comprises 22 patients with retinitis pigmentosa, of whom four (two brothers and two sisters) were siblings; sixteen were males and six females. Audiometric tests were carried out in all the patients and vestibular tests in fourteen.

In addition, four children of patients with retinitis pigmentosa showing only impairment of scotopic vision were tested audiometrically; three were children of a female patient and the other of a male patient. The ages varied from 11 to 56 years, and the four offspring were aged 6 to 16. All audiometric examinations were performed by the same technician using an E-1 Maico audiometer, routine clinical examination of the ears, nose and throat having previously been made. Both air and bone conductions were tested, masking being used when necessary. Vestibular tests were performed by caloric stimulation, using cold water.

Results

The main results are summarized in the Table. Fourteen of the 22 patients examined showed abnormal audiometric curves. Nine showed a typical deafness of perception type. The range of this impairment is illustrated in the Figures, Fig. 1 being taken from the mildest case, and Fig. 2 from one of the severe cases. One patient was totally deaf, two showed deafness of mixed type, and two of conduction type (Fig. 3). In all four children of patients, the audiometric tests were within normal limits.

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## TABLE

### AUDIOMETRIC FINDINGS IN RETINITIS PIGMENTOSA

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age</th>
<th>Sex</th>
<th>Audiometric Examination</th>
<th>Vestibular Examination</th>
<th>Remarks</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>44</td>
<td>F</td>
<td>Normal</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>31</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>28</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td></td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>32</td>
<td>F</td>
<td>Nerve deafness bilateral</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>40</td>
<td>F</td>
<td>Nerve deafness bilateral</td>
<td>Hypo-excitability bilateral</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>17</td>
<td>M</td>
<td>Normal</td>
<td></td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>19</td>
<td>M</td>
<td>Normal</td>
<td></td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>42</td>
<td>M</td>
<td>Normal</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>22</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td>Normal caloric reaction</td>
<td>Parents first cousins</td>
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<tr>
<td>10</td>
<td>45</td>
<td>M</td>
<td>Normal</td>
<td>Normal caloric reaction</td>
<td>—</td>
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<tr>
<td>11</td>
<td>26</td>
<td>F</td>
<td>Normal</td>
<td></td>
<td>—</td>
</tr>
<tr>
<td>12</td>
<td>35</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td>Hypo-excitability left</td>
<td>—</td>
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<tr>
<td>13</td>
<td>16</td>
<td>M</td>
<td>Conduction deafness bilateral</td>
<td>—</td>
<td>Laurence-Moon-Biedl syndrome</td>
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<tr>
<td>14</td>
<td>36</td>
<td>M</td>
<td>Mixed deafness bilateral</td>
<td></td>
<td>Chronic otitis media right Adhesive process left</td>
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<tr>
<td>15</td>
<td>56</td>
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<td></td>
<td>—</td>
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<tr>
<td>16</td>
<td>46</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td>Normal caloric reaction</td>
<td>Enucleation of right eye for glaucoma</td>
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<tr>
<td>17</td>
<td>33</td>
<td>M</td>
<td>Nerve deafness bilateral</td>
<td>Normal caloric reaction</td>
<td>Atypical retinitis pigmentosa</td>
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<tr>
<td>18</td>
<td>23</td>
<td>M</td>
<td>Normal</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
<tr>
<td>19</td>
<td>56</td>
<td>M</td>
<td>Mixed deafness bilateral</td>
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<td>—</td>
</tr>
<tr>
<td>20</td>
<td>13</td>
<td>M</td>
<td>Total deafness</td>
<td>Hypo-excitability bilateral</td>
<td>—</td>
</tr>
<tr>
<td>21</td>
<td>11</td>
<td>F</td>
<td>Conduction deafness bilateral</td>
<td>Hypo-excitability bilateral</td>
<td>Retinitis albescens</td>
</tr>
<tr>
<td>22</td>
<td>32</td>
<td>F</td>
<td>Normal</td>
<td>Normal caloric reaction</td>
<td>—</td>
</tr>
</tbody>
</table>
Fourteen patients out of the total of 22 also underwent vestibular tests. Four were abnormal, three showing hypo-excitability on both sides, and the fourth on one side only; all those with abnormal reactions also had impairment of hearing.

**Discussion**

The occurrence of simultaneous visual and acoustic lesions in the degenerative disease of retinitis pigmentosa has been widely discussed. Most
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FIG. 3.—Audiogram of a case of conduction type deafness.

authors (Franceschetti and Klein, 1951) are of the opinion that these lesions are due to the "polyphemic action of a mutated gene". It may be assumed (Kjerrumgaard, 1948) that developmental anomalies of widely different nature and localized in various parts of the body have a common point of time of attack of the gene-determined lesion in embryonic life. Thus such a lesion might as well occur in the receptive organ of sight as in that of hearing. Support for such a theory is afforded by the findings of Siebenmann and Bing (1907) who demonstrated similar histological changes in the retina and in the organ of Corti of a deaf mute with retinitis pigmentosa.

Of the 22 patients examined, two showed deafness of a conductive type with, however, normal otoscopic findings and no history of ear discharge. A clinical diagnosis of otosclerosis was made in these cases who were not related. One of them showed the syndrome of Laurence-Moon-Biedl and the other, retinitis punctata albescens. The combination of retinitis pigmentosa with otosclerosis in a family has been reported by Franceschetti and Klein (1948) who quote a previous report by Usher and Shennan (1930-31). It must be considered however that otosclerosis is quite common, and may occur in conjunction with a variety of other diseases, so that an association with retinitis pigmentosa could be fortuitous.

Ten of the fourteen patients examined showed normal caloric reactions. Six of them however had nerve deafness. These findings do not correspond with the statement of Fowler (1947), who writes that irritability of the vestibular apparatus is usually diminished or lacking in cases with nerve deafness and retinitis pigmentosa.

The fact that the four children of patients with retinitis pigmentosa, whose audiometric tests were normal, showed impairment of dark adaptation is
worthy of note. No definite conclusions can be drawn, however, from so few cases. It may be assumed that the impairment of dark adaptation, in families with retinitis pigmentosa, is more frequent than impairment of acoustic function.

Summary

Fourteen out of 22 patients, aged 11 to 56 years and suffering from retinitis pigmentosa, showed impairment of hearing when tested audiometrically. Vestibular tests were performed in fourteen cases and ten of these gave a normal caloric response.

Four offspring of two patients with retinitis pigmentosa showed normal audiometric values although their scotopic vision was impaired.

The relationship between the visual and acoustic lesions is discussed. It is suggested that impairment of scotopic vision is more common than disturbed acoustic function in the children of patients with retinitis pigmentosa.

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J. Landau and M. Feinmesser

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