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PIGMENTARY ABNORMALITY IN CHILDREN CONGENITALLY DEAF FOLLOWING MATERNAL GERMAN MEASLES.

By E. O. Marks, Brisbane.

It is with reluctance that one adds to the store of literature1 which has followed Dr. N. McAllister Gregg's original contribution on the subject of congenital cataracts following German measles in the mother. The observations submitted now seem, however, to have some fresh and material bearing on the subject, and indicate that ophthalmologists will need in future to keep in mind rubella and possibly other exanthemata also as causes of abnormal fundus conditions.

Epidemics of rubella which occurred in Sydney and Adelaide were followed by "epidemics" of congenital cataract, deafness and cardiac and other defects amongst the babies whose mothers had contracted the rubella. Rubella epidemics occurred in Brisbane also, but there was not the ensuing epidemic of cataracts. There were a very few cataractous babies with the maternal rubella history, but the opaque capsular type of cataract which these showed is as common here as in the mainland, and the numbers were not significantly increased. There did cause very serious "epidemics" of congenital deafness, as has been recorded by Dr. L. F. Winterbotham,2 who found associated cardiac defects.

I suggested to Mr. S. E. Holle, the superintendent of the Blind and Deaf School, and to Dr. Winterbotham, that though the children were in the school because of their deafness and not because of any visual disabilities, it would be of interest to examine their eyes for abnormalities.

The result has more than surprised me. While cataracts were very few, nearly half the deaf children born in the epidemic years showed marked pigmentary abnormality, while those born in other years had normal fundi. Of 26 deaf children with a history of maternal rubella 17 had abnormally pigmented fundi and four had lens opacities. Of 25 deaf children with abnormally pigmented fundi or cataracts 21 had the maternal rubella history, but it must be remembered that with such a mild sickness as rubella the absence of a history does not imply the absence of the disease.

The most striking fact is that the abnormal fundus occurred only in the epidemic years, as will be seen in the accompanying tables.

**Table I.**

<table>
<thead>
<tr>
<th>Group</th>
<th>Number of Males</th>
<th>Number of Females</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deaf children examined</td>
<td>56</td>
<td>45</td>
<td>101</td>
</tr>
<tr>
<td>Children with pigmentary abnormality</td>
<td>17</td>
<td>14</td>
<td>31</td>
</tr>
<tr>
<td>Children with lens opacities</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Children with normal eyes</td>
<td>16</td>
<td>80</td>
<td>96</td>
</tr>
</tbody>
</table>

I find it difficult to give a satisfactory word picture of the ophthalmoscopic appearance. The nearest I would compare it with is an atypical retinitis pigmentosa in which the pigment disarrangement has not the "bone-corpuscle" form. The maculae were coarsely mottled. The retinal vessels and the optic discs appeared normal, and the vision had not been affected sufficiently to interfere with the education or to be noticed as defective by the teachers.

**ACKNOWLEDGMENT.**

In conclusion, grateful acknowledgement is due to Mr. S. E. Holle both for arranging for the examination with the parents and for his help in working out the results.

**REFERENCES.**

1 See extensive list of references appended to paper by Charles Swan and A. L. Todd in The Medical Journal of Australia, May 17, 1946, page 446.


**DISCUSSION.**

The present paper added what was the relationship between deafness and cataract. In one year the proportion of cataract cases had been high and in another the...
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The most striking fact is that the abnormal fundi occurred only in the epidemic years, as will be seen in the accompanying tables.

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DISCUSSION.

The P hi o sh e asked what was the relationship between deafness and cataract. In one year the proportion of cataract cases had been high, but in another the
proportion of deafness cases. He referred to a report in a recent British Medical Bulletin.

E. O. Marks reported that there were four cataractous children amongst the 43 deaf children born in 1935. They had been included in the 20 of that year with pigmentary abnormality.

N. Mcdonald Green (Sydney) thanked Dr. Marks for his interesting paper. These pigmentary changes were quite common in the fundus of a child of particular interest. There were changes that had been noted by several observers in connection with these cases. In the original paper read by many years ago in 1939, the following words had appeared in the unhealthy appearance of the iris in some cases suggests that there might be possible some shafts in the choroid. Looking back into his notes for that paper, he had found that the arteriole in the affected vessels, his case of left-sided macular changes, had appeared pale, and some scattered irregular spots of pigment were observed. Credit therefore was given to him for the first observation of these changes.

Swan and his colleagues, in their paper in The Medical Journal of Australia of September 31, 1943, noted multiple small pigmented spots on the fundus of the unaffected eye in one case of unilateral cataract. The first opportunity the speaker had had of observing the fundus in 4 of these cases was in relation to the investigation carried out under the authority of the Director-General of Public Health, New South Wales, of 52 children, the majority suffering from deaf mutism, some from cataract, while some had both conditions. Marked pigmentary changes had been seen in sixteen cases, not only in cases of deaf mutism, but also in the unaffected eye in cases of unilateral cataract. In one case of cataract, in which treatment had been originally carried out by Macdonald and in which a good opening had been secured, the typical pigmentary changes were seen in the affected eye.

The speaker could not recall ever having seen a description of a fundal appearance similar to the one observed in this connection. In order to indicate to the members what they had tried to describe the appearance. "Monotony" was the term he would like to use, perhaps "blotchiness" would be preferable. The area affected included the macula and an area of about three to four arc diameters surrounding it. In a few cases the disturbance was more widely distributed. He hoped that in the near future his attention will be drawn to similar cases. It was like a piece of coarse Scotch tweed used for a sports coat over which a shirt had been thrown. The spots of pigment were discrete. An interesting feature of Dr. Marks' paper was that the time incidence of the epidemic of rubella in Queensland varied as compared with that in New South Wales. The big Queensland epidemic occurred in 1931; in New South Wales the first epidemic was 1932-1934 and the second 1941-1943. He would like to know from Dr. Marks the ratio of cases of deaf mutism to those of cataract. The incidence of both cases of deaf mutism in New South Wales was very high, especially in the first epidemic. He did not know whether the proportion was as high in the second epidemic; one had not heard of many such cases. It still appeared as if the time factor that was considered infection was of importance in the nature of the congenital defect in the infant. He had seen many patients showing cataracts and deaf mutism. These pigmentary disturbances had been noted in some recent articles in the American journals.

The condition did not appear to have any adverse effect on vision. He felt that the future some of these cases would be discovered by the course of an ocular examination, and that their distinctive appearance would lead to a correct diagnosis.

G. H. Barry (Adelaide) referred to a case in South Australia which would be described in a paper, at present in the hands of the publishers, by Swan and others. The child, now three or four years old, had a monocular cataract when first seen at an early age. Subsequently, though no operation had been performed, the cataractous lens was found to have absorbed spontaneously. Because he seemed to have had a traumatic vision in the "good" eye, the child was given a general anaesthetic and the fundus showed what might be regarded as a gross exaggeration of the condition described by Dr. Marks, there being patchy areas in the choroid, not unlike a disseminated choroiditis. The lens was removed and the macula seemed intact.

J. A. O'Brien (Melbourne) said that one point of diagnosis did not satisfy him. Many mothers had been seen with the alleged disease and the diagnosis was incorrect. Some complained of a severe sore throat and that at the time when the virus-infected thorns were rampant in military camps. Rubella had been known for centuries and this syndrome had never been noticed before. Was it not possible that they were dealing with some new infection altogether?

J. H. Keighley (Melbourne) said that in Townsville from the 1939-1940 epidemic the majority of the children suffered from deafness. Two deaf children examined had pigmentary changes in the retina similar to those described by Dr. Marks. Professor O'Brien had called the condition an ophthalmic-photographic. Older ophthalmologists would have said that it was an abnormal form of retinitis pigmentosa. It was regrettable that drawings were not made showing the condition, for it was worthy of record. There were difficulties in housing these children. They hoped to have a new deaf institution finished next month. It would be opened at the end of November and would provide for the modern treatment of deaf children. The Government had declared it a work of first priority and an excellent job had been done. It would be most interesting for the members.

A. L. Towers (Adelaide) referred to the article which was shortly to appear in The Medical Journal of Australia in regard to affected South Australian children. It was unfortunate that they were losing C. A. Swan, who had been appointed to the Royal London Ophthalmic Hospital. He wondered whether the grant under which Swan had worked could be made available to someone else, because he felt that they were only at the beginning of the solution of this interesting piece of research.

The President did not know how many Victorian cases had appeared during the year, but he himself had not seen one.

E. O. Marks said that the fundi were very similar to Gregg's description. Fair-complexioned children did not show so much pigmentary disturbance in the peripheral retina. M. O'Brien used the term "taytelmi"; he preferred "neth-taeten". He did not know of any cases of cataract which had appeared in Queensland during the previous year.

J. L. Day (Perth) said that the only epidemic which had occurred in Western Australia appeared in 1941. He had found cases in his own practice. One patient had died in fourteen years. Two bilateral congenital cataracts and two macular exudates, each with pigmentary changes. No deafness was noted in the four cases. He had not seen any cases since.

J. L. Towers (Adelaide) agreed to the question, stated that up to the present no of the patients he had seen had been sufficiently cooperative for any examination of the visual fields to be made. He quoted a psychologist as saying that the general mentality of the patients was too low for detailed examination and it was impossible now to make any further examination. He went on to say that the question raised by Dr. J. A. O'Brien had been well considered previously and the slightest doubt that hilheto these cases had occurred but had been missed. Professor O'Brien had seen a case in the Oxford Clinic in 1945. The patient had previously been seen by all the clinical assistants, not one of whom had thought to inquire into the maternal history.
proportion of deafness cases. He referred to a report in a recent British Medical Bulletin.

E. O. Marks reported that there were four cataractous children amongst the 63 deaf children born in 1888. They had been included in the 20 year data by pigmented abnormality.

M. A. SCULLY (Sydney) thanked Dr. Marks for his interesting paper. These pigmented changes in the fundus were of particular interest. He had been noted by several observers in connection with these cases. In the original paper read by him in the Medical Journal of Australia in 1923, he had noted that the unhealthy appearance of the iris in such cases suggests that there may be some pigment in the choroid. Looking back into his notes for that paper, he had found that A. C. Mitchell, reporting his case of left-sided macular cataract, had stated: "the fundus of the right eye appeared pale, and some scattered irregular spots of pigment were observed." Credit should therefore be given to him for the first observation of these changes.

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The speaker could not recall ever having seen a description of a fundal appearance similar to that one observed in this connexion. In order to make the point he had tried to describe the appearance. "Monkey" was the term he would like to use, perhaps "blotches" would be preferable. The area affected included the macula and an area of about three to four disc diameters surrounding it. In a few cases the disturbance was more widely distributed. He hoped that in the next "Joyce" Scarth would draw the fundus picture. It was like a piece of coarse Scotch thread used for a sports coat over which pepper had been thrown. The spots of pigment were discrete. An interesting feature of Dr. Marks' paper was that the time incidence of the epidemic of rubella in Queensland varied as compared with that in New South Wales. The big Queensland epidemic occurred in 1931; in New South Wales the first epidemic was 1942-1945 and the second 1941-1942. He would like to know from Dr. Marks the ratio of cases of deaf mutism to those of cataract. The speaker had no way of telling, the cases of deaf mutism in New South Wales was very high, especially in the first epidemic. He did not know whether the proportion was as high in the second epidemic; one had not heard of many such cases. It still appeared that the time factor in the maternal infection was of importance in deaf children. The general nature of the congenital defect in the infant. He had seen many patients showing cataracts and deaf mutism. These pigmented disturbances had been noted in some recent articles in the American journals.

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J. A. O'BRIEN (Melbourne) said that one point of diagnosis did not satisfy him. Many mothers had not been seen with the alleged disease and the diagnosis was inferred. Some complained of a severe sore throat and that at a time when the virus-infected throats were rampant in military camps. Rubella had been known for centuries and this syndrome had never been noticed before. Was it not possible that they were dealing with some new infection altogether?

J. L. DAVIES (Perth) said that in Townsend from the 1893-1896 epidemic the majority of the children suffered from deafness. Two deaf children examined had pigmented changes in the retina similar to those described by Dr. Marks. Professor Mann had called the condition an asthenopia. Older ophthalmologists would have said that it was an apparent form of retinitis pigmentosa. It was regrettable that drawings were not made showing the condition, for it was worthy of record. There were difficulties in housing these children. They hoped to have a new deaf institution finished next month. It would be opened at the end of November and would be ready for the modern treatment of deaf children. The Government had declared it a work of first priority and an excellent job had been done. It would be most interesting for the members to see.

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J. L. DAVIES (Adelaide) agreed that it was not easy to examine the patients after they had been given the first anesthetic.