

MULTIPLE ISLANDS OF RETINOBLASTOMA* INCIDENCE RATE AND TIME SPAN OF APPEARANCE

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IN the days before successful irradiation of bilateral retinoblastoma, the opportunities for studying the dates of appearance and the rate of growth of multiple islands of this neoplasm occurring in the remaining eye were rare, and indeed could only have occurred either when parental permission for excision of the second eye was refused or in the rare event of spontaneous recovery. The rarity of retinoblastoma in the white race (16·8 in 100,000 eye patients), the even greater rarity of spontaneous recovery, and the fact that the survival of those children for whom treatment was declined was unlikely to exceed one year or 18 months, doubtless explain the absence of any account in the literature of the incidence rate and the period during which multiple retinoblastoma islands may appear. Such information is of importance in the follow-up and prognosis of children affected by this tragic disease.

Hilgartner (1903) discovered that retinoblastoma was radiosensitive, and Lawson (1916), Knapp (1920), and Chase (1920) tried radiotherapy, using rather crude surface applicators of Columbia paste in which radium needles were set. The results of this technique were ineffective in destroying the neoplasm and often inflicted serious damage to the eye. So it was not till 1929, when Foster Moore (with whom I was privileged to be associated in this pioneer work) succeeded in destroying a retinoblastoma by the interstitial insertion of a radon seed, that a study of the subsequent fate of the second affected eye was possible.

Since 1929 improved radiotherapeutic technique, particularly with the use of radio-active discs made to fit exactly the curvature of a child's sclera, and the selection of patients in whom only one-third or less of the retina was involved in the neoplasm, have raised the successful results to 96 per cent. These favourable results have made it possible to study the course of 43 patients treated and followed-up during a period of 25 years.

Fig. 1 (overleaf) shows the ages (and therefore to some extent the span of time) at which multiple islands, two to five in number, appeared in 26 patients after the initial detection and successful treatment of this neoplasm at one or more sites. Absolute accuracy about the date of onset of the first island was only possible when a hitherto unaffected eye was being kept under observation, for in the majority when the child was first brought to hospital on account of one eye being almost full of retinoblastoma the second eye

*Received for publication January 3, 1955.

was also found to be affected. However, examinations thereafter at monthly intervals allowed fair precision in assessing the time of onset of the other islands.

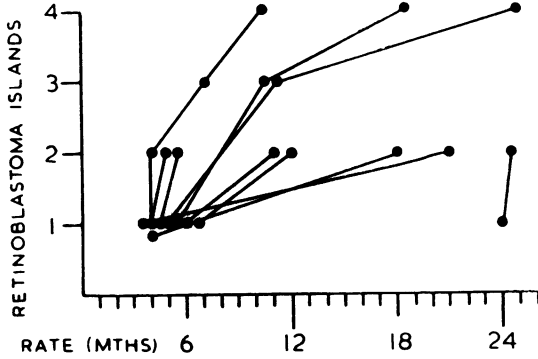


FIG. 1.—Age at which multiple islands of retinoblastoma appeared in 26 children.

It was interesting to note that the neoplasm might appear in some part of the fundus remote from the original site as early as 2 weeks after the treatment of the original growth.

With one exception these additional islands appeared in eyes in which the retinoblastoma was first noted between the 4th and 7th month of life. Indeed when both eyes are affected by retinoblastoma the neoplasm appears earlier than when the disease is unilateral. Dollfus and Auvert (1953) state that the average age at onset in bilateral cases is 1 year and 10 months, and remark that this is precocious compared with unilateral cases in which the average age is 3 years and 4 months. Unilateral cases of retinoblastoma sometimes occur as late as 10 or 11 years of age but these are rare. In my own series of 43 bilateral cases, the average age at which the children were brought for examination was 15 months, and 21 of these children were under 1 year of age. Fig. 2 shows the incidence of multiple islands in both eyes.

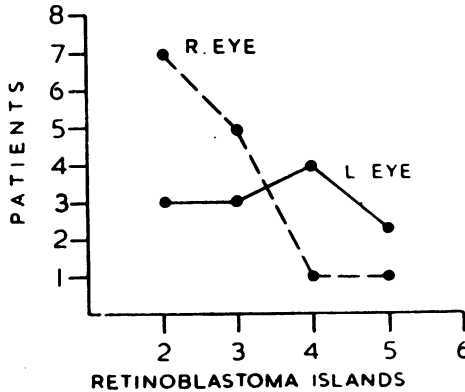


FIG. 2.—Bilateral incidence of multiple islands of retinoblastoma.

It is also interesting to note (see Fig. 1) that the appearance of other islands of retinoblastoma ceased at the age of 2 years and 1 month so far as my series is concerned. Nevertheless, I have considered it desirable to continue follow-up examinations at monthly intervals until the child is 5 years of age, after which the neoplasm is most unlikely to re-appear.

Summary

This paper discusses the incidence rate and the span of time during which multiple islands of retinoblastoma may appear after successful irradiation of an initial island or islands of retinoblastoma elsewhere in the fundus.

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