MARFAN'S SYNDROME WITH RETINAL DETACHMENT*

BY

JAMES R. HUDSON

London

Since the account of arachnodactyly by Marfan (1896), and the observation of its association with ectopia lentis by Salle (1912), a large number of cases has been described. The subject has been fully reviewed by Rados (1942), whose paper includes a statistical survey of all cases (numbering 204) described in the literature up until 1940, and an extensive bibliography. The prominent features of the syndrome are well known—elongation of the long bones, with long slender fingers and toes, spinal deformity, congenital heart disease, and ectopia lentis accompanied by miosis (Ormond, 1930). Opinion as to the hereditary nature of the condition varies. Strebel and Steiger (1915) reported a family in which ectopia lentis was associated with high myopia and congenital heart disease, Mann (1937) states that ectopia lentis linked with arachnodactyly is seldom an hereditary condition, and several other authors (Weve, 1931; Duke-Elder, 1937; Doggart, 1949) comment on the strongly hereditary character of the disease, which affects both sexes and is transmitted as a dominant characteristic. A perusal of the literature has revealed a description of only two cases of retinal detachment in association with the disease (Fischbach, 1937; Cassidy and McFarland, 1947). These were in a male child aged one-and-a-half years, and in a male adult aged 52 years, respectively. It is, therefore, hoped that the present case report may be of interest. The early recurrence of the detachment with spontaneous resolution following a short period of rest was a striking feature.

CASE REPORT

Miss D. B., aged 24 years, a typist, attended Moorfields Hospital on September 2, 1949, complaining of blurring of vision in the upper temporal field of the left eye during the previous three months, and of the appearance of a shadow over this area of the field, which had extended to obscure most of the field of the affected eye during the three weeks prior to her examination at the hospital. The patient exhibited the general physical features of Marfan's syndrome, including arachnodactyly, poor muscular development, and congenital heart disease.

Ocular Examination.—Movements full. Right eye 6/12 with correction (+ 4.00 D. sph.); eye white, anterior chamber deep, pupil active, iris tremulous, lens.

* Received for publication February, 15, 1951.

244
MARFAN'S SYNDROME WITH RETINAL DETACHMENT

透明和脱位向下，玻璃体混浊，视网膜正常，张力正常。

左眼6/36矫正 (+8.00 D. sph.); 眼白，前房加深，瞳孔活跃， iris tremulous，透明和脱位向下，玻璃体混浊，牵拉玻璃体下部通过瞳孔开大，几乎完全视网膜脱离深鼻侧和下部。

进一步检查显示视网膜内层没有异常。左眼显示几乎完全视网膜脱离的视网膜，深鼻侧和下部失明，张力正常。

手术治疗因两个原因延迟，首先是因为无法为可能的再循环提供时间，其次是因为患者对一般麻醉剂感到不安。医生J. S. Stead报告了主动脉瓣膜术后可能出现的窦性心率异常。虽然先天性心脏病患者不是由一种类型决定的，他考虑了一般麻醉剂的不必要性，而患者选择在局部麻醉下进行手术。

手术——手术于1949年9月19日进行，从顶部和视网膜下进行。内部凹陷腔被暴露并切开，玻璃体下部的表面电凝损伤在10点钟位置进行，15毫米。

如果电凝反应未显示出进一步的小电凝损伤，可能在切除内直肌时进行电凝。电凝损伤被插入了内直肌的区域。内直肌被重新附着，并且结膜被缝合。

患者在1949年10月7日从医院出院，四天后被允许出院。右眼未变。左眼被电凝，虹膜前部脱出，玻璃体混浊，视网膜在所有区域电凝损伤的周围区域有色素沉着，紧贴鼻侧，视觉指数6/18 + 7.00 D. sph. +1.00 D. cyl. at 10°。

复发和自发性恢复——两周后，患者再次出现症状。在患者出院后进行再电凝。电凝损伤被减少到4/60。在10月28日，这增加了总电凝损伤。在出院后9天，期间患者在家中休息，视网膜被观察到在正确的位置，尽管在患者被允许出院后，她被观察到在17月19日，视力指数6/18，戴上眼镜，左眼6/12。视网膜在所有区域。视网膜的张力是正常的。

患者不幸患心脏病，并于1950年2月23日去世。

我在此要感谢R. C. Davenport先生，感谢医疗委员会在Moorfields医院的许可，使我们可以使用医院的记录。

REFERENCES

Kinlton, London.