
In the past there has always been some dispute among those who believe in the value of arthrography of the knee whether a positive or a negative contrast medium is the better. The authors of this short monograph have combined the two procedures by injecting both media at the same time. For this they recommend 30 cubic centimetres of air and 5 cubic centimetres of diodone. The knee is then put through a full range of movement to distribute the diodone throughout the joint. There is no doubt that the result is excellent, giving an etched appearance to the menisci. Some of the illustrations, however, look like air arthograms, suggesting that the diodone has not been adequately distributed. It is mentioned that there must be no fluid in the joint but this point is not emphasised strongly enough. The method of taking the radiographs is basically the same as that described by others. Tangential views are taken of each meniscus, but the number of views—nine—would seem to be somewhat excessive, and the method of widening the side of the joint under examination is rather unnecessarily cumbersome although effective. The authors are wise to confine their diagnosis to lesions of the menisci, for which this procedure is best suited. Our criticisms in no way detract from the value of this book, which describes a good idea and illustrates it clearly.—E. W. SOMERVILLE.


Like other volumes in this valuable series of monographs on hereditary disease, this is a careful and critical review of the literature up to the present time. The author regrets that the bibliography is less complete than she would have liked, but in fact it must contain nearly all the important references to the Laurence-Moon syndrome, for 189 papers are listed here from 1864 to the present day, and 181 pedigrees are illustrated.

It has to be admitted that at present we have no clue at all as to the nature of the primary defect which produces such a varied and bizarre combination of abnormalities, namely adiposity, ocular defects (usually described as retinitis pigmentosa but often with considerable variation), some grade of mental deficiency, genital hypoplasia, and digital anomalies usually in the form of polydactyly. Although it is likely that retinitis pigmentosa is an abnormality occurring late in embryonic development, whereas polydactyly must occur early, the author nevertheless gives reasons for thinking that the syndrome is likely to be due to a single autosomal recessive gene. In all the pedigrees described there are only two in which a parent was affected as well as the offspring, and in one of these two the marriage was between second cousins. This of itself is strong evidence of the recessive nature of the defect. The fact that 45 per cent of siblings are affected in this series may well be due to selection for it is well known that authors tend to publish families in which more than one case has occurred, and, in any event, material collected from propositi will never show normal Mendelian ratios. In this particular disorder it is moreover difficult to know whether the siblings described as obese or mentally deficient were true examples of the syndrome; the occurrence of polydactyly or retinal defect is of course much stronger evidence, and digital abnormalities have often been recorded in siblings dying in infancy. The occurrence of the syndrome bears no relation to maternal age or to birth order. Another pointer to the recessive nature of the gene is the extremely high consanguinity rate amongst the parents, at least 25 per cent of whom were related in the published cases. No patient in the series was older than fifty-five years. It is very rare for either men or women with the Laurence-Moon syndrome to marry. No male patient became a father, though five affected women produced ten children, of whom six survived infancy. The relative sterility is probably only partly due to the hypogenitalism which is often a feature of the disease.

The syndrome varies a good deal from case to case. The diagnosis is usually first suggested by night-blindness and defective vision during childhood, but the actual ocular defects are very varied. Uncomplicated retinitis pigmentosa is probably not due to the same gene. The subnormal mentality cannot be purely ascribed to the visual difficulties. The digital anomaly sometimes takes the form of brachydactyly or syndactyly, though the most usual form is the occurrence of six digits on hands and feet. Associated defects in the nervous system, the heart and the urinary system have been described either during life or at post-mortem examination.

This valuable and authoritative review will become the essential starting point for future workers in this field.—Robert Platt.

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